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Nutrition delivery and growth outcomes in infants with long-gap esophageal atresia who undergo the Foker process

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ABSTRACT

Background: Predictors of growth outcomes in patients with long-gap esophageal atresia (LGEA) are not known. We examined nutrition and growth in-hospital and post-discharge in LGEA patients who underwent the Foker Process (FP).

Methods: Single-center, retrospective cohort study of infants with LGEA undergoing primary (non-rescue) FP from 2014 to 2020. Weight-for-age z scores (WAZ, 0 = average), macronutrient prescription, anthropometry, and clinical variables were collected. Longitudinal median regression evaluated differences in WAZ over time. Multivariable median regression examined variables associated with change in WAZ at 1 year.

Results: 45 patients met criteria, with median (IQR) age at repair of 4 (2, 5.8) months and WAZ of -0.96 (-1.55, -0.40). On admission, 11% were moderately (WAZ < -2) and 9% were severely (WAZ < -3) malnourished. Lower admission WAZ was significantly associated with improvement in WAZ at 1-year follow-up ($p = 0.002$); EA type (59% type A), esophageal leak (16%), median days paralyzed (13), ventilated (21), on parenteral nutrition (35), or to full enteral nutrition (35) were not associated with change in WAZ. Median WAZ remained stable while in-hospital, and patients maintained their growth curves through 3-year follow-up.

Conclusion: Throughout infancy, most primary FP LGEA patients have weight for age that is below average. Using targeted nutritional intervention, those who present with malnutrition can still achieve adequate growth despite prolonged and complicated hospital courses.

Abbreviations: Foker Process (FP), long-gap esophageal atresia (LGEA), weight-for-age z score (WAZ), parenteral nutrition (PN), enteral nutrition (EN), World Health Organization (WHO), Center for Disease Control (CDC), Functional Oral Intake Scale (FOIS), speech and language pathologist (SLP), interquartile ranges (IQR), confidence interval (CI), intensive care unit (ICU), central line associated blood stream infection (CLABSI), urinary tract infection (UTI), American Society for Parental and Enteral Nutrition (ASPEN), minimally invasive (MIS), Congenital Heart Disease (CHD), association of vertebral defects, anal atresia, cardiac defects, tracheoesophageal fistula, renal anomalies, and limb abnormalities (VACTERL), intrauterine growth restriction (IUGR), small for gestational age (SGA), head circumference (HC), post-operative day (POD), soybean oil based lipid emulsion (SOLE), mixed oil lipid emulsion (MOLE), fish oil lipid emulsion (FOLE), essential fatty acid deficiency (EFAD), parental nutrition associated liver disease (PNALD), recommended dietary allowance (RDA), mid-upper arm circumference (MUAC), establishment of esophageal traction system (Foker 1)

Level of Evidence: III

1. Introduction

Surgical management of long-gap esophageal atresia (LGEA) is challenging. Options for repair include delayed primary anastomosis, gastric pull-up, colonic interposition, jejunal interposition, magnet-induced elongation and anastomosis (magnamosis), or the Foker Process (FP). No single technique has yet been proven superior and it is likely that there is an institutional or surgeon bias toward a particular technique based on prior experience and available resources [1–4]. Furthermore, the LGEA patient popula-

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tion is so heterogeneous that one technique does not suit every patient. Though our center prefers a customized approach to each case, our institution has predominantly embraced the FP technique given our preference for esophageal preservation whenever possible [5,6].

The FP relies on traction-induced esophageal growth which requires prolonged periods of sedation and chemical muscle paralysis. During this time patients are maintained on intravenous nutrition to avoid reflux and pressurization of the lower esophagus from enteral nutrition, particularly in the setting of reduced gut motility while paralyzed, which might increase risk of esophageal leak. While paralyzed, FP patients often develop third spacing of fluids and clinically significant edema. FP patients are weighed infrequently during the FP and any weights obtained are likely influenced by fluid shifts. It is therefore difficult to monitor growth of these patients using standard metrics and more challenging to tailor nutritional recommendations. It can also be difficult to deliver optimal nutrition therapy via intravenous route due to fluid restrictions and intrinsic limitations to parenteral nutrition (PN) support.

