


Esophagogastrostomy with Linear Stapler Murny Rauf Procedure for Type III Achalasia – Case Report

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ABSTRACT

Introduction: Feeding difficulty is one of the main complaints of patients with achalasia. Achalasia is still considered a rare disorder with unknown etiology. The main goal of the treatment is to increase the patient's quality of life and treat dysphagia to improve food intake. A successful esophagogastrostomy Murny Rauf procedure for achalasia treatment in a limited-resource hospital is presented. **Case**: A 47-year-old woman was diagnosed with type III achalasia (Eckardt's score: 12) and severe malnutrition. The esophagogastrostomy with the Murny Rauf procedure was conducted to increase food intake. The patient gained weight due to improved feeding several weeks after the surgery. **Discussion:** Diagnosis of achalasia is made by clinical symptoms and confirmed by a diagnostic procedure**.** In this case, non-surgical medications were not considered due to the uncertainty of the benefit and difficulties of routine visits due to the far distance of the patient's residence to the hospital which may incur additional transportation costs. **Conclusion**: The esophagogastrostomy Murny Rauf procedure provides advantages in treating achalasia. This procedure gave a new insight into developing an efficient procedure for achalasia in rural areas.

Keywords: Achalasia, case report, feeding difficulty, Murny Rauf esophagogastrostomy.

ABSTRAK

Pendahuluan: Salah satu keluhan utama pasien *achalasia* adalah kesulitan makan. *Achalasia* masih dianggap sebagai gangguan langka dengan etiologi yang belum diketahui. Tujuan utama pengobatan adalah untuk meningkatkan kualitas hidup pasien dan mengatasi disfagia guna memperbaiki asupan makanan. Kami mempresentasikan kasus *achalasia* yang ditatalaksana dengan prosedur *esophagogastrostomy* Murny Rauf di rumah sakit dengan fasilitas terbatas. **Kasus:** Wanita berusia 47 tahun dengan diagnosis *achalasia* tipe III (skor Eckardt: 12) dan malnutrisi berat. Prosedur *esophagogastrostomy* Murny Rauf dilakukan dengan target meningkatkan kemampuan asupan makanan. Beberapa minggu setelah operasi, kemampuan asupan makanan meningkat yang ditandai dengan peningkatan berat badan. **Pembahasan:** Diagnosis achalasia didasarkan pada gejala klinis dan dikonfirmasi melalui prosedur diagnostik. Dalam kasus ini, obat-obatan non-bedah tidak dipertimbangkan karena ketidakpastian manfaatnya dan kesulitan dalam melakukan kunjungan rutin akibat jarak yang jauh antara tempat tinggal pasien dengan rumah sakit, yang dapat menimbulkan biaya transportasi tambahan. **Simpulan:** Prosedur *esophagogastrostomy* Murny Rauf memberikan keuntungan bagi pasien *achalasia*. Prosedur ini memberikan wawasan baru yang efisien untuk kasus achalasia di daerah terpencil. **Angela Djunaedi, Alders Alen Kusa Nitbani, Widhitomo Marino. Prosedur Esofagogastrostomi Murny Rauf untuk Tata Laksana *Achalasia* Tipe III.**

Kata Kunci: *Achalasia*, laporan kasus, kesulitan makan, esofagogastrostomi Murny Rauf.

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INTRODUCTION

Achalasia means "failure to relax" in Greek; it is considered one of the rare motility disorders of the esophagus. Historically, achalasia was first described in 1674 by Sir Thomas Willis. Achalasia was previously attributed to spasms in the cardia and was termed "cardiaspasm". In 1914, Dr. Arthur Hertz made an argument about the absence of normal relaxation at the cardiac sphincter, later proved by Rake in the post-mortem examination of patients with non-organic dysphagia. He found degeneration of the Auerbach plexus, then concluded that the esophageal dilatation was caused by some sort of neuromuscular dysfunction. Dr. Hertz changed the term "cardiaspasm" into "achalasia of the cardia" in 1915, even though the theory was not widely

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accepted at that time.[1]

Idiopathic achalasia is a rare disease that affects all ages and occurs equally in men and women with no racial predilection.[2] The annual incidence is estimated between 1.07 and 2.2 cases per 100,000 individuals.[3] In the United States, it affected 20,000–40,000 patients.[2] The prevalence studies of achalasia in Asia are still lacking. The prevalence of achalasia in Japan was calculated as 7 per 100,000 people, and the incidence is 0.81–1.37 per 100,000 people.[1,4,5]

Dilated esophageal condition in achalasia does have some effect on other mediastinal structures; the increasing pressure caused by dilated esophagus, specifically in the case of megaesophagus, might increase the risk of cardiovascular and respiratory problems, including secondary infection due to aspiration.[6–8] Long-term achalasia would also increase the risk of esophageal cancer due to chronic inflammation secondary to chronic incomplete esophageal emptying.[4] The incidence of esophageal cancer is 1.3% over 7,000 patients treated for achalasia.[9] Life-threatening condition of acute airway compression is also mentioned in some case reports of achalasia.[10,11]

