

Achalasia Management in Gastroenterology and Hepatology Teaching Hospital

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Abstract

Background: Achalasia is a primary disorder of the esophagus, of unknown etiology, characterized by degeneration of the myenteric plexus, resulting in impaired relaxation of the gastroesophageal junction (GEJ), together with loss of peristalsis of the body of the esophagus. Laparoscopic Heller's Myotomy is the first-line surgical therapy for achalasia; it has a response rate of 90–97% with recurrent dysphagia in 3–10% of patients. Laparoscopic incision is made anteriorly from 6 cm above the GEJ to 3 cm beyond, preserving cardio-oesophageal fat and the anterior vagus nerve. **Objective:** To assess the management of achalasia. **Patients and method:** A retrospective and prospective cohort study that carried out in the Gastroenterology and Hepatology Teaching Hospital in Medical city Complex in the period from the first of January 2019 to the end of Jun 2022. A sample of 25 patients who were diagnosed with achalasia according to clinical presentation and proved by barium swallow. **Results:** The current study included 25 patients, female was presented in 13 (52%) and male was found in 12 (48%) with female to male ratio (1.08:1). Dysphagia was presented in all patients (100%), then weight loss in 19/25 (76%) of patients, then regurgitation was found in 18/25 (72%) of cases. Complications happened as emphysema in one case 1/25 (4%) and simple chest infection in 2/25 (8%) cases (1 for male and other 1 for female). **Conclusion:** More than half of the achalasia patients were treated with Pneumatic dilatation (56%) while (44%) were treated by surgery. laparotomy done for (8.0%), and laparoscopy was done in (36.0%) of cases.

Keywords: Achalasia, Laparoscopic Heller's Myotomy, Dysphagia.

Introduction

Achalasia is an esophageal motor disorder of unknown etiology, characterized by esophageal aperistalsis and insufficient lower esophageal sphincter relaxation in response to swallowing. This results from loss of inhibitory nitrinergic neurons in the esophageal myenteric plexus ⁽¹⁾.

Epidemiology

Achalasia has been regarded as an uncommon disorder with an annual incidence of approximately 1.6 cases per 100,000 individuals and prevalence of 10 cases per 100,000 individuals ⁽²⁾. Although epidemiologic data on achalasia are limited, its frequency appears to be rising, with one study suggesting that, from 2004 to 2014, the incidence and prevalence of achalasia in central Chicago were two- to threefold greater than estimates from earlier years would have predicted ⁽³⁾. Men and women are affected with equal frequency. The disease can occur at any age, but onset before adolescence is rare. Achalasia is usually diagnosed in patients between the ages of 25 and 60 years. Achalasia may occur in association with adrenal insufficiency and acromegaly in patients with triple A syndrome or Allgrove syndrome, a rare autosomal recessive genetic disorder ⁽⁴⁾.

Diagnosis of achalasia

The diagnosis of achalasia is typically suspected in a patient with dysphagia to both solids and liquids with associated regurgitation of undigested food. Complimentary tests include esophagogastroduodenoscopy and barium

esophagram; however, definitive diagnosis should only be made following evaluation of esophageal motor function with manometric testing ⁽⁵⁾. The following are the diagnostic modalities often employed in achalasia.

Barium esophagram

Esophageal dilation with a gradual taper down to the gastroesophageal junction giving a "bird's beak" appearance is the classic description of achalasia on barium esophagram. Additional findings include aperistalsis and poor emptying of barium. Signs suggestive of late- or end-stage achalasia include tortuosity, angulation, and megaesophagus ⁽⁵⁾. An additional role for contrast radiography is the assessment of esophageal emptying following treatment of achalasia. Symptom relief often does not parallel esophageal emptying. Therefore, objective assessment of treatment response can be helpful in identifying patients at risk of failed treatment. This is accomplished by timed barium esophagram where the barium column height is measured at 1 and 5 minutes after upright ingestion of a large barium bolus. Subsequent studies have revealed the usefulness of timed barium esophagram for identifying patients at risk of failed treatment ⁽⁶⁾.

Computed tomography can also be used to support an underlying diagnosis of achalasia and show the extent of disease, especially in those with sigmoid esophagus; however, it is less sensitive and not commonly employed ⁽⁶⁾.

