Journal of Pediatric Surgery 50 (2015) 726–730

Contents lists available at ScienceDirect

Journal of Pediatric Surgery

journal homepage: www.elsevier.com/locate/jpedsurg

Treatment of esophageal achalasia in children: Today and tomorrow

Tamara Caldaro [a,*], Pietro Familiari [b], Erminia Francesca Romeo [a], Giovanni Gigante [b], Michele Marchese [b], Anna Chiara Iolanda Contini [a], Giovanni Federici di Abriola [a], Salvatore Cucchiara [c], Paola De Angelis [a], Filippo Torroni [a], Luigi Dall'Oglio [a], Guido Costamagna [b]

[a] Digestive Surgery and Endoscopy Unit, Bambino Gesù Children's Hospital, Rome, Italy
[b] Digestive Surgery and Endoscopy Unit, Catholic University, Rome, Italy
[c] Pediatric Gastroenterology and Liver Unit, Sapienza University, Rome, Italy

ARTICLE INFO

Article history:
Received 19 January 2015
Accepted 13 February 2015

Key words:
esophageal achalasia
esophageal manometry
Heller myotomy
POEM

ABSTRACT

Background: Esophageal achalasia (EA) is a rare esophageal motility disorder in children. Laparoscopic Heller myotomy (LHM) represents the treatment of choice in young patients. Peroral endoscopic myotomy (POEM) is becoming an alternative to LHM.
The aim of this study is to evaluate the effectiveness, safety, and outcomes of POEM vs LHM in treatment of children with EA.
Methods: Data of pediatric patients with EA, who underwent LHM and POEM from February 2009 to December 2013 in two centers, were collected.
Results: Eighteen patients (9 male, mean age: 11.6 years; range: 2–17 years) were included. Nine patients (6 male, mean age: 10.7 years; range: 2–16 years) underwent LHM, and the other 9 (3 males, mean age: 12.2 years; range: 6–17 years) underwent POEM procedure.
Mean operation time was shorter in POEM group compared with LHM group (62/149 minutes). Myotomy was longer in POEM group than in LHM group (11/7 cm). One major complication occurred after LHM (esophageal perforation). No clinical and manometric differences were observed between LHM and POEM in follow-up. The incidence of iatrogenic gastroesophageal reflux disease was low (1 patient in both groups).
Conclusions: Results of a midterm follow-up show that LHM and POEM are safe and effective treatments also in children. Besides, POEM is a mini-invasive technique with an inferior execution timing compared to LHM. A skilled endoscopic team is mandatory to perform this procedure.

Esophageal achalasia (EA) is a rare, chronic and progressive motility disorder, manometrically characterized by absence of esophageal peristalsis and incomplete or absent relaxation of the lower esophageal sphincter (LES) [1].

Pediatric achalasia is often misdiagnosed and may result in debilitating symptoms and failure to thrive.

The estimated incidence of EA in children is 0.11 per 100,000, without racial or gender predilection [2]. Although clinical, radiological and endoscopic results may raise the suspicion of EA, the definitive diagnosis depends on esophageal manometry.

Laparoscopic Heller myotomy (LHM) is the recommended procedure for adults with EA, while medical treatments and/or endoscopic esophageal pneumatic dilations are usually reserved for those older than 45 years and with high surgical risk [1].

Peroral endoscopic myotomy (POEM) is a minimally invasive procedure, that has been recently developed and introduced in clinical practice for the treatment of EA in adults [3–5].

POEM combines the benefits of an endoscopic procedure with the long-term efficacy of a surgical myotomy. Preliminary studies demonstrated that POEM is associated with a low rate of complications and a good effectiveness at midterm follow-up [6].

Up to now, isolated cases of children with EA have been treated with POEM, with excellent results at short-term follow-up [7,8].

The aim of this multicenter study is to compare the perioperative outcomes and clinical efficacy of POEM and LHM for the treatment of EA in children.

1. Patients and methods

Eighteen pediatric patients with EA, who had undergone LHM or POEM between 2009 and 2014, were retrospectively identified from the prospectively collected databases of two tertiary referral centers: Bambino Gesù Children's Hospital (Digestive Surgery and Endoscopy Unit) and Gemelli University Hospital (Digestive Surgery and Endoscopy Unit).

* Corresponding author at: Digestive Surgery and Endoscopy Unit, Bambino Gesù Children's Hospital, Piazza S. Onofrio 4, 00165, Rome, Italy. Tel.: +39 0668592841; fax: +39 0668592949.
E-mail address: tamara.caldaro@opbg.net (T. Caldaro).

http://dx.doi.org/10.1016/j.jpedsurg.2015.02.047
0022-3468/

Preoperatively, barium X-ray series, esophageal manometry and upper endoscopy were performed.

The Eckardt score system was used for the clinical evaluation before and after both procedures [9].

Since the introduction of POEM at Gemelli University Hospital in 2012, patients with EA and their parents were informed regarding the two available options (LHM and POEM) to freely decide which procedure to choose.

The surgical treatment (LHM) was performed in 9 patients (mean age at surgery: 10.8 years; range: 2–17 years) by the same team of three surgeons at Bambino Gesù Children's Hospital; while a single endoscopist performed POEM at Gemelli University Hospital in the other 9 children (mean age at POEM: 12.2 years; range: 6–17 years).

