

(RESEARCH ARTICLE)



Heller Cardiomyotomy in end stage of achalasia with mega esophagus: Results in five cases

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Abstract

Achalasia is a motility disorder of esophagus with poor emptying and, characterised by a lack of peristalsis and impaired relaxation of the lower esophageal sphincter with high LES pressure, diameter of esophagus can increase, and some patients progress to develop end-stage achalasia as megaesophagus or sigmoid esophagus and significant form dilation and tortuosity. These complications occur in between 5% and 20% of patients with achalasia. This group of patients if untreated are at risk of aspiration, aspiration pneumonia, and malnutrition.

The management of this type of achalasia, aims to palliate symptoms by improving esophagus emptying and reducing the pressure gradient. In this group of patients, pneumatic dilatation (PD), surgical myotomy, or peroral endoscopic myotomy (POEM) may be less or not effective.

Esophagectomy for the treatment of end-stage achalasia remains a controversial topic and has been recommended by some authors. We describe 5 patients with end-stage achalasia who was been successfully managed with laparotomy heller cardiomyotomy. In 4,12,18,24 months follow-up the condition of patients was good.

Keywords: Achalasia; Dysphagia; Esophageal motility; High-resolution manometry; Megaesophagus; Laparotomy heller cardiomyotomy; Esophagectomy

1. Introduction

Achalasia is a rare motility disorder of the esophagus and in this disease the lower esophageal sphincter (LES) cannot be to relax and the peristalsis in the esophageal is abescent. In the literatures the incidence and prevalence of achalasia is 1.63/100,000 and 10.82/100,000 respectively [1]. But in the new report's incidence increased in the world [2]. In recent years with improvement diagnosis of achalasia may show increase of incidence (1,2). Achalasia patients typically present with dysphagia for solids and liquids material, regurgitation, heartburn, and chest pain and nocturnal cough [3]. These clinical findings are used to evaluate the symptoms of achalasia and efficacy of achalasia treatment (Table 1) [4]. Dysphagia occurs in over 90% of achalasia patients [5]. Dysphagia can be seen others esophageal disorder as

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oropharyngeal dysfunction, esophageal stricture, malignancy, or extrinsic compression (5). In the achalasia, symptoms occur with both solids and liquids materials (3). However, symptoms from mechanical disorders usually occur with swallow of solids foods [6]. Nonspecific clinical presentation of achalasia, may mistake with gastroesophageal reflux disease (GERD), pseudo achalasia due to malignancy, and strictures (5,6). The definitive etiology of achalasia is unknown, and some studies have believed a multi-factorial cause [7]. Some studies suggested a viral or autoimmune inflammatory processes are the cause which leading to degeneration of inhibitor neurons of the esophageal myenteric plexus [3]. diagnosis of achalasia is maddened by barium swallow, chest CT, esophagogastroduodenoscopy and manometry (8). But usually, a definitive diagnosis is made with high-resolution manometry [9]. When achalasia progresses, dilation of the esophagus happen as sigmoidal shape or severs dilatation more than 10 cm in diameter and be tortuous shape from the chest portion towards the gastroenteric junction [10] (Tab 1). Treatment for late-stage achalasia with severely dilated, sigmoidal and tortuous shape of esophagus has been controversial (10,11). Some believe a laparoscopic or laparotomy and Heller myotomy with Dor fundoplication as our approach, (10). Some believe aggressive approach as esophagectomy [10,11]. This article wants show review of the literature and challenges with diagnosis, management, and outcomes with laparotomy heller cardiomyotomy and Dor fundoplication in severe achalasia, sigmoid esophagus and tortuous shape.

