A. INTRODUCTION

Achalasia is an uncommon motility disorder of the esophagus with a prevalence in the U.S. of <0.001% (Mayberry 2001). Primary, or idiopathic, achalasia includes the two categories of classic and vigorous distinguished by the amplitude of distal esophageal contractions on manometry. The cause of primary achalasia is unknown, but destruction of ganglion cells in Auerbach’s myenteric plexus is the pathologic hallmark leading to impaired relaxation of the LES. Esophageal body aperistalsis is believed to be a secondary event, though is a mandatory manometric finding to establish the diagnosis. Patients with idiopathic primary achalasia typically present between the ages of 20 and 40 years with an equal male to female distribution. Patients with secondary achalasia have similar symptoms and aperistalsis, with causes including Chagas’ disease, idiopathic intestinal pseudo-obstruction, infiltrative connective tissue disorders, diabetes and pseudo-achalasia from malignancy involving the region of the esophagogastric junction (Portale 2007).

The term pseudo-achalasia is frequently used interchangeably in the literature with “secondary,” to include not only cancer-related achalasia but also post-surgical loss of LES compliance, such as after a vagotomy or improperly constructed Nissen fundoplication. Chagas’ disease, a frequently encountered condition in South America, is the most common cause of secondary achalasia worldwide. In this disease, Trypanosoma
cruzi infects the ganglion cells resulting in inflammation and destruction (de Oliveira 1998). Patients with Chagas’ disease demonstrate esophageal body aperistalsis and LES nonrelaxation, but typically have a lower LES resting pressure than do patients with idiopathic achalasia because of differences in neuronal impairment (Dantas 1990).

B. DIAGNOSIS

Progressive dysphagia to both solids and liquids is the symptomatic hallmark of achalasia, occurring in more than 80% of patients, while regurgitation occurs in two-thirds (Bedgood 2007, Spiess 1998). Chest pain is a variable finding with an unpredictable response to treatment. Frequently, the diagnosis of achalasia is suggested by an upper endoscopy or barium esophagogram performed for dysphagia. Endoscopy is important to exclude the possibility of an intramural tumor causing pseudo-achalasia. The barium esophagogram typically demonstrates loss of peristalsis in the smooth muscle of the distal two-thirds of the esophagus, as well as the classic “bird’s beak” tapering at the LES or an epiphrenic diverticulum. A mega- or sigmoid esophagus may be detected and could change the treatment planning and patient discussion to include potential esophagectomy. A timed barium esophagogram has limited availability, though several centers have used this simple test for assessing the success of treatment (de Oliveira 1997; Vaezi 2002; Gockel 2005; Guardino 2004). While achalasia may be suspected based on endoscopic and radiographic findings, esophageal manometry establishes the diagnosis by showing failure of LES relaxation and esophageal body aperistalsis. Only about 60% of patients have an elevated resting LES pressure (Kraichely 2006,) so a
hypertensive LES may only support the diagnosis and help eliminate scleroderma from the differential diagnosis.

C. SURGICAL THERAPY

Before the wide-spread availability of minimally invasive myotomy, surgical treatment commonly was reserved for medical failures after patients had undergone one or more pneumatic dilations (Kadakia 2001; Parkman 1993). Dilation is associated not only with a risk of perforation, but also with the potential for recurrent dysphagia from inadequate therapy or the development of reflux symptoms from LES incompetence (Okike 1979). Given the better long-term symptom relief, low morbidity and short length of stay associated with laparoscopic Heller myotomy, surgery is now the favored treatment for primary achalasia in most centers.

The original esophageal myotomy, described by Heller in 1913, involved creating an anterior and posterior cardiomyotomy. The technique was modified by Zaaijer in 1923 to simply an anterior myotomy (Zaaijer, Frantzides, Payne) and has been performed via both transthoracic and transabdominal approaches. In 1992, Pellegrini reported the first series of minimally invasive thoracoscopic esophagomyotomy in 15 patients (Pellegrini 1992). The mean LES pressure decreased from 32 to 10 mm Hg in their patients after surgery. An excellent symptom response was noted in 70% of the patients, though the important conclusion was that the procedure was safe and feasible. A video-assisted thoracoscopic surgery (VATS) approach is not commonly utilized at present due to difficulties achieving an adequate distal myotomy and the reported experience with
persistent dysphagia, though may be useful if an intrathoracic esophageal diverticulum requires excision.

