Esophageal Strictures and Stenoses

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10.1 Introduction

The diagnosis and management of esophageal obstructions, namely strictures and stenoses, can be quite intimidating and challenging for the surgeon. We discuss in this chapter the many different types of esophageal obstructions and outline the major principles involved in their management. A careful understanding and thoughtful approach to the their treatment remains paramount to achieve optimal outcomes. Depending on the nature and etiology of the narrowing, the therapeutic options can range from pharmacotherapy alone to complex surgery, including esophagectomy with conduit reconstruction. More recently, endoscopic interventions have made a dramatic impact in many children previously thought to have a problem amenable only to major reconstructive esophageal surgery.

10.2 Epidemiology

Esophageal obstructions are relatively uncommon. The exact incidence within the pediatric population is not known. According to one estimate, there are >5,000 major caustic ingestions per year in the USA, of which ≈20–25% will result in an esophageal stricture [1]. Most caustic esophageal strictures occur in the setting of accidental ingestion, so most of these affected children are <5 years of age, with a peak incidence at 2 years of age [2]. Legislative efforts, including the United States Federal Hazardous Substances Act and the Poison Prevention Packaging Act of 1970, require that corrosive substances be packaged in such a way that it would be difficult for children <5 years to open them, yet not too difficult for adults to open them. Fortunately, such preventive measures have been effective in reducing the incidence of these tragic injuries. Nevertheless, in some "developing" countries, there continues to be an alarmingly high incidence of caustic esophageal injuries because of a lack of childproof containers.

Historically, the number of strictures secondary to gastroesophageal reflux disease (GERD), a common disorder in infants and children, has been estimated to occur in ≤15% of affected patients. However, more recent data suggests that this prevalence has probably decreased markedly with the widespread use of acid blockers, particularly proton pump inhibitors (PPIs) [3, 4]. Other causes of esophageal strictures and stenoses occur at a much lower frequency. For example, congenital esophageal stenosis is estimated to occur in 1 in 25,000–50,000 live births.

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10.3 Etiology and Pathogenesis

The more common disease processes (acquired and congenital) associated with esophageal strictures and stenoses are discussed below.

Caustic ingestion: Historically, caustic injuries have been the most common cause of esophageal strictures in children [5]. Many corrosive products contain sodium hydroxide or potassium hydroxide, and are therefore strong bases (pH >12). The usual alkaline agents include lye, caustic sodas, dishwashing detergents, disinfectants, degreasers, and drain cleaners.

Bases are particularly dangerous for young unsuspecting children because these substances are often tasteless and can cause extensive liquefactive necrosis, resulting in full-thickness esophageal damage and fibrotic strictures if ingested accidentally in small amounts. The consequences of these injuries, such as mediastinitis, tracheoesophageal fistula, and aortoesophageal fistula, are potentially lethal.

Acids (pH <1.5) can also be dangerous if ingested but are usually foul-tasting and cause less esophageal damage. The more common acidic agents are toilet-bowl cleaners, paint thinners, batteries, and metal cleaners. Most acids induce injury by coagulation necrosis, resulting in an eschar formation that is more superficial compared with injuries from bases. However, button batteries can cause significant esophageal damage, including tracheoesophageal fistula, from electrical discharge and direct pressure necrosis [6].

Gastroesophageal reflux (GER): Peptic esophageal strictures are the result of uncontrolled, chronic exposure of refluxed gastric acid onto the adjacent esophageal mucosa. Strictures of variable severity can develop over time secondary to recurrent inflammation with subsequent submucosal fibrosis. Most (but not all) of these strictures are located in the distal esophagus. Barrett's esophagitis, a potential precursor to esophageal adenocarcinoma, has been identified within some of these strictures in older children [7].

Eosinophilic esophagitis (EoE) has been an increasingly recognized cause of pediatric esophageal strictures over the last 15 years. In the past, many of these patients were thought to have GERD but showed a poor response to acid blockade medications. Pathology specimens of the esophagus in EoE are characterized by dense eosinophilic infiltrates with squamous hyperplasia.

Anastomotic strictures: The reported prevalence of strictures in neonates after repair of esophageal atresia is ≤37% depending on how these strictures are defined by authors [8, 9]. Known risk factors for stricture formation include increased tension, ischemia, acid reflux, and the use of silk sutures [10]. Additionally, stricture formation at the anastomosis is common after an anastomotic leak treated by non-surgical means.

Esophageal stenosis: Some infants are diagnosed with an intrinsic congenital stenosis of the esophagus. There are three histopathological subtypes of congenital stenosis: membranous webs, fibromuscular dysplasia, and ectopic tracheobronchial remnants [11]. The stenosis is usually located in the distal third of the esophagus, and is associated with esophageal atresia with tracheoesophageal fistula in about one-third of cases. Interestingly, the diagnosis of congenital stenosis is not commonly made during the neonatal period (Fig. 10.1). Instead, affected infants typically present with persistent vomiting, dysphagia, and/or failure to thrive, particularly with the introduction of solid foods between 4–10 months of age.

Mediastinal masses (e.g., esophageal duplications, bronchial duplications, leiomyomas) are rare but can cause local compression, thereby mimicking an esophageal stricture or stenosis.

Aberrant vascular anatomy: A vascular ring or an aberrant right subclavian artery can be a cause of proximal esophageal stenosis secondary to extrinsic compression. In the latter entity, children can present with dysphagia lusoria, described as such because of the unusual retro-esophageal course of the subcla-

vian artery. Contrast esophagography will often reveal a notching defect of the upper esophagus above the level of the aortic arch.

