

Achalasia: Heller's Myotomy, Surgical Procedure and Outcome

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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, loss of weight and other symptoms.

Objective: The objective of this study is to evaluate the outcome of surgical and laparoscopic management of achalasia in the gastroenterology and hepatology teaching hospital.

Patients and method: The study is a prospective, observational cohort study extending from November 2016 and for two years. All patients underwent a comprehensive preoperative assessment, which included of a thorough review of their medical history, a detailed physical examination, analysis of blood tests, an electrocardiogram, a chest X-ray, a barium swallow study, and an upper gastrointestinal endoscopy with biopsies, if deemed necessary. Standard esophageal manometry was conducted in the majority of individuals. A computed tomography (CT) scan of the chest and abdomen was conducted on a specific group of older individuals in order to rule out the presence of cancer.

In laparoscopic Heller's myotomy, 5 to 7 cm anterior myotomy extending for at least 2-3 cm distally into the stomach to gastric sling fibers is performed. By flooding the operative field with adequate amount of normal saline, and asking the anesthetist to push air slowly through nasogastric tube, sufficient myotomy, perforation of the esophagus were roughly assessed. Clinical evaluation and Eckardt score was performed at 1, 3, 6, and 12 months after operation, and then annually.

Results: thirty eight patients were enrolled of them 20 were male and 18 female, 1.3:1 ratio with median age of 31.5 and 30.93 of male and female respectively. Thirty patients managed by laparoscopic Heller's operation while 8 patients have been treated with open approach. The perforation risk was 10% in L.H.M. and 12.5% in open Heller operations. The median of hospital stay was 2.5 in L.H.M and 4.5 day in open operations. Postoperative recurrence of symptoms (Eckardt stage 3) was 2 patients (6.6%) from 30 patients treated by L.H.M

Conclusion: sustained symptoms relief have been obtained with the surgical Heller's operation. Laparoscopic Heller's operation has steadily positioned as the surgical approach of choice given its lower hospital stay, less analgesia use and early return to normal every day activities in comparison to open.

Keywords: Achalasia, Eckardt score, Heller operation

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1. INTRODUCTION

Idiopathic achalasia is a very uncommon primary esophageal motor illness that is distinguished by the absence of peristalsis and a malfunctioning relaxation of the lower esophageal sphincter (LES). This condition leads to compromised movement of food boluses and the accumulation of food in the oesophagus (1). Achalasia exhibits an identical occurrence rate in both males and females, with an incidence of 1 in 100,000 persons and a prevalence of 10 in 100,000. The highest occurrence rate is observed within the age range of 30 to 60 years (2,3). The predominant manifestations of achalasia encompass dysphagia for both solid and liquid substances, respiratory difficulties such as nocturnal cough and aspiration, chest pain, heartburn, and weight loss (4). Heartburn has the potential to imitate the symptoms of gastroesophageal reflux disease (GERD). Treatment is typically effective in managing dysphagia and regurgitation; however, the management of chest discomfort poses greater challenges (5). The Eckardt symptom score is commonly employed as the primary grading method for assessing symptoms, phases, and treatment effectiveness. A symptom score ranging from 0 to 1 is indicative of clinical stage 0, while a score ranging from 2 to 3 corresponds to stage 1. A score ranging from 4 to 6 is associated with stage 2, and a value over 6 is indicative of stage 3. Stages 0 and 1 are indicative of the state of remission of the disease. However, it should be noted that stages 2 and 3 are indicative of therapy failure (Table 1) (6,7). The criteria for failure of Heller myotomy were classified into two categories: primary failure, which was characterised by the inability of the Eckardt score to decline to 3 or below, and recurrence, which was identified by a decrease in the Eckardt score to 3 or below followed by an increase to a value greater than 3 within a one-year period of observation (Table 2). Achalasia is the most prevalent main disease of esophageal motility. The term "achalasia" originates from the Greek language and denotes the inability to relax. The origins and mechanisms behind the development of the disease are currently not understood. The most frequently observed neuro-anatomical alteration is a reduction or degeneration of the myenteric ganglion cells, accompanied by neural fibrosis and varying levels of chronic inflammation within the myenteric plexus. Recent research has revealed the targeted elimination of inhibitory neurons that are non-cholinergic and non-adrenergic in nature. The

