Published in final edited form as:
J Pediatr Surg. 2010 June ; 45(6): 1173–1177. doi:10.1016/j.jpedsurg.2010.02.086.

Outcomes of Treatment of Childhood Achalasia

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Abstract

Purpose—The optimal management of achalasia in children and adolescents remains unclear. The aim of this study was to review a single institution experience of endoscopic and surgical interventions in children with achalasia.

Methods—A retrospective review was conducted of the medical records of children treated for achalasia from 1978 to 2008. Patient demographics and interventions were reviewed. Outcomes after procedural intervention were evaluated.

Results—35 patients with achalasia were identified, and data were available for 34 (age 13±6 years, male 62%). 18 patients underwent esophageal dilation (ED), and 16 patients underwent Heller myotomy (HM). Follow-up was available for 30 patients (15 ED, 15 HM). There was symptom recurrence in 15/15 ED cases and 8/15 (53%) HM cases ($p<0.01$). Additional interventions were performed in 14/15 (93%) ED cases and 6/15 (40%) HM cases ($p<0.01$).

Conclusions—Heller myotomy may provide more durable long-term outcomes, as defined by symptom recurrence and need for subsequent intervention, and may be considered the procedure of choice.

Keywords

Pediatric; Achalasia; Heller myotomy; Esophageal dilation

Introduction

Achalasia is a disorder of the enteric nervous system characterized by degeneration of the inhibitory myenteric neurons that innervate the esophageal body and lower esophageal sphincter (LES) [1, 2]. The resulting imbalance between excitatory and inhibitory neurons results in failure of lower esophageal sphincter relaxation with swallowing and progressive loss of peristalsis of the esophageal body [1, 2]. Pathologically, achalasia is characterized by

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myenteric inflammation with fibrosis of myenteric nerves and loss of ganglion cells [3]. Clinically, the annual incidence of achalasia in children is 0.11 cases per 100,000 children [4, 5]. Achalasia most commonly presents with dysphagia, regurgitation, chest pain and symptoms of gastroesophageal reflux [1, 2]. Diagnosis is confirmed by barium esophagram, esophageal manometry, and upper endoscopy. The aim of treatment is reduction of LES pressure with resultant elimination of outflow obstruction and symptomatic relief. Medical therapy includes nitrates, calcium channel antagonists, sildenafil, injection of botulinum toxin into the LES, and dilation of the LES [1, 2, 6]. Surgical therapy consists of a Heller myotomy with or without an antireflux procedure. Both pneumatic balloon dilation and Heller myotomy have been shown to be safe and effective therapies in children [6, 7]. The goal of the present study was to review a single institution's experience with the treatment of childhood achalasia and evaluate short and long-term outcomes with both the treatment modalities.

Methods

With approval of the institutional review board at the University of Florida (project number 572-2008), a retrospective review was conducted on medical records of children treated for achalasia at our institution from May 1978 to June 2008. Patients were identified from the hospital database and cross-referenced to the surgery registry using ICD-9 code 530. Patients were excluded from the study if they had not received intervention for achalasia at our institution or had insufficient medical records for data collection. Pre-procedural data included age, gender, presenting symptoms, past medical and surgical history, previous medications and diagnostic methods. Procedural data included type of treatment intervention, operative time, operative complications, postoperative complications, and length of hospital stay. Post-intervention data included follow-up symptoms, medications, readmissions, and subsequent interventions. Early complications were defined as those occurring in-hospital or within 30 days of surgery. Late complications were defined as those occurring following hospital discharge or more than 30 days after surgery.

Surgical technique

Our institution does not follow a standardized algorithm for the management of pediatric achalasia. The decision to proceed with esophageal dilation or Heller myotomy is surgeon-specific and is based upon the patient's symptoms and overall health, as well as the preferences of the patient and patient's family.

During the 30 year study period there was an evolution of surgical technique. However, the majority of our esophageal dilation patients were treated with our current protocol. At present, we use a pneumatic balloon dilator which is endoscopically placed over a guidewire and positioned fluoroscopically in the region of the lower esophageal sphincter. The choice of dilator size is based on the size of the child and the stricture diameter; we typically use balloon sizes ranging from 12-mm to 35-mm. Following dilation the esophagus is evaluated with an esophagram to identify perforation. Our approach to the surgical myotomy has transitioned from open abdominal, to transthoracic or thoracoscopic, to our current laparoscopic approach. In brief, we perform a laparoscopic Heller myotomy using five 5-

mm ports with a liver retractor used to aid exposure. The pars flaccida of the lesser omentum is opened to allow for dissection of the right and left crura from the esophagus and transection of the phrenoesophageal ligament. A myotomy is performed using a combination of blunt dissection, cautery, and use of an ultrasonic scalpel. The myotomy is extended 2 cm onto the gastric wall and at least 6 cm along the anterior esophagus. Endoscopy is performed to evaluate the esophagus following the myotomy for both perforation and adequacy of the myotomy. It is our current practice to then construct a Dor anterior fundoplication.

