



Esophagus

Evolution, lessons learned, and contemporary outcomes of esophageal replacement with jejunum for children



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ABSTRACT

Background: The jejunal interposition is our preferred esophageal replacement route when the native esophagus cannot be reconstructed. We report the evolution of our approach and outcomes.

Methods: The study was a single-center retrospective review of children undergoing jejunal interposition for esophageal replacement. Outcomes were compared between historical (2010–2015) and contemporary cohorts (2016–2019).

Results: Fifty-five patients, 58% male, median age 4 years (interquartile range 2.4–8.3), with history of esophageal atresia (87%), caustic (9%) or peptic (4%) injury, underwent a jejunal interposition (historical cohort $n = 14$; contemporary cohort $n = 41$). Duration of intubation (11 vs 6 days; $P = .01$), intensive care unit (22 vs 13 days; $P = .03$), and hospital stay (50 vs 27 days; $P = .004$) were shorter in the contemporary cohort. Anastomotic leaks (7% vs 5%; $P = .78$), anastomotic stricture resection (7% vs 10%; $P = .74$), and need for reoperation (57% vs 46%; $P = .48$) were similar between cohorts. Most reoperations were elective conduit revisions. Microvascular augmentation, used in 70% of cases, was associated with 0% anastomotic leaks vs 18% without augmentation; $P = .007$. With median follow-up of 1.9 years (interquartile range 1.1, 3.8), 78% of patients are predominantly orally fed. Those with preoperative oral intake were more likely to achieve consistent postoperative oral intake (87.5% vs 64%; $P = .04$).

Conclusion: We have made continuous improvements in our management of patients undergoing a jejunal interposition. Of these, microvascular augmentation was associated with no anastomotic leaks. Despite its complexity and potential need for conduit revision, the jejunal interposition remains our preferred esophageal replacement, given its excellent long-term functional outcomes in these complex children who have often undergone multiple procedures before the jejunal interposition.

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Introduction

In children, the most common indications for esophageal replacement (ER) are complications from long-gap esophageal

atresia and refractory stricture due to caustic ingestion.¹ While it is optimal to preserve the native esophagus, this is not always possible, and ER may be necessary to allow for handling of oral secretions and facilitating adequate oral intake.² Options for ER include gastric (GI), colonic (CI), or small bowel-based interpositions (ie, jejunal [JI]), but there is no current consensus on the best strategy.³

Ideally, when deciding on a conduit for ER, the conduit should closely resemble the esophagus in size and function, should be performed with reproducible surgical techniques, have a low

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incidence of complications, and most importantly, provide consistent long-term functional results.^{4,5} The GI and CI are cited as the most common or preferred operations.^{6,7} Both are less technically demanding than a JI but come with their own set of complications and disadvantages, including significant rates of gastroesophageal reflux, decreased or absent conduit motility leading to stasis, dilation of the conduit, and aspiration with increased pulmonary morbidity.^{2,6–12} In comparison, the JI most closely resembles the esophagus in size and is thought to retain its intrinsic peristalsis, allowing for improvement in conduit emptying with decreased incidence of reflux. This may also decrease the burden of pulmonary disease.^{2,3,13,14}

Despite these potential advantages, the JI has not been widely adopted. Some reasons may include the technical and time-consuming nature of the procedure, the concern for conduit loss, and the reported high risk for anastomotic leak and stricture.^{5,8,10} Systematic reviews and meta-analyses, due to their small sample sizes, have failed to provide definitive conclusions about the benefits of the JI compared to the other conduits.^{5,6} Nonetheless, the International Network of Esophageal Atresia working group, in a position paper on long-gap esophageal atresia, proposed the JI as the best option for ER with the caveat that, due to its technical complexities, performance should occur at centers of expertise.²

Our group has adopted the JI as our preferred approach to ER since 2010, and our volume of JIs has grown substantially since our initial report.¹³ With increased experience, we have modified several aspects of our technique and perioperative care to decrease the surgical risks. In this report, we assess the benefits of these changes and present our outcomes.

Methods

Study design

This was a single center, institutional review board–approved, retrospective review of all patients who underwent a JI for ER between the years of 2010 and 2019. Patients who underwent surgery from January 2010 to December 2015 have been previously reported¹³ and were used as the historical cohort for comparison with the contemporary patients (surgery between Jan 2016 and Apr 2019).

Selection of operative approach

Patients are considered for a JI if they lack a functional esophagus, either secondary to refractory severe esophageal dysmotility or due to lack of adequate esophageal length from either a congenital (esophageal atresia) or acquired insult (caustic or peptic injury or postoperative complications). Patients referred to our Esophageal and Airway Treatment Center for ER are often complex and have typically undergone multiple prior surgical procedures. They undergo a multidisciplinary airway and gastrointestinal evaluation, which consists of a flexible and rigid laryngoscopy and dynamic 3-phase tracheobronchoscopy,^{15,16} as well as a flexible esophagogastroduodenoscopy with fluoroscopic contrast esophagogram (or gap-o-gram if the esophagus is discontinuous). The airway evaluation is as important as the esophageal evaluation, as it assesses for supraglottic issues, laryngeal cleft, vocal cord function, subglottic pathology, airway compression, tracheobronchomalacia, recurrent or acquired tracheoesophageal fistula, or other airway pathology that may need to be addressed concurrently or in anticipation of the planned esophageal work. All patients judged to be candidates for JI undergo a neck and chest computed tomography angiogram, occasionally including the abdomen if indicated, for surgical planning.

The length and condition of the proximal and distal functional esophagus, the patients' nutritional status, and associated comorbidities are key in determining the optimal approach and sequence of events for ER. Patients with some length of healthy proximal and distal functional esophagus are first considered candidates for traction-induced esophageal growth (Foker process).¹⁷ However, patients in whom the distal esophageal segment is missing or is not healthy, those who have repeatedly failed the Foker process, or those with a poorly functioning or failed prior ER are considered for a JI.

Single-stage, "short-segment" JI are considered for patients with a functional proximal esophagus that reaches at least to the mid-chest (roughly to the level of the carina); these patients do not receive microvascular augmentation (so called "supercharging") of the conduit. Select patients with a short but healthy upper esophageal segment can still be considered for a short-segment JI when traction-induced proximal esophageal growth to the level of the carina is achieved. We selectively apply this strategy for children who are under 10 kilograms, as we generally prefer to perform our long-segment, supercharged JI in children above 10 kilograms due to the blood vessel diameter needed for the anastomosis.

Given the complexity of the procedure, patients considered for a long-segment, supercharged JI are approached in either a sequential or a delayed fashion. Our sequential approach is reserved for patients with good nutritional status and an acceptable comorbidity profile that we believe will tolerate 2 major operative days in the same week. In this setting, the first operative day consists of a thoracic esophagectomy via a thoracotomy, any necessary posterior airway work, and a temporary cervical esophagostomy. The second operative day, often 2 days later, includes a laparotomy, sternotomy, neck dissection, harvest of the jejunal conduit, microvascular augmentation, and the restoration of gastrointestinal continuity. For patients in whom their comorbidity profile or nutritional status are considered high risk, we stage the repair by performing the thoracic esophagectomy first. We then allow the children time to recover, optimize their nutritional and pulmonary status, and have them return after their condition has improved (often months later) for the completion of their JI (delayed approach, see Fig 1 for treatment algorithm).