There is a perception among parents and providers that long-term growth of this patient population is below average compared to their peers without EA [7–10]. There is also a paucity of literature on nutritional needs and growth outcomes in EA patients. In particular, nutrition and growth in the LGEA-FP patient population have not yet been studied. In the past several years, there has been a focus on the importance of nutrition in pediatric surgical patients [11–14]. In addition, examining specific pediatric surgical cohorts has revealed modifiable risk factors for impaired growth that could facilitate nutrition optimization in those populations [15–17].

We aimed to describe in-hospital macronutrient prescription in patients who undergo the FP for LGEA management at our institution. We also describe growth in this cohort from birth through 3 years after repair and examine factors associated with poor growth at 1 year. We hypothesized that changes in weight-for-age z scores (WAZ) at 1 year in LGEA patients undergoing the FP would be associated with modifiable factors that could become targets or biomarkers for future nutritional interventions [15–17].

2. Methods

We conducted a single-center, retrospective cohort study. Approval with consent waiver was obtained from the institutional review board. We included all patients with LGEA who were admitted from 2014 to 2020 at Boston Children's Hospital for primary FP (no previous attempt at esophageal anastomosis). Because there is no consensus definition for what is considered LGEA [2], we defined LGEA as any type of EA with a perceived esophageal gap that was deemed not amenable to primary repair. Demographic, clinical, nutritional, operative, and perioperative data were collected from the electronic medical record.

Our growth outcome of interest was weight-for-age z score (WAZ), which is defined as the standard deviation above or below a statistical mean derived from population-based normative growth data (z score of 0 corresponds to the 50th weight percentile for a certain age) [18]. WAZ was recorded at birth, admission, time of surgical repair, hospital discharge, and every 6 months until 3 years after repair, or date of last follow-up. WAZ was calculated from the Fenton growth chart for premature infants (born less than 37 weeks gestational age) up until $<$ 40 weeks post-menstrual age (PMA). For full term infants and for premature infants $>$ 40 weeks PMA, WAZ was calculated from the World Health Organization (WHO) growth chart for infants and children less than 2 years of age. The Center for Disease Control (CDC) growth chart was used to calculate WAZ for children $\geq$ 2 years of age. Premature infants were corrected for gestational age up to 3 years of chronological age on the WHO or CDC growth charts per American Academy of Pediatrics recommendation [19,20]. For this study, a decline in WAZ by 0.5 was deemed clinically significant. Patients with WAZ $<$ -1 were classified as mildly malnourished, those with WAZ $<$ -2 as moderately malnourished, and those with WAZ $<$ -3 as severely malnourished [21].

We also recorded additional nutrition variables including parenteral nutrition (PN) and enteral nutrition (EN) macronutrient prescriptions, timing of PN/EN initiation and duration of support, type of IV lipid prescribed, and time to achieve full EN after anastomosis. Given the variability in the FP duration for each patient, we chose post-operative day 15 as the approximate mid-point of the FP for most patients. The Functional Oral Intake Scale (FOIS) was used to determine feeding status at 1 year after the FP and at each year subsequently up to 3 years post-FP when available [22].

2.1. Perioperative nutritional management

Perioperative management of these patients starts with a dietitian consult. Patients are screened for malnutrition, micronutrient deficiencies, and electrolyte derangements. Individualized nutrition and growth goals are identified, which take into account gestational age, birth metrics and classification, including the presence of small for gestation age and/or intrauterine growth restriction. Catch-up growth may be required to optimize patients prior to the first stage of the FP. Preoperative nutritional management often consists of enteral nutrition via a gastrostomy tube whenever possible. PN is considered when EN is contraindicated or if it is not possible to provide adequate EN to meet nutritional needs.

