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Endoscopic incisional therapy and other novel strategies for effective treatment of congenital esophageal stenosis

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ABSTRACT

Background & aims: Congenital esophageal stenosis (CES) is an inborn condition of the esophagus that can be refractory to endoscopic dilation. Surgical intervention is not curative, with patients experiencing frequent ongoing need for therapy for anastomotic stricture postoperatively.
We hypothesized that novel methods of endoscopic CES management including endoscopic incisional therapy (EIT) would lead to less surgical intervention.
Methods: We retrospectively reviewed the medical records of all patients with CES treated by our tertiary care center who had at least one endoscopy between July 2007 and July 2019. Statistical comparison of cohorts who underwent advanced endoscopic therapy involving EIT versus traditional endoscopic therapy with balloon dilation was performed. Primary outcome measure was need for surgical intervention.
Results: Thirty-six patients with CES met inclusion criteria. Thirty-four ever had at least one endoscopic intervention such as balloon dilation, steroid injection, stenting, and/or endoscopic incisional therapy (EIT) at their CES. Esophageal vacuum assisted closure (EVAC) was used for treatment or prevention of esophageal leak. Odds of surgical intervention were significantly lower in the group who received therapeutic endoscopy with EIT (odds ratio (OR) 0.1; $p = 0.007$). Clinical feeding outcomes were similar in the endoscopic and surgical management groups. Odds of complications after therapeutic endoscopies involving EIT were significantly greater than those without EIT (odds ratio 6.39; 95% confidence interval (2.34, 17.44); $p < 0.001$), though our rates of esophageal leak significantly decreased over time as our use of EVAC increased (Spearman's $\rho = -0.884$; $p = 0.004$).
Conclusion: Complementary endoscopic techniques such as EIT broaden the toolbox of the treating physician and may allow for avoidance of surgery in CES.
Level of evidence: Level III.

Congenital esophageal stenosis (CES) refers to a collection of congenital conditions of the esophagus caused by either tracheobronchial remnants (TBRs), fibromuscular thickening (FMT), or membranous webs (MWs) [1]. Management of CES often involves balloon dilation with reversion to myotomy, stricture resection or esophageal replacement in refractory cases [1–7]. Some centers have published surgical intervention rates as high as 71% [2], noting the often recalcitrant nature of CES to endoscopic dilation therapy alone [1,2,6–9]. However, surgical intervention often fails to produce durable response, with anastomotic strictures requiring ongoing dilation and stenotic symptoms continuing to plague more than half of patients in previously published series [8,10].

Complementary advanced endoscopic techniques such as esophageal incisional therapy (EIT), stenting, and vacuum-assisted closure significantly broaden the endoscopic toolbox of the treating physician, and may allow for avoidance of surgical intervention in some cases of CES. We have previously described our experience with EIT as a promising means of treating benign refractory esophageal strictures, even in severe cases [11].

We sought to review our experience with EIT, stenting, and EVAC in patients with CES treated at our institution to gain insights into the role of these advanced endoscopic techniques in this patient population. We hypothesized that the use of these advanced endoscopic techniques in their treatment would lead to less need for surgical intervention.

Abbreviations: CES, Congenital esophageal stenosis; TBR, tracheobronchial remnant; FMT, fibromuscular thickening; MW, membranous web; EA, esophageal atresia; EUS, endoscopic ultrasound; EIT, endoscopic incisional therapy; SEPS, self-expandable plastic stent; FCSEMS, fully covered self-expandable metal stent; EVAC, esophageal vacuum assisted closure; IQR, interquartile range.

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1. Methods

This study was approved by our institutional review board. We retrospectively reviewed the electronic medical records of all patients with CES treated by our tertiary care referral center who underwent at least one upper endoscopy between July 2007 and July 2019. All endoscopies were performed by one of two pediatric gastroenterologists using either Olympus XP-190N or Olympus GIF-H190 at the discretion of the endoscopist. All patients with a diagnosis of CES with at least one endoscopy during the study period were included for analysis.