Epidermolysis bullosa (EB) represents a spectrum of rare, inherited blistering disorders secondary to a genetic defect in type-VII collagen. EB primarily affects the mucosa of the skin, pharynx, and esophagus. Although infants with EB rarely develop esophageal symptoms in early childhood, most affected individuals (particularly those with recessive dystrophic EB) will develop symptomatic esophageal strictures by 25 years of age. Many EB strictures are located in the proximal cervical esophagus [12].

Schatski ring is a very rare entity in children. Although most cases are seen in male adolescents, it is unclear whether this is the same entity as the membranous web subtype of congenital esophageal stenosis. The hallmark finding of a Schatski ring is formation of a thin, circumferential fold of mucosa that protrudes into the lumen at the gastroesophageal junction. Many cases have been identified in association with GERD or EoE.

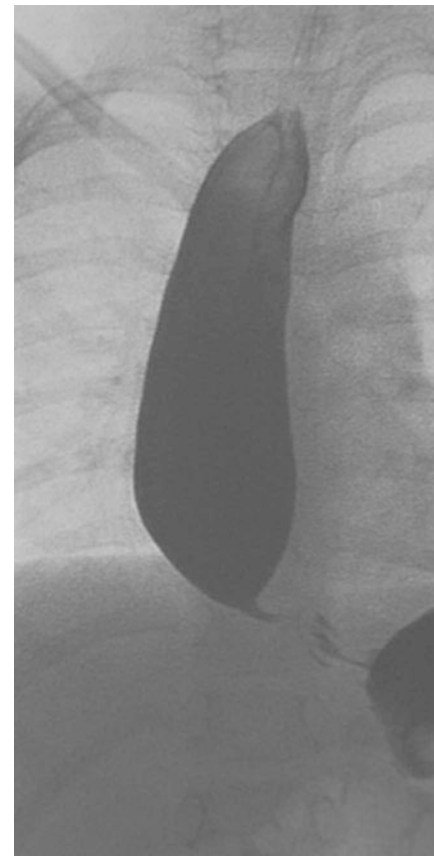

Fig. 10.1 Anterior contrast esophagram in a 6-month-old female with congenital esophageal stenosis showing marked dilation of the proximal esophagus with narrowing of the distal esophagus proximal to the gastroesophageal junction ("bird's beak" configuration)

10.4 Clinical Features

Although the presentation of an esophageal obstruction can vary depending on the etiology, most symptoms are specific to the upper alimentary tract. Most children will have some component of feeding intolerance, vomiting, dysphagia, or failure to thrive. Older children with GERD may experience heartburn. In young children with peptic esophageal strictures, it is important to realize that these symptoms can often be more insidious in nature, becoming more apparent over a 3–4 year period as they advance from a soft diet to more solid foods [3]. Children with EoE or Schatski ring often present with upper digestive tract symptoms complicated by impaction of a food bolus in up to 50% of cases [13, 14].

Extra-gastrointestinal tract symptoms may also be apparent in children with esophageal strictures and stenoses. For example, patients with a peptic esophageal stricture can be anemic secondary to chronic mucosal bleeding. Children with EoE often present with other associated manifestations of atopic diathesis (i.e., food allergy, asthma, eczema, chronic rhinitis, environmental allergies). Finally, because of the proximity of the esophagus to the airways, it is not uncommon for some patients with esophageal strictures or stenoses (particularly those secondary to GERD) to have concomitant respiratory symptoms because of chronic aspiration. Caustic ingestions may also cause acute and/or chronic injury to the larynx and trachea, leading to hoarseness, stridor, and dyspnea on presentation. All patients with suspected respiratory involvement should un-

dergo larynoscopy to fully assess the upper airway.

Children evaluated for a caustic ingestion in the acute setting should undergo immediate chest radiography if there is any concern for a possible esophageal perforation. Potential symptoms or signs of perforation include chest pain, fever, and tachycardia. Any radiographic evidence of a pleural effusion, pneumomediastinum, pneumothorax, or pneumoperitoneum mandates urgent esophagography with water-soluble contrast to further rule out perforation prior to undergoing esophagoscopy. Failure to recognize an occult esophageal perforation can be disastrous because insufflation during any esophagoscopy procedure may lead to worsening of the esophageal injury with the potential for mediastinitis, sepsis, cardiopulmonary arrest, and death.

10.5 Diagnosis

10.5.1 Esophagography

A contrast esophagram (often referred to as a swallow study) remains the "gold standard" for the diagnosis of an esophageal obstruction. During this test, water-soluble contrast (e.g., diatrizoic acid) or barium is delivered into the upper esophagus and followed distally past the gastroesophageal junction as it empties into the stomach. Barium gives superior mucosal detail but should not be used in a suspected perforation. In most cases, a carefully carried out study (including frontal and lateral projections) enables delineation of the precise location and length of the obstruction. Megaesophagus is highly suggestive of a chronic distal stricture. Evidence of dysmotility can often be ascertained from serial dynamic images. Based on the characteristics of the narrowing revealed by carefully carried out esophagography, a presumptive etiology for the stricture can often be made in conjunction with the clinical history.

10.5.2 Esophagoscopy

In addition to a contrast esophagography, complete endoscopic evaluation of the esophagus down to the gastroesophageal junction has an important and complementary diagnostic role in most types of esophageal obstructions. Although some surgeons prefer to use rigid esophagoscopes (which have a large operating channel), our preference is to use a flexible pediatric or neonatal endoscope for all but the most proximal cervical esophageal problems. We have found that modern, fiberoptic flexible endoscopes often give excellent visualization of the distal esophagus, induce less trauma to the oropharynx, and allow for evaluation of the entire stomach. The role of esophagoscopy is particularly vital in assessing obstructions in the settings detailed below.

