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Contemporary outcomes of the Foker process and evolution of treatment algorithms for long-gap esophageal atresia

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ABSTRACT

Background: Esophageal growth using the Foker process (FP) for long-gap esophageal atresia (LGEA) has evolved over time.

Methods: Contemporary LGEA patients treated from 2014–2020 were compared to historical controls (2005 to <2014).

Results: 102 contemporary LGEA patients (type A 50%, B 18%, C 32%; 36% prior anastomotic attempt; 20 with esophagostomy) underwent either primary repair (n=23), jejunal interposition (JI; n = 14), or Foker process (FP; n = 65; 49 primary [p], 16 rescue [r]). The contemporary p-FP cohort experienced significantly fewer leaks on traction (4% vs 22%), bone fractures (2% vs 22%), anastomotic leak (12% vs 37%), and Foker failure (FP→JI; 0% vs 15%), when compared to historical p-FP patients (n = 27), all $p \leq 0.01$. Patients who underwent a completely (n = 11) or partially (n = 11) minimally invasive FP experienced fewer median days paralyzed (0 vs 8 vs 17) and intubated (9 vs 15 vs 25) compared to open FP patients, respectively (all $p \leq 0.03$), with equivalent leak rates (18% vs 9% vs 26%, $p = 0.47$). At one-year post-FP, most patients (62%) are predominantly orally fed.

Conclusion: With continued experience and technical refinements, the Foker process has evolved with improved outcomes, less morbidity and maximal esophageal preservation.

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Introduction

The classification of esophageal atresia (EA) can be described by the Gross or the more comprehensive Kluth classification, which considers the length of both esophageal pouches [1,2]. Long-gap esophageal atresia (LGEA), thus, is a description of the length of separation between the esophageal ends. A “long gap” can occur with almost any type of EA; however, the term LGEA has become most associated with Gross type A or B esophageal atresia. The International Network of Esophageal Atresia (INoEA) working group has defined LGEA as “any esophageal atresia that has no intra-abdominal air” (which infers no distal tracheo-esophageal fistula) and “all other types that technically prove difficult to repair” [3]. This includes gap lengths as small as 2 cm during surgery to

greater than 4 vertebral bodies apart [4–6]. Thus, the preoperative perception and the intra-operative visualization of LGEA can be drastically different, with the ultimate decision on type of surgical intervention is made by the treating surgeon based on available resources and experience. However, in patients with a gasless abdomen on initial radiograph, the diagnosis of LGEA should not be an intraoperative surprise. Hence, to avoid such circumstances, it is ideal to measure the gap between the upper and lower esophageal pouches (gap-o-gram) prior to exploring the chest in order to 1) appropriately plan for the ideal surgical intervention, and 2) recognize when the length of the gap might be greater than the available resources and experience in order to make an early referral to a center with LGEA expertise. This last point is critical, as we have previously demonstrated that redo, or rescue, Foker procedures have inferior outcomes to first time, or primary, Foker procedures [7].

With the inability to achieve primary anastomosis, many techniques have been implemented to obtain esophageal continuity, including a waiting period to allow natural growth (delayed primary

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anastomosis), serial bougie dilation, esophageal myotomies, gastric pull-up, colonic or jejunal interposition, and the Foker procedure (FP) with or without Kimura advancement [6–15]. Our institution has been performing FP for patients with LGEA since 2005. We previously reported our outcomes from 2005–2014 for both patients undergoing a primary FP and those undergoing a secondary, or rescue, FP (patients who had a previous attempt at repair at an outside institution) [7]. This analysis found that patients undergoing rescue FP had a longer time to esophageal anastomosis with a higher leak rate and a longer length of stay, identifying them as having a more complex disease process due to redo operations, prior esophageal leaks, inflammation and scarring. In this study, we aimed to report how our treatment algorithm and operative techniques have evolved over time for patients with LGEA. Additionally, we sought to compare the outcomes between our more recent “contemporary” patient cohort (2014–2020) with our historical controls (2005–2014).

Methods

After Institutional Review Board approval, a retrospective review was performed of all contemporary LGEA patients who were treated at our Esophageal and Airway Treatment center over the past five years (2014–2020), along with three patients who were treated at a different institution by one of our recently departed former surgeons. We compared this group to a historical group that we treated with FP from 2005–2014 and has been previously reported [7]. All patients who were in esophageal discontinuity were included in the study. These patients had EA Gross type A, B, C or D with a gap that was beyond the capabilities of performing a primary anastomosis on the first attempt as determined by the referring surgeon and included those with a prior attempt at anastomosis at the referring hospital (as long as they were in discontinuity at arrival to our institution), as well as those with an esophagostomy.

Preoperative Evaluation and LGEA Treatment Strategy Selection

As not all LGEA patients are the same, we developed an algorithm of the multiple diagnostic and treatment pathways that we consider for each patient [Fig. 1]. We review every case in order to customize the best approach to address the specific set of problems with which each patient presents. All patients undergo pre-operative upper and lower esophageal contrast and endoscopic studies to assess the character and length of the esophageal segments and the gap between them. Gap lengths were measured both under pressure with the use of dilators or the endoscope and without tension using contrast injection [Fig. 2A–B]. A three-phase rigid dynamic bronchoscopy is undertaken to assess associated airway anomalies, such as laryngeal cleft, tracheobronchomalacia (TBM), and any associated tracheoesophageal fistula (TEF) [16,17]. Pre-operative echocardiogram and contrast-enhanced chest computed tomography (CT) are performed to evaluate arch location and great vessel anomalies [18]. Pre- and post-operative vocal cord status is determined via awake flexible nasolaryngoscopy to evaluate for recurrent laryngeal nerve injury which can be prevalent in this patient population [19]. Nutritional optimization is undertaken on all children prior to operative intervention. Although we do not have an age threshold to begin the LGEA operative treatment, we do wait for the child to weigh at least 3–3.5 kg prior to operative intervention given that we anecdotally experienced greater traction suture dislodgement problems with children that weighed less.

One of the main deciding factors in the treatment algorithm is the quality and length of the lower esophageal pouch. While we have successfully achieved successful growth for lower esophageal

pouches less than 1 cm in length, if this is absent or severely injured from prior interventions, then we consider this patient for a jejunal interposition. Other factors that lead us to consideration of esophageal replacement include larger patient size (we prefer over 10 kg for microvascular augmentation), presence of a cervical esophagostomy, the number of prior operations, and anatomic problems with the stomach (such as severe hiatal hernia from prior partial gastric pull-up). However, if there is a healthy lower esophageal pouch, we always explore the chest with the intent to perform a primary anastomosis but have a low threshold to place the esophagus on traction if we think we cannot achieve a quality anastomosis with acceptable tension. We also will perform traction induced growth to gain additional lower pouch length in order to create an anastomosis that is ideally at least 2 cm above the GE junction. Patients with severe symptoms of TBM (e.g. acute life-threatening events [ALTE]/ brief resolved unexplained events [BRUEs], inability to wean from respiratory support, etc.) or those with greater than 50% dynamic collapse of either the trachea or mainstem bronchi are considered for surgical repair of their TBM (posterior tracheopexy) at the time of their Foker procedure or primary anastomosis [16,20].

Patients are considered for a minimally invasive (MIS) approach if they do not have a major degree of TBM that would require surgical intervention and if they do not have a history of multiple prior operations. The decision to undergo internal or external traction depends on the length of the gap and associated patient comorbidity profile. Patients with a short gap (<2 cm) or those in whom a prolonged postoperative period of chemical muscle paralysis and intubation would be detrimental are placed on internal traction. Conversely, longer gaps are often best managed either with external traction or with internal traction via serial thoracoscopic traction adjustments (MIS strategy).

Although most of our LGEA repairs have been from the right chest, recently we have noted that a select group of patients may benefit from a left-sided approach. Patients with a large leftward-deviated upper pouch, without significant TBM, without an intrathoracic TEF, or who have had multiple previous surgeries in the right chest, are considered for a left chest repair, even in the presence of a left-sided aortic arch [21].