Endoscopy

Most patients presenting with dysphagia require

esophagogastroduodenoscopy primarily to rule out a mechanical obstruction due to cancer that can mimic achalasia both clinically and nanometrically. This is termed "pseudoachalasia."^{30–32} Similar to achalasia, mechanical obstruction can monometrically result in impaired LES relaxation and aperistalsis or spastic contractions of the esophageal body.³³ Endoscopic findings of achalasia range from a grossly normal appearance to a tortuous, dilated sigmoid esophagus with retained food and saliva. Oftentimes, the gastroesophageal junction will have a puckered appearance. Intubation of the stomach may give mild resistance; however, a strong resistance should raise suspicion of pseudoachalasia^(5, 6). Of note, patients with achalasia can develop candidiasis secondary to esophageal stasis. Endoscopic evidence of candidiasis in the setting of immunocompetence suggests an esophageal motility disorder⁽⁷⁾.

Esophageal manometry

Esophageal manometry is the gold standard for the diagnosis of achalasia. It functions to assess esophageal pressures along the length of a flexible catheter inserted into the esophagus. The classic manometric finding of aperistalsis of the esophageal body and incomplete LES relaxation without evidence of mechanical obstruction is strongly supportive toward the diagnosis of achalasia⁽⁸⁾. Other findings that include increased resting LES pressure and simultaneous non-propagating contractions are also suggestive of the diagnosis, however, these are not required⁽⁹⁾. Manometry, as a diagnostic tool, has evolved significantly over the past decade from conventional catheters with pressure sensors spaced 3–5 cm apart utilizing solid-state technology or a water-perfused extrusion catheter to high resolution manometry where pressure sensors are placed 1 cm apart with either water-perfused extrusion or solid-state technologies. High resolution manometry can display pressure data as esophageal topography plots. Esophageal pressure topography gives the clinician the advantage of differentiating achalasia into three distinct subtypes that have important therapeutic outcome implications. Subtype II has the most favorable prognosis, whereas subtype III is the most difficult to treat. The prognosis of subtype I is somewhere in between⁽⁸⁻¹¹⁾.

Aim of the study

To assess the management of achalasia in the Gastroenterology and hepatology Teaching Hospital in Medical City Complex

Patients and Method

Study design and setting

A Retrospective and prospective cohort study that carried out in the Gastroenterology and Hepatology Teaching Hospital in Medical city Complex in the period from the first of March 2020 to the end of Feb 2022. A sample of 25 patients who were diagnosed with achalasia according to clinical presentation and proved by BA. Esophagram And OGD which showed the clinical pictures of achalasia, which are dilated

esophagus with abnormal or absent motility of esophageal body and birdsbeak appearance of the lower end on esophagram.

OGD showed tight cardia with dilated esophagus. 14 patients had dilatation and 11 patients had esophageal Hellers myotomy done through laparotomy and laparoscopic approach.

Laparoscopy

Under general anesthesia, supine lithotomy position and the surgeon standing between the patients legs a 10 mm cannula the 30-degree laparoscope was inserted in the midline 5 cm above the umbilicus. After creation of pneumoperitoneum two of 10 mm trocars introduced 1 cm below the xiphoid and the other 2 cm below the left costal margin in the antero-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-oesophageal membrane was opened by hook. These esophageal separated from the cura by blunt dissection, the vagus nerve identified and preserved, esophagus using harmonic scalpel about 5 cm upward and 1 cm downward onto the stomach. No fundoplication done.

Laparotomy

Under general anesthesia, with patients on supine position the upper Medline incision done the left lobe of liver mobilized and retracted to the right, the cardia displayed by dividing the peritoneum over the gastroesophageal junction assessed by downward traction on the stomach. An anterior extra mucosal myotomy done with an upward extension 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.

Dilatation

Perform 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 guide wire is introduced into the stomach. The endoscope 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.

Ethical consideration

The study was approved by each of the local thesis committee Iraqi ministry of health, council of Iraqi board of health specialization.