After the first FP operation (establishment of an esophageal external or internal traction system via thoracotomy or thoracoscopy), patients are most often transitioned to PN and typically remain on PN throughout their traction course. After establishment of esophageal continuity, PN is continued until an esophagram is negative for leak (typically performed 7–14 days after anastomosis). During periods of chemical muscle paralysis (required for external traction patients and after high-risk anastomoses), fluid and caloric intake is restricted to an average of 80 kcal/kg and 80 mL/kg/day. When paralysis is discontinued or not required, fluids and caloric intake are liberalized to at least 90–100 kcal/kg and 100 mL/kg/day. After a satisfactory esophagram and depending on comorbidity profile, risk for reflux and aspiration, and need for ventilation or positive pressure respiratory support, patients are started on EN via a gastrostomy or gastrojejunostomy tube. Fluids and calories are typically liberalized further at this point as needed for promoting growth. Patients are evaluated by our speech and language pathologist (SLP) and begin oral feeds as soon as medically and developmentally appropriate.

2.2. Power and sample size

The sample of 45 patients undergoing the Foker Process, of which 32 patients had completed data as measured by WAZ at 1 year, provided 80% power for detecting a standardized change from admission to 1 year of 0.5 z score units (clinically significant) based on the Wilcoxon signed-ranks test for repeated growth measures over time within the same patient. Sample size calculations were performed using nQuery Advisor version 8.0 (Statistical Solutions Ltd., Cork, Ireland).

2.3. Statistical analyze

Demographic and nutrition variables were explored using appropriate descriptive statistics. Continuous data are presented as medians with interquartile ranges (IQR) and categorical data are presented as frequencies with percentages. Boxplots were created to show the distribution of WAZ at each measurement time point.

Table 1
Demographics and baseline clinical data ($n = 45$).*

	n (%)	Median (IQR)
Gender (Female)	23 (51.1)	
Gestational Age (weeks)		36.1 (33.9, 37.0)
> 37 weeks	18 (40.0)	
34–37 weeks	15 (33.3)	
32–34 weeks	6 (13.3)	
31–32 weeks	6 (13.3)	
Birth weight (kg)		2.30 (1.74, 2.69)
Birth WAZ		-0.96 (-1.55, -0.40)
Gross Classification A	26 (57.8)	
B	12 (26.7)	
C	7 (15.6)	
D	0 (0)	
E	0 (0)	
Referred from outside hospital	39 (86.7)	
CHD requiring operative intervention	10 (22.2)	
Trisomy 21	7 (15.6)	
Other chromosomal abnormality	1 (2.2)	
VACTERL	29 (64.4)	
Age at first operation (months)		3.97 (2.35, 5.84)
WAZ at admission		-0.70 (-1.59, -0.20)
WAZ at initial operation		-0.76 (-1.58, -0.14)

*IQR = interquartile range, WAZ = weight-for-age z score, CHD = congenital heart disease, VACTERL = association of vertebral defects, anal atresia, cardiac defects, tracheo-esophageal fistula, renal anomalies, and limb abnormalities

Table 2
Nutrient delivery.*

	n (%)	Median (IQR)
Post-op day PN initiated		0 (0, 1)
# PN courses		1 (1, 2)
% PN of hospital stay		42.6 (30.2, 51.4)
Total days on PN		35 (22, 43)
Post-op day enteral feeds initiated		26 (19, 32)
Post-op day full enteral feeds		35 (24, 40)
Total days on EN		45 (23, 78)
% EN of hospital stay		57.5 (48.7, 69.8)
Pre-op kcal/kg prescribed		120 (107, 131)
Post-op day 15 kcal/kg prescribed		81 (79, 89)
Discharge kcal/kg prescribed		112 (101, 120)
Sodium supplementation	14 (31.1)	
Pre-op g/kg protein prescribed		2.5 (1.8, 2.8)
Post-op day 15 g/kg protein prescribed		3.0 (3.0, 3.0)
Discharge g/kg protein prescribed		2.1 (1.6, 2.6)
Type of Lipid: Intralipid	29 (64.4)	
SMOF	15 (33.3)	
Omegaven	1 (2.2)	

