Long-term outcomes of peroral endoscopic myotomy for achalasia in pediatric patients: a prospective, single-center study CME

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Background: Peroral endoscopic myotomy (POEM) has been developed to provide a less-invasive myotomy for achalasia in adults but seldom has been used in pediatric patients.

Objective: To evaluate the feasibility, safety, and efficacy of POEM for pediatric patients with achalasia.

Design: Single-center, prospective study.

Setting: Academic medical center.

Patients: A total of 27 pediatric patients (mean age 13.8 years, range 6-17 years) with achalasia.

Interventions: POEM.

Main Outcome Measurements: The primary outcome was symptom relief during follow-up, defined as an Eckardt score of ≤ 3. Secondary outcomes were procedure-related adverse events, clinical reflux adverse events, and lower esophageal sphincter (LES) pressure on manometry before and after POEM.

Results: A total of 26 cases (96.3%) underwent successful POEM. A submucosal tunnelling attempt failed in 1 case because of serious inflammation and adhesion. No serious adverse events related to POEM were encountered. During a mean follow-up period of 24.6 months (range 15-38 months), treatment success was achieved in all patients (mean score before vs after treatment 8.3 vs 0.7; $P < .001$). Mean LES pressure also decreased from a mean of 31.6 mm Hg to 12.9 mm Hg after POEM ($P < .001$). Five patients developed clinical reflux adverse events (19.2%).

Limitations: Single center and lack of some objective evaluations.

Conclusion: This relatively long-term follow-up study adds to the evidence that POEM seems to be a promising new treatment for pediatric patients with achalasia, resulting in long-term symptom relief in all cases and without serious adverse events. (Gastrointest Endosc 2015;81:91-100.)

Achalasia is a primary esophageal motor disorder characterized by the absence of peristalsis and defective relaxation of the lower esophageal sphincter (LES), resulting in impaired bolus transport and stasis of food in the esophagus.[1] In children, it frequently presents with dysphagia, retrosternal pain, and vomiting after meals, and infants may demonstrate failure to thrive. Achalasia is an extremely uncommon pediatric disease, with an estimated annual

Abbreviations: EGJ, esophagogastric junction; HRM, high-resolution manometry; IRP, integrated relaxation pressure; LES, lower esophageal sphincter; POEM, peroral endoscopic myotomy; PPI, proton pump inhibitor.

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incidence of 0.02 to 0.11 cases per 100,000 children; however, it can lead to adverse events such as malnutrition as well as stunting of mental and physical development.[2,3]

To date, mainly because of a lack in pathophysiology insight, definitive treatments of achalasia address the symptoms but not the cause of the disease. Current treatments can be pharmacologic, endoscopic, or surgical. In part because of the limited number of pediatric patients with achalasia, the best treatment approach has been controversial. With respect to long-term efficacy, esophageal myotomy by using an open or a laparoscopic approach has gained wide acceptance in recent years as the procedure of choice for the management of pediatric patients with primary achalasia.[4]

Peroral endoscopic myotomy (POEM) has been described recently as a scar-free and less-invasive surgical myotomy option for treating achalasia.[5] This procedure incorporates concepts of natural orifice transluminal endoscopic surgery and achieves endoscopic myotomy by using a submucosal tunnel as the operating space. Thus, it is minimally invasive, less expensive than a surgical operation, and associated with a faster recovery. Initial published experience in adults is more than encouraging despite a relatively short follow-up.[6-8] However, POEM seldom has been used in pediatric patients.[9-11] The aims of this prospective study were to investigate the feasibility and safety of POEM for pediatric patients and to assess relatively long-term efficacy of POEM by using a validated achalasia symptom score and manometry.

PATIENTS AND METHODS

Patients

After institutional review board approval was obtained (approval number 2010-135, ethical committee of Zhongshan Hospital, Fudan University), the study included consecutive patients in the Endoscopy Center and Endoscopy Research Institute, Zhongshan Hospital, Fudan University between August 2010 and July 2012. The study also has been registered in ClinicalTrials.gov (identifier: NCT01649843). Patients aged <18 years were eligible for enrollment in the study if they reported persistence of symptoms, with an Eckardt symptom score[1] ≥ 4. The Eckardt score is the sum of the symptom scores for dysphagia, regurgitation, chest pain, and weight loss.[1] A score between 0 and 3 was assessed depending on the symptom of dysphagia, regurgitation, or chest pain (not present, score 0; occasionally, score 1; daily, score 2, and several times a day after each meal, score 3) or on the degree of weight loss (none, score 0; <5 kg, score 1; 5-10 kg, score 2; >10 kg, score 3). The symptoms were recorded from the patient's self-report or the report of the parents or caretakers. Achalasia was suspected because of regurgitation, vomiting, weight loss, recurrent cough, aspiration pneumonia, growth retardation, and retrosternal pain; it was confirmed by barium swallow, high-resolution manometry (HRM), and EGD. Exclusion criteria were severe cardiopulmonary disease or other serious disease leading to unacceptable surgical risk, pseudoachalasia, megaesophagus (diameter of >7 cm), and hiatal hernia (>2 cm). Written informed consent was obtained from the patients before POEM or from caretakers when patients were aged <16 years.

Outcomes measurements

The primary outcome of the study was therapeutic success (a reduction in the Eckardt score to ≤ 3) at the follow-up assessment. Secondary outcomes included procedure-related adverse events, LES pressure on manometry before and after POEM, clinical reflux adverse events, medication use before and after POEM, and procedure-related parameters such as procedure time, hospital stay, and myotomy length.

High-resolution manometry

Baseline and postmyotomy LES pressures and 4-second integrated relaxation pressures (IRP) were recorded by using an HRM system (Sierra Scientific Instruments Inc, Los Angeles, Calif) as previously described.[12] Briefly, the HRM assembly was placed transnasally and the manometric catheter positioned to record from the hypopharynx to the stomach, with approximately 5 intragastric sensors. Studies were performed with patients in a supine position after at least a 6-hour fast. The manometric protocol included a 5-minute period to assess basal esophagogastric junction (EGJ) pressure, 10 water swallows of 5 mL, and 1 water (Given Imaging, Yoqneam, Israel) swallow each of 1 mL (dry), 10 mL, and 20 mL. All manometric analysis was done by using Mano-View software applied to the data tracings viewed in the color pressure topography mode and referenced to intragastric pressure. The 4-second IRP, which is representative of the adequacy of deglutitive relaxation of the EGJ, was determined by adjusting the pressure on the isobaric contour tool to the lowest value at which a cumulative time period of 4 seconds was excluded along the axial plane of the EGJ.

POEM procedures

A standard, high-definition endoscope (GIF-H260 or GIF-Q260; Olympus Medical Systems Co, Tokyo, Japan) was used in all cases. A transparent cap (D-201-11802; Olympus) was attached to the tip of the gastroscope. Other equipment included a hybrid knife (ERBE, Erbe Elektromedizin GmbH, Tübingen, Germany), triangle-tip knife (KD-640L; Olympus), hook knife (KD-620LR; Olympus), injection needle (NM-4L-1; Olympus), hot biopsy forceps (FD-410LR; Olympus), clips (HX-610-90, HX-600-135; Olympus; Resolution, Boston, Mass), high-frequency generator (VIO 200D; ERBE), and argon plasma coagulation unit (APC300; ERBE). Room air was used for