Endoscopic Management of Congenital Esophageal Defects and Associated Comorbidities

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KEYWORDS
- Esophageal atresia
- Congenital esophageal stricture
- Dilation
- Surveillance
- Esophagitis

KEY POINTS
- Esophageal atresia patients are at risk for multiple comorbid pathologies, which may be identified and treated endoscopically.
- Dilation is the cornerstone of endoscopic management of anastomotic and congenital esophageal strictures.
- The endoscopic toolbox for management of refractory strictures includes intralesional steroid injection, stenting, and endoscopic incisional therapy.
- Endoscopic approaches for recurrent tracheoesophageal fistula have been described with mixed results.
- Routine endoscopic surveillance for mucosal pathology is critical in patients with esophageal atresia.

INTRODUCTION

Congenital esophageal defects are rare, medically complex problems. The endoscopist plays a critical role in the surveillance and treatment of these complex disorders. This review focuses on esophageal atresia (EA) and congenital esophageal strictures (CESs) and, in particular, the endoscopic management of comorbidities related to these conditions, including anastomotic strictures, tracheoesophageal fistulas (TEFs), esophageal perforations, and esophagitis surveillance. Practical aspects of endoscopic techniques for stricture management are reviewed, including dilation,
intralesional steroid injection (ISI), stenting, and endoscopic incisional therapy. Endoscopic surveillance for mucosal pathology is essential in this population, as patients are at high risk of esophagitis and its late complications such as Barrett’s esophagus.

ESOPHAGEAL ATRESIA

EA with or without TEF is the most common congenital anomaly of the esophagus. The overall incidence of EA/TEF ranges from one in every 2500 to 4500 live births.1 The first successful EA/TEF repair was performed by Dr Cameron Height in 1941.2 Survival rates for patients with EA, with or without TEF, have improved greatly over the past two decades with technical advances in surgery and critical care medicine. The most recent survival rates have ranged from 91% to 97%.3–6 The survival rates for infants born full-term with no associated congenital anomalies have been reported to approach 100%.3,7 Although survival rates are quite high, patients with EA may deal with significant comorbidities postoperatively, both in the immediate postoperative period and later in life (Box 1). In this review, the authors focus on the endoscopic management and surveillance for some of these comorbidities.

Tracheoesophageal Fistula

Most children with EA are also born with an associated TEF. TEFs are typically surgically addressed in infancy. In some cases, congenital fistulas—especially proximal fistulas—may be missed during the initial surgical repair of EA and persist. In addition, recurrence after initial surgical division has been described in up to 10% of cases,8 and acquired fistulas may occur after surgical leaks or perforations. TEFs can be difficult to diagnose and require a high index of suspicion, often requiring combinations of diagnostic modalities (eg, fluoroscopy, upper endoscopy, bronchoscopy) to detect them. For the gastroenterologist, a recently described method using capnography in an intubated patient while performing esophagoscopic insufflation with carbon dioxide can provide an additional method of identifying the presence of a TEF during upper endoscopy.9

There has been interest in endoscopic treatment of recurrent TEFs to potentially spare a patient the potential morbidity of an additional surgery. Various esophagoscopic and bronchoscopic methods for managing TEF have been described in case reports and case series, typically involving mechanical (eg, brushing), chemical (eg, polidocanol, aethoxysklerol), or thermal (eg, diathermy, laser) disruption of the fistula epithelium; application of an occlusive substance into the fistula tract (eg, fibrin adhesive, tissue adhesive); or both.10–13 The bronchoscopic approach is more commonly reported. This approach may take advantage of additional stability offered by the use of rigid instruments and technically more straightforward access into the fistula tract, as typically TEFs have a downward angle of takeoff from the trachea toward the esophagus. Reported pooled success rates of endoscopic TEF closure attempts are 60%, requiring a mean of 2.1 endoscopic procedures, with low morbidity reported.12,13 However, possible reporting bias in the literature may be inflating this statistic and in practice the durable success of endoscopic TEF closure is highly variable.