1.1. Laparoscopic Heller myotomy (LHM)

The patient is placed in the supine position and 4 trocars (one trocar: 10 mm; 3 trocars: 5 mm) are inserted in to the abdomen under direct visualization.

The esophagogastric junction is exposed for 6–8 cm proximally and the myotomy is performed longitudinally for a minimum of 6 cm, using a blunt dissector and scissors.

To prevent burning or perforation of the esophageal mucosa, the use of hook cautery or cautery scissors was avoided.

To exclude small esophageal perforations, air is inflated into the stomach using a nasogastric tube while a sodium chloride solution is introduced into the abdomen. An esophageal perforation is detected when bubbles can be seen into the abdomen.

The procedure is completed by performing a Dor fundoplication.

1.2. Peroral endoscopic myotomy (POEM)

POEM procedure is performed according to the technique described by Inoue et al [10] in 2010. Broad spectrum antibiotics (usually a cephalosporin or amoxicillin) are administered preoperatively. An upper endoscopy is performed using a high-definition endoscope (GIF-H180J; Olympus Tokyo, Japan) with carbon dioxide (CO_2) insufflation. The esophageal mucosa is cut on the anterior wall using an endoscopic cautery knife (Triangle-tip knife, Olympus), approximately 10–12 cm above the esophageal gastric junction (Fig. 1A). After the mucosal incision is completed, the esophageal submucosa is dissected using a spray coagulation current (VIO300D, ERBE Elektromedizin GmbH, Tubingen, Germany) (Fig. 1B). A long tunnel is created in the esophageal submucosa extending from the mucosal incision to 3 cm along the anterior gastric wall (Fig. 1C). The esophageal muscular layer is exposed and cut through the submucosal tunnel. The myotomy includes the circular bundles of the muscular layer and extends for 2–3cm up the gastric wall (Fig. 1D). At the end of the procedure, the mucosal incision is closed using endoscopic clips (EZ Clips, Olympus). Twenty-four or 48 hours after myotomy, an upper endoscopy under general anesthesia and a gastrografin esophageal study are performed to exclude mucosal tears or leaks; then patients are fed a soft diet.

1.3. Follow-up, data collection and analysis

Demographics, data on the clinical history and the results of preoperative studies (upper endoscopy, barium swallow and esophageal manometry) were collected from the medical charts and the prospective databases, recorded and compared.

Information on the perioperative outcomes, including procedure time, length of myotomy, perioperative complications and length of hospitalization, was analyzed.

After the procedures, patients underwent a rigorous follow-up.

Esophageal manometry was repeated after 6 months.

The onset of gastroesophageal reflux disease (GERD) was detected by pH-monitoring and upper endoscopy, 6 months postoperatively.

The Eckardt score was used to evaluate the clinical efficacy of the procedures during follow-up.

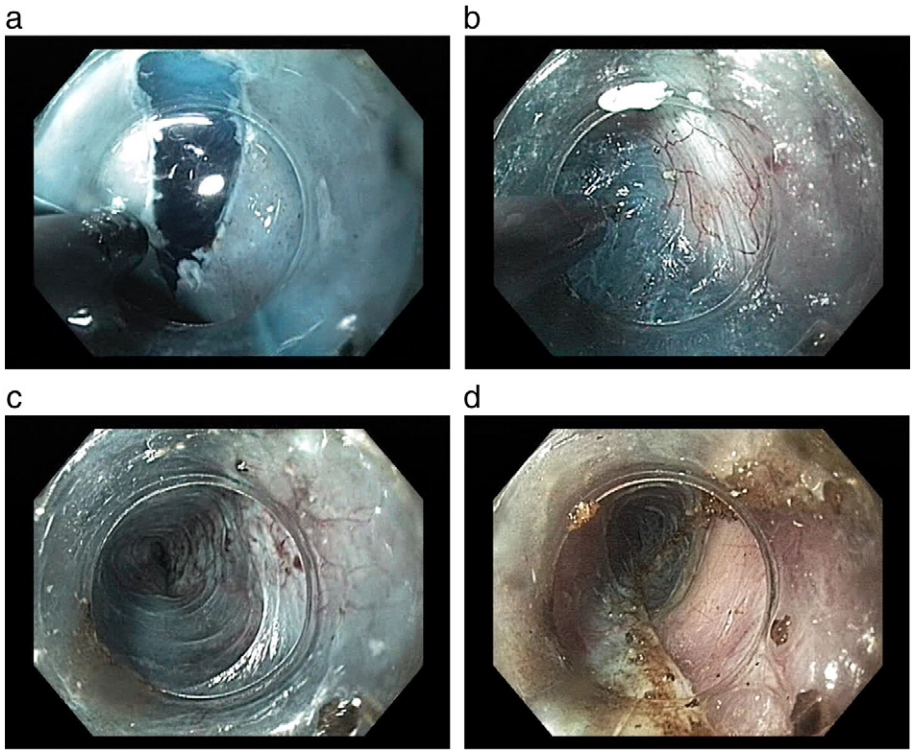

Fig. 1. POEM procedure. A) Mucosal incision. Esophageal submucosa is injected with saline solution and indigo carmine. B) Submucosal dissection. On the right side of the figure, the muscular layer. Submucosa is colored in blue. C) The submucosal tunnel is extended from the mucosal incision to the anterior gastric wall. The mucosal layer, on the left of the figure, is intact. D) Myotomy begins about 3 cm distal to the mucosal incision and it is extended until the anterior gastric wall.