1.1. Endoscopic ultrasound

In cases where endoscopic ultrasound (EUS) was performed, an Olympus 20 MHz endoscopic ultrasound probe UM-S20-17S (Olympus, Center Valley, PA USA) was advanced to the site of the stricture through the working channel of the endoscope. The esophagus was filled with normal saline to provide an adequate interface between the probe and the esophageal wall. Cartilaginous components (reported to be variably hypoechoic [12,13] versus hyperechoic [5,14] in the literature), thickness of the esophageal wall, and esophageal wall layers were identified by their respective characteristic ultrasonographic appearances by an experienced radiologist or endoscopist.

1.2. Endoscopic incisional therapy

We have previously described our method of endoscopic incisional therapy (EIT) [11]. Briefly, in cases where EIT was performed, a Huibregtse needle knife papillotome (Cook Medical, Winston-Salem, NC USA) was advanced into the working channel of the endoscope and directed towards the preferred area for incision. Current was generated by use of the ERBE ICC 200 or ERBE VIO 300 D electrosurgical generator (ERBE, Tübingen, Germany) with settings of 100 W (effect 2 or 3) or Endo Cut I (effect 2, cut duration 2, cut interval 3), respectively. Incisions were made preferentially along the posterior wall of the esophagus. An approximately 1 cm linear incision was first made in order to expose the underlying congenital fibrous and/or cartilaginous tissue. Once the underlying tissue was exposed, further incisions were made using a "lift and cut technique" in which the needle knife was carefully advanced underneath the fibrous or cartilaginous tissue and lifted anteriorly towards the center of the esophageal lumen (see Fig. 1). Then, the cut current was applied to incise the tissue. This technique was repeated until the ring of fibrous or cartilaginous tissue appeared nearly obliterated at that site. The entire method of exposing the abnormal congenital tissue followed by additional lifting and cutting was then repeated at a site approximately 2 cm adjacent to the first incision. On many occasions, after EIT was completed, balloon dilation was performed to further spread the tissue and open the esophageal lumen.

1.3. Esophageal stent

Stents were either self-expandable plastic stents (SEPSs) or fully covered self-expandable metal stents (FCSEMSs). Owing to patient size, airway stents, vascular stents, or biliary stents were sometimes placed (see Supplemental Materials).

1.4. Esophageal VAC therapy

We have previously described our method of esophageal vacuum assisted closure (EVAC) device assembly and placement (see Supplemental Materials) [15]. EVAC sponges were replaced every 4 to 7 days until sufficient mucosal healing as judged by bleeding granulation tissue and no fluoroscopic evidence of leak was achieved.

1.5. Treatment algorithm

Prior to the introduction of EIT at our center, endoscopic attempts at treating congenital strictures most often involved balloon dilation with or without steroid injection. Once EIT was introduced, our EIT treatment strategy for CES between 2012 and 2015 was to first perform EIT and then immediately place a fully covered stent to prevent or treat a leak and to allow the incisional area to heal without restenosis. After stent removal, follow up balloon dilations, intralesional steroid injections, and EIT were performed as needed. With the development of the EVAC technique in 2015, our treatment strategy shifted to EVAC placement instead of stent placement immediately after EIT. EVAC placement

