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Peroral endoscopic myotomy for treatment of achalasia in a patient with congenital osteogenesis imperfecta and scoliosis

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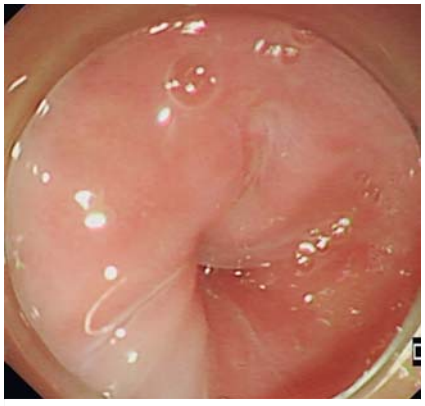
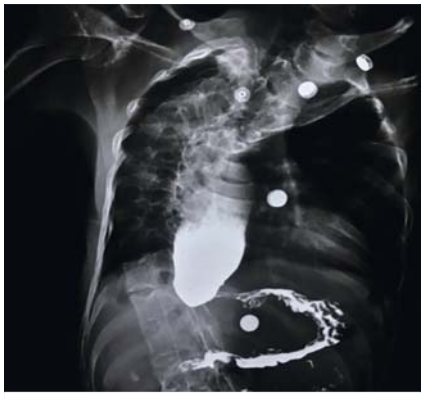
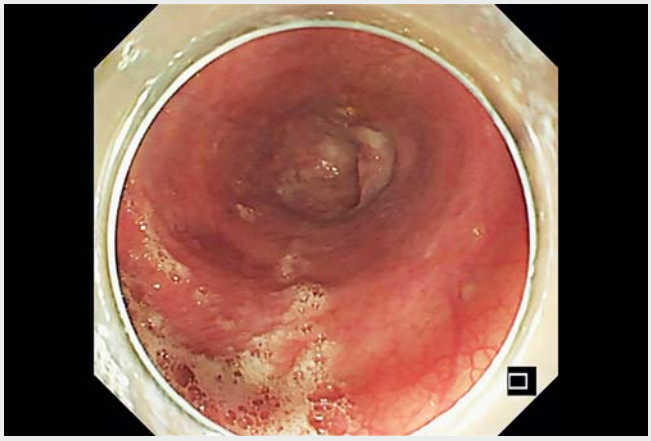


▶ **Fig. 1** Endoscopic image showing narrowing of the cardia consistent with achalasia.



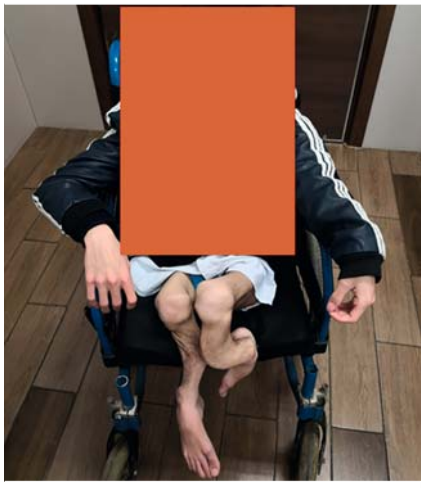
▶ **Fig. 2** Radiographic image from an upper gastrointestinal series showing the bird's beak sign of the cardia and dilatation and distortion of the esophageal lumen.



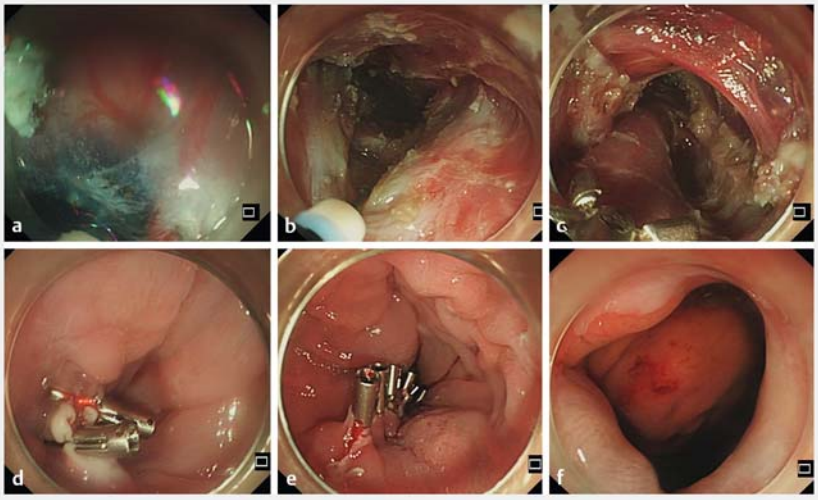
Video 1 The application of peroral endoscopic myotomy for a patient with multiple malformations, thoracocyllosis, and scoliosis, making the procedure more complicated and riskier than normal.

