

## Achalasia Cardia: Short-term Results of 40 Iraqi Patients

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### Summary:

**Background:** Achalasia is an uncommon but not a rare a malady. In Iraq, we lack true statistics about this condition.

**Objective:** Is to review the experience with trans-thoracic modified Heller operation for achalasia cardia in a major thoracic surgical centre in Iraq over a 4-year period.

**Study design:** a combined retrospective and prospective study.

**Setting:** Department of Thoracic Surgery in Baghdad Medical City/Baghdad/Iraq.

**Patients:** Forty patients (26 males and 14 females) with achalasia cardia who were admitted to the Department of Thoracic Surgery in Baghdad Medical City over a period of 4 years (2008 to 2012).

**Methods:** This study was both retrospective (20 cases) and prospective (20 cases); the information was collected from either patients, case sheets or obtained directly from patients, interviews. In both situations, relevant demographic and clinical information was obtained. The patients then were subjected to a standard diagnostic workup followed by preparation of patients for surgery. A trans-thoracic modified Heller's operation: esophagocardiomyotomy) was done following a standard technique. No anti-reflux procedure was added. Follow up was done to evaluate results of surgery.

**Results:** 26 males (65%) and 14 females (35%) with a mean age of 26.6 year. Patients aged 20 -40 years constituted the majority (47.5%). The commonest presenting symptom was dysphagia reported in 39 patients (75%). Cucumber dilated esophagus (rat tail sign) was the commonest appearance on contrast study found in 84% of studied patients. Ten patients (7 females and 3 males) had dilation therapy initially which failed to improve their symptoms thus scheduled for surgery. The remaining 30 patients were offered surgical treatment from the start; thus ultimately, operation was done to all 40 patients. Good to excellent results after surgery were obtained in 35 patients (87.5%).

**Conclusions:** Although good and excellent results followed Heller's operation for achalasia cardia versus multiple forceful dilatations, laparoscopic Heller oesophageal myotomy reverses the symptoms of achalasia with minimal morbidity. It is recommended to learn and practice this minimal invasive procedure rather than to continue doing an old operation.

**Key words:** Achalasia cardia, Heller's operation, Endoscopic dilatation.

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### Introduction:

Sir Thomas Willis, an English anatomist, first described a case of achalasia in 1674 and successfully treated the patient with a whalebone dilator. Von Mikulicz suggested in 1881 that oesophageal spasm might be causal in achalasia and named this disorder as (cardiospasm). However, this (spasm theory) was questioned by Hurst and Rake in 1915; instead, they advocated that the pathology was due to a failure of relaxation rather than spasm and, therefore, termed this disorder achalasia (from the Greek term meaning lack of relaxation). Russell, in 1898, performed the first successful pneumatic dilation of esophagus, whereas Ernst Heller, a German surgeon performed the first successful cardiomyotomy on April 14, 1913 (using both anterior and posterior incisions). The procedure was later modified to a single incision by Groenvedeldt and Zaaier.<sup>1</sup> The aim of this study was to review the experience with trans-thoracic modified Heller's operation for achalasia cardia in a major thoracic surgical centre in Iraq over a 4-year period.

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**Patients:** Forty patients (26 males and 14 females) with achalasia cardia admitted to the Department of Thoracic Surgery in Baghdad Medical City were included over a period of 4 years (2008 to 2012).

### Methods:

This study was both retrospective (20 cases) and prospective (20 cases); the information was collected from either patients' case sheets or obtained directly from patients' interviews. In both situations, the following information was looked for: age, sex, occupation, race, symptoms (dysphagia, regurgitation, pain, heartburn, weight loss, cough, etc), signs (cachexia, pallor) and family history of achalasia. For patients studied prospectively, a detailed history was taken and a thorough physical examination was performed. The patients then were subjected to a standard diagnostic workup. The specific investigations included plain chest radiography, barium esophagogram and endoscopy (rigid oesophagoscopy or flexible OGD). Oesophageal manometry was not done