Surgical Technique

The Foker procedure (FP) was first described by John Foker in 1997 and has been adapted by our institution [7,10]. Dissection can be performed either thoracoscopically or through an open incision. If the patient had a very short upper pouch or an upper pouch fistula, a cervical incision was performed to gain appropriate access and/or to repair the fistula. Though some have described repairing a cervical TEF via the chest with thoracoscopy [22], in the setting of an anticipated esophageal traction process, we prefer to approach a cervical TEF in a Gross type B esophageal atresia patient via an open incision in the neck to allow for full mobilization of the cervical esophagus, identification and protection of the recurrent laryngeal nerves (recently using intraoperative nerve monitoring), division of the TEF flush with the trachea and placement of a silastic sleeve in the neck to protect the esophagus from scarring as it comes through the thoracic inlet in order to optimize the traction process. This open cervical approach did not preclude use of MIS technique for the thoracic portion of the operation. Whenever possible, flexible endoscopic guidance (EGD) was used during traction suture placement to ensure that the sutures were not intraluminal. Silastic sleeves are placed to protect the esophageal pouches and traction system from adhesions, and the traction sutures are brought out through the skin and tied to a silicone disk. Small feeding tube segments are used to tighten sutures every 1 to 3 days. Chest radiographs are used to track esophageal growth

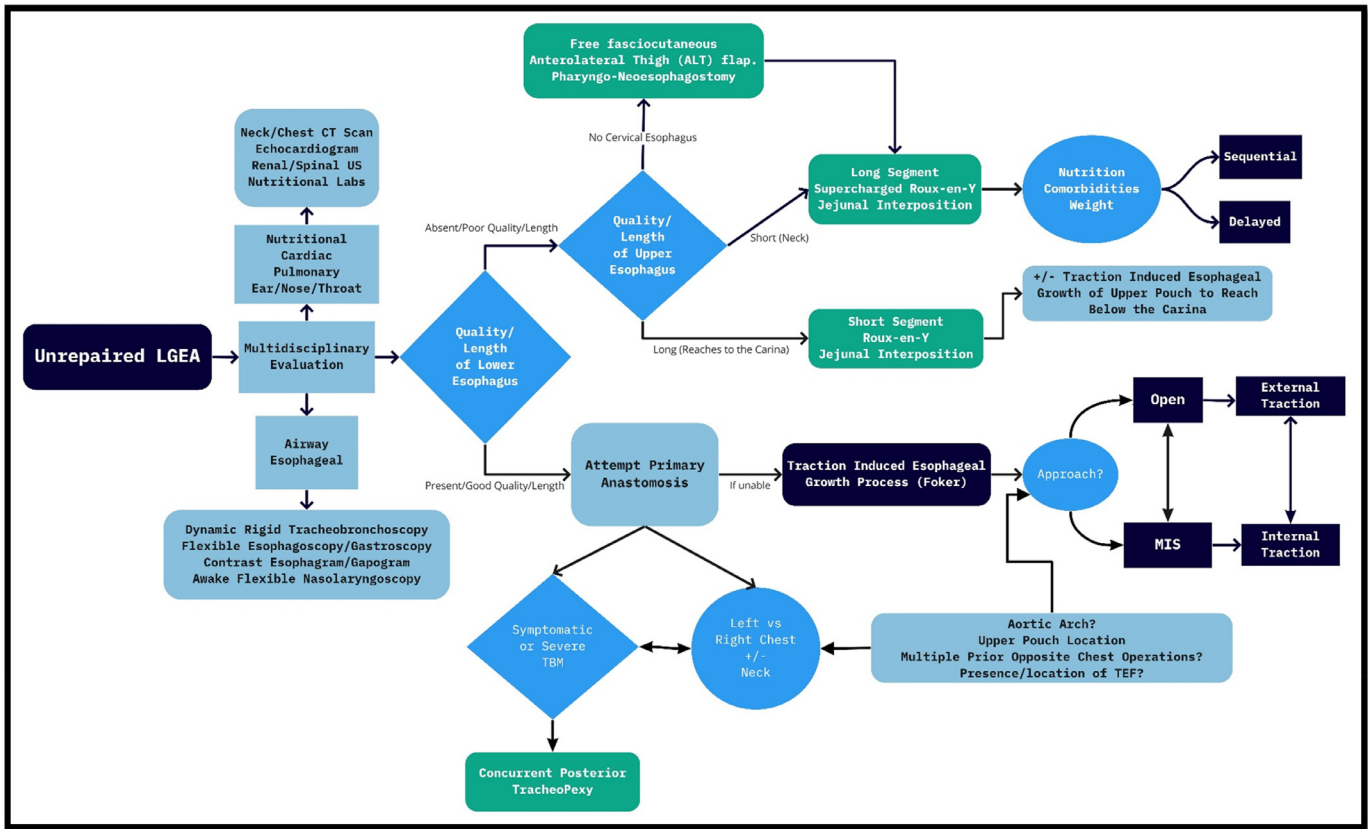


Fig. 1. Treatment Algorithm for Long-Gap Esophageal Atresia.

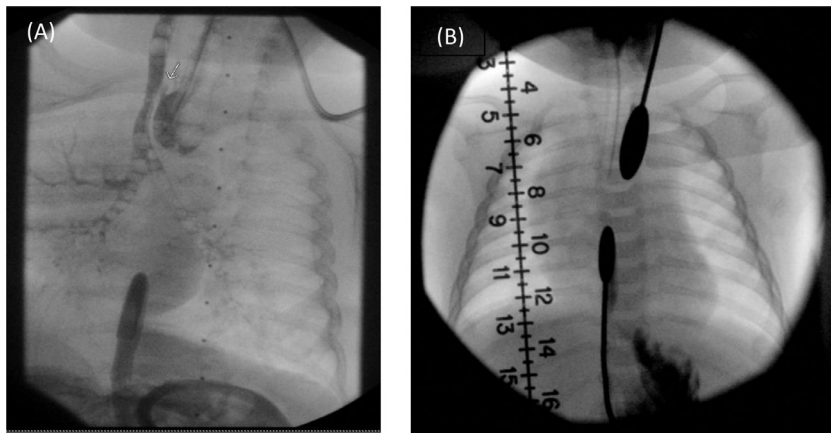


Fig. 2. A-B. Measurements of Gap Length. The gap length between the two esophageal ends can be measured passively with contrast (A) (note the proximal TEF and the tracheal compression by the proximal esophageal segment), or with pressure using dilators and/or an endoscope, (B) using a ruler (note the small 1 cm spaced dots along the spine), or by counting vertebral bodies.

by monitoring the traction system clips, and guide timing of return to the operating room for anastomosis once esophageal overlap is achieved [23].

For our thoracoscopic internal traction procedure, the esophageal traction sutures are secured either around a rib or one pouch to the other, as was similarly described by Tainaka [24]. [Fig. 3A–B] Given that the traction system of these patients is internal and not tightened frequently, no paralysis was used during the esophageal growth phase, and the patients were extubated and transferred to the floor when appropriate. The patients returned to the operating room on a weekly basis for traction system adjustment until the gap had closed and they were deemed ready for esophageal anastomosis. The esophageal anastomosis

was undertaken via an open or MIS technique depending on esophageal tissue quality and perceived tension.