10.5.2.1 Esophagoscopy in Caustic Ingestion

In the acute phase after ingestion, esophagoscopy serves as a useful tool for predicting the likelihood of a subsequent stricture. The absence of oropharyngeal burns on clinical examination or larynoscopy should never exclude the need for esophagoscopy in the presence of a good medical history [2]. The only major contraindication to endoscopic evaluation is esophageal perforation. Ideally, esophagoscopy should be done 12–48 h after ingestion. We discourage esophagoscopy done <12 h after ingestion because this may not allow sufficient time for full demarcation of the injury. We also do not advise delaying esophagoscopy for >72 h after ingestion because of the potential for encountering severe esophageal edema and early stricture formation, thereby increasing the risk of iatrogenic perforation.

During endoscopy, the entire esophagus should be assessed for the degree of injury according to a three-point grading scale modified from the more widely known classification system employed for thermal injuries to the skin [15]. First-degree injuries as demon-

strated by mucosal hyperemia and edema are superficial. Such findings predict a low likelihood of stricture formation. In contrast, second-degree injuries, characterized by patchy mucosal ulceration with vesicles, grayish exudates, and/or pseudomembranes, are suggestive of transmucosal (partial-thickness) involvement. Approximately half of all second-degree injuries will result in stricture. Findings of a third-degree esophageal injury include deep ulcerations with eschar formation. There can also be mucosal sloughing as well as thrombosis of submucosal vessels. The edema in third-degree injuries can sometimes be quite severe so as to obliterate the entire lumen. Signs of third-degree esophageal injury are consistent with transmural (full-thickness) damage and will therefore result in stricture formation in the vast majority of cases.

10.5.2.2 Esophagoscopy in GERD

Although peptic strictures classically occur in the distal esophagus, GERD-related strictures can also occur in the mid-esophagus at the anastomosis in patients after repair of esophageal atresia (Fig. 10.2). Evaluation of the esophageal mucosa by flexible esophagoscopy can be useful in confirming the etiology of the stricture. In older children, endoscopic biopsy of multiple areas within the distal esophagus can be done with minimal morbidity. Specimens will demonstrate chronic inflammatory changes within the mucosa. In older children with longstanding reflux symptoms, the surgeon should also look for salmon-red mucosa >2 cm above the gastroesophageal junction. Such findings are suggestive of Barrett's esophagus and merit endoscopic biopsy. Pathology specimens will show intestinal metaplastic columnar epithelium with goblet cells.

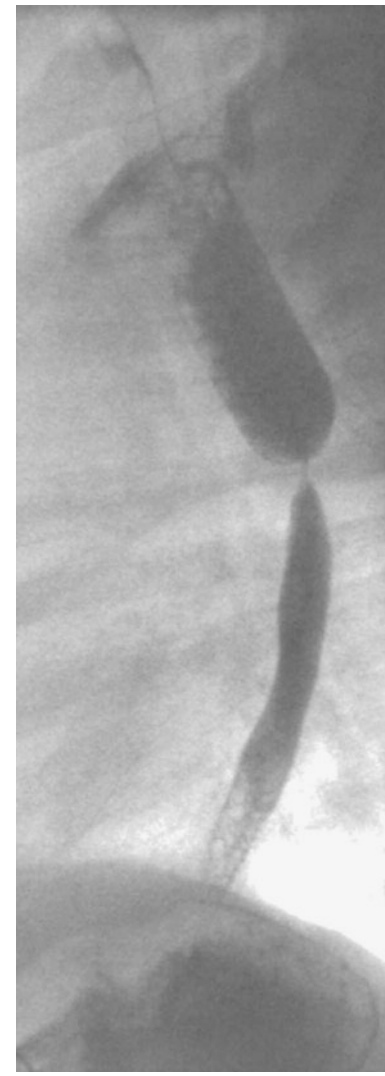

Fig. 10.2 Lateral contrast esophagram in a 4-month-old male after repair of esophageal atresia showing the interval development of a high-grade stricture at esophago-esophagostomy secondary to uncontrolled gastroesophageal reflux

10.5.2.3 Esophagoscopy in EoE

All children with a suspected esophageal narrowing secondary to EoE should undergo an endoscopic evaluation with possible biopsy. Findings on esophagoscopy consistent with EoE include a granular appearance of the esophageal mucosa with whitish exudates. A relatively specific finding of EoE is concentric mucosal rings, often referred to "trachealization" or "feline esophagus." However, not all patients with EoE will have obvious gross abnormalities, so a biopsy of multiple sites within the esophagus should always be done regardless of the appearance of the mucosa. Definitive cases of EoE will show >15 eosinophils per high-power field [16].

10.5.2.4 Other Diagnostic Studies

There are selected situations in which studies other than esophagography and esophagoscopy may be required to further delineate an esophageal stricture or stenosis. For example, angiography, computed tomography (CT) or magnetic resonance imaging (MRI) should be ordered to further evaluate patients with an obstruction secondary to a vascular ring. Similarly, an upper gastrointestinal contrast study and/or 24-h monitoring of esophageal pH may be indicated to help differentiate peptic esophageal strictures from esophageal stenosis or EoE. Prior to monitoring of esophageal pH, children need to temporarily discontinue all acid blockade medications to yield meaningful data that could guide further management. Finally, high-resolution esophageal manometry may be useful in some patients in whom achalasia or other functional disorders of the esophagus remain in the differential diagnosis.