Surgical technique

After the dysfunctional or unhealthy esophagus or prior ER conduit has been removed (as described above, ideally on a separate day), we begin the reconstruction. A heparin drip of 10 units per kilogram per hour (U/kg/h) is started at the time of the skin incision. The cervical esophagus is mobilized off the trachea through a left cervical incision unless an esophagostomy is present on the right side (Fig 2, A–C). The recurrent laryngeal nerve (RLN) is identified and protected. We currently use RLN monitoring to minimize the risk of injury to the nerve as the cervical esophagus is mobilized to the side of the trachea, under the strap muscles and sternocleidomastoid muscle. We use NIM TriVantage EMG endotracheal tubes (ETT) (Medtronic, Jacksonville, FL) for children able to fit an ETT size 5.0 mm (inner diameter) or greater. For children who need an ETT that is 4.5 mm or smaller, we employ Dragonfly (Neurovision, Ventura, CA) dual channel surface electrodes that are trimmed to size and fixed to a generic ETT. In addition to this, we use continuous vagal stimulation with Automatic Periodic Stimulation (Medtronic, Minneapolis, MN). All 2 systems feed into the NIM-Response 3.0 Nerve Monitoring System (Medtronic).¹⁸

A sternotomy, thymectomy (to make room for the conduit), and any great vessel or anterior tracheal work required to treat tracheobronchomalacia is then completed. Dissection of the left or right internal mammary vessels is performed (Fig 2, D). Vessels are

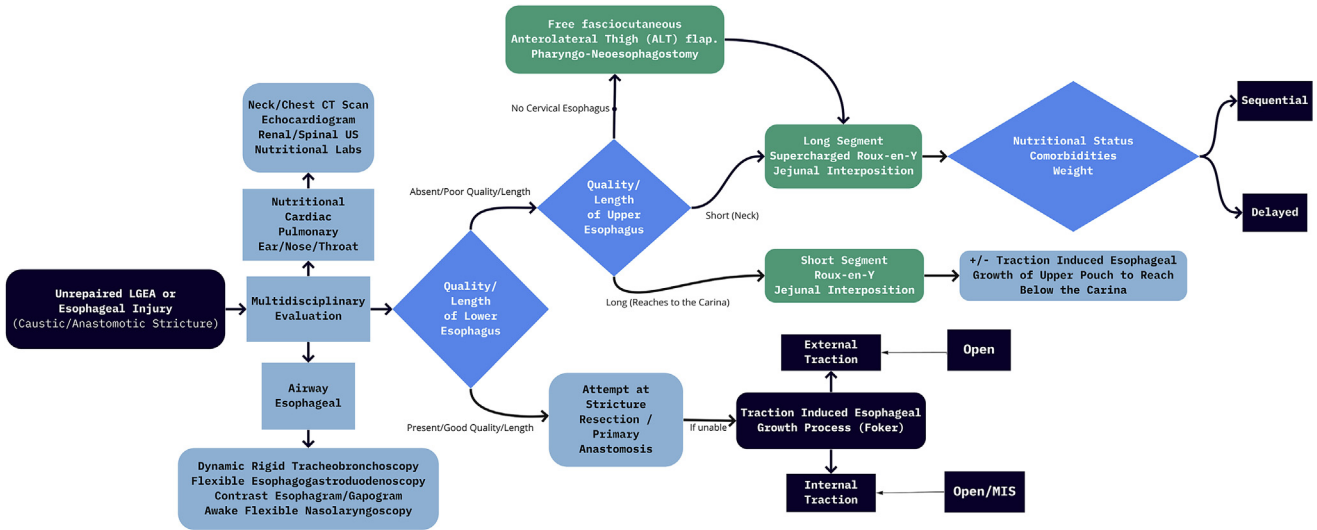


Fig. 1. Treatment algorithm for consideration of JJ. JJ, jejunum interposition; LGEA, long gap esophageal atresia; MIS, minimally invasive surgery.

