International Journal of Contemporary Pediatrics
Tamim T et al. Int J Contemp Pediatr. 2025 Oct;12(10):1733-1737
http://www.ijpediatrics.com
pISSN 2349-3283 | eISSN 2349-3291

DOI: https://dx.doi.org/10.18203/2349-3291.ijcp20252977

Case Report

Achalasia cardia in a child managed with per-oral endoscopic myotomy: a case report

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Received: 13 August 2025
Accepted: 09 September 2025

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ABSTRACT

We present a case of a 12-year-old girl with progressively worsening vomiting and respiratory symptoms, eventually diagnosed with achalasia cardia. Initial imaging and endoscopy suggested esophageal motility disorder, confirmed by high-resolution manometry as type I achalasia. The child underwent per-oral endoscopic myotomy (POEM) procedure after antifungal treatment for incidental esophageal candidiasis. The procedure was successful with complete symptom resolution and weight gain on follow-up. This case highlights the diagnostic challenges in pediatric Achalasia and emphasizes the role of POEM as an effective therapeutic modality.

Keywords: Achalasia cardia, Pediatric, POEM, High-resolution manometry, Esophageal motility disorder

INTRODUCTION

Achalasia is a rare oesophageal motility disorder with an annual incidence of 0.11 per 100,000 children.[1] It results from the degeneration of inhibitory neurons in the myenteric plexus, causing absent peristalsis and failure of the lower esophageal sphincter (LES) to relax. The disorder has a multifactorial origin, potentially linked to infections, autoimmune responses, and genetic predisposition.

Symptoms include progressive dysphagia for solids and liquids, regurgitation, heartburn, and chest pain. Diagnosis involves barium swallow (showing "bird's beak" and esophageal dilation), manometry (gold standard), and endoscopy to exclude malignancy.

Treatment options include pharmacotherapy (calcium channel blockers, nitrates, botulinum toxin), pneumatic dilation, laparoscopic Heller's myotomy, or POEM.

CASE REPORT

A twelve-year-old female child presented to our hospital with complaints of vomiting for the past 30 days, accompanied by fever and cough for 15 days. Initially, the vomiting occurred 1-2 times per day, but it gradually increased to 3-4 episodes daily. The vomitus primarily contained food particles, was non-projectile, non-bilious, and non-blood-tinged. Occasionally, it included sticky, white sputum. The child also reported a persistent wet-sounding cough associated with post-tussive vomiting for the past 15 days. The cough worsened at night, and caused significant sleep disturbance.

Additionally, the child had intermittent fever for 15 days, with moderate to high-grade spikes occurring 1-2 times per day, primarily in the early morning, relieved with medication and not accompanied by chills or rigors. The family also reported a history of weight loss, with the child losing approximately 2 kilograms over the past month.

The child presented with a lump sensation in the throat for the past month, difficulty eating at school over the last 4-5 months, and persistent symptoms despite being treated with cinitapride and pantoprazole. Previous treatments also included courses of amoxiclav for 5 days and azithromycin for 3 days, with endoscopy advised but not performed. The child had no history of contact with TB, chest pain, breathlessness, abdominal pain, rash, bleeding manifestations, ear pain, throat pain, or behavioral or psychiatric issues. She is the second order born to a non-consanguineously married couple with no significant family history of similar issues.

On examination, vitals were stable. General physical examination revealed no lymphadenopathy, normal oral cavity. Multiple hyperpigmented scars of old pyoderma were noted on the legs. Anthropometric measurements were underweighted for age and gender. Systemic examination was within normal limits. Investigations revealed a normal chest X-ray and normal USG of the abdomen and pelvis. Blood tests showed a hemoglobin level of 12.6 g/dL, total leukocyte count of 10,240 with neutrophilic predominance, and an elevated CRP. The child was admitted to the wards and started on symptomatic management. A Gastroenterology consultation was obtained, and an endoscopy was advised.

