Paparella: Volume III: Head and Neck

Section 2: Disorders of the Head and Neck

Part 5: The Larynx, Trachea, and Esophagus

Chapter 38: Esophageal Disorders

Allen M. Seiden

As head and neck surgeons, most of us are only superficially aware of the wide range of disorders that may affect the esophagus and the various methods of diagnosis and therapy that are currently available. Although the treatment of many of these diseases falls more appropriately within the realm of gastroenterology and thoracic surgery, these patients will often cross specialty lines when seeking medical attention.

Esophageal disorders are among the most common of the gastrointestinal tract, and many patients will have head and neck symptomatology. For example, reflux in some form affects roughly 10 per cent of the population, 80 per cent of whom will have associated dysphagia. Half of these patients will have so-called cricopharyngeal dysphagia and thereby may readily present to an otolaryngologist.

In fact, a thorough understanding of the sequence of events occurring during the course of a normal swallow is necessary so that we may distinguish those factors responsible for its dysfunction. In addition, esophagoscopy is routinely performed, whether to search for malignancy or to extract a foreign body. It behooves us then to be able to recognize any underlying pathologic condition and the implication this holds for further treatment.

The purpose of this chapter is to provide an overview of the esophagus and its maladies, emphasizing those areas that are important from a head and neck surgeon's perspective. Pertinent anatomy and physiology will be reviewed, followed by a discussion of the clinically relevant methods of diagnosis. The diseases that will be covered may be conveniently categorized as those caused by congenital factors, those caused by motility disorders, and those caused by structural disorders that cause obstruction, and they will be approached accordingly.

Anatomy

As it name implies, the esophagus serves as a conduit from pharynx to stomach, but such subtleties of purpose are accomplished with the utmost precision and adaptation. It propels almost a limitless variety of textures, temperature, and volume of food through its 20 to 25 cm, regardless of posture and opposing thoracic and abdominal pressures. It prevents reversal of this flow except when emesis becomes necessary. It maintains such rigorous functions by its specialized musculature and unique neuroanatomic relationships with surrounding structures.

Histologic Features

The entire digestive tract is composed of four basic layers that are modified slightly in different regions to accommodate individual functions. These layers include an inner mucosa, a submucosa, a muscularis externa, and an adventitia. There is no serosal or plural layer covering the esophagus, as compared with more distal areas of the gastrointestinal tract, which are covered by peritoneum. This makes it somewhat more sensitive to mechanical trauma, and anastomotic suture lines become more tenuous.

At rest, the esophageal lumen remains collapsed, with seven to ten longitudinal folds in the mucous membrane, which disappear during dilatation. As is true of any mucosa along the digestive tract, the esophageal mucosa is composed of an epithelial membrane; a supportive connective tissue, the lamina propria; and a thin outer smooth muscle layer or muscularis mucosae. These three layers usually are between 500 and 800 microm.

The epithelial membrane is a tough, nonkeratinizing stratified squamous epithelium that gives way to columnar epithelium at the gastroesophageal junction. This distinction is readily seen as an irregular line between the pale esophageal mucosa and reddened gastric mucosa, which appears so because of the more transparent and superficial columnar cells. This transition is generally 1.5 cm proximal to the true anatomic border with the stomach, and because of the saw-toothed pattern is also referred to as the Z-line.

The lamina propria is a loose areolar connective tissue layer containing collagenous and elastic fibers, blood vessels, and nerve fibers. The muscularis mucosae is somewhat thicker than that found more distally in the digestive tract and in fact is thickest in the lower end of the esophagus. It is composed of smooth muscle oriented longitudinally, which at the level of the cricoid cartilage is continuous with the elastic layer of the pharynx.

The submucosa is a thick layer of dense collagenous connective tissue that is pulled up into the longitudinal folds when the esophagus is at rest. It contains blood vessels, lymphatics, mucous glands, and autonomic fibers and parasympathetic ganglion cells that make up the myenteric plexus of Meissner.

The external muscular coat, or tunica muscularis, has classically been described as having an inner circular layer and an outer longitudinal layer, although this is perhaps an oversimplification. The longitudinal layer is in fact not directly vertical, but spirals around one quarter of the esophageal circumference. The so-called circular layer is not directly horizontal but is angled 10 to 20 degrees. The pattern thus corresponds to a screw-like configuration.

The separation between these two layers is not always distinct. At either end, the longitudinal layer largely blends with the circular layer, which assumes a more horizontal position in the area of the upper and lower portions of the sphincter. Anatomically, neither is a clearly defined sphincter, although the muscular bundles function as such. Scattered between the two layers is the myenteric plexus of Auerbach, composed of parasympathetic ganglion cells and neurons. In addition, this tunica muscularis is composed primarily of striated muscle in its upper third, mixed striated and smooth muscle in its middle third, and smooth muscle in its lower third. The delicate transition between these two muscle groups displays a remarkable degree of coordination and adaptation.

The connective tissue of the muscular layer is continuous with that of the outermost layer, the tunica adventitia. This layer contains both vascular and nervous elements and gives way to surrounding mediastinal structures, but perhaps its most important element is a complex network of elastic fibers. This network is continuous with a system of fibers interspersed within the inner layers of the esophagus. They ensure that the esophagus resumes its normal shape following deglutition and that it can withstand the distention and contortion often necessary to handle a bolus feeding.

Gross Anatomy

The esophagus begins just below the cricoid cartilage at the level of the sixth cervical vertebra. Endoscopically, this is 15 to 20 cm from the upper incisor teeth. It descends vertically along the anterior aspect of the spine and shifts to the left opposite the suprasternal notch. For this reason, incisions for approaching the cervical esophagus are usually made on the left side.

Once it enters the thorax, it is pushed back to the midline by the aorta, opposite the sternal angle and fourth thoracic vertebra. It passes through the posterior mediastinum, continuing to rest on the vertebral column, and is subjected to negative intrathoracic pressure. The aortic arch lies at the junction of the proximal and middle third of the esophagus, and the distal third then passes just behind the heart. At the seventh thoracic vertebra, it once again begins to curve to the left to pass through the esophageal hiatus in the diaphragm at the tenth thoracic vertebra. At this point, the phrenoesophageal ligaments inserts into the circular muscular layer and, in addition, fibers from the right crus of the diaphragm pass around the esophagus to form a sling, further stabilizing its position.

The abdominal portion of the esophagus is usually 2 to 4 cm in length, terminating at the cardioesophageal junction opposite the 11th thoracic vertebra. It lies in the esophageal groove of the left lobe of the liver and is covered by reflections of peritoneum on its anterior and left aspects.

From a more practical standpoint, although the esophagus is already the narrowest portion of the digestive tract, it contains in its length three important anatomic narrowings. They are important to remember, particularly when dealing with foreign body or caustic agent ingestion. The first is at the upper esophageal sphincter, corresponding to the cricopharyngeus muscle and cricoid cartilage. The second corresponds to anterior compression by the aortic arch and left mainstem bronchus, approximately 20 to 25 cm from the upper incisors. The third is at the gastroesophageal junction, measuring 40 to 45 cm from the upper incisor teeth.

Anatomically speaking, it is also worth remembering several areas of inherent weakness along the esophageal wall, particularly within the pharyngoesophageal segment. These naturally are areas in which diverticula may develop. Most worthy of mention is Killian's dehiscence or triangle, at which point the so-called Zenker's diverticulum develops. This is a potential space or weakness between the inferior pharyngeal constrictor muscle and the cricopharyngeus muscle. Remember that the cricopharyngeus muscle forms a muscular sling at the esophageal inlet, attaching to either side of the cricoid cartilage without a midline raphe. The inferior constrictor muscle extends from the oblique line of the thyroid cartilage to insert in the posterior midline pharyngeal raphe, a fibrous band extending from the base

of the skull to which all of the constrictor muscles are attached. Killian's dehiscence then lies just above the cricopharyngeal sling, below the raphe.

Actually, the particular anatomy in this area has been variously described. Some consider the cricopharyngeus muscle to have two parts: (1) the part fundiformis, or transverse portion, which forms the sling; and (2) the pars obliqua, which is continuous with the lower end of the inferior constrictor muscle. Others describe completely separate muscular units, and still others describe the cricopharyngeus muscle as being simply the lower portion of the inferior constrictor muscle. Although autopsy studies do indicate that individual differences may exist, the confusion is perhaps less anatomic and more physiologic, as there has been some disagreement as to the precise configuration of the upper esophageal sphincter. This will be discussed in more detail further on.

A second potentially weak area is also in the posterior midline, below the cricopharyngeus muscle. At the upper end of the esophagus, those longitudinal muscle fibers that do not converge with the circular fibers diverge laterally and anteriorly around the esophagus to insert in a common tendon behind the cricoid cartilage. This area of the upper posterior esophageal wall is covered simply by a layer of circular fibers and is known as Laimer's triangle or the Laimer-Haeckerman area. Although potentially weak, there have apparently been no reports of diverticula developing at this site. This would suggest that other precipitating factors are necessary as well.

Other potentially weak spots are those dehiscences that allow passage of blood vessels and nerves, such as the Killian-Jamieson area, which is a lateral dehiscence between the cricopharyngeus muscle and circular esophageal fibers through which the recurrent laryngeal nerve passes.

The blood supply to the esophagus can essentially be broken up into thirds. The upper third derives its blood supply from the inferior thyroid artery off the thyrocervical trunk from each side. The middle third is supplied by several direct branches from the thoracic aorta. The lower third is supplied by esophageal branches of the left gastric artery, which originates from the celiac trunk off the abdominal aorta.

In a parallel fashion, the upper third drains via the inferior thyroid veins to the thyrocervical trunks and subclavian veins. The middle third drains via venous plexuses along the surface of the esophagus into the hemiazygos vein on the left and the azygos vein on the right, both of which drain into the superior vena cava. The inferior pathway deserves special mention because of its relationship to the portal venous system. Lower esophageal veins are tributaries of the left gastric vein, but they also anastomose with the azygos system. Normally these anastomoses carry small amounts of blood under low pressure. The left gastric vein meanwhile drains to the portal vein, which, in turn, passes through the hepatic circulation and into the inferior vena cava. Valves in this portal system are insignificant or lacking. Therefore, portal hypertension and hepatic cirrhosis lead to shunting upward through the esophageal plexus to the azygos vein and superior vena cava. This increased blood flow and pressure causes distention of these poorly supported anastomoses, with the ultimate development of varicosities.

Lymphatic drainage of the esophagus can also be broken up into thirds. The cervical esophagus drains into the internal jugular chain as well as the paratracheal nodes. The middle third drains into tracheobronchial and posterior mediastinal nodes. The lower third drains into nodes following the left gastric vessels and the celiac nodes, which drain to the large intestinal trunk.

Physiology

A discussion of esophageal function would be incomplete without considering its role in the context of a normal swallow. As mentioned previously, it is imperative for an otolaryngologist to understand the sequence of events necessary for a safe and effective swallow. The average adult swallows 600 time each day: 35 swallows/hour while awake and 6 swallows/hour while asleep. Because the airway and food passage share the same pharyngeal space, coordination must be precise, and this requires a complex neuroregulatory network.

Traditionally, deglutition has been divided into three phases: oral, pharyngeal, and esophageal. The oral phase is preparatory, mixing the food with saliva and forming it into an appropriately sized bolus. This is largely under voluntary control, but it is dependent upon many other functions, such as olfaction, gustation, mastication, and salivary production. Preparation is complete with positioning of the bolus on the middle of the dorsum of the tongue. The tongue is then essentially responsible for delivering the bolus to the oropharynx, which it does by compressing it against the hard palate. A wavelike contraction or stripping wave beginning anteriorly propels the bolus posteriorly. This action is very much dependent on the intrinsic musculature of the tongue and the genioglossus, mylohyoid, styloglossus, and hyoglossus muscles. The importance of maintaining good tongue mobility and control then becomes clear, and its impairment is a primary reason for dysphagia in patients who have had strokes or surgical resection for oral and pharyngeal malignancy.

Passage of the bolus to the oropharynx then begins the pharyngeal phase, which elicits the reflex portion of deglutition. Once initiated, it carries through from start to finish in a stereotyped fashion. Receptors that initiate the reflex appear to be proprioceptive and are scattered over the base of the tongue, tonsillar pillars and fossa, uvula, and posterior pharyngeal wall. In humans, the most sensitive area seems to be the anterior tonsillar pillars. Receptors are also located in the larynx, but they perhaps serve more of a backup or protective function. These receptors are more sensitive to water than to mechanical stimulation and appear to be tied in with laryngeal taste buds.

It is interesting that some stimuli provoke swallowing, whereas others produce coughing and gagging. The mechanism whereby mechanical stimuli can produce these far different responses is not completely understood; however, animal studies seem to indicate that strong stimuli that produce higher neuronal firing rates are more likely to yield a gag response. Weaker stimuli more likely result in swallowing. In fact, electrical stimulation rates of 30 pulses/second seem to be optimal. It is likely that both spatial and temporal patterns of stimulation are important.

Afferent impulses are of course carried via the maxillary division of the trigeminal nerve and the glossopharyngeal and superior laryngeal nerves to converge in the tractus

solitarius of the brain stem. The information is then transmitted to the so-called swallowing center, which is located in the reticular substance of the upper medulla. Actually, there is a left and right center, each controlling ipsilateral deglutitory muscles except for the middle and inferior pharyngeal constrictor muscles, which are contralaterally innervated. There is also extensive communication between these two centers, which ultimately coordinate efferent impulses to appropriate cranial nerve nuclei, including cranial nerves V_3, VII, IX, X, and XII. The sequence of events thus elicited to effect a swallow is essentially fixed, irrespective of the particular type of afferent activation. Other activities, such as chewing, breathing, coughing, or vomiting, are concomitantly inhibited. Higher, or suprabulbar, centers likely have both a facilitative and an inhibitory effect on swallowing, although the specific role and pattern of this influence is not clear.

The first important goal, therefore, in the pharyngeal phase of swallowing is initiation of the swallowing reflex. That being done, there remain three important objectives: (1) propulsion of food through the pharynx into the esophagus, (2) protection of the airway, and (3) prevention of regurgitation into the oral cavity and nasopharynx.

Propulsion of the food bolus is dependent largely on gravity, posterior tongue thrust, and pharyngeal peristalsis. Gravity actually plays a small role, since an upright posture seems to facilitate the passage of liquids but not of more solid material. Tongue thrust generates a pressure of approximately 15 cm of water. Pharyngeal peristalsis begins with apposition of the soft palate with the contracting superior constrictor muscle (Passavant's ridge). The pharyngeal constrictor muscles then fire in overlapping sequence, innervated by the surrounding pharyngeal plexus. The duration of contraction in each constrictor muscle is approximately 0.5 second, and the total duration of pharyngeal activity is approximately 1 second. The pharyngeal peristaltic wave has a peak amplitude of 100 plus or minus 60 mm Hg, whereas the hypopharyngeal peristaltic wave has a peak amplitude of 200 plus or minus 150 mm Hg; this peristaltic wave travels down the pharynx at approximately 15 cm/second, reaching the esophageal inlet in 0.67 second, its speed being unaffected by the liquid or solid nature of the bolus. The bolus is, therefore, usually cleared from the pharynx within 0.5 to 1 second.

The prevention of regurgitation of food into the oral cavity and nasopharynx is accomplished by apposition of the tongue to the palate and the soft palate to the pharyngeal wall. These positions are maintained until the bolus has cleared the hypopharynx.

Protection of the airway is accomplished by competent glottal closure of both the true and false vocal cords. More important, however, is laryngeal elevation. As the deglutitory reflex begins, the suprahyoid muscles contract, drawing the hyoid toward the mandible. The thyrohyoid muscle contracts as well, bringing the larynx up even further. This protects the laryngeal inlet underneath the tongue base. In addition, it causes passive deflection of the epiglottis, which is turned downward 60 degrees below the horizontal to help shield the larynx as it directs the food bolus laterally.

Laryngeal elevation is also important for another reason. It serves to dilate the upper esophageal sphincter to facilitate passage of the bolus into the esophagus. As alluded to previously, the nature of the upper esophageal sphincter has generated a great deal of controversy. There have been conflicting data presented in the literature regarding its anatomic

and physiologic configuration. Anatomically, there is no clearly defined sphincter; however, manometrically there is an intraluminal area of elevated pressure, measuring 3 to 5 cm, centered around the cricopharyngeus muscle. The cricopharyngeal sling or pars fundiformis is only 1 cm wide, so therefore the lower inferior constrictor muscle and upper circular muscle fibers of the esophagus likely contribute. Electromyographic studies have verified that this area remains tonically contracted via vagal stimulation, thereby protecting the airway from refluxed gastric contents and preventing aerophagia.

Although there is some disagreement, it seems clear that during the swallowing reflex, the sphincter relaxes secondary to centrally induced inhibition of vagal impulses. Subsequently, a pressure of 10 mm Hg remains, most likely resulting from the inherent elasticity of the sphincter. This residual pressure is then abolished by anterior and upward displacement of the larynx. The bolus of food now descends into the esophagus ahead of pharyngeal peristalsis. As this peristaltic wave reaches the upper esophageal sphincter, the cricopharyngeus muscle contracts in sequence to carry the wave distally. It then resumes its normal resting, tonically contracted state. The larynx descends and resumes its resting position as well.

