

# Management of achalasia in the gastroenterology and hepatology teaching hospital

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## ABSTRACT

**Background:** Achalasia is the commonest specific primary esophageal motility disorder for which there is no cure. It causes dysphagia, regurgitation and loss of weight. The objective of this study is to evaluate the management of achalasia in the gastroenterology and hepatology teaching hospital. **Patients and Method:** A thirty patients were diagnosed to have achalasia based on clinical, radiological, and endoscopic criteria, during the period from December 2009 to January 2012. Thirteen patients had Heller's myotomy through trans-thoracic, abdominal and laparoscopic approaches and seventeen patients had pneumatic dilatation. The follow up of patients was done through clinic visits and the enquiry about relieving from dysphagia and any symptoms developed after intervention was done. The follow up period ranged between 4-19 months. **Results:** Fourteen patients were males with a median age of 30.41 years, and sixteen patients were females with a median age of 30.93 years. Dysphagia was the most common symptom followed by weight loss and regurgitation. In the surgery patients group excellent results occurred in 12 patients (92.3%), and 1 patient (7.6%) had poor result with reflux, while at the dilatation group 12 patients (70.5%) had excellent results, 4 patients (23.5%) with good results and 1 patient (5.8%) had fair result. **Conclusion:** Pneumatic dilatation and surgical myotomy both can be used as primary treatment for achalasia. Excellent results with long term relieve of dysphagia is better achieved with Heller's myotomy. **Keywords:** achalasia, management

## Introduction:

Idiopathic achalasia is the commonest specific primary esophageal motility disorder. The term achalasia is a Greek word means (failure to relax). The etiology and pathogenesis of the disease remain unknown. The commonest neuro-anatomical change seen is a decrease or degeneration of the myenteric ganglion cells with some neural fibrosis and variable degrees of chronic inflammation within the myenteric plexus. Recent studies showed selective destruction of non-cholinergic, non-adrenergic inhibitory neurons. The transmitters affected are nitric oxide and vaso-active intestinal peptide. This disorder is irreversible. Auto-immune and infectious factors are the most commonly accepted possible etiologies and auto-antibodies to myenteric neurons are present in as many as 50% of patients with idiopathic achalasia [1,2,3,4,5]. Loss of propulsive peristaltic contractions and defective lower esophageal sphincter (LES) relaxation result in stasis of food in the esophagus which progressively dilates and lengthens and thereby assumes a sigmoid shape in advanced cases. The mucosa of the esophagus often shows esophagitis with mucosal ulceration. This is largely due to stasis and bacterial proliferation and fermentation consequent to retention of food debris (2,3).

Autopsy studies revealed that patients with achalasia have a thicker esophageal muscle compared with that in normal subjects and using high frequency intra-luminal ultrasound imaging, it is found that LES and esophageal muscle thickness as well as esophageal muscle cross sectional area are greatest in achalasia than other esophageal motility disorders (4). Achalasia of the esophagus affects males and females at all ages with equal sex incidence. The main symptom of achalasia are dysphagia, regurgitation and chest pain. Dysphagia is mainly for solid food and of gradual onset with average duration of 2 years (2,3,6,7). The manometric profile that characterizes achalasia includes absence of peristaltic contractions within the esophageal body and incomplete relaxation of high pressure zone in response to swallowing (2,3,6). There appears to be some vague correlation between the degree of esophageal contraction and tortuosity seen on X-Ray film and the duration and severity of obstructive esophageal symptoms (7). The essential esophagogram features include bird's-beak appearance of LES with incomplete opening, loss of primary peristalsis and delayed esophageal emptying (6).

