**Name of journal: World Journal of Gastrointestinal Endoscopy**

**ESPS Manuscript NO: 8704**

**Columns: Minireviews**

**Childhood achalasia: A comprehensive review of disease, diagnosis and therapeutic management**

FranklinAL *et al*. Childhood achalasia: Disease, diagnosis, treatment

Ashanti L Franklin, Mikael Petrosyan, Timothy D Kane

**Ashanti L Franklin, Mikael Petrosyan, Timothy D Kane,** Department of Pediatric General and Thoracic Surgery, Children’s National Medical Center, Washington, DC 20010, United States

**Author contributions**: All authors made substantial contributions to the conception and design, acquisition of data, or analysis and interpretation of data; all three authors participated substantially in the drafting and revising of the manuscript for intellectual content; and all authors approved the final version to be published.

**Correspondence to: Timothy D Kane, MD,** Department of Pediatric General and Thoracic Surgery, Children’s National Medical Center, Washington, DC 20010, 111 Michigan Ave NW, United States. tkane@cnmc.org

**Telephone:** +1-202-4762151 **Fax:** +1-202-4764174

**Received:** January 03, 2014 **Revised:** [February](http://www.iciba.com/February) 25, 2014

**Accepted:** March 11, 2014

**Published online:**

**Abstract**

Achalasia is an esophageal motility disorder characterized by failure of lower esophageal sphincter (LES) relaxation and is rare in children. The most common symptoms are vomiting, dysphagia, regurgitation, and weight loss. Definitive diagnosis is made with barium swallow study and esophageal manometry. In adults, endoscopic biopsy is recommended to exclude malignancy however; it is not as often indicated in children. Medical management often fails resulting in recurrent symptoms and the ultimate definitive treatment is surgical. Laparoscopic Heller myotomy with or without an anti-reflux procedure is the treatment of choice and has become standard of care for children with achalasia. Peroral endoscopic myotomy is a novel therapy utilized with increasing frequency for achalasia treatment in adults. More experience is needed to determine the safety, efficacy, and feasibility of peroral endoscopic myotomy in children.

© 2014 Baishideng Publishing Group Co., Limited. All rights reserved.

**Key words:** Achalasia; Pediatrics; Surgical heller myotomy; Balloon dilatation; Lower esophageal sphincter

**Core tip:** Achalasia is a neourodegenerative disorder of the lower esophageal sphincter which occurs less commonly in children compared to adults and patients present with progressive dysphagia, vomiting, and weight loss. Medical therapy including botulinum toxin injection and endoscopic dilatation have been associated with only transient relief of dysphagia symptoms as is also seen in adults. While current evidence also suggests that the surgical approach of laparoscopic Heller myotomy provides lasting benefits for children with achalasia, future prospective evaluation will need to be conducted to ascertain whether peroral endoscopic myotomy is safe and equally effective in children.

Franklin AL, Petrosyan M, Kane TD. Childhood achalasia: A comprehensive review of disease, diagnosis and therapeutic management

**Available from:**

**DOI:**

**INTRODUCTION**

Achalasia is a rare esophageal neurodegenerative disorder in the pediatric population. The disease is even more infrequent in children less than 5 years of age. The incidence of achalasia in childhood is 0.11/100000 children annually[1,2]. Overall, less than 5% of patients with symptoms present under the age of 15[3]. The disease is more prevalent in males and is most commonly idiopathic. Achalasia has been associated with Trisomy 21, congenital hypoventilation syndrome, glucocorticoid insufficiency, eosinophilic esophagitis, familial dysautonomia, Chagas’ disease, and achalasia, alacrima, and ACTH insensitivity (AAA) syndrome[3].

Failure of the lower esophageal sphincter to relax leads to the sequelae of achalasia. The pathophysiologic basis of achalasia is characterized by the degeneration of the inhibitory myenteric plexus that innervates the lower esophageal sphincter (LES) and esophageal body[4]. This leads to an imbalance in the inhibitory and excitatory neurons resulting in the failure of the LES to relax with swallowing, absence of peristalsis of the esophageal body, and increased LES resting pressures[5]. Goldblum *et al*[6] found a depletion or absence of myenteric ganglion cells, destruction of myenteric nerves, and chronic myenteric inflammation in 42 esophageal specimens. It is supposed that abnormalities in the parasympathetic innervation of the esophagus result in the esophageal dysmotility seen in achalasia; however the precise etiology of this abnormality is unclear[7].

