Chapter 54 – Dysfunction of the Upper Esophageal Sphincter

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The cricopharyngeus muscle, first described by Valsalva in 1917, is the major component of the upper esophageal sphincter (UES). Jackson and Shallow subsequently distinguished the cricopharyngeus from other muscles of the UES, including the distal inferior constrictor and the proximal circular esophageal muscle.[1] The cricopharyngeus muscle is innervated by the pharyngeal plexus, which is composed of contributions from the vagus and glossopharyngeal nerves. Increasing evidence suggests that contributions from the recurrent laryngeal nerve also innervate the cricopharyngeus muscle.[2] The origin of the parasympathetic innervation is vagal, whereas sensory branches are derived from the glossopharyngeal nerve. Sympathetic fibers arise from the superior cervical ganglion and also join the pharyngeal plexus.

In its normal resting state, the cricopharyngeus muscle is in tonic contraction. When a swallow occurs, the cricopharyngeus relaxes during pharyngeal contraction (mediated by the vagus nerve), and anterior-superior movement of the larynx opens the UES to allow passage of the bolus into the esophagus. Dysphagia related to dysfunction of the UES results from (1) failure of cricopharyngeal relaxation, which can be due to lack of vagal innervation or loss of the normal viscoelastic properties of the muscle; (2) discoordination between cricopharyngeal relaxation and pharyngeal contraction; or (3) weakness of the suprahyoid musculature with poor elevation (anterosuperior movement) of the laryngeal complex.

In 1951 Kaplan described division of the cricopharyngeus muscle in treating dysphagia resulting from bulbar poliomyelitis.[3] Since then, cricopharyngeal myotomy has been used for treating cricopharyngeal dysfunction caused by lesions of the central and peripheral nervous systems, which affect laryngeal and pharyngeal muscle activity and coordination, as well as for the treatment of postsurgical dysphagia. Although indications for cricopharyngeal myotomy are controversial, in general, the procedure is most successful when treating patients with normal or nearly normal pharyngeal function, which is required for bolus propulsion, and with adequate laryngeal elevation, which is required for opening of the UES. For example, patients with amyotrophic lateral sclerosis (ALS), Parkinson's disease, or cerebellar atrophy have been treated by cricopharyngeal myotomy with generally poor results because these conditions are characterized by diffuse pharyngeal motor abnormalities and poor anterosuperior elevation.

One of the most widely recognized manifestations of UES dysfunction is Zenker's diverticulum. Although the first description of a pharyngoesophageal diverticulum is attributed to Ludlow in 1769, this entity became known as Zenker's diverticulum after it was described in Zenker and Ziemssen's *Krankheiten des Oesophagus,* a landmark compilation of esophageal pathology published in 1877. Killian clarified the anatomic site of origin as the space between the inferior constrictor muscle and the cricopharyngeus muscle, hence its name Killian's triangle (Fig. 54-1).[4] Zenker's diverticulum is a pulsion diverticulum that forms in the muscular dehiscence of Killian's triangle as the UES fails to open with pharyngeal contraction, thus raising pressure in the pharynx (Fig. 54-2). Other areas of weakness have been identified and can lead to variations of the site of origin of the diverticulum (Killian-Jamieson and Lamier triangles). Failure of UES opening may be the result of failure of cricopharyngeal relaxation or weakness of laryngeal elevation.

Standard treatment of Zenker's diverticulum includes a myotomy of the UES combined with resection of the diverticulum through a left-sided cervical incision, but other techniques are also in common use. Dohlman and Mattson described an endoscopic technique for cricopharyngeal myotomy that has been modified with the introduction of lasers, staplers, and bivalved endoscopes.[5] Advances in endoscopic instrumentation enabled Collard and associates to perform the first esophagodiverticulostomy with an endoscopic stapler in 1993.[6] This procedure involves endoscopic division of the cricopharyngeus muscle with marsupialization of the diverticulum into the pharyngoesophagus. Modification of the stapler as described by Lang and colleagues allows the division to extend as close as possible to the inferior aspect of the diverticulum.[7] Recent studies have shown that use of the endoscopic stapler technique results in shorter operative time, hospital stay, and time to resume oral feeding, as well as a lower complication rate than occurs with the open approach.[8,9] An alternative endoscopic approach uses the CO_2 laser to divide the party wall between the esophagus and diverticulum. A recent study, however, demonstrated a higher recurrence rate with this technique than with the open approach.[10] Furthermore, the open approach is still preferable for diverticula that are too small to accommodate the endoscopic stapler (<3 cm) or when endoscopic visualization is precluded by anatomic factors.

Since 1994, botulinum toxin A (BTX A) has been used as an alternative to surgical cricopharyngeal myotomy in the treatment of selected cases of UES dysfunction. Preliminary studies show BTX A to be a safe and effective treatment of UES dysfunction of multiple causes, including cerebrovascular accident, neuropathy, and laryngectomy.[11] BTX A may be injected transcutaneously in the office under electromyographic guidance or endoscopically in the operating room.[12] Given the preliminary nature of the studies, BTX A should generally be reserved for patients who refuse surgery or are not surgical candidates or when the diagnosis, prognosis, and expected response to myotomy are in question. In addition, BTX A improves only UES relaxation; thus, patients with poor pharyngeal propulsion or weak elevation of the larynx, such as those with Parkinson's disease or ALS, are unlikely to benefit from BTX A injection.

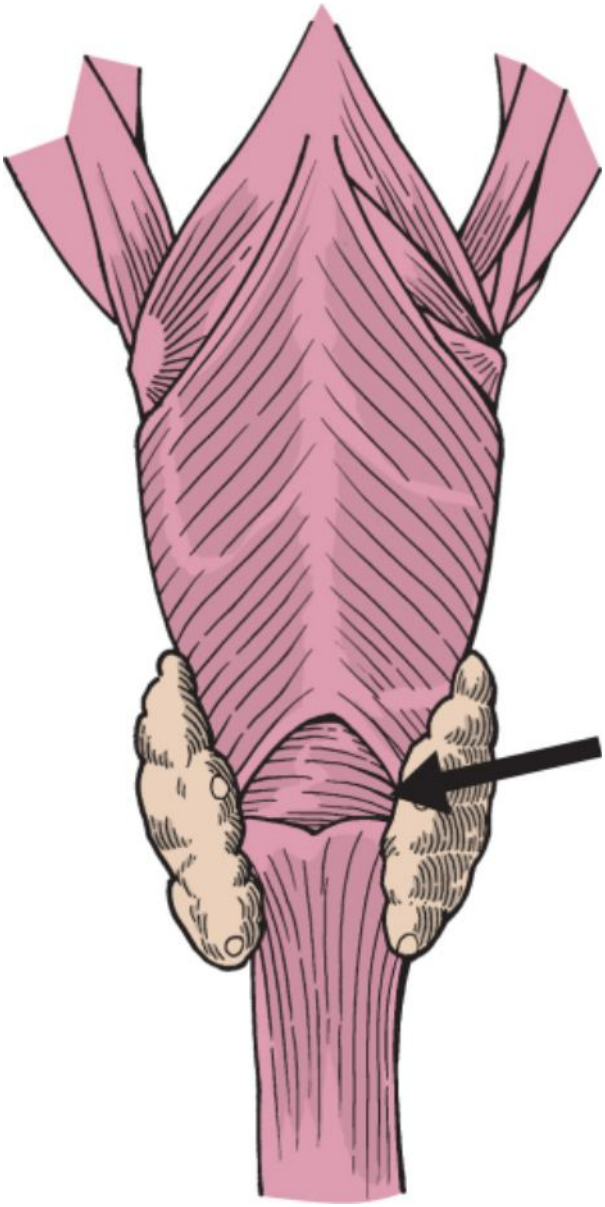

Figure 54-1 Killian's triangle is bounded superiorly by fibers of the inferior constrictor muscle and inferiorly by the cricopharyngeus muscle *(arrow)*.