10.6 Management

Appropriate management of esophageal strictures and stenoses depends largely on the underlying etiology. For example, in EoE, topical corticosteroids alone are the mainstay of therapy. For most other esophageal obstructions, endoscopic dilation is increasingly becoming the favored first line of treatment because of its minimal invasiveness and low prevalence of procedural morbidity. The specific management strategy based on etiology is detailed below.

10.6.1 Caustic Ingestion

In the acute setting after any suspected caustic injury, the patient should be placed on a strict *nothing per os* (NPO) diet. Blind placement of a nasogastric tube is contraindicated, and under no circumstances should the patient be given anything (including water) to dilute or neutralize the corrosive agent prior to diagnostic esophagoscopy. Such maneuvers may cause further esophageal injury by inducing an exothermic reaction and/or severe emesis.

First-degree injuries diagnosed by flexible esophagoscopy require no specific treatment because the risk of stricture formation is low. However, for all newly diagnosed second-degree injuries, the passage of a feeding nasogastric tube under direct visualization should be considered. In addition, all patients with second-degree injuries should be placed on a clear liquid diet as tolerated with esophagography ordered 2–3 weeks post-injury to assess for strictures. The indwelling nasogastric tube can serve as a guide for subsequent dilations if stricture formation occurs. If no stricture is present on follow-up esophagography, the tube can be removed and the child advanced to a regular diet as tolerated. There are surprisingly few data on the utility of any medication for a second-degree caustic injury. We advocate empirical parenteral antibiotics as well as chronic acid suppression in these patients. A seven-day course of piperacillin-sulbactam or a third-generation cephalosporin may be helpful in terms of protecting against potential direct bacterial translocation across the damaged esophageal wall and in diminishing the risk of aspiration pneumonia.

Third-degree caustic injuries should be treated initially with placement of a gastrostomy tube for two reasons. First, all of these patients are NPO, so the gastrostomy tube avoids the need for parenteral nutrition and provides stable access to the gastrointestinal tract for resumption of enteral feeds. Second, the gastrostomy tube facilitates placement of a transesophageal string guide that is passed through the gastrostomy site and one of the nares to maintain control of the esophageal lumen over the ensuing weeks of stricture formation. Commonly used guides include #3 braided silk suture, fishing wire, and silicone ventriculoperitoneal shunt tubing. The ends of the guide are then tied externally and taped on the child's back, leaving adequate laxity to prevent ulceration at the nose or gastrostomy sites while maintaining sufficient tension to keep

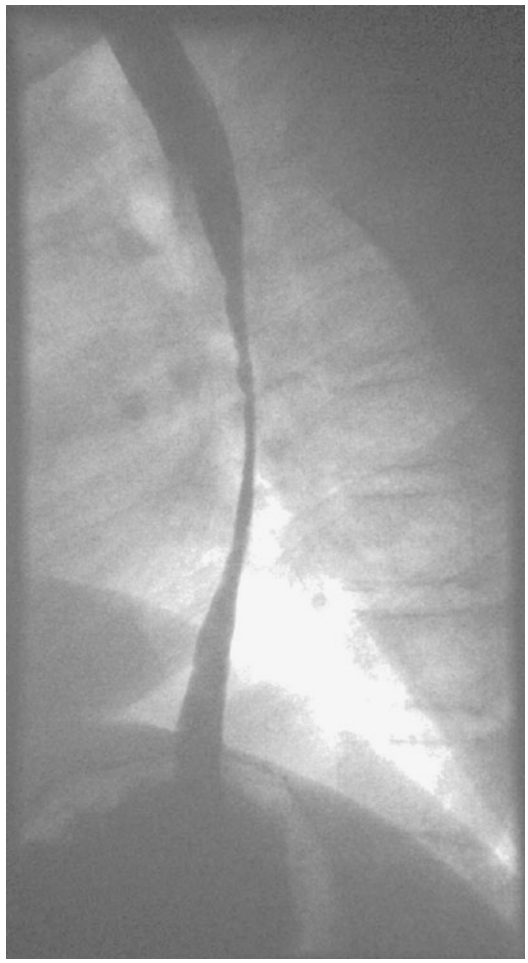

Fig. 10.3 Lateral contrast esophagram in an adolescent male 1 month after caustic injury demonstrating a long mid-esophageal stricture

from pulling out the guide. In the presence of a stricture on follow-up esophagography (Fig. 10.3), the guide allows for retrograde dilations through the site of the gastrostomy tube. Tucker dilators are specially designed for this purpose because they can be sequentially tied to the lower end of the string and passed through the stenosis in a retrograde fashion. String-guided retrograde dilation is associated with a lower prevalence of esophageal perforation and is therefore considered safer for high-grade and/or tortuous esophageal strictures [17]. One drawback of the retrograde approach is that the gastrostomy aperture is usually not well suited to accommodate larger-diameter bougies. However, in this situation one can simply reintroduce guided dilators in an anterograde fashion once a satisfactory lumen size has been established.