Postoperative neuromuscular paralysis was used for some patients in which the esophageal anastomosis was performed under moderate to severe tension, as judged by the operating surgeon. In addition, for some high-risk anastomoses during the year of 2018 (n = 9), we trialed the use of a trans-anastomotic prophylactic esophageal wound vac (EVAC) [25]. Given the efficacy of EVAC therapy to treat established esophageal leaks [26], we had hypothesized that the prophylactic use of an EVAC system at the time of the anastomosis could decrease the risk of leak and/or stricture formation. Our initial enthusiasm was tempered by our analysis indicating that the leak rate was similar, but the long-term risk of

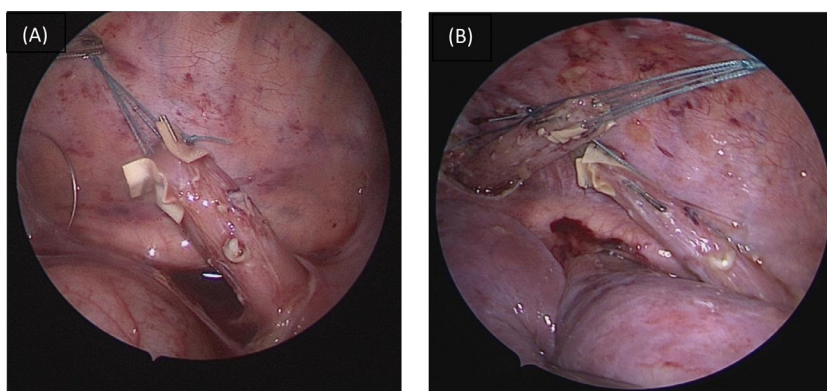


Fig. 3. A-B. Internal Traction Technique. Pledgeted sutures are placed on each end of the esophagus, incorporating the muscular and submucosa layer (A). The sutures are then secured around the rib using Roeder's knots, which are tightened weekly (B).

poor anastomotic outcome (i.e. need for stricture resection) was greater in the prophylactic EVAC group [25]; hence, this technique is no longer used in prophylactic approach.

For cases that require a jejunal interposition, we preferentially perform a long-segment, supercharged, pedicled, Roux-en-Y jejunal interposition as previously described [9,27]. In brief, the jejunal conduit is brought into the chest and/or neck through retro-colic, ante-gastric substernal route and anastomosed to the upper esophagus using an end-to-end hand-sewn, single layer approach. Microvascular augmentation, or “supercharging”, of the jejunal conduit is undertaken to augment arterial inflow and venous outflow via the internal mammary vessels.

Data Collection and Statistical Analysis

Demographic and clinical variables were collected from the electronic medical record. A primary Foker was defined as the first surgical attempt at esophageal continuity. A rescue Foker was defined in patients who had a previous attempt at esophageal continuity at an outside facility. The primary analyses focused on evaluating differences in post-operative outcomes between the contemporary and historical Foker patients. Secondary analyses focused on post-operative outcomes between patients who underwent a MIS versus an open FP, between primary and rescue Foker patients, and determining one-year post-FP feeding outcomes. Patients without a one-year follow-up were excluded from this feeding status evaluation. Feeding status was captured in four groups using a modified version of the Functional Oral Intake Scale (mFOIS) to include patients with an esophagostomy: full oral intake, consistent oral intake with some feeding tube supplementation, predominantly tube fed, and exclusively tube fed [28].

Descriptive and summary statistics are provided when applicable. Fisher's exact and Mann-Whitney U tests were used to compare demographic variables, post-operative outcomes, esophageal anastomotic complications and feeding outcomes between the historical and contemporary cohorts. To determine predictors of anastomotic leak and need for stricture resection, univariate analyses were performed using the following variables: historical vs contemporary, primary vs rescue Foker, type of EA, age at surgery, weight at surgery, internal vs external traction, open vs MIS approach, leak while on traction, time on traction and length of paralysis. For need of stricture resection, use of prophylactic EVAC and anastomotic leak were also included. Variables with a p-value of 0.10 or less were included in the multivariate logistic regression model. In general, we considered a p-value of <0.05 as statistically significant; however, in circumstances in which multiple hypothesis testing was present, we chose a more conservative p-value of $p < 0.01$ as significant to decrease the risk of rejecting the null

hypothesis just by chance [29]. Statistical analysis was carried out using STATA 15.2 (StataCorp 2017, College Station, TX).

Results

In total, 143 patients were treated for LGEA from 2005–2020, with 41 patients in the historical Foker cohort (2005–April 2014) and 102 LGEA patients in the contemporary cohort (May 2014–August 2020). Of these 102 contemporary LGEA patients, 65 (64%) underwent a FP, 23 (23%) received a primary anastomosis and 14 (14%) received a jejunal interposition [Table 1]. Three contemporary FP patients were treated at a different institution by one of our former surgeons (CJS). Thus, a total of 106 Foker patients (65 contemporary and 41 historical) were treated in the study timeframe. With respect to demographic and pre-operative clinical variables among the contemporary LGEA cohort, there was no difference in the percentage of patients in either the Foker or primary anastomosis groups who had previous surgery before transfer to our institutions (23% vs 35% had previous attempts at anastomosis, respectively, $p = 0.26$). All patients who underwent an immediate JI had an esophagostomy performed at another facility. The JI population was significantly older (33.0 months vs 5.0 months and 3.0 months) and weighed more (14.2 kg vs 5.5 kg and 4.3 kg) at the time of surgery than patients in the Foker group or primary repair group, respectively ($p < 0.001$). Although all patients who underwent a JI had an esophagostomy performed at another facility, two patients with an esophagostomy were able to undergo primary repair, and four achieved esophageal growth and anastomosis by FP (age range 3–22 months). There was a significant difference in gap length between the groups, with those undergoing primary repair having the shortest gap (1.5–2.9 cm) and those undergoing a JI having the longest gap length (6.5–8 cm; $p < 0.001$) [Table 1].

Historical Comparison

To analyze changes in outcomes and technique over time, our contemporary cohort of Foker patients was compared to our historical group [Table 2]. As a difference in outcomes between primary and rescue FPs in the historical cohort had been previously reported [17], post-operative outcomes were stratified into primary or rescue Foker process within both the historical and contemporary groups [Fig. 4, Table 3]. There was a significantly shorter length of paralysis ($p < 0.02$) and hospital length of stay ($p < 0.007$) in both the contemporary primary and rescue FP groups compared to their historical counterparts. Significantly fewer patients in both contemporary groups required a return trip to the operating room due to an esophageal leak while on traction compared to their historical counterparts [Table 3]. In the pri-

Table 1
Demographic and Intra-operative Variable Comparison Between Patients who underwent Foker procedure, primary repair, or jejunal interposition to establish esophageal continuity in the contemporary cohort.