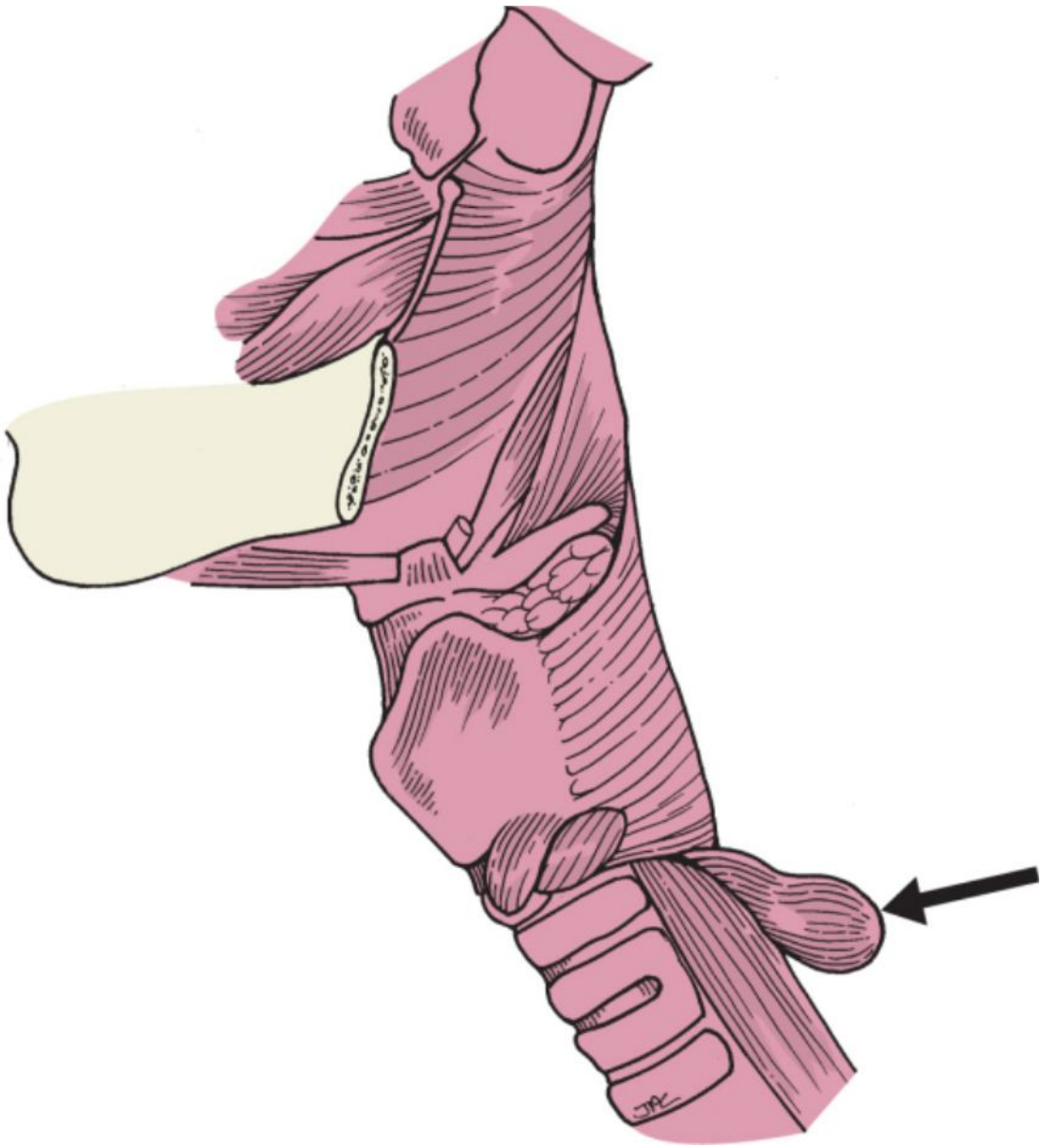

Figure 54-2 Zenker's diverticulum posterior to the esophagus *(arrow)*.

PATIENT SELECTION

Obtaining a thorough history in patients complaining of dysphagia is of great importance. The clinician should inquire whether the patient has experienced weight loss, has a history of pneumonia, or has dietary limitations as a result of dysphagia. A history of cerebrovascular accident or associated neurologic or neuromuscular disease helps exclude other types of problems such as malignancy. Previous major head and neck surgery or radiation therapy for cancer of the head and neck may be associated with UES dysfunction. Progressive dysphagia with associated regurgitation of undigested food and halitosis suggests Zenker's diverticulum. Signs and symptoms of gastroesophageal reflux disease (GERD) should also be elicited. After considering all these aspects in the history and completing the evaluation, the clinician must also consider the diagnosis of cricopharyngeal achalasia or “cricopharyngeal dysphagia,” the cause of which is unknown.

Radiologic studies, including barium swallow esophagography and modified barium swallow, are useful in evaluating UES function. The most common radiographic findings in cricopharyngeal achalasia, regardless of the cause, are prominence of the cricopharyngeal sphincter muscle, which is best seen on the lateral view at the level of the C5-C7 vertebral bodies, and failure of relaxation of the cricopharyngeus muscle during the pharyngeal phase of swallowing (Fig. 54-3). Likewise, barium esophagography is essential in confirming the diagnosis of Zenker's diverticulum (Fig. 54-4). Modified barium swallow can also identify and quantify associated laryngeal penetration of the bolus and aspiration and evaluate coordination of swallow and elevation of the larynx.

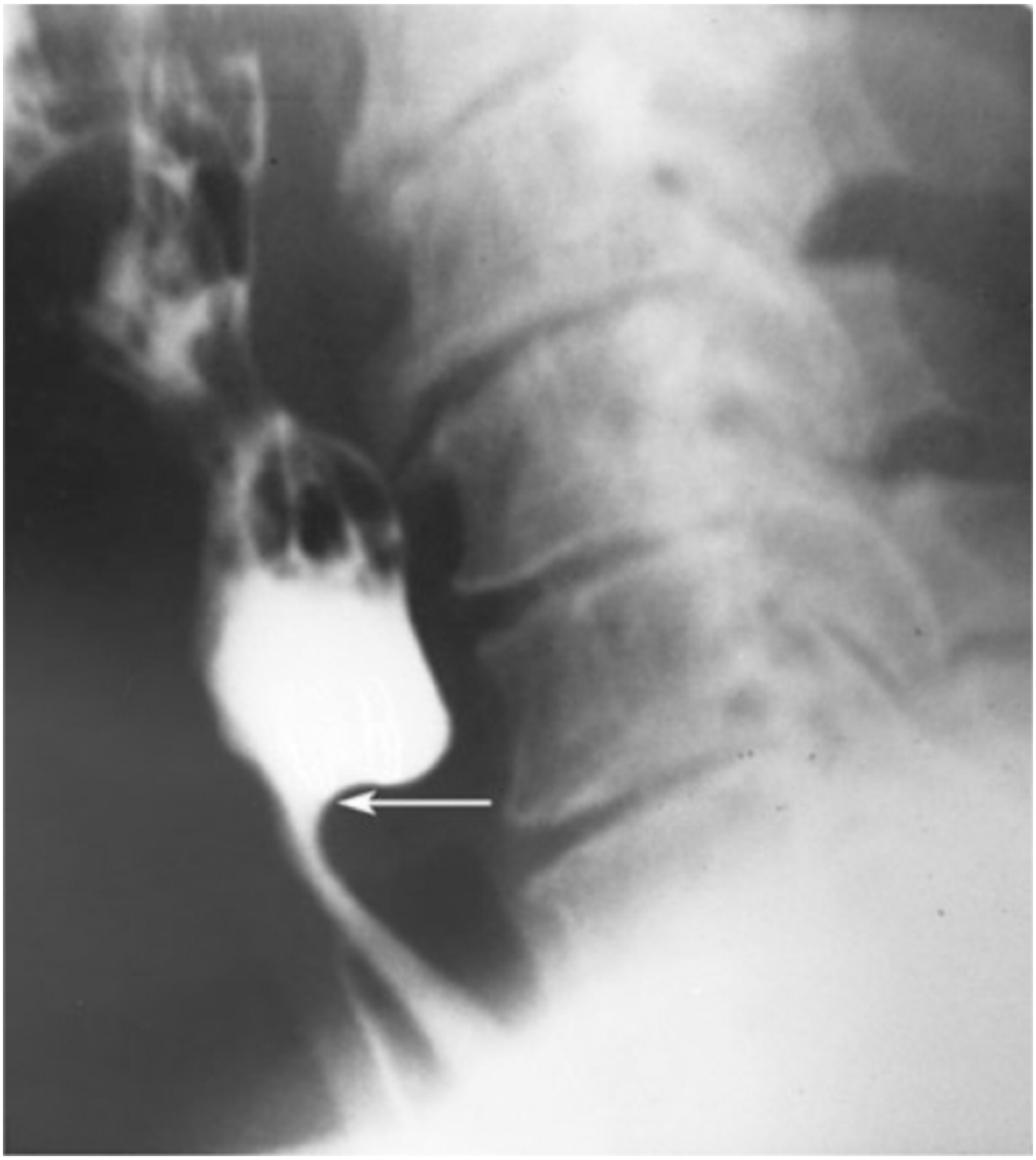

Figure 54-3 Barium esophagogram (lateral view) demonstrating failure of relaxation of the cricopharyngeus muscle *(arrow)*.