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All patients with suspected achalasia should undergo upper G.I endoscopy with close examination of the cardia and gastro-esophageal junction. At endoscopy, the esophageal body usually appears dilated, atonic and often tortuous. Sometimes the mucosa is reddened, friable, thickened or even superficially ulcerated. The LES region remains closed with air insufflation, however the endoscope will easily traverse this area with gentle pressure allowing examination of the stomach. If excess pressure is required the presence of pseudo-achalasia should be highly suspected (9). Currently there is no treatment that restores esophageal peristalsis and normalizes LES relaxation. Therefore, the existing treatment of achalasia focus on reducing the pressure gradient across the LES, thereby improving the gravitational esophageal emptying. The available treatments for achalasia includes: pneumatic dilatation, surgical myotomy and endoscopic injection of Botulinum toxin. Some benefits may be derived from the administration of long acting nitrates and calcium channel blockers with initial response ranging from 50-70% (1,2,3,4). Surgical therapy shows the best long term results, especially in young patients (8). Hellers introduced esophageal myotomy in 1914 by division of the muscle fibers of distal esophagus with an anterior and posterior incisions through abdominal approach (Double myotomy) this technique is later modified by Groeneveldt and then Zaajier to single anterior myotomy (7,8). There is actually no agreement about the length of myotomy in either its proximal or distal extent and there is no agreement whether an anti-reflux procedure should be added or not and even more controversy about what anti-reflux procedure to perform. The most common cause for imperfect late result was failure to relieve completely the obstruction (7). There are 3 ways to tell that the myotomy is complete:

- 1- visual inspection of the esophagus and cardia while the esophageal lumen is distended with a large dilator.
- 2- The use of intra-operative manometry which may be unreliable (9,10).
- 3- The use of intra-operative endoscopy to determine whether there is residual constriction of the channel between the esophagus and the stomach (10,11,12).

#### **Patients and method:**

This is a retrospective and a prospective study representing the treatment of achalasia in the GIT center during the period from December 2009 January 2012. The study includes 30 patients who were diagnosed to have achalasia on the basis of clinical presentation and proved by BA. and OGD which showed the classical pictures of achalasia, which are dilated esophagus with abnormal or

absent motility of esophageal body and birds beak appearance of the lower end on esophagogram. OGD showed tight cardia with dilated esophagus. Of the study group 17 patients had dilatation and 13 patients had esophageal Heller's myotomy done through thoracotomy, laparotomy and laparoscopic approach.

#### **Thoracotomy**

Under general anaesthesia with double lumen tracheal intubation and the patient in the right lateral position, incision done through the left 7th intercostal space. Division of the parietal pleura done with mobilization of the esophagus into sufficient length. Myotomy done for about 10 cm. proximally and for no more than 2 cm. at the stomach side with chest tube insertion.

#### **Laparotomy**

Under general anaesthesia, supine position, upper midline incision done. The left lobe of liver mobilized and retracted to the right. The cardia displayed by dividing the peritoneum over the gastro-esophageal junction assisted by downward traction on the stomach. An anterior extramucosal myotomy done with an upward extent for about 5-6 cm and downward for about 1-2 cm onto the stomach with Dor fundoplication in one case and Toupet in the other.

#### **Laparoscopy**

Under general anaesthesia, supine lithotomy position and the surgeon standing between the patient's legs, a 10 mm cannula for the 30 degree laparoscope was inserted in the midline 5 cm above the umbilicus. After creation of pneumoperitoneum 2 10 mm trocars introduced 1 cm below the xiphoid and the other 2 cm below the left costal margin in the anterior axillary line. 2 other 10 mm ports inserted 1 cm below the right costal margin in the right mid-axillary line and 7 cm to the left of umbilicus. Elevation of left lobe of liver by retractor done then, the phreno-esophageal membrane was opened by a hook. The esophagus separated from the crura by blunt dissection, the vagus n. identified and preserved. Myotomy done on the anterior surface of the esophagus using harmonic scalpel about 5 cm upward and 1 cm downward onto the stomach. No fundoplication done.

#### **Dilatation**

. Perform a complete endoscopy in the left lateral position with special attention paid to the cardia to exclude a tumor and assess for the presence of a hiatal hernia, then a guidewire is introduced into the stomach. The endoscope is then removed outside the patient and a balloon is passed over the guide wire. Still in the left lateral position, a 50 -cc syringe attached to the balloon port to pass air so as to inflate the balloon to 30 cc, under fluoroscopic control.

The disappearance of the balloons waist is the accepted sign of successful dilatation. .then we remove the balloon and the guide wire , and the patient is allowed to take liquids only in the first day.

