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Clinical factors and high-resolution manometry predicting response to surgery for achalasia in children

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ARTICLE INFO

Article history:
Received 9 January 2018
Received in revised form
5 April 2018
Accepted 13 April 2018
Available online 11 May 2018

Keywords:
Achalasia
Pediatric
High resolution manometry
Myotomy

ABSTRACT

Background: Esophageal achalasia is an uncommon condition in children. Although many interventions exist for the management of this disorder, esophageal (Heller) myotomy offers one of the most durable treatments. Our institution sought to review patients undergoing Heller myotomy concentrating on preoperative clinical factors that might predict postoperative outcomes.

Materials and methods: All patients from January 1, 2007, to December 31, 2016, who underwent surgical treatment for achalasia at our tertiary pediatric hospital were identified and included in the study cohort. Electronic medical records for these patients were reviewed for clinical presentation variables, nonsurgical preoperative treatment, surgical approach, clinical response to surgery, need for postoperative treatment for ongoing symptoms, and high-resolution manometry (HRM) data.

Results: Twenty-six patients were included in the study, and all underwent myotomy with partial fundoplication (median age: 14.4 y [interquartile range 11.6-15.5]). At a median follow-up of 9.75 mo (interquartile range 3.5-21 mo), 16 (61.5%) patients reported good resolution of their dysphagia symptoms with surgery alone. Two patients (7.7%) had perforation of the gastrointestinal tract requiring surgical intervention. Eight patients (30.8%) required additional treatment for achalasia, with 5 (19.2%) of these undergoing additional surgery or endoscopic treatment. Patients who had preoperative dilation did not have good resolution of their dysphagia ($n = 2$; $P = 0.037$). Two of four patients undergoing postoperative dilation had preoperative dilation. None of these patients underwent preoperative manometry. There was a statistically significant difference in the ages of patients who required postoperative intervention and those who did not (14.1 *versus* 15.2 y old, respectively; $P = 0.043$). In patients who reported improvement of gastroesophageal reflux disease/reflux type symptoms after Heller myotomy, lower esophageal residual pressure

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https://doi.org/10.1016/j.jss.2018.04.027

(29.1 *versus* 18.7 mmHg; $P = 0.018$) on preoperative HRM was significantly higher than in those who did not report improvement after surgery. Higher upper esophageal mean pressure (66.6 *versus* 47.8 mmHg; $P = 0.05$) also predicted good gastroesophageal reflux disease/reflux symptom response in a similar manner.

Conclusions: Current analysis suggests that preoperative dilation should be used cautiously and older patients may have a better response to surgery without need for postoperative treatment. In addition, preoperative HRM can aid in counseling patients in the risk of ongoing symptoms after surgery and may aid in determining if a fundoplication should be completed at the index procedure. Further research is needed to delineate these factors.

Level of evidence: Level III.

Introduction

Esophageal achalasia is an uncommon functional esophageal disorder, with an incidence of 0.5-1 per 100,000 per year, that is traditionally characterized by dysfunctional motility of the esophagus and delayed or absent relaxation of the lower esophageal sphincter (LES).[1,2] Only 5% of achalasia patients are under the age of 15 y.[2,3] Although dysphagia is reliably the primary chief complaint of adults with achalasia, the pediatric population can present with persistent emesis, weight loss, failure to thrive, and even recurrent respiratory infection in lieu of or in addition to dysphagia.[4] Although symptomatology creates suspicion for the diagnosis, esophageal manometry (the gold standard for diagnosis) should show increased basal pressure with lack of normal relaxation of the LES, pressurization of the esophageal body, and usually lack of esophageal peristalsis during swallowing.[5]

The array of treatment options for achalasia is wide and has developed significantly over the past century. Currently, the focus of treatment is on the removal of the functional obstruction at the LES, as there are currently no efficacious treatments for increasing motility in the esophageal body. Procedural treatment options include intrasphincteric botulinum toxin injection and endoscopic pneumatic dilation of the LES.[6,7] Botulinum injection is less effective, requires serial injections, and can make any posttreatment surgery more prone to complication due to scar formation in the submucosal plane; thus, it is most commonly used in patients who are poor candidates for dilation or surgery.[8-10] Pneumatic dilation has variable success rates reported to be 55%-70% for single dilations and up to 90% with serial dilations but is characterized by a risk of perforation in 3%-12% of patients.[10] Surgical intervention may provide the most durable treatment of achalasia, as it can offer a high rate of symptom relief and prolonged efficacy.[11-14] The laparoscopic modified Heller myotomy is now the standard surgical treatment for pediatric achalasia, as it provides the advantages of less pain, improved cosmetic results, and shorter hospital stays.[11,15-20] However, per-oral endoscopic myotomy has also been recently described in the pediatric population.[21]

The purpose of this study was to review a recent single-institution surgical experience treating children with achalasia to identify preoperative clinical factors or high-resolution manometry (HRM) results that may influence postoperative outcomes.

Methods

After obtaining Institutional Review Board approval, a retrospective review of all pediatric patients with achalasia undergoing surgical treatment at Riley Hospital for Children at Indiana University Health from January 1, 2007, to December 31, 2016, was completed. Patients were included if their preoperative and postoperative diagnoses were achalasia and they underwent surgery before 18 y of age. Patients were excluded if they underwent surgery at an institution other than Riley Hospital for Children or were 18 y of age or older at the time of operation.

Medical records were then reviewed for common preoperative (Table 1) and postoperative clinical factors related to their achalasia (Table 2). The study period was chosen to coincide with the adoption of HRM by the Department of Gastroenterology at our institution. Common data used for diagnosing achalasia were reviewed for patients who underwent HRM ($n = 20$; Table 3). Preoperative factors and HRM data were then analyzed for relationships to postoperative outcome. Mann–Whitney *U* tests and Fisher exact tests were used when appropriate.

Results

During the study period, 26 patients underwent surgical (Heller) myotomy for achalasia. All identified patients met inclusion/exclusion criteria. The median age at time of surgery was 14.4 y (interquartile range 11.6-15.5) with a median of

Table 1 – Patient preoperative information.

Age (median [IQR])	14.4 y (11.6-15.5)
Sex (*n* [% male])	16 (61.5%)
Duration of symptoms (median [IQR])	12 mo (5.8-21.0)
Age at symptom onset (median [IQR])	13 y (9.7-14)
Preoperative botox (*n* [%])	3 (11.5%)
Preoperative dilation (*n* [%])	2 (7.7%)
Any preoperative intervention (endoscopic or medical) (*n* [%])	5 (19.2%)
Presence of dysphagia (*n* [%])	26 (100%)
Presence of emesis/GERD (*n* [%])	20 (76.9%)

GERD = gastroesophageal reflux disease.