Abstract:
Achalasia is the commonest primary esophageal motility disorder, characterized by the failure of lower oesophageal sphincter (LOS) to relax along with loss of peristalsis in the oesophageal body and increased resting pressure of the LOS. Patients present with long-standing progressive dysphagia for both liquid and solid, other symptoms include regurgitation, respiratory symptoms due to chronic aspiration and weight loss due to nutritional failure. End stage achalasia is characterized by dialated and tortuous sigmoid oesophagus in patients previously treated with either pneumatic dilation or myotomy and failure of further such therapy to relieve symptoms. Oesophagectomy for such cases can help achieve good symptom control, improve nutrition and enable a good quality of life. We present a series of 3 cases of end stage achalasia managed by transthalal oesophagectomy and gastric pull up followed by cervical oesophago-gastric anastomosis and review the literature on management of end stage achalasia.

Keyword: ACHALASIA, OESOPHAGECTOMY

1. INTRODUCTION
Achalasia is the commonest primary esophageal motility disorder, characterized by the failure of lower oesophageal sphincter (LOS) to relax along with loss of peristalsis in the oesophageal body and increased resting pressure of the LOS. It results from progressive neuronal degeneration of the myenteric plexus of Auerbach.[1] The pathophysiology of the process represents a selective loss of inhibitory nerves that result in unopposed stimulation of the smooth muscle fibers of the LES. The etiology of primary achalasia remains controversial. Histological examination of the esophagus in achalasia suggests that the reduction in intramural ganglion cells may be a secondary change probably due to inflammation triggered by autoimmune mechanism or a chronic degenerative process of the central or peripheral part of the vagus nerve.[2] The primary lesion could also be a severe myopathy of the smooth muscle cells.[2,3] The precise etiology remains unknown.

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Achalasia is a rare disease, with an estimated annual incidence of 0.5 cases per 100,000 people, and a prevalence of 8 cases per 100,000 people per year. In developing countries the peak incidence is in the third decade, a decade earlier than in developed countries. Typically, patients present with long-standing progressive dysphagia for both liquid and solid, other symptoms include regurgitation and weight loss, and rarely respiratory symptoms due to aspiration. Retrosternal pain is a prominent feature and may result from regurgitation. It can also be due to food stagnation and bacterial growth in the distal esophagus or fermentation of retained food and subsequent oesophagitis.

In developed countries manometric diagnosis is obtained before therapeutic intervention. The most important finding is aperistalsis of the body of the esophagus in the distal smooth muscle segment of the esophagus. Other manometric findings that characterize achalasia are failure or incomplete relaxation of the LES, sustained hypertension of the LES and increased intra esophageal body pressure. Barium swallow is an important diagnostic aid in patients with symptoms suggestive of achalasia. With increasing duration of disease, there is continuous dilatation of the esophagus until it become tortuous or even ‘sigmoid’ in appearance. Oesophageal manometry, barium swallow and 24 hour pH-metry are also used for objective evaluation of outcome of treatment. Other diagnostic investigations include esophago-gastroduodenoscopy, and high frequency intraluminal esophageal ultrasonography. Chest x-ray may show mediastinal widening, presence of air-fluid level in the mid-esophagus, absence of gastric air bubble and abnormal pulmonary markings due to chronic aspiration. In our setting as in others with limited facilities, diagnosis of achalasia is made to a high degree of accuracy given the symptom complex of long-standing dysphagia, regurgitation and weight loss with barium swallow showing delayed emptying, dilatation of esophageal body and the classic bird beak deformity of the distal esophagus.

The treatment aims to palliate, through decreasing the pressure gradient across the LOS to and improve gravitational oesophageal emptying as no treatment is available that will restore oesophageal peristalsis. The two most commonly used therapeutic options for achalasia are pneumatic dilatation (PD) and surgical myotomy. Achalasia can present with a wide spectrum of disease severity, and while some patients achieve good symptom control from a pneumatic dilation, other patients will be needing other interventions to control symptoms. In the absence of intervention, or inadequate therapy, progressive dilatation with increasing tortuosity of the oesophagus can occur, resulting in end-stage disease. End stage disease, characterised by a markedly dilated and tortuous “sigmoid” oesophagus and recurrent symptoms which, may necessitate oesophageal resection in order to normalise gastrointestinal function, improve nutrition and reduce the risk of aspiration pneumonia.

In this report, we aimed to present our experience in the management of patients with end stage achalasia and review literature on oesophagectomy for end stage achalasia.

An Initiative of The Tamil Nadu Dr M.G.R. Medical University
University Journal of Surgery and Surgical Specialities
2. CASES
The following cases of end stage achalasia were managed by us from Jan 2009 to July 2011.

