Laparoscopic esophagomyotomy in children: is routine fundoplication necessary?
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**Purpose** Laparoscopic esophagomyotomy (LE) with or without fundoplication has been described as a modality for the treatment of achalasia in children. Our aim is to evaluate the safety and efficacy of LE without fundoplication in the management of achalasia in children.

**Patients and methods** A retrospective review of the medical record was carried out on patients with achalasia at our institution from January 2006 to March 2011. Eight children were diagnosed with esophageal achalasia and all of them underwent LE without fundoplication. These patients were reviewed to evaluate intraoperative and postoperative complications, as well as outcome.

**Results** Eight children with achalasia had LE without fundoplication successfully completed. There were four boys and four girls ranging in age between 1 and 13 years (mean 4.3 years). None of the patients had received therapy before LE. The mean operating time was 44 min. The mean follow-up was 40.3 months (range 6–62 months). There were no intraoperative or postoperative esophageal perforations or complications. The mean length of hospital stay was 2.7 days.

**Introduction**
Esophageal achalasia is a rare primary motor disorder of the esophagus, characterized by the triad of esophageal aperistalsis, failure of the lower esophageal sphincter (LES) to completely relax with swallowing, and a hypertensive or sometimes normal pressure LES [1–4]. The reported incidence of achalasia in the general population is four to six per 100,000, with 5% of patients being younger than 15 years of age [5]. Therefore, surgical experience in children is limited. A delay in diagnosis in children is common [6,7]. The diagnosis of achalasia in children may be delayed because of the rarity of the condition in this age group and the inability of younger children to describe their symptoms [7].

**Patients and methods**
From January 2006 to March 2011, eight children were diagnosed with esophageal achalasia, and all of them were treated with laparoscopic esophagomyotomy (LE) without fundoplication. Information collected for each patient included age, sex, medical and family history, presenting symptoms, diagnostic methods, previous treatments (such as pneumatic dilation, botulinum toxin injections, or prior surgery), surgical treatment, operative time, length of hospital stay, complications, postoperative investigations, and outcomes.

The research was approved by the ethical committee board.

None of the patients required redo esophagomyotomy or esophageal dilatation. Dysphagia symptoms improved uniformly and gastroesophageal reflux symptoms were not evident in follow-up.

**Conclusion** This technique of LE without fundoplication in the treatment of achalasia is safe, effective, and yields excellent cosmetic results. The routine addition of fundoplication to LE for the treatment of achalasia in our series appears to be unnecessary. Yet, a multicenter study with a large group of patients should be carried out. *Ann Pediatr Surg* 9:1–5 © 2013 Annals of Pediatric Surgery.

**Technique**
The child was placed in a supine lithotomy position. Younger children were placed in a frog-like position. The table was tilted in reversed Trendelenburg to allow the bowel to fall out of the way. The surgeon worked between the legs of the patient. The first 5 mm port was inserted just below the umbilicus using the open technique. The abdominal cavity was insufflated with carbon dioxide up to 12 to 15 mmHg.

A total of five ports were used: one positioned in the umbilicus for the telescope, one in high epigastrium to the right of the xiphoid for liver retraction, one in the left upper quadrant for the stomach retractor, and two more working ports were inserted into each subcostal lateral to the rectus muscle (Fig. 1). In infants, we used 3 mm ports and in older children, we used 5 mm ports. Yet, in older children, one 10 mm port may be used in the right subcostal to introduce a 4 × 4 gauze in order to dry the oozing of blood from the myotomy site. Retraction of the left lobe of the liver was carried out by the epigastric port. An appropriate-size bougie was introduced into the oropharynx and distal esophagus under laparoscopic vision. The stomach was pulled downward by the assistant to identify the gastroesophageal junction and to facilitate the dissection. Mobilization of the visceral peritoneal fold over the anterior esophagus was performed using an electrocautery hook. Minimal mobilization of the esophagus was performed anteriorly and laterally on each side. Identification, preservation, and protection of the anterior vagus nerve were ensured throughout the
dissection and myotomy. The site of the anterior esophageal myotomy was coagulated longitudinally using an electrocautery hook. The myotomy was begun on the anterior esophageal wall just above the gastroesophageal junction and continued above at least 6 cm along the anterior esophagus and then extended onto the stomach wall for 2 cm. Two Maryland dissecting forceps were used to grasp the edges of the esophageal muscles on each side and pulled on opposite directions to separate the esophageal muscles longitudinally. Subsequently, separation of the esophageal muscles was carried out by one Maryland holding one edge of the esophageal muscles and the second Maryland passed under the esophageal muscles to reach the submucosal plane. Hook diathermy was used to pick up and divide the circular fibers. Further separation and splitting of the esophageal muscles were carried out using two Marylands pulled out on opposite directions with preservation of the mucosa (Fig. 2). A flexible pediatric endoscopy was passed through the oropharynx and advanced into the myotomy site to check the integrity of the esophageal mucosa and determine the completeness of the myotomy. The endoscope was removed after aspirating air from the stomach. In two of our patients, approximation of right and left diaphragmatic crura by an intracorporeal 2–0 silk stitch was performed anteriorly (Fig. 3). All ports were removed after deflation of the abdomen. All ports sites were closed. A nasogastric tube was removed at the end of the procedure.