CONGENITAL ESOPHAGEAL STRicture

CESs are rare congenital anomalies of the esophagus, affecting approximately one in 25,000 to 50,000 live births.14 CES may be present in isolation or in association with other anomalies of the esophagus, particularly EA.15–17 Three subtypes of CES include tracheobronchial remnants (TBR), fibromuscular thickening, or membranous webs and are differentiated by histopathological and
anatomical configuration differences.\textsuperscript{18} Response to endoscopic therapy (in particular, dilations) is felt by some practitioners to depend on the subtype, with TBR potentially more refractory to dilations.\textsuperscript{15,18–22} Endoscopic ultrasound has been reported as an emerging tool for noninvasive subtype differentiation, though the ultrasonographic appearances in these reports are descriptively incongruous with each other (eg, cartilage reported as either hypo- and hyperechoic, depending on the report).\textsuperscript{23–25} Additional study of endoscopic ultrasound with definitively histopathological correlation would be helpful in making ultrasound a more robust diagnostic tool in CES, though unlikely given its rarity as a condition.

**Special Considerations for Management of Congenital Esophageal Stricture**

Management of CES is often first attempted via endoscopic therapy for all subtypes, with surgical intervention reserved for refractory cases.\textsuperscript{17–18,21,22,26,27} Dilation is considered first line, though potentially insufficient to produce durable response. Perforation rates for dilation of CES are high, reported anywhere from 9% to 44.4%.\textsuperscript{15,19} Endoscopic electrocautery incisional therapy (EIT), in which electrocautery is used to incise the congenital stricture in a selective fashion to create controlled weak points in the thickened stricture tissue, in combination with stenting and conventional dilation, has been described to allow for successful endoscopic therapy and avoidance of surgery.\textsuperscript{28} However, EIT is also high risk for perforation and should only be performed when the endoscopist is prepared to manage perforation with either endoscopic vacuum-assisted closure (EVAC) or with experienced surgical backup.\textsuperscript{29–33} Surgical intervention with myotomy, stricture resection, or in some cases esophageal replacement may ultimately be necessary. Patients undergoing surgical intervention must be monitored and treated for symptoms of anastomotic stricture, which may occur in over half of surgically managed CES patients.\textsuperscript{14,20}

**ENDOSCOPIC MANAGEMENT OF COMORBITIES RELATED TO ESOPHAGEAL ATRESIA AND CONGENITAL ESOPHAGEAL STRicture**

**Esophageal Anastomotic Strictures**

Surgical repair of congenital esophageal defects may be complicated by subsequent development of esophageal anastomotic stricture. The following sections describe the clinical presentation and endoscopic methods of treating anastomotic stricture.

**Pathophysiology and incidence**

Surgical creation of an esophageal anastomosis results in a wound, which heals by the natural process of granulation and scar tissue formation. During the tissue remodeling phase of wound healing, fibroblasts promote wound contraction.\textsuperscript{34} Tissue contraction of open wounds is beneficial in order to close the injury; however, wound contraction in the setting of a circular end-to-end anastomosis creates narrowing. Therefore, it is quite common to see a degree of narrowing at the site of the esophageal anastomosis after EA repair (Fig. 1).

The reported incidence of anastomotic stricture after EA repair has varied in case series from as low as 9% to as high as 80%.\textsuperscript{35–40} There are several factors implicated in the pathogenesis of anastomotic strictures, including creation of the esophageal anastomosis under excessive tension, ischemia at the ends of the esophageal pouches, creation of the anastomosis with two suture layers, use of silk suture material, anastomotic leak, esophageal gap length greater than 4 cm (long gap EA), and postoperative gastroesophageal reflux.\textsuperscript{3,36,41,42}
**Presentation**

When a swallowed food bolus becomes too large to pass through the narrowed portion of the esophagus, symptoms of dysphagia will occur. Although a lumen size does not always correlate with symptoms, esophageal lumen size at which dysphagia tends to occur in pediatric patients have been proposed based on expert opinion (Table 1). Typical symptoms of an esophageal stricture include feeding difficulties, coughing and choking during feeds, food impaction, and regurgitation of undigested material. In younger children, apnea may be a presenting symptom as well as feeding refusal. If a patient with EA develops any of these symptoms, they should undergo a contrast fluoroscopy study and/or endoscopy to evaluate for a possible

**Box 1**

**Significant comorbidities associated with surgically repaired esophageal atresia**

- Esophageal stricture
- Esophageal leak or perforation
- Anastomosis dehiscence
- Recurrent tracheoesophageal fistula
- Gastroesophageal reflux disease
- Dysphagia
- Esophageal dysmotility
- Aspiration
- Peptic esophagitis
- Eosinophilic esophagitis
- Barrett’s esophagus
- Esophageal cancer