Since the 1950s, there has been considerable interest in the use of systemic corticosteroids to modify the inflammatory response and prevent stricture formation in high-grade esophageal injuries [15]. At present, its routine practice cannot be widely endorsed because the efficacy of corticosteroids in this setting has not been demonstrated in a randomized trial [12]. Nevertheless, proponents of corticosteroids suggest that most studies have included only a small number of patients and that corticosteroids may be beneficial if given at the appropriate dose and duration. For example, in one uncontrolled study, dexamethasone (0.5–1.0 mg per kg per day for 4–6 weeks was associated with a low prevalence of stricture formation [2]. Opponents of corticosteroids argue that the potentially deleterious effects of the drug (including the masking of septic complications and delays in esophageal wound healing) should not be underestimated. Although we do not routinely place patients with higher-grade corrosive injuries on corticosteroids, those who do receive corticosteroids should be placed on an antifungal agent in addition to a PPIs because of the known association between corticosteroid use with mycotic infections and peptic ulcer disease.

Management of a known caustic stricture remains highly dependent on the degree of narrowing as well as the length of the injured segment. In general, every reasonable effort should be made to preserve the native esophagus before resorting to resection, particularly if this involves esophageal replacement. Therefore, the initial approach includes serial endoscopic dilations every 2–4 weeks using an anterograde and/or retrograde approach based on the degree of injury. The optimal instrument used for dilation (i.e., hydrostatic balloon: Hurst–Maloney, Savary–Gillard, Jackson, Tucker) should be individualized based

on the nature of the stricture as well as the experience of the surgeon. Although somewhat controversial, dense fibrotic strictures involving a long segment of the esophagus tend to be more responsive to bougeinage as opposed to hydrostatic balloon systems.

Management of recurrent caustic esophageal strictures represents one of the most challenging problems in pediatric thoracic surgery. Prophylactic dilations to prevent strictures have been explored by several authors but their role remains undefined [18]. In many cases, perseverance is all that is required because some patients may require >20 dilation procedures until the stricture resolves. Management of short-segment strictures prior to dilation with endoscopic-assisted intralesional corticosteroids (1% triamcinolone) or topical mitomycin C (an anti-neoplastic agent that has been shown to reduce fibroblastic collagen synthesis by inhibiting DNA-dependent RNA synthesis) has shown promise in several reports [19, 20] Unfortunately, the efficacy of these agents has not been examined prospectively.

Long (>5 cm) esophageal strictures, particularly those that are persistent, circumferential in nature, and located in the mid-esophagus, are reasonable candidates for a temporary, covered esophageal stent. Esophageal stents are expandable and therefore designed to provide continuous, radially oriented force vectors sustained over a prolonged period of time. Unfortunately, few centers have extensive experience with esophageal stents in the pediatric population [21–23]. Moreover, the use of esophageal stents has been controversial because many of these devices are not well tolerated for more than several weeks, are prone to migration, and can be difficult to remove in a safe manner. Nevertheless, covered stents may serve an important role in esophageal strictures with a concomitant fistula as a temporary measure before definitive surgery. Even if the stricture responds to dilations or stenting, eventual esophageal shortening can occur with the subsequent development of GERD. All of these patients should be on long-term antacid therapy and some may eventually require a fundoplication with or without a Collis gastroplasty. Although the treatment plan should be individualized for every child with a persistent esophageal stricture, we would generally advise esophageal surgery if serial endoscopic dilation or stenting fails to remediate the stricture after 6–12 months.

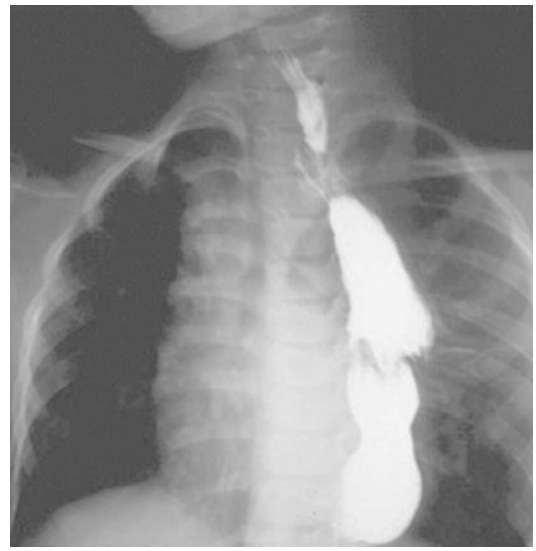

Fig. 10.4 Anterior contrast esophagram after esophagectomy with a gastric transposition placed through the posterior mediastinum

Short-segment strictures refractory to minimally invasive procedures can be considered for esophagoplasty or segmental esophageal resection. Colonic patch procedures have also been used with some success for less extensive disease [24]. Up to 60% of caustic strictures, particularly those that are long segment in nature, eventually require esophagectomy with conduit reconstruction. Based on our series of 9 patients with severe caustic strictures as well as our extensive experience with long-gap esophageal atresia, we favor the gastric transposition technique for esophageal reconstruction (Fig. 10.4) [25]. More recently, Javed reported similarly favorable outcomes in a large series of patients after gastric transposition when compared with other conduit procedures, including colon interposition [26].

Patients requiring esophagectomy are best cared for at major pediatric specialty centers with surgical expertise in the management of complex esophageal problems. A description of the surgical techniques involved in these procedures is beyond the scope of this chapter.

10.6.2 GERD

The initial principles of management for GERD-related strictures were established decades ago and include nutritional support, acid blockade, and serial bougienage [3]. Although some authors would recommend a primary anti-reflux procedure for peptic strictures, it may be technically more difficult to dilate the stricture just proximal to a fundoplication, and there have been additional concerns about causing trauma to the wrap itself after repeated dilations. Therefore, our research team as well as others advocate serial dilations while on a high-dose PPI (omeprazole 2 mg/kg/day) for several months [7]. Once the stricture has fully resolved and the child has been optimized from a nutritional and respiratory perspective, we would proceed with an anti-reflux procedure. Most children are excellent candidates for a Nissen fundoplication as definitive therapy to prevent further stricture recurrence [27]. Rarely, a Collis gastroplasty may also be required in conjunction with a fundoplication if there is significant esophageal foreshortening.

