

## Case Series

# Surgical management of achalasia cardia-analysis of immediate outcomes

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## ABSTRACT

Achalasia cardia is a motility disorder characterised by failure of relaxation of the lower oesophageal sphincter (LES) and associated with lack of peristalsis in the body of oesophagus. The mainstay of treatment in achalasia cardia is to reduce the LES pressure and improve oesophageal emptying by gravity. LHM is still considered the gold standard against which other methods are compared. The records of all patients who underwent surgery for achalasia cardia between August 2018 and December 2021 in the department of Surgical Gastroenterology, Government Rajaji Hospital and Madurai Medical College, Madurai, Tamil Nadu, India were reviewed retrospectively. Eighteen patients underwent surgery for achalasia cardia with median age of 32.5 years and the median duration of symptoms was 30 months. Seven patients had undergone pneumatic dilatation previously. Two patients who had mucosal injury had undergone pneumatic dilatation previously. The duration of surgery was significantly higher for the patients who underwent previous pneumatic dilatation when compared to those who did not undergo dilatation (200 minutes vs 170 minutes). The Eckardt score was zero in 12 patients in follow-up. One patient had a score of 1 and five patients had a score of 2. Laparoscopic Heller myotomy is the most effective current surgical management for achalasia cardia. It can be safely performed even in patients with prior pneumatic dilatations with lesser reintervention rates.

**Keywords:** LES, Laparoscopic Heller myotomy, Mucosal injury, Eckardt score

## INTRODUCTION

Achalasia cardia is a motility disorder affecting the body of oesophagus and the LES. It is characterised by failure of relaxation of the LES and commonly associated with lack of peristalsis in the body of oesophagus.<sup>1</sup> It occurs as a result of decreased or absent inhibitory neurons and ganglion cells in the myenteric plexus of the oesophagus and LES.<sup>2</sup> The loss of these neurons is thought to be caused by inflammation related to an underlying autoimmune, viral or degenerative process.<sup>3</sup> Patients usually present with dysphagia, chest pain, regurgitation and weight loss. Patients may also present with heartburn, vomiting and recurrent episodes of aspiration or pneumonia. It usually affects young adults and both genders equally.<sup>4</sup> Though achalasia cardia is relatively

rare, higher awareness about the disorder and wider availability of high-resolution manometry has helped readily diagnose the disorder.

Diagnosis is usually done using a combination of barium swallow, upper gastro-intestinal endoscopy and high-resolution manometry. Barium swallow demonstrates a dilated oesophagus with a smooth tapering of the distal oesophagus close to the gastro-oesophageal junction (bird-beak appearance). More severe cases may show a tortuous oesophagus or an associated epiphrenic diverticulum. Endoscopy show a dilated oesophagus with food stasis and a tight LES. Oesophageal manometry is the most sensitive to diagnose achalasia cardia indicating an absence of peristalsis, high pressure in the LES, and failure of the LES to relax on swallowing.<sup>5</sup> Endoscopy

and further cross-sectional imaging, usually a contrast enhanced CT scan help rule out pseudo-achalasia.

The mainstay of treatment in achalasia cardia is to reduce the LES pressure and improve oesophageal emptying by gravity.<sup>4</sup> Various methods are available to achieve LES degradation including pneumatic dilatation (PD), Laparoscopic Heller myotomy (LHM), botulinum toxin injection, pharmacotherapy and peroral endoscopic myotomy (POEM). But LHM is still considered the gold standard against which other methods are compared.

### CASE SERIES

The records of all patients who underwent surgery for achalasia cardia between August 2018 and December 2021 in the department of Surgical Gastroenterology, Government Rajaji Hospital and Madurai Medical College, Madurai, Tamil Nadu, India were reviewed retrospectively. The age and sex of the patients, a detailed history of clinical presentation, duration of symptoms and details of investigations in the form of Barium swallow, contrast enhanced CT scan Upper gastro-intestinal (GI) endoscopy and high-resolution manometry were noted. Details of previous interventions if any were recorded. The operative procedure was LHM with posterior Toupet (270°) fundoplication. The intra-operative details regarding duration of surgery, incidence of intraoperative mucosal injury, type of fundoplication and the need for a drain was noted. Post-operative recovery, hospital stay, improvement in symptoms and weight gain was noted. Patients were followed up for recurrence of symptoms and the need for re-intervention. The Eckardt score was noted at follow-up.

Out of 18 patients who underwent surgery for achalasia cardia nine were male and nine female. The median age of the patients was 32.5 years (range: 17-50 years) and the median duration of symptoms was 30 months (range: 7-240 months). Seven patients had undergone pneumatic dilatation previously. Of these, two patients underwent pneumatic dilatation twice and one patient dilatation with stenting presuming it to be a malignancy. Five out of 18 patients had megaesophagus on contrast enhanced CT scan (Table 1).