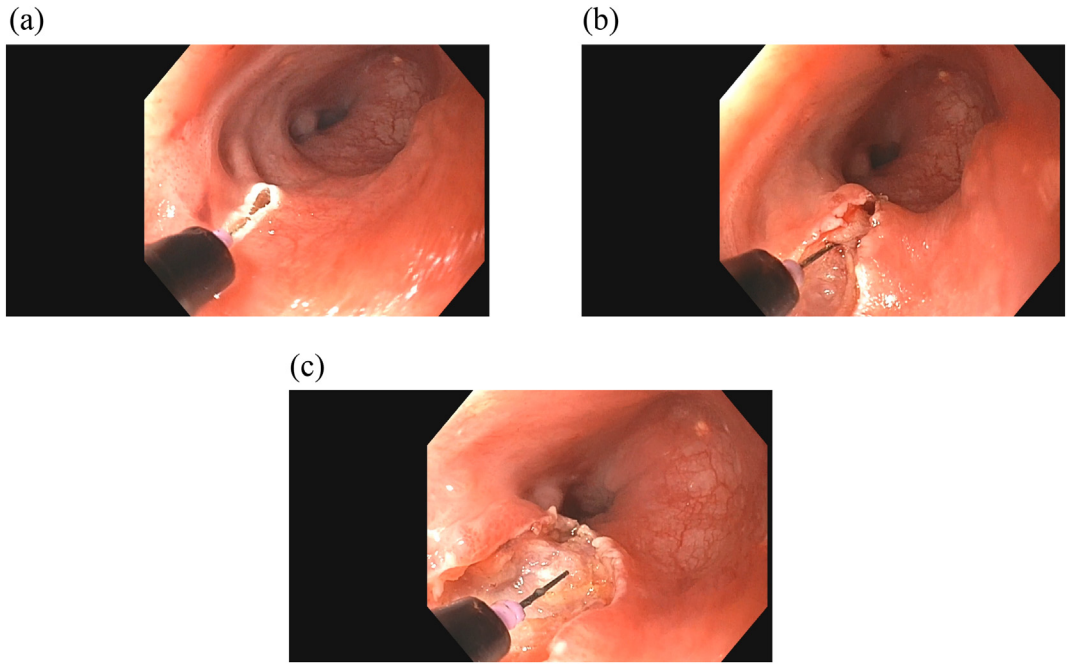

Fig. 1. Endoscopic incisional therapy (EIT) at a congenital esophageal stricture. After a 1 cm incision is made to expose the congenital fibers (A), the needle knife is carefully inserted into the congenital fibrous or cartilaginous tissue and lifted towards the center of the esophageal lumen prior to application of the cut current to incise the abnormal fibers (B). The cut is then spread open with balloon dilation (C).

was similarly performed to prevent or treat esophageal leak. After EVAC removal, follow up balloon dilations, stenting, intralesional steroid injections and EIT were performed as needed.

1.6. Outcome measures

Our primary outcomes were endoscopic and clinical success at most recent follow up endoscopy and clinic visit, respectively. Secondary outcome measures were need for surgical intervention and endoscopic complications.

Symptoms were assessed by caregiver interview at the time of routine clinic visits and recorded in the medical record. Clinical success was defined as full oral feeding of all age-appropriate textures with symptoms of dysphagia occurring once per month or less. Partial clinical success was defined as partial or full oral feeding with symptoms of dysphagia occurring at most 1–2 times per week. Clinical failure was defined as not meeting criteria for full or partial success.

1.7. Statistical analysis

Continuous data on demographics and clinical characteristics are presented as median (interquartile range (IQR)) and categorical data are presented as frequency (percentage). Fisher's exact test was used for the comparisons of rates of specific therapies among patient subgroups. Owing to the relatively small sample size, and therefore small number of patients experiencing the outcome variables (need for surgery and/or having complications), univariate regression results are presented. Logistic regression analysis was used to analyze the odds of reverting to surgery for each covariate among patients who ever received endoscopic therapy at their CES. Generalized Estimating Equations (GEEs) were used to analyze the risk of complications during therapeutic endoscopy performed at a native CES. A binomial family and logit link function were assumed in the GEE modeling for this dichotomous outcome. GEE modeling was used to take into account multiple observations per patient. Results of all regression analyses are presented with odds ratios, 95% confidence intervals and P values. The nonparametric Spearman's ρ (rho) correlation coefficient was used to assess the correlation between esophageal leak rate versus EVAC usage rate. A two-tailed alpha level of 0.05 was implemented to determine statistical significance, and Stata (version 15.0, StataCorp LLC., College Station, Texas) was utilized to perform all statistical analyses.

2. Results

2.1. Cohort characteristics

Patient characteristics are summarized in Table 1. A total of 36 patients with history of congenital esophageal stricture and at least one endoscopy at our institution were included. Twenty-one patients (58%) had a history of esophageal atresia (EA). Median age at CES diagnosis was 12 months (interquartile range (IQR) 2–19 months). Most patients were diagnosed with congenital esophageal stenosis after developing vomiting and feeding difficulties with introduction of solid foods in infancy; however, 10 patients (28%) were diagnosed within the first two months of life by studies performed for other reasons such as EA.

Most patients underwent more than one endoscopy at our center (94%). Patients underwent a median of 5.5 therapeutic endoscopies (IQR 2–8) at our center during the study period. Median length of follow up time from initial endoscopy through most recent endoscopy at our institution was 14 months (IQR 8–32) and median clinical follow up time was 25 months (IQR 11–50).

