

# Successful Outcome After Laparoscopic Heller Myotomy And Dor Fundoplication Avoiding Esophageal Resection In A Patient With Megaesophagus And Advanced Idiopathic Achalasia

J Carvajal Balaguera, J Camuñas Segovia, M Martín García-Almenta, S Oliart Delgado de Torres, L Albeniz Aguiriano, L Peña Gamarra, C Cerquella Hernández

## Citation

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## Abstract

Laparoscopic Heller myotomy and Dor fundoplication is the surgical procedure of choice for esophageal achalasia. However, treatment of megaesophagus for advanced idiopathic achalasia has been controversial. Some authors recommend a myotomy as initial treatment and reserve esophageal resection for cases with persistent symptoms, whereas others recommend esophagectomy. We report the case of a patient with an esophageal advanced dilatation due to idiopathic achalasia. We have performed laparoscopic Heller myotomy and Dor fundoplication. The postoperative course was uneventful. There is no evidence of recurrence of the dysphagia and the patient is asymptomatic after the first year follow-up. Based on our experience with this case and based on a review of the literature, we discuss the different treatment modalities of this pathology.

## INTRODUCTION

Idiopathic achalasia is a degenerative disease characterized by a defective peristaltic activity of the esophageal body and impaired relaxation of the lower esophageal sphincter, which leads to difficult progression of a bolus into the stomach. It affects 1 in 100.000 individuals. The age of maximum presentation oscillates between 20 and 40 years, although it can appear in all ages. It affects men and women equally. It is not a hereditary disease, although family cases have been described<sup>1</sup>.

Surgical treatment of achalasia has evolved dramatically over the past 15 years; since the first report of laparoscopic Heller myotomy by Cushieri et al<sup>2</sup>, and thoracoscopic Heller myotomy by Pellegrini et al<sup>3</sup>. Currently, laparoscopic myotomy is the gold standard for treatment of achalasia. The satisfactory results of this procedure are well documented in several large series<sup>4,5,6,7,8</sup>. In some cases with a great esophageal dilation or advanced mega-esophagus, myotomy can alleviate also the symptoms of these patients<sup>9</sup>.

We present a case of mega-esophagus due to achalasia of long evolution, in a patient that did not respond to endoscopic dilations. Laparoscopic ample Heller myotomy with Dor anterior hemi-funduplication was performed with good outcome after a year of follow-up.

## CASE REPORT

A 36-year-old male was admitted to our service with complaints of severe dysphagia for liquids and solids, sialorrhea, cough due to broncho-aspiration, recurrent respiratory infection, wrong breath and regurgitation, of eight years evolution. Despite the medical therapy (i.e., vasodilative drugs, proton pump inhibitors, and prokinetics) and repeated endoscopic dilations, the dysphagia did not improve causing a loss of 10kg of weight. Physical exploration showed a patient thin and pale. The abdomen was soft without masses and not painful.

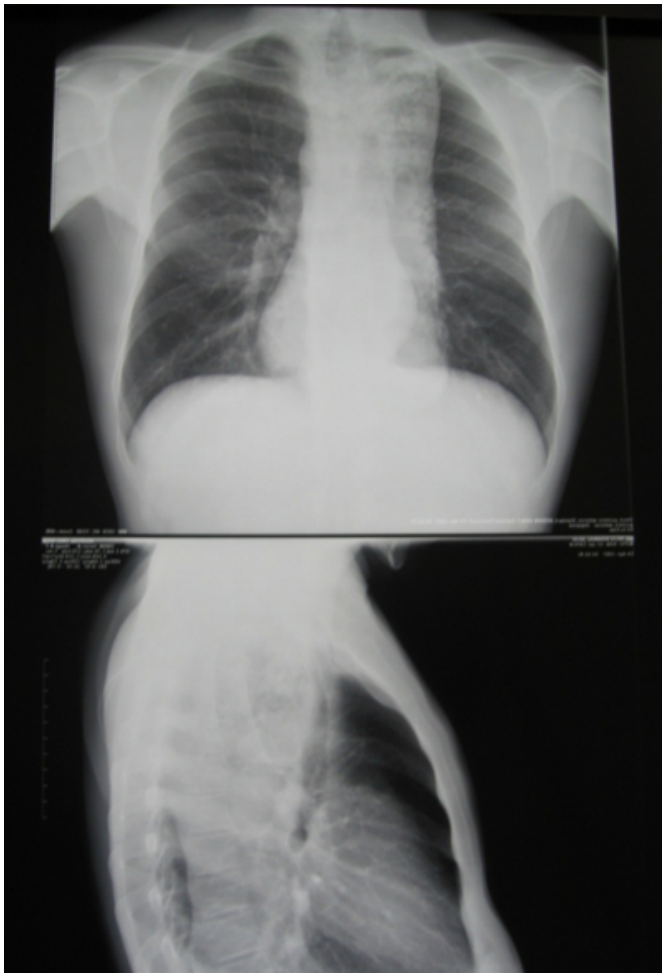
Laboratory data showed the following: Hemoglobin 12.2g/dl, hematocrit 35.3%, red blood count 3.700.000/ $\mu$ L, platelets 175.000/ $\mu$ L, white blood count 7.300/ $\mu$ L, partial