Statistical analysis

After the data were entered in a table developed by the researchers, the analysis was done by using the SPSS program, version 23 and for qualitative variables, we used frequencies and percentages, and for the quantitative variables, we used measures of central tendency and dispersion (standard deviation). For the inferential statistics, the tests were used of chi-square test (with a significance of $P \leq 0.05$).

Results

presented in 13 (52%) and male was found in 12 (48%) with female to male ratio (1.08:1) (table 1)

The current study included 25 patients, female was

Male		Female		Total	
No.	%	No.	%	No.	%
12	48.0	13	52.0	25	100.0

As shown in table 2, the range of age in male was between (12-65) years with mean age (30.7±7.4) years, while for female the range between (16-52) years and mean age was (31.4±6.2) years.

Gender	Range of age (years)	Mean± SD age (years)
Male	12-65	30.7±7.4
Female	16-52	31.4±6.2

Table 3 show, that dysphagia was presented in all

Symptom*	Male		Female		Total	
	No.	%	No.	%	No.	%
Dysphagia	12	48.0	13	52.0	25	100.0
Weight loss	8	42.1	11	57.9	19	100.0
Regurgitation	10	55.6	8	43.4	18	100.0
Cough	2	50.0	2	50.0	4	100.0
Chest pain	3	75.0	1	25.0	4	100.0
Epigastric pain	2	66.7	1	33.3	3	100.0
Chest infection	2	100.0	0	-	2	100.0

* More than one symptom may be found in each patient

As shown in table 4, dilatation was done for 14/25 cases: 7 in male and 7 in female, surgery was done for 11/25

patients: 5 (41.7%) for male and 6 (46.2%) for female.

Intervention	Male (n=12)		Female (n=13)	
	No.	%	No.	%
Dilatation	7	58.3	7	53.8
Surgery	5	41.7	6	46.2
Total	12	100.0	13	100.0

Regarding to the number of dilatations, one dilatation done for 9/14 patients (4 (57.1%) for male and 5 (71.4%) for female, two dilatations done for patients (1 (14.3%) for male and other one (14.3%) for

female, three dilatations was done for 2 patients (1 (14.3%) for male and other one (14.3%) for female, while four dilatations was done for 1 patient (male) (14.3%), as shown in table 5

No. of dilatation	Male (n=7)		Female (n=7)	
	No.	%	No.	%
One dilatation	4	57.1	5	71.4
Two dilatations	1	14.3	1	14.3
Three dilatations	1	14.3	1	14.3
Four dilatations	1	14.3	Nil	-
Total	7	100.0	7	100.0

Table 6 show, laparotomy done for 2/25 (8.0%) cases (male only), and laparoscopy was done in 9/25 cases (3 male (60.0%) and 6 (100.0) was female only.

Type of operation	Male		Female	
	No.	%	No.	%
Laparotomy	2	40.0	Nil	-
Laparoscopy	3	60.0	6	100.0
Total	5	100.0	6	100.0

Complications happened as emphysema in one case

1/25 and simple chest infection in 2/25 cases (1 for male and other 1 for female) (table 7).

Complications	Male		Female	
	No.	%	No.	%
Emphysema	1	50.0	Nil	-
Simple chest infection	1	50.0	1	100.0
Total	2	100.0	1	100.0

Discussion

In the current study there is gender difference in

which female was presented more than male with female to male ratio (1.08:1), in agreement with Khoudari G et al, study that revealed patients with achalasia were more likely to be females⁽¹²⁾. Kamal A et al., revealed that the majority of cases were women (72.3%) and men were (27.7%) from 155 patients enrolled in their study⁽¹³⁾. Also, it is in agreement with Li KF study, that found the female were more than male with female-to-male ratio was (1.3:1)⁽¹⁴⁾.

But it is not in agreement with et al., study that found achalasia was more common in male than female⁽¹⁵⁾. This may be attributed to the difference in sample size collection.

While achalasia can occur at any age, it is generally a disease of older age and rarely occurs before the 2nd decade of life. The mean of age in male was (30.7±7.4) years, while for female the mean age was (31.4±6.2) years which is in agreement with a previous study carried by Sadowski DC et al, that found the mean age at diagnosis was 53.1 years with no difference between male and female⁽¹⁶⁾. Symptoms are the hallmarks of achalasia; In the current study, dysphagia was presented in all patients (male and female), then weight loss, then regurgitation, this is similar to that found by Volcy J et al⁽¹⁷⁾.

