



Surgical management of esophageal achalasia: Evolution of an institutional approach to minimally invasive repair



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ABSTRACT

Background: Surgical management of esophageal achalasia (EA) in children has transitioned over the past 2 decades to predominantly involve laparoscopic Heller myotomy (LHM) or minimally invasive surgery (MIS). More recently, peroral endoscopic myotomy (POEM) has been utilized to treat achalasia in children. Since the overall experience with surgical management of EA is contingent upon disease incidence and surgeon experience, the aim of this study is to report a single institutional contemporary experience for outcomes of surgical treatment of EA by LHM and POEM, with regards to other comparable series in children.

Methods: An IRB approved retrospective review of all patients with EA who underwent treatment by a surgical approach at a tertiary US children's hospital from 2006 to 2015. Data including demographics, operative approach, Eckardt scores pre- and postoperatively, complications, outcomes, and follow-up were analyzed.

Results: A total of 33 patients underwent 35 operative procedures to treat achalasia. Of these operations; 25 patients underwent laparoscopic Heller myotomy (LHM) with Dor fundoplication; 4 patients underwent LHM alone; 2 patients underwent LHM with Thal fundoplication; 2 patients underwent primary POEM; 2 patients who had had LHM with Dor fundoplication underwent redo LHM with takedown of Dor fundoplication. Intraoperative complications included 2 mucosal perforations (6%), 1 aspiration, 1 pneumothorax (1 POEM patient). Follow-up ranged from 8 months to 7 years (8–84 months). There were no deaths and no conversions to open operations. Five patients required intervention after surgical treatment of achalasia for recurrent dysphagia including 3 who underwent between 1 and 3 pneumatic dilations; and 2 who had redo LHM with takedown of Dor fundoplication with all patients achieving complete resolution of symptoms.

Conclusions: Esophageal achalasia in children occurs at a much lower incidence than in adults as documented by published series describing the surgical treatment in children. We believe the MIS surgical approach remains the standard of care for this condition in children and describe the surgical outcomes and complications for LHM, as well as, the introduction of the POEM technique in our center for treating achalasia. Our institutional experience described herein represents the largest in the “MIS era” with excellent results. We will refer to alterations in our practice that have included the use of flexible endoscopy in 100% of LHM cases and use of the endoscopic functional lumen imaging probe (EndoFLIP) in both LHM and POEM cases which we believe enables adequate Heller myotomy.

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The incidence of esophageal achalasia in children is estimated to be 0.1–0.18 per 100,000 children per year [1] or a nearly a factor of 10 times less than that in adults [2]. The surgical approach in management of pediatric esophageal achalasia has progressed to exclusively involve minimally invasive surgery comprised of laparoscopic Heller's myotomy or a peroral endoscopic myotomy (POEM) techniques. Surgeons as well as gastroenterologists worldwide have increasingly adopted these approaches in the management of pediatric achalasia such that an open approach would no longer be the standard of care. Heller's myotomy offers a more definitive treatment of the primary

symptoms of esophageal achalasia which include dysphagia, weight loss, and regurgitation, when compared to pneumatic balloon dilation especially in younger patients [3,4]. Recently, dilation therapy has been found to require more frequent re-interventions for recurrent symptoms compared to primary Heller myotomy in children [5].

Although many of the published articles on the surgical management of pediatric achalasia describe the safety and efficacy of the MIS approach, a critical review of the literature reveals that there are significant operative complications and long-term issues in this population which have to be considered including intraoperative esophageal perforation, recurrent dysphagia, and need for repeat surgery or pneumatic dilations following Heller's myotomy.

In this report, we describe our institutional MIS experience with esophageal achalasia with regards to LHM and POEM procedures. The

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benefits of the use and adoption of flexible endoscopy during LHM and EndoFLIP during LHM and POEM will be described as adjuncts to MIS Heller's myotomy for ensuring adequacy of the esophageal myotomy and assessing for mucosal injury during these procedures.

1. Methods

An IRB (Institutional Review Board) approved retrospective study of all children who underwent surgical treatment for esophageal achalasia at single children's hospital from April 2006 to November 2015. Data including demographics, operative approach, Eckardt scores and weight changes pre- and postoperatively, complications, outcomes, and follow-up were evaluated. Mean values were analyzed using Student's *t*-test, and categorical data was compared using chi-square analysis. *P*-values <0.05 are considered statistically significant. Review of the literature was completed by searching PubMed utilizing the keywords 'achalasia in children' and peroral endoscopic myotomy'. There were 4 attending surgeons involved in the surgical management of the operative cases.

2. Diagnosis & surgical approach

All patients diagnosed with achalasia had at minimum, a timed barium esophagram demonstrating a narrowed lower esophageal sphincter and dilated proximal esophagus. About one-third also underwent esophageal manometry, or endoscopy with biopsy to contribute to the diagnosis. The majority of MIS cases were LHM with or without anterior fundoplication but beginning in 2015, patients were offered the POEM procedure as 2 surgeons (MP and TDK) had undergone prior training at an adult center to perform the technique. Demographics are shown in Table 1 with the most common symptoms being dysphagia, chest pain, and emesis.

