



## **Esophageal Peroral Endoscopic Myotomy in a Young Patient with Type II Achalasia**

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**Received: 20 May 2025**

**Published: 02 June 2025**

DOI: <https://doi.org/10.5281/zenodo.15654824>

**Abstract**

*Achalasia is a motor disorder of the esophagus characterized by incomplete relaxation of the lower esophageal sphincter and disordered esophageal peristalsis. Current treatments include pneumatic dilation, botulinum toxin injections, laparoscopic Heller myotomy, and esophageal peroral endoscopic myotomy (E-POEM). E-POEM is an advanced, minimally invasive non-surgical intervention for the treatment of achalasia. This procedure has repeatedly shown non-inferiority to the classic laparoscopic, surgical Heller myotomy, and is the preferred approach due to being an endoscopic procedure. In this article, we report a case of type II achalasia in a 21-year-old male that was treated by E-POEM which resulted in a significant reduction in Eckardt score and resolution of symptoms.*

*Keywords: esophageal peroral endoscopic myotomy (E-POEM), achalasia, dysphagia.*

**Case Report**

The patient is a 21-year-old male who was initially assessed for a three-year history of progressive dysphagia to liquids and solids with every meal, regurgitation, and chest pain without weight loss (Eckardt score of 8). The patient was initially seen by the surgery service and an Esophagogastroduodenoscopy was performed which showed increased resistance at the level of the lower esophageal sphincter muscle without any stricture or esophagitis. Esophageal dilation was performed using a Maloney dilator which did not result in resolution or improvement of symptoms. The patient was subsequently referred to the gastroenterology service. Initial esophagram showed the classic “bird beak” appearance at the distal esophagus with proximal dilation (Fig 1) suggesting the possibility of achalasia. High-resolution manometry showed an increased basal pressure of the lower esophageal sphincter of 51.6mmHg, a residual pressure of 26mmHg consistent with incomplete relaxation, 70% panesophageal pressurization, and 100% of swallows proving ineffective. These readings are consistent with type II achalasia. Various treatment options were discussed and the patient elected to undergo esophageal peroral endoscopic myotomy.

The E-POEM procedure was performed under general anesthesia. The patient was placed in supine position and the posterior approach was used. After esophagogastroduodenoscopy, the Functional Lumen Image

probe (FLIP) procedure was performed by passing an 8 cm Medtronic EndoFLIP balloon catheter into the esophagus to measure the pressure of the lower esophageal sphincter using various balloon volumes. Pre-procedure measurements are given in table 1. After completing FLIP study, the E-POEM procedure was performed. 7mL of saline with methylene blue was injected 10 cm above the gastroesophageal junction into the submucosal space at the 5 o'clock position. 1.5 cm mucosal incision was made using a triangular tip knife (TT J knife by Olympus America) on ENDO CUT settings using an erbe generator. The submucosal tunnel was done using spray coagulation electrical current. The tunnel was continued to 3cm into the gastric side. Blood vessels were cauterized by coagulation grasper (Olympus America) using the soft coagulation electrical current setting. The myotomy was started at 2 cm below the mucosal incision and continued all the way to 2 cm below the lower esophageal sphincter into the stomach. 7 cm of esophageal myotomy including lower esophageal sphincter and 2 cm of gastric myotomy were performed. The lower esophageal sphincter myotomy was full-thickness whereas the esophageal myotomy was partial thickness involving circular muscle only. After confirming the myotomy extended to the stomach the FLIP study was performed again (table 2) to confirm complete myotomy. The mucosal incision was closed using through-the-scope 17 mm clips (Boston Scientific). A total of six clips were used for mucosal closure. A final endoscopic evaluation was done to assure there was no inadvertent mucosal injury.

EndoFLIP data:

Pre-POEM: Table 1

	CSA ( $mm^2$ )	Diameter (mm)	Distensibility ( $mm^2/mmHg$ )	Pressure (mmHg)
Balloon 30mL	19	5	0.6	30.3
Balloon 40mL	34	6.6	0.6	57.2
Balloon 50mL	116	12.1	1.5	75

Post-POEM: Table 2

	CSA ( $mm^2$ )	Diameter (mm)	Distensibility ( $mm^2/mmHg$ )	Pressure (mmHg)
Balloon 30mL	120	12.4	13	9.3
Balloon 40mL	146	13.6	9.5	15.4
Balloon 50mL	233	16.2	8.6	23.6

One day following the procedure, the patient had mild post-procedure chest discomfort but was otherwise recovering well. A gastrografin swallow study was conducted and was unremarkable. The patient was discharged home with prophylactic oral antibiotic and proton pump inhibitor therapy, and a clear liquid diet for one week with transition to 2 weeks of soft diet afterwards. At the two-week follow-up appointment, the patient reported improved swallowing, minimal to no burning sensation in his chest, and no regurgitation with an Eckardt score of 1.