A 22-year-old man was admitted because of dysphagia and regurgitation for half a year. Gastroscopy showed narrowing of the cardia (▶ **Fig. 1**) and the upper gastrointestinal series showed the "bird's beak" sign of the cardia and dilatation and distortion of the esophageal lumen (▶ **Fig. 2**). The patient was diagnosed clinically as having achalasia. In addition, he had a previous history of osteogenesis imperfecta, a rare inherited bone disorder, from birth, with fragile bones that are easily broken. As a result, he had multiple malformations of his arms and legs, thoracocyllosis, and scoliosis, with a body weight of 55 kg and a sitting height of 50 cm (▶ **Fig. 3**). Preoperative pulmonary function tests showed a moderate restrictive ventilatory impairment. Peroral endoscopic myotomy (POEM) was proposed after a full multidisciplinary discussion with the anesthesia, orthopedic, and respiratory departments.

The procedure involved four steps (▶ **Video 1**). First, a mucosal incision was made at 6 o'clock about 8 cm proximal to the cardia. Second, submucosal longitudinal tunneling was performed across the cardia (▶ **Fig. 4 a**). Owing to the thoracocyllosis and distortion of the esophageal lumen, it was important during the tunneling to recognize the direction of the muscle fibers and tunnel, with the tunnel needing to be created more carefully along the muscle to avoid mucosal injury and misdirection. Third, circular muscle myotomy was performed from 1 cm distal to the mucosal entry to 2 cm beyond the cardia (▶ **Fig. 4 b**). After the myotomy, hemostasis was achieved with hot biopsy forceps (▶ **Fig. 4 c**). Finally, the mucosal entry and areas of mucosal injury were closed with clips (▶ **Fig. 4 d, e**). After the myotomy, the cardia was



▶ **Fig. 3** Photograph of the patient showing his multiple malformations, thoracocyllosis, and scoliosis caused by osteogenesis imperfecta, which made the endoscopic procedure more challenging.



▶ **Fig. 4** Endoscopic images showing: **a** the submucosal longitudinal tunnel; **b** circular muscle myotomy being performed; **c** hemostasis with a hot biopsy forceps; **d, e** the mucosal entry and areas of mucosal injury after their closure with clips; **f** the significant enlargement of the cardia after completion of the myotomy.

significantly enlarged (▶ **Fig. 4f**). The procedure duration was 30 minutes. The patient was discharged on postoperative day 6 after an uneventful recovery.

POEM has become widely accepted as a minimally invasive procedure for the treatment of achalasia. Here, we report the first case of achalasia in a patient with osteogenesis imperfecta that was managed by POEM. Owing to the patient's multiple malformations, thoracocyllosis, scoliosis, impaired pulmonary function, and the distortion of the esophageal lumen, POEM was more complicated and riskier than normal. Importantly, the preoperative preparation, intraoperative monitoring, and postoperative nursing needed to be more carefully carried out by multidisciplinary team.

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Competing interests

The authors declare that they have no conflict of interest.

The authors

Zu-Qiang Liu[1 *], Li Wang[1 *], Quan-Lin Li[1,2], Ping-Hong Zhou[1,2]

1 Endoscopy Center and Endoscopy Research Institute, Zhongshan Hospital, Fudan University, Shanghai, China
2 Shanghai Collaborative Innovation Center of Endoscopy, Shanghai, China

Corresponding author

Ping-Hong Zhou, MD
Endoscopy Center and Endoscopy Research Institute, Zhongshan Hospital, Fudan University, Shanghai, China
zhou.pinghong@zs-hospital.sh.cn

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Georg Thieme Verlag KG, Rüdigerstraße 14, 70469 Stuttgart, Germany

* Co-first authors