### Result:

There were 30 patients in our study ,14 males and 16 females .The range of age in the male group was 11-66 years ,the median was 30.14 years,while the range in females was between 17-50 years with amedian of 30.93 years.

**Table( 1 )Distribution of the study group according to sex**

| male | female | 0total |
|------|--------|--------|
| 14   | 16     | 30     |

**Table( 2) Distribution of study group according to age**

| sex    | Range of age | Median age |
|--------|--------------|------------|
| male   | 11-66 year   | 30.41 year |
| female | 17-50 year   | 30.93 year |

The most common symptom was dysphagia which occurred in all 30 patients .the second most common symptom was weight loss which occur in 19 patients, 8 males and 11 females, followed by regurgitation which occur in 17 patients,10 males and 7 females.

**Table( 3) Distribution of the study group according to symptoms**

| symptom         | male | female | Total no. |
|-----------------|------|--------|-----------|
| dysphagia       | all  | all    | 30        |
| regurgitation   | 10   | 7      | 17        |
| Epigastric pain | 2    | 1      | 3         |
| cough           | 2    | 2      | 4         |
| Weight loss     | 8    | 11     | 19        |
| Chest pain      | 3    | 1      | 4         |
| Chest infection | 2    | nil    | 2         |

The diagnosis of patients done by OGD and BA.Swallow in addition to clinical presentation. Both diagnostic procedures used to patients who had operation while only OGD ordered by the medical team

for those who had dilatation. The number of patients who underwent dilatation was 17 patients(56.66%),9 males and 8 females,and that who had operation was 13 patients(43.33%)5 males and 8 females.

**Table( 4) Distribution of the study group according to type of intervention**

| intervention | male | female | total |
|--------------|------|--------|-------|
| dilatation   | 9    | 8      | 17    |
| surgery      | 5    | 8      | 13    |
| total        | 14   | 16     | 30    |

Of the 17 patient who had dilatation 12 patient (70.58%) [ 7 males and 5 females] had one dilatation,2 patients (11.76%) ,[1 male and 1 female] had 2 dilatations, 2 female patients (11.76%)

had 3 dilatations and 1 male patient (5.88%) had 4 dilatations.The median time interval between dilatation sessions in patients with multiple dilatations was 3.7months.

**Table( 5) Distribution of the dilatation study group according to the no. of dilatations**

| No. of dilatation | male | female | total |
|-------------------|------|--------|-------|
| One dilatation    | 7    | 5      | 12    |
| Two dilatation    | 1    | 1      | 2     |
| Three dilatation  | nil  | 2      | 2     |
| Four dilatation   | 1    | nil    | 1     |
| Total no.         | 9    | 8      | 17    |

Among the 13 patients who underwent surgical intervention,10 patients (76.9%) had open trans-thoracic myotomy,[3 males and 7 females]. 2 patients (15.38%)

had open abdominal myotomy[1 male and 1 female],and finally 1 female patient (7.69%) had laparoscopic myotomy.

**Table(6) Distribution of the study group according to the type of operation**

| Type of operation | male | female | total |
|-------------------|------|--------|-------|
| Left thoracotomy  | 3    | 7      | 10    |
| laparotomy        | 2    | nil    | 2     |
| laparoscopy       | nil  | 1      | 1     |
| total             | 5    | 8      | 13    |

In the dilatation study group the procedure went smoothly without perforation in all patients. In the surgical group, 2 patients had esophageal mucosal perforation, 1 during open trans- thoracic myotomy ,

and 1 during laparoscopic myotomy. The patient with perforation during thoracotomy had previously 4 dilatations while that during laparoscopy with no previous dilatation.

**Table(7) showing the no. of patients who had perforation during various types of intervention**

| Type of intervention | male | female | total |
|----------------------|------|--------|-------|
| dilatation           | nil  | nil    | nil   |
| thoracotomy          | 1    | nil    | 1     |
| laparotomy           | nil  | nil    | nil   |
| laparoscopy          | nil  | 1      | 1     |
| total                | 1    | 1      | 2     |

Post operatively, 3 patients had simple chest infection 2 with thoracotomy ,and 1 with laparoscopy.1 patient had left pleural empyema[this patient had 4 previous dilatations followed by open thoracotomy].