## Discussion

Achalasia is a functional motor disorder of the esophagus characterized by dysfunctional relaxation of the lower esophageal sphincter and esophageal peristalsis. The severity and review of treatment success of patients with achalasia can be assessed by a severity scoring that utilizes the patient's symptoms known as the Eckardt score. Symptoms used for establishing the Eckardt score include chest pain, regurgitation, difficulty swallowing, and weight loss with each symptom contributing a score of 0-3 based on symptom severity and frequency. The lowest score is 0, the maximum score is 12, and a score of 3 or less following intervention is frequently used as an indicator of treatment success.

Diagnosis of achalasia begins with endoscopy to rule out pseudoachalasia, strictures, or other forms of mechanical obstruction including malignancy. Endoscopy may support achalasia with retained food products, esophageal dilation proximal to the LES, and a difficult to traverse LES, but these findings are not diagnostic of achalasia. Barium esophagram is a common test that is used in the dysphagia work-up that may show the classic "bird-beak" appearance of proximal esophageal dilation and tapered contrast travel across the LES in patients with achalasia as we saw with our patient (Figure 1). The gold standard test used to diagnose and type achalasia is high-resolution manometry. In high-resolution manometry, a manometry catheter with multiple pressure sensors is introduced into the esophagus to measure esophageal, LES, and proximal gastric pressures during 10 wet swallows and during esophageal peristalsis<sup>1</sup>. Achalasia on manometry will show absent or reduced relaxation of the LES during swallowing with absent or spastic peristalsis in the esophagus proximal to the LES. All three subtypes of achalasia will show reduced LES relaxation on manometry. Differentiation between the subtypes can be done by identification of low intraesophageal pressure or a relaxed esophagus in type I, intermittent pan-esophageal pressurization in type II, and spastic, premature contractions in type III<sup>2,3,4</sup>.

Current treatments for achalasia include pneumatic dilation, botulinum toxin injections, laparoscopic Heller myotomy, and E-POEM. Endoscopic botulinum toxin injection of the LES and pneumatic dilation were historically used in patients who are not candidates for E-POEM or Heller myotomy. Inoue and colleagues in 2010 reported their experience of E-POEM in 17 patients with a significant improvement in symptoms with no severe adverse events<sup>5</sup>. Since then, numerous studies have reported the non-inferiority of E-POEM to surgical, laparoscopic Heller myotomy with partial fundoplication. In a randomized, controlled trial of 221 patients of patients receiving E-POEM or Heller myotomy with partial fundoplication, 83% of patients in the E-POEM group and 81.7% of patients in the Heller myotomy group had clinical success at 2-year follow-up assessed by Eckardt score and manometric data over time. Secondary outcomes showed serious adverse events were slightly more common in the Heller myotomy group (2.7% vs 7.3%)<sup>6</sup>. Additionally, Awaiz, et al. conducted a meta-analysis of three studies which showed higher rates of short-term clinical failure in patients that received laparoscopic Heller myotomy<sup>7</sup>.

In this report, we present an interesting case of type II achalasia in a young, male patient and validate that esophageal peroral endoscopic myotomy for the treatment of achalasia is preferred method of treatment for achalasia due to its safety, treatment success, quick recovery time, minimal pain, and low rates of adverse events.



Figure 1. Double-contrast esophagram with proximal esophageal dilation and constricted lower esophageal sphincter (LES) giving the traditional “bird-beak” appearance of achalasia.

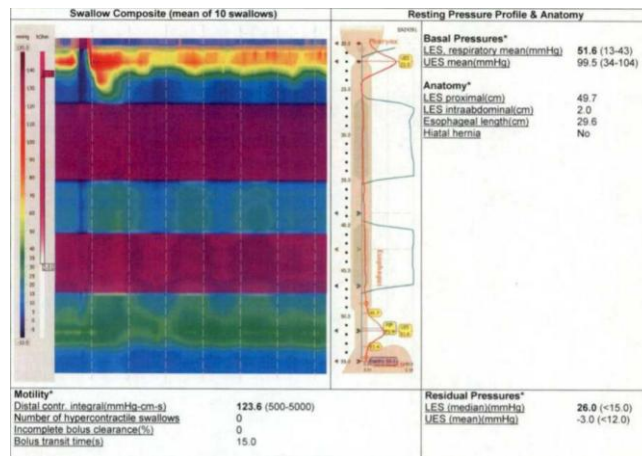


Figure 2. High-resolution manometry findings showing pan-esophageal pressurization with impaired relaxation of the LES consistent with type II achalasia.