10.6.3 EoE

The mainstay of therapy for EoE includes elimination of all food allergens and initiation of topical corticosteroids. Topical corticosteroids are most easily delivered by ingestion of the medication (e.g., fluticasone proprionate) *via* a metered-dose inhaler [28]. Unfortunately, almost all patients will develop recurrent symptoms and esophageal eosinophilia after discontinuation of medical therapy [13]. Systemic corticosteroids should be reserved only for EoE patients with severe dysphagia or failure to thrive. PPIs may be helpful in a small subset of patients but should not to be considered as the sole primary therapy. Based on the literature in adult subjects, endoscopic dilation should be reserved for failure of medical management because dilation alone does not address the underlying inflammatory process, and there may be an increased risk of esophageal perforation in EoE when compared with strictures secondary to other disease processes [16].

10.6.4 Anastomotic Strictures

Although anastomotic strictures can be more difficult to treat compared with many other types of strictures, most are responsive to repeated anterograde dilation [29]. The frequency of these dilations should be individualized, but usually every 2–3 weeks in the initial period. At our institution, we favor serial dilations under combined endoscopic and fluoroscopic guidance using a hydrostatic balloon catheter filled with contrast reagent (Fig. 10.5). For high-grade strictures, a 0.035-inch guidewire can be used to position the balloon across the narrowing. Many patients with recurrent anastomotic strictures have undergone esophageal atresia repair in the neonatal period, so 24-h pH probe testing or an empirical trial of a PPI should also be considered. Recurrent strictures may also benefit from intramural injections of corticosteroid delivered using a sclerotherapy needle. Finally, placement of a temporary, covered esophageal stent may also be an option in selected patients (Fig. 10.6), although this may exacerbate pre-existing GERD and lead to worsening pulmonary symptoms in some cases. Overall, <5% of anastomotic strictures after esophageal atresia repair fail endoscopic management and require further surgical intervention such as segmental esophageal resection or esophageal substitution [10].

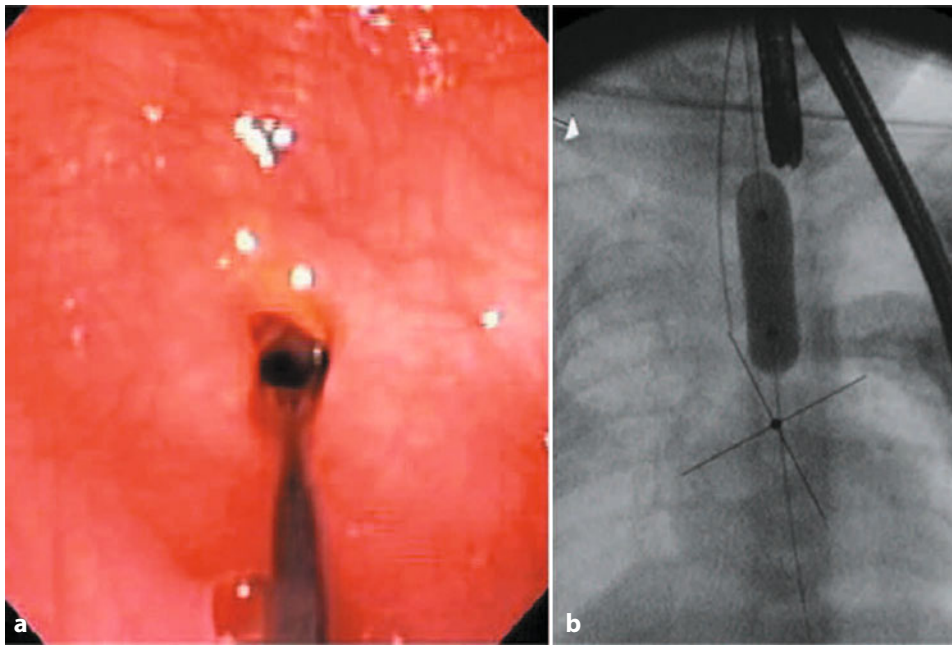

Fig. 10.5 Endoscopic view showing a high-grade anastomotic stricture after repair of esophageal atresia (**a**). A 0.035-inch guidewire is shown passing through the stricture at the 6 o'clock position. Anterior fluoroscopic image demonstrating successful dilation of the same stricture using a 10-mm hydrostatic balloon inflated with contrast medium (**b**)

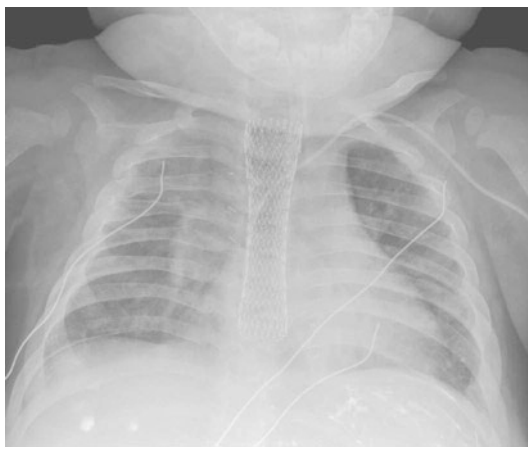

Fig. 10.6 Anterior chest radiograph of a fully covered esophageal stent (12 mm; Allimax) deployed in an infant referred for management of a recurrent esophageal stricture after repair of esophageal atresia