because it was neither available in thoracic surgery department nor in gastroenterology centre at the time of the study. After establishing the diagnosis the patients were prepared for surgical treatment (transthoracic modified Heller operation: esophagocardiomyotomy). Preoperative preparation involved treatment of any existing chest infection and correction of malnutrition if present. Prior to surgery, the esophagus was cleansed via rigid oesophagoscopy to minimize aspiration. Opportunity was taken also to exclude other pathological lesions simulating achalasia like strictures and tumours. Surgery was done following a standard technique as detailed in operative textbooks.<sup>2</sup> The NG tube was either removed immediately or left for 24 hours; in the latter situation, it was fixed to the nose and the nursing staff was instructed not to replace it in the event of inadvertent removal. Most of the times, no antireflux procedure was added. In smooth cases, oral liquid diet was started the next day and chest tube was removed in 24 hours. However, if perforation was encountered, NG tube was kept for 5 days, patient was given IV fluid and antibiotics. Oral

liquid diet was started after a normal Gastrograffin swallow performed on 5th postoperative day. Most patients were discharged home in a week period. Follow up was done to evaluate results of surgery.

**Results:**

Sex distribution: 26 males (65%) and 14 females (35%). The youngest patient was 1.5 year old while the oldest one was 65 years old. The mean age was 26.65 year. Patients aged 20 -40 years constituted the majority (47.5%). Children younger than 10 years constituted 25% of cases. Two patients had a family history of achalasia (5%). They were brothers (22 and 25 years old respectively had surgery with an interval of one year between them. Interestingly, the second patient; though recovered from general anesthetic smoothly, developed ptosis one week after operation and referred then to a neurologist who made a diagnosis of myasthenia gravis. His swallowing had improved after operation according to information obtained from the surgeon who did surgery for him.

**Table-1 Age and sex distribution**

Gender	Age (yr)							
	Up to 10	11-20	21-30	31-40	41-50	51-60	61 & above	
Males, n(%)	6 (15)	2 (5)	9(22.5)	3 (7.5)	3 (7.5)	2 (5)	1 (2.5)	
Females, n (%)	4 (10)	2 (5)	3 (7.5)	4 (10)	0 (0)	1 (2.5)	0 (0)	
Total, n(%)	10 (25)	4 (10)	12 (30)	7 (17.5)	3 (7.5)	3 (7.5)	1 (2.5)	

**Table-2 Symptomatology**

Symptom	Males, n (%)	Females, n (%)	Total, n (%)
Dysphagia Dysphagia due to foreign body impaction (food- related articles)	8 (20) 4 (10)	4 (10) 1 (2.5)	12 (30) 5 (12.5)
Dysphagia & Regurgitation	7 (17.5)	2 (5)	9 (22.5)
Regurgitation	2 (5)	1 (2.5)	3 (7.5)
Cough	5 (12.5)	3 (7.5)	8 (20)
Dysphagia with chest pain	1 (2.5)	2 (5)	3 (7.5)
Total	26 (65)	14 (35)	40 (100)

The duration of symptoms ranged between 2 months to 6 years as shown in Table 3.

Table 3 Duration of symptoms	Number of patients (Male)	Number of patients (female)	Total
0 -1 year	10 (25)	5 (12.5)	15 (37.5)
1-2 year	9 (22.5)	4 (10)	13 (32.5)
More than 2 years	6 (15)	6 (15)	12 (30)
Total	25 (62.5)	15 (37.5)	40 (100)

**Table-4: Plain radiographic appearances in the prospective group of patients.**

X-ray Findings	Patients, n (%)
Normal x-ray	13 (65)
Signs of bronchitis and pneumonic changes	7 (35)
Total	20 (100)

Barium swallow findings could be studied in 25 patients only. A common change in all these patients was narrowed lower end of esophagus and proximal dilation. Other appearances are listed in table 5:

**Table-5 Signs on barium swallow**

Sign on barium swallow	Patients, n (%)
Cucumber dilated esophagus	21 (84)
Sigmoid mega esophagus	4 (16)
Total	25 (100)

Treatment: Ten patients had dilation therapy prior to surgery. The remaining 30 patients were offered surgical treatment from the start; thus eventually, operation was done to all 40 patients. Four patients had a trial of dilatation via rigid oesophagoscopy

under GA using Maloney dilators performed by the surgeon. As the response was poor; surgery was offered after few weeks. Six patients had dilatation by gastroenterologists using flexible endoscopes. These patients consulted thoracic surgeons later due to poor response and thus were elected for surgery as well.