impacted transmitters include nitric oxide and vasoactive intestinal peptide. This illness is characterised by its irreversible nature. The prevailing theories about the causes of idiopathic achalasia primarily involve auto-immune and viral aspects. It has been observed that auto-antibodies targeting neurons are detected in around 50% of patients with this condition (8-12). The absence of propulsive peristaltic contractions and impaired relaxation of the lower esophageal sphincter (LES) lead to the accumulation of food in the oesophagus. Over time, this causes the oesophagus to expand and elongate, ultimately assuming a sigmoidal configuration in more severe instances. Esophagitis with mucosal ulceration is a common occurrence in the mucosa of the oesophagus. This is mostly attributed to the occurrence of stasis, as well as the multiplication of bacteria and subsequent fermentation resulting from the retention of food debris (9,10). Autopsy investigations have demonstrated that individuals diagnosed with achalasia exhibit a greater thickness of the esophageal muscle in comparison to those without the condition. Furthermore, the utilisation of high frequency intra-luminal ultrasound imaging has revealed that the thickness of the lower esophageal sphincter (LES) and esophageal muscle, as well as the cross-sectional area of the esophageal muscle, are notably higher in individuals with achalasia when compared to individuals with other esophageal motility disorders (11). The initial diagnostic procedure involves the exclusion of a benign or malignant obstruction through the utilisation of endoscopic or radiological techniques. During the initial phases, it is possible for both tests to exhibit entirely normal results. In instances with advanced pathology, the oesophagus may exhibit dilation due to the accumulation of food and saliva. Endoscopic examination will reveal a rosette-like appearance at the gastroesophageal junction, often accompanied by heightened resistance during the transit of the endoscope into the stomach (5). If excess pressure is required to pass esophagogastric junction by endoscopy, pseudo-achalasia should be highly suspected (13). The barium examinations reveal the presence of a "bird beak" look resulting from the non-relaxation of the lower esophageal sphincter (LES). Additionally, there is observed variation in the degree of esophageal dilatation, ranging up to sigmoid oesophagus. Other notable findings include aperistalsis, occasional presence of an air-fluid level, and the lack of the gastric air bubble (5). The diagnostic method of choice for achalasia remains manometry, which is widely regarded as the most reliable and accurate technique (14). An elevation in the

resting tone of the lower esophageal sphincter (LES) is frequently reported (4). A potential association has been observed between the extent of esophageal contraction and tortuosity observed on X-Ray imaging, and the length and severity of symptoms related to obstructive oesophagus (15). In recent times, the utilisation of high-resolution manometry (HRM) has emerged as the most accepted and preferred method for diagnosing achalasia. This preference stems from the objective and precise nature of HRM in detecting compromised relaxation of the esophago-gastric junction and evaluating peristaltic contraction, as evidenced by several studies (4,16). The utilization of human resource management (HRM) has resulted in the categorization of achalasia (according to the Chicago classification) into three clinically significant groups, which are determined by the contractility pattern observed in the esophageal body (17). The therapeutic options now utilised for achalasia encompass pneumatic dilatation, surgical myotomy, and endoscopic injection of Botulinum toxin. The treatment of long-acting nitrates and calcium channel blockers has been associated with potential advantages, as shown by an initial response rate ranging from 50% to 70% (8-11). Surgical intervention has been found to yield the most favourable long-term outcomes, particularly among individuals in younger age groups (18). There has been a growing interest in the exploration of a novel technique that combines an endoscopic approach with the concepts of natural orifice trans luminal endoscopic surgery (NOTES) for the purpose of conducting a myotomy. The technique is commonly referred to as POEM. In 1914, Heller invented the technique of esophageal myotomy, which involved dividing the muscle fibres of the distal oesophagus by an abdominal approach using both anterior and posterior incisions (known as double myotomy). This technique was subsequently improved by Groeneveldt and Zaajier, who developed a single anterior myotomy approach (references 15 and 18). The initial laparoscopic myotomy procedure was performed by Shimi et al in 1991, and it has since emerged as the favored approach because to its reduced morbidity and expedited recovery (19-21). Studies comparing open versus laparoscopic surgical access have yielded inconclusive results regarding the prevalence of dysphagia, since no significant changes have been observed between the two approaches. However, it has been demonstrated that patients undergoing laparoscopic surgery experience shorter hospital stays and recovery times compared to those receiving open surgery (22).