**Fig. 1.** An esophageal stricture visualized during upper endoscopy.
An esophageal stricture, therefore, is defined as an intrinsic luminal narrowing that leads to the patient becoming clinically symptomatic.\textsuperscript{45} A complex esophageal stricture is defined in adult patients as having one or more of the following characteristics: length $\geq 2$ cm, angulated, irregular surface, diameter $\leq 10$ mm and the presence of diverticulum.\textsuperscript{46}

### Treatment

#### Dilation

The cornerstone of esophageal stricture treatment is dilation. The goal of esophageal dilation is to increase the luminal diameter of the esophagus while also improving dysphagia symptoms. This is achieved through circumferential stretching and splitting of the scar tissue within the stricture.\textsuperscript{47,48} Even though there are many dilation techniques and a variety of available equipment, they fall into two main categories: mechanical (bougie or push-type) dilators or balloon-based dilators.

**Mechanical (bougie) dilation**

The basic technique of mechanical dilation involves the passage of a bougie dilator across the stricture (Fig. 2). This results in both longitudinal shearing force and radial force on the strictured area. The goal of mechanical dilation is to pass serial bougie dilators of incremental size across the stricture site. Although fluoroscopy is frequently recommended to confirm correct positioning as the bougie dilator is passed across the stricture, it is not mandatory in short strictures. It is generally recommended to use fluoroscopy in strictures longer than 1 cm and/or strictures that are angulated.

There are several different types of bougie-based dilators, the most common of which are guidewire-based. Presuming the wire position is checked frequently (using fluoroscopy or a fixed external landmark), this helps ensure the dilator will pass

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**Table 1**

<table>
<thead>
<tr>
<th>Age</th>
<th>Esophageal Lumen Diameter (mm)</th>
</tr>
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<tbody>
<tr>
<td>Less than 9 mo</td>
<td>8</td>
</tr>
<tr>
<td>9 to 23 mo</td>
<td>10</td>
</tr>
<tr>
<td>24 mo to 5 y</td>
<td>12</td>
</tr>
<tr>
<td>Greater than 6 y</td>
<td>14</td>
</tr>
</tbody>
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Fig. 2. A mechanical (bougie) dilator exerts a longitudinal and radial force, dilating the stricture proximal to distal, whereas a balloon dilator exerts a radial force delivered simultaneously across the entire stricture. (Adapted from Adler DG, Siddiqui AA. Endoscopic management of esophageal strictures. Gastrointest Endosc. 2017;86(1):35-43.)
correctly through the stricture. These dilators are tapered, cylindrical solid tubes made of polyvinyl chloride with a central channel to accommodate a guidewire.\textsuperscript{49} These dilating tubes have varying lengths of tapering at the tip and also have radiopaque markers to permit fluoroscopic guidance (eg, Savary-Gilliard dilators, American Dilators, and SafeGuide). There are also non-guidewire mechanical dilators that are tungsten weighted to allow for gravity assistance when the patient is in a seated position. The two commonly used non-guidewire bougie dilators are Hurst and Maloney dilators.\textsuperscript{49}

Another type of mechanical dilator is Tucker dilators, which are small, tapered silicone bougies with loops on each end. A string is attached to the loops to allow for the dilator to be pulled antegrade or retrograde across strictures. These dilators, therefore, require the patient to have a gastrostomy. Tucker dilators can remain inside the patient for periodic serial dilations.

Mechanical bougie dilation is a tactile technique.\textsuperscript{37,45,50} As the bougie is advanced across the stricture site, a degree of resistance should be appreciated by the operator. The object is to feel, and then overcome, the resistance across the strictured area. Once moderate resistance is encountered with the bougie dilator, it is generally recommended passing no greater than three consecutive dilators in increments of 1 mm in a single session for a total of 3 mm. This approach, known as the “rule of 3,” is a well-established approach for mechanical dilation.\textsuperscript{51} Strict adherence to the rule is not always necessary and there may be occasions that one may dilate larger than 3 mm in a single session. Two studies, one adult and one pediatric, have shown that nonadherence to the “rule of 3” was not associated with increased adverse events.\textsuperscript{52–54}

**Balloon dilation**

Balloons deliver equal radial force across the entire length of the stricture (see Fig. 2). They are designed to pass through the endoscope channel with or without a guidewire. Most commonly, they are 5 cm in length and 6 to 20 mm in diameter (some are multidiameter with increasing pressures). Through-the-scope (TTS) dilation allows the endoscopist to directly visualize the stricture during and immediately after the dilation. However, TTS balloon dilation requires the use of an adult gastroscope with a minimum working channel diameter of 2.8 mm, which is difficult to use in younger infants under 10 kg.