Children usually present with progressive dysphagia, vomiting, and weight loss. Younger children and infants may also present atypically with recurrent pneumonia, nocturnal cough, aspiration, hoarseness, and feeding difficulties[3,8]. Achalasia in children is often misdiagnosed as gastroesophageal reflux disease (GERD). Children frequently present with failure to thrive, eating disorders, eosinophilic esophagitis, or asthma, which then leads to a delay in diagnosis for as long as 6-10 years[3]. Up to 50% of children are treated with antacids or prokinetics before the diagnosis of achalasia is identified[2].

**DIAGNOSIS**

Achalasia is diagnosed with a barium swallow study and may be confirmed with esophageal manometry. Barium swallow studies classically demonstrate a dilated esophagus with “bird’s-beak” like tapering of the distal esophagus. Often, since there is a significant delay in diagnosis of achalasia in children, the esophagram study alone is diagnostic. Elevated resting LES pressure, absent or low-amplitude peristalsis, or non-relaxing LES upon swallowing are diagnostic findings on esophageal manometry in children with achalasia[1,2]. However, absence of these findings does not rule out the diagnosis of achalasia since LES function in children is heterogeneous. Partial relaxations are common and normal relaxations may also be present on manometry according to Morea *et al*[8]. Upper endoscopy and biopsy is reasonable to rule out esophagitis, Trypanosoma cruzi, malignancy, and other secondary causes of achalasia[1,4,5]. Our institutional protocol for work up consists of a barium swallow study, upper endoscopy, and endoscopic biopsy.

The various methods of treatment of achalasia involve reduction of LES pressure in order to facilitate esophageal emptying by: injection of botulinum toxin, oral administration of calcium channel blockers (Nifedipine), pneumatic dilatation, or esophageal myotomy (Heller) with or without an anti-reflux procedure.

**MEDICAL THERAPY**

Nifedipine, a calcium channel blocker, inhibits the trans membrane calcium influx in cardiac and smooth muscle and has been primarily used to treat achalasia in adults[5]. In children, the use of nifedipine has not been well studied. Maksimak *et al*[9] reported 4 children treated with nifedipine before meals who reported relief of symptoms likely related to a decrease in resting LES pressure. In either children or adults, nifedipine is not a definitive therapy and should only rarely be used as a bridge to relieve symptoms until pneumatic dilatation, Botox injection or myotomy can be performed[5,10].

**ENDOSCOPIC THERAPY**

Botulinum toxin injected into the LES acts on the excitatory terminal nerve endings of the myoneuronal junctions preventing acetylcholine release. Acetylcholine releasing neurons function in influencing the basal muscle tone[[1](#_ENREF_1)]. Injection of botulinum toxin into the LES can be both diagnostic and therapeutic. Optimal dosing and injection frequency of botulinum toxin to relieve achalasia symptoms in children has not been well defined. After botulinum injection, the mean duration of symptom relief is 4 months, often requiring multiple treatments within a year[11]. In addition, botulinum toxin injection only provides permanent relief in 10%-40% of cases in adult patients[12] thus, will often require definitive surgical management.

**PNEUMATIC DILATATION**

Pneumatic dilatation or dilation of the functionally obstructed esophagus has been used in children. Recommended balloon sizes in children > 8 years is 35 mm[13,14]. Multiple dilatations are often required to achieve successful relief of symptoms although initial response predicts the success or failure of subsequent dilatations[15]. Hamza *et al*[14] reported a 90% success rate in children treated with multiple pneumatic dilations. The advantages of balloon dilatation include shorter length of stay, quicker recovery time, and decreased cost[13]. Pneumatic dilatation can be complicated by substernal pain, prolonged epigastric pain, esophageal perforation, aspiration pneumonia, and GERD[13,16-19]. Multiple studies suggest that in older children, pneumatic dilation is effective and safe initial treatment for achalasia and may spare children with achalasia an operation[13,14,20]. There are no long-term follow up studies in children to document success rates of pneumatic dilatation for achalasia. For adult patients, Eckardt *et al*[21] reported recurrence rates in as high as 60% in patients who underwent a single pneumatic dilation. Recurrent symptoms in children following multiple dilatations may require surgical myotomy[17,18,22].

**SURGICAL**

Despite multiple treatments for achalasia, surgery is the most definitive and successful treatment of choice. Laparoscopic Heller myotomy (LHM) involves making a longitudinal incision in the muscle of the esophagus approximately 5 cm above the esophagogastric junction and extending 2-3 cm onto the cardia of the stomach. Laparoscopic Heller myotomy in children as in adults is the surgical treatment of choice[20,23-26].