This period of upper esophageal sphincter relaxation lasts less than 1 second, and it must be precisely coordinated with pharyngeal contraction. There are, of course, a variety of neurologic, neuromuscular, and postsurgical states that disrupt this relationship and lead to dysphagia. A complete discussion of these is beyond the scope of this chapter, but the reader is encouraged to become familiar with these problems, as well as the implication for medical and surgical therapy.

As the bolus of food descends into the esophagus, the esophageal phase begins. Despite the transition from striated to smooth muscle that occurs at the junction of the upper and middle thirds of the esophagus, the primary peristaltic wave sweeps down the esophagus in a regular, uninterrupted fashion. It travels more slowly in the esophagus, at approximately 3 cm/second, and therefore takes 8 to 12 seconds to conclude at the gastroesophageal junction.

The striated portion receives its innervation from the nucleus ambiguus via somatic efferents of the vagus nerve. The smooth muscle portion, of course, is innervated by autonomic nerves, which form both an extrinsic and intrinsic network. The extrinsic network consists of vagal and sympathetic fibers. Vagal fibers form a plexus over the body of the esophagus, the left vagus curving anteriorly and the right posteriorly. Preganglionic sympathetic fibers synapse in the cervical and thoracic sympathetic chains, and postganglionic branches accompany blood vessels to the esophageal wall.

Most of these extrinsic axons terminate in the myenteric plexus (Auerbach's) and the submucosal plexus (Meissner's), which form the intrinsic network. These intramural neurons are largely responsible for the adaptive capability of the esophagus by allowing the input of afferent stimuli. Thus, the speed and force of peristaltic contractions in this portion are greatly influenced by the nature of the bolus - a large volume precipitates a more forceful contraction, and higher temperatures accelerate its propagation. Esophageal distention, as might occur from residual bolus or reflux material, precipitates secondary peristalsis to clear the material. This is without an oropharyngeal component, and it is not induced by the swallowing reflex

(primary peristalsis)). Spontaneous secondary peristalsis may occur seven to ten times/hour and is important for clearing refluxed gastric acid.

Only one peristaltic wave may exist within the esophagus at a given time. Therefore, since esophageal peristalsis is much slower than peristalsis in the pharynx, during rapid successive swallows, esophageal peristalsis (smooth muscle portion) will be inhibited and will occur only intermittently. Pharyngeal peristalsis, conversely, is complete for each swallow and is mediated by a swallowing control center.

Tertiary contractions are nonperistaltic, simultaneous contractions, which are therefore nonfunctional. They may make up 5 per cent of swallows in normal individuals, but they do increase with age. They are usually mild in amplitude and can be seen as ripplelike effects on a barium swallow. A significant increase in frequency and amplitude leads to one of the spastic motility disorders, which will be discussed further on.

Similar to the upper esophageal sphincter, the lower sphincter is not demarcated anatomically but corresponds to an intraluminal zone of high pressure approximately 3 cm in length. It serves to separate the esophagus, surrounded by negative intrathoracic pressure, from positive intragastric pressure, which, of course, is important in preventing reflux. At rest, the mean sphincter pressure is approximately 10 to 30 mm Hg greater than intragastric pressure.

Unlike the upper esophageal sphincter, however, basal tone of the lower sphincter is not dependent upon tonic neural activity. Therefore, relaxation is not due to central inhibition of ongoing activity but is an active process mediated by the release of an unknown neurotransmitter activating inhibitory neurons. The mechanism whereby basal sphincter tone is maintained is a matter of some debate, but it seems to depend largely upon inherent smooth muscle tone within the circular muscular layer at the gastroesophageal junction. Other factors, however, are believed to contribute to sphincter competence: (1) the angle by which the esophagus enters the stomach, (2) positive intra-abdominal pressure compressing the intra-abdominal portion of the esophagus, (3) hormonal activity (such as gastrin), (4) mucosal rosetta caused by redundant folds of gastric mucosa at the gastroesophageal junction acting as a valve, and (5) diaphragmatic support from the right crus and phrenoesophageal ligament.

In any event, the sphincter relaxes roughly 2 seconds after initiation of the swallowing reflex and remains so for 8 to 10 seconds. As it is reached by the peristaltic wave, it too contracts. Contraction lasts 10 to 15 seconds, followed by a return to normal resting tone.

Diagnostic Evaluation

As in any diagnostic endeavor, a thorough history and physical examination are still the most important tools at a physician's disposal. Symptoms associated with esophageal disease are many, and although no one symptom is diagnostic of a specific disorder, a careful history can provide important clues as to probable cause and thus the appropriate workup. This includes not only specific complaints, such as dysphagia, odynophagia, chest pain, or heartburn, but also the duration and order in which symptoms developed. Association with specific foods, particularly liquids versus solids, is helpful in distinguishing an obstructive problem from a motility problem. Characteristic symptoms will be discussed in greater detail

with associated disorders.

A number of objective tests are then available to evaluate esophageal function (Table 1). It is worth stating at the outset that localization of symptoms by the patient is notoriously unreliable. This is particularly true when the patient points to the sternal notch region and above, which is often associated with lower esophageal pathologic conditions. This underscores the importance of a detailed history and the need to consider the entire swallowing chain, from oral cavity and pharynx to stomach.

Table 1. Diagnostic Evaluation

Imaging Techniques	**Intubation Procedures**
Barium esophagram	Manometry
Cinefluorography	Intraluminal pH studies
Videofluorography	Standard acid reflux test
CT, MRI	Acid infusion (Bernstein test)
Radionuclide scintigraphy	Acid clearance test
	24-hour pH monitoring
	Endoscopy.

Imaging Techniques

Barium Esophagram

Traditionally, the first and foremost objective test to evaluate the possibility of esophageal disease has been the barium swallow. In the typical single-contrast technique, the patient swallows a large amount of barium while in the horizontal position and may even perform a Valsalva maneuver in an attempt to attain maximal esophageal distention. Spot films are then taken under fluoroscopic control. This approach does well for demonstrating gross disease processes that alter the intraluminal or extraluminal contour of the esophagus, such as large carcinomas, significant strictures, and large ulcers. More subtle abnormalities and motility disorders, however, will be missed. Even the presence of a radiographic abnormality does not guarantee an accurate diagnosis. One study reports an incidence of false-negative and false-positive diagnoses for esophageal carcinoma as high as 21 and 37 per cent, respectively.

In an attempt to enhance diagnostic accuracy, the double-contrast technique was developed. This is performed with the patient in the upright position and turned obliquely to the left to avoid overlap with the spinal column. An effervescent agent is swallowed with a high-density barium preparation, and the combination serves to distend the esophagus and coat the epithelium. This provides much better mucosal detail, demonstrating small ulcers and esophagitis as well as early superficial spreading carcinoma.

If one suspects an esophageal perforation, it is best to use a water-soluble contrast material such as meglumine trizoate (Gastrografin). Since it can be absorbed from within the mediastinum, it tends to cause less inflammation. If results are negative, however, it should be followed with a small amount of barium, which remains somewhat more sensitive.

Conversely, patients who are suspected of aspiration should be given thin suspensions of barium, since the water-soluble materials are more irritating to the tracheobronchial tree.

The reflux of barium from the stomach into the esophagus may be indicative of gastroesophageal reflux disease; however, the clinical significance of such an observation is a matter of some debate. Generally, reflux observed in the supine position without provocative maneuvers is probably significant in a patient who is symptomatic. However, reflux induced by provocative steps, such as increasing intra-abdominal pressure, may generate a high rate of false-positive responses. In any event, there are better methods for assessing gastroesophageal reflux (Table 2).

Table 2. Accuracy and Specificity of Esophageal Tests

Indication	Test	Accuracy	Specificity
Gastrointestinal reflux			
	Standard acid reflux test	Excellent	Excellent
	24-hour pH monitoring	Excellent	Very good
	Radioisotope scintigraphy	Excellent	Excellent
	Barium swallow	Poor	Poor
Lower esophageal sphincter competence			
	Esophageal manometry	Excellent	Very good
	Endoscopy	Poor	Poor
Esophageal transport			
	Barium swallow	Excellent	Good
	Radioisotope scintigraphy	Excellent	Good

Peristaltic function
- Esophageal manometry
  - Excellent
    - Excellent
- Endoscopy
  - Poor
    - Poor
- Barium swallow, cineradiography
  - Fair
    - Fair

Mucosal integrity
- Barium swallow, air contrast
  - Good
    - Good
- Endoscopy, biopsy, cytologic examination
  - Excellent
    - Excellent.

Cinefluorography, Videofluorography

One problem with the barium swallow is that it records static pictures, whereas swallowing is very much a dynamic process. Now it is true that these spot films are always taken under fluoroscopic control. However, a cursory fluoroscopic glance is hardly adequate to appreciate the range of movements that occur so rapidly during a reflex swallow. To properly study motility, timing, and coordination, and even very subtle lesions, it is essential to record the movements on film so that they may be reviewed more slowly.

This usually involves cinefluorography, which uses motion picture film, or videofluorography, which uses vide tape. Most hospitals are now using video recording equipment, as the technology has improved and prices have come down. It is more versatile than cinematic recording and has the added advantage of exposing the patient to less radiation.

Swallowing movements should be followed from the voluntary oral phase down to the stomach. Regardless of the patient's symptoms, the entire sequence should be thoroughly evaluated, whereas certain areas may, of course, be highlighted. Each examination may be adapted somewhat to the needs of individual patients. To do this it may be helpful to have the input of a speech pathologist who has previously evaluated the patient if there is difficulty with the oral or pharyngeal phase of swallowing. A variety of barium consistencies should be used to stimulate the patient's clinical problem. A so-called modified barium swallow, using small amounts of varying consistencies of barium as described by Logemann, is particularly useful. A solid bolus, such as a barium tablet or barium-impregnated marshmallow usually elicits more useful information in the case of suspected esophageal disease.

In fact, a number of provocative maneuvers may be used in appropriate situations. For example, Donner and co-workers have shown that 90 per cent of patients who suffer from

reflux esophagitis will demonstrate abnormal esophageal motility when given acid barium.

Carrying this one step further, video recording may be very helpful in instituting swallowing therapy and rehabilitation for a variety of neuromuscular and motility disorders, including those in patients who have suffered a stroke or undergone surgical resection. This would include the application of test feeding techniques, bolus modification, patient positioning, and respiratory maneuvers. Such therapeutic steps may be monitored more accurately and thereby modified accordingly.

Computed Tomography, Magnetic Resonance Imaging

The use of computed tomography (CT) and magnetic resonance imaging (MRI) for evaluating esophageal disease has revolved largely around the pretreatment staging of esophageal carcinoma. There is disagreement, however, as to reliability. The problem lies in detecting tumor invasion through the muscular layer of the esophagus into periesophageal fat and the tracheobronchial tree. Quint and colleagues point out that there is not a complete fat plane between the esophagus and the aorta, which makes aortic invasion very difficult to assess. In addition, because the cartilaginous rings of the trachea are incomplete posteriorly, concavity of the tracheobronchial wall can certainly occur without invasion. In their study, both MRI and CT showed low accuracy (40 and 70 per cent, respectively) in staging these tumors.

In contrast, Donner feels that tissue planes between the esophagus and the descending aorta can be well visualized on CT and therefore claims it is a highly accurate method of staging not only mediastinal extension but also distant metastasis. Exceptional situations would be cachexia, a history of previous esophageal surgery, and previous mediastinal radiation, all of which tend to obliterate normal tissue planes.

Moss and colleagues agree, having separated the CT findings of esophageal carcinoma into the following: stage 1, intraluminal mass with esophageal wall thickening; stage 2, esophageal wall thickening greater than 5 mm; stage 3, spread into adjacent mediastinal structures; and stage 4, distant metastases.

The full utility of MRI of course remains to be seen. Its promise lies in its ability to distinguish among different soft tissues, as well as image in a variety of planes. As experience with this technology improves, so will its usefulness.

Radionuclide Scintigraphy

Radionuclide transit studies were first used to evaluate gastric emptying, but in 1972 esophageal scintigraphy was introduced by Kazem. The potential advantages were clear. This promised to be the first noninvasive quantitative approach to evaluate esophageal transit and thereby esophageal dysmotility.

Roentgenographic contrast studies are of course non-invasive. However, though they are essential for detecting obstructive abnormalities, they are regarded by some as relatively insensitive to subtle motor disturbances. In addition, they do require exposure to radiation and are very difficult to use quantitatively. Manometry measures intraluminal pressures but is

invasive and does not provide direct information about transit or reflux. The other intubation techniques (see Table 1) are also invasive and as such may introduce nonphysiologic artifacts.

That being said, the drawbacks of scintigraphy have revolved around questions of accuracy and reliability. The test works very simply. The patient fasts overnight and is then asked to swallow 15 mL of water mixed with Tc 99m-sulfur colloid in a single swallow while in the supine position. The patient then swallows at 15-second intervals for 10 minutes. Counts are picked up by a gamma camera and processed by computer. The technique may be adapted for specific needs, such as focusing on specific areas of interest or sequentially measuring counts over the pharynx and different areas of the esophagus. In addition, the radioisotope may be bound to specific foods.

The test provides no anatomic information but allows measurement of transit and emptying times. It may also be used to measure gastroesophageal reflux, which will be discussed in more detail later in this chapter. Because it is a quantitative study, it may be used effectively to gauge response to therapy.

Despite being rapidly performed, easily quantifiable, and noninvasive, its validity as a screening test for esophageal motility disorders has been questioned. Richter and associates have shown that radionuclide transit time may actually be independent of the amplitude and duration of esophageal contraction and lower esophageal sphincter pressure provided that contractions are peristaltic and the sphincter does relax. Therefore, patients who suffer from nutcracker esophagus, for example, may have normal radionuclide transit measurements.

In fact, nonspecific motility disorders seem to be relatively common and may occasionally be missed by any of the tests that measure motility. Caestecker and colleagues compared the sensitivity rates of radionuclide scintigraphy, manometry, and radiology for detecting esophageal dysmotility and found them to be 75, 83, and 30 per cent, respectively. Each test essentially measures a somewhat different aspect of esophageal function, and in that sense they are complementary. Which one is most appropriate will depend on the specific clinical situation, and this will be discussed in greater detail in subsequent sections.

Intubation Procedures

Manometry

Although manometry was long felt to be strictly a research tool, it has not become a well-established clinical technique for measuring esophageal motility. Intraluminal pressure measurements are made by means of a series of pressure-sensitive transducers, which may be one of two types. The one most commonly used over the last 2 decades has been a continuously perfused, triple-lumen catheter system attached proximally to transducers that measure changes in pressure transmitted through the water column. Three lateral openings in the catheter are spaced 5 cm apart and axially oriented 120 degrees. More recently, tiny transducers have been developed that are small enough to swallow. They are attached to a catheter assembly that measures the pressure directly within the esophageal lumen.

This pressure information, which is recorded simultaneously from different levels within the esophagus, is recorded on moving graph paper. One can therefore record not only

the amplitude and duration of contractions but also the rate of peristalsis by knowing the distance between recording points and the paper speed.

Typically, the examiner concentrates on three areas: the lower esophageal sphincter, the body of the esophagus, and the upper esophageal sphincter. The catheter assembly is passed transorally or transnasally down the stomach and is then gradually withdrawn. Location is determined by changes in pressure readings (eg, gastric pressure increases and esophageal pressure decreases with inspiration). Lower esophageal sphincter pressure is measured at rest (15 to 30 cm of water) and subsequent to swallowing. The esophageal body should show very little spontaneous activity, but it is evaluated during swallowing. The optimal swallowing rate is once every 30 seconds, since anything faster will tend to inhibit esophageal peristalsis (see section on physiology). One notes the peristaltic or nonperistaltic nature of contractions, the amplitude and duration, and whether or not spontaneous contractions occur. Upper esophageal sphincter pressure is measured at rest (40 to 80 cm water) and in response to swallowing; in addition, however, an attempt is made to measure the coordination of relaxation with pharyngeal contraction.

By gleaning this information, we can learn a great deal about esophageal physiology as well as the pathophysiology of esophageal dysmotility. In fact, this test may be diagnostic of a number of esophageal motility disorders. However, as pointed out by Hurwitz and colleagues, one needs to consider this information with a certain amount of circumspection. Variations in equipment as well as technique exist among laboratories, which will influence results and makes standardization and statistical comparisons difficult (Table 3).

Table 3. Variables Affecting Manometric Pressure Recordings

Variables Associated with Perfused Open-type Catheter
- Catheter diameter
- Catheter length
- Infusion rate
- Mechanical factors
  - Type of pump
  - Inherent system drag
  - Elasticity of tubing

Other Variables
- Spatial variables
- Respiratory variation
- Artifacts
- Dry versus wet swallows
- Intraobserver and interobserver variables
- Drugs, foods, hormones, and emotions.

An area in which this is perhaps most pronounced, and one that is of particular concern to the otolaryngologist, is the upper esophageal sphincter. Cricopharyngeal spasm, achalasia, or incoordination has been considered to be the possible cause of a number of swallowing disorders, including globus hystericus. Although a cricopharyngeal "bar" may be demonstrated radiographically, this cause has been difficult to confirm manometrically. In addition to the aforementioned variables, this area is inherently very difficult to measure

because it is not radially symmetric. This is largely because of the laryngeal cartilages and, in fact, manometric measurements in postlaryngectomy patients show this asymmetry to have disappeared. Therefore, depending upon the orientation of the catheter ports or transducers, pressure measurements will vary.