10.6.5 Esophageal Stenosis

The treatment of congenital esophageal stenosis remains controversial in large part because there are few large case series in the literature [11]. Historically, short, abrupt strictures were thought to contain rigid tracheobronchial remnants and were therefore not perceived to be amenable to endoscopic dilation without risk of perforation [30, 31]. In contrast, infants with stenosis secondary to fibromuscular dysplasia, as suggested by a short, tapered stricture within the lower esophagus, are considered to be excellent candidates for endoscopic dilation. In an effort to delineate the subtype of esophageal stenosis for further management guidance, the use of endoscopic ultrasound has been described but has not been studied extensively [11, 32].

More recently, our institution has favored empirical esophageal dilation using hydrostatic balloons or tapered, weighted (Hurst–Maloney) bougies as the first line of treatment in all cases of esophageal stenosis [11, 33]. Although the radial force induced by balloon dilators has some theoretical advantages with regards to wound healing, bougienage can provide greater tactile feedback, thereby reducing the risk of perforation. Depending on the de-

gree and duration of response after esophageal dilation, additional procedures can be done with potential long-term benefit. Iatrogenic perforations have been reported in ≤10% of cases, but many of these injuries can be treated medically depending on the size of perforation and clinical status [11]. Endoscopic ablation of a congenital membranous web represents another option that has been described recently [34].

If endoscopic maneuvers fail, the remaining treatment options for esophageal stenosis are segmental resection and esophagomyotomy. Resection of a congenital stenosis with primary esophago-esophagostomy *via* left thoracotomy or, more recently, left thoracoscopy, has been achieved with acceptable results [35]. However, segmental resection is recommended only if the stricture is <2.5 cm in length because longer resections often result in an intra-thoracic stomach and severe GERD that can be difficult to manage even with subsequent anti-reflux procedures. Postoperative anastomotic strictures after short-segment resections occur because of ischemia, tension, and/or untreated GERD. Fortunately, this complication usually responds to endoscopic dilation and anti-reflux medications. For distal lesions that involve the gastroesophageal junction, an anti-reflux procedure should be considered at the time of segmental resection. Esophagectomy for congenital stenosis is rarely, if ever, indicated.

10.6.6 EB

Historically, the management of EB esophageal strictures with dilations has been quite challenging because of the potential for creating severe iatrogenic mucosal sloughing. This can lead to significant chest pain and subsequent stricture formation after the procedure. Therefore, endoscopy and bougienage are now considered relative contraindications in the management of EB strictures, and hydrostatic balloon dilation under fluoroscopic guidance has become the preferred technique [36]. Balloon dilation allows for the application of uniform radial force at the site of the stricture while minimizing tangential forces that may cause inadvertent mucosal trauma to other areas of the esophagus. An added benefit of this approach is the potential avoidance of orotracheal intubation in this patient population.

10.6.7 Schatski Ring

Management of Schatski rings includes control of any underlying GERD using PPIs. For those who do not respond appropriately to acid blockade, recent data suggest that EoE should be considered with appropriate medical therapy such as topical corticosteroids employed in all cases in whom eosinophilia is identified on esophageal biopsy [37]. Endoscopic dilation of Schatzki rings has also been shown to be effective in refractory cases [14].

10.7 Outcome and Follow-up

Most children with successfully treated esophageal obstructions can resume complete oral nutrition with little or no dysphagia. Nevertheless, there are several long-term issues worth noting. For example, in children with significant residual scarring aftyer caustic ingestion who do not undergo esophageal replacement, there is an ≈1000-fold increased risk of esophageal carcinoma that can occur 30–45 years after caustic injury [38]. Because of the potential risk for malignant transformation, long-term endoscopic surveillance is advocated in this patient population. Similarly, most children managed by esophageal replacement require close follow-up because of significant short-term as well as long-term morbidities associated with esophagogastric anastomotic leak, recurrent stricture formation, and feeding intolerance [25]. These complications are yet another reason why esophagectomy patients are best cared for at major specialty centers equipped with surgical expertise and multidisciplinary care teams.