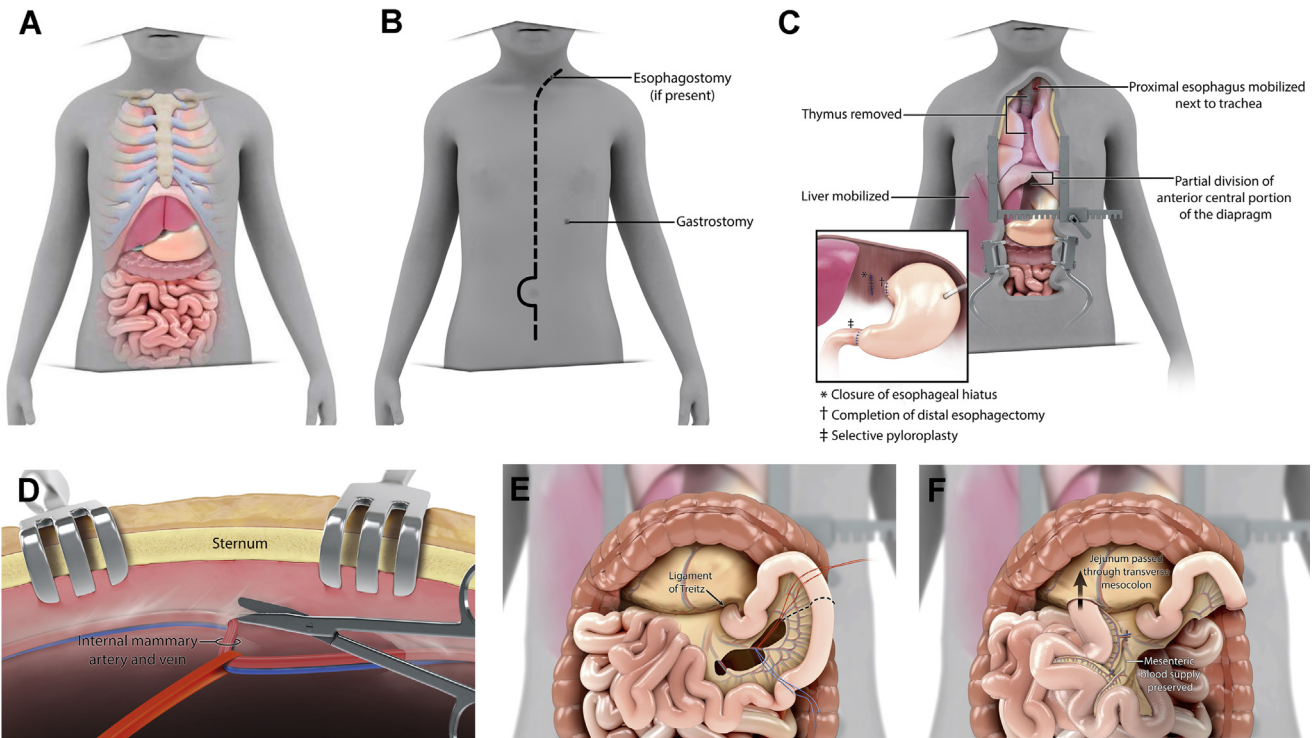


Fig. 2. (A) Normal anatomy. (B) Location of laparotomy, sternotomy, and extended cervical incisions with gastrostomy and esophagostomy locations. (C) Exposure after a cervical incision with proximal esophagus mobilized off of the trachea, sternotomy including thymectomy, and partial division of the anterior central portion of the diaphragm, laparotomy including liver mobilization, closure of the esophageal hiatus, completion distal esophagectomy, and selective pyloroplasty. (D) dissection of the internal mammary vessels (right or left) for use in microvascular augmentation of jejunal conduit. (E) The jejunal vascular arcade is carefully examined, the marginal arcade is left intact, and the appropriate jejunal vessels are selected for microvascular augmentation. (F) The jejunum is divided and passed via a retrocolic, antegastric, and substernal route into the anterior mediastinum. (G) With the jejunum in the anterior mediastinum, the microvascular anastomosis is performed between the internal mammary artery and vein and the jejunal artery and vein. The arterial anastomosis is performed in an end-to-end fashion using interrupted 9-0 nylon suture, while the venous anastomosis is performed using a venous coupler device to account for size mismatch between the jejunal and internal mammary veins. The device used is the Microvascular Anastomotic COUPLER Device and System IFU (Synovis Micro Companies Alliance, Inc, Birmingham, AL). (H) Liver mobilization to provide adequate space for the jejunal conduit. (I) Esophago-jejunal anastomosis performed in an end-to-end hand-sewn, single layer approach, using nonabsorbable, monofilament sutures (polypropylene). (J) Roux-en-Y jejunum-jejunum anastomosis fashioned in an end-to-side hand-sewn, double layer configuration. (K) Final configuration of jejunal conduit before closure. (L) Drains are left in the anterior mediastinum on the opposite side of the anastomosis and in the abdomen to monitor for chyle leak given the dissection at the root of the mesentery.

left in continuity until needed. Meanwhile, a laparotomy, lysis of adhesions (if required), takedown of any enteral tubes, and other intestinal reconstruction as needed is completed before the microvascular team prepares the jejunal vessels, leaving the

marginal arcade intact and typically only taking 1 (occasionally 2) major jejunal branches off the superior mesenteric artery and vein (Fig 2, E). A heparin bolus of 50 U/kg is given before dividing any vessels. The jejunum is divided and brought up across the anterior

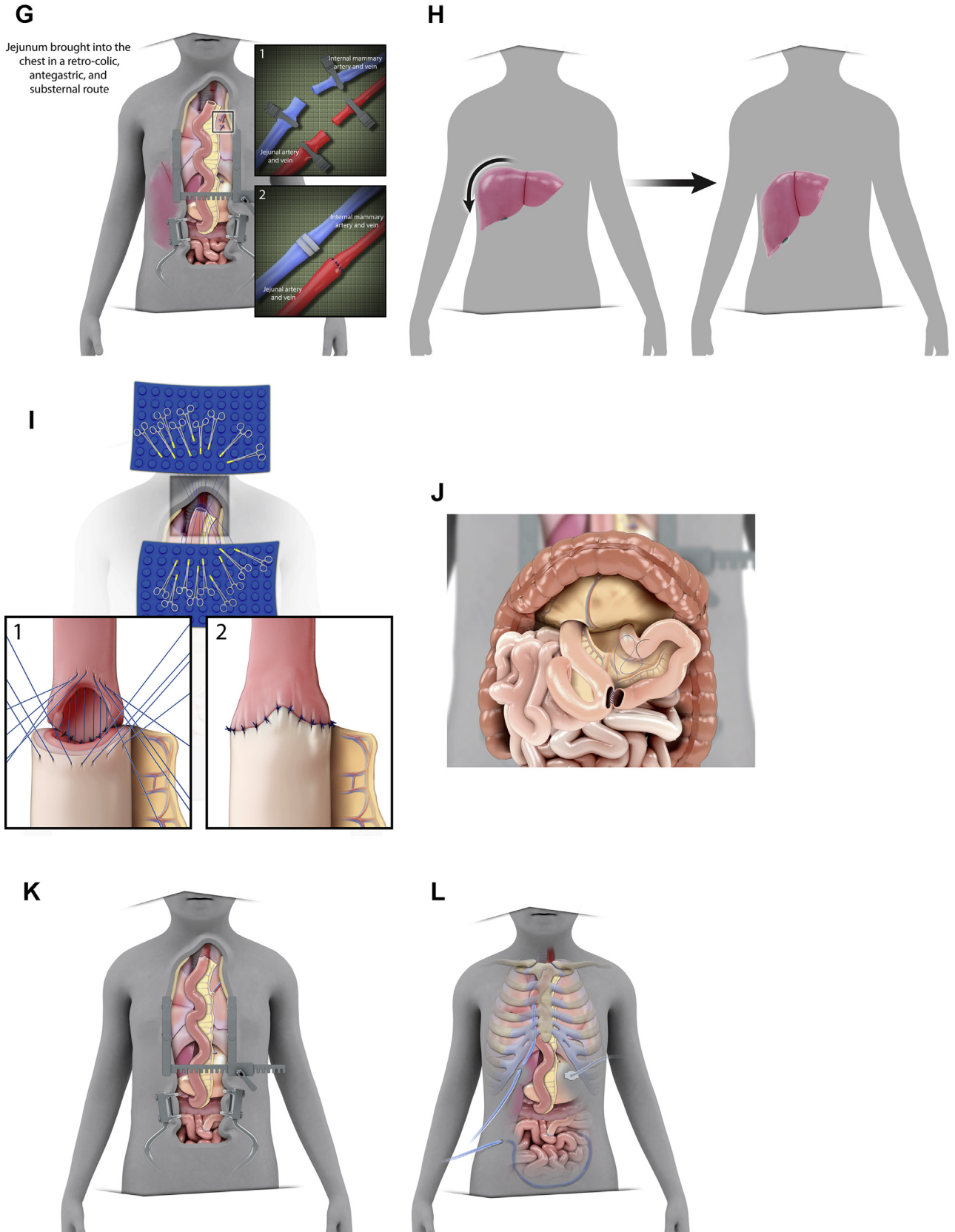


Fig. 2. (Continued).