A second problem is that the pharynx and upper sphincter are not stationary during swallowing but move up and down. Pressure measurements will thus reflect different anatomic segments.

To enhance the accuracy of this technique, McConnell and co-workers and others have combined the use of manometry and fluoroscopy, an approach that McConnell has labeled manofluorography. There seems to be some merit in this approach, and we await further studies.

The conclusion is that at this point, manometry is probably best for evaluating dysmotility of the esophageal body and lower esophageal sphincter, recognizing the aforementioned limitations. It is best used in conjunction with other tests and certainly with clinical correlation.

Intraluminal pH Studies

One of the most common questions that needs to be answered when evaluating esophageal function is the competency of the lower esophageal sphincter and the presence or lack of reflux. Symptoms tend to be non-specific, can be misleading, and usually implicate a number of causes. Simply measuring lower esophageal sphincter pressure manometrically is inadequate, since very often it will fall within the normal range. By the same token, although pressures less than 10 mm Hg are highly suggestive, reduced pressure does not necessarily correlate with symptomatic reflux. Further testing, which traditionally has involved the intraluminal measurement of pH will be necessary.

A certain amount of reflux is of course inevitable, occurring 2 per cent of the time while the patient is upright and 0.3 per cent of the time while the patient is supine, reflux being defined as a drop in pH to less than 4. The normal esophagus will generally rid itself of this acid material via primary or secondary peristalsis. Gastric pH may vary from 1 to 7; however, esophageal pH is usually maintained at 6 to 7.

A number of approaches have been devised for detecting the presence of significant reflux disease. The standard acid reflux test is considered by many to be the definitive diagnostic test. It has also been referred to as the Tuttle test. A pH electrode is placed 4 to 5 cm above the lower esophageal sphincter, and the patient undergoes a series of provocative maneuvers, such as changes in position, leg elevation, or Valsalva maneuver to increase intra-abdominal pressure. A pressure belt capable of insufflation worn around the upper abdomen has been described for this purpose, increasing intra-abdominal pressure to 15 mm Hg, which a normal sphincter should be able to handle. This may be done using basal acid secretion, but more often follows the infusion of 300 mL of 0.1 N hydrochloric acid into the stomach. A drop in pH to less than 4 is of course indicative of reflux.

The acid infusion or Bernstein test is an attempt to reproduce the patient's symptoms by infusing 0.1 N hydrochloric acid directly into the esophagus. It does not necessarily require the measurement of intraesophageal pH but relies on subjective responses. With the patient in the sitting position, a nasogastric tube is passed into the distal esophagus and acid is instilled at 5 mL/minute. As the patient begins to feel burning discomfort, the acid is changed to normal saline, which should provide relief. The test is discontinued after 20 minutes if no symptoms are produced.

Heartburn generally is associated with epigastric and retrosternal discomfort, which may be referred to the back when severe and to the left arm when very severe. This test should reproduce the patient's epigastric and retrosternal symptoms, but it is less reliable for referred pain. Pain reproduction will occur in 80 per cent of the patients with severe symptoms but in only 25 per cent or less of patients who have more minor symptoms merely suggestive of heartburn. The test is particularly useful in patients with atypical chest pain when the cause is not clearly esophageal or cardiac. Approximately 10 to 15 per cent of asymptomatic subjects may experience pain during the test, which is clinically insignificant.

The acid clearance test measures the patient's ability to bring esophageal pH back to normal following an acid infusion. A bolus of 15 mL of 0.1 N hydrochloric acid is instilled into the proximal esophagus with the patient supine, and the patient is asked to swallow every 30 seconds. The pH will initially fall to 1, but should rise to 6 within 10 to 13 swallows. Prolonged clearance indicates the presence of esophagitis. More specifically, 70 per cent of patients with ulcerative esophagitis will demonstrate prolonged clearance, as opposed to 36 per cent of patients with reflux but not ulceration. False-positive results may occur in patients with severe motility disorders who in fact do not have reflux esophagitis but will require more time to clear the acid bolus.

Twenty-four-hour pH monitoring gives the most comprehensive and perhaps the most physiologic information. A pH electrode is passed through the nose into the esophagus to 5 cm above the lower sphincter. The monitoring equipment is portable so that the patient may go about an ordinary routine, at the same time recording whenever symptoms are experienced. Normal subjects will demonstrate a total reflux time of not more than 1 hour in a 24-hour period. Symptomatic patients will experience reflux more often with more prolonged clearance times, and this will be reflected in the pH values recorded.

The advantage of this test is that it may verify symptomatic reflux during the course of normal physiologic activity without the need for provocation. It may also indicate whether symptoms such as chronic coughing, hoarseness, chest pain, or wheezing are associated with reflux. The disadvantage, of course, is the inconvenience to the patient.

Endoscopy

Esophagoscopy is routinely performed by both gastroenterologists and otolaryngologists, the former preferring flexible fiberoptic instruments, and the latter preferring rigid fiberscopes. Each has its advantages, with flexible equipment providing better optics and allowing air insufflation for distention, whereas rigid telescopes allow better facility for instrumentation. Any physician treating esophageal disease should be thoroughly familiar with endoscopic technique.

The diagnostic indications for esophagoscopy are a matter of some debate (Table 4). Some would argue that it adds little to a good radiologic examination, whereas others argue it should be the primary diagnostic modality. It certainly is the best method for assessing mucosal integrity, inflammation, and malignancy. The presence of a lesion detected by other means requires direct visualization and probable biopsy. Patients with persistent symptoms, such as dysphagia, whose cause has not been clarified by other means will probably require endoscopic examination.

Table 4. Indications for Esophagoscopy (Diagnostic)

Evaluation of symptoms
- Dysphagia
- Odynophagia
- Heartburn
- Hemorrhage
- Chest pain

Evaluation of Esophageal Diseases
- Esophagitis
- Caustic burn
- Achalasia
- Scleroderma
- Diffuse spasm
- Neoplasia

Evaluation of Radiologic Findings
- Diverticula
- Varices
- Stenosis
- Mucosal abnormalities
- Extrinsic compression
- Hiatal hernia

Postoperative Evaluation
- Anastomosis
- Recurrence
- Bleeding
- Dysphagia.

Once established, many disorders will require endoscopy for full evaluation. This is true of the congenital and obstructive disorders, descriptions of which follow. Disorders that have been associated with malignant degeneration, such as Barrett's epithelium, lye stricture, achalasia, or the Plummer-Vinson syndrome, should be considered for regular endoscopic follow-up and biopsy.

Motility disorders, however, lend themselves poorly to endoscopic assessment, since factors such as peristaltic contractions and sphincter competence are not easily visualized. Exceptions would be if complications were developing, such as reflux esophagitis and

stricture secondary to scleroderma, or if the clinical picture were atypical.

Specific Disorders

Congenital Disorders

Embryology

Congenital anomalies of the esophagus generally are recognized during the first few days of life because of associated respiratory or feeding difficulties. Occasionally, however, more subtle abnormalities are picked up in later childhood. Accurate diagnosis is important and will usually require surgery for correction.

The pharynx and esophagus, as well as the trachea, derive as ventral outpouchings from the primitive foregut. This separation into an anterior respiratory and posterior digestive tract is complex, and its failure is responsible for the majority of congenital anomalies found.

The esophagus develops from that part of the foregut extending from the primitive lung buds to a fusiform dilatation that ultimately will become the stomach. Between the third and fourth weeks of gestation, a tubular esophagus is recognizable, but short. It gradually will elongate as the fetus grows and differentiation occurs.

During the fourth to fifth week of gestation, a tracheoesophageal septum forms from paired lateral indentations that meet in the midline to separate the trachea and esophagus. This separation is ordinarily complete by the end of the second month.

The primitive esophagus contains an internal tube of entoderm, which will differentiate into the stratified squamous epithelial lining. Surrounding mesodermal elements differentiate into muscle and connective tissue to form the lamina propria, submucosa, and muscular layers. The inner circular muscular layer differentiates at the sixth week of gestation, and the longitudinal layer differentiates at the eighth week. In between, neuroblasts extending from the neural crest along the vagi reach the esophagus to form a plexus. The muscularis mucosae appears at the tenth week of gestation.

Major anomalies therefore occur between the third and sixth weeks of gestation. Usually they result from faulty separation of the esophagus from the trachea, yielding various types of fistulous tracts, or from faulty recannulation of the esophageal lumen, yielding various degrees of closure from web formation and stricture to atresia.

Atresias and Tracheoesophageal Fistulas

Esophageal atresia, with or without tracheoesophageal fistula, is by far the most common congenital anomaly affecting the esophagus, occurring once in every 3000 live births, with there being no apparent genetic predisposition. A number of variations are recognized and have been classified by different alphabetic and numeric schemes, but they are probably best remembered anatomically; in other words, whether a fistula is present and whether it is proximal or distal to the atresia.

The most common manifestations of this anomaly are depicted, in descending order of frequency. Esophageal atresia with a distal tracheoesophageal fistula occurs most often, making up to 86 per cent of these cases. Symptoms are predictable, characterized by fluids returning through the nose and mouth during feeding as it pools in the esophageal atretic sac. The infant will subsequently tend to aspirate. Between feedings, there is an inordinate amount of salivary pooling and drooling, whereas the stomach becomes distended with air. Gastric contents may occasionally be regurgitated through the fistula, and pneumonia is a frequent problem.

The second most common type, occurring much less often, is atresia without an associated fistula (7.7 per cent). It is characterized by similar symptoms, and the distinction generally needs to be made radiographically. An isolated tracheoesophageal fistula without atresia, the third most common type (4.2 per cent), is also referred to as the H-type of anomaly. In this case, the fistula may occur anywhere between the cricoid and tracheal bifurcation. Because the continuity of both the trachea and the esophagus is intact, symptoms may be minimal and the problem may go undetected for months or years. When regurgitation causes flow through the fistula into the trachea, the infant experiences paroxysmal coughing, choking, and perhaps subsequent pneumonia. Usually these episodes are associated with feeding. Recurrent pneumonia unresponsive to antibiotic therapy is typical and should suggest an underlying tracheoesophageal fistula.

When the proximal esophageal atretic segment communicates with the trachea, feeding immediately results in severe choking and coughing. If the distal segment is not connected to the airway, there is no gas in the abdomen. Otherwise, both the last two types will also result in excessive pharyngeal secretions.

The diagnosis should be suspected whenever a soft rubber catheter cannot be passed to the stomach, which is a useful diagnostic maneuver in the newborn and should be considered if there appears to be excessive mucus or signs of respiratory distress. Occasionally, obstruction will be detected at approximately 11 cm from the upper incisor teeth, but more often, the catheter will become coiled in the upper pouch. This will then need to be confirmed radiographically, which is pathognomonic for esophageal atresia. In addition, plain x-ray films will reveal the presence or lack of air in the stomach, which will indicate whether or not a distal fistula is present.

Definitive diagnosis is made with barium contrast, which needs to be performed very cautiously with small amounts (0.5 mL) of thin barium. Water-soluble contrast materials are contraindicated because of their propensity for causing pneumonitis.

Evaluation should include a search for associated anomalies, which are not uncommon in these patients. In a study of 1058 cases, Holder and colleagues found 48 per cent to have other congenital abnormalities, most commonly cardiovascular or gastrointestinal, which occurred in 19 and 13 to 27 per cent, respectively. Ultimate prognosis depends more on these associated anomalies, which are more often responsible for mortality and severe morbidity.

Survival from esophageal atresia and tracheoesophageal fistula has improved substantially with earlier detection and better perioperative care. Overall survival is reported to be between 75 and 82 per cent. Therapy initially is supportive, providing adequate nutrition

and preventing respiratory complications. This includes regular aspiration of the esophageal pouch, aggressive treatment of aspiration and pneumonitis, and usually prophylactic antibiotics. A gastrostomy will be necessary, which may be done during the definitive repair, or sooner if repair needs to be delayed. This not only provides a route for nutritional support but also allows gastric decompression, which helps prevent further pulmonary compromise and helps to avoid gastric reflux.

Surgical correction should usually be scheduled as soon as possible, which will depend upon the infant's general condition. Definitive repair includes division and closure of the fistula and direct esophageal anastomosis. Complications include anastomotic leaks, strictures, recurrent fistulas, and injury to the recurrent nerves. In 15 per cent of infants with esophageal atresia, the gap between the segments will be too long to close primarily, and an interposition graft will need to be used. Colon grafts are usually favored, in either a primary or delayed fashion.

Duplications

Duplication of the gastrointestinal tract in general is relatively rare, the ileum being the most common site, and the esophagus the second most common. Esophageal duplications compose approximately 15 per cent of those cases reported. The cause is uncertain, but several theories have been postulated. The most plausible would seem to be a failure of recannulation of the esophageal lumen. During development, the esophagus is essentially a solid tube. The lumen is formed by the development of scattered vacuoles that ultimately coalesce. If one or more of these vacuoles fails to join the esophageal lumen, it may remain as an isolated cyst attached to the esophageal wall. There is a reported incidence of associated vertebral anomalies.

Esophageal duplications may be tubular and communicate with the esophageal lumen or cystic and not communicating, with the cystic variety being much more common. They tend to occur most commonly in the lower third of the esophagus, which makes up 60 per cent versus 23 per cent in the upper third, and 17 per cent in the middle third.

Symptoms usually occur early; however, in milder cases, lesions may go undetected until being discovered incidentally. The usual presenting complaints are secondary to cyst expansion and compression of surrounding structures. Thus, for example, esophageal compression can lead to dysphagia and regurgitation, and tracheal compression can lead to dyspnea, stridor, cough, and pneumonia. Rupture of the cyst may lead to acute mediastinitis.

The diagnosis is usually suggested radiographically, revealing either a cystic mass in the posterior mediastinum or a barium-filled tubular duplication. Endoscopy will usually reveal only extraesophageal compression and is thus not diagnostic. The recommended treatment is surgical excision, which should also provide the definitive diagnosis. Palmer, cited by Pelot, has suggested three necessary criteria to make the diagnosis of an esophageal duplication, including definite attachment to the esophagus, the presence of a representative epithelial layer. The lining of these cysts may be squamous, respiratory, intestinal, or mixed. They may also be lined by gastric epithelium, which may lead to ulceration, bleeding, and perforation.

Dysphagia Lusoria

Congenital vessels of the great vessels of the superior mediastinum may cause esophageal compression, resulting in dysphagia. Most common is an aberrant right subclavian artery causing what has been termed *dysphagia lusoria.* The vessel arises from the descending aortic arch and passes obliquely upward toward the right arm, passing behind the esophagus instead of ventral to the trachea. Much less often, in 15 per cent of cases, it passes between the esophagus and the trachea. It represents an abnormality of the fourth branchial arch.

The incidence of this abnormality is relatively low, occurring in roughly 1 per cent of the population, although the incidence is somewhat higher in patients with congenital heart disease. Compressive symptoms will develop in 10 per cent of these patients, very often not until adulthood. It has been pointed out that symptoms are more likely to occur when the left and right carotid arteries originate either from a common trunk or very close together so that the trachea and esophagus are prevented from deviating forward between the two carotid arteries to escape the posterior compression.

If the compression is severe, the infant may display dysphagia from birth, with regurgitation and aspiration. However, more often symptoms present later as the diet begins to include solid food. In addition, the infant is apt to show signs of tracheal compression as well, with episodes of stridor and cyanosis that improve when the neck is hyperextended. In adults, symptoms, if present, are usually characterized by mild dysphagia with a sticking sensation at the level of the xiphoid process. They tend to present rather late, primarily as atherosclerosis develops along with progressive esophageal dysmotility.

The diagnosis is best made radiographically, particularly with a barium study. This will show an indentation of the posterior esophageal wall near the level of the aortic arch at the third and fourth thoracic vertebrae. Endoscopy may be helpful in excluding other causes and sometimes may reveal the compression to be pulsatile. Pressure with the tip of the endoscope may then obliterate the right radial pulse. Angiographic confirmation is usually not necessary unless surgery is required.

Treatment is usually supportive, particularly in adults. However, if the problem interferes with nutrition and is causing significant aspiration, surgical correction may be necessary. The surgery consists of simply ligating the aberrant subclavian artery provided that collateral circulation is adequate. Otherwise, a reanastomosis to the ascending aorta is necessary.

Chalasia

Chalasia is characterized essentially by incompetence or persistent relaxation of the lower esophageal sphincter without the presence of a hiatal hernia, thus resulting in persistent reflux. Otherwise healthy infants will regurgitate small amounts of food or formula and, in fact, one fourth of infants showed esophageal reflux after a barium meal. Therefore, some have called chalasia in infants simply an accentuation of a normal phenomenon rather than a specific disease entity.

The cause has been attributed to incomplete development of the lower esophageal sphincter and gastroesophageal barrier, including ancillary factors such as the angle of His and the pooling of gastric fluids, which increases gastric pressure.