Over the last 8 years at our institution, 24 patients were diagnosed with achalasia that subsequently underwent surgical treatment. Forty-six percent of the patients were male with a mean age of 11 (5-18 years). (Table 1) In this patient population, associated comorbidities included: mixed connective tissue disease scleroderma (1); Down’s syndrome (1); inflammatory bowel disease (1); Sjogren’s syndrome; and Pott’s disease (1). The most common presenting symptoms were dysphagia (83%), emesis (58%), weight loss (46%), and chest pain (42%). Average weight loss was 9.9 kg requiring supplemental nutrition. Mean duration of symptoms prior to surgical treatment was 2.8 years, which was consistent with multiple studies[16,26-31]. Upper endoscopy in our patients commonly showed a dilated esophagus with retained food products. Approximately one-third of our patients had an abnormal biopsy. Four patients had acute esophagitis one of which was treated for Candida. Esophageal manometry was done in only 38% of our patients secondary to inability to tolerate the procedure. Only 2 patients (8%) who underwent myotomy were treated with nifedipine with only temporary relief of symptoms. Four underwent pneumatic dilatation (17%). In 1 patient, pneumatic dilatation was complicated by esophageal perforation requiring video-assisted thoracoscopic surgery (VATS) drainage and prolonged hospital stay. This patient subsequently underwent a laparoscopic Heller myotomy (LHM) and Dor fundoplication with resolution of symptoms of achalasia at 3 month follow up. Most of our patients (88%) underwent laparoscopic Heller myotomy with a Dor or Thal fundoplication. Average age at the time of surgical treatment was 12.9 years of age (5-18) (Table 2). Average operating time was 124 min.

In our series, we had only 2 intraoperative mucosal perforations, which were repaired primarily laparoscopically in children that had had LHM without fundoplication. Two children who had LHM with Thal fundoplication developed recurrent dysphagia requiring pneumatic dilations several months later. One patient who underwent a LHM and Dor fundoplication required a laparoscopic redo LHM and Dor for recurrent dysphagia. All of our patients receive a barium swallow study and a clear liquid diet on the first postoperative day. We have had no incidence of leak on the esophagram in our patients postoperatively or delayed perforations. We routinely discharge our patients on postoperative day 2 and our average length of stay is 2.6 d. Eight percent of our patients had recurrent symptoms of dysphagia postoperatively. One patient required revision of the initial operation 10 mo after the first operation (Table 3). There was a significant improvement in symptoms after the second procedure. As seen in other centers, most patients with recurrent dysphagia after surgical treatment for achalasia undergo balloon dilatation with improvement in their symptoms (Table 3).

The laparoscopic approach is superior to the open approach secondary to the well-recognized benefits including minimal pain, better cosmesis, shorter hospital stay, and faster return to normal activity for the child and parent/guardian[26]. Common causes of surgical failure are GERD and recurrent dysphagia. A partial fundoplication is commonly used to prevent GERD in patients following Heller myotomy. In a randomized controlled trial, Rebecchi *et al*[33] determined that laparoscopic Dor fundoplication after a LHM was superior to Nissen fundoplication because the recurrence rate of dysphagia was significantly higher in patients who received a Nissen fundoplication in their adult patients. There is some controversy as to whether an anti-reflux procedure should be performed in children at the time of LHM. Corda *et al*[24] concluded that an anti-reflux procedure is not required with a LHM for the prevention of GERD. Other studies have shown benefits and it is our practice to perform LHM and partial fundoplication[27,28,31,32].

The two primary complications of surgical management of achalasia are esophageal perforation and recurrent dysphagia. In our experience and review of the literature, there was 0-26% recurrence rate of dysphagia after LHM with or without an anti-reflux procedure (Table 3)[16,24,26-30,32]. It is unclear if recurrent dysphagia is secondary to the nature of disease or failure of surgical treatment. Surgeon experience may contribute to decreasing rates of complications as suggested by Esposito *et al*[26] since their incidence of post-operative dysphagia dropped from 50%[32] to 16% with further experience. Our incidence of recurrent dysphagia is 8% compared to 11%, 16%, 25%, and 26%[29,25,26,31] in comparable sized series (19–31 patients). Perforation rates occur from 0-15% (8% in ours) in larger series[16,24,29,31] but rarely require re-operation (Table 3). Accordingly, in smaller series and those from longer time periods in the past, perforation rates were higher (22-50%) probably related to the establishment of a learning curve for the operation[30,32].

**PER ORAL ENDOSCOPIC MYOTOMY**

Peroral endoscopic myotomy (POEM) is a novel technique in the treatment of achalasia. POEM is one of few procedures utilizing natural orifice transluminal endoscopic surgery (NOTES) routinely in adults. POEM is an endoscopic procedure that directly treats the diseased tissue[23]. Pasricha *et al*[34] first described a submucosal endoscopic esophageal myotomy in animal studies for the treatment of achalasia. Inoue *et al*[35] coined the term peroral endoscopic myotomy and was the first to perform the procedure in 17 adult patients. Multiple studies have concluded that short-term outcomes of this procedure were safe[35-38].