Endoscopy revealed achalasia cardia along with gastric erosions (Figure 1-3). The biopsy report indicated cystic mucosa with focal erosions of the lining epithelium. The underlying lamina propria showed mild lymphoplasmacytic inflammatory infiltrates, which led to the impression of chronic gastritis. A pediatric surgery opinion was sought, and a barium swallow was performed, showing the characteristic bird beak appearance, which is often associated with achalasia (Figure 4). Based on these findings, Gastroenterology team recommended undergoing esophageal manometry for further evaluation and confirmation of the diagnosis.

High resolution esophageal manometry done showed achalasia cardia type 1, with complete absence of peristalsis (Figure 5).

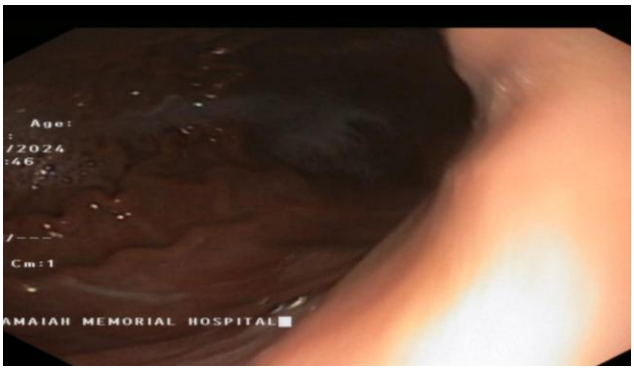

Figure 1: Endoscopy image showing dilated esophageal lumen.

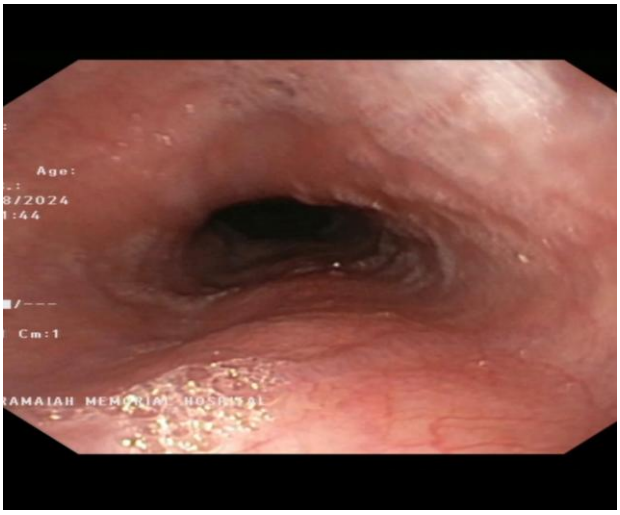

Figure 2: Endoscopy image showing dilated esophagus with retained secretions.

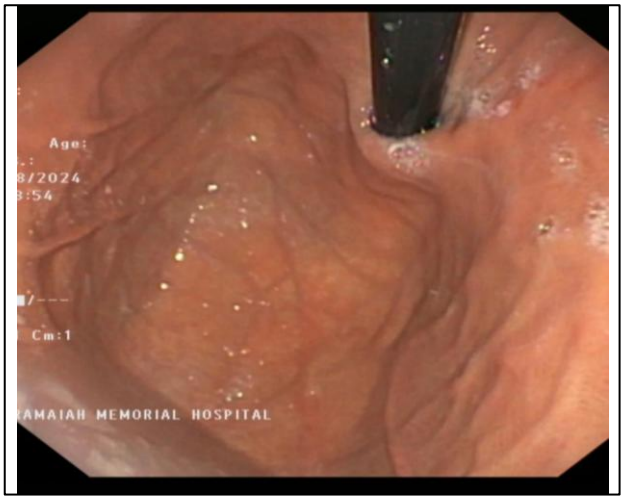

Figure 3: Endoscopy image showing dilated esophagus with narrowed Gastro esophageal junction.