Moreover, it is concordance with a previous Iraqi studies carried by Laith Alhadad,⁽¹⁸⁾ that found all patients with achalasia have dysphagia, then weight loss and regurgitation, also it is in agreement with El Hassani et al,⁽¹⁹⁾ showed that about 90% of patients with achalasia suffered from dysphagia, regurgitation then weight loss and dysphagia was for both solid and liquids at presentation. This contrast with patients having stricture or ring whose dysphagia was limited for solids. The treatment of esophageal achalasia is palliative, and it focuses on decreasing the outflow resistance of the GEJ caused by the dysfunctional LES. The evolution of achalasia treatment clearly shows that a fundoplication is required to prevent postoperative GERD⁽²⁰⁾.

In the present study, no fundoplication was done to all the trans-abdominal myotomy as the myotomy was extended distally to no more than 1 cm onto the stomach, this attitude goes with that of Streitz et al, who limited their myotomy to the esophago-gastric junction (EGJ), relying on the residual attachments of the phreno-esophageal ligament and the undisturbed configuration of diaphragmatic hiatus to prevent gastro-esophageal reflex⁽²¹⁾.

In the current study more than half of the achalasia patients were treated with dilatation (56%) while (44%) were treated by surgery. However, about one fourth of patients treated with Pneumatic dilation (PD) needed an additional dilatation. It is in fact known that patients treated with Pneumatic dilation finally need additional dilatations over time to control the achalasia symptoms. Which is not agreed with the current study as the dilatation were needed in 56% of patients.

In a large Korean study carried by Kim E et al., and throughout the period of the study, and from 975 patients that enrolled in their study, 719 patients were treated by balloon dilation therapy, while 17 patients were treated by esophago-cardio-myotomy, 256 of them received more than 2 treatments of balloon dilation therapy during the 5 yrs. period, indicating that 35% of patients treated with balloon dilation had a symptom recurrence within 5 yr. The most severe complication of balloon dilation is perforation of the esophagus. In recent studies, the perforation rate associated with balloon dilation is reported to be 1%-3%⁽²²⁾.

In the current study 11/25 (44%) patients underwent surgical myotomy of them, 8 had trans thoracic myotomy without anti reflex procedures, and all of them had excellent results regarding relieving of dysphagia and none of them develop reflux. which is same that found by an El-Hassani et al.,⁽¹⁸⁾ and Godispoti et al,⁽²³⁾ who showed that trans thoracic Hellers myotomy to be an effective mode of treatment

In the present study complications happened as empyema in one case 1/25 (4%) and simple chest infection in 2/25 (8%) cases. Kim E et al., revealed 6 cases of esophageal perforation were identified, yielding an estimated perforation rate of approximately 0.6%⁽²²⁾.

The main strength of the current study is the careful collection of the patient's sample. All patients with known esophageal motility disorders or prior gastro-esophageal surgery were excluded from the study, which may affect the results.

Conclusion

More than half of the achalasia patients were treated with Pneumatic dilatation (56%) while (44%) were treated by surgery. laparotomy done for (8.0%), and laparoscopy was done in (36.0%) of cases.

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References

- Palmieri O, Mazza T, Merla A, Fusilli C, Cuttitta A, Martino Get al. Gene expression of muscular and neuronal pathways is cooperatively dysregulated in patients with idiopathic achalasia. *Scientific reports*. 2016 Aug 11;6(1):1- 10.
- Sadowski DC, Ackah F, Jiang B, Svenson LW. Achalasia: incidence, prevalence and survival. A population-based study. *Neurogastroenterol Motil* 2010; 22:e256.
- Samo S, Carlson DA, Gregory DL, et al. Incidence and Prevalence of Achalasia in Central Chicago, 2004-2014, Since the Widespread Use of High-Resolution Manometry. *Clin Gastroenterol Hepatol* 2017; 15:366.
- Spechler SJ, Talley NJ, Travis AC. Achalasia: Pathogenesis, clinical manifestations, and diagnosis. *UpToDate*. 2022. Available at: <https://www.uptodate.com/consultation?search=achalasia>

uptodate.com/contents/achalasia-pathogenesis-clinical-manifestations-and-diagnosis. Accessed on 21/2/2022.