In the past 15 years, with the increased competence of surgeons in performing minimally invasive surgery, the gold standard treatment for achalasia has become a laparoscopic modified Heller myotomy. After a single case report from Scotland in 1991 of a laparoscopic cardiomyotomy for achalasia (Shimi S 1991), a series of 40 patients treated with laparoscopic cardiomyotomy was reported in the U.S. by John Hunter six years later (Hunter 1997). Recent operative controversies have included the myotomy length and the need for a concomitant antireflux procedure. When a modified Heller myotomy is performed via a left VATS and the phrenoesophageal ligament remains intact, an antireflux procedure should not be necessary. With a transabdominal Heller myotomy, however, the diaphragmatic attachments around the LES, critical to the antireflux mechanism, are disrupted and a hiatal hernia may be created, predisposing the patient to gastroesophageal reflux. Addition of a routine antireflux procedure is controversial because of the presence of an aperistaltic esophagus and concerns that a fundoplication will add to esophageal outflow obstruction.

Myotomy length

A longer extent of myotomy on to the gastric cardia (2-3 cm) appears to reduce persistent or recurrent symptoms of dysphagia or regurgitation. The extended myotomy recently endorsed by Pellegrini’s group involves lengthening the myotomy more than 3 cm on to the stomach (Wright 2007). An important technical maneuver is elevation of the anterior esophagogastric junction fat pad to provide accurate identification of the junction, facilitate completion of the myotomy and protect the anterior vagus nerve from
iatrogenic injury. Most surgeons will extend the myotomy proximally as far as can be visualized via laparoscopy. The overall myotomy length should be in the range of 6-9 cm.

A method for assessing completeness of myotomy is to perform intraoperative manometry with measurement of LES pressure (Chapman 2004). In a series of 132 patients, 34% had persistently elevated pressures after an initial myotomy, allowing immediate extension with a short-term postoperative success rate of 93% as assessed at one month. Such results, however, may represent surgeon inexperience at performing a myotomy of adequate length. The use of intraoperative manometry is a good educational tool, but is cumbersome and not typically available to surgeons. Probably a more common way of assessing the completeness of myotomy is with intraoperative endoscopy, noting the ease with which an endoscope can be passed across the gastroesophageal junction. Of course, this assessment must be carried out carefully so as to prevent inadvertent mucosal perforation at the myotomy site. Many surgeons perform no formal intraoperative assessment of the myotomy, other than assuring that the myotomy appears to extend in an adequate fashion both proximally and distally as based on external esophagogastric landmarks.

**The role of complete fundoplication**

In several early series from the minimally invasive era, and against conventional wisdom in the setting of an aperistaltic esophagus, a complete 360-degree (Nissen) fundoplication was performed with a laparoscopic Heller myotomy. In one review of 42 patients treated with various approaches, nine patients underwent a Heller and a “floppy” Nissen over a 40 French bougie. At a mean follow-up of 8.5 years in this small group of
patients, 22% (2/9) of patients complained of dysphagia to meat or bread and 30% complained of reflux, although there was no objective evidence of GERD on 24-hour ambulatory esophageal pH monitoring (Jordan 2001). Another study of patients in Australia included 49 who underwent a laparoscopic myotomy with Nissen fundoplication and 13 a myotomy with partial anterior fundoplication (Wills 2001). The dysphagia scores at 5 years for these two groups of patients trended toward less dysphagia in the partial fundoplication group (dysphagia score difference, p=0.08). The authors concluded that although partial fundoplication resulted in less dysphagia and chest pain compared to a complete wrap, there was no evidence to suggest that a total fundoplication was associated with a worse overall outcome. The authors recommended a controlled, randomized trial to answer the controversy (Wills 2001).