Demographics ^a	Total Patients (n = 102)	Foker Repair (n = 65)	Primary Repair (n = 23)	Immediate Jejunum (n = 14)	p-value
Male (%)	47 (46%)	32 (49%)	10 (44%)	5 (36%)	0.63
Birth Weight (kg)	2.20 (1.77, 2.62)	2.20 (1.77, 2.65)	2.22 (1.74, 2.49)	2.10 (1.89, 2.64)	0.47
Prematurity (%)	63 (62%)	42 (64%)	14 (61%)	7 (50%)	0.59
CHD* (%)	53 (52%)	40 (62%)	8 (35%)	5 (36%)	0.04
		40 (62%)	8 (35%)		0.03
		40 (62%)		5 (36%)	0.08
			8 (35%)	5 (36%)	0.95
Genetic Anomalies (%)	12 (12%)	11 (17%)	0 (0%)	1 (7%)	0.22
Type of Esophageal Atresia (Gross Classification)					
-Type A (%)	51 (50%)	35 (54%)	8 (35%)	8 (57%)	0.25
-Type B (%)	18 (18%)	14 (22%)	4 (17%)	0 (0%)	0.40
-Type C (%)	33 (32%)	16 (25%)	11 (48%)	6 (43%)	0.08
Previous Surgery at Outside Hospital	37 (36%)	15 (23%)	8 (35%)	14 (100%)	< 0.001
-No Prior Surgery (%)	11 (11%)	8 (12%)	3 (13%)	N/A	0.90
-G tube +/- TEF* ligation only	54 (53%)	42 (65%)	12 (52%)	N/A	0.27
-Previous Esophageal Anastomotic Attempt	37 (36%)	15 (23%)	8 (35%)	14 (100%)	< 0.001
		15 (23%)		14 (100%)	<0.001
			8 (35%)	14 (100%)	<0.001
Intra-Operative Variables	Total Patients (n = 102)	Foker Repair (n = 65)	Primary Repair (n = 23)	Immediate Jejunum (n = 14)	p-value
Gap Length					
-Pressure (cm)	2.3 (1.5, 3.5)	3 (2, 4)	1.5 (0.8, 1.9)	N/A	< 0.001
-Static/Contrast (cm)	4.5 (3.5, 6)	5 (4, 5.5)	2.9 (2.5, 3.5)	8 (6.4, 11)	<0.001
		5 (4, 5.5)	2.9 (2.5, 3.5)		<0.001
		5 (4, 5.5)		8 (6.4, 11)	<0.001
			2.9 (2.5, 3.5)	8 (6.4, 11)	<0.001
-Vertebral Bodies	5 (3, 6)	5 (4, 6)	1.9 (1.1, 3.7)	6.5 (6, 7)	<0.001
		5 (4, 6)	1.9 (1.1, 3.7)		<0.001
		5 (4, 6)		6.5 (6, 7)	0.005
			1.9 (1.1, 3.7)	6.5 (6, 7)	<0.001
Age (months) at Surgery	5.0 (3, 8)	5 (3, 6)	3.0 (1.0, 6.0)	33.0 (20, 51)	<0.001
Weight (kg) at Surgery	5.6 (4.1, 7.7)	5.5 (4, 6.5)	4.3 (3.7, 5.8)	14.2 (10.2, 16.5)	<0.001
Operative Approach					
-Open Thoracotomy	74 (73%)	43 (66%)	17 (74%)	14 (100%)	0.10
-Minimally Invasive	16 (16%)	11 (17%)	5 (22%)	N/A	0.60
-Part MIS, Part Open	12 (12%)	11 (17%)	1 (4%)	N/A	0.12
Side of Approach					
-Right Side	60 (59%)	40 (62%)	20 (87%)	N/A	0.03
-Left Side	28 (27%)	25 (38%)	3 (13%)	N/A	0.03
Median Follow-up Time (days)	648 (190, 1036)	647 (221, 1265)	685 (81, 921)	249 (88, 453)	0.01

^a All continuous variables are expressed in medians with inter-quartile range. Comparisons were performed between the Foker, primary repair, and jejunal interposition groups using an ANOVA test. For variables that were significant, specific 2-group analysis was performed to further delineate significant differences between individual groups; p-value ≤ 0.01 was considered statistically significant (bolded).

* CHD = congenital heart disease; TEF = tracheo-esophageal fistula

primary FP comparison, the contemporary cohort had fewer anastomotic leaks (12% vs 37%, $p = 0.01$), stricture resections (18% vs 37%, $p = 0.07$), funduplications performed (39% vs 67%, $p = 0.02$) and no subsequent conversions to a jejunal interposition (0% vs 15%, $p = 0.006$), although median length of follow-up was understandably shorter than in the historical group (1.8 years vs 5.3 years; $p < 0.001$). There was no difference in the number of dilations needed after anastomosis [Table 3]. In the rescue FP comparison, the contemporary group required fewer procedures to achieve esophageal anastomosis (2 vs 5.5, $p = 0.001$); a shorter length of paralysis (12 vs 35 days, $p < 0.001$), intubation (23 vs 50.5 days, $p = 0.006$), ICU stay (43.5 vs 94.5 days, $p = 0.005$), and hospital stay (71.5 vs 170.5 days, $p < 0.001$). Similarly, fewer contemporary rescue Foker patients had a fundoplication compared to their historical rescue FP counterparts (25% vs 82%, $p = 0.004$).

More patients in the contemporary primary and rescue Foker groups underwent concomitant airway work, most often a posterior tracheopexy, to correct tracheomalacia compared to historical cohorts, respectively (71% vs 41%, $p = 0.011$ for the primary groups

and 88% vs 21%, $p < 0.001$ for the rescue groups). Thus, fewer patients in both the contemporary primary and rescue cohorts required a subsequent operative intervention to correct symptomatic tracheomalacia ($p = 0.006$ and $p = 0.048$, respectively). [Table 3]

Two patients from the historical cohort and two in the contemporary cohort have died. One patient died from sequelae of a congenital heart defect (Epstein's anomaly), and one died from sepsis due to a urinary tract infection. Two patients died from respiratory issues related to chronic aspiration and residual tracheomalacia; this occurred early in our experience when we did not routinely perform direct posterior tracheopexies at the time of the Foker I procedure. Two of these patients died with a cervical esophagotomy in place.

Subgroup Analysis of Contemporary Foker Patients

Outcomes in the contemporary rescue Foker ($n = 16$) group were compared to patients who underwent a primary Foker pro-

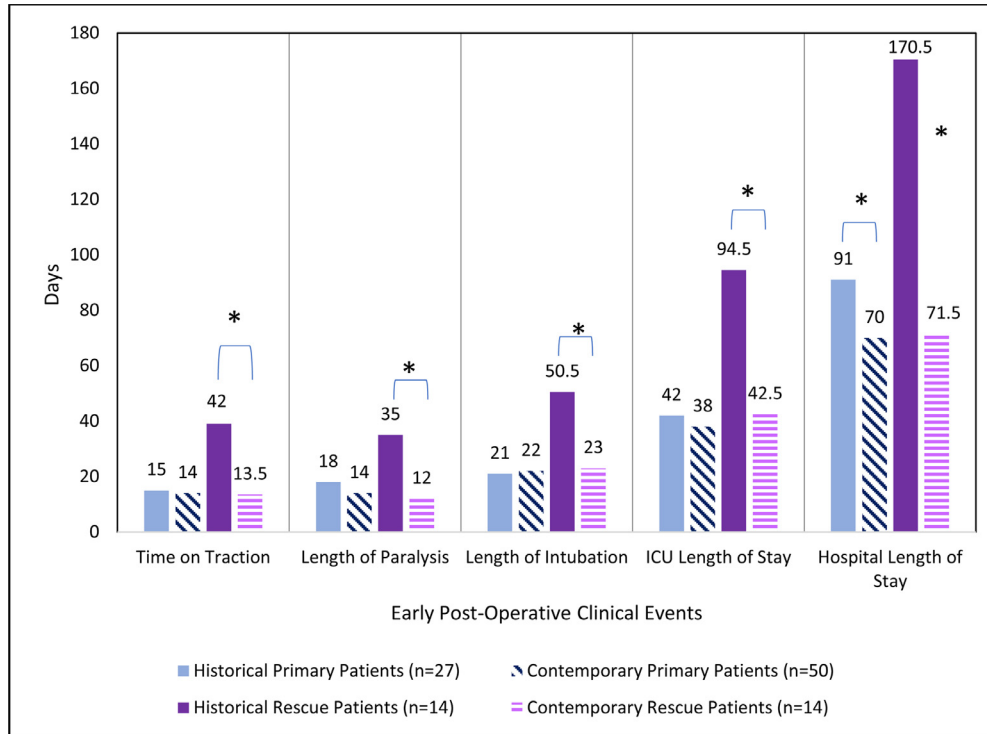


Fig. 4. Early post-operative outcomes between the historical and contemporary cohorts with stratification based on whether the patient had a primary or rescue Foker procedure. (ICU = intensive care unit) * = $p < 0.05$

Table 2
Demographic Comparison Between Historical and Contemporary Foker Patients. All continuous variables are expressed in medians and inter-quartile ranges.