**Table( 8) showing the Prevalence and type of post-operative complication**

| complication           | male | female | total |
|------------------------|------|--------|-------|
| empyema                | 1    | nil    | 1     |
| Simple chest infection | 1    | 1      | 2     |
| total                  | 2    | 1      | 3     |



Follow up of patients (range between 4 months-19 months) is conducted during clinic visits, and showed all patients to be benefited with regard to relieve of dysphagia. The effectiveness of intervention was evaluated following the criteria of Ellis et al [8]. In this criteria the results are divided into 4 groups :

- 1-Excellent: completely asymptomatic, gained weight, and return to daily activity.
- 2-Good: Gained weight, return to daily activity, but with occasional dysphagia
- 3-Fair: improved, but still complaining of some persistent dysphagia.
- 4-Poor: no improvement or the development of severe symptom other than dysphagia. In our study, with the surgical intervention group 12 patients (92.3%) had excellent results and 1 patient (7.6%) had poor result [the patient developed reflux], while at the dilatation group 12 patient (70.52%) had excellent results, 4 patients (23.54%) had good results and 1 patient (5.8%) with fair result

### Discussion

Esophageal achalasia remains a disorder of unknown etiology and with no definitive cure. It affects both sexes equally and the incidence peaks in the third and fourth decades of life<sup>(1,2,3)</sup>.

The same thing applies in our study for the age at presentation, with a mean age of 30.14 years for males and 30.93 years for females. Symptoms rather than signs are the hallmarks of achalasia. In our study dysphagia was the most common symptom followed by weight loss and regurgitation respectively which goes with other studies that showed a similar sequence of symptoms prevalence<sup>(3,6,7)</sup>. EL-Hassani et al showed that about 90% of patients with achalasia present with dysphagia, regurgitation and weight loss, and the dysphagia was for both solid and liquids at presentation. This contrast with patients having stricture or ring whose dysphagia is limited for solids<sup>(16)</sup>. Achalasia can be associated with other pathologies, such as esophageal diverticulum, hiatus hernia and peptic ulceration. In our study group there were no such associated pathologies.

Several therapeutic options have been adopted during the last century, such as pharmacological parasympathomimetic drugs, forceful dilatation and esophagomyotomy with variable patterns of safety and efficacy<sup>(10)</sup>.

Surgical myotomy of the distal oesophagus has been shown to yield the best results in relieving the symptoms of achalasia when compared to other non-surgical techniques<sup>(11,12,13)</sup>.

During the last decade, minimally invasive video-assisted techniques have enabled this procedure to be

carried out successfully from both a laparoscopic and thoracoscopic approach<sup>(14,15)</sup>.

However, there remains a paucity of data on the long-term results of Heller's myotomy via either of these minimally invasive approaches. Modified Heller myotomy, using either a trans-thoracic or a trans-abdominal approaches, has long been the surgical treatment of choice for achalasia. The criteria for successful treatment were based on subjective parameters by disappearance of symptoms and objective parameters by post-operative BA, swallow and OGD. Thoracic surgeons, and to a less extent general surgeons, carried out a left thoracotomy to perform a myotomy of the LES. If the caudal extent of the myotomy was carried on to the stomach for 2 to 3 cm, a partial fundoplication was performed<sup>(12)</sup>.

In our study no fundoplication was done to all the trans-thoracic Heller's myotomy, as the myotomy was extended distally to no more than 1 cm onto the stomach, this attitude goes with that of Streitz and colleagues<sup>(15)</sup>, who limited their myotomy to the esophago-gastric junction, relying on the residual attachments of the phreno-esophageal ligament and the undisturbed configuration of the diaphragmatic hiatus to prevent gastro-esophageal reflux. General surgeons usually use the abdominal approach for Heller's myotomy, the advantage of which is a more effective myotomy because the gastric portion of the myotomy can be performed quite easily with its concomitant anti-reflux procedure<sup>(10)</sup>.