2.2. Type of CES

Twenty-seven (75%) of 36 patients underwent EUS to attempt to identify their subtype of CES. Regions of altered echogenicity suggestive of cartilaginous tracheobronchial remnants were found in 2 (7%) patients; 25 patients (93%) had no cartilaginous remnants identified suggestive of fibromuscular stenosis.

Table 1
Clinical characteristics of CES cohort.

Clinical Data	Median (IQR) or *n* (%)
Male gender	19 (53%)
History of esophageal atresia	21 (58%)
Age at CES diagnosis, months	12 (2–19)
Endoscopic ultrasound of CES performed	27 (75%)
• Fibromuscular	25 (93%)
• Tracheobronchial remnant	2 (7%)
Length of clinical follow up, months	25 (11–50)
Length of endoscopic follow up, months	14 (8–32)
Number of endoscopies per patient	8 (5–12)
Number of endoscopies per patient with at least one therapeutic maneuver:	5.5 (2–8)
• Dilation	4 (2–7.3)
• EIT	1 (0–2.3)
• Corticosteroid injection	1 (0–2.3)
• Stent placement	1 (0–1)
• EVAC placement or replacement	0 (0–1)

Therapeutic maneuvers included balloon dilation, endoscopic incisional therapy (EIT), corticosteroid injection, stent placement, or esophageal vacuum assisted closure (EVAC) device placement or replacement. Therapeutic endoscopies are not necessarily independent events and some endoscopies involved more than one therapeutic intervention.

Six patients had histopathology specimens of their CES available from 5 stricture resections and 1 endoscopic biopsy of submucosal material exposed during EIT; 4 patients had tracheobronchial remnants and 2 patients were classified as fibromuscular thickening by histopathology. Four of these 6 patients had previously undergone EUS; 2 patients who were initially classified by EUS as FMT were subsequently classified by histopathology as TBR, and 2 patients had concordance between EUS and histopathology confirming FMT.

2.3. Endoscopic and surgical treatments

Median numbers of endoscopic therapeutic maneuvers per patient are summarized in Table 1. Twenty-one patients underwent EIT at their CES; 13 patients underwent non-EIT therapeutic endoscopy at their CES. All patients who underwent attempts at endoscopic therapy received at least one dilation.

Our practice involves use of stents and/or EVAC in conjunction with EIT to promote a larger diameter esophageal lumen and prevent the esophageal incisions from healing in a collapsed position; thus we expected EIT therapy to cluster with stenting and EVAC usage. Exposure to EIT was significantly associated with exposure to stenting (17/21 vs 0/13, $p < 0.001$) and to EVAC therapy (13/21 versus 0/13, $p < 0.001$).

A total of 13 patients (36%) ever underwent surgical intervention ($N = 5$ stricture resections, $N = 6$ surgical myotomies, $N = 2$ jejunal interpositions) for their CES. Only two patients underwent surgery without any attempts at endoscopic therapy at any institution owing to previously planned airway surgeries and decision to surgically manage the CES during the same thoracotomy.

2.4. Clinical outcomes

Of 21 patients who underwent EIT at their CES, 17 (81%) achieved full ($N = 16$) or partial ($N = 1$) clinical feeding success with endoscopic therapy alone; 3 (14%) needed surgical intervention to achieve full ($N = 3$) feeding success; 1 nonsurgical patient does not yet eat by mouth owing to oral aversion despite an endoscopically adequately treated CES. Of 13 patients who underwent non-EIT therapeutic endoscopy at their CES, 5 (38%) achieved full feeding success with endoscopic therapy alone; 8 (62%) required surgical intervention (3 full feeding success, 4 partial success, and 1 does not eat by mouth owing to airway

Table 2
Univariate logistic regression modeling for odds of surgical intervention in patients who ever received endoscopic therapy at their CES.