**Table 1: Demographic parameters.**

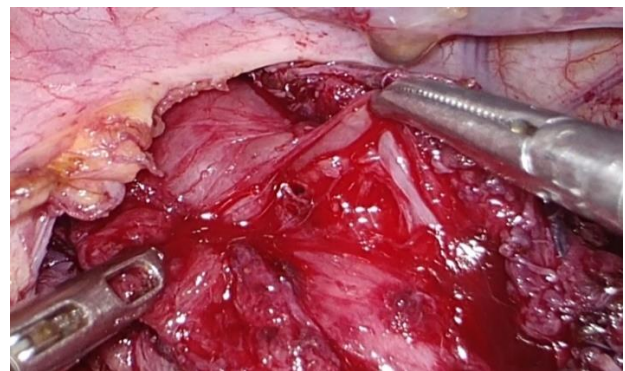
Parameters	Variables
No. of patients	18
Age	17-50 years, median (32.5 years)
Duration of symptoms	7-240 months median (30 months)
Previous treatment	2-Pneumatic dilatation 1-Dilatation and stenting

The preferred operative procedure was LHM with posterior Toupet fundoplication. The patient was placed with a leg split and the knees bent with support. The monitor was placed at the head end and the surgeon

standing between the patient's legs. The lesser omentum was divided and the proximal short gastric vessels were also divided. The lower oesophagus was looped and mobilised into the hiatus. A long myotomy was done extending at least 5 cm on the oesophageal side and 2 cm on the stomach side. A combination of blunt dissection, Ultrasonic shears and distraction was used for the myotomy. The hiatus was narrowed if wide. A posterior 270° Toupet fundoplication was done using 2-0 dacron sutures (Figure 1). The wrap was also fixed to the crus of the diaphragm to prevent wrap migration. In the two patients who had mucosal injury (Figure 2), the rent in the oesophageal mucosa was sutured with fine (3-0) vicryl interrupted sutures. The repair was buttressed by and anterior Dor fundoplication in these two patients (Figure 3). A drain tube was placed for these patients. Both patients who had mucosal injury had undergone pneumatic dilatation previously (Figure 4).



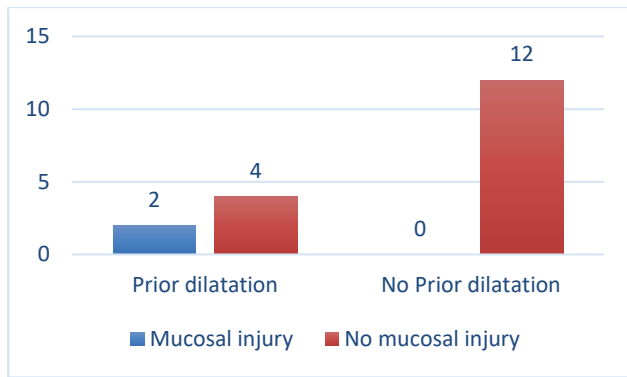
**Figure 1: Toupet fundoplication.**



**Figure 2: Mucosal injury during myotomy.**



**Figure 3: Dor fundoplication.**



**Figure 4: Relationship of prior dilatation to mucosal injury.**

One patient had underwent pneumatic dilatation five years back and later stenting presuming it to be a malignancy. The stent had migrated into the stomach and further endoscopy, imaging and follow-up did not reveal malignancy. patient underwent LHM with Toupet fundoplication and gastrotomy to remove migrated stent.

The median duration of the procedure was 187.5 minutes (range: 140-240 minutes). The duration of surgery was significantly higher for the patients who underwent previous pneumatic dilatation when compared to those who did not undergo dilatation (200 minutes vs 170 minutes). Patients were started on oral liquids on the first post-operative day as a routine. In the patients who had mucosal injury, oral liquids were started after the third post-operative day. The median post-operative hospital stay was 4 days (range: 3-8 days).

Patient were under followed up for a median of 12 months (range: 1-41 months). During follow-up 17 patients had complete resolution of dysphagia. One patient had persistence of mild dysphagia. Five patients had occasional regurgitation which was managed with drugs. The Eckardt score was zero in 12 patients in follow-up. One patient had a score of 1 and five patients had a score of 2 which was the maximum among patients (Table 2). Three out of 5 patients who had an Eckardt score of 2 had megaesophagus in pre-operative imaging.