Results

Eight children with achalasia had LE without fundoplication successfully completed. There was no conversion to open surgery. There were four boys and four girls ranging in age between 1 and 13 years (mean 4.3 years). All patients presented with dysphagia, failure to gain weight, and regurgitation. Five of them had vomiting. One episode of pneumonia was found in two patients. All of the patients underwent preoperative diagnostic investigations including chest radiograph, upper gastrointestinal (GI) contrast study, and esophagoscopy. Upper GI contrast study indicated aperistalsis in a dilated esophagus with the characteristic ‘bird beak’ deformity with no gastroesophageal reflux (GER) in all patients (Fig. 4). Flexible upper endoscopy in all patients showed a proximal dilated esophagus with lower esophageal stricture, where the esophagoscope could pass with minimal resistance. None of the patients had received therapy before LE. The mean operating time was 44 min. The mean follow-up was 40.3 months (range 6–62 months). Upper GI contrast study was carried out on the first postoperative day to confirm mucosal integrity and to
check for GER (Fig. 5). Esophageal mucosa was intact and there was no evidence of GER on the upper GI study in any of the cases. There were no intraoperative or postoperative esophageal perforations or complications. The duration of hospital stay ranged from 2 to 3 days (mean 2.7 days). Oral feedings were resumed on the first postoperative day after upper GI study. None of the patients required redo esophagomyotomy or esophageal dilatation. Dysphagia symptoms improved uniformly and GER symptoms were not evident at follow-up.

Discussion

There is no therapy for achalasia that can reverse the underlying neuropathology or associated impaired LES relaxation and aperistalsis [8]. Therefore, all reported methods of therapy are directed to reduce the LES pressure. To improve esophageal emptying, esophageal myotomy is the most effective modality of treatment for achalasia [3,9,10]. Since the introduction of minimal invasive surgery in children, it has gained widespread acceptance. LE avoids the need for major surgery, is effective, and associated with rapid recovery, short hospital stay, and good long-term outcome [6,11–15].

The surgical objective in the treatment of achalasia is to disrupt the LES enough to eliminate dysphagia without causing excessive reflux [8,9]. If the incision is not performed adequately on the distal esophagus and stomach, dysphagia will recur and persist after surgery [16,17]. However, if the myotomy is too long on the gastric side, the risk of GER is high [17]. A myotomy that extends more than 1 cm onto the cardia of the stomach has a higher incidence of postoperative GER [7,18]. Indeed, extension of the myotomy more than 2 cm onto the cardia of the stomach is reportedly associated with 100% reflux [19]. Therefore, it has been recommended to carry out intraoperative endoscopy at the time of LE to determine the exact site of gastroesophageal junction that allows extension of the myotomy on to the gastric body to be monitored precisely and to check for integrity of the esophageal mucosa [10,17,20]. Extension of the myotomy only far enough to ensure complete division of the encircling musculature of the esophageal sphincter (LES) is recommended [7,18].

There is a debate on whether a fundoplication should be performed in addition to esophagomyotomy, and which is the best one [6,12,21,22]. In several pediatric series, a concomitant fundoplication has been used selectively [7,23,24]. Lemmer et al. [7] performed partial fundoplication in addition to esophagomyotomy in the cases where division of the obstructing gastroesophageal junction fibers required continuation of the myotomy well onto the stomach or when extensive dissection about the hiatus was performed. Payne and King [24] have been carrying out a concomitant fundoplication when a hiatus hernia is present. Richards et al. [9] have reported certain situations in which anterior fundoplication should be added to LE: when there is mucosal perforation, sliding hiatus hernia, or where preoperative 24 h pH studies indicate pathologic reflux.

Many reports recommend a routine antireflux procedure to be added to esophagomyotomy [6,21,22,25–31]. They have reported the advantages of a concomitant fundoplication, including a reduced risk of postoperative GER, protection of the herniated mucosa from the feeding injuries, and reduced risk of postoperative perforation [6,21,25–31]. There is no consensus on the ideal operation to prevent GER, because of the controversy in terms of the relative merits of the procedures: the Floppy Nissen is believed to be potentially obstructing, the Toupet procedure is satisfactory, but requires additional time and dissection, and the Dor procedure is criticized as an unsatisfactory antireflux operation [32]. Among the various fundoplications, anterior fundoplication is the most effective in reducing the risk of stenosis or recurrence of achalasia, and in preventing reflux [29,32,33]. Patti et al. [34] have reported that 17% of their patients who had LE and anterior fundoplication developed postoperative GER documented by a 24-h pH study. Even after the addition of partial fundoplication to

Fig. 4

Preoperative upper gastrointestinal.