*IQR = interquartile range, PN = parenteral nutrition, EN = enteral nutrition, Pre/Post-op = Pre/Post-operative

Table 3
Clinical course and post-operative complications.*

	n (%)	Median (IQR)
Total length of stay		79 (55, 130)
Length of initial ICU course**		39 (24, 78)
# operations to anastomosis		2 (2, 3)
Total # procedures in operating room**		8 (5, 10)
Days on traction		14 (10, 20)
External	26 (57.8)	
Internal	14 (31.1)	
Both	5 (11.1)	
Days of mechanical ventilation		21 (11, 30)
Days of paralysis		13 (6, 19)
Esophageal leak requiring intervention	7 (15.6)	
Chyle leak	3 (6.7)	
CLABSI	8 (17.8)	
UTI	9 (20.0)	

*IQR = interquartile range, ICU = intensive care unit, CLABSI = central line associated blood stream infection, UTI = urinary tract infection; **Includes endoscopies and other non-operative procedures.

Longitudinal median regression was used to evaluate differences in WAZ over time while accounting for repeated measurements within patients over time [23]. Median regression is more appropriate for analyzing WAZ than a linear regression model because the median is less sensitive to outliers than the mean and a better summary statistic.

Predictors of changes in WAZ from admission to 1 year follow-up were assessed using multivariable median regression modeling, with results presented as adjusted coefficients with 95% confidence intervals and *p*-values [24]. Statistical significance was assessed as two-tailed $p < 0.05$ for all analyses. Data were analyzed using Stata (version 16.0, Stata Corp LLC., College Station, Texas).

3. Results

Data from 45 consecutive, eligible patients with LGEA were included in the analyses. The median (IQR) gestational age was 36 (34, 37) weeks; 51% were female. Median (IQR) age at the time of the FP was 4 (2.3, 5.8) months (Table 1). Less than half of this cohort ($n = 18$, 40%) was full term, although the majority of all patients were born at greater than 34 weeks gestation (73%). Most patients were either type A (58%) or B (27%), with only 15% of patients classified as Gross type C. Median (IQR) birth weight was 2.3 (1.7, 2.7) kg, which corresponded to a median (IQR) WAZ of -0.96 (-1.55, -0.40). At admission, mild malnutrition was identified in 9 (20%) of the patients, moderate malnutrition in 5 (11%), and severe malnutrition in 4 (9%), by WAZ criteria.

After initial operation (establishment of traction system), patients were started on PN on median (IQR) post-operative day 1 (0, 1), and received PN for 35 (22, 43) days, which accounted for 43% (30, 51) of their total hospital stay. Enteral nutrition was initiated on post-operative day 26 (19, 32) and full EN was achieved by post-operative day 35 (24, 40). All patients had a surgical feeding tube placed prior to beginning the FP.

Pre-operative prescription of energy was median (IQR) 120 (107, 131) kcals/kg, with 2.5 (1.8, 2.8) g/kg of protein. On post-operative day 15 (roughly the mid-point in the FP for most patients), PN provision was typically stable and at goal, with median (IQR) 81 (79, 89) kcals/kg and 3 (3, 3) g/kg of protein. At time of discharge, EN was prescribed for median (IQR) 112 (101, 120) kcals/kg, with 2.1 (1.6, 2.6) g/kg of protein (Table 2).

Median (IQR) length of hospital stay was 79 (55, 130) days, of which 57% (36, 72) was spent in the intensive care unit (ICU) with a median stay of 39 (24, 78) days. Of this time, patients spent a median (IQR) of 14 (10, 20) days on traction, 21 (11, 30) days mechanically ventilated, and 13 (6, 9) days paralyzed. Post-operative complications included esophageal leak requiring intervention ($n = 7$, 16%), chyle leak ($n = 3$, 7%), central line associated blood stream infection (CLABSI; defined by bacteremia, $n = 8$, 18%), and urinary tract infection (UTI; $n = 9$, 20%) (Table 3).

