Peroral Endoscopic Myotomy for the Treatment of Achalasia in a 10-Year-Old Male Patient

Jörg Filser1  Anke Dick2  Thomas Meyer1  Christoph-Thomas Germer1  Burkard H.A. von Rahden1

1 Department of General, Visceral, Vascular and Paediatric Surgery, University Medical Center, Würzburg, Germany
2 University Children’s Hospital, University Medical Center, Würzburg, Germany


Abstract
Peroral endoscopic myotomy (POEM) is a new endoscopic treatment for achalasia with very good short-term results in adults. Data about POEM in pediatric patients are missing. We present the case of a 10-year-old male patient with type I (classic) achalasia, successfully treated with POEM. The procedure was accomplished in a similar fashion to the technique used in adults. Short-term results were fine, with a complete control of dysphagia and absence of reflux. We suggest that POEM is a suitable option in pediatric patients—similar to adults—but long-term results must be awaited.

Keywords
- achalasia
- POEM
- peroral endoscopic myotomy

Introduction
Achalasia is a rare disease with an incidence of 1 per 100,000 patients in adults.1 The disease also occurs in children—through all age groups—however, it is estimated even less frequent in the pediatric population (0.11 cases per 100,000 patients).2 Achalasia is a rare esophageal motility disorder characterized by failure of the lower esophageal sphincter (LOS) to relax and aperistalsis of the tubular esophagus. Pneumatic dilation (PD) and laparoscopic Heller myotomy (LHM) are well established and currently used as standard treatment for achalasia in children.3,4 Peroral endoscopic myotomy (POEM) is a new, entirely endoscopic procedure for the treatment of achalasia. Myotomy is performed exclusively through the transoral endoscopic route and is therefore a true NOTES procedure (Natural Orifice Endoscopic Surgery). Current data regarding POEM in adults demonstrate excellent postoperative outcome.5–7 Data about POEM in pediatric patients are still missing.

Case Report
We present here the case of a 10-year-old male patient admitted to our department with an 18-month history of dysphagia, recurrent vomiting, and weight loss. The patient complained about regurgitations appearing immediately after food intake. General pediatric workup including gastroscopy showed no morphologic correlative for these symptoms. High-resolution manometry (HRM)/esophageal pressure topography (EPT) study using Sierra (Sierra Scientific Instruments Inc., Los Angeles, California, United States), according to the standards defined in the Chicago classification8 was performed to check for esophageal motility disorder. EPT data were analyzed using ManoView analysis software (Given Imaging Ltd., Yokneam, Israel). This investigation revealed an increased integrated relaxation pressure (IRP) of the LOS (15.5 mm Hg) and aperistalsis of the tubular esophagus, with 100% simultaneous (tertiary) contractions (∫ Fig. 1). According to the Chicago classification of esophageal motility disorders, this result had to be classified as type I (classic) achalasia.8

Additional timed-esophagography (Fluo Loop [3/s; Global Siemens Healthcare, Erlangen, Germany] and fluoroscopy with last-image-hold-technique) showed a dilated esophagus with slow transit of the contrast medium across the esophagogastric junction (OeGJ) (∫ Fig. 2). The Eckardt dysphagia score9 was 8 points (weight loss 1; dysphagia 3; retrosternal pain 1; and regurgitation 3).

The different available treatment options were discussed with the parents of the patient. Due to the convincing results
of POEM in adults—in our own experience as well as the available literature— we decided to offer the POEM procedure.

The POEM procedure was performed using the technique suggested by Inoue et al as follows: (1) creation of a mucosal entry, (2) creation of a submucosal tunnel using an endoscopic submucosal dissection technique, (3) myotomy, and (4) clip closure of the mucosal entry (Fig. 3A–E).