Table 1 Stages of esophageal achalasia.

| Stages | Description |
|-----------|--|
| stage 0 | esophageal width of 4 cm or less |
| stage I | esophageal width of between 4 and 6 cm |
| stage II | esophageal width of greater than 6 cm |
| stage III | marked dilation of the distal esophagus >10 cm in diameter, tortuous course, angulation ± axis deviation |

1.1. CASE 1

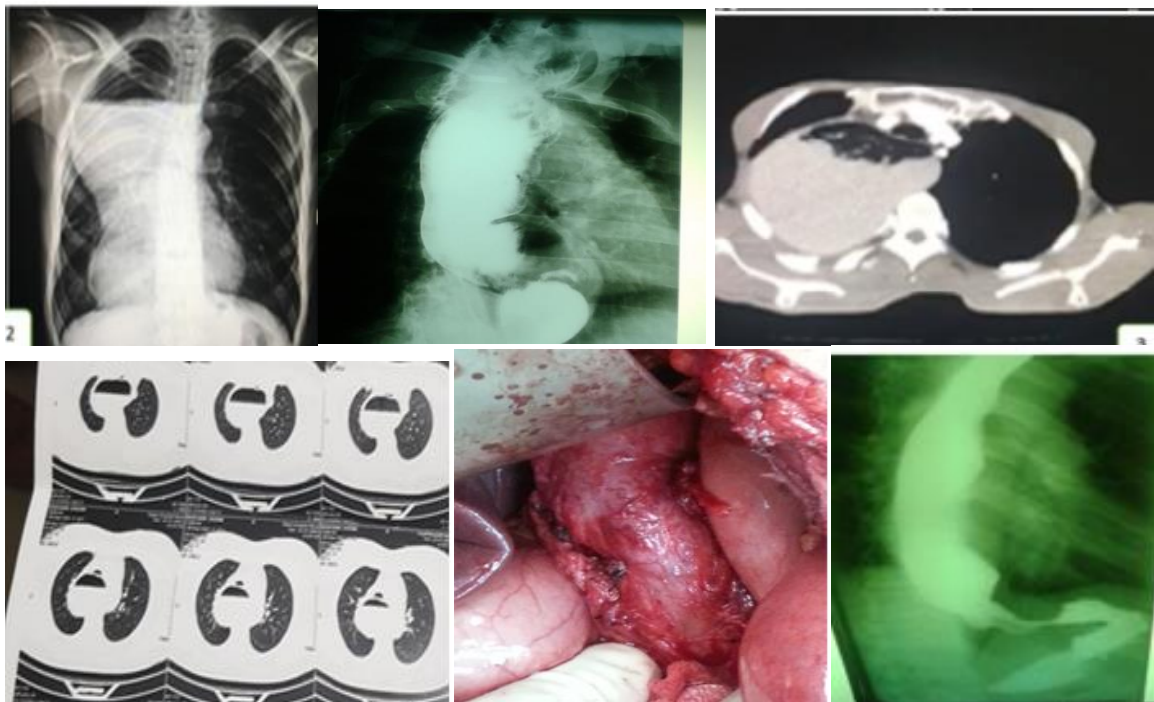


Figure 1 CXR, show barium swallow, CT-scan of chest with sever dilation, cardiomyotom and show post operative barium swallow

A 42-year-old man referred to our department for 3 years of progressive dysphagia, cough, aspiration and regurgitation. Worsening of symptoms in the last 4 months associated with a weight loss. On admission, the patient showed signs of severe malnutrition and halitosis. A chest x-ray and Barium swallow revealed a severely dilated esophageal, with a sigmoid-like pattern was performed in which an apparent mediastinal mass displacing the midline was observed. and a chest computed tomography with oral contrast (CT) scan was performed, in which show megaesophagus. During the esophagogastroduodenoscopy, esophagus was full of food. and a high-resolution esophageal manometry was performed finding evidence of 100% failed waves which diagnosis was type I or classic achalasia. Decision was esophagectomy that is best treatment option for this patient. But patient don't accept this high-risk operation So he underwent to laparotomy heller cardio myotomy (Fig5). After operation tolerance to soft diet and better emptying of esophagus and improvement of his nutritional status (Figure 1).