The third report of a “floppy” Nissen combined with myotomy was a recent review by Frantzides and colleagues (Frantzides 2004) covering their ten-year experience dating back to 1992. In this retrospective study of 53 patients, 48 (90%) had a concomitant Nissen fundoplication with the laparoscopic myotomy. After performing a long myotomy with up to 7 cm on the cardia, a 50 French bougie was placed and the 360-degree wrap completed. Laxity of the wrap was assessed by placing a 1 cm instrument next to the esophagus. At a median 3-year follow-up, 2/48 (4%) of the Nissen patients had dysphagia as determined by a modified Visick scoring system. Persistent dysphagia was attributed to an incomplete myotomy in one case and an excessively tight wrap in the other. The authors attributed their low postoperative dysphagia rate to the use of a lighted bougie, which allowed performance of a complete myotomy with improved visualization and division of intact muscle fibers.
Several series have evaluated Heller myotomy with and without partial fundoplication, with most of the reports involving an anterior or Dor fundic wrap (Table) (Wills 2001; Patti 2001; Luketich 2001; Donahue 2002; Lyass 2003; Wright 2007; Richards 2004). One study looking at quality of life (QOL) and dysphagia after myotomy used an SF-36 QOL and dysphagia scoring system (Youssef 2007). The authors found that, at an average follow-up of 3.3 years, the mean dysphagia score was not statistically worse when a Dor was added to the myotomy. There was also no difference in the mean SF-36 scores in the two groups at 3 years, as the 100 total respondents who underwent a Heller with or without a Dor had an equal satisfaction rate of 92%. Although the authors did not report rates of GERD in their cohorts, a reasonable conclusion from the data is that dysphagia is not a significant long-term complication after addition of a partial anterior fundoplication.

Due to the controversy about whether an antireflux procedure routinely should be added to a myotomy, a prospective, randomized clinical trial of myotomy with and without Dor was completed at Vanderbilt University and reported in 2004 (Richards 2004). In this study, 43 patients undergoing laparoscopic myotomy for achalasia were randomized to Dor versus no Dor. Patients underwent manometry and 24-hour esophageal pH monitoring 3-5 months after surgery. Pathologic reflux was significantly less in the Dor group (9% vs. 48% in patients without the Dor). In addition, distal esophageal acid exposure was significantly higher in the Heller-only group. Dysphagia was not a frequent long-term complication, with similar dysphagia scores noted between the two groups.
Fewer total patients have been studied following the use of a partial posterior, or Toupet, fundoplication as the antireflux procedure following myotomy. An advantage of the Toupet is that the fundic wrap is secured to the myotomy edges, exerting a distracting force that may minimize the risk of recurrent achalasia. On the other hand, an anterior fundoplication is technically easier to perform in that no posterior dissection is required and short gastric vessel division may not be necessary. In addition, the myotomy is covered by the fundic wrap, potentially buttressing any unsuspected mucosal microperforations.

The only report directly comparing Dor and Toupet partial fundoplications comes from the University of Washington (Wright 2007). The authors, however, compared two non-contemporaneous cohorts, the first from 1994-1998 utilizing a standard myotomy (1-2 cm on the cardia) with a Dor fundoplication while the second, from 1998-2003, underwent an extended myotomy (>3 cm on the cardia) with a Toupet fundoplication. Using telephone surveys, they determined that dysphagia severity was lower in the extended myotomy and Toupet group, although the frequency of dysphagia was similar in both cohorts. Postoperative heartburn and regurgitation frequencies were similar with the two different approaches. The contributions of the extended myotomy, the use of a posterior partial fundoplication, and surgeon experience to the improved results in the later group are difficult to discern from the data.

Other studies looking at fundoplication and rates of postoperative dysphagia did not separate Dor from Toupet, Nissen or even Belsey patients, so one can only conclude that rates of dysphagia after partial fundoplication range from 2 – 38% (Table). In the University of Pittsburgh report, a patient who complained of any degree of
dysphagia at a mean follow-up of 19 months was considered as having persistent
dysphagia, contributing to their high (38%) postoperative dysphagia rate. More
importantly, though, the dysphagia severity score was significantly improved in >95% of
patients and >92% of patients were satisfied with the operation (Luketich 2001). These
data highlight the difficulty in assigning success or failure to a treatment that is palliative
in nature and for which the results are subjectively defined.