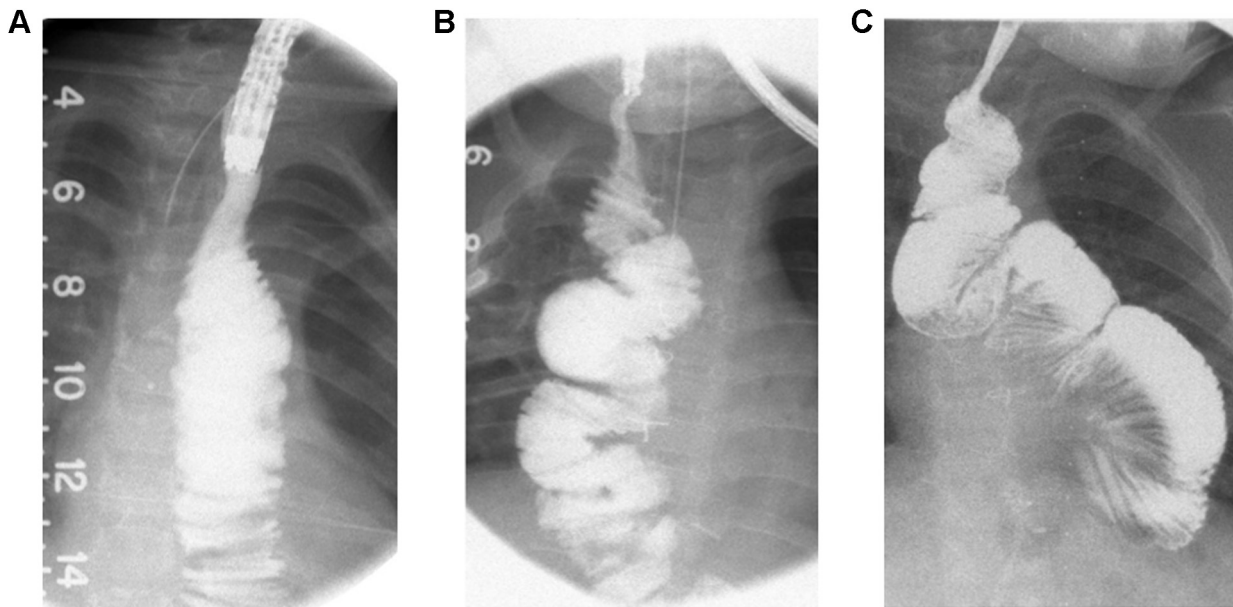


Fig. 3. JI tortuosity: Contrast studies of JI at time of surveillance upper endoscopy. These were graded based on their degree of tortuosity and classified as (A) straight, (B) mildly tortuous, and (C) tortuous. JI, jejunal interposition.

mediastinum via a retro-colic, ante-gastric, substernal route (Fig 2, F).

For supercharging, the internal thoracic artery and vein are most frequently used with an end-to-end microsurgical anastomosis to the jejunal mesenteric vessels^{19,20} (Fig 2, G). When bringing the jejunum into the mediastinum, if full mobilization of the left lobe of the liver and division of the midline anterior portion of the diaphragm is not adequate to provide space for the conduit, a partial resection of the left lobe of the liver may be performed to prevent stretching of the jejunal mesentery as it enters the lower anterior mediastinum (Fig 2, H). All esophago-jejunal anastomoses are performed in an end-to-end, hand-sewn, single-layer approach, using nonabsorbable, monofilament sutures (polypropylene), while our Roux-en-Y jejuno-jejunostomy is fashioned in an end-to-side, double-layered configuration (Fig 2, I and J). Occasionally, an additional mesentery-sparing segmental resection of the jejunal conduit is undertaken, when there is too much redundancy of the conduit in the mediastinum. Flexible endoscopy is performed to assess patency and integrity of the anastomosis with CO₂ insufflation. We then assess anastomotic perfusion quality with indocyanine green image capture technology (SPY-PHY; Stryker, Kalamazoo, MI).²¹ Most patients have had some form of gastrostomy tube in the past, but if not, one is placed and a pyloroplasty is considered to facilitate gastric emptying if there is any prior evidence of gastric feeding intolerance. Once the reconstruction is complete, meticulous hemostasis and closure of all mesenteric gaps and potential internal hernia locations is performed. If a pleural space was entered, we attempt to close it to avoid the conduit being pulled into a pleural cavity, as this can lead to dilation or poor functional outcome due to increased tortuosity. One drain is left in the abdomen to monitor for a chyle leak given the dissection at the root of the mesentery; 1 drain is left in the anterior mediastinum on the side opposite the microvascular work, and 1 drain is placed in the pleural cavity if it was entered (Fig 1, K and L).

Perioperative management

Postoperatively, patients initially are monitored and managed in the intensive care unit (ICU). We strive to extubate patients as

soon as possible. The heparin drip (10 U/kg/h) is maintained for the first 3 days as patients are transitioned to aspirin prophylaxis (81mg/day) for 1 month. We try to avoid use of vasopressors and prefer volume for resuscitation guided by central venous pressure and arterial pressure monitoring. Patients are fed via their gastrostomy tube as soon as they have bowel function. An esophagogram is performed a week postoperatively, and endoscopic surveillance of the anastomosis is performed approximately 1 month postoperatively, and then annually for at least the first few years. Our feeding team begins working on oral feedings as soon as it is safe from a respiratory standpoint. We attempt to perform a flexible nasolaryngoscopy on all patients to evaluate their postoperative vocal cord function.

Data collection

Demographic, intraoperative, and postoperative variables, including outcomes and complications, were collected from the medical record for this review. Anastomotic leaks were defined as any contrast extravasation seen on the postoperative contrast study. Refractory strictures were defined as those requiring advanced interventions (>8 dilations, stent placement, endoscopic incisional therapy, or stricture resection).^{22–24} Complications were graded based on the modified Clavien-Dindo classification.²⁵ We defined clinically serious complications as grade III (surgical, endoscopic, or radiologic intervention) subdivided as grade IIIa (not under general anesthesia) or grade IIIb (under general anesthesia) and grade IV (life-threatening complication requiring ICU management). Contrast studies of the JI were reviewed to determine the degree of tortuosity of the JI and classified as straight, mildly tortuous, and tortuous (Fig 3). The preoperative and postoperative feeding status of patients was determined using a scale adapted from the functional oral intake scale (Table I)^{26,27} to account for sham feeding in patients who had an esophagostomy before undergoing a JI. Follow-up information and the most recent feeding status were determined based on their most recent clinic visit. Follow-up occurs at 3 months postoperatively and yearly for at least the first few years.

Table I
Preoperative and postoperative oral feeding status*

Feeding status	Description	Historical cohort (n = 14)		Contemporary cohort (n = 41)	
		Preop	Postop	Preop	Postop
0	NPO	8 (57%)	n/a	6 (15%)	n/a
1	Esophagostomy, with PO tastes	0	n/a	7 (17%)	n/a
2	Esophagostomy, consistent PO intake limited consistencies	1 (7%)	n/a	10 (24%)	n/a
3	Esophagostomy, consistent PO intake, all consistencies	0	n/a	6 (15%)	1 (2%)
4	Feeding tube dependent, with PO tastes	1 (7%)	2 (14%)	1 (2%)	9 (22%)
5	Feeding tube dependent, consistent PO intake, limited consistencies	2 (14%)	3 (21%)	5 (12%)	9 (22%)
6	Feeding tube dependent, consistent PO intake, all consistencies	0	1 (7%)	0	7 (17%)
7	Full PO intake with limited consistencies	0	0	3 (7%)	0
8	Full PO intake of all consistencies	2 (14%)	8 (57%)	3 (7%)	15 (37%)

NPO, nothing by mouth; PO, by mouth; *postop*, postoperative; *preop*, preoperative.

* Scale modified from the functional oral intake scale^{26,27} to account for sham feedings (feeding status 1–3).