1.2. Case 2

A 59-year-old woman the referred to our hospital with presentation of worsening aspiration and regurgitation, dysphasia and cough for several months. He had an 8-year history of dyspepsia with dysphagia of solid and liquid food. An upright barium esophagram. showed a dilated proximal esophagus with distal narrowing in a bird's beak morphology of esophagus. A computed tomography (CT) scan showed a markedly dilated esophagus with collection of food in the lumen with this picture suspecting achalasia, esophageal strictures and others lesion as pseudo achalasia. Patient undergo upper GI endoscopy, which showed esophagus is full of remanent food particle and saliva, no evidence of tumor or stricture and esophagoscope passes from distal of esophagus. Diagnosis was achalasia. She underwent esophageal manometry and the achalasia was the definitive diagnosis. After tracheal intubation, for prevention of aspiration, we putt a tracheal tube in the esophagus and aspiration of all content of esophagus. With laparotomy a heller caediomyotomy with Dor fundoplication was performed (3). At 2-year follow-up, she was doing well, with good oral intake and every 6-month esophacogram was performed, the dilatation of esophgouse decreased (Fig2). and the patient.s wight was increase.

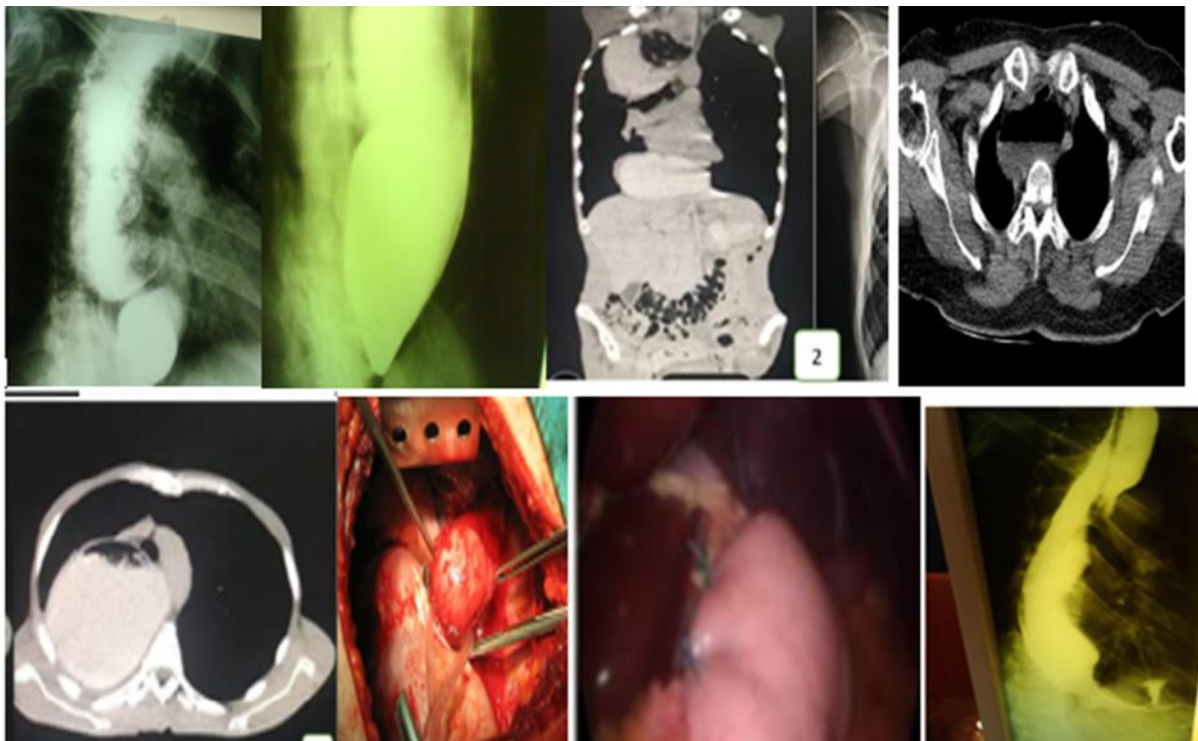


Figure 2 Barium swallow with sever dilation, CT-scan of chest with sever dilation, cardio myotomy, fundoplication and post operative barium swallow

1.3. CASE 3

A 65-year-old woman admit to our hospital due to 5 years of progressive dysphagia to solid or liquid food. Worsening of symptoms as night coughing, aspiration and regurgitation and weight loss in the last 6 months. On admission, the patient showed signs of malnutrition and halitosis. First A barium swallow was performed which show severely dilated

esophageal body, with a sigmoid-like dilatation of esophagus and an enhanced chest computed tomography (CT) scan was performed, in which show megaesophagus.