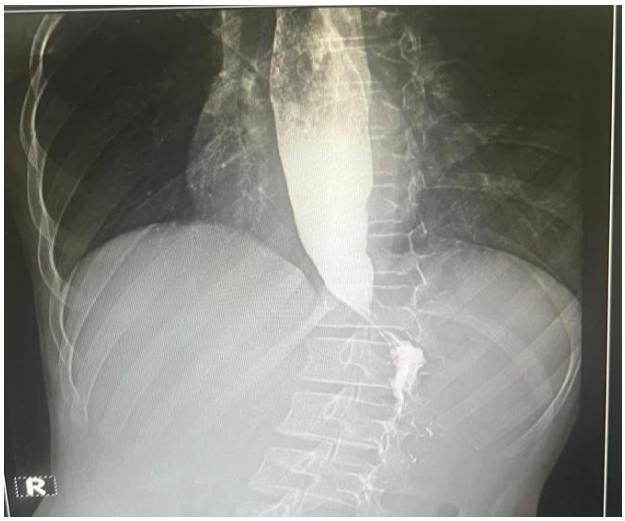

Figure 4: Barium swallow-bird beak appearance.

Figure 5: Esophageal manometry report-showed absent esophageal peristalsis-achalasia cardia type 1.

During the hospital stay, the child had persistent fever spikes and elevated CRP, raising suspicion of pneumonia. She was treated with IV ceftriaxone for five days, which led to improvement. Gastroenterology recommended POEM. The child was taken to tertiary gastroenterology centre for POEM. However, endoscopy revealed esophageal candidiasis, delaying the surgery. The child was started on antifungal treatment for 15 days, and after confirming the resolution of candidiasis with a follow-up OGD, the child underwent POEM surgery on 28.09.2024.

The procedure revealed a dilated esophagus with a tight gastroesophageal junction (GEJ) located at 35 cm. At 25 cm from the incisors, 10 cc of normal saline mixed with methylene blue was injected submucosally at the 5 o'clock position to create a submucosal bleb. A 2 cm long mucosal incision was then made using a T knife on the bleb. The endoscope was carefully advanced into the submucosal space, and a tunnel was created from the 25 cm mark to the GEJ, extending 2 cm beyond it. A myotomy was performed starting at 25cm and continued to the GEJ, with an additional 2 cm beyond the junction. The mucosal incision was closed with hemoclips. Following the procedure, the GEJ appeared relaxed, indicating successful treatment.

Post-procedure, the child's chest X-ray was within normal limits. A check OGD was performed on 30.09.2024, confirming that the POEM procedure clips were in place. A small opening was noted at the incision site, which was successfully closed using an endoscopic-Z clip. No resistance was observed at the GEJ, and there was no mucosal injury. A nasogastric tube (RT) was placed under endoscopic guidance and the child was started on RT feeds which she tolerated well.

A repeat endoscopy after 10 days showed complete healing. Nasogastric tube was removed; patient was started on oral feeds. At the next follow up a month later she displayed a weight gain of 2 kg and was symptom free.

DISCUSSION

This study highlights the use of esophageal manometry for diagnosing achalasia, as X-ray, ultrasound (USG), and endoscopy did not confirm the condition. This also highlights that POEM procedure in children achieves good results. Clinical symptoms of achalasia can be assessed by the Eckardt score which is a standardized and verified scoring system that rates four symptoms of achalasia (dysphagia, reflux, chest pain, and weight loss) based on severity, each on a scale of 0 to 3 on a final 12-point scale, where higher scores indicate more severe symptoms (Figure 6). A score <3 is used to define symptom remission or successful remission.[2]

Achalasia Severity: Eckardt Score

	Score for each symptom/sign			
Symptom/Sign	0	1	2	3
Recent weight loss (Kg)	none	< 5	5-10	>10
Dysphagia	none	occasional	daily	each meal
Chest pain	none	occasional	daily	several times/day
Regurgitation	none	occasional	daily	each meal

Figure 6: Severity of achalasia-Eckardt score.