The occurrence of symptomatic relapse following treatment of achalasia is observed in 32% of individuals who undergo balloon dilation and in 10% of patients who undergo Heller myotomy within the initial year (23,24). The recurrence rate in young individuals may go up to 72% over a period of 10 years (25). There is currently a lack of consensus over the optimal length of myotomy, both in its proximal and distal extent. Additionally, there is a lack of agreement on whether an anti-reflux technique should be included, and if so, which specific procedure should be performed. These matters have generated significant controversy within the medical community. The primary factor contributing to suboptimal delayed outcomes was the inability to fully alleviate the blockage (15). There exist three discernible indicators that can be employed to ascertain the completion of the myotomy procedure:

1. Visual inspection of the esophagus and cardia while the esophageal lumen is dilated with a large dilator.
2. The use of intra-operative manometry which may be unreliable (13,26).
3. The use of intra-operative endoscope to determine whether there is residual constriction of the channel between the esophagus and the stomach (26- 28).

Table 1. Eckardt score clinical scoring for achalasia

Score	Dysphagia	Regurgitation	Retrosternal pain	Weight loss(Kg)
0	None	None	None	None
1	Occasional	Occasional	Occasional	<5
2	Daily	Daily	Daily	5-10
3	Each meal	Each meal	Each meal	>10

Table 2. clinical staging of achalasia

Stage	Eckardt score	Clinical implication
0	0-1	Remission
1	2-3	Remission
2	4-6	Treatment failure
3	>6	Treatment failure

2. METHODOLOGY

The study is a prospective, observational cohort study. The hospital registries were reviewed to identify all patients who were surgically treated for achalasia between November 2016 and 2018. Patients were eligible for this study if they have a minimum 1 year follow up and had pre-operative Eckardt score, barium study, and oesophagogastroscopy (OGD). Thirty eight patients included in this study as they fulfill above mentioned criteria. All patients underwent a comprehensive preoperative assessment, which encompassed a thorough review of their medical history, a comprehensive physical examination, analysis of blood tests, an electrocardiogram, a chest X-Ray, a barium swallow study, and an upper gastrointestinal endoscopy with biopsies, if deemed necessary. Standard esophageal manometry was conducted in the majority of individuals. A computed tomography scan of the chest and abdomen was conducted on a specific group of older individuals in order to rule out the presence of malignancies. The preoperative assessment of the Eckardt symptom score is contingent upon the frequency of esophageal symptoms, including dysphagia, regurgitation, chest pain, as well as the extent of weight loss. Hence, individuals who shown no symptoms whatsoever were assigned a score of 0, whilst those who exhibited the most severe symptoms were assigned a score of 12. Based on the obtained scores, the subsequent clinical stages were delineated:

1. stage 1 (score 2-3)
2. stage 2(score 4-6)
3. Stage 3 (score<6). Of the study group, 30 patients underwent laparoscopic Heller myotomy, one transthoracic and seven by laparotomy.

The laparoscopic Heller's myotomy:

The surgical procedure was conducted with the patient under general anaesthesia in the supine lithotomy position, while the surgeon positioned themselves between the patient's legs and utilised a 5-port access. The establishment of the pneumoperitoneum occurs at a pressure of 14 mmHg. Following the incision of the peritoneal reflection, a focused anterior dissection is conducted to reveal the crura, the left vagus nerve, and the gastroesophageal junction. A surgical procedure is conducted with an anterior myotomy measuring between 5 and 7 centimetres. This myotomy extends at least 2 to 3 centimetres distally into the

stomach, specifically targeting the gastric sling fibres. By flooding the operative field with adequate amount of normal saline, and asking the anesthetist to push air slowly through nasogastric tube, sufficient myotomy, perforation of the esophagus were roughly assessed. Dor fundoplication securing the anterior fundic wall to both crura and to the edges of the myotomy site were performed in some patients.