Figure 3. Endoscopic assessment of EndoFLIP balloon placement

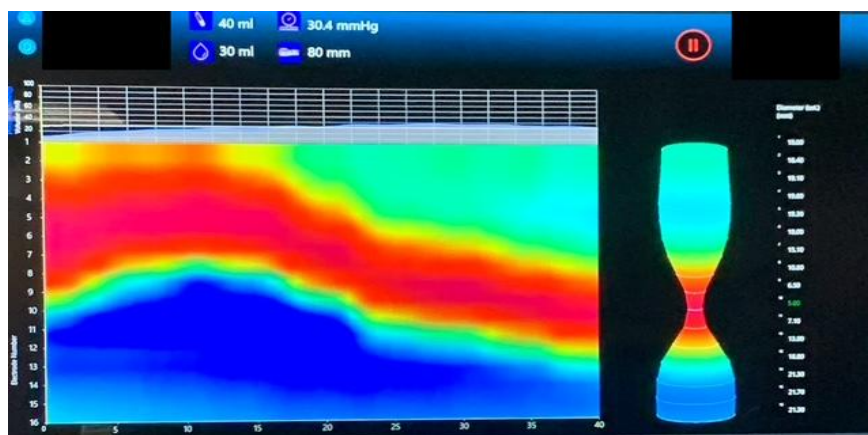


Figure 4. Pre-procedure EndoFLIP measurement showing decreased distensibility index consistent with achalasia at various balloon volumes



Figure 5. Mucosal incision to the submucosal space to form the submucosal tunnel.

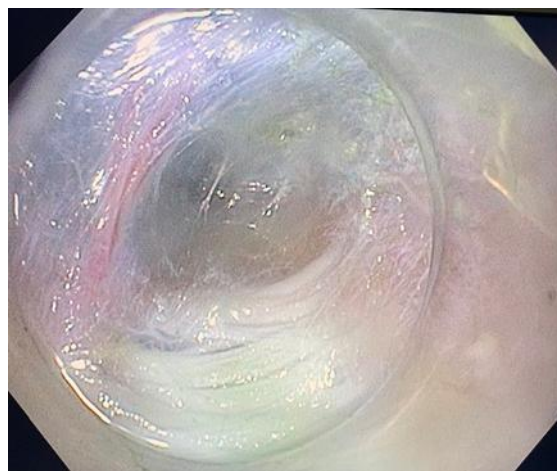


Figure 6. Submucosal tunnel with the intrinsic muscle layer inferior and the esophageal mucosa superior.

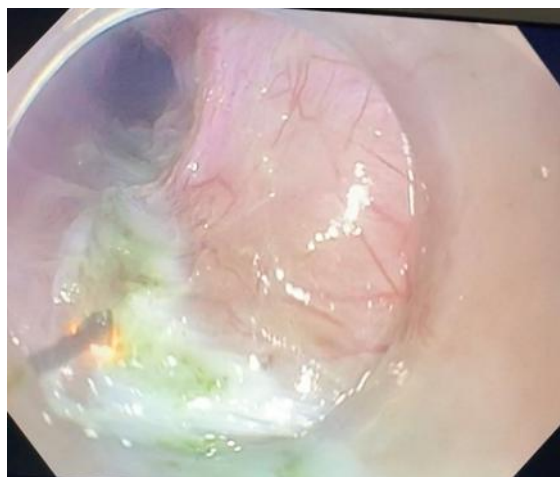


Figure 7. Spray coagulation with a triangle tip knife to dissect the circular muscle fibers of the lower esophageal sphincter.

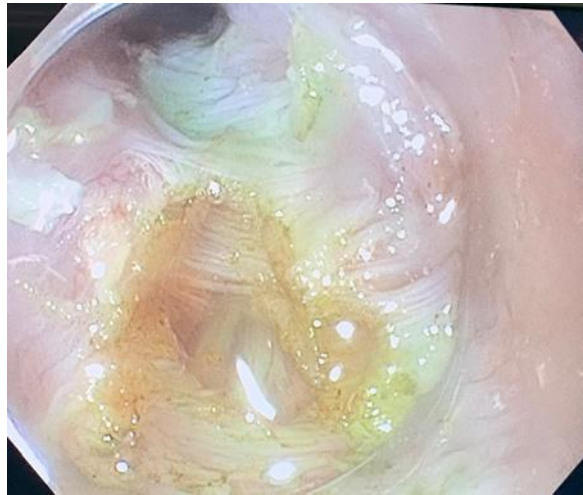


Figure 8. Complete thickness myotomy maintaining longitudinal muscle and adventitia.

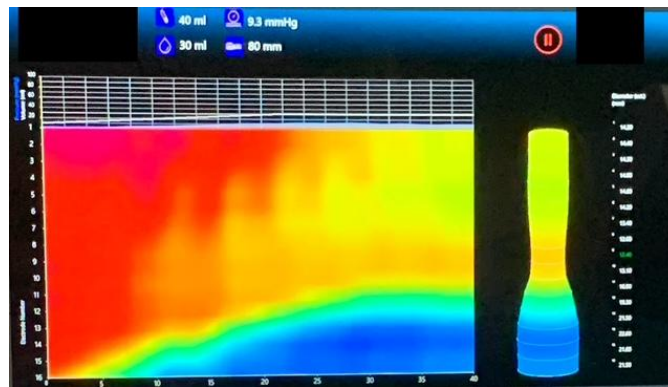


Figure 9. EndoFLIP following completion of the myotomy showing decreased pressurization across the LES with relief of proximal dilation.



Figure 10. Six clips were placed to close the mucosal incision following myotomy completion.

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