Although upper gastrointestinal endoscopy and barium swallow may suggest achalasia, esophageal manometry remains the gold standard for diagnosis.[3]

The key features of esophageal manometry include incomplete relaxation of a frequently hypertensive LES and the absence of peristalsis in the tubular esophagus.[4] Esophageal manometry is performed using a low-compliance capillary perfusion system or a solid-state assembly with pressure sensors spaced 1 cm apart. During the procedure, the LES resting pressure and its relaxation response to five wet swallows are measured using a sensor placed in the middle of the LES high-pressure zone. The response to ten wet swallows, each separated by at least 30 seconds, is also tested. The diagnosis of classic achalasia is characterized by the complete absence of peristalsis in the esophageal body, with simultaneous contractions that have amplitudes less than 40 mmHg or no apparent esophageal contraction,

alongside incomplete relaxation of a hypertonic or normotonic LES. These findings confirm the presence of achalasia and help differentiate it from other motility disorders, guiding further treatment options like pneumatic dilation, surgical myotomy, or pharmacologic therapy.[5,6]

We found POEM to be safe and effective in this child, with no major adverse events. Complete symptom resolution and a significant decrease in baseline LES pressure were noted. POEM offers several advantages over laparoscopic Heller myotomy (LHM), including shorter operative time, reduced blood loss, a shorter hospital stays, and the ability to extend the myotomy based on the type of achalasia. Additionally, since POEM does not involve the dissection of the diaphragmatic hiatus or division of the crura, the risk of post-procedure gastroesophageal reflux disease (GERD) is theoretically lower than with LHM. However, follow-up pH studies are necessary to determine the exact incidence of GERD after POEM. Overall, POEM is a minimally invasive procedure with faster recovery compared to LHM.[4] Esophageal motility disorders has evolved many major changes in conceptualization, diagnostic instruments, diagnostic algorithms, and therapeutics since initial use. High resolution manometry (HRM) has proved superiority globally and the Chicago classification, is uniformly accepted as the consensus statement on HRM interpretation. Classification of achalasia in the Chicago classification system has grading as type I-elevated median IRP, 100% failed peristalsis, type II-elevated median IRP, 100% failed peristalsis, panesophageal pressurization with 20% of swallows, type III-elevated median IRP, no normal peristalsis, premature contractions with 20% of swallows.[7]

Therapeutic POEM has largely replaced LHM and pneumatic dilation (PD) as first-line therapy for achalasia, especially spastic achalasia, in esophageal tertiary care centers.[8,9]

As per the American college of gastroenterology guidelines, and recommendation based on the grading of recommendations assessment, development, and evaluation (GRADE) process[8] in young male patients with achalasia, PD with larger balloon sizes (3.5 cm then 4 cm) or myotomy or POEM may be more effective than initial PD with the 3 cm balloon size. They recommend that patients who are initially suspected of having GERD but do not respond to acid-suppressive therapy should be evaluated for achalasia.

The pathophysiology has been explained that the primary etiology of achalasia is believed to be selective loss of inhibitory neurons in the myenteric plexus of the distal esophagus and LES, resulting in a neuronal imbalance of excitatory and inhibitory activity. Excitatory neurons release acetylcholine, whereas inhibitory neurons primarily release vasoactive intestinal peptide and nitric oxide. A localized decrease of vasoactive intestinal peptide and nitric oxide with unopposed excitatory activity causes failure of LES relaxation and disruption of esophageal peristalsis.[10]

Markar et al showed an early success rate of 89%-100% for POEM and that it is highly effective in the management of type III achalasia cardia. A study comparing POEM with LHM found that POEM was safer and even superior to LHM in terms of cost-effectiveness, length of hospital stays, and dysphagia relief.[11,12]

It is important to note that long-term follow-up is necessary for patients with achalasia cardia who undergo POEM, which can be used to monitor clinical, radiological, and manometry therapy success; functional changes in GEJ; and pathological gastroesophageal reflux. The follow-up duration is usually 3-6 months after discharge. The Eckardt score should be assessed, and endoscopy, manometry, and timed barium meal examinations should be performed.

CONCLUSION

Pediatric achalasia is a rare but treatable motility disorder. High clinical suspicion and HRM are essential for diagnosis. POEM is a safe and effective option, offering symptom resolution and improved quality of life with minimal invasiveness.

Funding: No funding sources
Conflict of interest: None declared
Ethical approval: Not required

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Cite this article as: Tamim T, Rao C, Kumar S, Avinash B, Vari KR, Rao A. Achalasia cardia in a child managed with per-oral endoscopic myotomy: a case report. Int J Contemp Pediatr 2025;12:1733-7.