Statistical analysis

Categorical variables were expressed as proportions and compared using Fisher's exact test. Continuous variables with nonparametric distribution were expressed as medians with ranges and compared using the Mann-Whitney U test. Continuous variables with normal distribution were expressed as means with standard deviation. In addition, wherever appropriate, when assessing differences between the means, a Student's unpaired t-test was used. A p-value < 0.05 was considered significant for all the statistical tests.

Results

Demographics

Thirty-four patients were identified with a mean age of 13 ± 6 years (range 7 weeks to 20 years). There were 21 (62%) male and 13 (38%) female patients. The 7 week-old infant in the series presented with vomiting and choking; an esophagram revealed a dilated esophageal body and a lower esophageal sphincter that failed to relax with swallowing. The infant was given a clinical diagnosis of achalasia and was treated with esophageal dilation.

Presenting Symptoms

The most common symptom was dysphagia, followed by vomiting, weight loss, and chest pain. The median duration of symptoms prior to evaluation was 7.2 months (range 1 month to 6 years) overall, 18 months in esophageal dilation patients (range 1 month to 6 years), and 6.8 months in Heller myotomy patients (range 1 month to 1.5 years).

Associated Diseases

Within this series, one patient was diagnosed with Allgrove syndrome, and one patient was diagnosed with a mitochondrial disorder. By systems, comorbidities were most often gastrointestinal (32%), followed by neurologic (24%) and pulmonary (24%), and cardiac (6%).

Prior Treatment

Prior to procedural intervention at our institution, 17 (50%) patients had been treated with either a prokinetic or acid-reducing medication. Four (11%) patients had been treated with a calcium channel blocker. Eleven (32%) patients had undergone esophageal dilation at an outside hospital; within this sub-population the median number of prior dilations was 1 (range 1 to 6). One patient had undergone prior injection with botulinum toxin. A history of

prior intervention did not significantly influence our method of treatment, the incidence of early complications, symptom recurrence or the need for additional surgery.

Diagnostic Tests

Most patients were diagnosed with achalasia using a combination of barium esophagram, upper endoscopy and esophageal manometry. In the ED group the median LES pressure was 43 mmHg (range 21 – 74 mmHg). In the HM group the median LES pressure was 38.5 mmHg (range 19 – 67 mmHg). There was no significant difference between ED and HM LES pressures (p=0.33).

Operative Treatment

Eighteen (53%) patients underwent esophageal dilation (ED) as their first intervention at our institution. Of the ED patients, 15 patients (83%) underwent dilation alone, 3 (17%) received injection of botulinum toxin with esophageal dilation. Sixteen (47%) patients underwent a Heller myotomy (HM) as their first intervention at our institution. There were no intraoperative complications.

Comparing the patients that received esophageal dilation as the initial procedure at our institution to patients that received Heller myotomy, there was no significant difference in gender, symptom duration, or incidence of previous dilation at an outside facility.

Follow-Up and Outcome

Post-intervention follow-up was available for fifteen (83%) ED patients and fifteen (94%) HM patients. The ED group had a median follow-up duration of 9.1 months (range 0.6 to 275 months). The HM group had a median follow-up duration of 7.3 months (range 0.5 to 186 months). There was no mortality during the index hospitalization or within the first 30 days. There was one (7%) late death in the HM group, in a patient with history of end-stage renal disease on hemodialysis. This patient had symptom recurrence, underwent an esophagectomy and splenectomy with distal pancreatectomy, was discharged to home and at follow-up was noted to be doing well, but a subsequent note documented that he died at home 6 months after the index surgery. There were two (13%) early complications in the ED group, including immediate symptom recurrence requiring a second dilation during initial hospital stay, and a small bowel obstruction. Four (27%) patients had early complications in the HM group, including pneumonia after an abdominal HM and a transthoracic HM, a submucosal esophageal leak and small persistent pneumothorax after a thoracoscopic HM that were treated conservatively, and a deep venous thrombosis secondary to a peripherally inserted central catheter after a laparoscopic HM. There was no significant difference in the incidence of complications between the ED and HM groups (p=0.65).

All esophageal dilation patients had symptom recurrence, compared to 53% of Heller myotomy patients (p<0.01). Readmission for achalasia diagnosis was required by 80% of ED patients, compared to 33% of HM patients (p=0.03). Additional intervention was required by 93% of esophageal dilation patients, compared to 40% of Heller myotomy patients (p<0.01). In the ED group, 47% of patients eventually required surgical intervention to treat recurrence. The median intervention-free time after esophageal dilation was

significantly shorter than after Heller myotomy (p=0.04). Of note, all fundoplications in the ED group occurred after those patients had received a Heller myotomy.