Clinically, infants show persistent regurgitation and vomiting beginning within the first week of life; it is usually worse in the supine position. In an upright position, the infant will more often feed normally. Poor weight gain as well as dehydration may occur in severe cases.

Once again, a barium study is diagnostic, showing free reflux of barium into a larger than normal esophagus up to the pharynx when the infant is laid supine. No other associated gastroesophageal abnormalities should be present. If motility studies are performed, they will show a patulous lower sphincter.

The problem usually disappears as the child grows older, after approximately 6 weeks. Until then, the infant will need to be fed sitting up and will need to be kept in that position most of the time. Thickening the feedings may prove helpful, but only rarely is parenteral nutrition necessary. Significant reflux needs to be treated aggressively, however, to avoid serious complications such as esophagitis and stricture. The problem has also been linked to respiratory arrest and sudden infant death syndrome.

Motility Disorders

As stated earlier in the section on esophageal physiology, swallowing is a complex process requiring delicate precision among as many as 26 muscles and five cranial nerves. In addition to bolus preparation and airway protection, proper timing and strength of muscle contraction are necessary to generate effective peristalsis. The main purpose of the esophagus then is to transport food from the pharynx to the stomach and prevent its return, except in appropriate circumstances.

Failure to carry out this purpose can usually be attributed to one of two problems: (1) a motility disorder, characterized by a failure of peristalsis or sphincter function, or both; or (2) a structural defect, causing obstruction. This is obviously an oversimplification, but it presents a convenient way for looking at esophageal disorders. Motor disturbances of the esophagus tend to produce intermittent dysphagia, which progresses very slowly or may not progress at all. The difficulty is seen with both liquids and solids right from the outset. Obstructive disorders, in contrast, tend to produce constant and progressive symptoms, at first with solids and then progressing to liquids as the condition worsens.

Most motility disorders are associated with a decrease in the number of actual propagative waves, as well as either an increase or decrease in contraction amplitude. Actually, aperistalsis alone will not necessarily produce symptoms sufficient to warrant medical intervention. The esophagus is capable of emptying by gravity. If, however, there is associated lower esophageal sphincter dysfunction or high-pressure contractions, symptoms result.

Typical symptoms associated with motility disorders are dysphagia, odynophagia, and chest pain. Odynophagia and episodic chest pain are often associated with high-pressure, nonperistaltic contractions. Such contractions may be typical of primary motor disorders but

may also occur secondary to esophageal obstruction. In addition, a neoplastic or inflammatory process may invade the submucosal plexus, resulting in severe pain. So-called esophageal colic, characterized by crushing anterior chest pain, and usually associated with diffuse esophageal spasm, simulates the pain of myocardial infarction, and should be considered in the differential diagnosis of chest pain.

The point once again is that symptoms tend to be non-specific, and a thorough history is crucial to elucidate the probable cause. In general, it is probably best to approach any patient with esophageal symptoms with the assumption that there is an underlying structural lesion until proved otherwise.

Basically, motility disorders can be broken down into primary disorders, which affect just the esophagus, and secondary disorders, in which esophageal dysfunction is part of a systemic illness (Table 5).

Table 5. Esophageal Motility Disorders

Primary Disorders	Secondary Disorders
Achalasia	Scleroderma
Diffuse esophageal spasm	Other connective tissue disorders
Nutcracker esophagus	Diabetes mellitus
Nonspecific dysfunction	Alcoholism
Hypertensive lower esophageal sphincter	Central nervous system disorders
Diminished amplitude of esophageal peristalsis	Presbyesophagus
	Chagas' disease.

Achalasia

Achalasia is a generalized motor disorder of the esophagus that, as a primary disease, has an incidence of 1/100.000 population. Classically, it is characterized by a hypertensive lower esophageal sphincter (resting tone), incomplete relaxation of the sphincter with swallowing, and lack of esophageal peristalsis. This, of course, results in delayed esophageal emptying, which leads to dilatation and elongation

The cause of this disorder remains unknown. There seems to be no clear genetic predisposition, nor is there any predilection associated with sex or race. Histologic findings have been consistent in showing loss or complete lack of ganglion cells in Auerbach's plexus. This is particularly prominent in the distal portion and lower esophageal sphincter. Degenerative changes are also noted in the preganglionic axons and their respective brain stem nuclei, presumably secondary to retrograde degeneration.

The majority of patients are in the third and fourth decades of life. The most prevalent symptom is dysphagia, secondary to the functional obstruction of the lower esophageal sphincter and aperistalsis. It is usually painless but may vary in severity. Patients may, in fact, point to the level of the xiphoid process as the place they feel food becoming stuck or hanging up. A number of maneuvers - such as the Valsalva maneuver, throwing the head and shoulders back, or washing the food down with liquids - may facilitate passage. Halitosis may

be a significant problem, as may regurgitation, because of retained material in the dilatated esophagus. Regurgitation may lead to aspiration, and a picture of chronic bronchitis or recurrent pneumonia may ensure. In considerably advanced cases, there have been reports of severe esophageal dilatation causing laryngeal and tracheal compression, resulting in upper airway obstruction. The esophagus must then be emergently evacuated.

Odynophagia or chest pain are unusual in achalasia, since generally they are associated with forceful contractions that typically do not occur. However, a variant of achalasia has been described in which the esophagus demonstrates high-amplitude, nonperistaltic contractions. It has been referred to as vigorous achalasia and tends to occur in the earlier stages of disease when the esophagus is only partially dilatated. In fact, it may represent a transitional state between diffuse esophageal spasm and achalasia, as some have considered the two to be opposite extremes of the same basic process.

Diagnosis is best made with a barium study and manometry. Simply a plain chest x-ray film, in advanced cases, may reveal an air-fluid level in a dilatated esophagus, noted posterior and superior to the cardiac silhouette. Lack of a gastric air bubble may also be noted, as may concomitant acute or chronic lung disease. A barium esophagram will generally show no peristalsis, although tertiary contractions may be present to a variable degree. Depending upon severity and duration of disease, varying amounts of dilatation will be noted. In severe cases, marked dilatation tapering to a "bird's beak" deformity at the lower esophageal sphincter is classic.

Although these x-ray findings are very suggestive for achalasia, a similar picture may be noted in other conditions, such as carcinoma at the gastroesophageal junction or scleroderma with lower esophageal stricture. Therefore, manometry is a helpful adjunct and should be performed. Several characteristic findings should be noted. The lower esophageal sphincter will demonstrate a high resting pressure, usually more than 35 mm Hg above gastric pressure. The sphincter will fail to relax or will only partially relax with swallowing. Examination of the esophageal body will show a lack of peristalsis, although spontaneous, nonpropagative contractions may occasionally be noted. Similar nonfunctional contractions of low amplitude will occur in response to swallowing. It is pathognomonic if the resting esophageal pressure is greater than gastric pressure.

Another characteristic finding in achalasia is a hypersensitivity of the esophageal body and lower sphincter to cholinergic stimuli. Methacholine given subcutaneously will precipitate vigorous contractions, occasionally associated with chest pain. It is believed to be secondary to denervation hypersensitivity, and it is not observed in other conditions, such as carcinoma.

Endoscopy should be performed in all patients believed to have achalasia to rule out the possibility of an underlying malignancy or even a benign stricture. During this examination, significant inflammation and ulceration may be seen secondary to stasis. The lower esophageal sphincter will appear narrowed, but it should allow passage of the endoscope with only minor resistance. Tumors extending from the gastric fundus have been associated with a clinical picture of achalasia, and therefore this area should be carefully evaluated. In addition, patients with long-standing achalasia have a higher incidence of esophageal carcinoma developing, which is found in 5 to 10 per cent of cases, presumably because of chronic stasis, inflammation, and subsequent metaplasia. As one might imagine,

the diagnosis is often delayed, and consequently these patients tend to have a poorer prognosis. Therefore, those who have long-standing disease need to be followed and evaluated regularly.

Therapy for achalasia has centered primarily around decreasing lower esophageal sphincter pressure. The deficiency in peristalsis has been found to be largely irreversible. Some conservative measures may be helpful, such as a softer diet or keeping the head of the bed elevated, but usually an invasive procedure will be necessary.

There are virtually three ways to open the lower esophageal sphincter: (1) pharmacologically, (2) with dilation, and (3) by myotomy. Quite a number of pharmacologic preparations are known to decrease sphincter pressure and have been studied with respect of achalasia. They include hormones such as glucagon, nitrates, anticholinergics, beta-adrenergic agonists, and calcium antagonists. They have essentially proved to be ineffective or produce too many side effects. They are usually recommended only for patients who have failed, have refused, or are not suitable for dilation or surgery.

For dilation to be effective, it usually requires forceful tearing of the esophageal musculature. Therefore although mercury-tipped bougienage may provide some palliation, it is often not enough. Vigorous pneumatic dilation, with a fluoroscopically placed insufflated bag dilator, is the method of choice. There is disagreement as to the range of pressure and duration of insufflation used, but good results have been reported after one or two dilations. It is more effective if performed when disease is less advanced. The most significant complication is esophageal rupture or perforation, reportedly occurring in 2 to 6 per cent of cases.

An alternative is surgical myotomy, referred to as the Heller procedure. It involves a thoracotomy and sectioning of the longitudinal and circular muscles of the esophagus from the gastroesophageal junction to a proximal distance of 7 to 10 cm. Although certainly more invasive than dilation, it is the preferred technique by many because approximately 85 per cent of patients show significant improvement. The major complication is postoperative gastroesophageal reflux, occurring in as many as 30 per cent of patients, so now many recommend that an antireflux procedure be performed at the time of the myotomy. The issue of whether dilation or myotomy is most appropriately the initial treatment remains controversial, and much depends upon the experience of the individual gastroenterologist and surgeon.

Diffuse Esophageal Spasm

The syndrome of diffuse esophageal spasm is characterized by repetitive, high-amplitude contractions of the smooth muscle portion of the esophagus. The striated portion functions normally, and the lower esophageal sphincter essentially relaxes appropriately. These contractions occur intermittently and are of variable severity but are associated with more than 30 per cent of swallows.

Once again, the cause is not known. Histopathologically, muscular hypertrophy is noted along with lymphocytic infiltration of Auerbach's plexus and degenerative changes in esophageal branches of the vagus nerve. Hypersensitivity to cholinergic stimulation has been

observed in these patients, and therefore denervation may be part of the problem, as seen with achalasia. For this reason, as well as the recognized syndrome of vigorous achalasia and reports of diffuse esophageal spasm progressing to achalasia, the two conditions are considered by many to be related, with diffuse esophageal spasm manifesting in early stage of achalasia. However, the evidence for this relationship as yet is insufficient.

Patients tend to be older than 50 years of age, although symptoms may present at any age. The most prevalent symptom is chest pain or esophageal colic, which may occur with or without swallowing. It may vary in its severity but is substernal and may radiate to the arms and back. This makes it very difficult to distinguish from cardiac pain, which must be considered in the differential diagnosis. However, there should be no exertional pain, and the occasional relationship with swallowing and the secondary dysphagia are helpful clinical clues. Patients will complain of intermittent dysphagia and odynophagia.

The typical patient tends to be somewhat anxious and in fact, symptoms can be triggered by emotional stress. Other precipitating factors include hot or cold liquids and gastroesophageal reflux.

When evaluating patients with diffuse esophageal spasm, a routine chest x-ray film and cardiac evaluation will usually be necessary. Evaluation of the esophagus will include at least a barium study and manometry. Characteristic radiographic findings have been labeled as corkscrew esophagus, or beading or curling of the barium column. In other words, the nonperistaltic, simultaneous contractions produce segmentations of the barium column in the distal two thirds of the esophagus. An acid barium swallow may be helpful in eliciting this picture. The esophagus generally is not dilated and, in fact, may appear diffusely narrowed, but it should ultimately empty. Radionuclide scintigraphy may be useful in demonstrating prolonged transit time (more than 20 seconds), along with fragmentation of the radioactive bolus.

Manometry will demonstrate prolonged, high-amplitude nonperistaltic or tertiary contractions following a swallow and occurring spontaneously. Some normal peristalsis may remain in the proximal striated portion and occasionally in the distal portion of the esophagus. Both upper and lower esophageal sphincter function should be normal, although elevated lower esophageal sphincter resting pressure with incomplete relaxation may occasionally be found.

Since symptoms in this disorder tend to be intermittent, diagnostic studies may sometimes be normal. This has generated interest in provocative testing - for example, with anticholinergic agents and edrophonium. It is important to point out, however, that abnormal findings may occasionally be found in asymptomatic subjects, both with and without provocation. It is, therefore, very important to correlate objective findings with clinical history.

Treatment of diffuse esophageal spasm should begin by reassuring the patient that he or she is not about to suffer a heart attack. Avoiding factors that trigger symptoms is important, particularly during meals. This may include treating any associated gastroesophageal reflux.

Medical therapy has revolved largely around nitrates, including sublingual nitroglycerin for acute attacks and longer acting preparations for prophylaxis. Improvement has been reported clinically and manometrically. Calcium channel blockers (nifedipine) and anticholinergic agents have also been used with limited success.

Dilation has been used, both bougienage and pneumatic dilation, but it seems to help only in patients who actually are found to have lower esophageal sphincter dysfunction. It improves the dysphagia temporarily but does little to alleviate the chest pain.

When conservative measures have failed, and the patient's symptoms are incapacitating, a surgical myotomy could be considered. The surgery involved is extensive, since the myotomy usually needs to include the entire smooth muscle portion of the esophagus, which extends up to the level of the aortic arch. This is not as effective as a myotomy done for achalasia, nor is therapy for diffuse esophageal spasm generally as satisfactory as that for achalasia.

Nutcracker Esophagus

The disorder of nutcracker esophagus has been described as the most common motility disorder seen in patients evaluated for noncardiac chest pain and has been labeled nutcracker esophagus by Benjamin and colleagues. Like diffuse esophageal spasm, it is associated with substernal chest pain and dysphagia, which are secondary to high-amplitude esophageal contractions. It differs, however, in that the contractions remain peristaltic.

Like other primary esophageal motility disorders, it cause is unclear, as is its relationship to other manifestations of esophageal dysmotility. It has been associated with gastroesophageal reflux, as well as an increased incidence of psychiatric problems. Therapeutic success usually requires addressing these underlying problems. Otherwise, management of the painful contractions is similar to that for diffuse esophageal spasm. Initial medical therapy favors one of the calcium channel blockers, with surgery reserved only as a last resort.

Many patients may present with dysphagia and even chest pain and demonstrate nonspecific manometric patterns. Isolated abnormalities of the lower esophageal sphincter and esophageal body may be noted. Very often they will be discovered incidentally, without producing significant symptoms. Most of these nonspecific motility disorders have not been clearly defined, and treatment will therefore need to be individualized.

Connective Tissue Disorders

A number of connective tissue disorders can affect esophageal motility, including scleroderma, systemic lupus erythematosus, dermatomyositis, polymyositis, and Raynaud's disease. For the most part, esophageal dysmotility as a presenting symptom is unusual, but it can cause significant functional impairment during the later stages of disease.

Polymyositis, a degenerative disease involving primarily striated muscle, and dermatomyositis, involving the skin as well, affects the esophagus in approximately 30 per cent of cases. However, dysmotility is observed primarily in the pharynx and upper striated

portion of the esophagus.

The remaining connective tissue disorders produce a pattern very similar to that found in scleroderma, which is the most prevalent disorder. It has been estimated that 80 per cent of patients with scleroderma will have head and neck manifestations, and 52 per cent report some type of dysphagia as a primary symptom. Postmortem studies have found histologic involvement of the esophagus in 74 per cent of cases.

Scleroderma, also known as progressive systemic sclerosis, causes a generalized small vessel arteritis with excessive collagen production and deposition. In the esophagus, smooth muscle atrophy and collagen deposition in the submucosa have been found. The proximal striated portion usually remains normal. The result is a decreased amplitude of contractions with loss of coordinated peristalsis and a decrease in lower esophageal sphincter resting pressure. Raynaud's phenomenon, which occurs in 80 to 90 per cent of patients with scleroderma and may be seen in other connective tissue diseases as well, is closely correlated with the presence of esophageal involvement.

Despite the distal aperistalsis, patients usually remain asymptomatic, as the esophagus empties with gravity. Because of the decreased lower esophageal sphincter pressure, however, they are prone to significant reflux. In addition, the lack of peristalsis disrupts the ability of esophagus to clear the acid, making these patients highly susceptible to esophagitis and its complications, which include ulceration, bleeding, and stricture that may involve one third to one half of the esophagus and is found in as many as 48 per cent of patients. This compares with an incidence of 11 per cent in patients with idiopathic reflux. Dysphagia, particularly in response to solid food, will then become an increasing problem.

Evaluation of these patients should begin with a barium study. This will show diminished peristalsis or tertiary contractions of the distal two thirds of the esophagus, with a patulous lower sphincter. The esophagus may be variably dilated. In more advanced disease, stricture formation may be present and is usually long and severe. This may cause proximal esophageal distention.

The progression of disease is highly variable and does not correlate with duration. Therefore, varying patterns of esophageal motility will be evident, particularly when performing manometry. Classically, however, these patients will demonstrate aperistalsis of the distal two thirds of the esophagus and decreased lower esophageal sphincter pressure. Studies measuring distal esophageal pH will be consistent with reflux.