**Table-6 Treatment Options**

Treatment option	Patients, n (%)
Repeated dilatation followed by surgery	10 (25)
Modified transthoracic Heller's operation	30 (75)
<b>Total</b>	<b>40 (100)</b>

Ten patients underwent dilation therapy in this study (7 females and 3 males). Their ages ranged from 2 yrs to 60 yrs; the mean age was 28.45 yrs. Three patients were children while the remaining were 16 yrs and older. The youngest patient underwent dilatation in this study was a 2 yrs old child with dysphagia and recurrent chest infection for few months. She had bougienage under GA. The second was also a child 2 and a half year old with a similar history; underwent bougienage under GA also. The oldest patient was a 60 yrs old lady with history of dysphagia of 4 yrs duration. She had a megaesophagus as shown by contrast study. She received a pneumatic dilatation by a gastroenterologist. Six patients received a pneumatic dilatation by a gastroenterologist (Balloon sizes 3, 3.5 and 4 cm distended for 0.5 to 1 minute through an endoscope under fluoroscopy) and 4 patients had a bougienage under GA done by surgeons. All patients had a long-standing dysphagia. Results of surgery were considered excellent when symptoms completely relieved, good when symptoms became mild, fair when symptoms became moderate and poor when symptoms persisted or complications ensued.

**Table-7 Results of surgery**

Result	Excellent	Good	Fair	Poor	Total
No. & % of pt	20 (50)	15 (37.5)	2 (5)	3 (7.5)	40 (100)

Three patients had poor results: the first had persistent dysphagia and was referred for gastroenterologist for dilation therapy. The second had postoperative heartburn and offered Belsey mark IV operation later on. The third developed hiatal hernia and sought treatment abroad.

**Discussion:**

Achalasia is an uncommon but not a rare a malady.<sup>3</sup> In Iraq, we lack statistics about this condition. However, Husein WM in a study on achalasia in Baghdad Medical City Teaching Hospital could collect only 50 cases in 6 years period i.e., 8.3 case per year.<sup>3</sup> The present study which is conducted in the same centre, involved 40 patients in 4 years i.e., an annual incidence of 10 patients per year which is just slightly higher than the previous study.

Though literature states that achalasia occurs equally in males and females<sup>4</sup>, our study showed a higher incidence among males (1.8:1 males to females). Therefore, we think that no specific sex predisposition can be claimed. The age distribution of the patients in the present study revealed 2 peaks; the first was among children younger than 10 years (25%) and the second was among young adults in the third decade of life (30%). Previous studies did show a peak among young adults<sup>3</sup> but not so in children. It is clear that achalasia can affect people from infancy<sup>3</sup> to extreme adulthood. The youngest patient in the present study was one and a half year while there was an infant of 30 days in Husein WM, study.<sup>3</sup> Two patients had a family history of achalasia (5%) quite similar to the previous study of Husein WM (6%).<sup>3</sup> It is very interesting to have an association between achalasia and myasthenia gravis (MG) in this study. Henry J. Kaminski described achalasia and MG in a patient with thymoma suggesting that achalasia may occur as a paraneoplastic immune disorder.<sup>5</sup>

**Radiography:** Seven out of 20 patients in the present study (35%) had signs of pneumonitis on plain chest X-ray; higher than reported by Harley (13.33%).<sup>6</sup> This difference is explained by the late presentation and negligence of our patients. Barium swallow is of great importance in the differential diagnosis of achalasia. Cucumber esophagus was the commonest finding in our study (21 out of 25 patients 84%); a similar finding in other studies.<sup>3,6</sup>

**Esophagoscopy:** Esophagoscopy examination in 24 patients i.e., 60% (from whom information could be obtained) revealed retained food debris cleared by esophagoscopy irrigation and suction. Esophagoscopy is mandatory and useful to confirm the diagnosis and to exclude other conditions simulating achalasia like carcinoma of the cardia.<sup>3</sup> Most of the patients in this study had esophagoscopy examination using the rigid instrument. In view of the liability for perforation of the achalasic esophagus, flexible OGD could be safer beside its added benefit of examining the stomach and duodenum as well. **Endoscopic ultrasonography** enhances our ability to detect achalasia; the lower esophageal sphincter (LES) is about 31 mm thick in achalasia compared with 22 mm seen in normal persons. Furthermore, it adds in-depth view of the esophageal wall to detect malignancies by showing thickening in mucosal and sub-mucosal layers and lymph nodes and is proven to be better than CT scan for detecting sub-mucosal disease.<sup>1</sup>

**Esophageal manometry:** This is the gold standard for the diagnosis of achalasia.<sup>1</sup> None of the patients in this study had Endoscopic ultrasonography or esophageal manometry; probably they were not available at the time of the study.