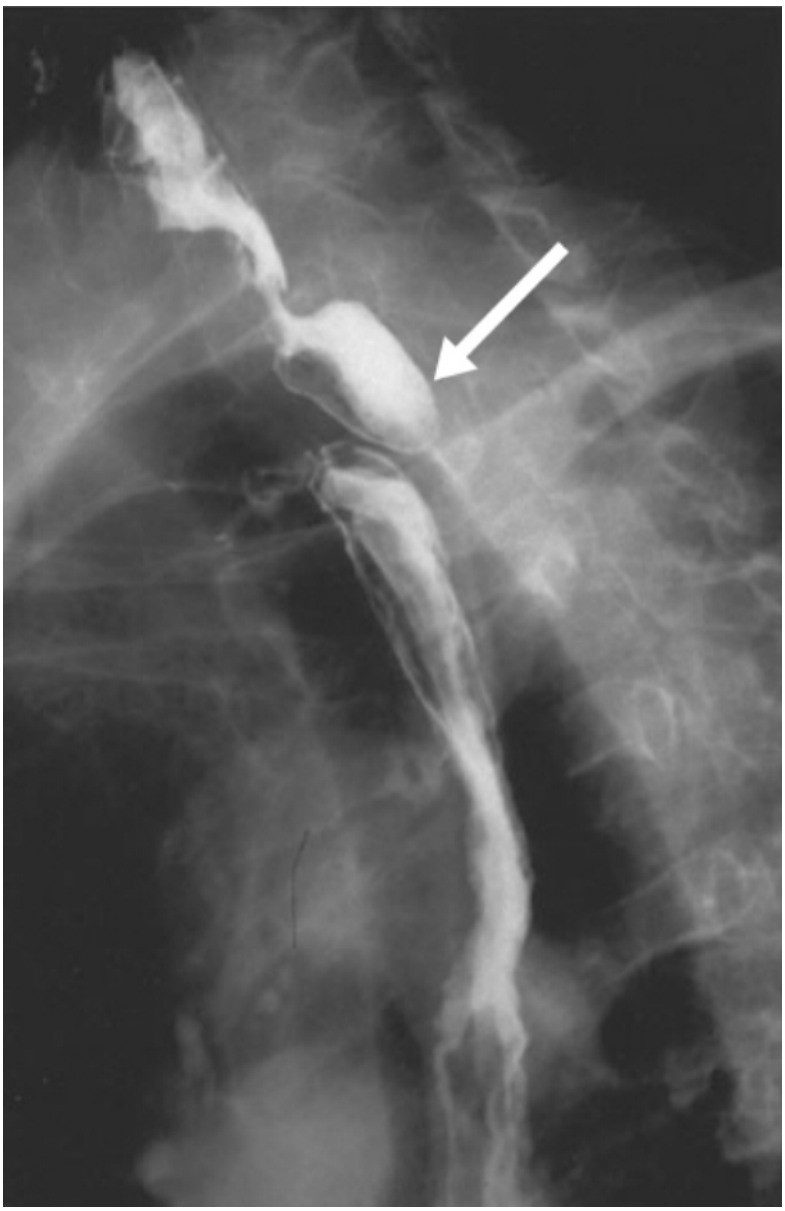

Figure 54-4 Barium esophagogram demonstrating Zenker's diverticulum *(arrow)*.

Functional endoscopic examination of swallowing (FEES) is a complementary tool in assessing swallowing function but does not provide the necessary information to evaluate UES function. FEES may provide useful information, such as postcricoid pooling of secretions (Fig. 54-5) and ingested material, as well as regurgitation consistent with UES dysfunction. Weakness of the palate, pharynx, and base of the tongue or discoordinated initiation of deglutition may suggest more global oropharyngeal dysphagia as opposed to isolated UESpathology. FEES also helps identify laryngeal penetration or aspiration of saliva or ingested material. Occasionally, regurgitation into the left piriform sinus is directly observed and is highly suggestive of Zenker's diverticulum.

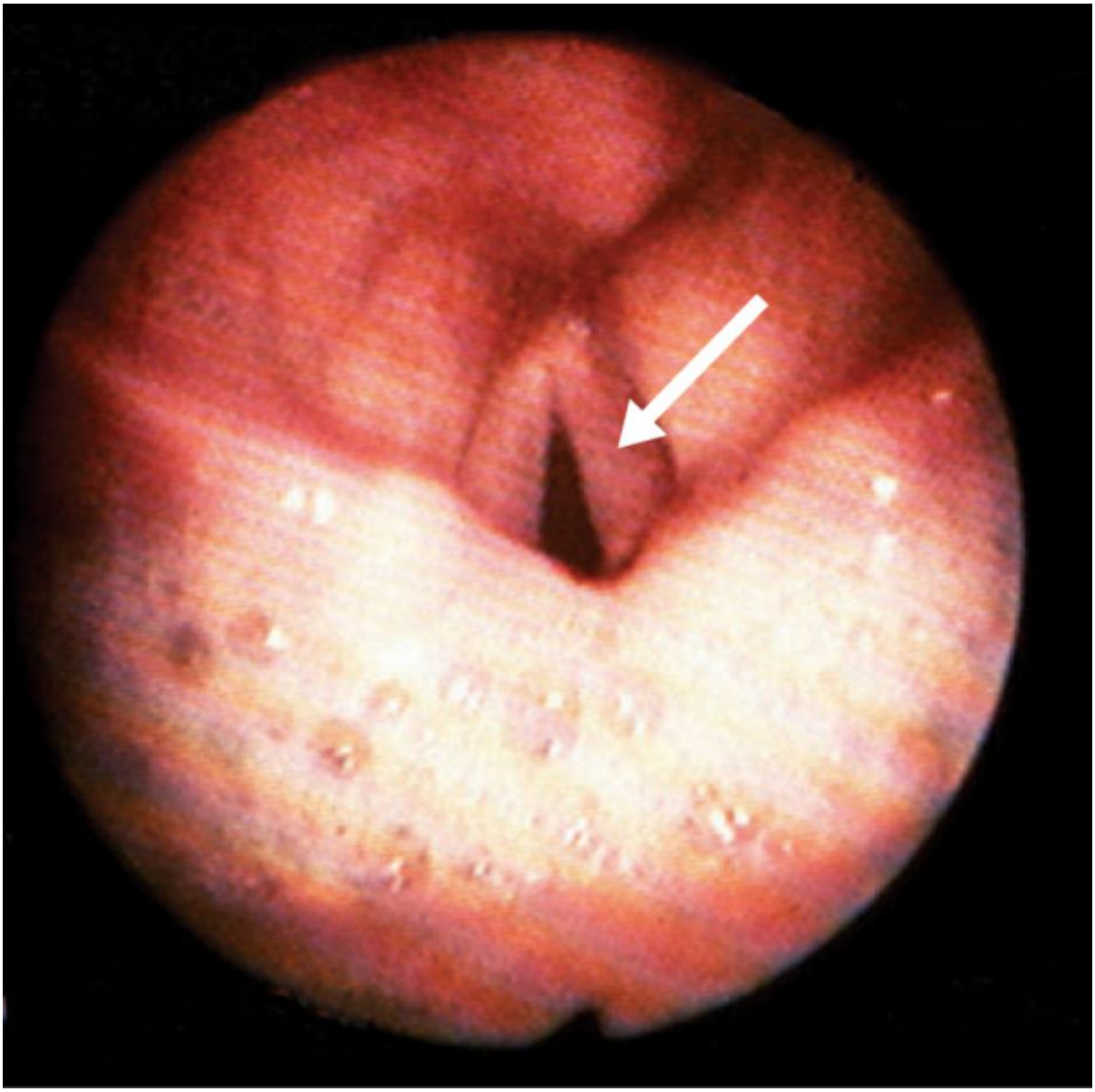

Figure 54-5 Postcricoid pooling of secretions detected by functional endoscopic examination of swallowing.

The role of manometry in assessing the UES is more controversial. Though frequently used as a research tool, manometry has not been widely applied clinically for the evaluation and management of patients.

PREOPERATIVE PLANNING

Patients with global neurologic or neuromuscular problems should be carefully evaluated by a neurologist and anesthesiologist before surgery. A precise diagnosis is important because patients with ALS or Parkinson's disease do not benefit from cricopharyngeal myotomy and should not undergo this procedure.

Patients with significant pulmonary problems should be prepared with extensive respiratory therapy preoperatively. Patients with uncontrolled GERD should be treated both preoperatively and postoperatively to minimize this problem.

Intravenous perioperative antibiotics are administered to minimize postoperative wound infection in the event that the pharynx is entered.

SURGICAL TECHNIQUE

Cricopharyngeal Myotomy

This operation is usually carried out under general anesthesia but local anesthesia may also be used if the patient's condition does not permit general anesthesia. The neck is extended with a rolled blanket placed under the

shoulders. Rigid esophagoscopy is performed to exclude neoplasm or inflammatory esophageal disease as a cause of the dysphagia. A transverse incision is carried out in a skinfold close to the level of the cricoid cartilage, which marks the cricopharyngeal muscle (Fig. 54-6). After the incision is made and carried down through the platysma muscle, the flapsare elevated inferiorly to the clavicle and superiorly above the thyroid notch (Fig. 54-7). Sharp separation of the sternocleidomastoid (SCM) muscle from the strap muscles exposes the carotid sheath. The SCM and carotid sheath are separated from the strap muscles, thyroid gland, and laryngotracheal complex and re-tracted laterally to allow entry into the retropharyngeal-prevertebral space. The middle thyroid vein and the omohyoid muscle may be divided to facilitate the exposure and decrease the need for retraction. A large double hook is placed in the posterior aspect of the midthyroid lamina, and the larynx, trachea, and thyroid gland are rolled medially (Fig. 54-8). The loose areolar tissue is bluntly dissected down to the prevertebral fascia. Bipolar cauterization of the fascia covering the pharyngeal musculature will reveal the UES muscles. This maneuver should be performed close to midline to avoid injury to the recurrent laryngeal nerve. The cricoid cartilage will mark the level of the cricopharyngeal sphincter; therefore, if the myotomy is performed at the level of the cricoid cartilage and extended above and below this level, it will be successful.