Worsening progression of achalasia and nonspecific symptoms leads to significant lifestyle changes; anxiety and depression are mainly reported in all achalasia patients due to significant social and daily life changes.[12] Kalantari, *et al*., (2021) explained two main phases during the patient's journey in experiencing the achalasia condition: before diagnosis and after diagnosis.[13] Before diagnosis, many patients have been stuck in a loop of unsuccessful treatments due to the similarity and ambiguity of achalasia symptoms to other conditions. Most achalasia patients had to repeatedly visit health care until being referred for diagnosis. As symptoms worsen, some patients try to manage their symptoms by adjusting their diet and eating habits and treating themselves with indigestion remedies. After diagnosis, patients start to understand their conditions and do self-management. Along with medical intervention suggestions, patients start to change their dietary habits, such as the food texture, timing of the meals, and sleeping posture at night; struggling to adapt to the chronic condition of achalasia needs some coping strategy. The uncertain prognosis of achalasia leads to some behavioral changes that might cause patients to feel helpless.[13]

Achalasia is still considered a rare disorder with unknown etiology. Due to the complexity of achalasia, providing a comprehensive treatment in every case would be a special challenge for surgeons. The main goal of the treatment is to increase the patient's quality of life and treat dysphagia to improve food intake. A resource-limited diagnostic modality should not be a hindrance to providing the best treatment. This report presented a case of achalasia treated with the esophagogastrostomy Murny Rauf procedure in a limited-resource general hospital.

CASE

A 47-year-old woman living in a rural area of East Nusa Tenggara suffered from worsening dysphagia over the last two years, accompanied by heartburn and regurgitation for a year. The patient also had difficulty swallowing both liquid and solid food, followed by frequent vomiting. The patient had a history of long-term use of proton pump inhibitor (PPI) with no relief of symptoms.

The patient came in a malnourished condition with a BMI under 18.5 due to her feeding difficulty. No other abnormalities were detected during the physical examination. The Eckardt's score was 12. Plain radiology imaging of *the esophagus-stomach-duodenum* (OMD) in an initial evaluation showed the classic bird's beak deformity, a narrowed lower esophageal sphincter (LES), and a dilated esophagus **(Figure 1)**. According to the Rezende classification, the patient was classified as grade III based on the esophageal dilatation measurement. Due to limited resources, high-resolution manometry couldn't be performed. Esophagogastroduodenoscopy (EGD) found a dilated esophagus starting from medial to distal with pooled undigested food and a narrowed LES without any evidence of tumor **(Figure 2)**. Based on these diagnostic approaches, the patient was diagnosed with achalasia type III. Due to her severe condition of difficulty feeding and living in a remote area, surgical intervention was decided. Esophagogastrostomy Murny Rauf procedure combined with pyloroplasty **(Figure 3)** was conducted. No complication was found during post-surgery follow-up.

The patient was discharged on the 10th day post-surgery. An OMD for post-treatment evaluation at follow-up one week later showed better barium flow than the pre-operative OMD and decreased width of the dilated esophagus **(Figure 4).** The patient had significant clinical improvement in both liquid and solid food intake and gained weight up to 1 kg per week. No complaint of regurgitation, heartburn sensation, or vomiting.

Photo documentation by Angela Djunaedi, Alders Alen Kusa Nitbani, Widhitomo Marino.

Figure 1. Pre-operative OMD showed a dilated distal esophagus and narrowing LES with a "bird beak" appearance.

DISCUSSION

The diagnosis of achalasia is based on clinical symptoms and confirmed by a diagnostic procedure.[14] Clinical symptoms such as dysphagia, heartburn, regurgitation, vomiting of undigested food, and a history of unrelieved dyspepsia with long-term usage of antacid or PPI medication (proton-pump inhibitor) could be suspected as achalasia. Esophageal manometry is the gold standard of diagnostic procedure for achalasia, with findings of peristalsis, poor LES relaxation, and elevation in the baseline of LES pressure.[3,14,15] Unfortunately, esophageal manometry is unavailable in our region.