Discussion

This study reviews a series of 34 pediatric patients at a single tertiary center treated for achalasia with a combination of endoscopic and surgical techniques over a 30-year time period. Achalasia is a rare disorder with an annual incidence of 1 case per 100,000, of which childhood achalasia accounts for less than 5% [8]. Therefore, neither pediatric nor adult literature has provided sufficient evidence to conclude that either endoscopic or surgical therapy is superior. Both esophageal dilation and Heller myotomy have been shown to be safe and effective in the treatment of achalasia in children [6, 7, 9-13]. A review of 260 dilations in children for esophageal strictures resulting from a variety of causes including achalasia, reported a perforation rate of 1.5%, similar to rates reported in adult achalasia literature [14, 15]. There were no esophageal perforations in our 18 index esophageal dilations, and there was no significant difference in overall complication rates between our dilation and Heller groups.

An adult study has shown that younger patients have a less sustained long-term clinical response to esophageal dilation [16]. In our series, all esophageal dilation patients developed recurrent symptoms; 93% required additional intervention in the form of repeat dilations or Heller myotomy. This recurrence rate is high compared to previously published data [9, 17]. It is possible that the LES had not been sufficiently disrupted during the procedure. However, the recurrence rate remained high even in the current era, when all dilations have been performed using a pneumatic balloon specifically designed for achalasia. Importantly, our data support the adult literature which demonstrates that a history of esophageal dilation does not affect the success of future Heller myotomy [18]. Furthermore, we did not note more complications of surgery following dilation. Prior studies of pediatric Heller myotomy report a symptom recurrence rate between 0% and 80% [2, 6, 9, 10, 19]. Our overall symptom recurrence rate of 53% after Heller myotomy may be improved with the use of intraoperative manometry as a guide, as shown in a recent study [19]. In our series, Heller myotomy was also associated with the benefits of a longer symptom-free duration, fewer hospital admissions and fewer subsequent interventions than treatment with esophageal dilation.

We do recognize that there are several significant limitations with this small, retrospective study that reduce the impact of our findings. With regards to study design, there was no standardized evaluation of symptoms or interventional success, and no systematic follow-up. In addition, this study spans a 30-year period during which time multiple surgeons and endoscopists used a variety of treatment protocols as the management of this disease evolved. Given these limitations and the rarity of achalasia in children, our study is not adequately powered to make a conclusion regarding the benefit of a simultaneous anti-reflux procedure at the time of Heller myotomy. However, based on the authors' clinical experience we believe that the Dor fundoplication may reduce the incidence of reflux and has the benefit of covering the exposed mucosa while avoiding a posterior dissection of the

esophagus. Despite these restrictions on our results, we believe that this series contributes additional data on a rare condition for which the therapy is continually progressing.

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Table 1

Demographics & Clinical Symptoms

Number of patients	34
Age (y), mean ± SE	13 ± 6
Male gender (n, %)	21 (62%)
Presenting symptoms (n, %)	
Dysphagia	27 (79%)
Vomiting	20 (59%)
Weight loss	15 (44%)
Chest pain	13 (38%)
Regurgitation	12 (34%)
Chronic cough	8 (24%)
Reflux	4 (12%)
Duration of symptoms (month), median and range	7.2 (1 – 72)

Table 2

The Impact of Prior Esophageal Dilation

	Prior Dilation N (%)	No Dilation N (%)	P value
Total number of patients	11	23	<0.01
Patients with follow-up	10 (91)	20 (87)	1.0
Esophageal dilation	4 (36)	14 (61)	0.27
Heller myotomy	7 (64)	9 (39)	0.27
Early complications	2 (20)	4 (20)	1.0
Symptom recurrence	7 (70)	16 (80)	0.66
Additional surgery	6 (60)	14 (70)	0.69

Table 3
Intraoperative Details of Heller Myotomy

Type of HM	Number (%)
Total number HM	16
Abdominal HM	3 (19%)
Alone	2
With Nissen fundoplication	1
Transthoracic HM	5 (31%)
Thoracoscopic HM	1 (6%)
Laparoscopic HM total	7 (44%)
Alone	2
With Dor fundoplication	4
With Nissen fundoplication	1

Table 4
ED vs. HM: Patient Characteristics & Prior Dilation

	ED	HM	P value
Age (y), median (range)	10.7 (0.13 – 20)	15.5 (9 – 20)	0.10
Male (N, %)	12 (67%)	9 (56%)	0.73
Symptom duration (months), median (range)	6.8 (1 – 24)	18 (1 – 72)	0.20
Prior dilation (N, %)	4 (22%)	7 (44%)	0.27

Table 5
Symptom Recurrence and Subsequent Intervention

	Esophageal Dilation	Heller Myotomy	P value
Symptom recurrence (N, %)	15 (100)	8 (53)	<0.01
Additional admission (N, %)	12 (80)	5 (33)	0.03
Intervention-free duration (d), median (range)	96 (5-1455)	1112 (98-4921)	0.04
Additional intervention (N, %)	14 (93%)	6 (40%)	<0.01
Esophageal dilation	11 (73%)	4 (27%)	0.03
Heller myotomy	7 (47%)	1 (7%)	0.04
Nissen fundoplication	2 (13%)	2 (13%)	1.0
Esophagectomy	1 (7%)	1 (7%)	1.0