Endoscopy is valuable to assess esophagitis and its complications. The possibility of malignancy will need to be excluded.

Endoscopy is valuable to assess esophagitis and its complications. The possibility of malignancy will need to be excluded.

Therapy in these patients is usually directed toward prevent gastroesophageal reflux and its complications. This includes patients who may not be symptomatic but who demonstrate hypotension of the lower sphincter. Patients with scleroderma should therefore be thoroughly evaluated to detect esophageal involvement. Aggressive antireflux maneuvers,

which will be discussed later in this chapter, as well as antacids and H_2 antagonists, should be recommended. Metoclopramide (Reglan) may be helpful by increasing lower esophageal sphincter pressure and gastric emptying. Once stricture has occurred, it is usually best managed conservatively with dilation.

Other Secondary Disorders

Actually, quite a number of diseases have been associated with esophageal dysmotility. Table 5 is by no means all-inclusive, and quite a variety of classification schemes is possible.

Both diabetes and alcoholism can produce a peripheral neuropathy resulting in disordered esophageal peristalsis. In fact, 80 per cent of diabetic individuals with autonomic neuropathy will have abnormal esophageal motility. For the most part, however, these cases tend to remain asymptomatic.

Certainly central nervous system diseases, including the degenerative and demyelinating diseases, as well as vascular lesions, can lead to disruption of esophageal motility. Although symptoms in these cases tend to be more pronounced in the pharynx and striated portion of the esophagus, abnormalities in the distal smooth muscle portion may become significant as well.

Presbyesophagus refers to those abnormalities of esophageal motility seen with aging. Elderly patients will tend to show varying degrees of reduced peristalsis, a greater frequency of tertiary contractions, and occasional failure of lower esophageal sphincter relaxation. These findings may be associated with incoordination of the upper esophageal sphincter. Symptoms are variable, but many of these patients tend to be asymptomatic. Histopathologically, partial denervation with a decrease in ganglion cells in Auerbach's plexus has been detected.

Chagas' disease is a systemic infection caused by the parasite *Trypanosoma cruzi*. It is endemic to South America and may involve multiple organ systems. When the esophagus is involved, it presents a picture virtually indistinguishable from achalasia, with the same manometric findings. Histologically, ganglion cell destruction of Auerbach's plexus is evident.

Patients with Chagas' disease complain primarily of dysphagia, without associated pain. It may be differentiated from achalasia by a history of exposure to endemic areas and evidence of other organ involvement, for example, megacolon, megaureter, and bronchiectasis. Since diagnosis is usually made when there is no longer active infection, treatment usually must be symptomatic, and for the esophageal manifestations it is the same as that for achalasia.

Structural Disorders

As alluded to previously, a dysfunction can usually be made by history whether a dysphagic patient suffers from a motor abnormality or structural problem of the esophagus. A mechanical obstruction will tend to produce dysphagia with solids, which may progress to include liquids; it is therefore important to obtain a detailed history regarding any dietary adjustments the patient may have made. If a history of dysphagia with liquids and solids was preceded by difficulty with solid food only, this would suggest progressive obstruction rather

than a motility disorder.

Specific information regarding food consistency will indicate the severity of the dysphagia. A history of whether the problem has been constant, intermittent, or progressive will direct the physician toward an accurate diagnosis.

A distinction needs to be made among reflux, regurgitation, and vomiting. *Reflux* refers to the flow of material from the stomach to the esophagus, whereas *regurgitation* describes the effortless passage of material from the esophagus into the pharynx or oral cavity. Patients may confuse both with *vomiting,* which, of course, is the forceful retching of gastric contents. Making this distinction can provide important clues. For example, if a patient suffers from regurgitation that tastes bitter or burns, its source is very likely the stomach, which indicates gastroesophageal incompetence and obviates the presence of a severe esophageal stricture. Regurgitation simply of mucus or undigested food suggests obstruction or a diverticulum. Also important would be the timing with meals and association with position.

Esophageal colic is characteristic of a motility disorder, but chest pain may also occur secondary to obstructive lesions. Food impaction upon a stricture as well as gastroesophageal reflux may produce substernal chest pain that radiates to the neck, jaw, ear, and both arms and therefore may also simulate myocardial ischemia. Retrosternal pain that is constant but is exacerbated by swallowing solid food, hot liquids, alcohol, or fruit juice suggests ulceration, usually secondary to reflux. Pain occurring within 10 seconds after swallowing solid food and lasting until the bolus is either regurgitated or eventually passed suggests a severe stricture. In a malignant stricture, this pain tends to dissipate as the lesion progresses, secondary to destruction of mucosal receptors and impaired peristalsis.

Table 6 lists structural problems that may involve the esophagus, to which the remainder of this chapter will be devoted. Again, it is by no means all-inclusive, but it does include the more common disorders with which the otolaryngologist may be confronted.

Table 6. Structural Disorders

Extrinsic compression
Webs and rings
Diverticula
Caustic agent ingestion
Reflux esophagitis and stricture
Hiatal hernia
Neoplasms.

Extrinsic Compression

Because the esophagus remains mobile throughout most of its length, it can withstand significant displacement and still perform its intended function. It is not uncommon to note distortion of the barium column secondary to extraluminal compression of some sort. It is uncommon, however, for such compression to be symptomatic. Therefore, it is important that the clinician be wary of such findings and rule out other possibilities before making a

diagnosis.

Nevertheless, there are a variety of cervical and mediastinal lesions that can cause esophageal compression to the point of dysphagia. In the neck and thyroid and parathyroid glands, enlargement may impinge upon the esophageal lumen. In the chest, the most common cause is cardiomegaly or an enlarged, tortuous aorta. Once again, these findings are readily detected on a barium esophagram, but only in the extreme case will they cause obstruction. An enlarged left atrium or left ventricle, usually secondary to mitral stenosis, may lead to dysphagia. Reports of large aortic aneurysms producing dysphagia have also appeared in the literature.

Mediastinal tumors or metastases to mediastinal lymph nodes may cause esophageal compression. Inflammatory processes involving mediastinal lymph nodes are becoming less common but may occasionally compress and fix the esophagus. The cause is no longer likely to be tuberculosis but rather fungal infections, such as histoplasmosis, or sarcoid.

Another source of compression that deserves particular mention is cervical osteophytes, which are rather common in the aging population and easily blamed as a cause of dysphagia. The incidence of asympatomatic hypertrophic osteophytes of the anterior cervical spine is estimated at 20 to 30 per cent. Saffouri and Ward looked at 116 patients with significant osteophytes and found only 6 per cent with dysphagia, so the incidence of these being symptomatic is quite low. They have been recognized as a possible cause of dysphagia since the early part of the 20th century, and there are now approximately 70 to 80 cases reported in the world literature.

There are a variety of conditions that may cause cervical osteophytes, but the one that seems most often associated with dysphagia is known as diffuse idiopathic skeletal hyperostosis. This is presumably because the bony abnormalities in this disorder tend to reach a larger size. The osteogenic proliferation is probably secondary to skeletal stress, although the precise cause is not known. The spaces between the fourth and fifth and the fifth and sixth cervical vertebrae seem to be most often involved.

The mechanism whereby dysphagia is produced would seem to be direct mechanical compression of the esophagus. This is supported by the fact that symptoms tend to result when encroachment occurs at points of fixation, that is, at the cricoid cartilage and diaphragm. In addition, thoracic osteophytes are less likely to cause dysphagia because the thoracic esophagus is so mobile. Other investigators claim that the mechanical obstacle alone is not sufficient, and additional factors must occur to cause decompensation, such as muscle weakness or trauma.

It is likely that pressure from the osteophytes causes irritation and periesophagitis, which contributes to the dysphagia and odynophagia. This periesophagitis can cause the symptoms to fluctuate, which may, in turn, become transient or progressive. Other than dysphagia, patients may complain of a foreign body sensation, referred otalgia, and neck stiffness.

A lateral neck x-ray film will clearly show cervical osteophytes, and a barium study will reveal narrowing of the cervical esophagus. Again it must be emphasized, however, that

the mere presence of spurs is not pathognomonic and may simply be coincident with symptoms rather than cause them. Zerhouni and colleagues have suggested that objective evidence of impaired function should be demonstrable on cineradiography to definitively make the diagnosis. In other words, patients with asymptomatic osteophytes should maintain normal mobility of the barium column. Symptomatic patients should show local fixation or decreased laryngeal elevation.

Endoscopy may also be helpful in that a bulge may be noted along the posterior wall, and palpation with the endoscope may reveal the bony nature of the lesion. It is risky though because the bony obstruction will impede passage of the endoscope and thinning of the esophageal wall secondary to pressure and inflammation will increase the chance for perforation.

Treatment should initially be conservative, including reassurance, dietary adjustments, and sedation. Antibiotics and anti-inflammatory drugs may relieve the periesophageal inflammation. Those patients with persistent dysphagia that does not respond to medical therapy may be candidates for surgical excision. This is best accomplished through an anterolateral extrapharyngeal approach, although a transoropharyngeal and posterolateral extrapharyngeal approach have been described for superiorly located osteophytes.

Webs and Rings

Esophageal webs and rings are terms that are often used interchangeably to denote a bandlike esophageal narrowing. However, this is actually incorrect. A web is a thin membrane projecting into the esophageal lumen that is covered with squamous epithelium. It may be concentric or eccentric, single (more common) or multiple, and may occur anywhere in the esophagus. Conversely, rings, specifically mucosal rings, occur at the gastroesophageal junction with squamous epithelium on the upper surface and columnar epithelium on the lower surface. These are otherwise known as Schatzki's rings.

As radiographic techniques have improved, so too has the diagnosis of these lesions, very often in asymptomatic individuals. Detection must therefore be correlated with symptoms to determine clinical significance.

These lesions may be conveniently classified based on location, which is discussed in the following sections.

Upper Esophageal Web. Located primarily along the postcricoid area or anterior aspect of the cervical esophagus, upper esophageal webs have traditionally been associated with the Plummer-Vinson syndrome (otherwise known as the Paterson-Kelly or Paterson-Brown-Kelly syndrome). Typically, these patients are of Scandinavian descent and suffer from iron deficiency anemia, dysphagia, achlorhydria, atrophic gastritis, and hiatal hernia. There is an increased incidence of postcricoid or cervical esophageal carcinoma. Ninety per cent of patients are women, usually between the ages of 20 and 50 years. The incidence of cervical webs in this syndrome has been reported to be 86 per cent.

The cause of the Plummer-Vinson syndrome is unclear. Although a number of theories have been postulated, iron deficiency anemia seems to be most significant. The anemia

generally precedes dysphagia and web formation. In fact, the dysphagia is more likely due to secondarily induced mucosal atrophy and disturbance of afferent stimulation than to the cervical web. This is supported by the fact that the dysphagia has been noted to improve following iron therapy, without the need for dilation.

The dysphagia associated with the Plummer-Vinson syndrome tends to vary in severity. It may be intermittent or progressive, but it usually occurs with solid food and not with liquids. Patients will complain that food gets hung up above the sternal notch and may feel as though they are about to choke. Rarely do they complain of pain. Weakness, weight loss, and generally poor health are not uncommon as this disease progresses. Further findings in advanced cases include pallor; brittle and spoon-shaped nails; cheilosis; a red, smooth, atrophic tongue; and friable mucous membranes. Splenomegaly may be present, and patients tend to be edentulous.

Radiographic detection of a cervical esophageal web requires an attentive barium study and can easily be missed unless the radiologist has been cautioned to look for it. It is best seen in the lateral projection with the esophagus distended, and it generally appears as a thin projection into the barium column from the anterior esophageal wall at the level of the fifth to seventh cervical vertebrae. It must be distinguished from indentations caused by the cricopharyngeus muscle or the postcricoid venous plexus. This differentiation is facilitated by video recording, since a web will not change its shape during the course of the study.

Endoscopically, these webs are usually located around 15 cm from the upper incisors. They may be easily missed and in fact may be inadvertently ruptured. They tend to be thin and eccentric. Endoscopic evaluation is essential in these patients to rule out associated malignancy and assess the caliber of the web and any associated inflammation.

Treatment centers around iron therapy and dilation, which in the majority of cases will relieve the dysphagia. Because of the greater likelihood of upper esophageal carcinoma developing, which is reported to be as high as 50 per cent, patients with the Plummer-Vinson syndrome require regular long-term follow-up.

Middle Esophageal Web. These webs tend to be much less common and may occur anywhere along the middle length of the esophagus. They have been classified as congenital or acquired, the acquired form usually occurring secondary to an inflammatory process, for example, from ingestion of a caustic agent or from reflux esophagitis.

When congenital, these webs tend to be asymptomatic until the infant begins eating solid food, at which time he or she may begin to spit up. This usually takes place between 5 and 10 months of age. Adults may remain asymptomatic or may complain of intermittent dysphagia to solid foods. At times they may be able to localize the site of obstruction.

The lesion may be diagnosed radiographically, but again, careful scrutiny is required. Diagnosis is usually easiest when the esophagus is fully dilated and viewed obliquely or in profile. It has a characteristic appearance, which is a sharp, thin, regular indentation of the barium column. It is easily distinguished from stenosis, which has tapered edges; or muscular spasm or contractions, which are transient; and carcinoma, which presents a broader, more irregular narrowing. It may be helpful to have the patient ingest a barium tablet to better

delineate the web and its functional significance.

Treatment may remain conservative, depending upon the severity of symptoms. Dilation or even endoscopic lysis is usually adequate. Any associated underlying problem, such as gastroesophageal reflux, should be treated. Case reports of excision through a thoracotomy have been described.

Lower Esophageal Web and Schatzki's Ring. Lower esophageal webs are very unusual, although the exact incidence is unknown. They relate to the lower 2 cm of the esophagus, which normally lies below the diaphragm. They are very often mistaken for lower esophageal rings, which are much more common, but by definition webs are covered only by squamous epithelium.

The cause is believed to be at least in part inflammatory, which is supported by histologic evidence of submucosal inflammation. Clinically, symptoms are indistinguishable from those of a lower esophageal ring, and the diagnosis is usually made on radiographic and histologic grounds.

As mentioned previously, a lower esophageal ring or Schatzki's ring is located at the gastroesophageal junction, and thereby is covered by squamous epithelium on its upper surface and columnar epithelium on its lower surface. It was originally described as a cause of dysphagia in 1953 by Schatzki and Gary, and since then it has been increasingly recognized as one of the most common causes of dysphagia as well as a common coincidental findings. It may be seen on as many as 14 per cent of routine barium studies, approximately a third of which will be from symptomatic individuals. Symptomatic patients tend to be male and older than 40 years of age.

The cause is not clear, but a number of theories have been proposed, including the plication theory, which holds that the esophageal mucosa is simply redundant; the developmental theory, which claims the ring is simply a natural expression of the junction between the esophagus and stomach; and the inflammatory theory, which proposes that the ring is merely secondary to peptic esophagitis. Histologically, it is a mucosal ring, the core of which is composed of a variable amount of connective tissue with essentially no contribution from the muscular wall.

The caliber of the ring is what largely determines whether symptoms occur. Generally, if the intraluminal diameter of the ring is less than 13 mm, symptoms are inevitable. If the diameter is larger than 20 mm, symptoms are unlikely, and anything in between is variable. Characteristically, the dysphagia that results is intermittent, attacks often being separated by weeks or months. Symptoms often occur when the patient is excited, distracted, and eating a hurried meal so that the food is poorly chewed. A bolus will become stuck and cause obstruction, which the patient may relieve with vomiting or forceful swallowing with liquids. Subsequently, he or she will be able to eat normally, which is atypical of any other disorder. This has been dubbed the "steak-house syndrome", because steak is so often responsible and steak houses are frequently the site of hurried business lunches.

Episodes gradually become more frequent, which ultimately brings the patient to medical attention. Associated chest pain or heartburn is uncommon.

Radiologically, a lower esophageal ring appears symmetric, as opposed to an esophageal web. It will appear narrow, usually 2 to 4 mm in thickness, but will be missed unless the esophagus is dilated with barium beyond the size of the ring. A barium tablet or barium-coated bread may be useful in making the diagnosis.

Endoscopy is useful to rule out esophagitis or any other underlying abnormality. Here, too, however, a ring will be difficult to visualize unless the esophagus is distended. An advantage of fiberoptic flexible endoscopy in this situation is the larger caliber of the scope and its ability to insufflate air.

Treatment is tailored to the patient, depending upon the frequency and severity of symptoms. Weight loss is unusual because of the intermittent nature of the problem. Oftentimes reassurance and more thorough mastication and slower eating habits may suffice. A better fitting pair of dentures may be helpful. Otherwise, dilation is indicated, whether by bougienage or endoscopy. In some instances, more aggressive pneumatic dilation will be necessary, but only rarely is surgical excision necessary.

Diverticula

Esophageal diverticula are best classified by location. They tend to occur at the pharyngoesophageal junction (Zenker's diverticulum), in the midesophagus just below the level of the carina, and in the distal esophagus. Most are acquired secondary to pulsion forces, rather than traction, and by and large tend to be false diverticula. In other words, they consist primarily of mucosa that has herniated through the muscularis propria, rather than containing all layers of the esophageal wall.