In smaller patients, the balloon can be passed over a guidewire under fluoroscopic guidance. This technique is performed by passing a 0.035-mm guidewire across the stricture through the endoscope working channel, followed by a wire exchange under fluoroscopy, leaving the wire in place as the scope is removed. The balloon is then passed over the wire and positioned across the stricture under fluoroscopic guidance.

When possible, TTS balloon dilation should be performed as it can provide direct visualization of the tissue during and immediately following the dilation and permit monitoring of the degree of developing mucosal disruption. It also allows the endoscopist to minimize fluoroscopy time as balloon placement can be done by endoscopic view. However, a balloon should not be passed blindly through a stricture if the scope cannot traverse the stricture beforehand and instead a wire should be used to prevent unintentional perforation from the tip of the dilator.

Dilating balloons expand by the injection of liquid (eg, water, radiopaque contrast) under pressure using a handheld inflation device. A manometer on the device will measure the fluid pressure in the balloon to allow for accurate radial expansion force.\textsuperscript{49} Balloon dilators are either designed to inflate to a single target diameter or
to allow for sequential inflation to multiple sizes (typically three incremental diameters per balloon, depending on the pressure delivered into the balloon).

The basic approach to balloon dilation is to first estimate the size of the stricture. This can be done by performing an intraoperative contrast esophagogram immediately before dilation to estimate stricture diameter, length, and possible underlying contraindicative pathology (e.g., TEF or preexisting esophageal leak). To guide decision-making in regard to balloon size, further assessment of stricture size can be estimated by using a visual reference such as the biopsy forceps of known dimensions as a measurement tool and/or by estimating size based on the outer diameter of the endoscope and ability of the endoscope to pass the stricture.

Once the size is estimated, the “rule of 3” can similarly be applied to balloon dilators by choosing a balloon that will increase in size by increments of 1 mm in a single session for a total of 3 mm above the originally estimated stricture size. A recent pediatric study found that dilating up to 5 mm above the stricture diameter did not increase the risk of perforation compared with dilating only to 3 mm. This study points out that these rules are meant as a guide rather than a replacement for clinical judgment based on inspection of the stricture post-dilation. It is important to carefully inspect the tissue in between dilations, and in situations, where the endoscopist notes an unsatisfactory response to standard dilation increments with no evidence of perforation, there is now precedent to support dilating further if indicated.

Before dilation, the balloon is advanced across the stricture either with endoscopic and/or fluoroscopic guidance. Ideally, the balloon should be positioned so that the middle of the balloon is centered across the stricture. Balloons are available with or without a wire. Our recommended approach is to have a wire passed across the stricture and typically into the stomach. The goal of the wire is to make certain that the tip of the balloon remains within the lumen of the esophagus, as the tip of the balloon is sharp, and it is possible for the tip to dissect through the esophageal wall if blindly passed without a guidewire.

Once the balloon is properly positioned, the balloon is inflated to the desired size. The optimal inflation time has not been established. Balloon inflation times of 30 to 60 seconds are generally accepted. A randomized study of patients with strictures who underwent dilation using different balloon inflation times showed no significant difference in dilation effectiveness based on inflation time. Therefore, it seems that the act of inflation, which tears the scar tissue, is more important than the duration of the balloon is inflated.

The use of fluoroscopy during balloon dilation is helpful. In the setting of a complex stricture, fluoroscopy is useful in advancing the wire and balloon safely across the stricture. In addition, inflating the balloon with contrast will allow the endoscopist to see if the stricture is being effectively dilated. It is useful to see the appearance of the stricture forming a waist around the balloon and the subsequent obliteration of a waist as the balloon is further inflated (Fig. 3). It is a practice of these authors to use fluoroscopy with our esophageal balloon-based stricture dilations. In addition, there is an added benefit of using fluoroscopy to conduct a post-dilation contrast study to evaluate for a post-dilation esophageal leak or perforation.