Variable	Odds Ratio	95% CI	P value
EA Type			
• No EA	Reference	.	.
• Type C / LGEA	0.92	(0.22, 3.92)	0.914
Exposure to EIT at CES	0.1	(0.02, 0.55)	**0.007***
Presurgical exposure to stent	0.12	(0.02, 0.69)	**0.017***
Presurgical exposure to EVAC	0.09	(0.01, 0.84)	**0.034***
Initial stricture size (millimeters)	0.71	(0.51, 0.98)	**0.040***
Type of CES			
• FMT	Reference	.	.
• TBR	1.63	(0.26, 10.09)	0.602

Statistically significant results are denoted by boldface type and an asterisk (*). EA, esophageal atresia; LGEA, long-gap esophageal atresia; EIT, endoscopic incisional therapy; CES, congenital esophageal stenosis; EVAC, endoscopic vacuum assisted closure; FMT, fibromuscular type; TBR, tracheobronchial remnant; CI, confidence interval.

comorbidities). The two patients who went straight to surgical intervention without ever having therapeutic endoscopy attempted at their congenital strictures are clinical feeding failures; one patient with history of EA and esophageal dysmotility tolerated only liquids by mouth and was unable to advance to purees or solids, and the other patient does not eat by mouth owing to airway comorbidities.

Need for surgical intervention was significantly less likely in the group who received EIT compared to patients who received non-EIT endoscopic therapy (OR 0.1; $p = 0.007$). However, rates of full or partial clinical feeding success were not statistically significantly different between the groups who were managed with EIT (17/18) versus surgical therapy (10/13) (Fisher's exact test, $p = 0.284$).

All 13 patients who ever had surgical intervention at their CES required ongoing endoscopic dilations at their surgical anastomoses or myotomy sites, undergoing a median of 2 postsurgical dilations (IQR 1–4) and 1 postsurgical nondilation endoscopic therapy (IQR 0–2) such as steroid injection ($N = 7$ patients), stenting ($N = 4$ patients), or EIT ($N = 5$ patients) during the study period.

2.5. Clinical predictors of need for surgical intervention

Since all patients who had endoscopy performed prior to any CES surgical intervention (if ever performed) experienced at least one endoscopy with dilation and at least one endoscopy with steroid injection, these maneuvers were not included in the regression analyses. In a univariate logistic regression analysis performed on the subset of patients who ever had attempts at endoscopic therapy at their CES ($N = 34$), exposure to EIT, stenting, and EVAC were significantly associated with reduced odds of need for surgical intervention (Table 2). In addition, increasing initial diameter size of the congenital stricture was significantly associated with reduced odds of need for surgery.

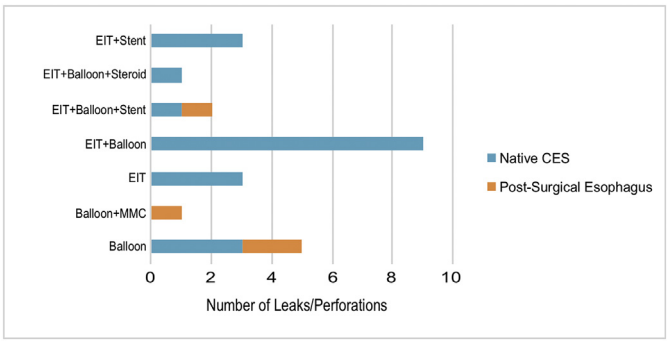

Fig. 2. Combinations of endoscopic therapies associated with esophageal leaks or perforations. EIT, endoscopic incisional therapy; balloon, balloon dilation; MMC, application of mitomycin C; CES, congenital esophageal stenosis.

Multivariable logistic regression performed with the same set of predictors did not identify any significant independent effects of each of the predictors of need for surgical intervention owing to lack of power (data not shown). Type of CES and history of EA were not significantly associated with surgical reversion in any model.

2.6. Complications

Twenty-eight endoscopies (9%) were associated with complications in 18 unique patients. Twenty-four complications occurred in native esophagus prior to any surgical intervention (24/245 endoscopies); 4 complications occurred during therapeutic endoscopy at an anastomosis from prior surgery for CES (4/84 endoscopies). Complications included esophageal leak ($N = 18$), stent migration ($N = 4$), and stent edge erosion with perforation ($N = 2$) (Fig. 2).

Of 24 endoscopies performed at a native CES associated with a complication, 18 (75%) involved EIT. Odds of complications after therapeutic endoscopies involving EIT were significantly greater than therapeutic endoscopies without EIT (odds ratio 6.39; 95% confidence interval (2.34, 17.44); $p < 0.001$).