During the esophagogastroduodenoscopy, esophagouse was full of food and foamy saliva .

Esophageal manometry was performed, finding was evidence of 100% failed waves which was diagnosis of type I or classic achalasia.

For treatment our approach was esophagectomy which was the best treatment option for this patient. But patient don't accept this high-risk operation. She accepted lesser approach as cardiomyotomy (Fig3) and tolerance operation and soft diet. In six months follow -up, improvement of her nutritional status (4).

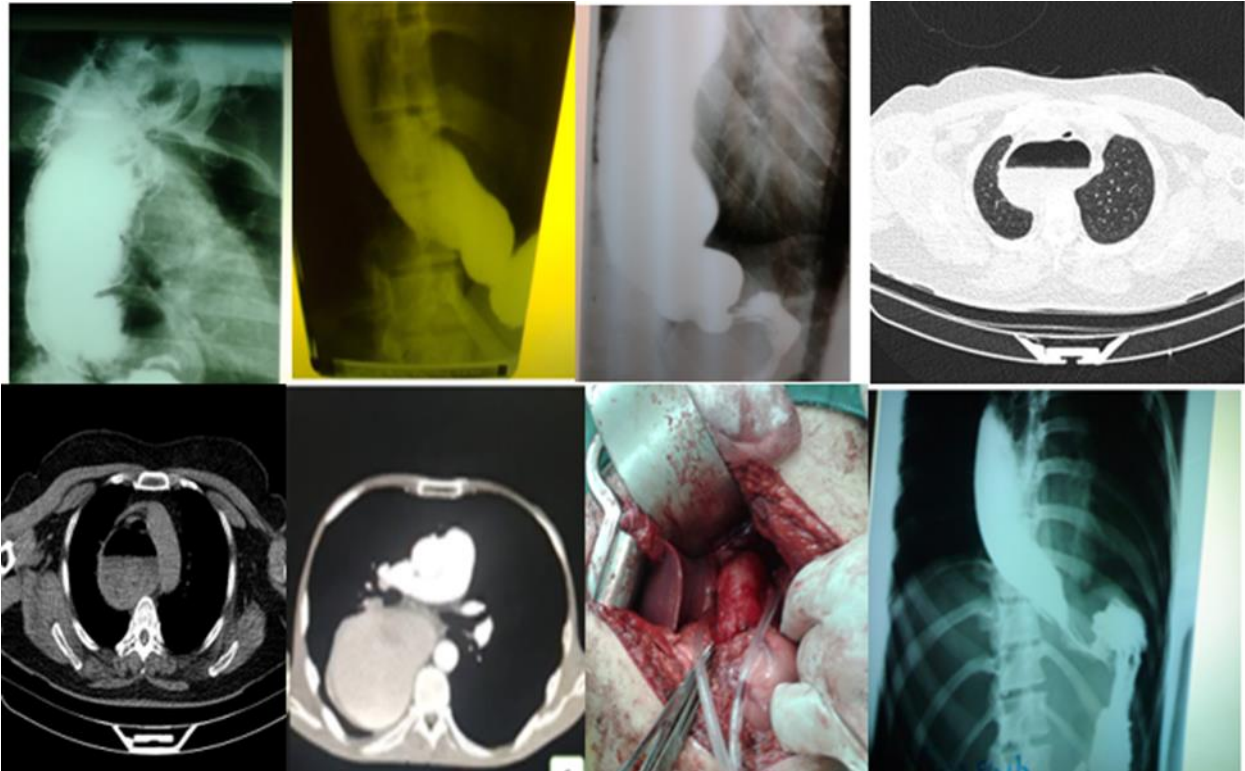


Figure 3 Barium swallow with sever dilation, CT-scan of chest with sever dilation, cardio myotomy, show post operative barium swallow 8-month latter.

1.4. Case 4

A 64-year-old woman with a history of achalasia, referred to our department with grad five of dysphagia on solids and liquids materials, regurgitation, cough, dyspnea and substernal pain after meals for the previous one month. The onset of dysphagia had been 15 years ago, the diagnosis of achalasia was established only at 8 years age. she disagrees with surgery and thus underwent treatments with botulin in two time and two-time pneumatic dilatation and medicated with nifedipine and isosorbide. All this treatment improved the symptoms temporary. On physical examination, hemodynamically was stable and peripheral oxygen saturation of 92% with air room.