Thoracotomy Heller myotomy:

The patient was placed under general anaesthesia and positioned in the right lateral position. An incision was made via the 7th intercostal gap. By performing a division of the parietal pleura, a satisfactory length of the lower oesophagus is acquired. A myotomy procedure was performed, extending approximately 10 centimeters in a proximal direction and no more than 2 centimeters in a distal direction towards the stomach. Additionally, a chest tube was introduced.

Laparotomy:

Under general anesthesia supine position, upper midline incision done. The left liver lobe was mobilised and then retracted towards the right side. The cardia is visualised through the division of the peritoneum covering the gastroesophageal junction, aided by applying downward traction on the stomach. An anterior extra mucosal myotomy was performed, with an upward extension spanning approximately 5-6 cm and a downward extension spanning approximately 1-2 cm onto the stomach.

A soft diet on postoperative day one was allowed for most of the patients. Clinical evaluation and Eckardt score was performed at 1, 3, 6, and 12 months after operation, and then annually. Upper OGD endoscopy were performed as needed postoperatively

A thirty eight patients were diagnosed to have achalasia based on clinical, radiological, and endoscopic criteria, during the period from November 2016 to December 2018. Thirty patients had Heller myotomy through laparoscopy and seven patients had open myotomy trans-abdominal and one patients underwent thoracotomy. The follow up of patients was done by clinical interview visits and enquiry about dysphagia symptoms in details, Eckardt score and any symptoms developed after the intervention. The follow up period ranged from 4-19 months. The criteria for classifying a failure of Heller myotomy included two scenarios: main failure, which was characterised by the inability of the Eckardt score to decline to 3 or

below, and recurrence, which involved a decrease in the Eckardt score to 3 or below followed by an increase to a value greater than 3, both observed within a one-year follow-up period.

Table 3. Clinical staging of achalasia

Stage	Eckardt score	Clinical implication
0	0-1	Remission
1	2-3	Remission
2	4-6	Treatment failure
3	>6	Treatment failure

3. RESULTS

A total of 38 achalasia patients were consecutively enrolled in the database. There were 20 male and 18 females the ratio was 1.3:1. The range of ages in male group was 12-75 years, the median was 31.5 years, while the range in females was between 15 to 60 years with median of 30.93 years. Pre-operative Eckardt score class distribution according sex and age (**Table 4**). The diagnosis of patients done by OGD and Barium swallow in addition to clinical presentation. Both diagnostic procedure used for patients who had operation. Manometry study done in 27 patients and as shown in (**Table 5**). Among the 38 patient who underwent surgical myotomy, 30 patients (79%) had laparoscopic approach, 16 male ,51% and 14 female 49%, 7 patients had laparotomy, 3 male 42% and 4 female 58%, and 1 patient transthoracic approach. Duration of hospital stay range was 4-7 days in 7 patients treated with laparotomy, 4 days in one patient with thoracotomy and 2-4 days for 30 patients treated laparoscopic Heller myotomy. Post-operatively, 4patients had simple chest infection, one with thoracotomy and one with L.H.M. and two patients with laparotomy.

Table 4. Distribution of study group according to gender and age.

Gender	No.	%	Range of age (years)	Median age (years)
Male	20	52.6	12-75	31.5
Female	18	47.4	15-60	30.9

Table 5. Pre-operative positive OGD distribution

Gender	Eckardt class	No.
Male	3	20
Female	3	18
Total		38

Table 6. Demographic and pre-operative clinical characteristics

Variable	No.	%	
Gender	Male	20	52.6
	Female	18	47.4
Age mean (years)	31		
Eckardt stage 3	38	100	
Positive OGD.	Esophagitis	5	13.2
	Hiatus hernia	2	5.2
Manometry	27	70	
Barium swallow	38	100	
Co-morbidity	C.V.S	5	13.5
	Metabolic	2	5.2
	Hematology	1	2.6
Pre-operative dilation	1	2.6	

Table 7. Surgical approach

Type of operation	Male	Female	Total
L.H.M	16	14	30
Laparotomy	3	4	7
Transthoracic	1	0	1
Total	20	18	38

Table 8. Surgical approach and perforation risk

Variable	No.	%
L.H.M.	30	79.0
L.H.M. Perforation	3	10.0
Open H.M.	8	21.0
Open H.M. Perforation	1	12.5

Table 9. Duration of hospital stay, in days, and surgical approach.