**Treatment options:** Good and excellent results are obtained among patients with achalasia treated primarily by Heller's operation versus those treated with multiple forceful dilatations. The Mayo Clinic had reported a comparison involving 431 patients treated by forceful dilation and 456 who underwent esophagomyotomy. A successful outcome was achieved in 81% of the former and 94% of the latter. Not

only did myotomy produce better results, but it was safer than dilation. There were a higher mortality rate and a greater risk of perforation for patients undergoing dilation.<sup>7</sup> Despite this fact; it is very interesting that physicians still favor dilation as a first step for achalasia. One surgeon from France commented on this by saying that myotomy has almost disappeared from their operating program because of forceful dilation done by physicians. (We do not see them anymore, except late in the evening or on Saturdays, when physicians have used forceful dilation and ruptured the esophagus and then we are needed) he says. He adds that in 2 to 3 years he hadn't seen one primary indication for myotomy. Instead, he used to operate on patients who had had prior unsuccessful dilatations. Thus surgeons who still see achalasia cases coming for surgery as a primary option are considered very lucky.<sup>7</sup> In the present study, all patients who were managed initially by dilatation except 2 were studied retrospectively. This explains the paucity of information relevant to these patients. It was not clear for us the basis on which dilatation therapy was offered to them. The patients who received pneumatic dilatation were dealt with by gastroenterologists who believe that this method of therapy is the ideal primary treatment option for achalasia; thus were managed in this way without a surgical consultation. Unfortunately, all of them did not benefit from dilation therapy and thus consulted surgeons later to have surgery which cured or improved their symptoms. Surgeons performed bougienage for 4 patients in this study (2 of them were very small children). We think that these patients, children in particular should have surgery without a trial of dilatation. As symptoms used to be severe in children and respiratory complications are frequent, surgery should be considered from the start rather than wasting time in unsuccessful dilation.<sup>3,8-11</sup> In Europe most Heller operations are done through an abdominal approach, and if one has to bring down the esophagus to perform the myotomy, one destroys its posterior and lateral attachments. Therefore, the rate of reflux cases through an abdominal approach goes up to 15%; so although dysphagia is cured; reflux is promoted, which is not quite a success. That is why anti-reflux procedures have been advocated by most European surgeons, because they are using the abdominal approach. It is very illogical to destroy the hiatus and then try to repair it. Furthermore, during an abdominal approach, there is a tendency for the incision to be carried down too far on the stomach, thus increasing the likelihood of postoperative GERD. Richard Earlham's literature review of the results of myotomy for achalasia emphasized that the incidence of postoperative reflux following the abdominal approach was high unless an anti-reflux procedure was employed. The thoracic approach, on the other hand, incurs minimal trauma to the hiatus leaving the esophagus within its berth. When a short transthoracic myotomy is employed, an anti-reflux procedure is not necessary.<sup>7</sup> End stage achalasia, characterized by a markedly dilated and tortuous "burned-out" esophagus and recurrent obstructive symptoms, may require esophageal resection in

order to restore gastrointestinal function, reverse nutritional deficits and reduce the risk of aspiration pneumonia.<sup>12</sup> After the popularization of minimally invasive surgery, Pellegrini and coworkers in 1992 reported the first use of thoracoscopic esophagomyotomy for achalasia. Rosati and colleagues in 1995 first reported excellent relief of symptoms after a laparoscopic myotomy and anterior partial fundoplication for achalasia.<sup>1,13-15</sup> A follow-up of thoracoscopic esophagomyotomies showed a high incidence of dysphagia and reflux. Stewart and colleagues have shown that laparoscopic myotomy results in less mucosal perforations and postoperative dysphagia and a GERD than the thoracoscopic approach. The incidence of inadequate myotomy and GERD has led to favor laparoscopic Heller's myotomy for the primary surgical therapy of achalasia.<sup>1,16,17</sup>

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