Vaezi MF, Pandolfino JE, Vela MF. ACG clinical guideline: diagnosis and management of achalasia. *Am J Gastroenterol*. 2013;108(8):1238–1249. quiz 1250

Krill JT, Naik RD, Vaezi MF. Clinical management of achalasia: current state of the art. *Clinical and Experimental Gastroenterology*. 2016;9:71.

Pandolfino JE, Gawron AJ. Achalasia: a systematic review. *JAMA*. 2015;313(18):1841–1852.

Pandolfino JE, Kahrilas PJ. American gastroenterological A. AGA technical review on the clinical use of esophageal manometry. *Gastroenterology*. 2005;128(1):209–224.

Spechler SJ, Castell DO. Classification of oesophageal motility abnormalities. *Gut*. 2001;49(1):145–151.

Salvador R, Costantini M, Zaninotto G, et al. The preoperative manometric pattern predicts the outcome of surgical treatment for esophageal achalasia. *J Gastrointest Surg*. 2010;14(11):1635–1645. [PubMed] [Google Scholar]

Pratap N, Reddy DN. Can achalasia subtyping by high-resolution manometry predict the therapeutic outcome of pneumatic balloon dilatation?: author's reply. *J Neurogastroenterol Motil*. 2011;17(2):205.

Khoudari G, Abou Saleh M, Sarmini MT, Parikh MP, Vega KJ, Sanaka MR.

463 The Prevalence and Epidemiology of Achalasia in the USA: A Population-Based Study. *Official journal of the American College of Gastroenterology | ACG*. 2019 Oct 1;114:S270-1.

Kamal A, Shakya S, Lopez R, Thota PN. Gender, medication use and other factors associated with esophageal motility disorders in non-obstructive dysphagia. *Gastroenterology Report*. 2018 Aug;6(3):177-83.

Li KF. Ten-year review of epidemiology, clinical features, and treatment outcome of achalasia in a regional hospital in... *Hong Kong Med J*. 2010 Oct;16(5):362-6.

Brintha K, Enestvedt, J. Lucas Williams, Amnon Sonnenberg. Epidemiology and Practice Patterns of Achalasia in a Large Multi-Center Database. *Aliment Pharmacol Ther*. 2011 Jun; 33(11): 10.1

Sadowski DC, Ackah F, Jiang B, Svenson LW. Achalasia: incidence, prevalence and survival. A population-based study. *Neurogastroenterol Motil*. 2010;22:e256–61

Volcy J, Ajose T, Ivonye C. Gradually Progressive Dysphagia, Reflux, Weight Loss, and Chest Discomfort. *American Family Physician*. 2019 Jul 15;100(2):113-4.

1. Laith Alhadad, Abdulruzak Alhilo. Management of achalasia in the gastroenterology and hepatology teaching hospital. *Iraqi journal of Gastroenterology*. 2006; 1 (6): 25-32.

2. El-Hassani NB, Mustafa W: achalasia of the cardia. *Iraqi postgraduate medical journal*. 2004;

1:15-17

3. Nurczyk K, Patti MG. Surgical management of achalasia. *Annals of gastroenterological surgery*. 2020 Jul;4(4):343-51.

4. Streitz Jr JM, Ellis Jr FH, Williamson WA, Glick ME, Aas JA, Tilden RL. Objective assessment of gastroesophageal reflux after short esophagomyotomy for achalasia with the use of manometry and pH monitoring. *The Journal of Thoracic and Cardiovascular Surgery*. 1996 Jan 1;111(1):107-13.

5. Kim E, Lee H, Jung HK, Lee KJ. Achalasia in Korea: an epidemiologic study using a national healthcare database. *Journal of Korean Medical Science*. 2014 Apr 1;29(4):576-80.

6. Godispoti M, Soon SY. Clinical results of thoracoscopic Hellers myotomy in the treatment of achalasia. *Eur J cardiothorac Surg* 2003; 24:620-24.