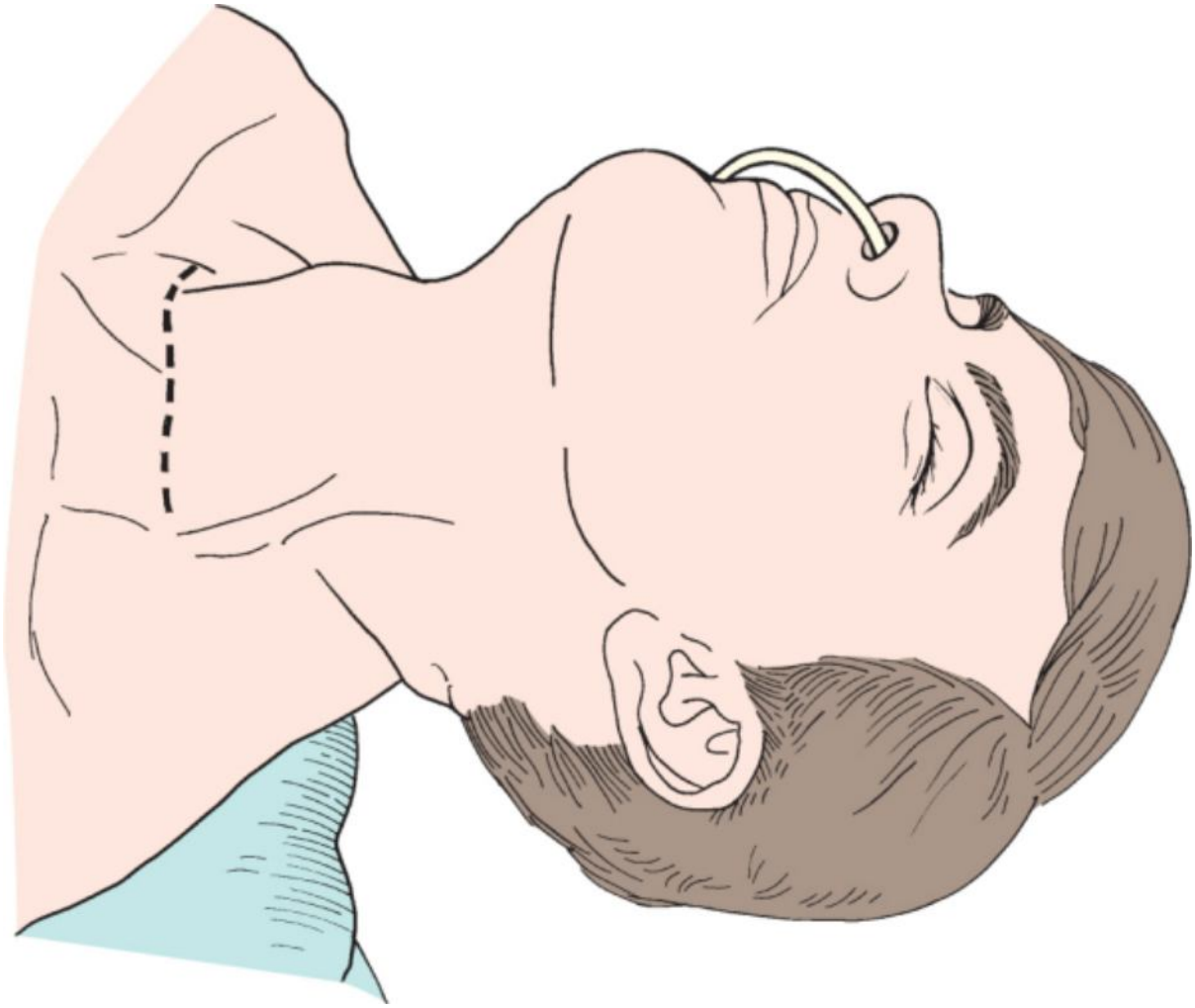

Figure 54-6 A transverse incision is made in a skin crease at the level of the cricoid cartilage.

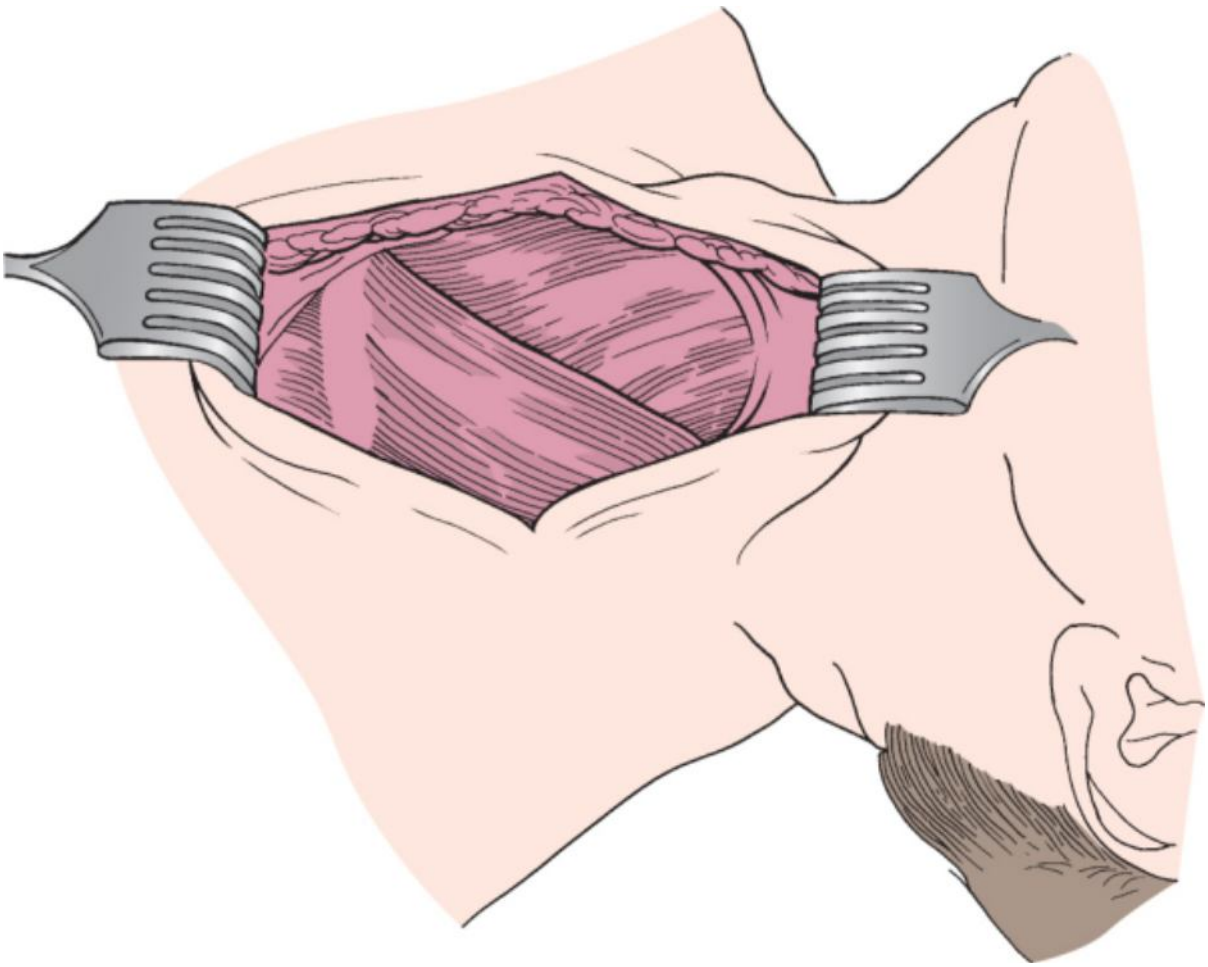

Figure 54-7 Subplatysmal flaps are undermined to expose the sternocleidomastoid and strap muscles.