Not all patients are suitable candidates for POEM. Contraindications include severe pulmonary disease, coagulation disorders, prior esophageal mucosal resection, or any prior therapy that has compromised the integrity of the esophageal mucosa[37]. POEM is performed utilizing flexible endoscopy, mucosal incision and dissection of a submucosal tunnel distally in the esophageal wall to approach the esophagogastric junction. A 2-3 cm longitudinal incision in the inner circular muscle approximately 4 cm from the LES, will produce similar results to Heller myotomy[36,38]. A contrast esophagram is routinely obtained on the first postoperative day and the patient is started on a pureed diet if esophagram is normal[36-39].

Ren *et al*[40] reported 119 cases of achalasia treated with POEM, the most common postoperative complications included subcutaneous emphysema (55.5%), pneumothorax (25.2%), pneumomediastinum (29.4%), pleural effusion (48.7%), segmental atelectasis (49.6%), pleural effusion (48.7%), and pneumoperitoneum (39.5%). In this study, 13 patients with pneumothorax were treated with thoracic drainage and 2 patients with pleural effusion were treated with thoracentesis. The high incidence of pneumothorax, pneumomediastinum, subcutaneous emphysema, and pneumoperitoneum was attributed to the use of air insufflation during the procedure and subsequently this group now utilizes CO2 insufflation[23]. Swanström *et al*[36] reported pneumoperitoneum in 3 out of 5 patients that were treated with Veress needle. Inoue and associates reported pneumomediastinum in multiple patients, however these patients did not require treatment although another patient in that series underwent thoracostomy drainage tube placement[39]. Feasibility of POEM is highly dependent on surgeon’s experience, duration of symptoms, prior pneumatic dilatations, and endoscopic therapies[41]. Nonetheless, multiple studies have reported POEM provides favorable outcomes and is relatively safe for the treatment of achalasia in adults[35-37,39-43]. Long-term outcomes (> 6 mo) for POEM in adult patients have been reported by Swanström *et al*[44] as significant in relieving dysphagia in 83%. Maselli *et al*[45] reported the first case of POEM performed in a 3-year-old with achalasia complicated by failure to thrive. At 1 year follow up, the patient was asymptomatic and had an appropriate weight for her age[45]. Familiari *et al*[46] reported 3 children treated with POEM for achalasia. There were no postoperative complications. In this study, 2 out of 3 patients had complete resolution of symptoms and the third patient had improvement in symptoms after 1-year follow up[46]. Although POEM is effective, minimally invasive, and safe in adults, there is also more recent evidence to suggest that the surgical approach (laparoscopic Heller myotomy) is more definitive and long lasting in relieving symptoms in these patients compared to endoscopic dilatation or botulinum toxin injection techniques[47]. It is apparent that effective therapy for children with achalasia is needed. Marlais *et al*[48] reported that children with achalasia have a significantly lower quality of life (QOL) compared to both children with inflammatory bowel disease and healthy children. While current evidence also suggests that the surgical approach provides lasting benefits for children with achalasia, future prospective evaluation will need to be conducted to ascertain whether POEM is safe and equally effective in children. For now, it is unclear; however pediatric surgeons are interested in learning this novel technique and employing its use in the management of pediatric achalasia.

**REFERENCES**

1 **Walzer N**, Hirano I. Achalasia. *Gastroenterol Clin North Am* 2008; **37**: 807-25, viii [PMID: 19028319 DOI: 10.1016/j.gtc.2008.09.002]

2 **Lee CW**, Kays DW, Chen MK, Islam S. Outcomes of treatment of childhood achalasia. *J Pediatr Surg* 2010; **45**: 1173-1177 [PMID: 20620315 DOI: 10.1016/j.jpedsurg.2010.02.086]

3 **Hallal C**, Kieling CO, Nunes DL, Ferreira CT, Peterson G, Barros SG, Arruda CA, Fraga JC, Goldani HA. Diagnosis, misdiagnosis, and associated diseases of achalasia in children and adolescents: a twelve-year single center experience. *Pediatr Surg Int* 2012; **28**: 1211-1217 [PMID: 23135808 DOI: 10.1007/s00383-012-3214-3]

4 **Park W**, Vaezi MF. Etiology and pathogenesis of achalasia: the current understanding. *Am J Gastroenterol* 2005; **100**: 1404-1414 [PMID: 15929777]

5 **Chuah SK**, Hsu PI, Wu KL, Wu DC, Tai WC, Changchien CS. 2011 update on esophageal achalasia. *World J Gastroenterol* 2012; **18**: 1573-1578 [PMID: 22529685 DOI: 10.3748/wjg.v18.i14.1573]