At 1-year follow-up, the majority of patients still had a surgically placed feeding tube (gastrostomy tube = 66%, gastrojejunostomy tube = 5%). However, of the 71% with a surgical tube in place, only 58% were utilizing the access (Table 4).

Median (IQR) growth by WAZ over time is depicted in Fig. 1 (Appendix *A*). Longitudinal analysis using median regression indicated no significant differences in WAZ during hospital stay and following discharge at any time point (Appendix *B*).

Multivariable analysis confirmed that patients with lower WAZ at admission had greater improvement at 1 year (coefficient = 0.91 per 1-unit smaller WAZ at admission (CI 0.37, 1.45), $p = 0.002$) (Appendix *C*). Of the 18 patients with WAZ < -1, 13 were seen at 1-year follow-up. Of these, 85% ($n = 11$) had improvement in WAZ from admission to 1 year. Longitudinal median regression of

Table 4
Feeding outcomes.

	n (%)
Functional Oral Intake Score (FOIS)[22] at 1 year	
FOIS = 1, No oral intake	3/38 (7.9)
FOIS = 2, Tube dependent, some oral intake	7/38 (18.4)
FOIS = 3, Tube dependent, consistent oral intake	12/38 (31.6)
FOIS = 4, All oral intake, single consistency	0/38 (0)
FOIS = 5, All oral intake, requires special preparation	4/38 (10.5)
FOIS = 6, All oral intake, must avoid some specific foods/liquids	3/38 (7.9)
FOIS = 7, all oral intake, no restrictions	9/38 (23.7)
Gastrostomy Tube at 1 year	25/38 (65.8)
Gastrojejunostomy Tube at 1 year	2/38 (5.3)
Reliant on feeding tube to some degree at 1 year (FOIS1–3)	22/38 (57.9)
All oral intake at 1 year (FOIS 4–7)	16/38 (42.1)
Fundoplication	20/45 (44.4)

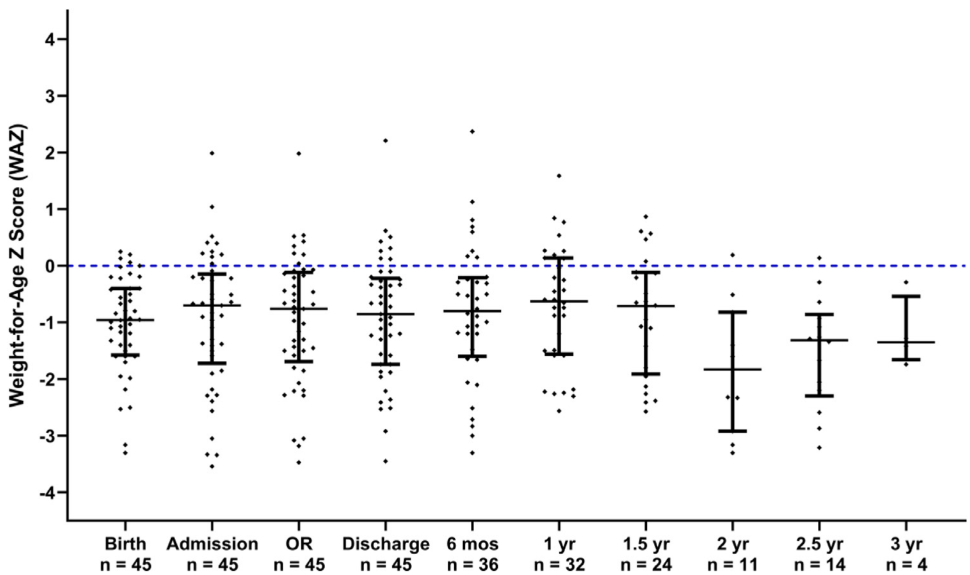

Fig. 1. Growth as described by WAZ from admission to 3-year follow-up. Longitudinal analysis using median regression indicated no significant differences in WAZ during hospital stay and following discharge, for all time points. Median WAZ remained below the 50th percentile (blue) of weight for age from birth through 3 years (For interpretation of the references to color in this figure legend, the reader is referred to the web version of this article.).