Variable	Range	mean
L.H.M.	1-4	2.5
Open H.M	2-7	4.5

Table 10. Post-operative recurrence of symptoms (Echardt stage 3) and surgical approach.

Intervention	NO. of cases	Echardt stage 3		Duration of follow up (month)
		No.	%	
L.H.M	30	2	6.6	6-18
Open H.M.	8	0	0.0	6-18
Total	38	2	5.2	

4. DISCUSSION

Achalasia is a primary esophageal motor disorder with a variety of causes. Fei L, Rossetti G, Moccia F et al., Vaezi M F, Pandolfino J E, Vela M F., Moonen A J, Boeckstaens G E and others found that achlasia has an incidence of 0.03-1.63/100.000 population and a prevalence of approximately 0.01% with no difference between genders(29,30,31,32,33). The incidence peaks in the third and fourth decades of life as stated by Mcfadden DW, Michael J.,Zinner MJ. and Marelo FV.(8,9,10), and mean age was 38.4-+ 11.6 years(2,3). This result same applied in our study for age at presentation, with a mean age of 31.5 for male and 30.6 for female with no sex difference. Emanuel Asti et al, by a retrospective, observational cohort study and data analysis observe that achalasia can be associated with other pathologies such as esophagitis, esophageal diverticulum, hiatus hernia and peptic

ulceration of the esophagus(34). In our study group there was such an association, 7 patients (18.4%) had positive findings as detected by OGD, 5, (13.15%) with esophagitis and 2, (5.26%) patients with hiatus hernia. Single-center experience and systematic review of the literature done by Lynch KL, Pandolfi no JE, Howden CW, Kahrilas PJ, the most common complication of Heller myotomy is perforation of the esophageal or gastric mucosa (average 6.3%) during the myotomy, usually repaired without clinical consequences(35). The perforation occurred in 4 out of 38 patients who underwent Heller myotomy, the average was 10.52%. The perforation rate in L.H.M. was 3 out of 30 patients, 10%, whom underwent L.H.M. and they recognized intra-operatively and immediately repaired and this result goes with Borges A A, Lemme EM, Abrahao L J et a(36), Hamdy E, El Nakeeb A, El Hanfy E et al(44), Comparative study and a prospective randomized study, Moonen A, Annese V, BelmansA et a(37)I. a multicentre randomised controlled trial. One intraoperative mucosal tear occurred in open approach, laparotomy, and had been repaired 14.28%. Surgical myotomy of distal esophagus has been shown to yield the best results in relieving the symptoms of achalasia when compared to other non-surgical approach as concluded by Csedes A, Braghetto I, Henriques A, late result prospective study(27) , Malthaner RA, Todd TR, Miller I, Griffith Pearson F et al(,28), and Ellis Jr FH (38). However, there remain a paucity of data on long-term results of Heller,s myotomy via either of trans-thoracic or trans-abdominal minimal invasive technique. The assessment of therapy efficacy for achalasia has traditionally relied on a combination of subjective and objective criteria. Subjective parameters involve evaluating the elimination of symptoms reported by the patient, while objective measures involve the use of post-operative Barium-swallow tests and OGD(28). Thirty six patients out of 38, (94.73%), had symptom improvement over eighteen months follow-up period as assessed clinically by Eckardt score stage. Two patients, (5.26%), developed Eckardt stage 2 in 6-18 months follow-up both of them were after L.H.M. then they submitted to pneumatic dilation to improve their symptoms. This result is comparative to study of V.A. Williams and J. H. Peters(39) as they report that approximately 5% of patients will need another intervention and result of endoscopic dilation or reoperation as both safe and effective options also reported by Yokohama group.