6 **Goldblum JR**, Whyte RI, Orringer MB, Appelman HD. Achalasia. A morphologic study of 42 resected specimens. *Am J Surg Pathol* 1994; **18**: 327-337 [PMID: 8141427]

7 **Goldblum JR**, Rice TW, Richter JE. Histopathologic features in esophagomyotomy specimens from patients with achalasia. *Gastroenterology* 1996; **111**: 648-654 [PMID: 8780569]

8 **Morera C**, Nurko S. Heterogeneity of lower esophageal sphincter function in children with achalasia. *J Pediatr Gastroenterol Nutr* 2012; **54**: 34-40 [PMID: 21694632 DOI: 10.1097/MPG.0b013e3182293d8c]

9 **Maksimak M**, Perlmutter DH, Winter HS. The use of nifedipine for the treatment of achalasia in children. *J Pediatr Gastroenterol Nutr* 1986; **5**: 883-886 [PMID: 3794905]

10 **Cheatham JG**, Wong RK. Current approach to the treatment of achalasia. *Curr Gastroenterol Rep* 2011; **13**: 219-225 [PMID: 21424734 DOI: 10.1007/s11894-011-0190-z]

11 **Hurwitz M**, Bahar RJ, Ament ME, Tolia V, Molleston J, Reinstein LJ, Walton JM, Erhart N, Wasserman D, Justinich C, Vargas J. Evaluation of the use of botulinum toxin in children with achalasia. *J Pediatr Gastroenterol Nutr* 2000; **30**: 509-514 [PMID: 10817280]

12 **Pasricha PJ**, Ravich WJ, Hendrix TR, Sostre S, Jones B, Kalloo AN. Intrasphincteric botulinum toxin for the treatment of achalasia. *N Engl J Med* 1995; **332**: 774-778 [PMID: 7862180]

13 **Babu R**, Grier D, Cusick E, Spicer RD. Pneumatic dilatation for childhood achalasia. *Pediatr Surg Int* 2001; **17**: 505-507 [PMID: 11666045]

14 **Hamza AF**, Awad HA, Hussein O. Cardiac achalasia in children. Dilatation or surgery? *Eur J Pediatr Surg* 1999; **9**: 299-302 [PMID: 10584188]

15 **Boyle JT**, Cohen S, Watkins JB. Successful treatment of achalasia in childhood by pneumatic dilatation. *J Pediatr* 1981; **99**: 35-40 [PMID: 7252667]

16 **Pastor AC**, Mills J, Marcon MA, Himidan S, Kim PC. A single center 26-year experience with treatment of esophageal achalasia: is there an optimal method? *J Pediatr Surg* 2009; **44**: 1349-1354 [PMID: 19573660 DOI: 10.1016/j.jpedsurg.2008.10.117]

17 **Nakayama DK**, Shorter NA, Boyle JT, Watkins JB, O'Neill JA. Pneumatic dilatation and operative treatment of achalasia in children. *J Pediatr Surg* 1987; **22**: 619-622 [PMID: 3612456]

18 **Di Nardo G**, Rossi P, Oliva S, Aloi M, Cozzi DA, Frediani S, Redler A, Mallardo S, Ferrari F, Cucchiara S. Pneumatic balloon dilation in pediatric achalasia: efficacy and factors predicting outcome at a single tertiary pediatric gastroenterology center. *Gastrointest Endosc* 2012; **76**: 927-932 [PMID: 22921148 DOI: 10.1016/j.gie.2012.06.035]

19 **Wang L,** Li YM, Li L, Yu CH. A systematic review and meta-analysis of the Chinese literature for the treatment of achalasia. *World J Gastroenterol* 2008; **14:** 5900-6 [PMID: 18855991[DOI: 10.3748/wjg.14.5900](http://dx.doi.org/10.3748/wjg.14.5900)]

20 **Hussain SZ**, Thomas R, Tolia V. A review of achalasia in 33 children. *Dig Dis Sci* 2002; **47**: 2538-2543 [PMID: 12452392]

21 **Eckardt VF**, Gockel I, Bernhard G. Pneumatic dilation for achalasia: late results of a prospective follow up investigation. *Gut* 2004; **53**: 629-633 [PMID: 15082578]

22 **Jung C**, Michaud L, Mougenot JF, Lamblin MD, Philippe-Chomette P, Cargill G, Bonnevalle M, Boige N, Bellaïche M, Viala J, Hugot JP, Gottrand F, Cezard JP. Treatments for pediatric achalasia: Heller myotomy or pneumatic dilatation? *Gastroenterol Clin Biol* 2010; **34**: 202-208 [PMID: 20303225 DOI: 10.1016/j.gcb.2009.10.022]