2.1 Case 1
A 46 year old male who had 15 year history of achalasia was referred with severe dysphagia and regurgitation. He had undergone laparoscopic cardiomyotomy with Dor Fundoplication 8 years back with good symptom control for 6 months. Following symptom recurrence, the patient learned to live with symptoms till he developed weight loss and nutritional disturbances. At this time, he was referred to our unit. Initial investigations included a barium swallow, a CT thorax and an endoscopy. The barium study revealed a markedly dilated tortuous "sigmoid" oesophagus with food debris, a tapered narrowing oesophago-gastric junction and markedly delayed emptying of the oesophagus (fig 1). Endoscopy confirmed a fluid-filled mega-oesophagus. The CT scan demonstrated the megaesophagus and signs of aspiration pneumonitis. A transhiatal oesophagectomy was carried out via abdominal midline and left neck incisions with blunt mobilisation of the mega-oesophagus (fig 2), gastric pull-up and a cervical anastomosis. Despite moderate peri-oesophageal adhesions, there were no significant intraoperative difficulties, and a transhiatal approach was safely performed. The approximate intraoperative blood loss was 400 ml. Routine post-oesophagectomy care was provided, including naso-gastric tube drainage, adequate analgesia, and enteral feeding via a jejunostomy tube placed intra-operatively. He was discharged home twelve days postoperatively. Histopathological examination of specimen showed hypertrophy of circular muscle fibers of distal oesophagus with aganglionosis. Patient had good symptom control and gained weight.

2.2 Case 2
A 57 year old female was admitted for recurrent respiratory tract infection 2 years back and was evaluated. She had complained of regurgitation for the past 2 years without any obvious dysphagia. During that period, UGI endoscopy revealed distended esophageal lumen with diverticula in mid oesophagus, impaired peristalsis and a tight oesophago gastric junction that does not distend with air insufflation. Pneumatic dilatation was proceeded with, following which the patient had symptom control for 9 months. She developed dysphagia and was again re-hospitalised with respiratory infection. Barium oesphagogram (fig 3) was ordered, which showed hold up of contrast in the lower 1/3 of oesophagus, tertiary contractions and a Diverticula in retrocardiac segment of thoracic esophagus. Ct scan (fig 4,5) of the thorax revealed bronchiectatic left lingular lobe, acinar opacification in left

![Fig 2 transhiatal oesophagectomy specimen showing mega oesophagus](image)
lower lobe aspiration pneumonitis and a dilated oesophagus. A transhiatal oesophagectomy was carried out via abdominal midline and left neck incisions, gastric pull-up and a cervical anastomosis. The approximate intra-operative blood loss was 450 ml. The resected specimen revealed a dilated oesophagus with a diverticulum. Histopathological examination was consistent with achalasia.

Routine post-oesophagectomy care was provided, including naso-gastric tube drainage, adequate analgesia, and enteral feeding via a jejunostomy tube placed intraoperatively. She required post operative ventilator support for 6 days and had developed left recurrent laryngeal nerve palsy. Patient was discharged on 20 days postoperatively and remains symptom free with good quality of life.

2.3 Case 3

A 77 year old male, known case of coronary artery disease on pacemaker therapy with history of coronary artery bypass grafting presented with dysphagia for 3 months. Evaluation was consistent with diagnosis of achalasia cardia and pneumatic dilatation was carried out. He had symptom recurrence a month later and was referred to us, the patient wanted alleviation of his symptom with a single therapeutic intervention. Barium oesophagogram revealed a dilated and tortous distal oesophagus, a tapered narrowing oesophago-gastric junction and markedly delayed emptying of the oesophagus (fig 6,7). UGI endoscopy showed distended esophageal lumen with impaired peristalsis.

A transhiatal oesophagectomy was carried out via abdominal midline and left neck incisions with blunt mobilisation of the dilated oesophagus (fig 8), gastric pull-up and a cervical anastomosis. No significant intra-operative difficulties were encountered, and a transhiatal approach was safely performed. The approximate intra-operative blood loss was 450 ml. Histopathological examination was consistent with achalasia. Routine post-oesophagectomy care was provided, including naso-gastric tube drainage, adequate analgesia. He was discharged home twelve days postoperatively. The patient continues to remain symptom free with a good quality of life.