The OMD was performed as an initial

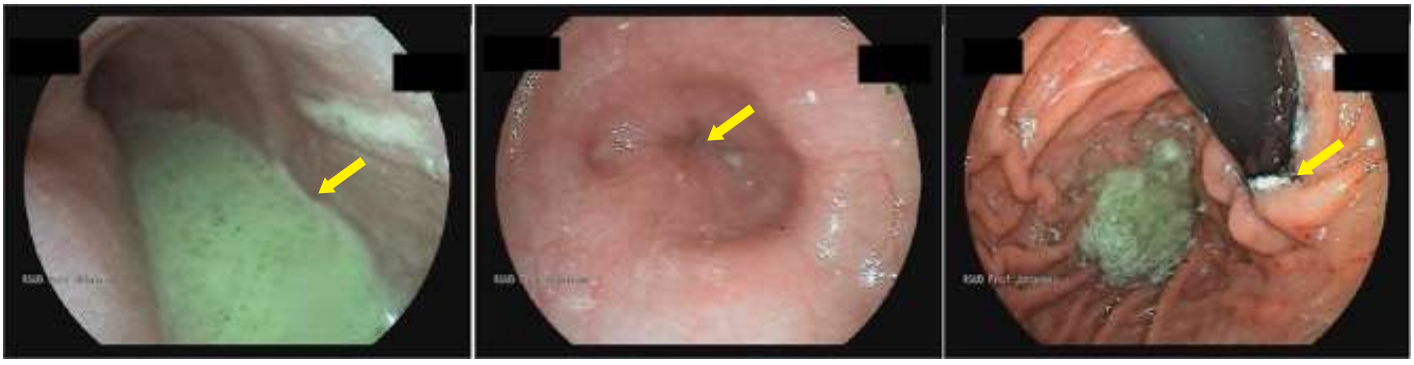

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Figure 2. Esophagogastroduodenoscopy: pooled undigested food at distal esophagus (left), puckering of the gastroesophageal junction (center), rigid scope getting into the LES (right).

Photo documentation by Angela Djunaedi, Alders Alen Kusa Nitbani, Widhitomo Marino.

Figure 3. Intraoperative use of linear stapler to join the distal site of the esophagus and the site of the fundus, replacing the use of sutures.

diagnostic procedure and continued with EGD. Despite the diagnosis confirmation, the EGD was performed to rule out pseudoachalasia and other malignancies.[3] A barium meal OMD radiograph is the imaging procedure suggested by the United Gastroenterology and European Society of Neurogastroenterology and Motility Recommendation.[3] According to Rezende, based on the maximum esophageal dilatation in the barium esophagogram, the esophageal dilatation of 7–10 cm is classified as third grade.[14]

Photo documentation by Angela Djunaedi, Alders Alen Kusa Nitbani, Widhitomo Marino.

Figure 4. Post-surgery OMD showed a better flow of contrast liquid.

The palliative aspect of surgical treatment in achalasia is to reduce LES hypertonicity,[2] with the hopes of increasing the ability to swallow both solid and liquid food to improve the patient's quality of life.[3] In this case, non-surgical medications were not considered due to the uncertainty of the benefit and difficulties of routine visits due to the far distance of the patient's residence from the hospital, which may incur additional transportation costs.

Depression related to achalasia is mainly due to the decreased quality of life.[16] Many studies has found that feeding difficulty, as one of the main symptoms of achalasia, has impacted the patient's psychological status, especially in women.[17,18] Psychological evaluation was not performed in our case due to no signs of depression or anxiety; an early psychological assessment or psychological consultation could be suggested to reduce the risk of psychological distress.[12]

An effective treatment procedure wouldn't directly improve the psychological status.[17] The reduced achalasia-related quality of life was also found to be related to the increased symptoms of anxiety and depression, independent of the severity of the main symptoms.[17] This suggests the relation of increasing psychological burden since the early manifestation of achalasia symptoms.[13] Peroral endoscopic myotomy (POEM) was found to be effective in improving the clinical status of achalasia and suggested to influence psychological effects.[16,19] Most treatment modalities of achalasia, including peroral endoscopic myotomy (POEM),[20] laparoscopic Heller myotomy,[21] pneumatic dilation, and endoscopic injection of botulinum toxin, are unavailable in our region. The options for this patient were through open surgical treatment: open Heller myotomy with Dor fundoplication, Heyrovsky's procedure, esophagogastrostomy, and esophagectomy. Although esophagectomy is required in type III or end-stage achalasia, the most recent International Society for Diseases of the Oesophagus (ISDE) guidelines in 2018 suggest that less intrusive treatments should be the primary focus of management for recurrent symptoms.[15] The care of end-stage achalasia should proceed to esophagectomy if these approaches prove ineffective.[15,22] In this case, the esophagogastrostomy


Heyrovsky's procedure was the surgeon's sole preference.

The esophagogastrostomy Murny Rauf procedure is a modification of the esophagogastrostomy Heyrovsky's procedure using a linear stapler, introduced by Murny Abdul Rauf, an Indonesian surgeon.[23] The Murny Rauf procedure uses a linear stapler instead of a conventional suture to join the distal site of the esophagus and the site of the fundus **(Figure 3).**

In our opinion, as a modification of Heyrovsky's esophagogastrostomy procedure, the indications for the Murny Rauf procedure are similar; it may include type III achalasia with severe malnutrition in regions lacking adequate medical providers. In Deng, *et al*., (2015) study of esophagogastric anastomosis in esophageal cancer, the use of a linear stapler showed a significant decrease in anastomotic leakage and stricture in comparison with hand-sewn anastomosis.[24] Our study showed that the Murny Rauf procedure could greatly impact the patient's quality of life in a rural area with limited resources.

CONCLUSION

The esophagogastrostomy Murny Rauf procedure provides advantages in treating achalasia, including the short duration of the surgical procedure, minimal bleeding, low morbidity, and short post-operation length of stay. This procedure is a new alternative to an efficient procedure for type III achalasia in a rural area with limited resources.

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Conflict of Interests

No potential conflicts of interest to declare.

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