The causes of early failure or persistent dysphagia in both approaches may not only related to incomplete myotomy, but there are other causes like: extremely poor esophageal body function (especially in long standing cases), peri-esophageal fibrosis and mechanical obstruction related to anti-reflux procedure. The causes of the late recurrence of dysphagia has been attributed to: bridging of muscle gap with scar tissue due to insufficient haemostasis, linear healing of the myotomy incision due to incomplete circumferential dissection of the incision and a stricture due to reflux esophagitis secondary to associated or a created hiatus hernia<sup>(7,16)</sup>.

In our study 13 patients underwent surgical myotomy of them, 10 had trans-thoracic myotomy without anti-reflux procedures, all of them had excellent results regarding relieving of dysphagia and none of them developed reflux. These results agreed with that of EL-Hassani et al and Godispoti et al who showed that trans-thoracic Heller's myotomy to be an effective mode of treatment<sup>(16,17)</sup>.

M. Codespoti and Andreollo et al found that GERD can occur after trans-thoracic Heller's myotomy regardless whether an anti-reflux procedure added or not, therefore they concluded that it is not

necessary to concentrate to add this procedure which might cause functional obstruction especially if the myotomy was not carried to long distally<sup>(17,32)</sup>.

2 patients in our study had trans- abdominal esophagealmyotomy with partial fundoplication both had excellent results with no dysphagia or reflux .Although their number is small and may be inappropriate to compare them to other studies, however O'Sullivan et al , Jekler J. et al showed that abdominal Heller smyotomy with partial fundoplication had excellent results regarding relieve of dysphagia<sup>(18,19,20)</sup>. The same thing is true for the only case in our study how had laparoscopic Hellersmyotomy . Abir et al and Douard and colleagues showed the effectiveness of this approach in treating achalasia,<sup>(21,22,23,24)</sup>.

In our study 17 patients had forceful balloon dilatation .Dilatation was the mainstay for non-surgical treatment of achalasia since 1674. However the benefits were for a short period of time in many patients .In our study 12 patients (70.58%) had excellent results ,while 4 patients (23.52%) required subsequent dilatation and 1 patient (5.88%) had no benefit at all from frequent dilatation. Our results are supported by that of Fellows JW. et al, Jacobs JB. Et al and several other studies which showed that 69.7% of patient had excellent results with single dilatation ,while 22.1% of patient required subsequent dilatations and 8% of patients had no benefit at all<sup>(24,25,26,27,28,29,30,31)</sup>.

In a study done at the GIT center in Baghdad, Abdullah F. et al found that excellent results were achieved by a single balloon dilatation in 69% of patients, and 19.4% had recurrent symptoms that required another sessions<sup>(33)</sup>.

However achalasia is a progressive disease with progressive functional deterioration of the esophageal body, Mineo et al found that in long standing cases with severe esophageal body functional deterioration and hugely dilated sigmoid shape esophagus Hellersmyotomy should be the first line of treatment as the response to dilatation in such delayed cases is most probably not encouraging and poor (33).In our study 2 patients(15.3%) had mucosal perforation during operation , one patient during trans-thoracic approach and the other during laparoscopic Hellers, both treated by immediate suturing , and oral intake delayed until the 7th post operative day ,this percentage of perforation in our study goes with that done by J . Black and colleagues who had a 16% mucosal perforation during myotomy(35) . post operatively 2 patients(15.3%) had simple chest infection and one patient (7.69%) had empyema which is similar to the results of J. Blak and Ermanno et al (35,36) . On follow up of the patients one patient (7.69%) had reflux symptoms treated successfully by PPI, in a

Study done by Ermano A. et al one patient out of 17 (5.8%) had reflux symptom and treated with PPI (36)

### Conclusion and recommendations:

- 1-There is no definitive cure for achalasia.
- 2-The aim of treatment is to relieve dysphagia and permit the patient to swallow effectively.
- 3-Both pneumatic dilatation and surgical myotomy can be used as primary treatment.
- 4-Surgical myotomy is associated with high rate of definitive and long term relieve of symptoms.
- 5-We recommend that More attention should be paid for minimally invasive surgery (laparoscopy, thoracoscopy) in the treatment of achalasia , as it was proven to be an effective and safe method of treatment.

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