Leaks or perforations were managed with EVAC therapy ($N = 11$), stent placement ($N = 4$), antibiotic monotherapy ($N = 4$), sump placement ($N = 2$), clips ($N = 1$), or surgical intervention ($N = 2$).

2.7. Changes in practice patterns over time

As our experience with EIT advanced over the years, our rate of need for surgical intervention significantly decreased over time (Spearman's ρ correlation coefficient $= -0.894$, $p = 0.001$; Fig. 3).With increasing use of EVAC over time, our leak rate has also significantly decreased (Spearman's ρ correlation coefficient $= -0.884$, $p = 0.004$; Fig. 4).

3. Discussion

The rarity of CES has made it difficult to study, and optimal management of CES is still debated [2,4,5]. Some advocate for early surgical intervention owing to the observation that CES (especially TBR subtypes) can be refractory to endoscopic dilation and may experience greater rates of perforation with dilation attempts [1,2]. Other series have described high rates of success with dilations with relatively low complication rates, regardless of CES subtype [3].

Here we report our experience with both endoscopic and surgical management of CES. We have found EIT in conjunction with stenting and EVAC in particular to be transformative in the way we approach

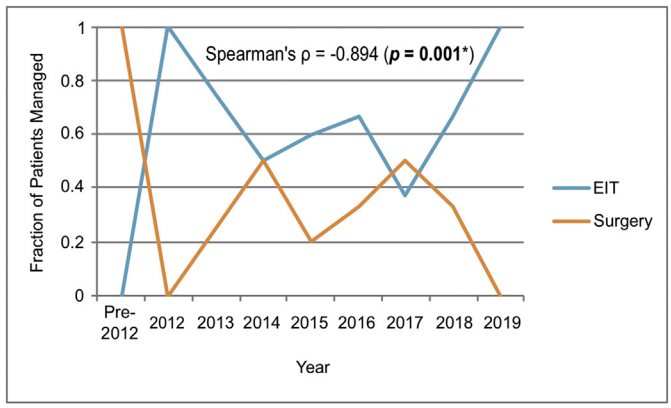

Fig. 3. Rates of CES management with EIT and surgery over time. For each time point, the number of patients managed with each respective modality (EIT or surgery) was normalized to the number of CES patients treated at our institution that year. For patients managed over multiple calendar years, only the year of their initial endoscopic intervention is plotted. Statistical significance is denoted by boldface type and an asterisk.

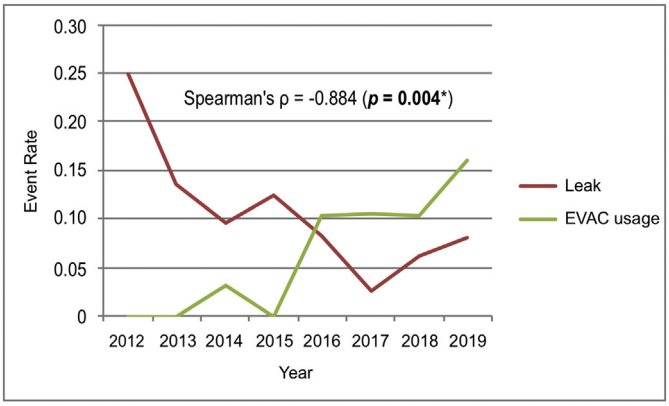

Fig. 4. Esophageal leaks and unique episodes of esophageal vacuum assisted closure (EVAC) usage, normalized to total number of endoscopies performed in at native esophagus congenital esophageal stenosis (CES) sites per year. Spearman's ρ correlation coefficient comparing paired data of esophageal leak rate each year versus EVAC usage rate each year is −0.884 ($p = 0.004$). Statistical significance is denoted by boldface type and an asterisk.

management of CES, allowing us to achieve full clinical feeding success and avoid surgical intervention in a significant number of cases.

Our study is strengthened by our relatively large numbers of CES patients for such a rare diagnosis, though for statistical analysis our numbers are relatively small and limit our ability to identify significant effects of multiple clinical predictors in our multivariable models. Limitations of our study include its retrospective nature at a single referral center with a biased population towards children with a history of EA. In addition, there were 5 patients with history of endoscopic therapy attempts with balloon dilation at outside institutions prior to transfer to our center, skewing our ability to fully judge their response to endoscopic therapy owing differences in institutional practices and endoscopic capabilities; none of these 5 patients underwent EIT at our center. Just more than half of our patients also carried a diagnosis of EA, whereas rates of EA among CES patients have been reported to be 25% in a systematic review of the literature [4]. In our univariate logistic regression model of clinical predictors, history of esophageal atresia was not significantly associated with need for surgery for the CES.