Comparative studies of mechanical and balloon dilation

There is little data comparing bougie (mechanical dilators) versus balloon dilators in both the pediatric and adult literature. In adult studies, there has been no significant difference in safety or efficacy between wire-guided bougie dilation and balloon dilation. Pediatric studies show mixed results, although they are limited by small sample sizes. There have been two pediatric studies that favored balloon dilation over...
bougie in both safety and efficacy, whereas a third study only evaluated safety and found no difference between both groups.\textsuperscript{59–61} Although further investigation is needed, the authors recommend based on the existing literature that the provider should use the technique with which they are most comfortable and experienced when performing a dilation. Wire-guided bougie dilation is generally recommend over non-wire-guided push bougie dilators in complex strictures due to higher rates of esophageal perforation without wire guidance.\textsuperscript{46}

**Refractory Strictures and Adjunct Treatments**

Refractory strictures are defined in adults as a failure to remediate the stricture successfully up to diameter of 14 mm over five sessions at 2-week intervals as well as maintaining a satisfactory diameter for 4 weeks once the desired diameter has been achieved.\textsuperscript{62} A modification for pediatrics has been suggested by the current authors, whereby a refractory stricture is defined as the inability to remediate the esophageal lumen with five dilatations performed within 5 months to the desired size for age.\textsuperscript{45} Alternatively, a North American and European Societies of Pediatric Gastroenterology, Hepatology and Nutrition EA guideline report based on expert opinion defined three or more clinically relevant stricture relapses as a recurrent stricture.\textsuperscript{50} There is a need for consensus around the definitions of refractory stricture to help standardize study outcomes and accurately evaluate the efficacy of different therapies.\textsuperscript{63} Regardless of the definition, once a stricture becomes refractory to esophageal dilation, there are several treatment therapies

Fig. 3. An esophageal stricture visualized with fluoroscopy.
available as adjuncts to dilation therapy that should be considered before surgical resection.

**Intralesional steroid injection**

ISIs are typically used in conjunction with dilation to facilitate larger post-dilation esophageal stricture diameter. The proposed mechanism of ISI in the treatment of esophageal strictures is to locally inhibit the inflammatory response that promotes collagen formation and scarring within a stricture. Triamcinolone acetonide 40 and 10 mg/mL is commonly used. The authors prefer 10 mg/mL concentration as the 40 mg/mL is viscous and typically needs to be diluted before injection; dilution is not necessary with the 10 mg/mL concentration. ISI is administered via a sclerotherapy needle in 0.1 to 0.2 mL aliquots. Four quadrant injections are common; however, if the scar tissue is uneven a preponderance of steroid can be injected into targeted scar tissue areas. The dose of triamcinolone acetonide used is 1 to 2 mg/kg per dose, up to a maximum dose of 80 mg in adults. ISI may be injected before or after dilation therapy.

The efficacy of ISI in peptic strictures was demonstrated in a randomized double-blind placebo controlled trial in which patients received four quadrant injections of 0.5 mL of triamcinolone acetate (40 mg per mL) for total of 80 mg or a sham. Two of 15 (13%, 95% CI 4%–38%) patients in the steroid group and 9 of 15 (60%, 95% CI 36%–80%) in the sham group required repeat dilation ($P = 0.021$). There have been multiple studies that have shown the benefit of ISI in reducing recurrent stricture formation in other types of strictures. However, most reports are small uncontrolled studies evaluating strictures of diverse etiology. In a multicenter double-blind placebo control trial involving 60 patients with benign esophagogastric anastomotic strictures, the authors reported no statistically significant decrease in frequency of repeat dilations with a median number of two dilations (range, 1–7) performed in the corticosteroid group compared with three dilations (range, 1–9) in the control group ($P = 0.36$). A pediatric retrospective study evaluating 158 patients with anastomotic strictures showed that ISI combined with dilation was well tolerated with no increased incidence of perforation, and statistically significant improvement in stricture diameter was observed when compared with dilation alone. In addition, this study showed that the effectiveness of ISI injections appeared to peak at three injections, with no significant gains in diameter beyond three injection sessions.

Potential complications of ISI include adrenal suppression. Therefore, some authors suggest surveillance for adrenal suppression. However, this is currently not standard of care. In addition, there have been reports of increased Candida esophagitis. Last, there has been one report of intralesional steroids contributing to the spontaneous rupture of a right aortic arch presumably secondary to the steroids weakening the arterial wall.