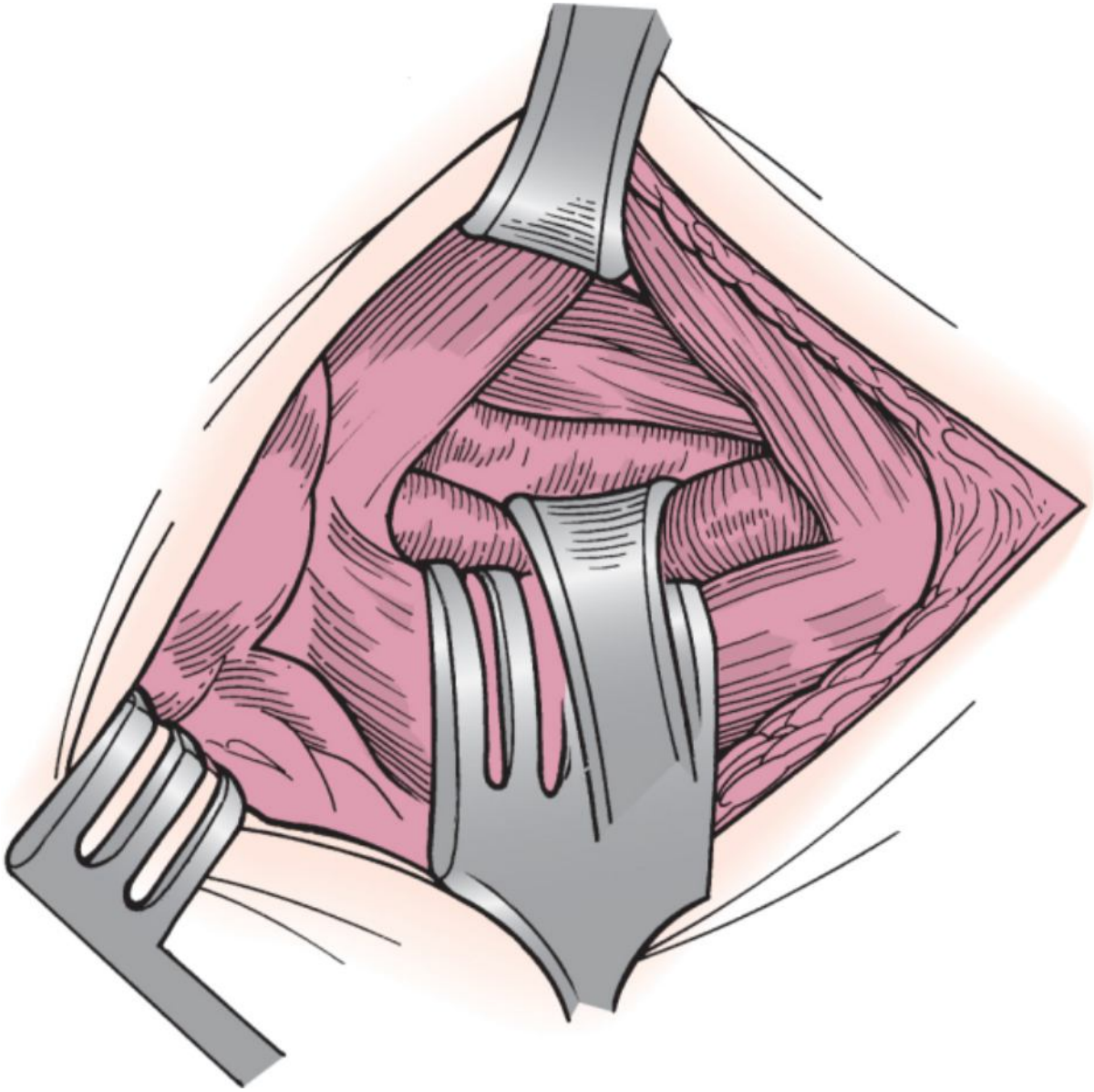

Figure 54-8 The carotid sheath is retracted laterally to expose the prevertebral fascia.

Using a small hemostat, the oblique muscle fibers of the inferior constrictors are separated from the hypopharyngeal mucosa (Fig. 54-9). The muscle fibers are divided with a bayonet-type, insulated bipolar electrocautery as the hemostat is advanced along the plane between the muscles and mucosa. Change from an oblique to a horizontal direction of the muscle fibers marks the change from the inferior constrictor to the cricopharyngeal muscle. These steps are repeated until the entire height of the cricopharyngeus is divided. After the transection has been carried out, a strip of muscle is undermined and excised to prevent recurrence. The wound is then irrigated with normal saline solution and closed in layers.

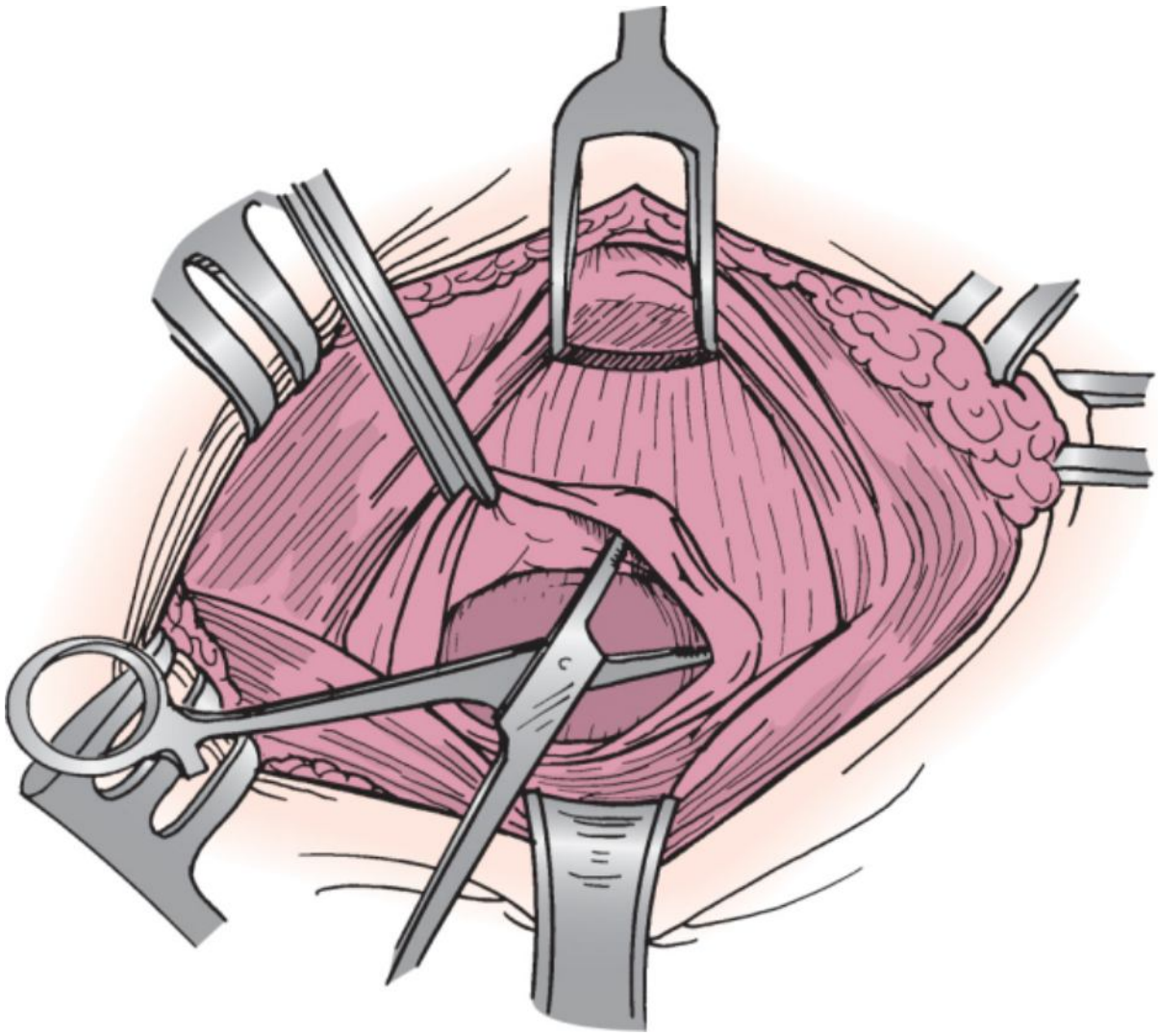

Figure 54-9 Muscle fibers of the upper esophageal sphincter are separated from the hypopharyngeal mucosa in the midline with a hemostat.

If the pharynx is entered inadvertently, a nasogastric tube is introduced and secured to the nose. Any opening into the pharynx is closed with absorbable suture. The wound is then irrigated and closed in layers over a suction drain, which is inserted into the area of the prevertebral fascia between the operative site and the carotid sheath.

Excision of Zenker's Diverticulum

External Approach

Direct laryngoscopy is performed and the contents of the diverticulum aspirated. The mucosal surface is thoroughly examined to rule out squamous cell carcinoma,which occurs in a small percentage of patients with Zenker's diverticulum. Cricopharyngeal myotomy is accomplished as described in the previous section. For most patients a myotomy suffices to alleviate symptoms caused by Zenker's diverticulum, especially when the diverticulum is small. Large pouches, however, are excised to prevent retention of secretions or food and regurgitation on assuming a recumbent position. The diverticulum is dissected from the surrounding tissue as the myotomy is performed (Fig. 54-10). If a diverticulectomy is to be performed, the neck of the diverticulum may be treated in one of two ways. The classic technique is to grasp the tip of the sac with a clamp and excise the sac (Fig. 54-11). Excessive traction on the sac may lead to over-resection of hypopharyngeal mucosa and result in a stricture. A 3-0 or 4-0 gastrointestinal suture is then used to close the mucosa. Alternatively, a stapler may be used to excise the sac and repair the pharyngeal mucosa. The muscles should not be closed over the mucosa. Diverticulopexy was developed to avoid the risks inherent in entering the pharynx. If diverticulopexy is to be carried out, the fundus is inverted and sewn to the prevertebral fascia (Fig. 54-12). The wound is then irrigated and closed in layers over a suction drain, which is inserted into the area of the prevertebral fascia between the operative site and the carotid sheath.