	Historical Controls (n = 41)	Contemporary Cohort (n = 65)	p-value
Male (%)	24 (58%)	32 (49%)	0.17
Birth Weight (kg)	2.4 kg (1.9, 3.1)	2.20 kg (1.77, 2.65)	0.03
Prematurity (%)	19 (45%)	42 (64%)	0.06
Type of Esophageal Atresia			
-Type A (%)	23 (56%)	35 (54%)	0.76
-Type B (%)	6 (15%)	14 (22%)	0.15
-Type C (%)	11 (27%)	16 (25%)	0.74
Gap Length			
-Static/Contrast (cm)	4.5 cm (3.5, 5)	5 cm (4, 5.5)	0.24
-Vertebral Bodies	5.5 cm (4, 7)	5 cm (4, 6)	0.55
Operative Approach			
Open Thoracotomy (%)	41 (100%)	43 (66%)	<0.001
-Minimally Invasive	0 (0%)	11 (17%)	0.006
-Part MIS, Part Open	0 (0%)	11 (17%)	0.006
Type of Foker Repair			
-Primary Foker	27 (66%)	50 (77%)	0.22
-Rescue Foker	14 (34%)	15 (23%)	0.22
Concurrent Airway Procedure	16 (39%)	43 (66%)	0.007
Age at Surgery - months	4 months (2, 7)	5 months (3, 6)	0.73
Weight at Surgery - kg	4.5 kg (3.9, 8)	5.5 kg (4, 6.5)	0.96

cedure (n = 49). Between groups, gender, birth weight and incidence of prematurity were similar [Table 4]. More patients in the primary Foker group were a Gross classification Type A ($p = 0.04$), while more patients in the rescue Foker group were a Gross classification Type C ($p < 0.001$). Rescue Foker patients were also older ($p = 0.008$) and weighed more ($p = 0.01$). Early post-operative outcomes, including length of paralysis ($p = 0.77$), in-

tubation ($p = 0.22$) and hospital stay at our institution ($p = 0.71$) were similar between groups [Fig. 5]. Fewer primary Foker patients developed an anastomotic leak (12% vs 50%, $p = 0.003$) but rates of stricture resection (18% vs 19%, $p = 0.62$) were similar [Table 5].

A completely minimally invasive (MIS) approach for both traction and anastomosis was performed in 11 (17%) of the 65 patients undergoing a FP in the contemporary cohort. An additional 11 patients (17%) had either part or their entire FP traction performed thoracoscopically with an open anastomosis, while the remaining 43 patients underwent their traction and anastomosis via an open approach. In comparing the three groups, there were no significant differences in demographic variables between the complete MIS, part MIS, and open FP groups [Appendix 1]. Despite having similar traction times, both the complete MIS-FP group and the part MIS-FP group had significantly shorter lengths of paralysis ($p \leq 0.003$), intubation ($p \leq 0.002$), and ICU stay ($p \leq 0.001$ and $p = 0.049$, respectively) than those who underwent open thoracotomy [Fig. 6]. The complete MIS-FP group also had a significantly shorter length of paralysis compared to the part MIS group ($p = 0.005$), and a shorter hospital stay compared to the open group ($p = 0.007$). No difference in leak rate (18% vs 9% vs 26%, $p = 0.47$) was found between the complete MIS-FP, part MIS-FP and open FP groups, respectively; however, a greater percentage of patients in the complete MIS-FP group (45%) required a stricture resection compared to either the open FP group (14%, $p = 0.02$) or the part MIS-FP group (9%, $p = 0.06$). The number of dilations were also similar: 36%, 55%, and 35% ($p = 0.48$) of patients required three or fewer dilations, 27%, 45%, and 44% ($p = 0.57$) required 4–7 dilations, and 36%, 0%, and 21% ($p = 0.26$) required eight or more dilations in the complete MIS, part MIS, and open groups, respectively. While the time to most recent follow-up was slightly shorter in the part MIS-FP group (median: 367 days, range: 116, 928 days) than in the open FP group (median: 648 days, range: 221–1371 days) or the complete MIS group (median: 705 days, range: 105–1002 days), this was not statistically significant ($p = 0.28$).

Table 3
Comparison of outcomes between the contemporary and historical cohorts, stratified based on whether the patient had a primary or rescue FP.

	Historical Primary (n = 27)	Contemporary Primary (n = 49)	p-value	Historical Rescue (n = 14)*	Contemporary Rescue (n = 16)	p-value
Number of Procedures to Anastomosis	2 (2, 3)	2 (2, 3)	0.70	5.5 (4, 13)	2 (2, 3)	0.001
-Number of Patients with Leak on Traction	6 (22%)	2 (4%)	0.01	10 (71%)	3 (19%)	0.005
-Number of Patients Requiring Procedure due to Suture Pull-out	5 (19%)	11 (22%)	0.78	2 (14%)	0 (0%)	0.13
Time on Traction	15 days (12, 22)	14 days (11, 19)	0.21	42 (27, 49)	13.5 (8.0, 21)	0.004
Length of Paralysis	18 days (14, 27)	14 days (6, 20)	0.02	35 (33, 55)	12 (9.5, 21.5)	<0.001
Length of Intubation	21 days (19, 32)	22 days (10, 29)	0.42	50.5 (38, 74)	23 (15, 50.5)	0.008
Length of ICU Stay	42 days (30, 76)	38 days (28, 58)	0.28	94.5 (66, 192)	43.5 (15, 75)	0.005
Length of Hospital Stay	91 days (64, 144)	70 days (50, 95)	0.007	170.5 (112, 462)	71.5 (27.5, 103)	<0.001
Anastomotic Leak	10 (37%)	6 (12%)	0.01	5 (45%)	8 (50%)	0.80
Stricture Resection	10 (37%)	9 (18%)	0.07	4 (36%)	3 (19%)	0.33
Anastomotic Dilations						
-3 or Fewer Dilations	9 (33%)	17 (36%)	0.79	2 (20%)	8 (750%)	0.12
-4 to 7 Dilations	9 (33%)	20 (41%)	0.49	5 (50%)	7 (44%)	0.76
-8 or More Dilations	9 (33%)	9 (18%)	0.14	3 (30%)	1 (6%)	0.10
Fundoplication	18 (67%)	19 (39%)	0.02	9 (82%)	4 (25%)	0.004
-360° Fundoplication	16 (59%)	17 (35%)	0.04	9 (82%)	4 (25%)	0.004
-Partial Posterior Fundo	0 (0%)	1 (2%)	0.46	0 (0%)	0 (0%)	N/A
-Partial Anterior Fundo	0 (0%)	1 (2%)	0.46	0 (0%)	0 (0%)	N/A
-Intussusception-type	2 (7%)	0 (0%)	0.06	0 (0%)	0 (0%)	N/A
Jejunal Interposition	4 (15%)	0 (0%)	0.006	5 (45%)	3 (19%)	0.15
VTE	3 (11%)	6 (12%)	0.90	4 (29%)	0 (0%)	0.02
Chyle Leak	0 (0%)	3 (6%)	0.20	2 (14%)	0 (0%)	0.13
Fracture	6 (22%)	1 (2%)	0.004	9 (64%)	1 (6%)	<0.001
Airway Work	11 (41%)	35 (71%)	0.011	3 (21%)	14 (88%)	<0.001
-At Foker Procedure only	0 (0%)	26 (53%)	<0.001	0 (0%)	8 (50%)	0.002
-After FP	9 (33%)	4 (8%)	0.006	3 (21%)	2 (13%)	0.54
-During and After FP	2 (18%)	5 (10%)	0.66	0 (0%)	4 (25%)	0.048
Mortality	0 (0%)	2 (4%)	0.30	2 (14%)	0 (0%)	0.13
Length of Follow-up	5.3 years (97 days–8.1 years)	1.8 years (45 days–5.7 years)	<0.001	7.0 years (1.7–9.6 years)	1.5 years (28 days–5.8 years)	<0.001

* One patient ultimately underwent a jejunal interposition due to the inability to obtain esophageal anastomosis with FP.

Table 4
Demographic Comparison between contemporary Primary and Rescue Foker Patients.