Pharyngoesophageal (Zenker's) Diverticulum. The Zenker's diverticulum is the most common and occurs at the junction of the hypopharynx and the cervical esophagus. A mucosal sac protrudes between the oblique and transverse fibers of the cricopharyngeus muscle, an inherently weakened area known as Killian's triangle, in the posterior midline.

Although the diverticulum had been described previously, Zenker in 1878 was one of the first to recognize its relationship to an underlying anatomic defect and repeated pulsion forces. Since then, the pathophysiology has been a matter of substantial debate. Since Killian's dehiscence is located just above the transverse portion of the cricopharyngeus muscle, many favor the theory that spasm or failure of relaxation of the upper esophageal sphincter results in a proximal high-pressure zone as the pharyngeal constrictor muscles contract. Killian's area gradually gives way to this continued pressure and herniation eventually occurs. Manometric studies, however, have not corroborated this theory.

Ellis found that although resting tone and onset of relaxation remained normal in these patients, relaxation ended prematurely before pharyngeal contraction was complete. The problem, therefore, was one of incoordination. Ellis readily admits, however, that others have not confirmed his findings.

More recently, Knuff and colleagues evaluated nine patients with Zenker's diverticula, using modern low-compliance manometric recording equipment. They found no evidence of spasm, achalasia, or incoordination. They did find low upper esophageal resting pressures, but

the significance of this finding remains unclear.

Such contradictory evidence is most likely due in large part to the variable manometric techniques used. In addition, as mentioned earlier in this chapter, the upper esophageal sphincter area is very difficult to measure accurately. The picture is further confused by individual patient variability. Therefore, although the prevailing theory still seems to favor some form of incoordination, documentation is lacking.

The chief symptom, of course, is dysphagia, which tends to be gradual in onset and of long duration. A sensation of food sticking in the throat may be noted, frequently localized to the suprasternal notch. Material retained in the pouch will lead to regurgitation of undigested food, a foul taste in the mouth, and foul breath. This regurgitation may be worse at night and may lead to nocturnal aspiration and chronic coughing, and a picture of pneumonitis may develop in older, debilitated patients. Seventy per cent of patients in whom Zenker's diverticula develop are older than 60 years of age.

As the pouch distends, compressive symptoms may occur, both on the esophagus and in the airway, and associated pain may be present. The sac may then become palpable as it projects usually into the left side of the neck, posteroinferior to the sternocleidomastoid muscle. This seems to be the path of least resistance, since the cervical esophagus normally bulges to the left. Indirect laryngoscopy may reveal significant salivary pooling.

Plain x-ray films may show a retropharyngeal fullness with an air-fluid level, but a barium study is diagnostic, as it will show a variably sized pouch that may demonstrate multiple radiographic defects from retained food. Although this may suggest malignancy, which is a potential possibility in long-standing diverticula, it is distinctly uncommon. The incidence of malignant degeneration in a Zenker's diverticulum has been reported at 0.31 per cent.

Endoscopy adds little to the diagnosis unless one is concerned about malignancy or sac decompression is necessary. Otherwise, it is hazardous and best avoided. Because the sac has no muscular compartment, it is very thin and easily perforated. The preceding discussion on pathophysiology notwithstanding, manometry also has little benefit in the routine diagnosis of these lesions.

If symptoms are mild and intermittent, conservative therapy may suffice. This would include more thorough chewing of food and copious flushing with liquids. Periodic dilation may also be helpful. Typically, however, surgical intervention will be necessary.

The recommended surgical approach in these cases has varied, largely revolving around the issue of whether a cricopharyngeal myotomy is necessary. There are those who claim that this procedure alone is adequate therapy without the need for diverticulectomy, since it supposedly eradicates the underlying cause. By the same token, there are those who claim that diverticulectomy without adjunctive cricopharyngeal myotomy leads to unacceptable recurrence rates. Conversely, there are those who have found the addition of cricopharyngeal myotomy to make no difference. Still others have recommended diverticulopexy rather than diverticulectomy, which is simply inverting the sac and tacking its base superiorly to the anterior longitudinal ligament of the cervical spine. This avoids a

mucosal incision and allows for sac decompression.

Once again, there is contradictory information, which stems largely from patient variability, as well as varying methods of follow-up and surgical technique. It would seem that for small but symptomatic diverticula, cricopharyngeal myotomy alone is adequate, particularly since these diverticula are often difficult to locate. Larger diverticula, however, should be resected, which is preferable to suspension and is probably best combined with cricopharyngeal myotomy.

One further surgical approach deserves mention. In the early part of this century, an endoscopic technique was described, but one case of mediastinitis occurred and the procedure was abandoned. In 1960, Dohlman and Mattson revived the technique using diathermy and a specially adapted bivalved esophageal speculum. The idea is to lyse the party wall between the esophagus and diverticulum to effect better drainage. They reported a series of 100 patients with no significant complications and a recurrence rate of 7 per cent. This compares with a recurrence rate of 3.6 per cent reported for diverticulectomy. Because it is done endoscopically, however, it is a lesser operative procedure with less morbidity. For this reason, the Dohlman procedure is recommended for older, more debilitated patients who may not withstand an external approach, remembering that it does require experience to be performed safely and effectively. The external approach is still preferred in healthier, younger patients.

Midesophageal Diverticula. Traditionally, diverticula appearing in the midportion of the esophagus, at about the level of the fourth and fifth thoracic vertebrae, have been believed to result from traction by surrounding inflammatory nodes. The usual cause was tuberculosis. More recent data, however, relate these more to pulsion forces or abnormalities of esophageal motility. Increasing intraluminal pressure between spastic segments results in diverticula at areas of inherent weakness. A predilection for this site has been theorized to be secondary to residual congenital attachment of the esophagus to the trachea in the region of the carina on its anterior wall.

Generally, these diverticula incorporate the entire thickness of the esophageal wall. For that reason, because of the muscular component, they tend not to reach a very large size. In addition, they usually maintain a wide opening into the esophagus and thus are often asymptomatic. Symptoms may result from the underlying motility problem, or in extreme cases patients may complain of dysphagia or substernal chest pain. Suppurative complications, such as mediastinitis and tracheoesophageal or bronchoesophageal fistula, were more apt to occur with traction diverticula and are less common today.

Diagnosis is made radiographically and often may be an incidental finding. Manometry may be helpful if an underlying motility disorder is suspected. Treatment is then indicated. Otherwise, the diverticula usually do not warrant specific therapy unless, of course, complications develop. A thoracotomy will then be necessary.

Epiphrenic Diverticula. These diverticula occur in the distal 10 cm of the esophagus and are uncommon, making up less than 10 per cent of esophageal diverticula. They are twice as common in men and can be found in a younger age group.

The cause is once again most likely related to pulsion forces from a motility disturbance. Forceful contractions with abnormal lower esophageal sphincter relaxation cause elevated intraluminal pressures. This pressure is dissipated by the herniation of a mucosal sac through an area of muscular weakness. Epiphrenic diverticula have most often been associated with diffuse esophageal spasm and achalasia but have been correlated with reflux esophagitis and hiatal hernia as well. They usually protrude posteriorly and extend into the right side of the thorax.

Symptoms usually result more from the underlying disorder; however, the diverticulum may itself produce mild dysphagia. Regurgitation of undigested food with associated aspiration may occur, which is worse when the patient is supine. With significant distention, compression of the esophagus with obstruction may occur, as may compromise of pulmonary function. Ulceration with perforation and mediastinitis has been reported, but is unusual. Malignant degeneration is also a rare complication.

Treatment is usually directed toward the underlying disorder, and therefore manometry usually plays an important diagnostic role. If significant retention occurs in the sac, liquids and postular maneuvers may be helpful. Surgery becomes necessary in severe unresponsive cases. This requires a left-sided thoracotomy and diverticulectomy, and a concomitant myotomy is usually recommended.

Ingestion of Caustic Substances

Caustic injuries of the esophagus tend to occur by accident in children, in whom they are more common, and in adults who attempt to commit suicide. For that reason, adult injuries tend to be somewhat more severe. The goal in these cases is accurate assessment of the location and extent of injury and the prevention of significant complications such as stricture and perforation. The means to achieving these goals has been the subject of much discussion and controversy. Unfortunately, it is difficult to develop well-controlled, randomized studies to explore these issues, and although animal studies shed some light, the clinician faces a substantial challenge when treating severe corrosive injuries of the esophagus.

It is estimated that more than 5000 cases of caustic agent ingestion occur in the United States each year, 50 to 80 per cent occurring in children. The most common agents are alkaline, such as sodium and potassium hydroxides, which are found in many household cleaners, and acid corrosives. In addition to the pH, the degree of injury is determined by the quantity, viscosity, and contact time with mucosal surfaces.

Alkali tends to penetrate more rapidly than acid, thereby more often causing oropharyngeal and esophageal burns. It causes a liquefaction necrosis and can rapidly extend through the submucosa into underlying muscle. Conversely, because of its alkaline pH, the esophagus tends to be more resistant to acid burn. In addition, acid burn produces a coagulation necrosis and superficial eschar, which helps prevent deeper penetration. More severe acid damage, therefore, occurs in the stomach.

Crystalline substances are more difficult to swallow and tend to produce more severe damage proximally, in the oral cavity, pharynx, and hypopharynx. The rapid pain produced

tends to inhibit further ingestion. Liquids are much more easily swallowed and tend to produce the most damage at normal anatomic narrowings at which point there is greater contact time. As discussed earlier in this chapter, this would include the area of the cricopharyngeus muscle, the aortic arch, the left main-stem bronchus, and the gastroesophageal junction. Once in the stomach, pooling generally occurs in the antrum if the patient is upright.

Acutely, patients usually experience severe pain, odynophagia, and dysphagia from esophageal spasm and inflammatory edema, which gradually peaks over the first 48 hours. Supraglottic edema may occur, causing stridor and hoarseness. These severe symptoms usually subside within several days so that by the end of the first week, the patient is able to swallow. Symptoms will of course depend upon the severity of injury, and if esophageal perforation occurs patients may suffer severe chest pain, subcutaneous emphysema, sepsis and shock.

As the acute reaction subsides, the patient may enter a quiescent period lasting 3 to 6 weeks in which symptoms are relatively nonexistent. In those with extensive injury, there is fibroblastic and collagen proliferation with scar contracture during this period. Subsequently, stenosis may develop, resulting in severe dysphagia.

The most important prognostic factor regarding ultimate outcome is the depth of injury. First-degree burns, resulting in hyperemia and superficial desquamation, rarely result in serious complications. Second-degree burns penetrate the mucosa to expose submucosa and muscle, and significant scarring and contracture may result. Third-degree burns result in transmural damage, which may lead to perforation.

The best method of establishing depth of injury is with endoscopy. Unfortunately, no definitive staging criteria exist, and therefore it becomes very subjective and arbitrary. Assuming the patient's respiratory and cardiovascular status is stable, however, endoscopy should be the first diagnostic step. It remains the most sensitive approach to assess the extent of injury and the need for further therapy.

Most recommend early endoscopy, preferably within the first 24 to 48 hours, because of the advantages of instituting prompt therapy. It is widely recognized that there is little correlation between the presence or lack of oropharyngeal burns and esophageal burns. Many feel, however, that if extensive injury is encountered endoscopically, particularly a circumferential second or third-degree burn, further passage of the endoscope is ill-advised because of the risk of perforation. Others would disagree. Thompson has pointed out that this risks the failure of detecting more distal, possibly life-threatening burns that may require debridement or repair. He therefore recommends complete endoscopic evaluation of the esophagus and stomach, acknowledging the need for experienced hands. Some have even advocated early laparotomy and thoracotomy when there is second- or third-degree burn to assess the condition of the esophageal wall and stomach.

The early use of radiographic contrast studies is of limited value other than to detect perforation. They are not sensitive for establishing the depth of injury. However, they are recommended routinely at 3 weeks and during regular follow-up to assess the development of stenosis.

Therapy for caustic injuries has focused largely on prevention of subsequent complications. Acute measures should be supportive, such as ensuring an airway and volume replacement. Some have recommended efforts to neutralize or dilute alkaline ingestants; however, there is little evidence that this is of any benefit. Emesis and gastric lavage should be avoided because of the risk of repeated esophageal exposure.

If significant burn injury is found, most would favor prompt administration of steroids and antibiotics. Steroids have been found to help prevent stricture formation if administered within 24 to 48 hours of injury. However, they are best used in cases of moderately severe alkaline injuries. Superficial burns will not become stenosed, and severe transmural burns will be little affected. In addition, as previously mentioned, acid ingestion is less likely to cause significant esophageal injury. The steroids should be continued for at least 3 to 4 weeks.

Antibiotics are also of benefit to prevent intramural spread and possibly mediastinitis, as well as to minimize granulation tissue. They should be continued concomitantly with the steroids.

There are those who favor early prophylactic dilation in patients with severe burns. However, since this is associated with a greater risk of perforation, and since strictures do not actually develop in most patients, it is not widely advocated. Dilation may subsequently be required in patients in whom stenosis does eventually develop, which will be approximately 20 to 40 per cent of patients with significant corrosive injuries. More about specific dilation techniques will be discussed later in this chapter.

Indwelling tubes or esophageal stents have also been recommended but have met with little success. By the same token, the early placement of nasogastric tubes has been controversial and is not generally practiced, although it is strongly advocated by some investigators.

There has been some experimental work with medications that interfere with collagen synthesis, such as penicillamine and beta-aminopropionitrile, which are known as lathyrogens. Preliminary animal work is encouraging, but further investigation is necessary.

One final note regarding caustic agent ingestion that is often overlooked is that of medication-induced esophagitis. A variety of medications when retained in the esophagus can cause a localized irritation and ulceration. In a review of the literature, Kikendall and associates, found 221 cases that were reported secondary to 26 different medications, and they acknowledge that the syndrome has only been widely recognized since 1970. Very likely, many more cases go unreported.

Although pre-existing esophageal disease may affect esophageal transit and predispose to retention of medications, it is by no means a prerequisite for development of this problem. Interestingly, only eight of ten normal subjects will clear radiolabelled capsules from the esophagus after one swallow with water in a sitting position. This becomes worse in the supine position or without water, and many swallows may be required. Most are unaware of this retention. Patients who take their medication at bedtime, with little or no water, are at significant risk. Swallowing decreases at night, and in the recumbent position retention occurs with prolonged mucosal contact. Ensuing esophagitis and ulceration further impair esophageal

motility, aggravating the problem.

The most common medications accounting for esophagitis have been antibiotics, particularly doxycycline (Vibramycin) (Table 7). Emepronium bromide is an anticholinergic agent used to relax the bladder, but it is not available in the United States. Other commonly prescribed medication that have been implicated include potassium chloride, ferrous sulfate, quinidine, and some steroidal as well as nonsteroidal anti-inflammatory agents.

Table 7. Drugs Causing Esophagitis

Tetracycline compounds (especially doxycycline)
Emepronium bromide
Potassium chloride (especially slow-release formulations)
Iron supplements
Alprenolol chloride
Quinidine
Aspirin
Clindamycin
Indomethacin
Ascorbic acid
Steroids
Other antibiotics (ampicillin, erythromycin, phenoxymethyl penicillin, lincomycin, tinidazole).

Symptoms are consistent with esophagitis and include odynophagia (74 per cent), continuous retrosternal pain (72 per cent), and dysphagia (20 per cent). Ulceration, stricture, and even perforation may occur.

Endoscopy will usually demonstrate areas of discrete ulceration and esophagitis. The site within the esophagus most often injured is at the level of the aortic arch; this is caused by compression and is also the site of transition from striated to smooth muscle so that the peristaltic wave is reduced in amplitude.

Barium esophagograms will vary depending upon the severity of inflammation and may reveal extensive ulcerations. In severe cases, they may demonstrate a significant stricture that can easily be mistaken for malignancy. In fact, failure to confirm a radiologic and endoscopic diagnosis of malignancy by histologic examination should raise the possibility of underlying drug-induced esophagitis.

Treatment includes stopping as many of the medicines as possible or replacing them with liquid preparations. Medications should be taken in the sitting or standing position with plenty of liquids and not just before bedtime. Most symptoms will resolve after these simple measures, although occasionally more aggressive intervention, such as dilation will be necessary.

Gastroesophageal Reflux

The lower esophageal sphincter and supporting structures are remarkably effective in preventing the reflux of gastric acid into the esophagus. The precise mechanism whereby this

barrier functions has been subject to some speculation and was discussed earlier in the section on physiology of the esophagus. Under normal circumstances, however, it allows the reflux of material less than 1 per cent of the time when the individual is supine and 2 per cent of the time when the individual is upright. Most of this occurs after meals and is considered physiologic.

Reflux becomes pathologic when it occurs in excess and begins to cause symptoms in association with esophagitis. Exactly why this occurs is oftentimes not clear, but usually a variety of neural, humoral, and anatomic factors culminate in the breakdown of the barrier mechanism. Certainly, abnormal relaxation or hypotension of the lower esophageal sphincter will allow excessive reflux to occur, but it will be aggravated by delayed gastric emptying and distention. Esophageal dysmotility will fail to clear the esophagus readily of refluxed material, which will then exacerbate mucosal damage and subsequent symptoms.