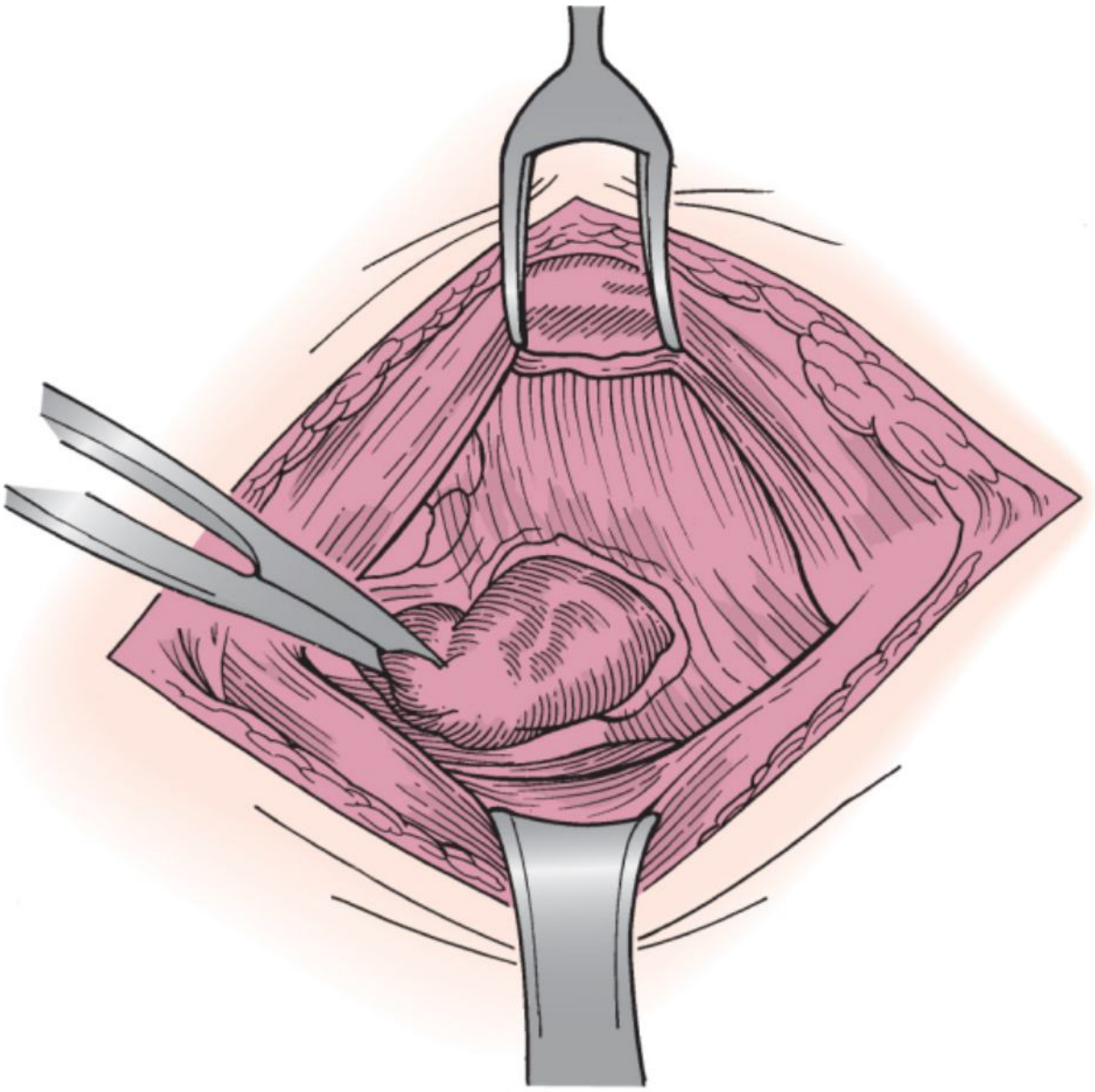

Figure 54-10 The diverticulum is dissected from surrounding tissue.

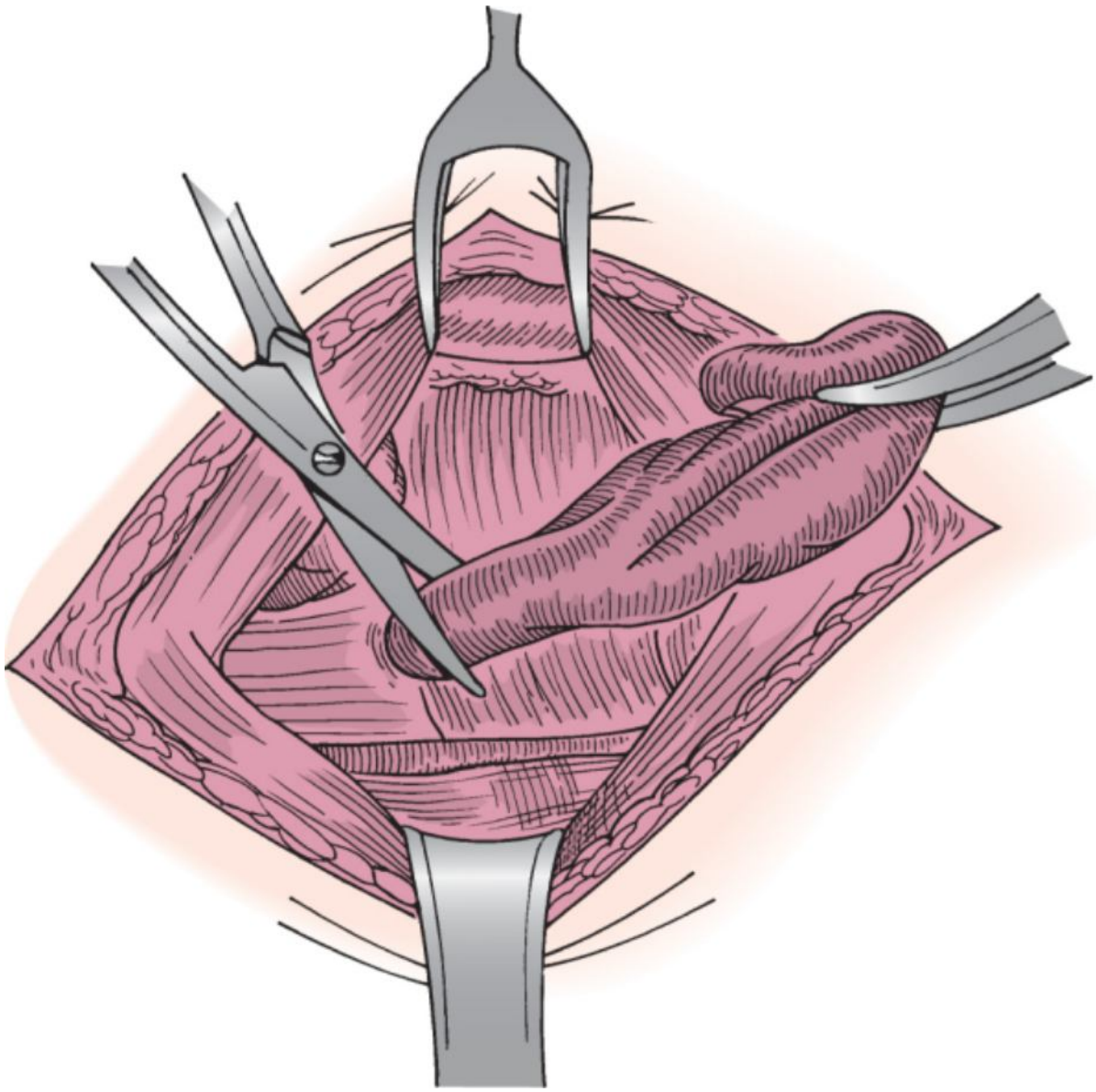

Figure 54-11 Diverticulectomy is performed by excising the sac.

