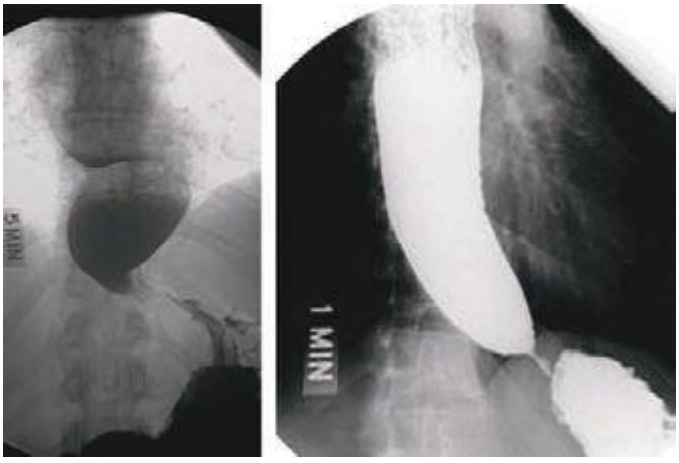

FIGURE 347-5 Achalasia with esophageal dilatation, tapering at the gastroesophageal junction, and an air-fluid level within the esophagus. The example on the *left* shows sigmoid deformity with very advanced disease.

The only durable therapies for achalasia are pneumatic dilatation and Heller myotomy. Pneumatic dilatation, with a reported efficacy ranging from 32–98%, is an endoscopic technique using a noncompliant, cylindrical balloon dilator positioned across the LES and inflated to a diameter of 3–4 cm. The major complication is perforation with a reported incidence of 0.5–5%. The most common surgical procedure for achalasia is laparoscopic Heller myotomy, usually performed in conjunction with an antireflux procedure (partial fundoplication); good to excellent results are reported in 62–100% of cases. A European randomized controlled trial demonstrated an equivalent response rate of approximately 90% for both pneumatic dilation and laparoscopic Heller myotomy at 2-year follow-up. Occasionally, patients with advanced disease fail to respond to pneumatic dilatation or Heller myotomy. In such refractory cases, esophageal resection with gastric pull-up or interposition of a segment of transverse colon may be the only option other than gastrostomy feeding.

An endoscopic approach to LES myotomy has been introduced, referred to as per oral esophageal myotomy. This technique involves the creation of a tunnel within the esophageal wall through which the circular muscle of the LES and distal esophagus are transected with electrocautery. Short-term studies of efficacy have been favorable. Potential advantages over the conventional laparoscopic approach include avoidance of surgical disruption of the diaphragmatic hiatus and more rapid recovery.

In untreated or inadequately treated achalasia, esophageal dilatation predisposes to stasis esophagitis. Prolonged stasis esophagitis is the likely explanation for the association between achalasia and esophageal squamous cell cancer. Tumors develop after years of achalasia, usually in the setting of a greatly dilated esophagus with the overall squamous cell cancer risk increased 17-fold compared to controls.

DIFFUSE ESOPHAGEAL SPASM (DES)

DES is manifested by episodes of dysphagia and chest pain attributable to abnormal esophageal contractions with normal deglutitive LES relaxation. Beyond that, there is little consensus. The pathophysiology and natural history of DES are ill defined. Radiographically, DES has been characterized by tertiary contractions or a "corkscrew esophagus" (Fig. 347-7), but in many instances, these abnormalities are actually indicative of achalasia. Manometrically, a variety of defining features have been proposed including uncoordinated ("spastic") activity in the distal esophagus, spontaneous and repetitive contractions, or high-amplitude and prolonged contractions. The current consensus, derived from high-resolution manometry studies, is to define spasm by the occurrence of contractions in the distal esophagus with short latency relative to the time of the pharyngeal contraction, a dysfunction indicative of impairment of inhibitory myenteric plexus neurons.