**Mitomycin C**

Mitomycin C is an antineoplastic agent that disrupts base pairing of DNA molecules, inhibits fibroblast proliferation, and reduces fibroblastic collagen synthesis by inhibiting DNA-dependent RNA synthesis. It also induces apoptosis at higher doses by suppressing cellular proliferation during the late G1 and S phases. It has been proposed as an adjunct treatment to manage esophageal strictures. Mitomycin C has been mainly placed topically in the literature; however, there are also reports of injection of mitomycin C. There have been numerous described methods of topically placing mitomycin C, such as soaking pledgets or cotton swabs and placing them topically on the stricture area, dripping mitomycin C via an injection needle onto the affected area,
or using a spray catheter.\textsuperscript{73–76} The concentration of mitomycin C used in these studies is also variable, ranging from 0.004 to 1 mg/mL.\textsuperscript{77}

The efficacy of mitomycin C has been a controversial topic in patients with EA. A recent study reported a 71\% success rate in EA patients with the majority of them being type C, with success defined \textit{a priori} as any reduction in the number of dilations over the same period from before to after the application.\textsuperscript{78} This is in stark contrast to another study on EA patients that showed a 27\% success rate with no significant difference in dilations compared with historical controls.\textsuperscript{79} The lack of standardized definitions of refractory strictures and treatment success may contribute to different outcomes. In addition, the timing of treatment early or late in the course of dilations may also be a factor. Mitomycin C has been shown to be effective in some prospective studies looking at strictures secondary to caustic ingestion.\textsuperscript{80–82} However, a recent meta-analysis looking at the efficacy of mitomycin C in caustic strictures did not show a statistical difference in the overall number of dilations in treatment and nontreatment groups.\textsuperscript{83}

There is a hypothetical risk of secondary malignancy with mitomycin C, so this must be taken into account and should be discussed with the patient and caregivers before use.\textsuperscript{84} There have been reports of de novo gastric metaplasia around the areas of the anastomosis in two of the six cases that received topical mitomycin C.\textsuperscript{84} Therefore, long-term follow-up with esophageal biopsies at the site of mitomycin C application should be recommended.

\textbf{Esophageal stents}

The rationale for stenting strictures, in theory, seems sound. By applying dilation forces to the esophagus for prolonged periods of time, stenting may reduce the risk of recurrent stricture formation and thus may be an alternative treatment option to serial esophageal stricture dilations. The first externally removable stents were self-expandable plastic stents. These have largely been replaced by fully covered self-expandable metal stents (FCSEMSs). These stents are composed of a memory shape metal nitinol (an alloy of nickel and titanium) and are available in various diameters and lengths. Esophageal FCSEMSs are designed for adult patients and thus are too large for the majority of pediatric patients. For most children, either biliary or airway FCSEMSs can be used in the pediatric esophagus.

Stent deployment requires the use of radiography to ensure proper placement. Many experts suggest that vascular imaging should be done before stenting to look for an aberrant right subclavian artery as this may increase risk of complication.\textsuperscript{37,85} Each stent has their own unique deployment mechanism that separates the stent from the housing sheath. As FCSEMS have a degree of foreshortening during deployment, the placement should be done under fluoroscopy in order to reposition the stent in real time, whereas it is deploying.

The utility of stent treatment for esophageal strictures is unclear. The success of esophageal stenting in the pediatric literature is also variable with rates ranging from 0 to 86\% success.\textsuperscript{86–89} A pooled analysis of seven pediatric studies with a total of 69 patients with esophageal strictures of multiple etiologies reported a pooled success rate of 52\%.\textsuperscript{90} A single-center pediatric study looking at 49 esophageal strictures secondary to EA reported clinical success in 41\% of patients.\textsuperscript{91} Patients whose procedures were successful underwent a median of 0.5 dilations (Interquartile range [IQR] 0, 1) during follow-up period, which was a median duration of 5 years (IQR 2–6). This study found the greatest predictor of stent success was the degree of re-stricturing seen at the time of follow-up endoscopy performed at a median of 2 weeks after stent removal, with shrinkage of the stricture \(\geq 4\) mm from the starting stent diameter.
predicting endoscopic treatment failure. Although the utility of stenting to treat strictures is still debatable, it may serve as a bridge to surgery.

Esophageal stenting has been associated with numerous adverse events, including life-threatening events such as bleeding, perforation, and erosion into the vascular system or airway. There has also been reported mortality secondary to esophageal stenting. Additional adverse events include stent migration, tracheal compression, gastroesophageal reflux, aspiration pneumonia, and new esophageal stricture development due to the edges of the stent or ischemia secondary to overly aggressive stent diameter selection.