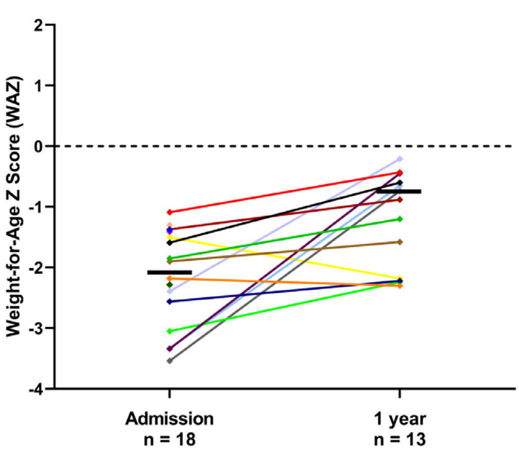

Fig. 2. Growth of patients who presented with WAZ < -1 at admission. Median WAZ at admission and 1 year (horizontal bar) were below average (dashed line). Longitudinal median regression showed improvement in WAZ of median 1.3 z score units (95% CI 0.3, 2.3; $p = 0.01$).

growth in this subgroup demonstrated improvement in WAZ of median 1.3 z score units (95% CI 0.3, 2.3; $p = 0.01$) (Fig. 2).

While the majority of patients improved or maintained their WAZ from admission to 1 year, three patients, who had no evidence of malnutrition on admission (WAZ > 0), had mild to moderate malnutrition at 1 year. Of these patients, 1 had an esophageal leak requiring intervention, and 1 a CLABSI. The third patient had persistent feeding intolerance requiring jejunostomy feeds, was also found to have a type *B* tracheoesophageal fistula (TEF) on post-operative esophagogastroduodenoscopy (EGD), and underwent a Nissen fundoplication and repair of the TEF, four months after the FP.

By univariate analyze, there was no statistically significant association with any of the post-operative complications collected (leak requiring intervention, chyle leak, CLABSI or UTI) and weight or WAZ at time of first operation. In addition, univariate analyze did not reveal a significant association between time to full EN or total days on PN and weight or WAZ at the time of the FP.

There were 10 patients who had CHD requiring intervention, which might increase basal metabolic demands and result in poor growth. While our study is not well powered for such a small subgroup analyze, there was no association between this cohort and admission WAZ, WAZ at time of FP, or a change in WAZ from admission to 1 year.

4. Discussion

Patients with LGEA treated with the FP tend to remain below average in WAZ throughout infancy. However, patients with lower

WAZ on admission demonstrate the most improvement in WAZ by 1 year. Our institution's nutritional strategy (Appendix *D*) likely identifies these high-risk patients promptly and through targeted nutritional rehabilitation helps these patients "catch up" to their peers prior to and during their complex surgical courses.

Despite complicated hospital courses, with relatively long periods of mechanical ventilation, sedation, and paralysis, patients are able to maintain their EN as soon as growth curves in-hospital by providing early PN support and transition to possible. After admission, patients are able to maintain their individual growth curves through careful outpatient follow-up and monitoring, often with ongoing EN support via surgical tube access. Though our long-term follow-up is limited by attrition, the patients seen at 2 and 3 years post-operatively were also able to maintain their WAZ. While the majority are discharged with a surgical feeding tube in place, most are transitioned to full oral feeds between 1 and 2 years after surgery, at which point their feeding tubes are removed.

4.1. Impact of the FP on nutrition delivery

There are many inherent features to the FP that likely affect adequate nutrition delivery and impair growth. Long periods of mechanical ventilation and paralysis can decrease accretion of lean body mass, and restricted caloric provision is needed to accommodate the assumed lower resting energy expenditure. In contrast, increased metabolic demands associated with critical illness and inflammation can increase caloric requirements and the need for catch-up growth or presence of pre-existing or hospital-acquired malnutrition can require hypercaloric prescriptions. Frequent procedures in the operating room (*e.g.*, need for repeat surgical intervention or requirement for surveillance EGD) and/or complications related to the FP can interrupt both PN and EN therapy.