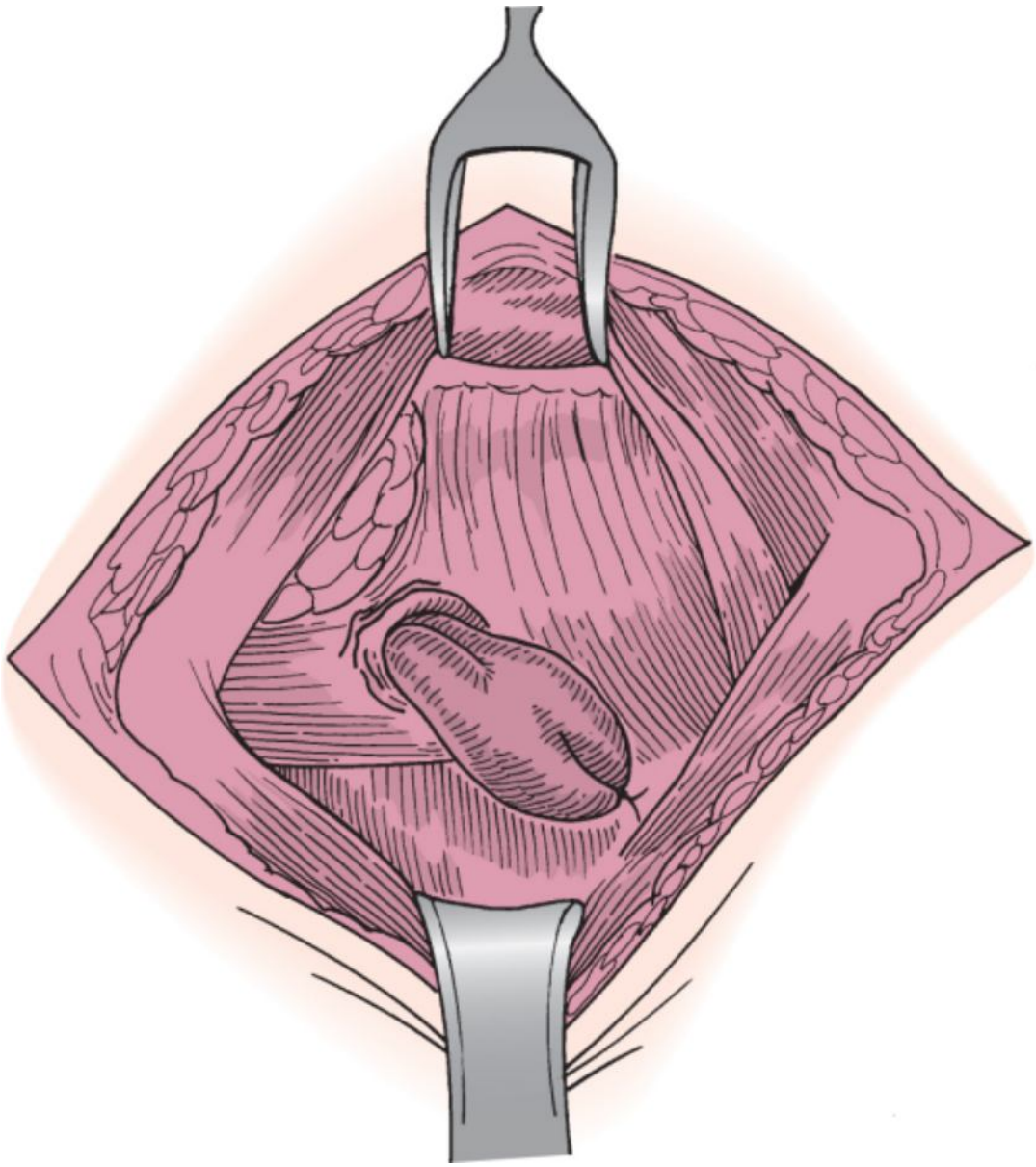

Figure 54-12 Diverticulopexy is performed by sewing the sac to the prevertebral fascia.

Endoscopic Approach

The endoscopic approach for the treatment of Zenker's diverticulum requires a bivalved endoscope, one blade of which is inserted into the cervical esophagus and the other into the diverticulum (Fig. 54-13). The endoscope is suspended. If an endoscopic stapling device is to be used, visualization is achieved with a 0-degree rigid endoscope. The “bar” between the pouch and the esophagus, which consists of the cricopharyngeus muscle, is stabilized by grasping it with alligator forceps or by using endostitches. The Endo-GIA stapler is then used to divide the party wall between the esophagus and diverticulum, with several rows of staples being deposited on each side in the process (Figs. 54-14 and 54-15).[13] A modification of the Endo-GIA stapler as described by Lang and colleagues includes shortening the anvil so that it is as close as 1 mm to the metal clips. This allows the division of the party wall and marsupialization of the diverticulum to a level close to its fundus.[7]

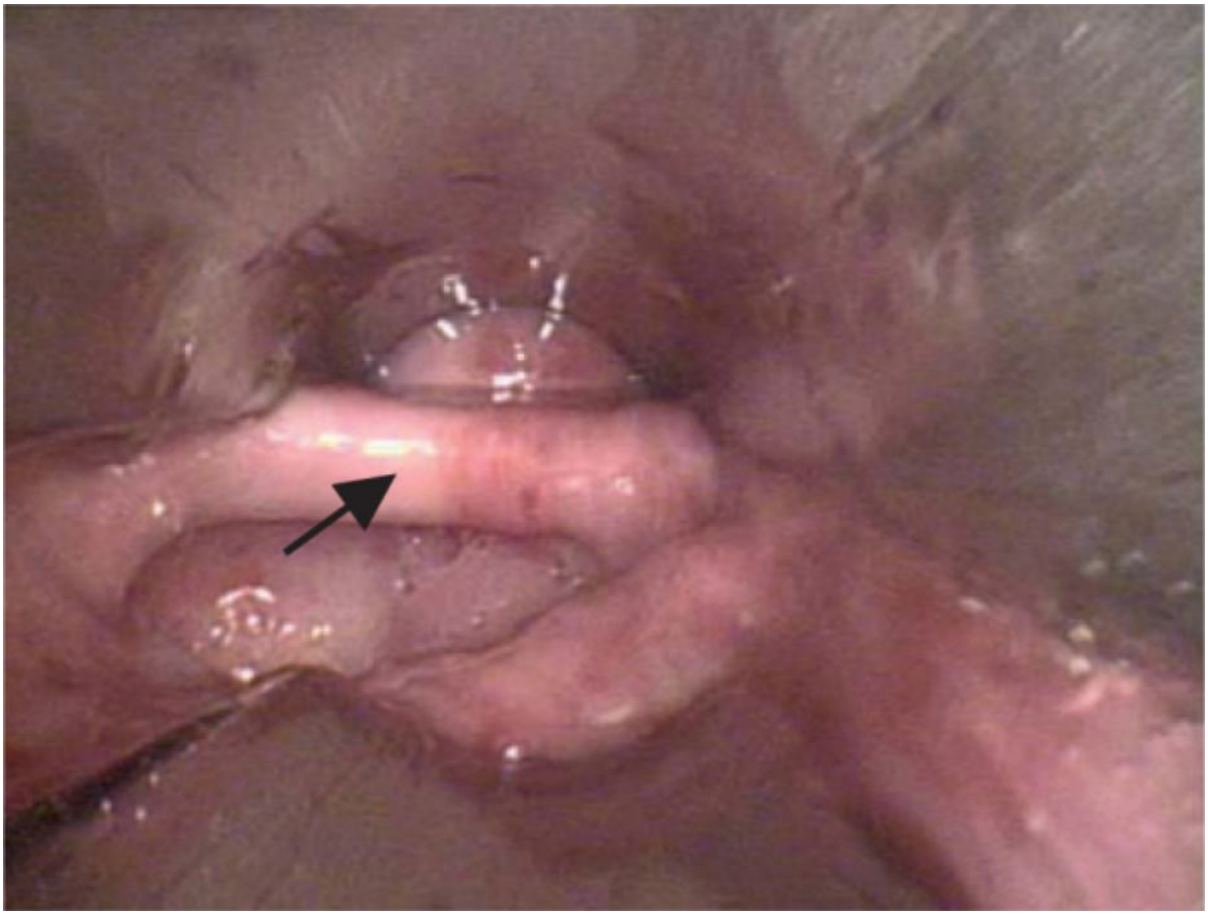

Figure 54-13 Endoscopic view of the "bar" dividing the esophagus and pouch (the esophagus is anterior) *(arrow)*.