**Endoscopic electrocautery incisional therapy**

EIT is a technique based on understanding that not all strictures are symmetrical. Many strictures, particularly anastomotic strictures, are asymmetric with areas of varying degrees of scar tissue. Thickened scar tissue may have the appearance of bands or shelves of tissue. Dilation alone in an asymmetric stricture tends to tear the stricture at areas of thinner scar tissue, and thus may lead to a less effective dilation. EIT is a technique that involves applying electrocautery via a needle knife to make small incisions into the scar tissue at its thickest areas to create preferential weak points (Fig. 4). Once the incisions are made, a balloon dilation can then be performed to preferentially dilate the areas that were weakened by the incisions. This technique tends to be better for strictures less than 1 cm in length.

In a pediatric study of 58 patients, EIT was successful in remediating the stricture in 76% of patients with 2 year follow-up. In subgroup analysis of patients who met criteria for a refractory stricture, EIT was successful in 61% (N = 36) of patients. This same study reported a perforation rate of 2.3%. As EIT has a higher adverse event rate than balloon dilation, it should be performed by a highly skilled endoscopist who has access to fluoroscopy and surgical backup at the time of the procedure to recognize an adverse event.

**Esophageal Perforation or Leak**

Esophageal perforation or leak is a potentially life-threatening problem if not quickly diagnosed and treated appropriately (Fig. 5). The rate of developing an esophageal anastomotic leak after surgical EA repair has been reported to range from 11% to 16%. In addition, esophageal perforation may occur after esophageal dilation or another endoscopic therapeutic intervention at a stricture. In a systematic review with pooled analysis of 5 pediatric studies comprising 139 patients and 401 dilations, the perforation rate was 1.8%. A large single-center pediatric study of 284 patients with 1384 dilations reported an esophageal perforation rate of 1.6% for balloon dilation. Of note, steroid injection in combination with dilation had no statistically significant increase in perforation compared with dilation alone. Traditional management of esophageal perforations or leaks in children includes making the patient nil per os, intravenous broad-spectrum antibiotics, and esophageal decompression with the placement of a nasoesophageal tube to low wall suction. External wound drainage with a chest tube is considered in the setting of a large fluid collection in the chest.

Esophageal stenting has been shown to be effective in adult patients with esophageal perforations and has become a first-line treatment, with a reported clinical success rate of 85% with a mean stent duration time of 6 to 8 weeks. In pediatrics, the use of esophageal stents to treat esophageal leaks has a reported success rate from 64% to 100%. The median number of days that stents were left in situ in the pediatric studies ranged from 8 to 36 days with the longer duration of stent time being associated with higher success. In our experience, esophageal stents
**Fig. 4.** Endoscopic incisional therapy involves incising the thickened scar bands of the stricture using electrocautery. After incisions are created, a balloon dilation is performed.

**Fig. 5.** A fluoroscopic image confirming extravasation of radiopaque contrast outside the lumen of the esophagus, consistent with esophageal leak.
also have drawbacks, especially in children with a surgically repaired esophagus, as in the EA population. Esophageal stents may lead to local pressure necrosis of the esophagus, which may worsen the existing esophageal perforation and may lead to erosion into surrounding structures such as the airway and major blood vessels. Last, stenting does not facilitate drainage of the fluid collection around the esophagus and can in fact trap infection in the chest; thus, stents may facilitate abscess formation unless external drainage with a chest tube is initiated at the time of stent placement.29

EVAC is an adaptation of traditional vacuum-assisted closure devices (Fig. 6). It is based on the principles of negative pressure wound therapy, which stimulates wound healing by removal of fluid from the perforation site, source control for infection, reduction of tissue edema, and promotion of blood flow to the area stimulating granulation tissue formation.101 In a pediatric study of 17 patients with EA who underwent EVAC therapy for esophageal perforation secondary to either postsurgical anastomotic leak or endoscopic therapy, the success rate of EVAC to seal all esophageal perforations was 88%.29 The success rate was similar in both subgroups (surgical anastomotic leaks at 88% [7/8] and endoscopic therapy leaks at 89% [8/9]), with a median duration of EVAC treatment of 8 days. This same study compared EVAC with a cohort of esophageal perforations treated with esophageal stents (n = 24) and found a statistically significant difference in favor of EVAC in sealing surgical anastomotic perforations (P = 0.032); however, there was no statistical difference in sealing endoscopic therapy perforations (P = 0.360).