In fact, reflux damage is probably greater at night despite reflux episodes being more prevalent during the day. This is because swallowing occurs much less frequently during sleep, averaging 7 times/hour as opposed to 72 times/hour when the individual is awake. Refluxed material is therefore not cleared as rapidly and tends to pool in the lower esophagus.

Thus the damaging effects of reflux depend in large part upon the duration of exposure, as well as the frequency and volume of refluxed material. In addition, studies have shown that bile-contaminated gastric juice is much more corrosive than gastric juice alone. As esophagitis develops, sphincter tone tends to decrease further, and disordered motility of the lower esophagus occurs, both of which exacerbate the problem.

The most common symptom then becomes heartburn, a retrosternal burning pain that tends to occur after large meals, after lying down, or after any activity that tends to increase intragastric pressure, such as bending over or heavy lifting. The pain is predominantly retrosternal and epigastric but may be referred to the back, arm, pharynx, and ear when severe. In fact, isolated pharyngeal or ear pain may occasionally be the only presenting complaint. Large meals that are particularly fatty or spicy most often produce symptoms, which last from 20 minutes to 2 hours and can usually be relieved with antacids. Interestingly, the severity of pain often does not correlate histologically with the amount of esophageal inflammation. As mentioned earlier, however, pain that is aggravated by hot or cold liquids, coffee, citrus juices, or alcohol suggests ulcerative esophagitis.

At times the pain from gastroesophageal reflux may be atypical and must be differentiated from cardiac pain. Although nitroglycerin has on occasion been reported to relieve reflux pain, Henderson and co-workers point out that if relief occurs rapidly, that is, within 3 minutes, it is unlikely to be esophageal in origin.

Other symptoms include a sour (acid) or bitter (bilious) taste in the mouth from regurgitation. This occasionally may be associated with a burning sensation of the tongue, lips, and buccal mucosa. Excess salivation may occur secondary to lower esophageal irritation, which is referred to as waterbrash. Nocturnal aspiration, choking, and coughing may develop, which, if chronic, may lead to recurrent bronchitis or pneumonia. Laryngeal irritation may cause hoarseness or cough.

Patients with gastroesophageal reflux may also complain of varying degrees of dysphagia, which is usually due to one of three causes. Esophageal irritation may lead to spasm and dysmotility, as already mentioned. Chronic esophagitis may lead to stricture formation, which will be discussed further on. Reflux has also been associated with upper esophageal sphincter dysfunction or achalasia in as many as 50 per cent of patients with significant reflux. Presumably, closure of the sphincter initially represents a protective mechanism to prevent aspiration of gastric contents. Its dysfunction represents a response to chronic reflux. Cherry and colleagues were one of the first to recognize that pharyngeal symptoms could result from gastroesophageal reflux, in which case the cause tends to be rather obscure. It is estimated that the majority of cricopharyngeal problems seen by otolaryngologists are in fact related to gastroesophageal reflux.

The diagnosis of gastroesophageal reflux can often be made on a good history, and therapy can be initiated empirically. If the patient fails to respond to initial therapy, or if symptoms are atypical, diagnostic tests are indicated to substantiate reflux disease. A barium esophagram is typically the first test performed, preferably with fluoroscopy. This can be used to detect spontaneous reflux or reflux secondary to provocative maneuvers. However, as stated earlier, this test alone tends to be terribly inaccurate, picking up only 40 to 50 per cent of patients with reflux. The reason has been ascribed to the intermittent nature of lower esophageal sphincter relaxation in these patients as opposed to sustained hypertension. Still, changes secondary to esophagitis may be revealed with barium - from superficial erosions in some cases to severe ulcerations and strictures.

Tests that demonstrate more sensitivity and specificity are those that incorporate intraluminal pH measurements; these tests were discussed previously in the section on diagnostic tests. They are indicated if reflux disease needs to be substantial, and they can be readily combined with manometry. Lower sphincter pressures less than 10 mm Hg strongly support a diagnosis of gastroesophageal reflux, whereas pressures greater than 20 mm Hg suggest an alternative cause. The validity of sphincter pressure measurement as a diagnostic tool is controversial, however, because of a reportedly high incidence of normal values in otherwise symptomatic patients.

Another test to consider is radionuclide scintigraphy, which is rapid and noninvasive and may provide quantitative as well as diagnostic information. Delayed esophageal transit time has been demonstrated in patients with reflux esophagitis. In addition, once esophageal emptying has occurred, intra-abdominal pressure can be increased by an abdominal binder or other maneuvers. Radioactivity crossing the gastroesophageal junction at less than 35 mm Hg is considered evidence of pathologic reflux. The test has been found to have a sensitivity rate of 90 per cent, with a 10 per cent false positive rate.

Endoscopy is not necessary in every patient, but it is helpful to verify the presence and severity of esophagitis. Certainly, if preliminary evaluation shows ulceration or stenosis or suggests the possibility of malignancy, endoscopic evaluation is essential. If the distal esophageal mucosa appears normal, this does not rule out the possibility of reflux disease, since it has been reported that as many as 55 per cent of patients with symptomatic reflux will have normal endoscopic examinations. Erythema alone is not sufficient to diagnose esophagitis, and one should note the friability of the mucosa with the presence of exudate and ulceration. Biopsy is indicated in this situation, and a number of staging systems have been

established to assess the degree of injury.

The treatment for gastroesophageal reflux should proceed in a stepwise fashion (Table 8), but most patients will respond to conservative management. This includes elevating the head of the bed, which is preferably done on blocks rather than on pillows. The patient should be instructed not to eat at least 2 hours prior to retiring, also to help prevent nocturnal reflux. Maneuvers that foster reflux by increasing intra-abdominal pressure - such as tight clothing, bending forward, heavy lifting, and straining - should be avoided. By the same token, weight reduction is important if the patient is overweight, and small meals are preferable to avoid gastric distention.

Table 8. Treatment of Reflux Esophagitis

Step 1:
- Elevate head of bed 6 to 8 inches.
- Discontinue smoking.
- Diet.
  - Lose weight, if overweight.
  - High-protein, low-fat (less than 45 gm/day) diet.
  - Eat small meals.
  - Avoid problem foods.
- Do not lie down after eating.
- Avoid tight clothing, bending, heavy lifting.

Step 2:
- Antacids.
- Alginic acid (Gaviscon).

Step 3:
- Bethanechol.
- Metoclopramide.
- Cimetidine, ranitidine.

Step 4:
- Fundoplication.
  - Nissen (abdominal).
  - Belsey (thoracic).

Dietary adjustments are essential and should be strongly encouraged. This would include not only avoiding those foods that obviously exacerbate symptoms but also avoiding those foods that tend to decrease lower esophageal sphincter pressure, such as fat, alcohol, and coffee. In fact, there are a number of foods and medications that have been found to decrease lower esophageal sphincter pressure, which are best avoided if possible (Table 9). Conversely, high-protein meals have been found to enhance lower esophageal sphincter pressure (Table 10).

Table 9. Agents Decreasing Lower Esophageal Sphincter Pressure

Fat
Chocolate
Ethanol
Peppermint
Theophylline
Caffeine
Gastric acidification
Smoking
Valium
Calcium blockers
Meperidine, morphine
Hormones
    Secretin, cholecystokinin, glucagon
Beta-adrenergic agonists
    Isoproterenol
Alpha-adrenergic antagonist
    Phentolamine
Dopamine
Anticholinergics
    Atropine.

During the initial stages of therapy, antacids will usually be a necessary adjunct and may be instituted as needed or on a regular schedule. They are best given after meals and at bedtime. They help to neutralize gastric acid as well as promote the release of gastrin, which enhances lower esophageal sphincter tone. Agents such as Gaviscon contain bicarbonate, which reacts with gastric acid to produce carbon dioxide. This produces a foam that helps to mechanically impede reflux.

Table 10. Agents Increasing Lower Esophageal Sphincter Pressure

Protein meals
Histamine
Antacids
Metoclopramide
Indomethacin
Hormones
    Gastrin
Alpha-adrenergic agonists
    Norepinephrine, phenylephrine
Cholinergic drugs
    Bethanechol
Anticholinesterase
    Edrophonium.

If the patient fails to respond, or if the symptoms are severe, one of the H_2-receptor antagonists is indicated. Both cimetidine and ranitidine have been found to reduce gastric acid

output. However, although they improve symptoms, they do not affect lower esophageal sphincter tone and thus do not prevent reflux. Drugs such as metoclopramide and bethanechol act as a cholinergic stimulus to increase lower esophageal sphincter tone as well as to enhance peristalsis, which improves gastric emptying. They can help to improve reflux symptoms, although bethanechol also stimulates gastric acid secretion and metoclopramide can cause drowsiness and ataxia. They are therefore recommended only if the aforementioned therapies have failed.

When the patient continues to have severe symptoms despite thorough medical therapy, or if complications ensue, including ulceration, stricture, Barrett's esophagus (to be discussed further on), malnutrition, bleeding, or aspiration, surgical therapy should be considered. A number of fundoplication procedures have been described, with and without associated hiatal hernia repair. Nissen and Belsey procedures are the most widely used and have been shown to relieve esophagitis and increase lower esophageal sphincter pressure. Over time, however, these procedures may lose their effectiveness and there may be recurrences. Henderson and Marryatt have described a fundoplication technique combined with gastroplasty that achieved excellent results in 93 per cent of patients participating in a large series who were followed for more than 5 years.

Hiatal Hernia. The more common sliding type of hiatal hernia has long been implicated as a causative factor in reflux disease. It seems logical that displacement of the lower esophageal sphincter into the negative pressure of the thoracic cavity would predispose to incompetence and reflux.

The esophagus passes through a hiatus in the diaphragm at which point it is stabilized by the diaphragmatic crura and attachment of the phrenoesophageal membrane. This membrane is simply a coalescence of the endothoracic and endoabdominal fasciae that line the diaphragm. The hiatal opening through the crura is a narrow slit and compresses the esophagus slightly at this point to an oval shape. This probably plays a small secondary role in maintaining competency of the lower esophageal sphincter. Actually, as discussed earlier in this chapter, a number of factors are believed to contribute to the lower esophageal sphincter, but the precise mechanism is still debated.

Essentially two types of hernias can develop through the hiatus secondary to its enlargement and laxity of its membranous attachments. The first type, the sliding or axial hiatal hernia, involves herniation of the proximal gastric cardia through the hiatus. The phrenoesophageal membrane remains attached, though lax, so the herniated segment is not a free peritoneal sac. The cause remains unknown but has been related to congenital, traumatic, and iatrogenic mechanisms. It is estimated that 10 per cent of adults will demonstrate varying degrees of sliding hiatal hernia during routine barium swallow. In contrast, only 5 per cent of patients with such a radiologic diagnosis will have symptomatic gastroesophageal reflux.

In fact, more recent investigations support the notion that it is the competency of the lower esophageal sphincter, independent of its relationship to the diaphragm, that determines whether reflux will occur. Although the two may be coincidental, the presence of a hiatal hernia does not necessarily predispose to the development of symptomatic reflex. The majority of patients with a sliding hiatal hernia are asymptomatic, although larger hernias may

occasionally be associated with bloating and increased belching. Therefore, a diagnosis of gastroesophageal reflux needs to be made independently of the presence of a hiatal hernia.

The second type of hernia is the paraesophageal hiatal hernia, in which portion of the gastric fundus herniate through the hiatus adjacent to the esophagus. It generally occurs through a weakened portion of the phrenoesophageal membrane, whereas the lower esophageal sphincter remains in its subdiaphragmatic position. With continued exposure to negative intrathoracic pressure, as well as positive intra-abdominal pressure, the sac gradually enlarges. Eventually, the gastroesophageal junction is pulled upward through the defect, producing a combined sliding and paraesophageal hiatal hernia.

Approximately 20 per cent of patients with a para-esophageal hernia will complain of varying degrees of dysphagia. With obstruction of acid drainage from the sac and ischemia, gastritis, ulceration, and bleeding can occur, although associated esophagitis is unusual. A dilatation continues, volvulus, infarction, and perforation may eventually result. For this reason, surgical repair is recommended for paraesophageal hernias even if the patient is asymptomatic.

Sliding hiatal hernias, in contrast, do not require any specific therapy. If gastroesophageal reflux is present, it needs to be addressed; otherwise, there is no correlation with subsequent enlargement and production of symptoms. There are even reports that small sliding hiatal hernias may regress with time.

Complications. The surest way to treat complications arising from gastroesophageal reflux is to begin therapy early and prevent their occurrence. Once complications have ensued, they can be difficult to treat effectively. Table 11 lists some of the more common complications.

Table 11. Complications of Gastroesophageal Reflux

Esophageal ulceration
Hemorrhage
Aspiration
Stricture
Barrett's esophagus

Esophageal ulceration is perhaps the most common complication and tends to occur in those patients with delayed esophageal and gastric emptying. Ulcers tend to occur on the esophageal side of the squamocolumnar junction and tend to remain superficial. However, they may cause a significant inflammatory reaction in the deeper muscular layers of the esophagus. These patients are more likely to complain of dysphagia and odynophagia, which will fluctuate with the degree of inflammation. The pain can become quite severe, characterized as a deep, boring pain radiating to the back, which may be relieved with antacids. It is exacerbated by positional changes that increase intra-abdominal pressure. Although the risk of perforation is real, it is very uncommon.

Significant bleeding from esophagitis is unusual, but when it occurs it can be profuse. It may occur secondary to diffuse esophagitis or a penetrating esophageal ulcer and must be

differentiated from varices. Endoscopy is thus indicated. Control can usually be achieved with upright positioning and administration of iced saline and antacids via a nasoesophageal tube, but occasionally immediate surgery may be necessary. Skinner and Belsey reported that 3 per cent of their patients who required antireflux surgery had major bleeding as their presenting symptom.

Aspiration and the subsequent development of pulmonary complications often presents a problem in diagnosis. The two may often coexist, and it is difficult to substantiate that reflux, in fact, is causing the respiratory symptoms. Morning hoarseness is a frequent complaint, as is a chronic cough that seems to be worse during the night. These are easily complaints that may bring referral to an otolaryngologist. In more advanced cases, vocal cord inflammation may be noted, particularly in the posterior commissure, and secondary pulmonary fibrosis, bronchiectasis, and recurrent pneumonia may occur. An association has long been reported between gastroesophageal reflux and exacerbation of asthma. The best objective method for establishing cause and effect seems to be 24-hour pH monitoring, which can relate reflux to symptoms. In addition, radionuclide scintigraphy has been used for this purpose.

Chronic reflux esophagitis that goes on to ulceration will ultimately tend to heal with scarring and contracture, resulting in stricture formation. The time course for this to occur is highly variable, which emphasizes the need to treat ulcerative esophagitis aggressively. Approximately 10 to 15 per cent of patients with reflux esophagitis will subsequently experience a stricture. Progressive dysphagia is characteristic, occasionally localized by the patient to the lower esophagus, and occasionally associated with substernal chest pain. Gradually, as the stricture worsens, it will allow less reflux; partial healing will then occur and the stricture will stabilize. Pain will tend to decrease, and the patient will tolerate liquids. Weight loss therefore is usually not significant. A history of prolonged heartburn may not be forthcoming, as a number of patients have significant symptoms only when stricture and dysphagia ensue. The clinician, of course, should always be suspicious of the possibility of an underlying malignancy.

As reflex esophagitis and ulceration persist, healing may occur by upward migration of gastric columnar epithelium. This phenomenon was first diagnosed and subsequently reported by Barrett, who initially ascribed a congenital cause, and it has become known as Barrett's esophagus. Although isolated islands of columnar epithelium do occur (often as high as the cervical esophagus) and are believed to be congenital in origin, it is now well documented that secondary healing by upward migration does occur. It is easily recognized endoscopically because of its salmon red appearance, which contrasts with the more pale-appearing stratified squamous epithelium of the esophagus. Although this type of epithelium is more resistant to gastric acid, when ulceration does occur it tends to be more severe with deeper penetration, very much like gastric ulcers. Bleeding may occur, and strictures are very common. The main clinical significance of Barrett's esophagus, however, lies in its potential for malignancy. The incidence of adenocarcinoma in Barrett's esophagus is approximately 10 per cent, whereas adenocarcinoma composes less than 15 per cent of all esophageal malignancies. It is therefore important that a transition to columnar epithelium be recognized and aggressively followed. Treatment will include antireflux measures, which when successful may yield a reversion toward normal squamous epithelium. If the patient fails to respond, however, surgery should be considered.

Dilation. Conservative treatment for esophageal stricture usually involves some form of dilation (Table 12).

Table 12. Methods of Dilation

Tucker esophagoscope
Rigid esophagoscope
Eder-Puestow metal olives with guide wires
Hurst and Maloney mercury bougies
Distensible balloons

Esophageal dilation dates back to the 16th century when blind bougienage was performed for foreign body impaction. A wax taper was usually used for this purpose and, in fact, the word bougie is derived from an Algerian town named Boujiyah, which was the center for the medieval wax candle trade.

Bougienage at this time was done primarily for foreign body impaction and caustic agent ingestion. Other devices were developed, including a lead instrument with an olive-shaped tip that was passed with the aid of gravity rather than pushing blindly.