**Table 2: Outcomes.**

Parameters	Variables
Type of fundoplication	Toupet-16, Dor-2
Mucosal injury	2
Duration of surgery in patients without previous treatment	170 minutes
Duration of surgery in patients with previous dilatation	200 minutes
Hospital stay	3-8 days (median 4 days)
Follow up	1-41 months (median 12 month)
Eckardt score	0-2

## DISCUSSION

The main goal of treatment in patients with achalasia cardia is to relieve the functional obstruction of the lower oesophagus caused by the high LES pressure and the failure of the LES to relax in response to deglutition.<sup>6</sup> The common treatment modalities available at present are endoscopic pneumatic dilatation, Laparoscopic Heller myotomy (LHM) and peroral endoscopic myotomy (POEM).<sup>7-11</sup> Botulinum toxin injection and pharmacotherapy is not common modalities of treatment and are reserved for patients who are at high risk for surgery.<sup>12</sup> POEM is the latest modality of treatment available for achalasia cardia claiming efficacy equal to LHM. But all studies are based predominantly on small retrospective studies and long-term effects are yet to be established.<sup>13</sup> Pneumatic dilatation is effective in elderly patients and the effects are short-lived in younger patients requiring repeated dilatations. Pneumatic dilatation can cause scarring around the LES making surgery difficult if need be later.<sup>14</sup>

LHM is the primary surgical management of achalasia cardia with a response rate exceeding 90%.<sup>15,16</sup> Heller myotomy involves linear division of the circular muscle fibres of the lower oesophagus for a length of 6 cm above the oesophago-gastric junction and 3 cm below it anteriorly. The anterior vagus is identified and preserved during the procedure to prevent injury. The advent of laparoscopy and the increasing skills of laparoscopic surgeons to perform complex procedures has facilitated the successful and effective execution of LHM. The extension of the myotomy to at least 3 cm on the gastric side is considered to be important. Recurrence of symptoms is usually due to a shorter myotomy on the stomach side. An adequate myotomy is usually associated with a significant gastro oesophageal reflux and hence a fundoplication is added to the procedure.<sup>8,17</sup> The type of fundoplication used is controversial. Incidence of post operative dysphagia is higher with Nissen's fundoplication when compared to partial fundoplication.<sup>18</sup> So, either an anterior 'Dor' or a posterior 'Toupet' fundoplication is usually performed. It is argued that a posterior Toupet is better as helps to hold the myotomy apart preventing re-approximation.

The complications of LHM are inadequate myotomy leading to persistence of dysphagia, gastro oesophageal reflux disease (GERD) due to the myotomy, symptoms due to inadvertent vagotomy and mucosal injury which is usually identified intra operatively and sutured without consequences.<sup>19-28</sup> In our centre posterior Toupet fundoplication is routinely performed following LHM. But in patients who had mucosal injury identified intra operatively and sutured (2 patients), an anterior Dor fundoplication is done. We believe it helps to buttress and support the repair. None of the two patients had a leak and both of them had a smooth recovery. We also noted that both the patients who had mucosal injury had undergone pneumatic dilatation. Previous PD made



dissection around the oesophago-gastric junction and lower oesophagus difficult. The risk of mucosal injury was higher in these patients. LHM may be the preferred first line management in appropriate young and fit individuals. PD is less effective with a response rate of around 68% as compared to about 89% with LHM.<sup>8</sup> PD also made subsequent surgery difficult with higher complication rate. In other studies, more than 50% patients who underwent PD required subsequent intervention at 5 years, whereas only about 30% of patients who underwent LHM needed subsequent intervention at 5 years.<sup>29</sup>

The effect of LHM is perceived to be suboptimal in patients with sigmoid oesophagus and type III achalasia cardia. Oesophagectomy is thought to be a better surgical option for a sigmoid oesophagus. But in our series two patients had sigmoid oesophagus. One of the patients had complete resolution of dysphagia and the other had mild persistence of symptoms. We find, LHM may still be effective in patients with sigmoid oesophagus. Oesophagectomy is a major procedure with significant morbidity and is reserved as a last resort.

Proponents of POEM claim better improvement of symptoms after POEM over LHM in short term.<sup>30,31</sup> But there are not many studies comparing long term outcomes after LHM and POEM. POEM is also associated with significantly higher incidence of GERD.<sup>9</sup>

## CONCLUSION

Laparoscopic Heller myotomy is the most effective current surgical management for achalasia cardia. As most patient with achalasia cardia present at a relatively young age, LHM may be adopted as the first line management in these fit patients. It can be safely performed even in patients with failed pneumatic dilatation or botulinum toxin injection. Even in patients who present late with megaesophagus or sigmoid oesophagus, LHM is reasonably effective and may avoid a major morbid procedure like oesophagectomy. LHM is associated with high efficacy and a low reintervention rate. The incidence of post-procedure troublesome GERD in the long term is also low. So LHM is still the gold standard in the treatment of achalasia cardia.

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