According to V.A., incomplete myotomy on the stomach side, which involves a more complex dissection, as well as myotomy fibrosis or a combination of both, are the primary factors contributing to post-surgical failure. Williams and J. H. Peters(39) Richter, J.E. and Boeckstaens, G.E. The treatment options for managing achalasia mostly consist of surgical intervention or pneumatic dilation (40). The authors of the study are Zaninotto G, Costantini M, Portale G, et al. This paper aims to discuss the aetiology, diagnosis, and treatment options for failures following laparoscopic Heller myotomy in patients with achalasia. The management of symptom recurrence following Heller myotomy has demonstrated favourable outcomes by the utilisation of pneumatic dilatation. In instances when pneumatic dilation proves ineffective, a novel laparoscopic approach has been employed as an alternative therapeutic intervention. Heller myotomy (41). The causes of early failure or persistent of dysphagia in both approach (open and minimally invasive) may not only related to incomplete myotomy, However, there are additional factors that might contribute to this condition, such as severely impaired esophageal body function, particularly in cases that have persisted for a long duration. Additionally, peri-esophageal fibrosis and mechanical obstruction resulting from anti-reflux procedures can also be underlying causes. Late dysphagia is believed to be caused by several factors. One factor is the formation of scar tissue that bridges the muscle gap, which occurs due to inadequate hemostasis. Another factor is the linear healing of the myotomy incision, which happens when the incision is not fully dissected circumferentially. Additionally, a stricture can develop as a result of reflux esophagitis, which is secondary to a preexisting or surgically created hiatus hernia (15,42).

Prior research on cardiomyotomy revealed minimal clinical reflux in the absence of fundoplication, however aberrant pH tests were observed in 40% of the patients (43). Four scholarly studies have documented the amelioration of symptoms two years post-treatment. These articles include the works of Bogres et al. (31), Hamdy et al. (44), Moonen et al. (32), as well as Persson J, Johnsson E, Kostic S, Lundell L, and Smedh U. This study presents the long-term outcomes of a prospective, randomised trial comparing laparoscopic myotomy and pneumatic dilatation as treatment options for achalasia. (45), and they were match as in this study. Post-operative morbidity was 3.5%, two patients, and consists of atelectasis and simple wound infection.

Post-operative hospital stay mean was 2.5 days in L.H.M., 4.5 days in open procedure. Numerous comparative investigations examining open and laparoscopic surgical approaches have yielded inconclusive evidence on the occurrence of dysphagia, since no significant differences have been observed between the two methods. However, it has been consistently demonstrated that patients who undergo laparoscopic surgery experience shorter hospital stays and recovery periods compared to those who receive open surgery (22).

Advantages of minimal invasive surgery include:

1. A few small cuts versus a large incision
2. Less traumatic to the muscle, nerves and organs
3. Less bleeding
4. Less scarring
5. Less pain and reduced use of narcotics
6. Less hospital time
7. Less effect on immune system.

For all the fore mentioned advantage, patients with laparoscopic Hellers myotomy spend less time in the hospital as seen in our study.

5. CONCLUSIONS

In summary, the surgical Heller operation has been found to offer enduring alleviation from symptoms based on a one-year follow-up period. Laparoscopy has emerged as the preferred surgical method for treating achalasia because to its advantages over open procedures, including reduced hospitalisation, decreased bleeding, decreased analgesic usage, and shorter recovery time for resuming regular activities..

Heller myotomy has been found to yield symptom improvement in around 90% of patients, with a significant proportion of individuals experiencing sustained relief even throughout extended periods of postoperative monitoring. However, it is important to note that a small proportion of cases may exhibit persistent or recurring symptoms. In such instances, a comprehensive evaluation of these patients is vital to ensure appropriate therapies, such as

pneumatic dilation or reoperation, are administered. Esophagectomy should be reserved for patients with really severe esophageal dilation.

Limitations:

Limitations of presented study are the small sample size due to rarity of the disease, prospective study design, the potential selection bias, the limited duration of follow-up period not exceeding 18 months, and the lack of postoperative manometry and 24 hours pH monitoring assessment.

Ethical Approval:

All ethical issues were approved by the author. Data collection and patients enrollment were in accordance with Declaration of Helsinki of World Medical Association , 2013 for the ethical principles of researches involving human. Signed informed consent was obtained from each participant and data were kept confidentially.

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