Table II
Demographic, surgical, and intraoperative characteristics

Demographics	Historical (n = 14) median (IQR) Or n (%)	Contemporary (n = 41) median (IQR) Or n (%)	P value
Gestational age (wk)	35 (33,38)	36 (32.5,38)	.78
Male sex	10 (71%)	22 (54%)	.27
Type of esophageal abnormality			.51
Type A EA	7 (50%)	11 (27%)	
Type B EA	1 (7%)	4 (10%)	
Type C EA	5 (36%)	20 (49%)	
No EA (peptic stricture or caustic injury)	1 (7%)	6 (15%)	
VACTERL	4 (29%)	11 (27%)	.89
Major chromosomal abnormality*	4 (29%)	5 (12%)	.14
Number of previous surgeries	6 (4,12)	4 (2,6)	.03
History of Foker procedure at OSH	4 (29%)	7 (17%)	.33
History of Foker procedure at BCH	7 (50%)	9 (22%)	.049
Esophagostomy before JI	4 (29%)	27 (66%)	.02
History of gastric pull-up	2 (14%)	10 (24%)	.44
History of colonic interposition	3 (21%)	2 (5%)	.07
Age at surgery (y)	6.6 (2.3, 14) [†]	4 (2.2, 6.5) [‡]	.36
Surgical characteristics [§]	Historical (n = 15 procedures) n = 15	Contemporary (n = 41 procedures) n = 41	
Duration of operation (min)	827 (598, 991)	700 (627, 749)	.03
Supercharged anastomosis	6 (40%)	33 (80%)	.004
Esophagojejunal anastomosis			.01
End-to-end	10 (67%)	38 (93%)	
End-to-side	5 (33%)	3 (7%)	
Length of jejunal conduit			.01
Short	7 (47%)	6 (15%)	
Long	8 (53%)	35 (85%)	
1 d long jejunum	1 (12.5%)	2 (5%)	
2 d long jejunum	2 (25%)	8 (20%)	
Delayed long jejunum	5 (62.5%)	25 (61%)	
Distal jejunal conduit anastomosis			.07
Straight to stomach	6 (40%)	7 (17%)	
Roux-en-Y	9 (60%)	34 (83%)	
Other surgical adjuncts			
Sterno-clavicular resection	7 (50%)	25 (61%)	.46
Partial liver resection	4 (28.6%)	16 (39%)	.48
Pyloroplasty	4 (28.6%)	16 (39%)	.48
Mesentery-sparing segmental resection	0	3 (7%)	.30

Bold values indicate statistical significance.

BCH, Boston Children's Hospital; CLOVES, congenital lipomatous (fatty) overgrowth, vascular malformations, epidermal nevi and scoliosis/skeletal/spinal anomalies; EA, esophageal atresia; IQR, interquartile range; JI, jejunal interposition; OSH, Outside Hospital; VACTERL, vertebral, anorectal, tracheoesophageal, renal and limb anomalies.

* eg, Trisomy 21, anophthalmia-esophageal-genital syndrome, q11 deletion, CLOVES syndrome.

[†] Range—8 mon to 23 y.

[‡] Range—12 mon to 25 y.

[§] One patient underwent jejunal interposition twice due to failed conduit on the first attempt; therefore, n = 15 in this group.

^{||} Short JI—esophagectomy and esophagojejunal anastomosis performed same day, esophagojejunal anastomosis in the right chest near carina, often not supercharged. Long Sequential—esophagectomy and esophagojejunal anastomosis performed same day or within 72 hours, esophagojejunal anastomosis in the neck, often supercharged. Long Delayed—esophagectomy and esophagostomy done during prior hospitalization or at outside hospital, esophagojejunal anastomosis in the neck, often supercharged.

Statistical analysis

Descriptive and summary statistics are provided when applicable. Categorical variables are expressed as frequencies and percentages, while continuous variables are expressed as medians (interquartile ranges [IQR]). Fisher exact and Mann-Whitney *U* tests were used to

compare variables between groups (historical versus contemporary) and to examine predictors of consistent oral intake postoperatively (ie, preoperative feeding status and degree of JI tortuosity). Statistical analyses were carried out using STATA 15.2 (StataCorp, College Station, TX) with some graphical representations performed in Microsoft Excel (Microsoft Corporation, Redmond, WA).

Table III
Complications

Complication	Historical (n = 14)	Contemporary (n = 41)	P value
Overall (at least 1 complication)	9 (64%)	24 (59%)	.76
Pneumonia/tracheitis	5 (36%)	9 (22%)	.30
Subcutaneous wound infection	4 (29%)	8 (19.5%)	.46
Readmission within 30 d	1 (7.5%)	5 (12%)	.64
Chyle leak	0	5 (12%)	.18
Reintubation	2 (14%)	5 (12%)	.85
Internal hernia*	2 (14%)	5 (12%)	.85
Transdiaphragmatic	2 (14%)	4 (10%)	.68
Mesenteric	0	1 (2%)	.60
Pleural/pericardial/effusion†	0	4 (10%)	.22
Clostridium difficile infection	2 (14%)	3 (7%)	.43
DVT	1 (7%)	3 (7%)	.99
CLABSI	1 (7%)	3 (7%)	.99
Small bowel obstruction*	1 (7%)	2 (5%)	.78
Adhesive	1 (7%)	1 (2%)	.37
Ischemic stricture	0	1 (2%)	.60
Deep space infection‡	0	2 (5%)	.40
Bile leak	0	2 (5%)	.40
Anastomotic leak	1 (7%)	2 (5%)	.78
Conduit loss	1 (7%)	1 (2%)	.37
UTI	1 (7%)	1 (2%)	.37
Horner's syndrome	0	1 (2%)	.60
Drain erosion‡	0	1 (2%)	.60
Pressure ulcer §	0	1 (2%)	.60
Major bleeding event*	2 (14%)	1 (2%)	.08
Chest washout	0	1 (2%)	.60
Seizure	2 (14%)	0	.02
Meningitis	1 (7%)	0	0.09

Bold values indicate statistical significance.

CLABSI, central line associated blood stream infection; DVT, deep vein thrombosis requiring anti-coagulation; UTI, urinary tract infection.

* Required operative intervention.

† Required intervention (eg, drain).

‡ Required multiple endoscopic interventions for a drain erosion through the duodenum.

§ Required prolonged wound care.

Results

We identified 55 patients, with a median age of 4 years (IQR 2.4–8.3 years, and range 8 months to 25 years), who underwent 56 JIs for ER between 2010 and 2019. Fourteen patients (25%) underwent surgery between the years of 2010 and 2015 (historical cohort); the remaining 41 patients (75%) underwent surgery between the years of 2016 and 2019 (contemporary cohort). Our median overall length of follow-up for the entire cohort was 1.9 years (IQR 1.1, 3.8), with 1.5 years (IQR 0.9, 2.3) for the contemporary cohort and 5.4 years (IQR 4.3, 5.9) for the historical cohort ($P < .001$). Two patients were lost to follow-up.

There was no significant difference in the demographic variables between the historical and contemporary cohorts (Table II). Patients with an esophagostomy before the JI were more commonly in the contemporary cohort (29% vs 66%; $P = .02$), whereas those in the historical cohort had, on average, more operations before their JI (median 6 vs 4.9; $P = .039$) and more often had undergone the Foker process before JI (50% vs 22%; $P = .049$). There were significant differences between cohorts in how the esophago-jejunal anastomosis was performed; it was more likely for the contemporary cohort to have an anastomosis be end to end rather than end to side (93% vs 67%; $P = .01$) and created in the neck rather than in the chest (85% vs 53%; $P = .01$), likely reflecting shorter proximal esophageal segments. In the contemporary cohort, the JI also was more frequently supercharged (80% vs 40%; $P = .004$). Although more patients in the contemporary cohort had a Roux-en-Y reconstruction rather than one straight to the stomach, this difference was not statistically significant (83% vs 60%; $P = .07$). Only 1 of the last 31 JI was performed directly to the stomach; the remaining 30 were all in a Roux-en-Y configuration.

The contemporary cohort had significantly shorter median duration of postoperative chemical paralysis (0 vs 5 days; $P < .001$), intubation (6 vs 11 days; $P = .01$), ICU stay (13 vs 22 days; $P = .03$), and hospital stay (27 vs 50 days; $P = .004$) when compared to the historical cohort; the operative time (11.7 hours to 13.4 hours; $P = .024$) was also shorter in the contemporary group.