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thromboplastin time 32 seconds, prothrombin time 13.9 seconds, blood urea nitrogen 13 mg/dl, creatinine 0.4 mg/dl, aspartate aminotransferase 34 IU/l, alanine aminotransferase 45 IU/l, total bilirubin 0.8 mg/dl, sodium 140 mEq/l and potassium 4.5 mEq/L. Serum carbohydrate antigen 19-9, carcinoembryonic antigen and alpha-fetoprotein antigen were normal. Breath test for *Helicobacter pylori* was negative. Abdominal ultrasonography did not show any alteration. Thorax x-ray revealed a para-cardiac double shade on the right side with a hydro-aerial level and a mediastinum enlarged by esophageal dilatation (Fig. 1).

### Figure 1

Figure 1: Thorax x-ray revealed a para-cardiac double shade with hydro-aerial level and a mediastinum enlarged by esophageal dilatation.

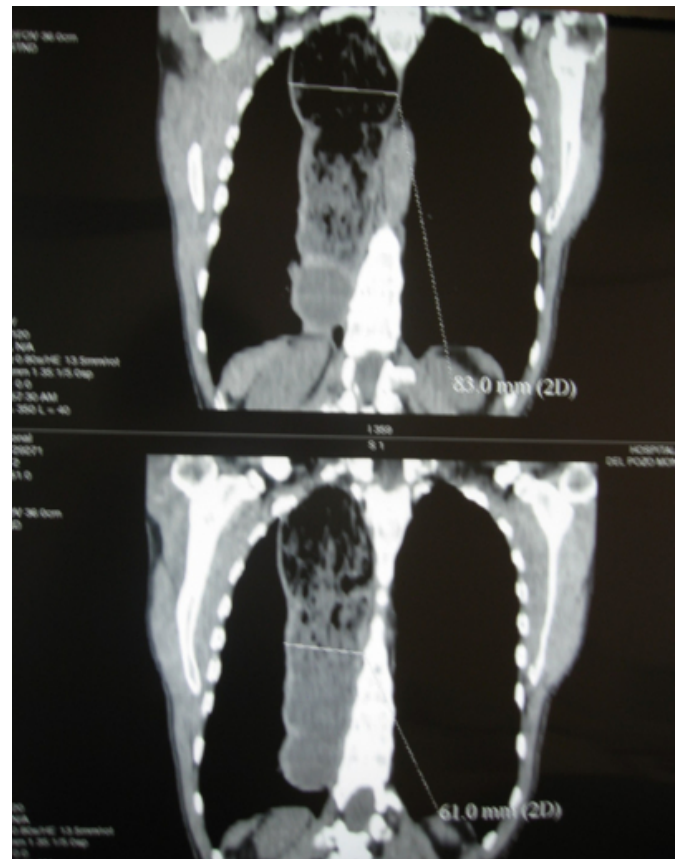


Barium esophagogram identified a residual mega-esophagus with a stenosis at the distal segment. Thoracic CT showed an extensive esophagus compatible with achalasia and also revealed absence of air in the stomach. There were not signs of tumoral infiltration (Fig. 2). Upper GI tract endoscopy confirmed the dilated esophagus for advanced achalasia. At

manometry, the lower esophageal sphincter (LES) resting pressure was 12mmHg. Absence of esophageal body peristalsis was noted in 100% of the swallows. Superior esophageal sphincter pressure was normal and pharyngo-esophageal coordination was appropriate. The 24-hour pH monitoring showed a long acidification, but without any drop of pH below 3.5.

### Figure 2

Figure 2: Thoracic CT showed a residual mega-esophagus with a maximum diameter of 8.3cm and a stenosis at the distal segment of the esophagus.