References

1 Rothstein FC (1986) Caustic injuries to the esophagus in children. Pediatr Clin North Am 33:665–674
2 Riffat F, Cheng A (2009) Pediatric caustic ingestion: 50 consecutive cases and a review of the literature. Dis Esophagus 22:89–94
3 O'Neill JA, Jr., Betts J, Ziegler MM et al (1982) Surgical management of reflux strictures of the esophagus in childhood. Ann Surg 196:453–460
4 Guda NM, Vakil N (2004) Proton pump inhibitors and the time trends for esophageal dilation. Am J Gastroenterol 99:797-800
5 Haller JA, Jr., Andrews HG, White JJ et al (1971) Pathophysiology and management of acute corrosive burns of the esophagus: results of treatment in 285 children. J Pediatr 6:578–584
6 Yardeni D, Yardeni H, Coran AG et al (2004) Severe esophageal damage due to button battery ingestion: can it be prevented? Pediatr Surg Int 20:496–501
7 Numanoglu A, Millar AJ, Brown RA et al (2005) Gastroesophageal reflux strictures in children, management and outcome. Pediatr Surg Int 21:631–634
8 Manning PB, Morgan RA, Coran AG et al (1986) Fifty years' experience with esophageal atresia and tracheoesophageal fistula. Beginning with Cameron Haight's first operation in 1935. Ann Surg 204:446–453
9 Serhal L, Gottrand F, Sfeir R, et al (2010) Anastomotic stricture after surgical repair of esophageal atresia: frequency, risk factors, and efficacy of esophageal bougie dilatations. J Pediatr Surg 45:1459–1462
10 Chittmittrapap S, Spitz L, Kiely EM et al (1990) Anastomotic stricture following repair of esophageal atresia. J Pediatr Surg 25:508–511
11 Romeo E, Foschia F, de Angelis P et al (2011) Endoscopic management of congenital esophageal stenosis. J Pediatr Surg 46:838–841
12 Anderson KD, Rouse TM, Randolph JG (1990) A controlled trial of corticosteroids in children with corrosive injury of the esophagus. N Engl J Med 323:637–640
13 Furuta GT, Liacouras CA, Collins MH et al (2007) Eosinophilic esophagitis in children and adults: a systematic review and consensus recommendations for diagnosis and treatment. Gastroenterology 133:1342–1363
14 Buckley K, Buonomo C, Husain K, et al (1998) Schatzki ring in children and young adults: clinical and radiologic findings. Pediatr Radiol 28:884–886
15 Estrera A, Taylor W, Mills LJ et al. Corrosive burns of the esophagus and stomach: a recommendation for an aggressive surgical approach. Ann Thorac Surg 41:276–283
16 Liacouras CA, Furuta GT, Hirano I et al (2011) Eosinophilic esophagitis: updated consensus recommendations for children and adults. J Allergy Clin Immunol 128:3–20
17 Hawkins DB (1988) Dilation of esophageal strictures: comparative morbidity of antegrade and retrograde methods. Ann Otol Rhinol Laryngol 97:460–465
18 Tiryaki T, Livanelioglu Z, Atayurt H (2005) Early bougienage for relief of stricture formation following caustic esophageal burns. Pediatr Surg Int 21:78–80
19 Berenson GA, Wyllie R, Caulfield M et al (1994) Intralesional steroids in the treatment of refractory esophageal strictures. J Pediatr Gastroenterol Nutr 18:250-252
20 Uhlen S, Fayoux P, Vachin F et al (2006) Mitomycin C: an alternative conservative treatment for refractory esophageal stricture in children? Endoscopy 38:404–407
21 de Jong AL, Macdonald R, Ein S, et al (2001) Corrosive esophagitis in children: a 30-year review. Int J Pediatr Otorhinolaryngol 57:203–211
22 Zhang C, Yu JM, Fan GP, et al (2005) The use of a retrievable self-expanding stent in treating childhood benign esophageal strictures. J Pediatr Surg 40:501–504
23 Kramer RE, Quiros JA (2010) Esophageal stents for severe strictures in young children: experience, benefits, and risk. Curr Gastroenterol Rep 12:203–210
24 Othersen HB, Jr., Parker EF, Smith CD (1988) The surgical management of esophageal stricture in children. A century of progress. Ann Surg 207:590–597
25 Hirschl RB, Yardeni D, Oldham K et al (2002) Gastric transposition for esophageal replacement in children: experience with 41 consecutive cases with special emphasis on esophageal atresia. Ann Surg 236:531–539
26 Javed A, Pal S, Dash NR, et al (2011) Outcome following surgical management of corrosive strictures of the esophagus. Ann Surg 254:62–66
27 Rode H, Millar AJ, Brown RA et al (1992) Reflux strictures of the esophagus in children. J Pediatr Surg 27:462–465
28 Konikoff MR, Noel RJ, Blanchard C et al (2006) A randomized, double-blind, placebo-controlled trial of fluticasone propionate for pediatric eosinophilic esophagitis. Gastroenterology 131:1381–1391
29 Lan LC, Wong KK, Lin SC et al (2003) Endoscopic balloon dilatation of esophageal strictures in infants and children: 17 years' experience and a literature review. J Pediatr Surg 38:1712–1715
30 Shorter NA, Mooney DP, Vaccaro TJ et al (2000) Hydrostatic balloon dilation of congenital esophageal stenoses associated with esophageal atresia. J Pediatr Surg 35:1742–1745
31 Vasudevan SA, Kerendi F, Lee H et al (2002) Management of congenital esophageal stenosis. J Pediatr Surg 37:1024–1026
32 Takamizawa S, Tsugawa C, Mouri N et al (2002) Congenital esophageal stenosis: therapeutic strategy based on etiology. J Pediatr Surg 37:197–201
33 Jones DW, Kunisaki SM, Teitelbaum DH et al (2010)

Congenital esophageal stenosis: the differential diagnosis and management. Pediatr Surg Int 26:547–551
34 Roy GT, Cohen RC, Williams SJ (1996) Endoscopic laser division of an esophageal web in a child. J Pediatr Surg 31:439–440
35 Martinez-Ferro M, Rubio M, Piaggio L et al (2006) Thoracoscopic approach for congenital esophageal stenosis. J Pediatr Surg 41:E5–E7
36 Anderson SH, Meenan J, Williams KN et al (2004) Efficacy and safety of endoscopic dilation of esophageal strictures in epidermolysis bullosa. Gastrointest Endosc 59:28–32
37 Nurko S, Teitelbaum JE, Husain K et al (2004) Association of Schatzki ring with eosinophilic esophagitis in children. J Pediatr Gastroenterol Nutr 38:436–441
38 Hopkins RA, Postlethwait RW (1981) Caustic burns and carcinoma of the esophagus. Ann Surg 194:146–148