Complications

The majority of patients in the contemporary and historical cohorts experienced at least 1 complication (59% vs 64%, respectively; $P = .74$, Table III), of which 37% and 29% were deemed clinically serious ($P = .59$). The proportion of clinically serious complications did not vary by type of JI ($P = .57$, Table IV). Similarly, almost half of all patients have undergone a reoperation (47%) at a median of 82 days after JI (IQR: 15, 372). Patients undergoing a delayed long JI were significantly less likely to require a reoperation (30%) than those undergoing a sequential short-interval long jejunum (69%) or a short-length jejunum (69%; $P = .01$). Reoperations varied from operative management for wound infections to stricture resections or small bowel obstructions (Table V). Most reoperations (62.5%) were elective operations that were undertaken to improve the functional status of the conduit, such as conversion from straight interposition to Roux-en-Y configuration (poor conduit emptying, $n = 3$; poor gastric emptying, $n = 1$; severe reflux and aspiration, $n = 1$) or vice versa (improve satiety and facilitate gastrostomy tube removal, $n = 1$; treatment of a recurrent transdiaphragmatic hernia, $n = 2$), while 37.5% of the reoperations were unplanned (emergency or semielective), of which the

Table IV
Major Complications for historical and contemporary cohorts combined

Complication	Short JI* (n = 13)	Long JI sequential (n = 13)	Long JI delayed (n = 30)	P value†
Anastomotic leak	3(23%)	0	0	0.01
Esophagojejunal anastomotic stricture resection	3 (23%)	1 (8%)	1 (3%)	0.11
Refractory stricture‡	4 (31%)	2 (15%)	2 (7%)	0.12
Reoperation	9 69%	9 (69%)	9 (30%)	0.01
Complication (any)	9 69%	9 (69%)	16 (53%)	0.48
Grade III or IV Clavien-Dindo complication	4 31%	6 (46%)	9 (30%)	0.57

Bold values indicate statistical significance.

ANOVA, analysis of variance; JI, jejunal interposition.

* Short JI—esophagectomy and esophagojejunal anastomosis performed same day, esophagojejunal anastomosis in the right chest near carina, often not supercharged, Long Sequential—esophagectomy and esophagojejunal anastomosis performed same day or within 72 hours, esophagojejunal anastomosis in the neck, often supercharged. Long Delayed—esophagectomy and esophagostomy done during prior hospitalization or at outside hospital, esophagojejunal anastomosis in the neck, often supercharged.

† P value refers to the comparison of the 3 groups using ANOVA.

‡ Refractory stricture—anastomotic stricture requiring at least 1 of the following: resection, >8 dilations, or advanced endoscopic management (stent, steroid injection, endoscopic incision).

Table V
List of reoperations for both historical and contemporary cohort

Reoperations	Historical (n = 14); n (%)	Contemporary (n = 41); n (%)	P value
Conduit revision	6 (43%)	10 (24%)	.18
Esophagojejunal anastomotic stricture resection	1 (17%)	4 (40%)	
Change in configuration	4 (67%)	4 (40%)	
Straight to Roux-en-Y*	2	3	
Roux-en-Y to Straight†	2	1	
Revision of esophagojejunal anastomosis end-side to end-end	1 (17%)	1 (10%)	
Revision of jejuno-gastric anastomosis	0	1 (10%)	
Internal hernia	2 (14%)	5 (12%)	.85
Transdiaphragmatic	2	4	
Mesenteric	0	1	
Wound management for SSI	1 (7%)	3 (7%)	.99
SBO	1 (7%)	2 (5%)	.78
Adhesive	1	1	
Ischemic stricture	0	1	
Pyloroplasty	1 (7%)	1 (2%)	.37
Resection of jejunal conduit	1 (7%)	1 (2%)	.37
Bleeding	2 (14%)	1 (2%)	.08
Vascular clip retrieval	1 (7%)	0	.09
Vocal cord granuloma excision‡	0	1 (2%)	.60

SBO, small bowel obstruction; SSI, surgical site infection.

* Reasons for conversion from straight to Roux-en-Y: poor conduit emptying, $n = 3$; poor gastric emptying, $n = 1$; severe reflux and microaspiration, $n = 1$.

† Reasons for conversion from Roux-en-Y to straight: improve satiety and facilitate gastrostomy tube removal, $n = 1$; treatment of a recurrent transdiaphragmatic hernia, $n = 2$.

‡ Multiple failed extubations, successful extubation after granuloma excision from vocal cord

majority were for an internal hernia (number or percentage to stay consistent).

The anastomotic leak rate (5% vs 7%; $P = .78$), rate of stricture resection (10% vs 7%; $P = .74$), and need for reoperation (46% vs 57%; $P = .48$) were similar for the contemporary versus historical cohorts, respectively. However, for all supercharged JIs, not a single leak occurred (0% vs 18% without supercharging; $P = .007$). Overall, there were only 2 patients who experienced conduit loss (3.6%). The first, early in our experience, did not undergo supercharging, has since received a successful redo-JI, and is fully orally fed. Our second patient was a young adult with a history of 2 prior failed ERs (CI and JI) at other institutions with limited reconstructive options and very poor pulmonary status. We attempted a JI in her but were unable to get satisfactory additional blood flow to her conduit and her JI failed, leaving her with an esophagostomy.

Vocal cord assessments were performed in 27 patients (49%). There was no significant difference in assessments performed in the contemporary cohort compared to the historical cohort (56% vs 29%; $P = .14$). Five patients had a preexisting tracheostomy at the

time of their JI, 1 for bilateral vocal cord paresis, 2 due to caustic injury with severe scarring of the larynx, and 2 for inability to wean off the ventilator. Six additional patients, without a tracheostomy, had either unilateral or bilateral vocal cord paresis preoperatively that remained unchanged after their JI. Postoperatively, the majority (74%) of patients assessed had a normal or unchanged exam from baseline. However, 2 patients (7%) had a new temporary unilateral paresis that resolved before their repeat evaluation, while 7 patients (26%) had a new unilateral paresis that had either not resolved by the time of their repeat evaluation or had not been re-evaluated.

Feeding and motility outcomes

All patients ($n = 55$) successfully handle their oral secretions. Most patients ($n = 43$; 78%) are either fully (40%) or predominantly (38%) orally fed (Table I). Patients who had oral intake preoperatively, including those with an esophagostomy (sham feedings), were more likely to achieve full or consistent oral intake

Table VI
 JI modifications/lessons learned

Modifications/lessons learned	Rationale/alternative
1. Microvascular augmentation or supercharging is critical if long JI is needed	Decreased anastomotic leak rate likely from improved perfusion and venous drainage
2. End to end esophagojejunal anastomoses	Avoids the progressive dilation of the blind end and poor functional outcome associated with the end-to-side configuration
3. Avoid hemi-manubriectomy, partial first rib resection whenever possible	Prevents bullfrogging. Thymectomy provides extra space for conduit
4. Roux-en-Y drainage configuration (end to side jejuno-jejunostomy)	Better conduit drainage, possibly less reflux when compared to straight to stomach configuration. However, dumping symptoms may occur, hence dietary counseling and monitoring is imperative
5. Meticulously close all potential internal hernia locations	Internal hernias are common after JI, particularly transdiaphragmatic
6. Consider JI earlier in the course of a patient after failed EA repairs	Repeated Foker process attempts carry significant risks with diminishing returns
7. Consider delayed JI strategy for a high risk patient (particularly those with poor nutritional or pulmonary status)	Less risk of reoperation. Improved nutritional status. May provide the opportunity to improve oral feeding skills
8. Routine use of intraoperative RLN monitoring, and pre and postoperative assessments of vocal cord function	JI patients are at high-risk for pre-existing or new RLN nerve injury. RLN injury can impact ability to reach full oral intake
9. Preoperative oral feedings whenever safe and possible	Patients with preoperative oral intake (even sham feedings in patients with esophagostomy) can more rapidly reach consistent postoperative oral intake