2.1. Laparoscopic Heller myotomy

Depending upon surgeon preference, patients underwent laparoscopic Heller myotomy with or without partial anterior fundoplication (25 Dor, 2 Thal fundoplications). During laparoscopic myotomy, typically an esophageal myotomy was performed from the gastroesophageal junction (GEJ) proximally for at least 6 cm up onto the esophagus (5–8 cm). Distally, the myotomy was extended for 3 cm on the gastric side (2–4 cm). The myotomy was performed with hook electrocautery and blunt disruption of the longitudinal and circular muscle fibers down to the submucosal layer. In 26 cases, flexible endoscopy was performed in conjunction with Heller myotomy to ensure adequate release at the esophagogastric junction, as well as to assess for mucosal perforation. Since 2010, endoscopy has been utilized in 100% of cases. An anterior fundoplication (primarily Dor type) was done in 76% of cases where the stomach is attached to the edges of both sides of the myotomy anteriorly.

Table 1
Patient data & presentation

	Number	% (Range)
Gender		
Female	20	61%
Male	13	39%
Age of Diagnosis	11	(5–18 years)
Duration of Symptoms	2.8 years	(1–11 years)
	<i>n</i>	Percentage
Presenting symptoms		
Dysphagia	28	85%
Chest pain	19	58%
Emesis	17	52%
Weight loss	13	39%
Regurgitation	8	24%
Odynophagia	6	18%

2.2. Peroral endoscopic myotomy (POEM)

The POEM procedure is performed as originally described by Inoue [6] with the patient in supine position. Diagnostic endoscopy is performed to make measurements for landmarks such as GEJ, mucosotomy site, myotomy start/end landmarks and esophagus clear of debris. An Apollo OverTube™ is placed and a GIF-H180 Olympus endoscope with CO₂ insufflation is passed through this to perform the submucosal injection of 10 mL saline with mixture of methylene blue is placed at about 10–12 cm above the GEJ. A triangle knife is used to make a 2 cm longitudinal incision in the mucosa in the 2 o'clock position (mucosotomy). A submucosal tunnel is created down 3 cm past the GEJ. The myotomy is started 5–6 cm above the GEJ and continues 2–3 cm past the GEJ. The mucosotomy is closed with large clips (Boston Scientific).

Since 2014, we have used the functional lumen imaging probe (EndoFLIP; Crospon Ltd., Galway, Ireland) during both LHM and POEM procedures to aid in identifying the adequacy of myotomy using cross-sectional diameter measurements of the GEJ before and after myotomy as described by Teitelbaum et al. [7]. All patients underwent postoperative day number one esophagram (even patients who underwent concomitant intraoperative endoscopy) and there were no leaks demonstrated. The main purpose of postmyotomy contrast study is to obtain a baseline evaluation of the esophagus and GEJ in order to have a comparison study if the patients become symptomatic in the future.

3. Results

A total of 33 patients underwent 35 operative procedures to treat achalasia. For the initial operative procedures, the mean age at surgery was 12.9 years (5–18 years); length of stay was 1.9 days (1–6 days); and follow up 30.9 months (7–84 months) [Table 2]. Of the 35 MIS operative procedures; 25 patients underwent laparoscopic Heller myotomy (LHM) with Dor fundoplication (LHM + Dor); 4 patients underwent LHM alone; 2 patients underwent LHM with Thal fundoplication (LHM + Thal); 2 patients underwent primary POEM [Table 3]. Mean operative time in minutes and range for each procedure was 122 (97–180) for LHM; 137 (105–169) for LHM/Thal; 128 (45–213) LHM/Dor; and 212 (92–332) for POEM.

Preoperative and postoperative Eckardt scores were obtained for all operations (including 2 redo LHM). Fig. 1 demonstrates the graph of Eckardt scores for each patient, which shows improvement from baseline Eckardt score preoperative 9.7 (3–11) to 0.9 (0–3) postoperative Heller myotomy (*p* < 0.002). All Eckardt scores postmyotomy were less than 3, which is considered normal. The majority of patients (34 of 35) also demonstrated positive weight gain following MIS Heller myotomy, although not statistically significant; this may be considered as a surrogate marker for symptomatic relief.

Complications and treatment shown in Table 4 included 2 mucosal perforations (6%), 1 aspiration, 1 wound infection, and 1 pneumothorax (POEM procedure). There were 5 patients with recurrent dysphagia which required 1–3 dilations (3 patients) and 2 patients who underwent redo LHM with takedown Dor fundoplication (at 10 months and 7 years)

Table 2
Initial operative therapy

	Mean	Range
Age at surgery (years)	12.9	5–18
OR time (minutes)	113	45–352
LHM	122	97–180
LHM + DF	128	45–213
LHM + TF	137	105–169
POEM	212	92–352
Length of Stay (days)	1.9	1–6
Follow up (months)		7–84

LHM - laparoscopic Heller myotomy; DF - Dor fundoplication; TF - Thal fundoplication; POEM - peroral endoscopic myotomy.

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