A barium swallow performed and show dilation of esophagus (8 cm). These findings were suspicious diagnosis with a megaesophagus due to achalasia which esophagous filled with food. A computed tomography (CT) scan showed a markedly dilated of esophagus with food in it . Upper gastrointestinal endoscopy showed a dilated and tortuous esophageal lumen and multiple particles of small piece of meat and others food was identified and removed some of them, endoscopic balloon dilatation until 25 mm was performed again uneventfully. After two weeks all symptoms recurred. Patient candid for esophagectomy and she refuse the operation because of high complications. she accepts the minor surgery as laparotomy and heller cardiomyoty.

At one-year follow-up, she was doing well, with good oral intake and every 6-month esophagogram performed the dilatation of esophagus was decreased (Fig4). and the patient wight was increased

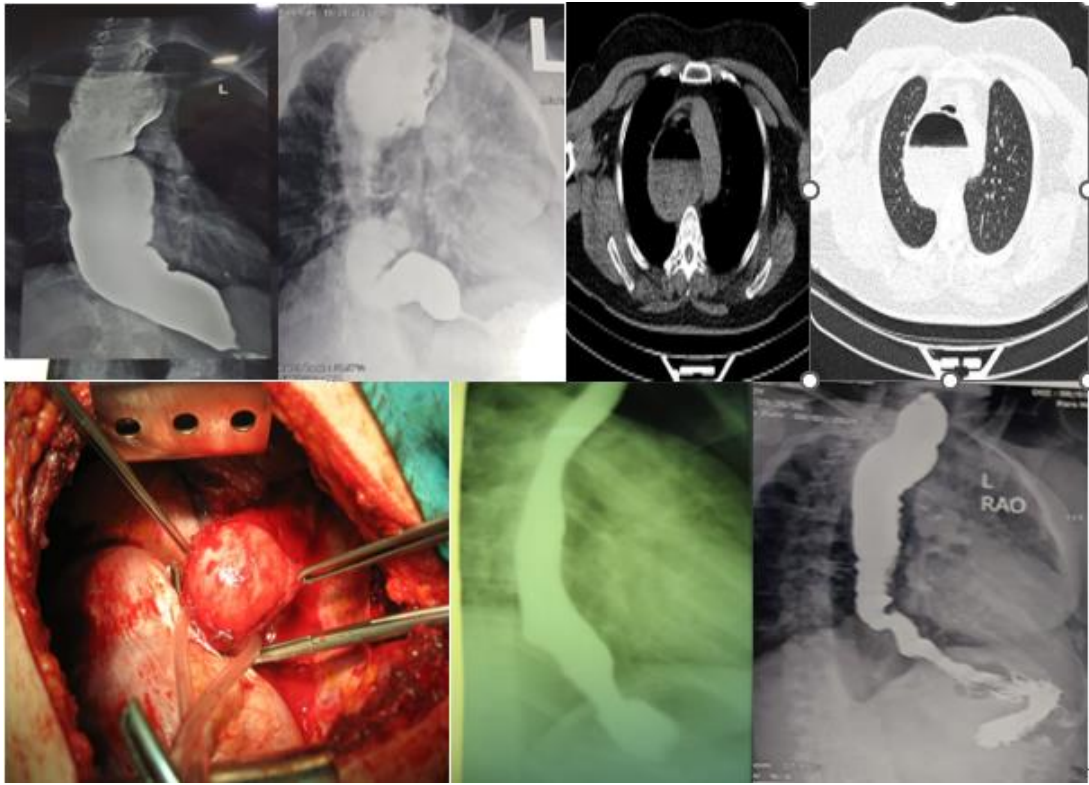


Figure 4 Barium swallow with sever dilation, CT-scan of chest with sever dilation, cardio myotomy, post operative barium swallow 6-month latter.