EA, esophageal atresia; JI, jejunal interposition; RLN, recurrent laryngeal nerve.

postoperatively (preoperative feeding 87.5% vs not 64%; $P = .04$). Twelve patients (all in the Roux-en-Y configuration) have reported symptoms consistent with dumping syndrome; most ($n = 11$) are mild and are being managed with dietary changes. However, 1 patient still has lifestyle-limiting symptoms and is being considered for conversion from a Roux-en-Y to direct to stomach configuration.

Review of contrast esophago-jejunograms showed that the majority of conduits are straight (48.1%) or mildly tortuous (44.4%), with only 7.4% being classified as tortuous. The degree of conduit tortuosity was not significantly different between JI configurations (Roux-en-Y versus direct to stomach; $P = .49$) or JI cohorts (historical versus contemporary; $P = .49$) and did not appear to affect a patient's ability to reach full or consistent oral intake (straight versus mildly tortuous or tortuous; $P = .83$). Eight patients who were making either slow or no progress with oral feeding, and without obvious endoscopic or radiographic mechanical obstruction, underwent a nuclear medicine esophageal transit study to evaluate motility of the JI. All 8 patients demonstrated varying degrees of delayed transit through their JI. Five patients ($n = 3$, straight to stomach, $n = 2$ Roux-en-Y) had normal or near normal emptying of the proximal portion of the JI, whereas the mid or distal portion demonstrated retention but cleared with time or with additional swallows. These patients are being managed with anticipatory guidance and feeding therapy. Three patients ($n = 1$, straight to stomach; $n = 2$, Roux-en-Y) demonstrated poor transit through their entire graft; of these, 1 patient improved clinically and has achieved full oral intake, while the other 3 underwent conduit revisions ($n = 1$ straight to Roux-en-Y conversion, $n = 1$ diaphragmatic hernia repair) with improvement in symptoms.

Discussion

We report the largest experience to date in the literature of JI for ER in children. Over time, several technical and perioperative refinements have substantially decreased ICU and hospital duration of stay and decreased the risk of anastomotic leak. Future considerations need to address our complication rate and rate of reoperation. Although the JI is technically challenging, we have previously shown feasibility,¹³ and we continue to demonstrate excellent function long term.

Anastomotic leak, stricture, and conduit loss

One of the major criticisms of the JI has been its perceived increased risk of anastomotic leak, stricture, and conduit loss. Reported leak rates have varied widely from 0% to 60%.^{1,8,10,28–30} We have shown that it is possible to achieve a very low rate of anastomotic leak (overall 5.5%). Our leaks were all in nonsupercharged short JI. Notably, none of our patients with a supercharged long JI experienced a leak, highlighting the importance of blood flow and venous drainage to an anastomosis. However, supercharging may not be the only factor, as adult JI series with supercharging have reported leak rates of 13% to 32%.^{29,31} Although it is important to note that not all supercharging is the same, adult JI series advocate division of the marginal arcade to straighten the conduit^{29,31,32}; we prefer to preserve the marginal arcade and only divide 1 (or rarely 2) jejunal branches. These differences make transposition more difficult but preserve perfusion and avoid intraoperative ischemia. We also do not leave an external monitoring flap because we assess intraoperative perfusion with indocyanine green and maintain the patients on a low dose heparin drip until conversion to 30 days of aspirin prophylaxis.

Similarly, reported stricture rates vary widely from 16% to 73%,^{4,5,7,30} and although variability in the definition of a stricture exists, our rate of refractory anastomotic strictures remains very low, particularly for our long JI cohort (9%). Given that anastomotic leaks are a significant risk factor for refractory strictures, efforts to improve rates of leak are likely to result in lower rates of stricture. Furthermore, conduit loss appears to be strongly related to suboptimal blood flow to the conduit, as demonstrated in the 2 patients who lost their conduits. Our rate of conduit loss (3.6%) is at the lower range of reported rates (0%–37.5%).^{1,10} With improved perfusion by supercharging of our conduits, we expect this to become a rare occurrence.

Complications, reoperations, and lessons learned

Our complication rate, although not trivial, is also not surprising given the magnitude of the operation that is performed on a patient population that has significant risk factors at baseline. Patients often come for an ER as their last option, after having had multiple failed operations to achieve esophageal continuity. Only 13% of our population had an esophagostomy as their only operation, and none underwent a JI as their initial operation. Other JI series in

children have reported complication rates that range from 45% to 93%.^{8,10,30} Although the majority of our complications were infection related, requiring treatment with antibiotics alone (ie, pneumonias, tracheitis, or wound infections), and were not clinically serious, nearly half of the patients required a reoperation, the most common being a conduit revision. About 15% of these patients required a change in their drainage configuration from gastric to Roux-en-Y or vice-versa. An additional 13% of patients developed an internal hernia requiring operative intervention with nearly all of these being transdiaphragmatic, either at the esophageal hiatus if a short JI or substernal if a long JI was performed. Because of these issues, we have switched to performing predominantly Roux-en-Y reconstructions and spending additional time meticulously closing all potential internal hernia defects, in particular the retroperitoneal dissection area, and anchoring the conduit to the diaphragm and substernal space.

It is possible that some of our wound morbidity is related to the length of our incision. Those who perform JI in adults advocate doing so without a sternotomy.^{29,32} We prefer the sternotomy approach because it (1) allows for thymectomy and wide exposure of the anterior mediastinum, (2) permits harvest of the internal mammary vessels for supercharging the jejunal conduit, (3) provides access for concomitant anterior great vessel or airway work that is sometimes needed to treat associated tracheomalacia or vascular compression, and (4) provides excellent visualization of the entire conduit to ensure that it is as straight as possible, with minimal tension, without pressure points, and with excellent perfusion.

One must be cautious with the use of the “short” JI, although they are appealing for patients who have an upper esophageal length that reaches the mid-chest. The jejunal conduit in these cases can be brought with relative ease through the esophageal hiatus into the posterior mediastinum via the right chest and avoids the associated sternotomy and microvascular work used in the long JI. Ironically, with the short JI, the tension, blood flow, and venous drainage are often suboptimal, and our outcomes in this cohort are worse (increased leak and stricture rates) than what occurred in the cohort who received the long supercharged JI. Nonetheless, it is an option to consider in carefully selected patients.

Although some of our patients came to us with established vocal cord injuries, it is clear that the JI (as well as other forms of ER) can risk injury of both RLNs. Some of our injuries have been transient, but others have persisted after repeat evaluations and are likely permanent. These injuries carry significant morbidity as they are often associated with aspiration and may limit the potential for full oral intake despite a fully functional JI conduit. Although the use of RLN monitoring systems in routine, nonoperative thyroid surgery remains controversial,³³ there is evidence to suggest that they do add value to high-risk patients.³⁴ Hence, we now routinely employ RLN monitoring systems.