3. Review of Literature

Thorough search for reference of literature review were made using Pubmed, Medline and Cochrane database using the keywords “oesophagectomy” and “achalasia”. Identified articles were investigated for reference and are presented in table 1.
4. Discussion
End stage achalasia occurs in less than 5% of all achalasia patients, and may be characterised both clinically and radiologically. Radiological features include a tortuous (sigmoid) and enormously dilated oesophagus, usually more than 6 cm in diameter. Clinically, patients present with severe dysphagia or regurgitation, and nutritional failure.

The management of patients with end stage achalasia is difficult because of previous interventions, nutritional disturbances, and reflux leading to chronic bronchopulmonary aspiration.

Patients reporting with symptom recurrence following primary therapy either in the form of “myotomy” or “pneumatic dilatation” should be evaluated with endoscopy, manometry and contrast studies for identifying the cause of recurrence, which is mostly due to treatment failure. About 10% of post-myotomy patients will have recurrence, and 5% of them need reoperation. Patients who have only undergone pneumatic dilatation should be offered surgical myotomy. Patients with failed myotomy should be offered re-do myotomy, or oesophagectomy in those patients with a highly tortuous and dilated sigmoid oesophagus. Patients with failed re-do myotomy should be offered oesophagectomy if symptoms are severe and quality of life is affected.

Treatment options for patients with end stage achalasia are unfortunately limited and oesophageal resections can be offered for selected people. Patients with massively dilated oesophagus (>6cms), highly tortuous sigmoid oesophagus, with severe dysphagia or reflux associated with nutritional failure are considered to be better candidates. All of our patients, who underwent oesophageal resections show excellent symptom control, which is similar to the results observed in review of literature.

Oesophageal resection in end stage achalasia is technically more demanding as the anatomy is altered due to previous interventions in the form of myotomy which can produce adhesions and scarring making mobilization difficult. Also mega-oesophagus as a rich blood supply, making hemostasis more difficult in mediastinal dissection. Mortality rates of upto 5-10% have been reported, hence the need for proper patient selection is needed. Anastomotic leaks form the major part of morbidity along with anastomotic strictures in the long run.
Several approaches have been suggested including transhiatal, transthoracic, thoraco-abdominal. Advocates for transthoracic approach emphasis, the better visualization and hemostatic control. But we routinely follow transhiatal technique with gastric pull up and cervical anastomosis. For reconstruction, gastric pull through is most commonly followed, although colonic interposition as also been suggested. Recently laparoscopic transhiatal approach as been presented by some centres, and it is feasible along with the advantages of minimal access surgery.\cite{10,18,21} Recently published studies have tried laparoscopic Heller myotomy for extreme megaesophagus as an alternative to esophagectomy and they report relief of preoperative symptoms, but the study sample is small (n=4). \cite{24} Also, Laparoscopic Hellers Myotomy with Dor’s fundoplication (LHM + Dor), as also been reported as a alternative to oesophagectomy, and suggested as a first treatment option for end stage achalasia before oesophagectomy can be contemplated. \cite{25} But Tra shiatral esophagectomy with gastric pull-up should be considered the preferred procedure for end stage achalasia as it can be performed with low morbidity. \cite{14,15,16,17,19,26}

![fig 7 barium oesophagogram showing dilated oesophagus with delayed emptying](image7)

In conclusion, our experience in management of patients with end stage achalasia, combined with results of several larger studies highlight the therapeutic value of oesophagectomy in resolution of symptoms, improving nutrition and enabling a good quality of life.

![fig 8 THE specimen showing the lumen of a dialated oesophagus](image8)

References


<table>
<thead>
<tr>
<th>First Name</th>
<th>Year</th>
<th>Procedure</th>
<th>Description</th>
<th>Mortality</th>
<th>Morbidity</th>
<th>Outcome</th>
<th>Methodology</th>
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<tr>
<td>Peters</td>
<td>1995</td>
<td>Left Thoracoabdominal</td>
<td>Colon interposition</td>
<td>0%</td>
<td>21%</td>
<td>93% felt “cured” with excellent quality of life</td>
<td>Oesophagectomy is a safe and effective therapy in end stage achalasia</td>
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<td>Tark</td>
<td>2009</td>
<td>Transhiatal</td>
<td>Gastric, cervical anastomosis</td>
<td>5%</td>
<td>50%</td>
<td>100% had normal swallow</td>
<td>Oesophagectomy can be performed with acceptable mortality in patients with achalasia</td>
</tr>
<tr>
<td>Pailanveu</td>
<td>2008</td>
<td>Laparoscopic transhiatal</td>
<td>Gastric, cervical anastomosis</td>
<td>9%</td>
<td>Anastomotic leak: 10%</td>
<td>Symptometric improvement in 52%</td>
<td>Laparoscopic oesophagectomy is a safe and effective procedure in specialised centres</td>
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