	Primary Foker (n = 49)	Rescue Foker (n = 16)	p-value
Male (%)	25 (51%)	7 (44%)	0.97
Median Birth Weight – kg ^a	2.2 kg (1.8, 2.7)	2.1 kg (1.5, 2.5)	0.26
Prematurity (%)	30 (61%)	12 (75%)	0.31
Gross Type of Esophageal Atresia			
-Type A (%)	30 (61%)	5 (31%)	0.04
-Type B (%)	12 (24%)	2 (12.5%)	0.33
-Type C (%)	7 (14%)	9 (56%)	<0.001
Gap Length			
-Pressure (cm)	2.75 cm (2, 4)	3.25 cm (2, 4)	0.55
-Static/Contrast (cm)	4.5 cm (4.0, 5.5)	5.5 cm (4, 6)	0.13
-Vertebral Bodies	5.75 (5.0, 7.0)	4 (3, 6)	0.06
Age at Surgery - months	4 months (3, 6)	6.5 months (3.5, 10)	0.008
Weight at Surgery – kg	5.1 kg (3.9, 6.2)	6.3 kg (5.1, 9.2)	0.01

^a All continuous variables are in medians with interquartile range unless otherwise stated.

Table 5
Comparison of Outcomes Between Patients in the contemporary Primary Foker group and the Rescue Foker group.

	Primary Foker (n = 49)	Rescue Foker (n = 16)	p-value
Number of Procedures Until Anastomosis	2 (2, 3)	2 (2, 3)	0.74
-Number of Patients with Leak on Traction	2 (4%)	3 (19%)	0.05
-Number of Patients Requiring Procedure due to Suture Pull-out	11 (22%)	0 (0%)	0.04
Anastomotic Leak (%)	6 (12%)	8 (50%)	0.003
Anastomotic Dilations (%)			
-3 or Fewer Dilations	17 (36%)	8 (50%)	0.32
-4–7 Dilations	20 (41%)	7 (44%)	0.83
-8 or More Dilations	12 (24%)	1 (6%)	0.12
Median Number of Dilations (IQR)	5 (3, 7.5)	3.5 (1, 7)	0.29
Stricture Resection (%)	9 (18%)	3 (19%)	0.62
Jejunal Interposition (%)	0 (0%)	3 (19%)	0.01
VTE (%)	6 (12%)	0 (0%)	0.17
Chyle Leak (%)	3 (6%)	0 (0%)	0.57
Fractures (%)	1 (2%)	1 (6%)	0.44
Full Oral Intake at 1 Year ^b	15/39 (38%)	3/12 (25%)	0.41

^aAll continuous variables are expressed in medians with inter-quartile range.

^b Only patients who had at least one year of follow-up were included in this analysis (Primary Foker = 39, Rescue Foker = 12).

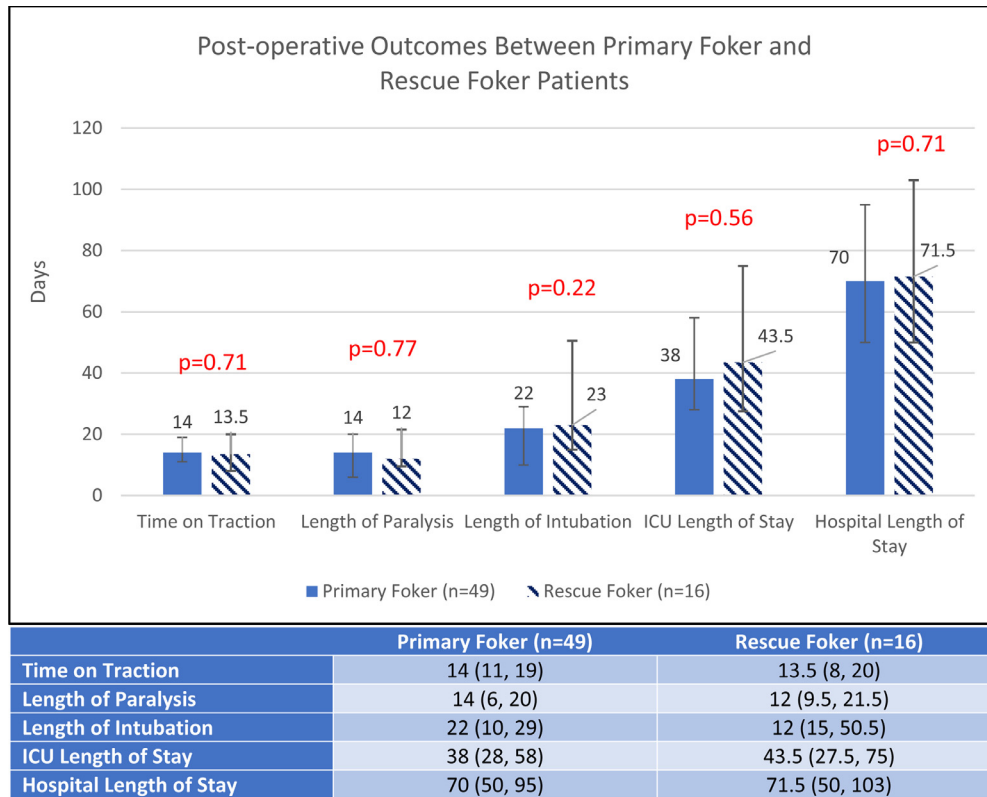


Fig. 5. Early post-operative outcomes compared between contemporary patients who underwent a primary Foker procedure versus those who underwent a rescue Foker procedure. All outcomes are expressed as medians with inter-quartile ranges.

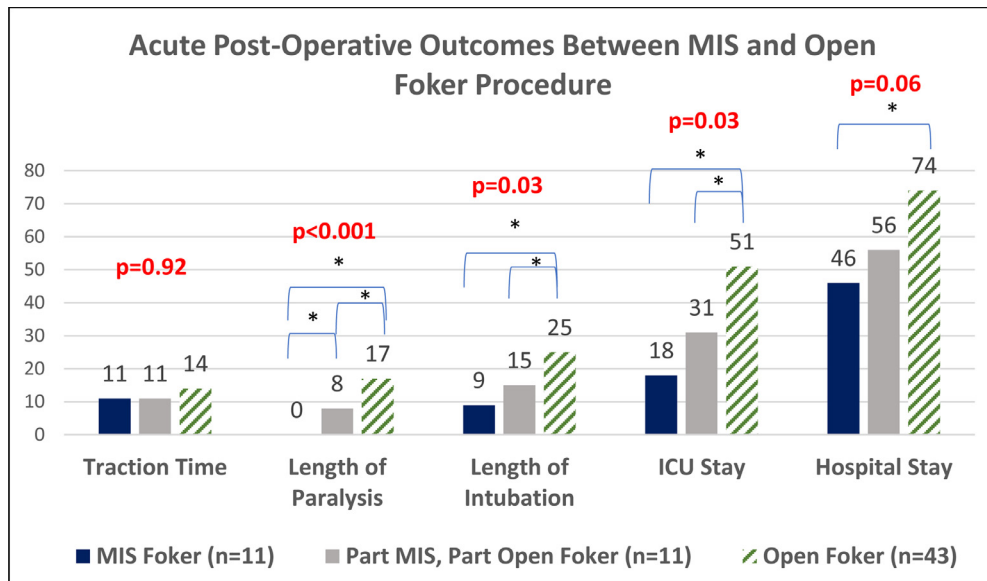


Figure 6. Acute post-operative outcomes between patients who had a complete minimally invasive Foker procedure, a part minimally invasive and part open Foker procedures, and an open Foker procedure. All values are the median length of time required for each outcome. *p*-values represent the ANOVA comparison between all three groups, * indicates individual *p*-value < 0.01; MIS = minimally invasive surgery, ICU = intensive care unit

Feeding Outcomes

One-year post-FP feeding outcomes were assessed among the contemporary FP patients who had at least one year of follow-up after anastomosis (n = 51). Using the mFOIS, 18 (35%) of all Foker patients were fully orally fed, while another 14 (27%) achieved consistent oral intake with some tube feed supplementation, 11 (22%)

were predominantly tube fed, and 8 (16%) were exclusively tube fed. Looking specifically at the subgroups of Primary and Rescue Foker patients, 15/39 (38%) of patients in the Primary Foker group were fully orally fed by one-year of follow-up, while 3/12 patients (25%) in the Rescue Foker group were fully orally fed at that time point (*p* = 0.41).