23 **Rosemurgy AS**, Morton CA, Rosas M, Albrink M, Ross SB. A single institution's experience with more than 500 laparoscopic Heller myotomies for achalasia. *J Am Coll Surg* 2010; **210**: 637-45, 645-7 [PMID: 20421021 DOI: 10.1016/j.jamcollsurg.2010.01.035]

24 **Corda L**, Pacilli M, Clarke S, Fell JM, Rawat D, Haddad M. Laparoscopic oesophageal cardiomyotomy without fundoplication in children with achalasia: a 10-year experience: a retrospective review of the results of laparoscopic oesophageal cardiomyotomy without an anti-reflux procedure in children with achalasia. *Surg Endosc* 2010; **24**: 40-44 [PMID: 19495877 DOI: 10.1007/s00464-009-0513-4]

25 **Salvador R**, Costantini M, Cavallin F, Zanatta L, Finotti E, Longo C, Nicoletti L, Capovilla G, Bardini R, Zaninotto G. Laparoscopic Heller myotomy can be used as primary therapy for esophageal achalasia regardless of age. *J Gastrointest Surg* 2014; **18**: 106-11; discussion 112 [PMID: 24018591 DOI: 10.1007/s11605-013-2334-y]

26 **Esposito C**, Riccipetitoni G, Chiarenza SF, Roberti A, Vella C, Alicchio F, Fava G, Escolino M, De Pascale T, Settimi A. Long-term results of laparoscopic treatment of esophageal achalasia in children: a multicentric survey. *J Laparoendosc Adv Surg Tech A* 2013; **23**: 955-959 [PMID: 24073839 DOI: 10.1089/lap.2013.0308]

27 **Tannuri AC**, Tannuri U, Velhote MC, Romão RL. Laparoscopic extended cardiomyotomy in children: an effective procedure for the treatment of esophageal achalasia. *J Pediatr Surg* 2010; **45**: 1463-1466 [PMID: 20638525 DOI: 10.1016/j.jpedsurg.2009.08.023]

28 **Patti MG**, Albanese CT, Holcomb GW, Molena D, Fisichella PM, Perretta S, Way LW. Laparoscopic Heller myotomy and Dor fundoplication for esophageal achalasia in children. *J Pediatr Surg* 2001; **36**: 1248-1251 [PMID: 11479868]

29 **Lelli JL**, Drongowski RA, Coran AG. Efficacy of the transthoracic modified Heller myotomy in children with achalasia--a 21-year experience. *J Pediatr Surg* 1997; **32**: 338-341 [PMID: 9044149]

30 **Rothenberg SS**, Partrick DA, Bealer JF, Chang JH. Evaluation of minimally invasive approaches to achalasia in children. *J Pediatr Surg* 2001; **36**: 808-810 [PMID: 11329595]

31 **Askegard-Giesmann JR**, Grams JM, Hanna AM, Iqbal CW, Teh S, Moir CR. Minimally invasive Heller's myotomy in children: safe and effective. *J Pediatr Surg* 2009; **44**: 909-911 [PMID: 19433168 DOI: 10.1016/j.jpedsurg.2009.01.022]

32 **Esposito C**, Cucchiara S, Borrelli O, Roblot-Maigret B, Desruelle P, Montupet P. Laparoscopic esophagomyotomy for the treatment of achalasia in children. A preliminary report of eight cases. *Surg Endosc* 2000; **14**: 110-113 [PMID: 10656938]

33 **Rebecchi F**, Giaccone C, Farinella E, Campaci R, Morino M. Randomized controlled trial of laparoscopic Heller myotomy plus Dor fundoplication versus Nissen fundoplication for achalasia: long-term results. *Ann Surg* 2008; **248**: 1023-1030 [PMID: 19092347 DOI: 10.1097/SLA.0b013e318190a776]

34 **Pasricha PJ**, Hawari R, Ahmed I, Chen J, Cotton PB, Hawes RH, Kalloo AN, Kantsevoy SV, Gostout CJ. Submucosal endoscopic esophageal myotomy: a novel experimental approach for the treatment of achalasia. *Endoscopy* 2007; **39**: 761-764 [PMID: 17703382]

35 **Inoue H**, Minami H, Kobayashi Y, Sato Y, Kaga M, Suzuki M, Satodate H, Odaka N, Itoh H, Kudo S. Peroral endoscopic myotomy (POEM) for esophageal achalasia. *Endoscopy* 2010; **42**: 265-271 [PMID: 20354937 DOI: 10.1055/s-0029-1244080]