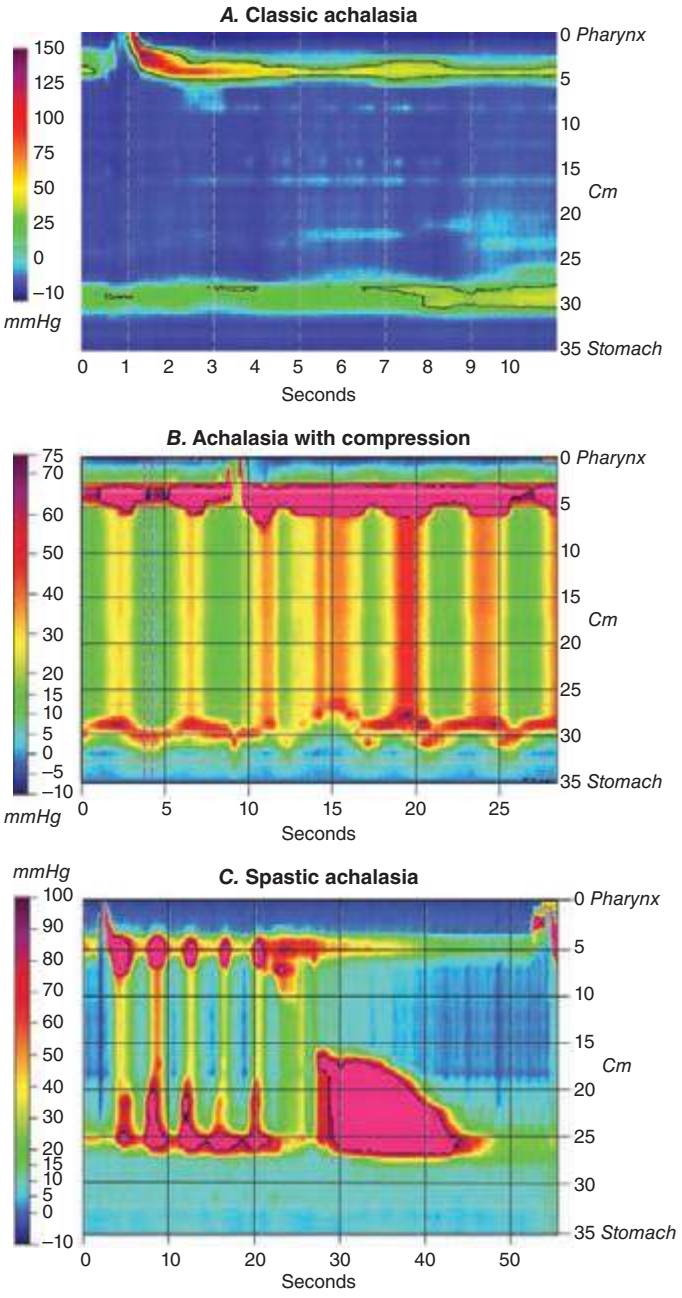

FIGURE 347-6 Three subtypes of achalasia: classic (*A*), with esophageal compression (*B*), and spastic achalasia (*C*) imaged with pressure topography. All are characterized by impaired lower esophageal sphincter (LES) relaxation and absent peristalsis. However, classic achalasia has minimal pressurization of the esophageal body, whereas substantial fluid pressurization is observed in achalasia with esophageal compression, and spastic esophageal contractions are observed with spastic achalasia.

When defined in this restrictive fashion (Fig. 347-8), DES is actually much less common than achalasia.

Esophageal chest pain closely mimics angina pectoris. Features suggesting esophageal pain include pain that is nonexertional, prolonged, interrupts sleep, meal-related, relieved with antacids, and accompanied by heartburn, dysphagia, or regurgitation. However, all of these features exhibit overlap with cardiac pain, which still must be the primary consideration. Furthermore, even within the spectrum of esophageal diseases, both chest pain and dysphagia are also characteristic of peptic or infectious esophagitis. Only after these more common entities have been excluded by evaluation and/or treatment should a diagnosis of DES be pursued.