1.5. Case 5

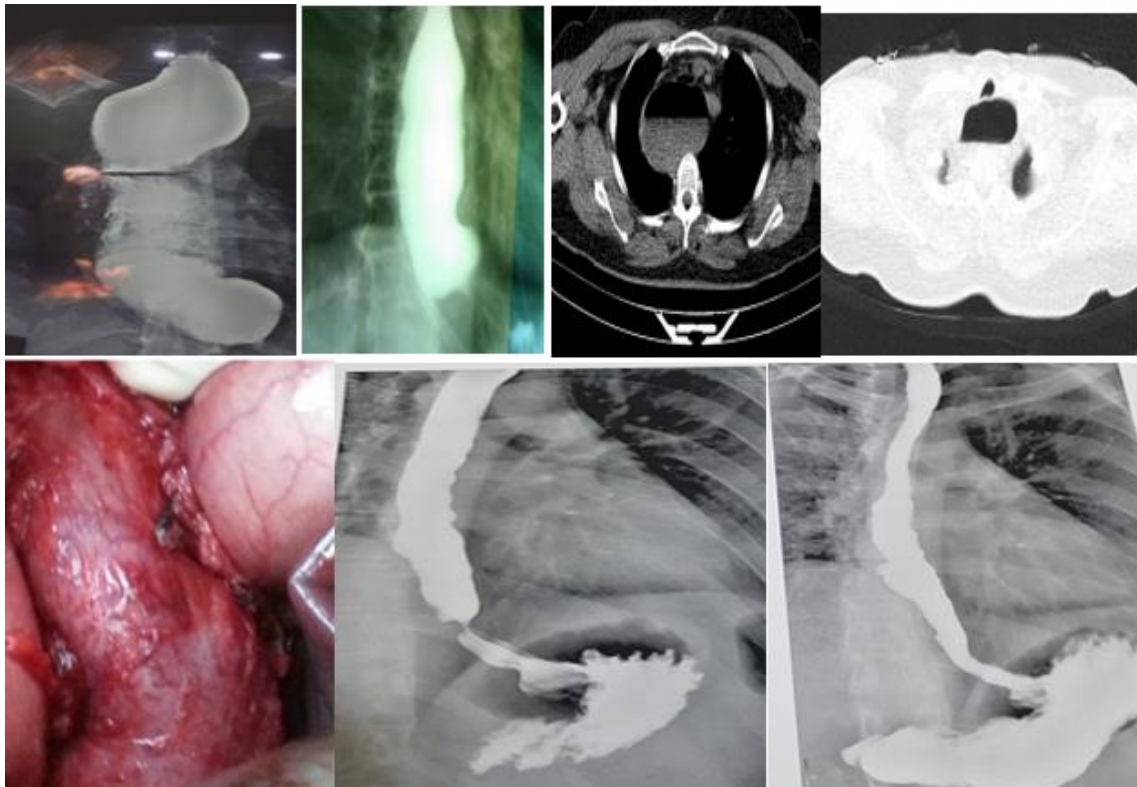


Figure 5 Barium swallow with sever dilation, CT-scan of chest with sever dilation, show cardio myotomy and Dor fundiption, post operative barium swallow 6 and 12months latter.

A 55-year-old man with type I achalasia was referred to our unit with progressively dysphagia, night coughing and regurgitation and aspiration. Other co-morbidities included Type 1 diabetes mellitus which was difficult to control and gastroparesis.

Initial investigations included barium swallow a show dilatation of esophagus. Esophageal manometry was performed show type 1 achalasia A computed tomography (CT) scan showed a markedly dilated esophagus with food fluid material. A endoscopy was performed showing large amounts of fluid and food particle in the dilated esophagus despite a 36-hour fast. The decision was to perform to an esophagectomy after discussion of complication and mortality for patient and his family. The patient his family were disagreed of this operation, and accepted for minor operation.

After laparotomy a long heller cardiomyotomy 3 cm from stomach and ten centimeters on the esophagus and Dor fundoplication was performed . The patient's symptoms improvement gradually in 6-month follow-up. A barium swallow was also repeated showing good emptying of esophagus (Fig 5). In 8,12, and 18 months follow - up the patient was in good condition and satisfaction

2. Discussion

Achalasia is the results of destruction of the myenteric plexus in the wall of the esophagus, which result in fibrous replacement (8). But complete etiology of achalasia unclear (8). Some study believes multifactorial immunological and genetic factors may be the cause of achalasia (9). If achalasia left untreated, can lead to dilatation, and functional of the esophagus loss (10). As a result, end-stage of disease as sever dilated and tortuous esophagus, (greater than 6cm), or sigmoid esophagus can induce (11).