Endoscopic Surveillance for Mucosal Pathology

Long-term, patients with EA are at increased risk of esophagitis. Historically, most esophagitis in this population has been attributed to acid reflux; in recent years, increased attention has been paid to higher rates of allergic eosinophilic esophagitis in EA patients as well.102,103 Dysmotility with poor esophageal clearance is also a likely contributor to long-term risk of esophageal mucosal pathology.104,105 In the setting of higher rates of chronic esophageal inflammation and injury, individuals with EA have been noted to be at significantly higher risk of the precancerous condition Barrett’s esophagus compared with the general population.50,104,106 Chronic inflammation may also contribute to dysphagia and stricturing.

Chronic acid suppressive therapy is commonly prescribed and has been linked to lower odds of abnormal esophageal biopsy,107 though optimal duration of treatment is uncertain and esophageal inflammation remains common even in those on acid suppression.107 Thus, consensus guidelines advocate for endoscopic surveillance with multiple levels of esophageal biopsies, even in asymptomatic patients on acid suppression, to proactively monitor for esophagitis before its late complications such as Barrett’s esophagus develop.50 However, the optimal interval for surveillance has not been defined.50 Current recommendations advocate for at least three
endoscopies in childhood: one after stopping Proton Pump Inhibitor (PPI) therapy, one before age 10 years, and one on transition to adulthood. In the authors’ experience, this approach to surveillance may lead to delayed diagnosis of important pathology as the rate of endoscopic pathology in this patient population is high. Indeed, the authors have identified erosive esophagitis in 6% of patients and high rates of significant histologic esophagitis (>15 eosinophils/high-powered field) in over 25% of endoscopies as well as biopsy-confirmed Barrett’s esophagus in patients as young as 5 years old, despite nearly 90% of endoscopies being performed on chronic acid suppressive therapy.

Thus, the authors’ current practice is to perform surveillance endoscopy every 1 to 3 years throughout childhood, with more frequent follow-up for patients with significant or refractory esophagitis, Barrett’s esophagus, or other pathology; our initial surveillance endoscopy is performed around approximately 1 year after the surgical creation of the esophageal anastomosis.

SUMMARY

Dilation remains the first-line treatment for many types of esophageal strictures including anastomotic strictures that can develop after repair of EA. Adjunctive therapies such as ISI, mitomycin C application, stenting, and incisional therapy may be useful in treating strictures that do not adequately respond to dilation alone. CESs require special consideration and potentially referral to a center of expertise given the high risk of complications with all forms of endoscopic therapy. Dilation alone may be less effective in treating some congenital strictures and may be associated with relatively high rates of perforation. Esophageal perforations can often be managed via endoscopic means with stenting or EVAC devices. Rates of mucosal pathology in children with congenital esophageal defects are high, and routine endoscopic surveillance is warranted to prevent long-term complications of uncontrolled inflammation.

CLINICS CARE POINTS

- When performing dilation, the endoscopist should perform visual inspection to determine the degree of mucosal disruption. Some degree of mucosal disruption is expected and even desired in treating most strictures, though excessive or asymmetric mucosal disruption increases the risk of perforation.
- The use of real-time fluoroscopy during dilation allows the endoscopist to assess for obliteration of a radiographic “waist,” which can aid in assessing anastomotic stricture response to dilation.
- In contrast, obliteration of the radiographic waist should not be the sole endpoint goal of a dilation session of a CES, which is high risk for perforation. CESs should be visually inspected frequently during the endoscopy session to assess for any evolving asymmetry in mucosal disruption that may indicate a region at risk for perforation should more aggressive dilation diameters be attempted.
- Asymmetric strictures often benefit from endoscopic incisional therapy to create select weakened points in the thicker scar bands to then more evenly spread the dilation force around the circumference of the stricture. Incisional therapy should only be attempted by a skilled endoscopist with appropriate backup plan in case of perforation (including availability of surgical backup, an intensive care unit bed, and an ability to place an endoscopic vacuum-assisted closure device or stent).
- Intraläsional steroid injection may be performed in four quadrants around a stricture or can be focused in areas of apparent thickened scar tissue.
Endoscopic treatment of esophageal perforation should be accompanied by medical management to address microbial contamination of the extraluminal space and should include broad spectrum intravenous antibiotics. In cases of persistent fevers or clinical deterioration, the addition of antifungal therapy should be considered.

Surveillance endoscopy should be periodically performed in patients with history of EA, even if asymptomatic, as symptoms do not predict the presence of abnormal findings.

CONFLICTS OF INTEREST

The authors have no conflicts of interest or funding sources to disclose.

REFERENCES