It can be challenging to balance nutritional support during periods of transition between EN and PN, which are sometimes repeatedly required by the clinical course. Limitations to safely weighing patients during and sometimes after the FP can preclude prompt titration of energy prescription and can lead to excessive weight loss or gain. Close oversight by a dietitian and careful monitoring of nutritional variables by the surgical and medical teams is essential to overcome these challenges and provide optimal nutrition to support growth over time during hospitalization.

From a micronutrient perspective, patients undergoing the FP are at risk of bone demineralization and fracture which can result in osteopenia and poor linear growth velocity [25]. Periods of muscle paralysis are inherently associated with a lack of weight-bearing activity, which is a stimulator of bone mineral deposition, though this can be ameliorated by gentle physical therapy with passive range of motion exercises. Loop diuretics, which are often needed to mitigate fluid shifts and pulmonary edema, can further contribute to bone demineralization due to renal calcium and phosphate wasting. Lastly, long term use of PN has been associated with poor bone health, in part due to obligatory limitations in calcium and phosphate prescriptions to avoid risk of precipitation at higher concentration. It is therefore important to optimize calcium and phosphorous provision in PN, to be judicious about the use of diuretics, and to consider transition to EN as soon as clinically feasible [25].

4.2. WAZ trends from admission to 3-year follow-up

In this heterogeneous study population, there was significant variability between each patient's clinical course and individual growth trajectories. We chose a longitudinal analysis to evaluate changes in WAZ from admission to 3-year follow-up to account for repeated measurements within patients over time. Our study results suggest that patients who undergo FP for LGEA management begin their journey smaller than the 50th percentile and some with a certain degree of baseline malnourishment based on below average WAZ on admission (Table 1). With our current nutrition strategy, most patients are able to maintain or achieve adequate (defined as WAZ $>$ -1) growth at least through 1.5 years post-operatively. Notably, there appears to be a down-trend at 2 years postoperatively, although this was not statistically significant (Fig. 1). This downtrend may be associated with transition to full oral feeds and timing of gastrostomy tube removal, which typically occurs between 1 and 2 years post-operatively.

4.3. Factors associated with change in WAZ from admission to 1-year follow-up

When designing our model to evaluate potential predictors associated with WAZ, we considered a threshold WAZ as an outcome; in other words, a model evaluating predictors of WAZ greater or less than -1, which is our definition for adequate WAZ. However, this approach does not take into account improvement over time. For example, a patient with an admission or pre-operative WAZ of -3 would be "penalized" for reaching a WAZ of -1, despite significant growth. On the other hand, evaluating changes in WAZ could theoretically give "credit" to patients who move from a "good" WAZ, for example between 0-1, to a higher WAZ, which may be unnecessary and lack clinical benefit. But because the median WAZ for the entire cohort was less than 0, we felt that evaluating predictors of change in WAZ from admission to 1 year had the potential to provide the most substantive conclusions.

The only predictor of change in WAZ from admission to 1-year follow-up was admission WAZ. When examining this association, those with the lowest admission WAZ scores demonstrated the most significant change over time. These patients were likely identified at admission as needing catch-up growth, and this subset of patients (WAZ $<$ -1) did demonstrate statistically significant improvement in their WAZ (Fig. 2). The few patients whose WAZ had declined at 1 year follow-up had post-operative complications as described above.

It is also reassuring to see that temporary interruptions in nutrition delivery and changes in metabolic demand over time as well as the multiple other challenges to providing adequate nutrition support during the FP as mentioned above do not hinder growth in the long-term.