In patients with delay in treatment and deterioration of function can lead to significant complications as malnutrition, pulmonary problem from repeated aspiration, and chronic esophagitis (12). The incidence esophageal squamous cell cancer due to the stasis risk are 3 to 10% (13,14). A study by Tustumi *et al.* show an absolute risk of esophageal adenocarcinoma in patients with achalasia is 18 cases per 100,000 population year (15).

Management of end stage achalasia is controversy because less research about the best treatment is in the literatures (10,11,16). The conserve management are failed, because many patients having undergone multiple failed therapeutic procedures (16). In the literature there are some recombination: First,if pneumatic dilatations failed the patients should undergo a cardiomyotomy, either laparoscopic ,laparotomy or endoscopic (POEM) .Second, if achalasia recurrent despite a laparoscopic or laparotomy Heller's myotomy, pneumatic dilatation is advised. Third, if (POEM) fails to improve symptoms, the patient should undergo either a laparoscopic,laparotomy myotomy or pneumatic dilation. Finally, if of all above strategies fail, esophagectomy should be performed. (31,32,33). In Our hospitals laparoscopic or laparotomy cardiomyotomy used for end-stage achalasia because in esophagectomy morbidity and mortality are high as others reports (16,19,20).

Some authors believe that esophagectomy is an appropriate option for patients with end-stage achalasia (19,20). There are some researches are disagreed with esophagectomy, and still believes less aggressive approach can relive as our study (21,22).

Esophagectomy for management of end-stage megaesophagus in achalasia can be technically difficult with several changes in esophagus anatomy in the pleural and abdominal cavities as Deviation of the megaesophagus into the right chest and these changes increasing the risk of pleural and tracheal injury (16,19,20). Vascularity hypertrophied of esophageal muscle in achalasia haemostasis of the mediastinal vessels is difficult (19,20). Some studies showed slow mediastinal bleeding requiring reoperation (13,16,23,24). Adhesions and scarring of the lower esophagus and proximal stomach, adhesions to the adjacent aorta and left lung can complicate aggressive a esophagectomy (16,31). If esophagectomy is preferred approach is an Ivor-Lewis esophagectomy with an open thoracotomy, and laparotomy is performed (25,26,27,28).

A 3-stage procedure reported, with a thoracoscopic approach in the chest, has also been performed in our institution. However, this approach is only possible if (I) the original myotomy was done by way of the abdomen and (II) an anterior partial fundoplication was performed at the original procedure (26,27,28). In some research they always perform a pyloromyotomy or pyloroplasty to help with gastric emptying, and they prefer a handsewn anastomosis because of dilatation of esophageal lumen, and do not use stapler anvil because of too dilatation of esophagus (26,27,28). But in our report, we use only laparotomy and heller cardiomyotomy usually 3 centimeter over stomach to 8 to 10 CM on esophagus with Dor fundoplication without pyloroplasty or pyloromyotomy.

Open transthoracic or transhiatal is another approach which reported (16,20,23,24,28), and laparoscopic transhiatal also reported (29,30) .and laparoscopic transhiatal approach was reported (29,30). But we believe heller cardio myotomy can alleviate all symptoms with less morbidity and mortality in compression to esophagectomy. Miller *et al.* report that transhiatal esophageal resection was associated with increased morbidity and mortality (20). In contrast, Orringer and co-workers reported that a transhiatal approach is a technique and could be done safely with a lower level of morbidity (24).

Hsu *et al* reported, a left sided thoracoabdominal approach has been rememorated, it provided an excellent operative field with easier mobilization of intrathoracic esophagus with better fundoplication (27).

In esophagectomy patients the morbidity rates are up to 50% (30) and mortality from 0% (11,24-29) to 9% (30).