When considered together in multivariable analysis, we could not identify any significant independent effect of each of our examined clinical variables as predictors of needing surgical intervention. This lack of significance is almost surely related to our low number of observed events (i.e. 13 of 36 patients requiring surgery) and we do not have a large enough sample size to fit a multivariate model with all of our suspected associated clinical predictors to determine independent associations between the predictors and need for surgery. We suspect that the univariate effects of EIT, stenting and EVAC are likely interrelated given the high degree of concordance of performing these therapeutic maneuvers within the same patient in our practice.

While the natural tendency is to view surgical intervention as a definitive therapy for CES, patients are often reported to have ongoing symptoms with ongoing requirement for dilations at anastomotic strictures. We found that all patients who underwent surgical treatment of CES at our center required at least one anastomotic balloon dilation and commonly required other adjunct therapies such as steroid injections and stenting. Five of 13 postsurgical patients subsequently underwent EIT to break difficult scar bands at their anastomoses. Others have reported similar outcomes regarding ongoing need for dilation; one study found that 15 of 18 patients who underwent surgical intervention required balloon dilation after their procedure, and 8 of 18 patients continued to be symptomatic [8]. Another study found that 16 of 24 of patients who underwent surgical intervention still had stenotic symptoms [10]. Alternatives to surgery that spare the morbidity associated with thoracotomy are highly desirable in treating CES.

Surgical intervention rates vary considerably by institution, with some centers reporting success rates as high as 96% with endoscopic therapy alone [3] while others report surgical intervention rates as high as 71% [2]. In our cohort, subtype of CES (i.e. FMT vs TBR) was not significantly associated with need for surgery; this is in contrast to others who have reported difficulty treating TBR type strictures with endoscopic therapy alone [4,5,8]. Our numbers of identified TBR ($N = 6$) are likely too low to identify significant effects. We suspect that we underdiagnosed the TBR subtype, especially early on in our care of these patients, since there is disagreement in the literature regarding the ultrasonographic features of the TBR subtype and we were initially specifically looking for hypoechoic areas within the CES. Consensus regarding ultrasonographic features of the TBR subtype would be helpful for future study of the response of TBR to therapy to ensure accurate classification of CES subtypes by EUS.

Of 5 patients with TBR type strictures in our cohort who underwent attempts at endoscopic therapy, 2 had endoscopic attempts that included EIT; only 1 patient of all 5 was managed successfully with endoscopic therapy alone and this patient required 7 repeat sessions of EIT. In contrast, Romeo et al. report successful endoscopic management of 5 of their 6 confirmed cases of TBR with dilation alone [3]. Multicenter analysis of institution-specific practices and referral populations may help clarify the reasons for such dramatically variable experiences with surgical intervention, though it seems our data are consistent with others in that the TBR subtype may be more resistant to an endoscopic approach.

Perforation rates for dilation of CES are highly variable, reported anywhere from 9% [2] to as high as 44.4% [9]. Our perforation rate during balloon dilation without EIT at native CES was 2.5% (3/118 endoscopies) and our perforation rate during endoscopic procedures involving EIT at native CES was 29% (17/58 endoscopies). Unsurprisingly, odds of complication were significantly increased in procedures involving EIT. Owing to high risk of procedures involving EIT, we highly recommend that endoscopist be prepared to intervene for esophageal leak with appropriate endoscopic tools (e.g. EVAC or stent) and have surgical backup available. At our center, EVAC therapy is now our standard first-line practice in cases of esophageal leak or perforation and we have applied EVAC prophylactically in cases deemed at the time of endoscopy to be high risk for development of leak; combining EVAC placement with EIT cases led to significantly decreased leak rate over time.

In summary, we found that EIT allows for successful avoidance of surgery in cases of CES. Further study is needed to identify clinical factors that predict success of endoscopic management of CES.

Appendix A. Supplementary data

Supplementary data to this article can be found online at https://doi.org/10.1016/j.jpedsurg.2020.01.013.

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