4.4. Long-term enteral access and growth

We used the Functional Oral Intake Scale to measure feeding outcomes (Table 4). We found that although 73% of patients had a surgical feeding tube at 1-year follow-up, only 58% of patients were utilizing it. This suggests that some patients are transitioning from some degree of tube feed dependence to full oral feeding between 1 and 2 years postoperatively. The decline in WAZ during this time may reflect premature removal of a feeding tube or a temporary sacrifice in optimal nutrition delivery in the interest of ongoing oral progression. Closer follow-up, particularly until patients are on full oral nutrition, could help clarify this issue. Follow-up beyond 1 year was poor in our cohort and limits our ability to examine this on a granular level. The transition and timing from tube dependence to full oral autonomy should be examined more closely in future studies.

4.5. Nutrition strategy

We have a dedicated multidisciplinary Esophageal Atresia and Airway Treatment Center team. Our dietitians work closely with our surgeons and other providers to become familiar with the physiology and clinical course of patients undergoing the FP and have developed a successful nutrition strategy specific to these patients derived from current American Society for Parental and Enteral Nutrition (ASPEN) guidelines as well as center standards of care (Appendix *D*) [26].

Our findings suggest that achieving a target WAZ prior to the first FP operation may not be necessary. We typically wait to start the FP until the infant is at least 3.5 kg to allow for more robust nutritional status, better tissue quality, and improved cardiopulmonary reserve during staged repair. However, we recognize that this is an arbitrary weight cutoff, that we chose based on anecdotal experience with traction related complications in patients who weighed less. While our study is not well powered to stratify complications by WAZ, univariate analyses of leak, chyle leak, CLABSI, and UTI were not associated with either weight at time of initial operation or WAZ with thresholds of -1 and -2 considered. In addition, there was no difference in total days on PN or time to EN for these cohorts. Further research with larger patient populations is necessary to clarify an ideal pre-operative target WAZ, but it is reassuring that our current data does not suggest there are increased rates of post-operative complications with lower weight or WAZ at time of initial operation.

We did find that the few patients whose WAZ declined by 1 year had more complicated courses. These patients should be identified as needing closer interval nutritional assessment both in-hospital and after discharge.

4.6. Limitations

We recognize that our study has several limitations. The data were collected retrospectively and are limited by the completeness of the medical record. We were not able to collect actual calories or protein delivered and instead rely on the macronutrient prescriptions. Though the size of the cohort is not large, given the rarity of the diagnosis, it is still the largest experience reported to date on LGEA patients undergoing the FP. We examine data from a single academic medical center with a highly specialized referral practice of LGEA patients, which limits the generalizability of the study as the FP is not performed by all pediatric surgeons. Nonetheless, we believe that the lessons gleaned from this patient population may apply to other patient populations undergoing prolonged periods of sedation and paralysis, critical illness, or staged surgical repairs.

We included internal traction patients and patients who underwent the FP via a minimally invasive (MIS) approach. These cohorts, as opposed to the external traction FP cohort, are not routinely kept paralyzed during their esophageal growth process and are sometimes fed enterally (post-pyloric) prior to their anastomosis. This difference could have contributed to the heterogeneity of individual patient hospital courses and growth trajectories, but we mitigated this by using longitudinal analysis.

5. Conclusion

We have described our institution's nutrition strategy as applied to a complex patient population, LGEA patients who undergo the FP, which involves reliance on PN during staged repair, careful titration of fluids, judicious use of diuretics, and advancement to EN as early as clinically appropriate. We identified maintenance or improvement in WAZ in a majority of this cohort in-hospital and to 1 year post-operatively, with a small cohort followed to 3 years. It appears that early identification and aggressive nutritional rehabilitation of patients who present with malnutrition allows them to catch up to their peers despite complex surgical courses. There were no statistically significant associations between patient characteristics, perioperative care or complications and long-term growth. However, we recognize the need for closer short-term follow-up to evaluate why WAZ scores remain below average and to ascertain why there is a down-trend at 2 years. While the FP is a unique operation, many aspects of our approach to nutrition could be extrapolated to optimize growth in other critically ill pediatric surgical patients.

Declaration of Competing Interest

The authors have no conflicts of interest relevant to this article to disclose.

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Supplementary materials

Supplementary material associated with this article can be found, in the online version, at doi:10.1016/j.jpedsurg.2021.07.014.

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