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: esophagocardiomityomy) 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 dilatation 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 esophageal 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.

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 (cardiospasms). 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 dilatation of esophagus, whereas Ernst Heller, a German surgeon performed the first successful cardiomityomy on April 14, 1913 (using both anterior and posterior incisions). The procedure was later modified to a single incision by Groenvedeldt and Zaajar.1 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.

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 oesophagoscope 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 (transabdominal modified Heller operation: esophagogastrectomy). Preoperative preparation involved treatment of any existing chest infection and correction of malnutrition if present. Prior to surgery, the oesophagus 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. 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 Gastrografin 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 years. 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) who had surgery with an interval of one year between them. Interestingly, the second patient, though recovered from general anesthetic smoothly, developed postis 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>
<thead>
<tr>
<th>Table-1 Age and sex distribution</th>
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<tbody>
<tr>
<td>Gender</td>
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<tr>
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</tr>
<tr>
<td>Males, n (%)</td>
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<tr>
<td>Females, n (%)</td>
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<tr>
<td>Total, n (%)</td>
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</table>

<table>
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<tr>
<th>Table-2 Symptomatology</th>
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<tbody>
<tr>
<td>Symptom</td>
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<tr>
<td>---------</td>
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<tr>
<td>Dysphagia</td>
</tr>
<tr>
<td>Dysphagia due to foreign body impaction (food-related articles)</td>
</tr>
<tr>
<td>Dysphagia &amp; Regurgitation</td>
</tr>
<tr>
<td>Regurgitation</td>
</tr>
<tr>
<td>Cough</td>
</tr>
<tr>
<td>Dysphagia with chest pain</td>
</tr>
<tr>
<td>Total</td>
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</table>

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

<table>
<thead>
<tr>
<th>Table 3 Duration of symptoms</th>
<th>Number of patients (Male)</th>
<th>Number of patients (Female)</th>
<th>Total</th>
</tr>
</thead>
<tbody>
<tr>
<td>0-1 year</td>
<td>10 (25)</td>
<td>5 (12.5)</td>
<td>15 (37.5)</td>
</tr>
<tr>
<td>1-2 year</td>
<td>9 (22.5)</td>
<td>4 (10)</td>
<td>13 (32.5)</td>
</tr>
<tr>
<td>More than 2 years</td>
<td>6 (15)</td>
<td>6 (15)</td>
<td>12 (30)</td>
</tr>
<tr>
<td>Total</td>
<td>25 (62.5)</td>
<td>15 (37.5)</td>
<td>40 (100)</td>
</tr>
</tbody>
</table>

<table>
<thead>
<tr>
<th>Table-4: Plain radiographic appearances in the prospective group of patients.</th>
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<tbody>
<tr>
<td>X-ray Findings</td>
</tr>
<tr>
<td>----------------</td>
</tr>
<tr>
<td>Normal x-ray</td>
</tr>
<tr>
<td>Signs of bronchitis and pneumonia changes</td>
</tr>
<tr>
<td>Total</td>
</tr>
</tbody>
</table>

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

<table>
<thead>
<tr>
<th>Table-5 Signs on barium swallow</th>
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<tbody>
<tr>
<td>Sign on barium swallow</td>
</tr>
<tr>
<td>-------------------------</td>
</tr>
<tr>
<td>Cucumber dilated esophagus</td>
</tr>
<tr>
<td>Sigmoid mega esophagus</td>
</tr>
<tr>
<td>Total</td>
</tr>
</tbody>
</table>

Treatment: Ten patients had dilatation 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

<table>
<thead>
<tr>
<th>Treatment option</th>
<th>Patients, n (%)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Repeated dilatation followed by surgery</td>
<td>10 (25)</td>
</tr>
<tr>
<td>Modified transthoracic Heller’s operation</td>
<td>30 (75)</td>
</tr>
<tr>
<td><strong>Total</strong></td>
<td><strong>40 (100)</strong></td>
</tr>
</tbody>
</table>

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 megasophagus as shown by contrast study. Six patients 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

<table>
<thead>
<tr>
<th>Result</th>
<th>Excellent</th>
<th>Good</th>
<th>Fair</th>
<th>Poor</th>
<th>Total</th>
</tr>
</thead>
<tbody>
<tr>
<td>No. &amp; % of pt</td>
<td>20 (50)</td>
<td>15 (37.5)</td>
<td>2 (5)</td>
<td>3 (7.5)</td>
<td>40 (100)</td>
</tr>
</tbody>
</table>

Three patients had poor results: the first had persistent dysphagia and was referred for gastroenterologist for dilatation 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 malady.² 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.³ 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 females⁴, 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 adults⁵ but not so in children. It is clear that achalasia can affect people from infancy⁶ 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.³ Two patients had a family history of achalasia (5%) quite similar to the previous study of Husein WM (6%).³ 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 as a paraneoplastic immune disorder.⁷

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%)³. 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.³,⁶

Esophagoscopy: Esophagogoscopic examination in 24 patients i.e., 60% (from whom information could be obtained) revealed retained food debris cleared by esophagogoscopic irrigation and suction. Esophagoscopy is mandatory and useful to confirm the diagnosis and to exclude other conditions simulating achalasia like carcinoma of the cardia.³ Most of the patients in this study had esophagogoscopic 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.¹

Esophageal manometry: This is the gold standard for the diagnosis of achalasia.¹ 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 Hellers operation versus those treated with multiple forceful dilatations. The Mayo Clinic has 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.7 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 dilations. Thus
surgeons who still see achalasia cases coming for surgery as
a primary option are considered very lucky.7 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 dilatation 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 dilatation.5,8-11 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
recession 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.7 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.12 After
the popularization of minimally invasive surgery, Pellegreni
and coworkers in 1992 reported the first use of thorascopic
esophagomyotomy for achalasia. Rosati and colleagues in 1995
first reported excellent relief of symptoms after a laparoscopic
myotomy and anterior partial fundoplication for achalasia.13-15
A follow-up of thorascopic 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 thorascopic approach. The incidence of inadequate
myotomy and GERD has led to favor laparoscopic Heller’s
myotomy for the primary surgical therapy of achalasia.13,16,17

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