Table 6A

Univariate Screen and Multivariate Analysis to Determine Predictors of Anastomotic Leak. This analysis included the historical vs contemporary cohort variable. Variables with a p -value ≤ 0.10 on univariate screen were added to the multivariate analysis.

Predictors of Leak	Univariate p -value	Multivariate p -value	Odds Ratio	95% CI
Historical/ Contemporary	0.004	0.037	0.32	0.11–0.93
Primary/ Rescue	0.014	0.051	3.24	1.0–10.60
Type of EA	0.36			
Age at Surgery	0.029	0.527		
Weight at Surgery	0.032	0.972		
Type of Foker	0.129			
Open/ MIS	0.100	0.252		
Gap Length	0.196			
Leak on Traction	0.004	0.059		
Time on Traction	0.325			
Length of Paralysis	0.012	0.451		

Table 6B

Univariate Screen and Multivariate Analysis of the Contemporary Foker Population to Determine Predictors of Anastomotic Leak. As many technical changes have been made over the study period, the contemporary cohort was analyzed to determine if any new changes in long-gap esophageal atresia management were associated with anastomotic leak.

Predictors of Leak	Univariate p -value	Multivariate p -value	Odds Ratio	95% CI
Primary/ Rescue	0.003	0.014	7.63	1.51–38.63
Type of EA	0.518			
Age at Surgery	0.033	0.923		
Weight at Surgery	0.017	0.427		
Type of Foker	0.427			
Open/ MIS	0.672			
Gap Length	0.607			
Leak on Traction	0.09	0.041	5.10	1.07–24.29
Time on Traction	0.143			
Length of Paralysis	0.376			

Table 7

Univariate Screen and Multivariate Analysis of all Foker Patients to Determine Predictors of Needing an Anastomotic Stricture Resection.

Predictors of SR	Univariate p -value	Multivariate p -value	Odds Ratio	95% CI
Historical/ Contemporary	0.066	0.076		
Primary/ Rescue	1.00			
Type of EA	0.443			
Age at Surgery	0.842			
Weight at Surgery	0.883			
Type of Foker	0.317			
Open/ MIS	0.171			
Gap Length	0.329			
Leak on Traction	0.011	0.016	3.79	1.28–11.24
Time on Traction	0.207			
Length of Paralysis	0.927			
Prophylactic EVAC	0.04	0.003	12.21	2.37–63.97
Anastomotic Leak	0.014	0.164		

Predictors of Anastomotic Leak and Need for Stricture Resection

On multivariate analysis, patients in the historical cohort and those requiring a rescue Foker procedure had significantly greater odds of developing an anastomotic leak [Table 6A]. While focusing on only the contemporary cohort, patients who underwent a rescue Foker procedure and those who developed an esophageal leak while on traction were significantly more likely to develop an anastomotic leak [Table 6B]. Patients who developed an esophageal leak while on traction and those who had a prophylactic EVAC were found to be significantly more likely to require an anastomotic stricture resection [Table 7].

Discussion

Our results highlight the evolution and improvement in outcomes for LGEA over a 15-year period. Understanding the vari-

ables that create complexity for patients with LGEA such as airway problems, prior esophageal operations and leaks, the presence of tracheoesophageal fistulas, and different gap lengths has allowed customization of our approach. The ability to offer a full range of procedures that best address the totality of issues has proven to be critical, as the need to change tactics can often present even during the course of the operations. We believe that the goal of repairing the native esophagus remains paramount for optimizing long term outcomes; yet, expertise with esophageal replacement is necessary for certain cases. Our prior report on primary versus rescue Foker cases highlighted the added morbidity of long paralysis and ICU times that can occur when traction cases go awry from suture dislodgment and leaks, which included increased risk for long bone fractures and venous thromboembolism events (VTE). As our program has evolved, we have learned both better techniques to salvage these problems, and better patient selection for rescue Foker procedures versus jejunal interposition.

To start, a better definition of LGEA is needed. Despite ongoing controversy about the best definition for LGEA [3–6], there remains many patients with all types of esophageal atresia who have been referred to us for an inability to achieve esophageal anastomosis because of the gap length between the upper and lower pouches. Additionally, gap measurement (by any of the available techniques) does not always correlate with ability to achieve an anastomosis when attempted, especially a quality anastomosis with acceptable tension. EA cases with prior operations often have pouches that are scarred in place, which further complicate accurate gap measurement. Ultimately, the ability to utilize multiple modalities of techniques to achieve native esophageal connection can negate the preoperative uncertainties that may exist for cases of LGEA. It also means that one does not have to wait for a certain weight or time period or amount of perceived esophageal growth before undertaking the process of either esophageal anastomosis or growth induction.

Another key aspect of FP evolution has been the understanding that tension-induced esophageal growth works well with less frequent traction adjustment. Whereas we adjusted traction sutures daily, or even twice daily, as a rule in the beginning of our experience, it became apparent that suture adjustments can be spaced out with very similar esophageal growth and less risk of suture dislodgement. This concept contributed to the use of internal traction and MIS approaches, which can greatly decrease the morbidity that has been historically associated with this procedure, as has been described by Till, Tainaka and van der Zee [14,24,30]. Patients on traction through a MIS approach in our cohort had much shorter lengths of paralysis and hospital stay without increasing the complication rate, which has helped to change our practice for select patients. Not having to provide chemical paralysis and sedation for intubation safety allows for a faster narcotic wean, which may have contributed to the shorter length of stays in both the ICU and the hospital. The ability to be extubated between procedures also allowed for more movement and less use of furosemide, decreasing the risk of developing a DVT or fracture [31,32]. While the MIS approach is not optimal for all patients, as some require more complicated airway work and/or redo operations in very scarred and inflamed chests, we anticipate that its utilization will likely continue to increase.

Additionally, a select group of patients who have a large leftward-deviated upper pouch without thoracic tracheomalacia or who have had multiple prior surgeries in the right chest may be candidates for a FP through the left chest. Our previous research focusing on this cohort found that those who underwent a left-sided approach had equivalent anastomotic outcomes to the right-sided FP, while requiring less airway interventions and had shorter hospital stays from the increased use of the MIS-FP strategy in the left-sided FP group [21].

Stadil et al. performed a systematic review looking at outcomes specifically in patients with Gross type A or B esophageal atresia [32]. In this review, the most common method of esophageal anastomosis was delayed primary repair, with 38% of patients undergoing some kind of lengthening procedure prior to anastomosis. In the review, 53.7% developed an anastomotic stricture, while 22.7% developed a leak [32]. While these numbers are worse than our contemporary primary FP cohort (12% developed a leak and 18% required a stricture resection), the numbers approach those in our combined rescue FP experience (43% developed a leak and 23% required a stricture resection). Similarly, a recent systematic review performed by the APSA Outcomes committee evaluated multiple LGEA treatment strategies [4]. Again, they found that though delayed primary repair appears to be the most common strategy, it can be associated with high stricture (60%) and leak rates (30%) [33–35].

More recently, the use of magnets to treat LGEA has resurfaced as a strategy in select patients with a gap of less than 4cm. Unfortunately, not all patients are candidates, and given the size of the currently available magnets, there is concern of significant stricture rates, need for multiple endoscopic dilations (average of 10–13), as well as refractory stricture rates (requiring stents and/or reoperations) of up to 46% [36]. In contrast, only 18% of our contemporary primary Foker cohort required a stricture resection, and the median number of endoscopic dilations was 5 (IQR 2, 7). Given the potential deleterious effects of repeated anesthetic exposures on the development of a child's brain [37–39], one must be cautious about LGEA therapies that may predispose a child to a large number of endoscopic dilations by trying to avoid one or two definitive operations upfront.