For this procedure we used the “Silverscope” by KARL STORZ, Tuttingen, Germany, type 13821PKS, 9.3 mm. The whole procedure was performed using carbon dioxide insufflation (UCR, Olympus Medical Systems Corp., Tokyo, Japan). A cap attachment with straight orifice (DH28GR, Fuji film, Tokyo, Japan) was used for all steps of the procedure (submucosal tunnel, myotomy). Closure of the mucosal incision, was performed with an oblique cap attachment. The POEM procedure was carried out under general anesthesia with the patient in supine position. Submucosal injection of 10 mL indigocarmine-stained saline solution, which was applied with a special injection device (InjectorForceMax, NM-401L-0425, Olympus) was performed 2 cm below the aortic arch (clearly visible at 30 cm from the incisors). The mucosal entry was created with the triangle tip (TT)-knife (KD-640L; Olympus) and the energy source VIO300D (ERBE Elektromedizin GmbH; Tübingen, Germany) in “cut” mode (80 W; effect 2).

Dissection of the submucosal space was also performed with the TT-knife. Stained saline solution was repeatedly injected into the submucosal space, using a blunt catheter (PW-205V; Olympus) and dissection was performed with the energy source in “spray” mode (50 W; effect 2). The submucosal tunnel was created in the tubular esophagus and carried forward across the OeGJ (35 cm below the dental line). Myotomy of the circular muscle fibers was started approximately 3 cm below the mucosal entry and conducted across the OeGJ until 2 cm at the gastric site (31–37 cm below the dental line), whereas the longitudinal muscle layer was preserved to serve as “safety margin.”

For clip closure of the mucosal entry, we used a cap attachment with oblique orifice (MH-588; Olympus). Seven endoclips (HX-610–090L; Olympus) were applied with the standard multiuse clip applicator (HX110QR; Olympus). The operating time was 93 minutes.

Postoperatively the patient was allowed to drink small sips of water. Proton pump inhibitors (PPI) as well as cefuroxime were administered intravenously for 3 days and then switched to per oral. Postoperatively, a slightly elevated C-reactive protein levels (3.6 mg/dL) with normal white blood cell were noted. On postoperative day 1 a timed esophagography was performed, which demonstrated prompt delay of the esophagus (Fig. 4) and no signs of leakage. After an unremarkable postoperative course, the patient was discharged home on day 3 in good general condition with...
very good symptom relief, that is, absence of dysphagia, and no clinical reflux symptoms. Short-term follow-up (2 months) showed no clinical impairment with complete control of dysphagia (Eckardt score 0) and no sign of reflux.

Discussion

POEM is a promising new technique for treatment of achalasia, with more than 1,400 cases having been performed in specialized centers worldwide to date. The procedure is attractive due to the myotomy being performed entirely through the endoscopic route and its excellent results, that is, good control of dysphagia and low reflux rates in the short term. However, little is known about the potential role of POEM for treatment of pediatric achalasia patients. Only two previous reports have addressed this issue: Maselli et al have reported briefly about a successfully accomplished POEM procedure in a 3-year-old achalasia patient with growth retardation and Down syndrome. Familiar et al reported about successful POEM procedures in three female achalasia patients (9, 9, and 11 years of age) with no intra- and postoperative complications and complete symptom relief (normalized Eckardt score) at 1-year follow-up. These reports as well as our experience suggest that POEM might be well suited as treatment option for pediatric achalasia patients as well. Due to our excellent results with POEM in adult achalasia patients, we decided to recommend this well-suited technique for treatment of pediatric achalasia as well.
technique for our pediatric patient. At this time we had an
experience of altogether 11 POEM procedures performed in
adults. Before introducing POEM in clinical practice we had
undergone a special training in Yokohama/Japan and had
trained the procedure in a well-suited animal model. There-
fore, we had discussed the circumstances, a new procedure
without long-term experience and rather no experience in
children, but excellent short-term results in adults with the
parents, who agreed to have POEM done on their child.

Technical aspects as well as the procedure itself did not differ
from our experience in adults. Anatomical landmarks were
identified easily, especially the OeGJ, which is a crucial landmark
for this procedure, and the impression of the aortic arch. To
confirm a save intragastric position, we followed the suggestions
made by Inoue et al.5 which are also nicely summarized in the
IPOEM (International Per Oral Endoscopic Myotomy) survey by
Stavropulos et al.6: the myotomy has completely addressed the
muscle responsible for achalasia, when the endoscope reaches a
wide submucosal space with larger vessels. Further indicators
are detection of indigocarmine stain in retroflexed view, level
from the incisors and smooth passage of the endoscope through
the previously tight OeGJ.