36 **Swanström LL**, Rieder E, Dunst CM. A stepwise approach and early clinical experience in peroral endoscopic myotomy for the treatment of achalasia and esophageal motility disorders. *J Am Coll Surg* 2011; **213**: 751-756 [PMID: 21996484 DOI: 10.1016/j.jamcollsurg.2011.09.001]

37 **Friedel D**, Modayil R, Iqbal S, Grendell JH, Stavropoulos SN. Per-oral endoscopic myotomy for achalasia: An American perspective. *World J Gastrointest Endosc* 2013; **5**: 420-427 [PMID: 24044040 DOI: 10.4253/wjge.v5.i9.420]

38 **von Renteln D**, Inoue H, Minami H, Werner YB, Pace A, Kersten JF, Much CC, Schachschal G, Mann O, Keller J, Fuchs KH, Rösch T. Peroral endoscopic myotomy for the treatment of achalasia: a prospective single center study. *Am J Gastroenterol* 2012; **107**: 411-417 [PMID: 22068665 DOI: 10.1038/ajg.2011.388]

39 **Inoue H**, Tianle KM, Ikeda H, Hosoya T, Onimaru M, Yoshida A, Minami H, Kudo SE. Peroral endoscopic myotomy for esophageal achalasia: technique, indication, and outcomes. *Thorac Surg Clin* 2011; **21**: 519-525 [PMID: 22040634 DOI: 10.1016/j.thorsurg.2011.08.005]

40 **Ren Z**, Zhong Y, Zhou P, Xu M, Cai M, Li L, Shi Q, Yao L. Perioperative management and treatment for complications during and after peroral endoscopic myotomy (POEM) for esophageal achalasia (EA) (data from 119 cases). *Surg Endosc* 2012; **26**: 3267-3272 [PMID: 22609984 DOI: 10.1007/s00464-012-2336-y]

41 **Teitelbaum EN**, Soper NJ, Arafat FO, Santos BF, Kahrilas PJ, Pandolfino JE, Hungness ES. Analysis of a learning curve and predictors of intraoperative difficulty for peroral esophageal myotomy (POEM). *J Gastrointest Surg* 2014; **18**: 92-8; discussion 98-9 [PMID: 24002767 DOI: 10.1007/s11605-013-2332-0]

42 **Li QL**, Chen WF, Zhou PH, Yao LQ, Xu MD, Hu JW, Cai MY, Zhang YQ, Qin WZ, Ren Z. Peroral endoscopic myotomy for the treatment of achalasia: a clinical comparative study of endoscopic full-thickness and circular muscle myotomy. *J Am Coll Surg* 2013; **217**: 442-451 [PMID: 23891074 DOI: 10.1016/j.jamcollsurg.2013.04.033]

43 **Costamagna G**, Marchese M, Familiari P, Tringali A, Inoue H, Perri V. Peroral endoscopic myotomy (POEM) for oesophageal achalasia: preliminary results in humans. *Dig Liver Dis* 2012; **44**: 827-832 [PMID: 22609465 DOI: 10.1016/j.dld.2012.04.003]

44 **Swanstrom LL**, Kurian A, Dunst CM, Sharata A, Bhayani N, Rieder E. Long-term outcomes of an endoscopic myotomy for achalasia: the POEM procedure. *Ann Surg* 2012; **256**: 659-667 [PMID: 22982946 DOI: 10.1097/SLA.0b013e31826b5212]

45 **Maselli R**, Inoue H, Misawa M, Ikeda H, Hosoya T, Onimaru M, Yoshida A, Eleftheriadis N, Suzuki K, Kudo S. Peroral endoscopic myotomy (POEM) in a 3-year-old girl with severe growth retardation, achalasia, and Down syndrome. *Endoscopy* 2012; **44 Suppl 2 UCTN**: E285-E287 [PMID: 22933258 DOI: 10.1055/s-0032-1309924]

46 **Familiari P**, Marchese M, Gigante G, Boskoski I, Tringali A, Perri V, Costamagna G. Peroral endoscopic myotomy for the treatment of achalasia in children. *J Pediatr Gastroenterol Nutr* 2013; **57**: 794-797 [PMID: 23941997 DOI: 10.1097/MPG.0b013e3182a803f7]

47 **Krishnamohan P**, Allen MS, Shen KR, Wigle DA, Nichols FC, Cassivi SD, Harmsen WS, Deschamps C. Long-term outcome after laparoscopic myotomy for achalasia. *J Thorac Cardiovasc Surg* 2014; **147**: 730-76; Discussion 730-76; [PMID: 24239236 DOI: 10.1016/j.jtcvs.2013.09.063]