The INoEA position paper on the management of long-gap esophageal atresia recommends that these high-risk patients might be best served at either centers of excellence or regional pediatric surgical centers, where a multidisciplinary team is available for all aspects of pre-operative, operative, post-operative, and long-term care [3]. To our knowledge, our surgeons have successfully treated a greater number of LGEA patients than any of the 57 studies that were included in Stadil's systematic review (range: 1–33) [32]; comparing our data to such a review highlights that centers that specialize in these complex patients with a greater patient volume will have improved outcomes with decreased complications.

For cases that required a secondary/rescue Foker procedure, the hospital courses were longer and more complicated. Our goal is to preserve the native esophagus; however, over the course of our practice, we have come to recognize that older patients who present with multiple previous surgeries may have an esophagus that has been scarred and devascularized, leading to an increased risk for leaks and strictures. Therefore, we have performed fewer rescue Fokers in this population and have opted for esophageal replacement using jejunal interposition in more contemporary patients. As a result, our failed Foker rate (patients who underwent a rescue FP but ultimately required a jejunal interposition) decreased from 19% in the historical group to 0% and 5% in the contemporary primary and rescue FP cohort, respectively. The jejunal interposition, although technically challenging, can maintain better peristalsis and more closely resembles the esophagus in size – which may lend itself to less progressive dilation over time as is often seen with colonic interpositions [3,40]. The jejunal interposition also does not incur the increase in reflux or potential upper esophageal mucosal changes (i.e. Barrett's) that can be seen with a gastric pull-up; hence, the jejunal interposition graft has become our preferred conduit when esophageal replacement is indicated [3,9].

While associated with technical challenges, FP provides direct esophago-esophageal anastomosis in young infants, without the need to wait months to allow the esophagus to potentially grow enough to achieve primary repair. Earlier anastomosis allows for earlier attempts at oral intake and consultation with feeding therapists, which can be very beneficial, as oral aversion can start within the first few months of life. This was seen in our study, as 39% of patients who underwent a primary FP were fully orally fed at one year of follow-up, while 25% in the secondary FP group, whose median age at anastomosis was five months later than the primary FP group, were fully orally fed by the same time point. In the effort to decrease rates of oral aversion, a recent study describing the use of the Foker process for esophageal growth and anastomosis without initial gastrostomy placement has also been published [41].

Over time we have changed our approach to the management of gastro-esophageal reflux post-FP. Initially, we routinely

performed a fundoplication in nearly all patients within 3–4 weeks after their esophageal anastomosis as a matter of protocol. However, we found that fundoplication at this early stage often added significant morbidity and we began to question the benefit compared to the potential negative impact of an additional large operation in lengthening the hospitalization and ICU stay. Furthermore, the evolution of GERD diagnostic modalities (e.g. pH-impedance) and medical therapies for acid suppression and promotility agents have allowed us to monitor and medically treat GERD more effectively. Furthermore, we have become more experienced with the use of gastro-jejunostomy tubes for post-pyloric feeds; a strategy that gives reflux-prone LGEA patients time to gradually transition to gastric and oral feeds [42]. Similarly, a greater emphasis on routine endoscopic surveillance has allowed us to closely monitor the health of the esophagus [43] as a result, we are now more selective and reserve fundoplications for patients with true failure to thrive or in those in whom their reflux appears to be injurious to their anastomotic outcome. With this selective approach, it appears that only about a quarter of the FP patients need a fundoplication.

Another important component that needs to be considered when preparing a child with LGEA for surgery is to determine the presence and severity of tracheomalacia. The reported incidence of tracheomalacia in patients with LGEA is varied, with values ranging from 11–87% [12,18,20,44,45]. For all patients, identification of significant tracheomalacia via dynamic 3-phase bronchoscopy prior to esophageal atresia repair is imperative. As we have recognized the high incidence of airway anomalies, a greater percentage of patients have undergone a posterior tracheopexy at the time of their initial traction procedure in the contemporary cohort, with fewer patients subsequently requiring additional procedures and anesthetics for symptomatic tracheomalacia compared to the historical controls. In each of our LGEA patients, we look at all aspects of their esophageal and airway anatomy and function in order to determine the optimal operative plan that addresses all issues from the onset. Decisions about open vs MIS, right vs left side, and neck and/or chest approach are made in the context of this comprehensive evaluation.

Our study has limitations that we acknowledge. Our results are reflective of a very specialized and focused referral center for children with complex esophageal and airway problems and may not be applicable to other centers. Given that the MIS approach is a more recent technique, and their follow-up is shorter than the open Foker group, we recognize the possible lead time bias in certain long-term outcomes (e.g. need for stricture resection or jejunal interposition) for our MIS Foker patients. Nonetheless, early outcomes have proven to be at least equivalent if not better (i.e. shorter hospital stays), hence we anticipate that these benefits will persist in the long-term. Certainly, selection bias is present in our MIS cohort. Due to it being a new technique, we purposely selected patients for the MIS strategy that had a more favorable LGEA profile to being with (e.g. fewer prior operations, no significant TBM, etc.). Hence, the outcome differences between the MIS-FP and the open FP groups may be related in part to their underlying baseline differences and not entirely due to the technique used. Furthermore, our data is retrospectively collected, which has limitations in terms of granularity of detail. We recognize that our outcomes are incomplete and that we are missing important patient reported outcomes such as quality of life, healthcare resource utilization, and other long-term issues. This represents an area of future research in which we are actively engaged. Other questions that remain to be addressed include the optimal patient age and weight for beginning the Foker process, the best frequency and intensity of traction adjustments and the timing or even necessity for fundoplication in the post-Foker child.

Conclusion

With continued experience and technical advancements, we have refined our algorithm to treat the many complexities associated with patients who have long-gap esophageal atresia. Advances in technical skill, including increased use of MIS techniques, to achieve esophageal continuity has led to improved outcomes and less morbidity. Our results highlight the benefits that patients with long-gap esophageal atresia receive when cared for at referral centers with the volume and multi-disciplinary expertise to provide the intensive pre-operative, operative, post-operative, and long-term care needed for these patients.

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Appendix 1 Demographic Comparison between Foker patients who were repaired through a minimally invasive approach at all stages versus patients who required an open procedure for at least one stage of repair

	MIS Foker (n = 11)	Part MIS, Part Open Foker (n = 11)	Open Foker (n = 43)	p-value
Male (%)	3 (27%)	7 (64%)	22 (51%)	0.22
Median Birth Weight – kg*	2.41 kg (2.16, 2.70)	2.33 kg (1.80, 2.66)	1.94 kg (1.58, 2.64)	0.15
Prematurity (%)	6 (55%)	5 (45%)	31 (72%)	0.19
Gross Type of Esophageal Atresia				
-Type A (%)	8 (73%)	4 (36%)	23 (53%)	0.23
-Type B (%)	2 (18%)	5 (45%)	7 (16%)	0.10
-Type C (%)	1 (9%)	2 (18%)	13 (30%)	0.30
Gap Length				
-Pressure (cm)	2.0 cm (2.0, 3.5)	3.0 cm (1.5, 3.5)	3.5 cm (2, 4)	0.24
-Static/Contrast (cm)	5.5 cm (4.0, 6.0)	5.0 cm (4.0, 5.5)	4.5 cm (3.7, 5.5)	0.77
-Vertebral Bodies	5.5 (4, 6)	7.0 (6.0, 7.0)	5 (4, 6)	0.48
Age at Surgery – months	4 months (3, 8)	3 months (2, 5)	5 months (3, 6)	0.13
Weight at Surgery – kg	5.77 kg (5, 7.90)	4.57 kg (3.67, 6.45)	5.50 kg (3.45, 6.40)	0.35
Primary Foker Repair	9 (82%)	8 (73%)	32 (74%)	0.86
Rescue Foker Repair	2 (18%)	3 (27%)	11 (26%)	

*All continuous variables are in medians with interquartile range unless otherwise stated.

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