As a new procedure for the treatment of achalasia POEM
must exhibit some outstanding characteristics to be a serious
alternative to the current procedures PD and LHM in children.
The free choice of length and localization of the myotomy may
be regarded as a fundamental advantage of the POEM pro-
dure. A longer myotomy may be beneficial in patients suffer-
ing from chest pain, and perhaps especially in patients with
type III achalasia, where PD and LHM are potentially inferior.

POEM could also be an option for esophageal motility
disorders in which pharmacological treatment has failed.13
As a pure NOTES procedure the manipulation in the abdomi-
nal cavity as well as visible scars are avoided. This could be a
weighty argument for this procedure referring to the future
body image of adolescents.

Also, the previous operations in the abdominal cavity are
assessed as a reasonable indication for POEM.6,14,15 But which
side effects should be mentioned particularly in the consult-
tation with patient and the parents? The focus of interest
should be the potential appearance of gastroesophageal
reflux after POEM.

Despite all the promising early experiences with POEM,
some aspects warrant controversial discussion. One of these
is the exact rate of postoperative reflux, which is currently not
known. One might expect a higher reflux rate after POEM
compared with LHM, because the myotomy is not combined
with a fundoplication, which is standard when performing
LHM.16,17 However, an antireflux procedure might—on the
other hand—be unnecessary after POEM, because natural
antireflux barriers such as the phrenoesophageal membrane
and the angle of His remain untouched. The current available
literature is controversial regarding reflux rates after POEM,
ranging from 5.9 to 46%.5,7 The overall reflux rates of 20 to
30% are probably the correct estimate10 and are pretty similar
to with reflux rates after LHM and PD.17,18 Most reported
reflux-associated problems are minor complaints, which are
well treatable with PPI. Nevertheless, this issue will be of the
special interest because of the longer exposition of the (distal)
esophagus to reflux in children compared with adults.

One other aspect regarding differences between POEM and
LHM is the type of myotomy: during LHM, all muscle layers
(circular and longitudinal) are cut. During POEM we cut only
the circular muscle fibers, whereas the longitudinal muscle is
preserved, following the suggestion by Inoue et al.5 A recent
randomized study from China19 has shown, that there is no
difference with respect to inclusion or exclusion of longitu-
dinal fibers in the myotomy. This supports the current view of
protagonists of the POEM procedure, that only the circular
muscle layer is responsible for achalasia.

All currently available publications reported no serious
local infections although the setting of this procedure
would suggest this. To reduce the potential risk of infection
we perform an application of a topical antibiotic (genta-
mycin) and administer intravenous antibiotics for 1 week
postoperatively.

If CO2 insufflation is used20 our experience as well as
reported data revealed predominantly minor complications
such as pneumoperitoneum, cutaneous emphysemas, pneu-
mothorax, which are well treatable intraoperative or are
mostly self-limited.5,7,14,20,21 Mucosal lesions, which
were normally depict and provide at the postoperative
endoscopy cannot be regularly identified in our described
setting. Therefore, other strategies have to be developed to
prevent serious events as described above. The risk of major
bleeding especially in the submucosal tunnel seems to be low
but also difficult to manage.22

One final issue to discuss is the question what to do after a
potential treatment failure of POEM. Again all three options
for treatment of achalasia (LHM, PD, and POEM) may be
considered: (1) LHM might be an option but might well be
more difficult, because there might be severe scarring in the
submucosal tunnel and at the outer surface of the mucosa,
which might show stronger adherence to the muscular layers.
(2) PD might also be an option. However, the risk for esopha-
geal perforation might be higher after POEM, when the
muscular layer at the esophagogastric junction is already
weakened. (3) The best option in this setting might again be a
(redo) POEM procedure. This could be performed at the
posterior esophageal wall, which is untouched at this time.
Only recently, Inoue et al have reported POEM as a redo
procedure after failed myotomy.23

Conclusion

Technical aspects of the POEM procedure in children do not
differ from adults. Pre- and postoperative treatment concepts
must be tailored to children conditions and detailed informa-
tion of the parents is crucial. In an adjusted setting, POEM
seems to be a suitable alternative for the treatment of
achalasia. Larger case series and long-term follow-up are
required.

Conflict of Interest

None.
References