Fig. 5

Day 1 postoperative upper gastrointestinal.
LE, patients with achalasias may have postoperative pathologic GER [9]. Paston et al. [35] have reported no significant difference in their patients who had LE with floppy Nissen fundoplication on achalasia outcomes including the subsequent development of GER.

The actual incidence of GER following surgery for achalasia is difficult to establish because it depends on the surgical approach, length of myotomy, associated pathologies, length of follow-up, and the methods used for assessment of postoperative GER [3,16]. Moreover, pseudoreflux may occur before and after esophagomyotomy [33]. This occurs when food stasis in the lower esophagus results in fermentation and the production of lactic acid, which reduces esophageal pH [32,33]. Heartburn and regurgitation symptoms might be related to acid fermentation from poor emptying of the esophagus rather than true GER [32]. There is an \( \sim 13.2\% \) risk of GER following esophagomyotomy alone by laparotomy, whereas with the addition of an antireflux procedure, the risk of GER is 7.4\% [3,23,36,37]. A recent meta-analysis study reported that the difference in the rate of GER diagnosed in post-LE pH studies in wrapped and nonwrapped patients was not statistically significant (7.9 vs. 10\%, respectively) [33]. Kumar et al. [13] reported an incidence of 6.7\% of GER after LE, and attributed this low incidence to the fact that lateral and posterior attachments of the esophagus are maintained intact. Iatrogenic GER will occur if extensive hiatal dissection is performed and if the incision onto the stomach is generously performed more than 2 cm onto the cardia of the stomach [16,18]. Mobilization of the posterior esophageal wall will increase the incidence of postoperative GER [12,29]. Limited anterior and lateral esophageal dissection eliminates the need for fundoplication [12].

The diagnosis of achalasia can be difficult in children as the symptoms can be confused with feeding aversion, failure to thrive, or GER [38]. However, in patients older than 5 years of age, the clinical presentation is very similar to adults, as follows: vomiting, dysphagia, weight loss, recurrent pneumonia, failure to thrive, or nocturnal cough [38]. In the current study, clinical evaluation, upper GI contrast study, and flexible upper endoscopy were enough to reach the diagnosis in all the cases. This approach was also reported by Tannuri et al. [22]; they used manometry only when esophageal dilatation was mild (stage I megaesophagus). They performed manometry in three out of 15 patients in their series, sparing most patients from esophagomanometry, which is a difficult, lengthy, and uncomfortable test, especially for children.

There has been a major concern of development of persistent or recurrent dysphagia after the addition of an antireflux procedure to esophagomyotomy in the treatment of achalasia, which is attributed to aperistalsis of the esophagus [3,9,13,18]. Esposito et al. [28] reported that one of the 10 patients who underwent LE and Dor fundoplication developed dysphagia 1 year after surgery. The first valve was released, and a Thal antireflux mechanism was performed. Donahue et al. [32] have reported that 15 out of 58 patients who had undergone LE and Toupet or Dor fundoplication developed postoperative dysphagia. Two of these patients required reoperation and 13 patients underwent pneumatic dilatation. Richards et al. [39] have reported that two patients out of 16 patients who underwent LE alone developed significant dysphagia postoperatively, and additional treatment in both these patients is being considered. The effective antireflux procedures (Nissen, Toupet, and Hill) all aim to increase LES pressure, which of course would be counterproductive in achalasia with an aperistaltic esophagus [32]. It seems odd to perform a Heller’s myotomy, which weakens the LES, and then add an antireflux procedure that will increase the sphincteric pressure [3].

GER may occur in patients with achalasia whether they have undergone LE alone or LE plus fundoplication [9]. Patients found to have pathologic reflux after esophagomyotomy with or without fundoplication can be effectively treated medically, but persistent dysphagia requires a more drastic and potentially hazardous treatment such as pneumatic dilatation or reoperation [9,20,32].

In the current series, we elected not to perform routine refundoplication with LE for achalasia for the following reasons:

1. Limited anterior and lateral esophageal dissection during LE to preserve the antireflux mechanism.
2. Adequate myotomy without extension of the myotomy more than 2 cm onto the cardia of the stomach to avoid GER.
3. The aim of the treatment of achalasia is to relieve the distal esophageal obstruction. Nissen fundoplication and Toupet fundoplication may cause partial distal esophageal obstruction and recurrent dysphagia.
4. Dor fundoplication has been used in the belief that it will protect the exposed esophageal mucosa postoperatively, and it is not considered as an antireflux procedure. Dor fundoplication can be useful in case there is esophageal perforation during LE.
5. There is little evidence that the GER is a relevant complication of the esophagomyotomy, and even if it occurs, it can be controlled by medical treatment.

**Conclusion**

This technique of LE without fundoplication in the treatment of achalasia is safe, effective, and yields excellent cosmetic results. The routine addition of fundoplication to LE for the treatment of achalasia in our series appears to be unnecessary. Yet, a multicenter study with a large group of patients should be carried out.

**Acknowledgements**

There are no conflicts of interest.

**References**

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