Early in our experience, we performed an end-to-side esophago-jejunal anastomosis. In addition, in order to avoid any sort of pressure on the anastomosis, we would perform an ipsilateral hemi-manubriectomy, as advocated by the adult JI literature.^{29,32} What we encountered, however, was that the residual “blind-end” jejunum beyond the anastomosis would become progressively dilated causing both dysphagia and food impaction. Similarly, the lack of structural support over the anastomosis resulted in an unpleasant “bullfrogging” or neck distention with larger boluses of food or during Valsalva. Many of these initial patients have required partial revision of their conduit with conversion to an end-to-end anastomosis and either nearby tissue transfer (ie, muscle) or biologic mesh placement to address the bullfrogging, a problem similarly dealt with by Ring et al.³⁵ To prevent this, we have transitioned to performing all of our esophago-jejunal anastomoses

end-to-end, placing the conduit under the strap muscles and moving the conduit to run adjacent and parallel to the trachea. We avoid the hemi-manubriectomy whenever possible.

As our experience with the JI has grown and we have recognized the significant risks encountered by patients who have undergone multiple attempts at a “rescue” Foker procedure,¹⁷ we now consider a JI as an earlier option. This is particularly true if the quality of the remnant lower esophagus is suboptimal. This transition in our approach is reflected in our data, as we are performing fewer rescue Foker procedures, and patients are receiving a JI with fewer prior operations. As a result, a greater proportion of patients receive their JI with an established esophagostomy in place. This last point also exemplifies our transition to more readily consider a delayed long jejunum strategy if we think a patient’s comorbidity (particularly infectious and pulmonary status) and nutritional profile would improve with esophageal diversion as opposed to pursuing a sequential JI with 2 large operations within the same week. This extra time to recover between diversion and restoration of esophageal continuity has allowed several patients to acquire substantial feeding skills, which in turn allows them to achieve full oral intake more rapidly after the JI. This delayed strategy is further supported by the lower reoperative rate in this cohort, likely related to better nutritional and pulmonary status.

Our multidisciplinary team has been able to streamline the JI process significantly despite the complexity of the operation and the patients’ frequent comorbidities. Our operating time has decreased by more than 2 hours, although it is still a median duration of nearly 12 hours. Patients in the historical cohort remained intubated, sedated, and chemically paralyzed after surgery out of concern for anastomotic leak. In our current regimen, no routine postoperative paralysis is used, and patients are extubated as soon as possible based solely on their respiratory needs. As a result, the duration of intubation has been reduced nearly in half. This transition has led to decreased duration of sedation weans, quicker tolerance of enteral diet, and shorter hospital stays.

The most important aspect of the JI process may be the critical review of the procedures and outcomes, as well as review of other centers’ results, in a process of constant improvement, which can only occur in high-volume centers. Critical input in the planning stages from gastroenterology, nutrition, radiology, social work, and nursing are considered in the surgical plan, which is then discussed extensively with the critical care, pulmonary, and feeding teams to create a final comprehensive plan. The lessons we have learned in this review process are outlined in [Table VI](#). The core surgical team consists of experienced Esophageal and Airway Treatment Center pediatric general surgeons and pediatric plastic and microvascular surgeons, flexibly augmented by pediatric cardiac surgery and pediatric ear, nose, and throat specialists as indicated to optimize surgical outcomes.

Feeding outcomes

Our feeding outcomes are consistent with that of other JI series in children,^{10,30,35} in which the majority of JI patients can achieve a full or predominant oral intake status. Feeding outcomes are likely strongly related to the age at which patients undergo restoration of esophageal continuity and their degree of underlying oral aversion. To address this, patients should be encouraged to sham feed whenever safely possible when they have an esophagostomy, and they should be referred to a center of excellence for complex esophageal reconstruction as early as possible in their clinical course to minimize the development of oral aversion. In addition, avoiding RLN injury and its attendant increased risk of aspiration is critical to facilitating oral intake and explains our recent dedication to having RLN monitoring during every case.

Limitations

This is a retrospective single-center design, which comes with its inherent biases. Our population is very heterogeneous, and patients were referred to us after failed operations elsewhere; hence, our results may not be reflective of other centers. Nonetheless, this study represents the largest series of children, adolescents, and young adults undergoing an ER that has been reported. This provides us the ability to identify trends that have led to multiple changes in our surgical management and subsequent improved outcomes. Due to geographic restrictions, not all patients have had a recent clinic follow-up; therefore, some of the feeding outcomes might actually be better than reported since only the last reported feeding assessment was recorded for this analysis. Though we largely attribute the improvement in the rate of anastomotic leak to supercharging, it is possible that it may be due to other factors, such as advancements in critical care, operating team experience, or patient characteristics. One could attempt to statistically adjust for such potential confounders, but given that there were few leaks, multivariable modeling would lead to unreliable estimates. We also recognize that given the multiple univariate associations tested, it is possible that some of the *P* values could have been significant due to chance; however, given the exploratory nature of these analyses and the strong associations observed, we believe that statistical adjustments are not likely to influence the results or conclusions of this study. We have only recently begun to implement more formal pre and postoperative vocal cord examinations on all our patients; hence, our true incidence of vocal cord injury is unknown. Lastly, we have not standardized the motility assessments of our JI patients. Only patients with poor feeding status underwent nuclear medicine transit studies; hence, we cannot make firm conclusions on the motility status of the JI conduits in patients with consistent oral intake. Nonetheless, as a proxy for motility, we evaluated conduit tortuosity and found that most conduits were straight or only mildly tortuous; however, it is clear that these JI will need to be monitored long term.

Future directions

We have established procedural success, improved clinical outcomes, and shown excellent long-term functional feeding outcomes. Nonetheless, several areas require additional research. For example, patient-reported outcomes, such as patient's quality of life, caregiver burden, and financial impact, need further attention. Nutritional outcomes, such as growth, vitamin deficiencies, and dumping syndrome, are also topics that require additional assessment.

Because our children will carry these JI conduits for their lifespan, we need to develop systems for long-term monitoring to detect and address issues as they become older. For example, Lee et al have developed a report card that is used in adult JI patients to keep track of areas such as emotional well-being, body image, diet, and swallowing issues, dumping symptoms, and effects on their social life in order to longitudinally follow these patients and to identify issues if they arise.³⁶ We are in the process of developing such metrics and longitudinal follow-up capabilities.

Finally, it has been hypothesized that the improved emptying potential of the JI conduit could lead to improved pulmonary status, an area in which many GI or CI patients suffer due to reflux and aspiration resulting from poor emptying of a dilated conduit.¹¹ Hence, future studies must evaluate long-term pulmonary status.

In conclusion, the JI is a complex and challenging operation for children, adolescents, and young adults. We have demonstrated that microvascular augmentation improves the perfusion of the proximal jejunal conduit and has drastically decreased the risk for

anastomotic leak. We have made multiple modifications based on our experience as outlined above. We have decreased the duration of paralysis, intubation, and ICU and hospital stay. Preoperative oral intake, even if sham feeding, is a significant determinant for early consistent oral intake after reconstruction. Despite the complexity and potential morbidity of the procedure, we have shown that excellent outcomes can be achieved when children with complex esophageal pathology are treated at high-volume centers by an experienced multidisciplinary team.

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Conflict of interest/Disclosure

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