Post-operative complications as pneumonia, anastomotic leak, bleeding, chylothorax, and wound infection have all been reported (16). Anastomotic leaks or fistulae were noted in 4% to 18% of the patients in the post-operative period. (24,30). The average length of hospital stay in esophagectomy patients in multiple study, was 20 days (11,13,16,20,23,24,26-30). Only 8.3% of patients with the most megaesophagus (for 6 cm) require an esophagectomy after LHM +Dor fundoplication [45,46].

Some authors believe esophagectomy is a viable and safe approach to manage end-stage achalasia (23,24,26-30).

In our study Some patients disagree with esophagectomy because of high morbidity and mortality but agree with lesser aggressive surgery as laparoscopy or laparotomy heller cardiomyotomy after discussion. Also, we believe gastric pullup(stomach) and remnant esophagus in myotomy patients work as a conduit organ without any functions.

Successful treatment of sigmoid-shaped esophagus with laparotomy Heller myotomy (LHM) has been show in some studies, agreed with LHM, as a primary surgical, we performed this operation as a first step in five of our patients with acceptable results [34,35]. To prevent postoperative gastroesophageal reflux (GER) after LHM, addition of an anti-reflux procedure like Dor fundoplication is needed (10,36,37) as in our patients. Dor's fundoplication is more effective and safer procedure for avoiding GER, dysphagia, mucosal perforation, and pseudo-diverticulum [36] as our patients in this report in a study they believe (LHM) had less favorable outcome in patients [37]. As our five patients which had a good outcome.

Sweet et al, reported that Heller myotomy with anterior Dor fundoplication was highly successful even mega or sigmoid esophagus [37]. Swallowing improved in 90% of patients and acceptable palliation of dysphagia, regurgitation, chest pain, heartburn, retrosternal pain, weight loss and recurrent respiratory infections in more than 80% of patients and Patients also saw significant improvements in quality of life [38,39].

Faccani et al. reported the pull-down technique to improve the clinical outcomes of LHM with Dor fundoplication for sigmoid achalasia (aspiration diameter >6 cm), when esophagus kinked to the left and esophageal axis changed [40]. Prior to performing the Heller–Dor procedure, our recommendation is putting a tracheal tube in the esophagus after tracheal intubation and aspired all contented of esophagus for prevention of aspiration during operation time and was successful. Some authors are offered esophagectomy in patients if the size of esophagus (>6 cm), younger age <55 years, recurrent infection secondary aspiration, severe mucosal inflammation and moderate to severe dysplasia [41]. Esophagectomy may be offered after Heller myotomy which cancer present in esophagus and postsurgical scar stenosis of the GEJ those with failure a redo myotomy and if dysphagia is severe and quality of life is not good [41,47].

In some studies, the mean interval from present of dysphagia in patients to establish of esophageal squamous cell carcinoma was 17–21.5 years [43]. In patients with myotomy and present carcinoma was a mean period of 17 years follow-up time [44].

Minimally invasive esophagectomy offers similar symptom relief but hospital stay relative more than to LHM [45,48]. But we do not use this procedure in our patents in this study.

3. Conclusion

Management of end stage of achalasia with sigmoid and tortuous esophagus are a significant challenge and highly controversial in patients. Literature has demonstrated successful treatment of sigmoid or tortuous shaped esophagus with laparoscopic or laparotomy Heller myotomy (LHM). multiple studies show LHM is a primary surgical treatment for sigmoid or tortuous achalasia. It is considered effective procedure are to avoid gastroesophageal reflux, dysphagia,

mucosal perforation or pseudo-diverticulum. Patient education and discussions for patients before this procedure is essential. Like reported in literature, we prefer performing Heller myotomy with Dor fundoplication for patients with severe achalasia and sigmoid or tortuous esophagus as it provides satisfactory relieve symptom improvements and outcomes.

Compliance with ethical standards

Acknowledgments

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Disclosure of conflict of interest

No conflict of interest to be disclosed.

Statement of ethical approval

The study was performed in accordance with the declaration of Helsinki and approved by the Ethics Committee of Guilan by the Local Ethical Committee of Arya hospital. Guilan University of Medical Sciences, Iran

Statement of informed consent

Informed consent was obtained from all individual participants included in the study.

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