Although the goal of an antireflux procedure with the myotomy is to prevent
postoperative symptomatic and objective reflux and associated complications of
esophagitis, the randomized study by Richards, et al provides the best evidence that the
heartburn rates improved with a Dor fundoplication compared to no fundoplication. The
rates of heartburn following fundoplication have ranged from 8-26% (Table ).

**Robotic-assisted myotomy**

Although robotic surgery entered the surgical arena in 1994 with FDA-approval of the
AESOP robotic arm to control a laparoscope (Computer Motion, Goleta, CA), the results
of robotic- or computer-assisted advanced minimally invasive procedures began to fill the
The first surgical system approved by the FDA was the da Vinci robot in 2000 (Intuitive
Surgical Inc., Sunnyvale, CA). From several large series of minimally invasive
procedures, surgical outcomes utilizing robotics have compared favorably to those
following conventional laparoscopic techniques. The robotic approach appears feasible,
safe and increasingly available, while the appropriate indications continue to be clarified
and refined. Advantages of robotic-assisted surgery include improved imaging utilizing
three-dimensional and high-definition optics, articulating instruments with multiple
degrees of freedom to enhance precision and accuracy of surgical maneuvers, and the
ability to filter tremor and scale movements. The primary disadvantages of the robot
include expense, technical training of operators, bulk of the equipment, loss of tactile
sensation, and the fact that the surgeon is separated from the patient and positioned
outside of the sterile operative field.

Use of the robot for laparoscopic antireflux procedures has not improved operative
times or outcomes over traditional laparoscopic approaches (Cadiere, Himpens et al
2001; Melvin 2002). For patients with achalasia, however, advantages of the robot for a
laparoscopic myotomy include three-dimensional imaging, which may reduce the risk of
esophageal perforation. A large series was reported of 104 patients who underwent a
robotic Heller myotomy with partial fundoplication at three institutions (Melvin 2005).
The authors cited an unacceptably high incidence of intraoperative esophageal
perforation complicating standard laparoscopic myotomy of up to 15% as reported in the
literature. This high perforation rate, however, was observed in the first series of
laparoscopic Heller reported in 1997 (Hunter 1997). The average rate from over 600
patients in nine series reported subsequently was less than 10% (Melvin 2005, Galvani
2006). Considering the fact that perforation infrequently results in conversion to an open
approach and is typically remedied with delicate suturing and patching with the
fundoplication, one would not consider this risk a strong indication for performing
myotomies with a robot. On the other hand, a postoperatively diagnosed or partially
treated perforation can result in significant morbidity and even mortality for the patient.
On that note, in the series of 104 patients treated with robot-assisted myotomy, no
esophageal perforations occurred. In addition, significant improvement in dysphagia was
noted in all 79 patients available for follow-up (Melvin 2005). Table x summarizes the results from several large series of robotic Heller myotomy. Some reports directly compared outcomes after robotic procedures to those following standard laparoscopic approaches. Importantly, robotic-assisted myotomy continued to be associated with no esophageal perforations (Melvin 2005; Galvani 2006; Iqbal 2006; Huffman 2007).

D. CONCLUSIONS

Idiopathic achalasia is an uncommon primary motility disorder of the esophagus. Laparoscopic modified Heller esophagocardiomyotomy has become the standard treatment for this benign but incurable disease. Perioperative mortality approaches 0% and long-term patient satisfaction exceeds 90%. The optimal length of the cardiomyotomy on to the stomach is two to four cm. An antireflux procedure can reduce postoperative heartburn rates by 80% and reduce the risk of esophagitis and peptic stricture. A Dor or Toupet partial fundoplication reduces reflux and should be a routine adjunct to laparoscopic myotomy; the choice between the two is a matter of surgeon preference. The robotic Heller myotomy may result in a lower intraoperative esophagogastric perforation rate but is not widely utilized.