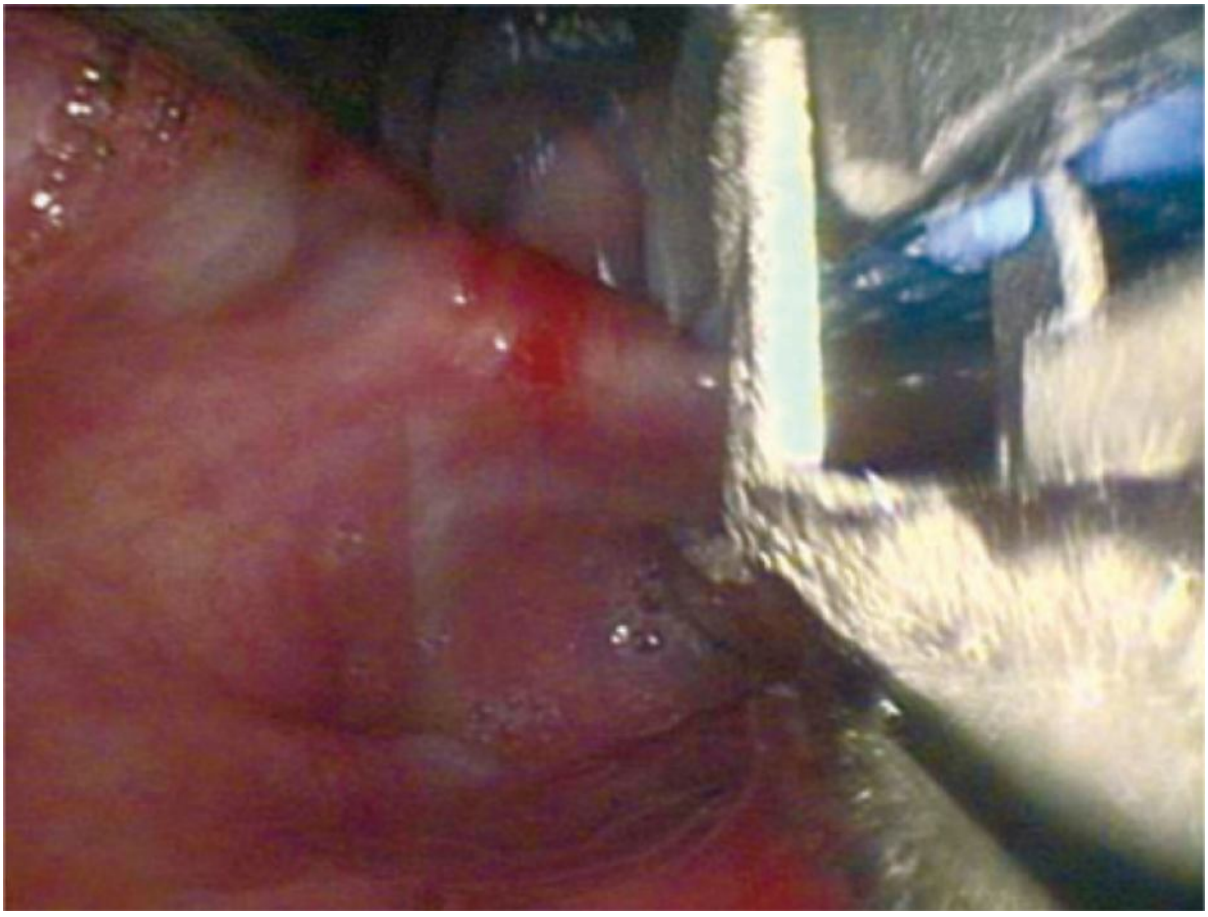

Figure 54-14 The Endo-GIA stapler is positioned on the "bar" between the esophagus and pouch before stabilization with alligator forceps.

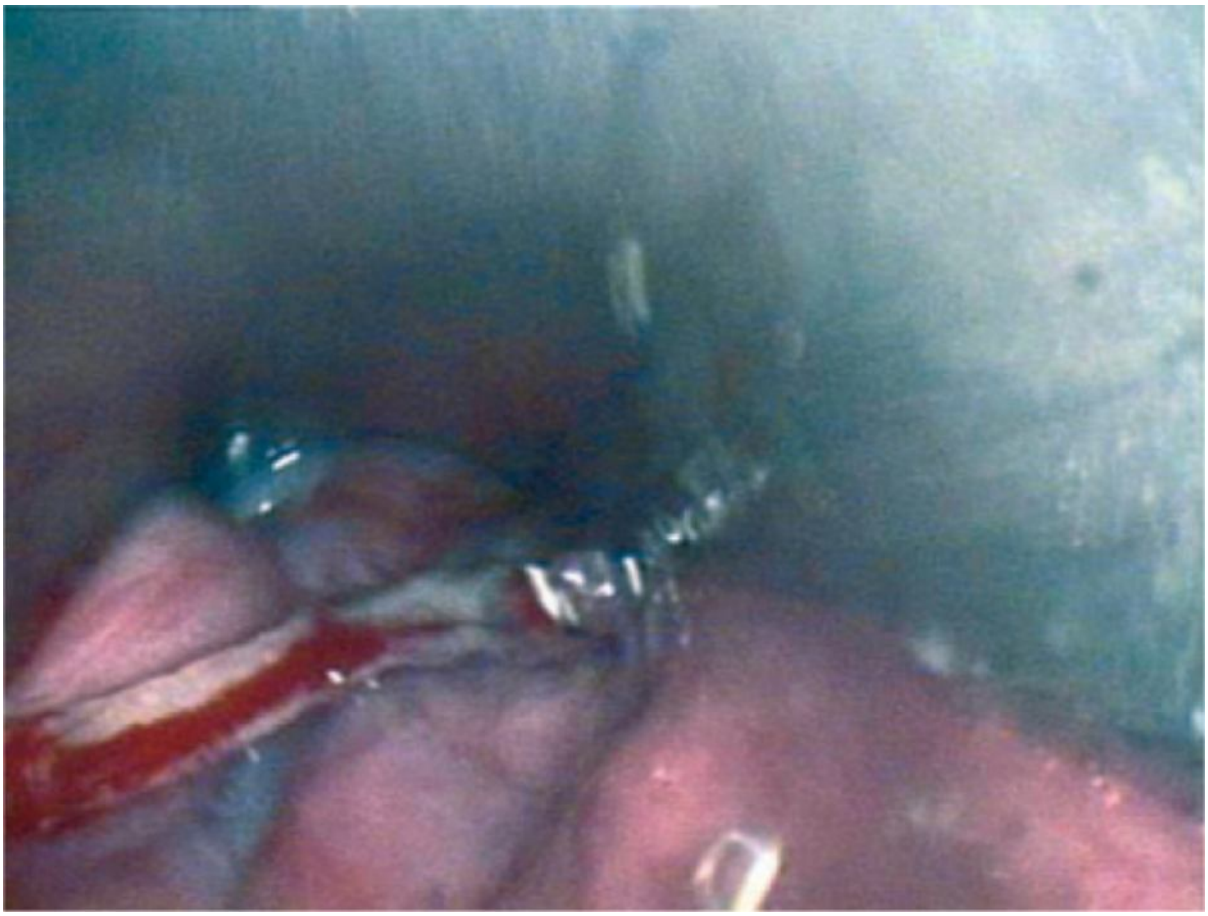

Figure 54-15 The party wall between the esophagus and diverticulum has been divided and the resulting esophagostomy stapled in the process.

POSTOPERATIVE MANAGEMENT

Inadvertent entry into the pharynx is ruled out with a diatrizoate meglumine (Gastrografin) esophagogram. For patients who have undergone primary cricopharyngeal myotomy without pharyngeal entry, a soft diet is started and advanced to solid as tolerated; the patient is usually discharged the day of surgery. If a stapler diverticulectomy is performed, the patient is kept in the hospital overnight and the suction drain is removed on the first postoperative day. Diverticulectomy repaired with traditional suture requires a longer hospital stay with no oral intake (3 to 7 days).

If the pharynx was entered, a nasogastric tube should be passed to decompress the stomach over the first 48 hours and then used for tube feeding. On the fifth day, the tube may be withdrawn after confirming the lack of a leak by a Gastrografin esophagogram. The patient is given a soft diet for 1 week and then advanced to more solid food as tolerated.

If the patient complains of a change in voice, laryngoscopy should be performed to rule out vocal cord paralysis/paresis from injury to the recurrent laryngeal nerve.

PEARLS

- Vocal fold paralysis after cricopharyngeal myotomy is avoidable by incising the muscle fibers posteriorly in the midline.
- Wound dehiscence and a salivary fistula are always possible in patients in whom the pharynx has been entered.

- There are now a variety of techniques available to the surgeon for treating Zenker's diverticulum, and it is prudent to select a technique with which the surgeon is experienced and is best suited to the patient's needs.
- A significant percentage of patients with Zenker's diverticulum will have a second cause of dysphagia, and a thorough history and physical examination along with FEES, barium esophagography, and rigid esophageal endoscopy will help disclose the correct diagnosis.
- The mucosal surface of Zenker's diverticulum should be examined endoscopically to rule out squamous cell carcinoma.

PITFALLS

- Endoscopic techniques for the treatment of Zenker's diverticulum may not be possible in some patients because of anatomic limitations.
- Patients with diffuse neurologic problems, such as Parkinson's disease or ALS, will not benefit from cricopharyngeal myotomy because of diffuse weakness of the pharyngeal muscles.
- Recurrence of symptoms must be prevented by careful patient selection.
- Retraction of thyroid cartilage near the cricothyroid joint at the point where the recurrent laryngeal nerve enters the larynx may lead to paralysis of the vocal fold.
- A salivary fistula is detected by observance of saliva in the drain, erythema and edema of the skin flaps, and drainage of saliva through the incision.[14]