48 **Marlais M**, Fishman JR, Fell JM, Rawat DJ, Haddad MJ. Health-related quality of life in children with achalasia. *J Paediatr Child Health* 2011; **47**: 18-21 [PMID: 20973860 DOI: 10.1111/j.1440-1754.2010.01884.x]

**P-Reviewers:** El-Radhi A, Wang R **S-Editor:** Song XX **L-Editor: E-Editor:**

**Table 1 Patient demographics**

|  |  |  |
| --- | --- | --- |
|  | **Mean** |  |
| **Gender** |  |  |
| Female | 13 | 54% |
| Male | 11 | 46% |
|  |  |  |
| **Age of Diagnosis** | 11 | 5-18 |
| **Duration of Symptoms** | 2.8 years | 1-11 years |
|  |  |  |
|  | **n** | **Percentage** |
| **Presenting Symptoms** |  |  |
| Dysphagia  | 20 | 83% |
| Emesis | 14 | 58% |
| Weight loss | 11 | 46% |
| Chest pain | 10 | 42% |
| Regurgitation | 4 | 17% |
| Odynophagia | 2 | 8% |

**Table 2 Surgical approach**

|  |  |  |
| --- | --- | --- |
|  | **Mean** |  |
| **Age at Surgery** | 12.9 | 5-18 |
| **OR Time** | 124 min | 45-213 min |
| **LOS** | 2.7 d | 1-6 d |
| **Follow up** | 3.5 mo | 1-12 mo |
|  | **n** | **%** |
| **LHM** | 3 | 12.5% |
| **LHM+TF** | 2 | 8.3% |
|  |  |  |
| **LHM+DF** | 19 | 79.2% |

 LOS: Length of stay; LHM: Laparoscopic heller myotomy; TF: Thal fundoplication; DF: Dor fundoplication.

**Table 3 Surgical management of pediatric achalasia**

|  |  |  |  |  |  |  |  |  |  |
| --- | --- | --- | --- | --- | --- | --- | --- | --- | --- |
| Ref. | N= | Age-years | Symptom Duration (mo) | Procedure | OR time (min) | Complications | Treatment | Length of stay(d) | Follow up (mo) |
| Pastor *et al*[16] | 40 | 12.4 | 10.7 | 6 OHM3 LHM11 LHM + Nissen21 Dilation | 186156 | 1 perforation2 perforations | SuturedSutured  | -- | 75 |
| Corda *et al* [24] | 20 | 12(5-15) | 24 | 20 LHM | 96(60-160) | 4 conversions OHM5 dysphagia | 1 lap LOA 1Redo LHM 1Redo OHM | 3(1-5) | 60 |
| Esposito *et al* [26] | 31 | 8.4(5-15) | >12 | 31 LHM/Dor | 120 | 3 perforations5 dysphagia | 2 sutured1 Redo HM2 dilated1 redo OHM | 4(3-8) | 9-156 |
| Tannuri *et al* [27] | 15 | 12 (9-17) | 30 | 15 LHM/Dor | 90(150-260) | 2 dysphagia | 1 Botox injection | 2.5(1-4) | 32.5(2-96) |
| Patti *et al*[28] | 13 | 15(6-17) | 24 | 13 LHM/Dor | 144 +/-35 | - | - | 2 | 19 |
| Lelli *et al* [29] | 19 | 10 (1-17) | - | 14 OHM5 OHM + Belsey | - | 2 dysphagia | 2 dilation | 8 | 108 (6-252) |
| Rothenberg *et al*[30] | 9 | 12 (5-17) | 6-24 | 4 THM5 LHM/Dor | 9562 | 1 perforation1 dysphagia2 GERD1 delayed perforation | SuturedRedo LHMMedical RxLap repair | 21 | - |
| Askegard-Giesmann *et al* [31] | 26 | 15(4-18) | - | 1 LHM2 LHM/Dor23 LHM + Toupet | - | 1 perforation1 perforation /aspiration7 dysphagia1 GERD | SuturedSutured3 Redo LHM3 dilation1 BotoxMedical Rx | 2.7(1-4) | 0-75(20) |
| Esposito *et al* [32] | 8 | 6.3(2-13) | >121 LHM | 6 LHM/Dor2 LHM/Thal | 120(90-150) | 3 perforations | 3 sutured | 4 (3-31) | 6-60 |
| Current Study | **24** | **12.9 (5-18)** | **>24** | **3 LHM****2 LHM/Thal****19 LHM/Dor** | **124** **(45-213)** | **2 perforations****2 dysphagia** | **2 sutured****2 dilations****1 redo LHM** | **2.7** **(1-6)** | **4** **(4-24)** |

OHM: Open heller myotomy; LHM: Laparoscopic heller myotomy; THM: Thoracosopic heller myotomy; Rx: Therapy.