By the 19th century, the technique was well established, then in the later part of that century, esophagoscopy was developed, and with it Chevalier Jackson developed instruments to be manipulated through the scopes for dilation, biopsy, and foreign body extraction. This formed the beginning of otolaryngology as a specialty, with an emphasis on bronchoscopy and esophagoscopy.

During the 20th century, with the development of thoracic surgery, many chronic lower esophageal problems have become amenable to surgical treatment. Still, dilation is often indicated, particularly in the early stages of therapy. A number of techniques are available, depending upon the severity of the stricture.

Tight strictures are best dilated over some sort of guide or under direct visualization. Plummer was one of the first to suggest passing a bougie over a previously swallowed thread. This technique was carried further by the Tucker method, which uses retrograde dilatation. The string is brought out through a gastrostomy, and dilation can be performed in a retrograde and then prograde fashion, pulling the string and dilator back and forth. Because the esophagus tends to distend above the stricture, retrograde dilation is theoretically safer in severe cases by avoiding a false passage.

The rigid esophagoscope can be used as a dilator, taking advantage of the bevel tip. Otherwise, a variety of bougies have been devised for passage through the scope. Most have adapted the olive-shaped tip which was popularized by Jackson, which seems to be safer than conical or blunt tips. The Eder-Puestow dilator system combines both techniques, passing guide wires under direct visualization and passing graduated metal olives over these guides.

The technique of using weighted bougie dates back to the 18th century, but Hurst in 1915 was the first to devise one made from mercury and popularized its use for the treatment of achalasia. Maloney modified the technique by giving the bougies a tapered end. This is the

only method whereby the dilators can be passed blindly, which has the advantage of allowing the patient to perform self-dilation at home. It is best used for milder soft strictures.

There are thus a number of techniques and a wide variety of instruments available for esophageal dilation. This underscores the fact that the procedure has not been well standardized, which perhaps is related to the high degree of individual variability. It needs to be done as often as necessary to provide symptomatic relief, which, of course, will vary. The optimal diameter for which dilation should strive is also not clear. Dilators are usually measured in French gauge, which is the circumference in millimeters. They usually range from No. 9 to No. 39 French, but larger ones are certainly available. Again, symptomatic relief is the goal.

Failure is generally defined in terms of the failure to alleviate the dysphagia, or if the dilation has to be performed too often, a schedule that needs to be determined by the patient and his physician. Certainly, caution and extreme patience are the rule.

Perforation. Esophageal perforation may result from a variety of causes, but it most commonly occurs secondary to endoscopic manipulation and dilation. It is certainly the most common serious complication of dilation. In the series by Michael and co-workers, 68 per cent of their perforations were iatrogenic versus 13 per cent that were spontaneous, 11 per cent that were secondary to foreign body ingestion, and 8 per cent that were secondary to external trauma.

When performing endoscopy, the most common sites of injury are the cervical esophagus at the cricopharyngeus muscle and the thoracoabdominal esophagus at the diaphragmatic hiatus. The former tends to occur because of improper introduction of the scope into the pyriform sinus, narrowing of the upper esophageal sphincter, or prominent cervical osteophytes. This area tends to be poorly visualized with fiberoptic esophagoscopy. The latter tends to occur as the esophagus angles anteriorly toward the diaphragm, which is often not appreciated by the endoscopist.

There is some controversy as to whether fiberoptic or rigid endoscopy is safer. In actual fact, the incidence of perforation from diagnostic esophagoscopy is relatively low, reported to be around 0.11 per cent with rigid instruments and 0.025 per cent with flexible instruments. Much depends upon the experience of the endoscopist and the anatomy of the patient.

Dilation, in contrast, carries a much greater risk. In this case, perforation tends to occur at the site of the lesion, and the incidence will depend upon the type of stricture and method of dilation. Chronic lye strictures and malignant strictures are generally associated with a higher likelihood of perforation.

Pneumatic dilation for achalasia carries the highest risk of perforation, ranging from 1 to 5 per cent. The Maloney and Hurst dilatores have the lowest risk, from 0.1 to 0.4 per cent, and the Eder-Puestow dilators fall in between, with a risk of 0.3 to 0.6 per cent. Complications are avoided by proceeding cautiously and advancing dilator size gradually. Fluoroscopic control may be helpful in difficult cases and certainly when performing pneumatic dilation.

When esophageal perforation occurs, it is of course important to make the diagnosis rapidly. Symptoms will vary depending upon the location of injury. Persistent pain after esophagoscopy or dilation should alert the physician to its possibility.

Perforations of the cervical esophagus generally produce neck pain (95 per cent) and cervical crepitance (55 per cent). Fever within the first 24 hours and leukocytosis typically occur. Thoracoabdominal perforations will produce retrosternal or occasionally pleuritic chest pain and shoulder pain if there is diaphragmatic irritation. Fever and leukocytosis also will follow.

Diagnosis is best confirmed radiographically. A plain chest x-ray film may show a thin streak of prevertebral air, subcutaneous or cervical emphysema, mediastinal widening, or pneumomediastinum. During the first 12 to 24 hours, a left pleural effusion and pulmonary infiltrates may develop. A contrast study should be obtained with a water-soluble agent such as meglumine diatrizoate (Gastrografin), which tends to cause less mediastinal inflammation. If the results are negative, however, it should be followed with a barium study, which remains somewhat more sensitive.

The treatment for esophageal perforation continues to be highly controversial, revolving largely around the adequacy of medical versus surgical therapy. Certainly there is ample documentation in the literature to support either approach. Medical therapy generally includes high-dose antibiotics, nasogastric suction, intravenous hydration and alimentation, and of course nothing by mouth. Surgical therapy usually involves external drainage and primary closure of the perforation if it is less than 24 hours old.

Perforations of the cervical esophagus are usually associated with less morbidity and mortality, and conservative therapy is generally appropriate. In contrast, quoting a mortality rate as high as 15 per cent, Shockley an colleagues advocate the need for surgical drainage. Certainly cases will need to be individualized, and any patient showing signs of sepsis should be treated more aggressively. Whether or not surgery is ultimately necessary, medical therapy should begin as soon as possible in every instance, since it has been well documented that mortality increases with delays in diagnosis and treatment.

Regarding perforation of the thoracic esophagus, most would seem to favor surgical drainage. However, Wesdorp and associates differentiate between instrumental perforations and those secondary to other causes, since the former tend to be recognized early before major contamination has occurred. As part of their treatment, they recommend nasoesophageal suction, using a catheter with multiple holes positioned above and below the perforation site. They reported a mortality rate of 6 per cent in 49 patients treated in this fashion and recommended conservative therapy for instrumental perforation provided that it was detected within 24 hours and not located below the diaphragm.

Once again, treatment will need to be individualized, but it should not be delayed. The clinician should maintain a high index of suspicion and if in doubt should initiate therapy prior to actual documentation of a perforation. Avoiding this complication is best accomplished by thorough familiarity with the anatomy, exercising extreme caution during instrumentation, and never performing blind manipulation.

Esophageal Neoplasms

Benign Tumors. These tumors of the esophagus are distinctly uncommon, making up 0.5 to 0.8 per cent of all esophageal neoplasms. Probably a good number go undetected, since they are often asymptomatic and are picked up only as incidental findings on x-ray films or endoscopy. Postmortem studies, however, corroborate this small incidence.

These benign lesions can be readily broken down into nonepithelial, epithelial, and heterotopic tumors (Table 13). By far the most common is the leiomyoma, which accounts for approximately 36 per cent of the benign neoplasms; as a group, the various myomas account for more than 50 per cent.

Table 13. Benign Esophageal Tumors

I. Nonepithelial tumors
- A. Myomas
  1. Leiomyomas
  2. Fibromyomas
  3. Lipomyomas
  4. Fibromas
- B. Vascular tumors
  1. Hemangiomas
  2. Lymphangiomas
- C. Mesenchymal and other tumors
  1. Reticuloendothelial tumors
  2. Lipomas
  3. Myxofibromas
  4. Giant cell tumors
  5. Neurofibromas
  6. Osteochondromas

II. Epithelial tumors
- A. Papillomas
- B. Polyps
- C. Adenomas
- D. Cysts

III. Heterotopic tumors
- A. Gastric mucosal tumors
- B. Melanoblastic tumors
- C. Sebaceous gland tumors
- D. Granular cell myoblastomas
- E. Pancreatic gland tumors
- F. Thyroid nodules.

Esophageal leiomyomas occur predominantly in men and occur at a much younger age than is found in carcinoma. Since they arise from smooth muscle, they are most commonly found in the distal two thirds of the esophagus. They tend to develop in the outer muscular coat, as opposed to the muscularis mucosae, and remain intramural and generally do not disrupt the mucosa. They are usually well circumscribed and oval and may be multiple,

although this is uncommon.

Many of these patients are asymptomatic, but as the tumor grows, dysphagia will often develop, which tends to be slowly progressive, occurs with solid food, and is often associated with substernal pain. If the tumor reaches a large size, tracheal or bronchial compression may occur, with secondary cough, wheezing, or dyspnea. Since the overlying mucosa usually remains intact, ulceration and bleeding are very unusual, in contrast to gastric leiomyomas.

On barium esophagram, a smooth, crescent-shaped mass is noted, with no evidence of ulceration or irregularity. A sharp demarcation between the tumor and the adjacent mucosa is clearly evident on the lateral view. The mass will be seen to move with swallowing, which verifies it is not fixed to surrounding structures. By the same token, esophagoscopy will reveal a freely movable mass compressing the esophageal lumen, with normal-appearing mucosa. Palpation will reveal a rubbery consistency. Since the lesion is submucosal, biopsy would be difficult and is contraindicated, as it may lead to subsequent perforation.

In many cases, no specific therapy is necessary. Malignant degeneration has been reported but is extremely rare. If the patient is symptomatic, conservative enucleation through an external approach is the procedure of choice.

Polypoid lesions are perhaps the next most common benign tumor seen in the esophagus. The fibrovascular polyp in particular is composed of a variable amount of fibrous connective tissue, vascular tissue, and adipose tissue, covered by squamous epithelium. It is intraluminal and usually pedunculated, most frequently arising in the upper portion of the esophagus. It varies in size but may actually grow quite large and, depending upon the length of its stalk, may actually be regurgitated into the mouth. This can be followed by its aspiration and subsequent asphyxiation. For that reason, these tumors should always be excised, which can be done endoscopically with very little chance of recurrence.

Malignant Tumors. These tumors of the esophagus are far more common than benign tumors, but they make up only 1.1 per cent of all cancers. They usually occur between the ages of 50 and 70 years and are more frequent in men, particularly black men. Although the prevalence of malignant esophageal neoplasms in the United States is estimated at 10/100.000 population (especially in the southeastern United States), a greater predilection has been demonstrated in other geographic areas, notably Japan, Iran, and certain regions of China.

No obvious environmental factors have been linked to this geographic preference. Certainly, excessive alcohol and tobacco use are well-known risk factors, although the precise pathophysiologic mechanism is debated. Nutritional deficiencies have been suggested, but the only clear relationship is seen in the Plummer-Vinson syndrome, in which there is a greater risk for developing postcricoid and cervical esophageal carcinomas. Chronic esophagitis of any cause seems to predispose to squamous cell carcinoma of the esophagus; therefore, increased risk has been noted with caustic injury, long-standing achalasia, and some of the connective tissue disorders. As mentioned earlier, Barrett's esophagus is associated with a higher incidence of adenocarcinoma.

Esophageal carcinoma may grow in a polypoid fashion and obstruct by means of a mass effect, or it may infiltrate and circumferentially constrict the lumen. There is typically

rather extensive submucosal spread. There appears to be a predilection for the middle third of the esophagus, at the level of the tracheal bifurcation and aortic arch, with prevalence in the upper and lower thirds being about equal.

Squamous cell carcinoma accounts for 90 to 95 per cent of esophageal malignancies, whereas primary adenocarcinoma accounts for most of the remainder (5 to 10 per cent). Adenocarcinoma may develop anywhere within the esophagus, most likely from embryonal columnar remnants or glandular elements. However, it is most commonly seen in the lower esophagus arising in a Barrett's columnar metaplasia or secondary to extension of a gastric fundus carcinoma.

Other tumors, such as carcinosarcoma, spindle cell sarcoma, leiomyosarcoma, adenoid cystic carcinoma, and lymphoma may occur but are exceedingly rare. Leiomyosarcoma is the most common nonepithelial malignant tumor of the esophagus. Primary malignant melanoma may also occur and in fact is more common than the sarcomas; however, it is found more often to be a metastatic lesion.

Aside from malignant melanoma, metastases to the esophagus are rarely reported, most commonly occurring with adenocarcinoma of the breast. Tumor may involve the esophagus secondarily by direct extension or extension from mediastinal lymph node metastases, or less commonly by direct hematogenous spread. These tumors usually surround the esophagus, producing a short segmental narrowing while the mucosa remains intact.

In general, patients with esophageal carcinoma present with advanced disease. Symptoms initially tend to be mild and easily ignored, and it is only after significant narrowing has occurred that the patient seeks medical attention. The average time between onset of symptoms and diagnosis is roughly 6 months. The tumor grows rapidly, and since the esophagus has no serosal layers, it spreads readily to adjacent structures and lymphatics. The lymphatic drainage follows the segmental blood supply, which was discussed earlier in this chapter. Therefore, lesions of the upper third of the esophagus drain to lower internal jugular and supraclavicular nodes; the middle third drains to mediastinal nodes, including paratracheal, hilar, and paraesophageal nodes; and the lower third drains to retroperitoneal celiac, and left gastric nodes. Nevertheless, metastases may readily skip nodal groups so that 44 per cent of midesophageal and 10 per cent of upper esophageal lesions are found to have celiac node involvement. Visceral sites of spread include most commonly the lung and liver.

When patients do present, dysphagia is the most common symptom, and it will vary in severity. However, it will be characterized as having been chronic, persistent, and beginning with certain solid foods but gradually becoming worse. The patient may have adapted the diet or eating habits to compensate, and it is important to elicit this sort of information.

Many patients will complain of a dull, substernal pain that will occur intermittently with swallowing or will be persistent. Weight loss is generally significant and may be associated with an iron deficiency anemia from occult bleeding. Chronic aspiration or even tracheoesophageal fistula secondary to tumor erosion may occur, causing pulmonary complications. Hoarseness of course can occur from compression of the recurrent nerve.

The diagnosis is usually apparent in a barium esophagram, which may show an exophytic mass creating a filling defect, a narrowed, irregular segment, or an annular constricting mass. Early lesions may require a double-contrast technique for better delineation.

Whenever there is suspicion of an esophageal mass, endoscopy should be performed. This allows for a tissue diagnosis and helps to assess the extent of disease. Although perforation is unlikely in the hands of an experienced endoscopist, one should never force the instrument through an area of narrowing. If the lesion is not clearly seen, brushings for cytologic examination might be helpful, which has an accuracy reportedly ranging from 75 to 90 per cent. It is traditionally performed under direct vision but can be done indirectly or by blind lavage, and it has been recommended as a screening procedure in high-risk patients.

Once the diagnosis is made, it is important to assess the extent of disease. This will usually require evaluation by at least CT scan and bone scan. A full endoscopic evaluation of the upper aerodigestive tract will be necessary to evaluate for tracheobronchial invasion and to rule out the presence of a second primary cancer. Penetration of the esophageal muscular wall and lymphatic spread have each been shown in a significant fashion to adversely affect survival. Conversely, tumor size per se, based on x-ray studies and degree of tumor differentiation, do not seem to affect prognosis.

As previously stated, the majority of patients present with advanced lesions so that on the average only 30 per cent are actually considered resectable and curable. For this reason, despite advances in surgical techniques and radiation therapy and the constant investigation of new chemotherapeutic drugs, cure rates for esophageal cancer remain extremely low. Five-year survival rates with combination surgery and radiation therapy are generally in the range of 15 to 25 per cent, with chemotherapy as yet effecting little change.

This, of course, emphasizes the need for accurate screening methods to detect early disease. It also means that palliation becomes a very real issue in many of these patients. The primary goal of any palliative approach when dealing with esophageal carcinoma will be to alleviate the dysphagia, with the least possible morbidity. Unfortunately, it is difficult if not impossible to make specific comparisons of different methods of palliation, since there are no well-controlled studies in this regard. The method of choice will often depend upon the expertise of the particular physicians involved, as well as the general condition of the patient.

One of the simplest options is to bypass the obstruction with a gastrostomy and pharyngostomy. However, it is certainly preferable to maintain the continuity of the gastrointestinal tract. Therefore, an alternative would be palliative resection or intestinal bypass, both of which carry a high degree of morbidity but may be reasonable in some patients.

Radiation therapy may be effective in some instances and has the advantage of being noninvasive, although it can cause mucositis and injury to mediastinal structures. Dilation alone is usually not effective because the dysphagia tends to be progressive, and constant dilation becomes necessary. It is helpful, however, when used in conjunction with other modalities, such as radiation therapy. A number of indwelling stents have been developed to maintain esophageal patency following dilation. They are generally made of silicone in a variety of shapes to facilitate push-through from above or pull-through from below and to

prevent migration. In addition, the neodymium-yttrium aluminium garnet (Nd-YAG) laser has been applied endoscopically to re-establish the esophageal lumen, with few complications in experienced hands.