Laparoscopic ample Heller myotomy and Dor anterior hemifundoplication were performed. Briefly, after the phrenoesophageal ligament was divided and the fat pad excised exposing the anterior gastroesophageal junction, the myotomy was performed by incising the distal 10cm of esophageal musculature. The myotomy was extended 2 cm into the gastric cardia using a cautery hook. Intraoperative endoscopy was performed simultaneously to assess the adequacy of the myotomy, to gauge how far to carry the myotomy into the gastric cardia, and to detect mucosal perforations. We added a Dor anterior hemifundoplication. Operative time was 145 minutes. Blood loss was not relevant. On postoperative day 3, after a normal medical

prescription swallow with gastrografin, a liquid diet was started. The patient was discharged home on the 6<sup>th</sup> postoperative day and returned to normal feeding in 8 weeks. Two months afterwards, the medical prescription swallow confirmed good functioning of the esophageal myotomy. During the first postoperative year, she reported complete resolution of her dysphagia and she did not present reflux with a weight gain of 8 kg.

## DISCUSSION

The advanced form of megaesophagus is characterized by dilatation with an increase in the organ diameter (dolicho-megaesophagus), aperistalsis associated with tertiary contractions, an amplitude of contraction of the esophageal body lower than 20mmHg and by incomplete or absent opening of the lower sphincter. Because of these findings, the resolution of dysphagia is the main objective of treatment, when performing any procedure in the esophagogastric transition<sup>10</sup>.

The treatment of patients with achalasia and megaesophagus has been controversial. Some surgeons recommend a myotomy as initial treatment and reserve esophageal resection as a last resort for the patients with persistent dysphagia and malnutrition<sup>1,9,10,11,12,13,14</sup>, whereas other authors<sup>10,15,16,17,18,19</sup> recommend esophagectomy as first treatment.

The authors who advise esophagectomy as first therapy for those patients with the most advanced stages of achalasia, argue with the following reasons:

Esophagectomy is the only surgical procedure that theoretically would cure advanced megaesophagus.

Multiple prior therapies are associated with a poorer functional outcome. These results are not superior to 70% although the intervention has been carried out by expert surgeons.

Another reason for the resection of this pouch is that emptying of the dilated esophagus continues to be incomplete after surgical procedures (Heller, Thal and Merendino) performed above the esophagus-gastric transition and, consequently, the risk of bronchial aspiration of stasis fluid rich in gram-negative bacteria and fungi continues to be present.

Another problem is the possibility of an association between advanced megaesophagus and neoplasia, which has been

observed in between 3.2%<sup>20</sup> and 18.9%<sup>21</sup> of the cases. Furthermore, Brucher et al<sup>22</sup>, observed that the risk of developing esophageal cancer was about 140 times higher in patients with achalasia than in the general population.

On the contrary to these arguments, Patti et al., affirm that laparoscopic Heller myotomy and Dor funduplication should be considered the primary surgical treatment, regardless of the diameter and shape of the esophagus, because in presence of esophageal dilatation the surgical intervention is not longer and is not more difficult, did not cause more complications or longer hospital stay and gave just as good relief of dysphagia as in patients without esophageal dilatation. The only difficulties during the myotomy resulted from side effects of previous treatment, not from the size of the esophagus per se. Pneumatic dilatation or botulinum toxin injections produced fibrosis at the gastroesophageal junction, which obscured the plane between the mucosa and the circular muscular layer.

In the opinion to Wang et al.<sup>23</sup>, the laparoscopic Heller-Dor operation in patients with end-stage achalasia had the advantages of reduced compromise of the cardiopulmonary function, with less disruption of the supporting structures (phrenoesophageal membrane) of the antireflux mechanism, requiring simpler general anaesthesia, less pain and reduced morbidity, shorter hospitalization and faster convalescence in relation to laparoscopic transhiatal esophagectomy and a cervical esophagogastric anastomosis.

In addition, in patients who have severe cardiac or respiratory disease and in whom esophagectomy is contraindicated due to high surgical risk, the laparoscopic Heller-Dor operation could become a valid option for a conservative treatment.<sup>9</sup>

In our opinion, the patients with this pathology demand careful evaluation before considering radical surgery. Because advanced megaesophagus is a benign incapacitating disease that affects young individuals, its treatment requires a surgical approach with low morbidity and mortality and good and long-lasting outcome. Laparoscopic myotomy satisfies these conditions, whereas the morbidity and mortality rate associated with radical procedures is much higher, as demonstrated in the following study: Analysis of a total of 348 patients submitted to open or laparoscopic transhiatal subtotal esophagectomy and cervical esophagogastric (91%) or coloesophagic anastomosis<sup>10</sup> showed that the mortality was 3.7%, postoperative

complications were 45.5% and postoperative outcomes were acceptable in 96%. Indications for esophagectomy included tortuous megaesophagus in 64%, failure of prior myotomy in 63% and associated reflux stricture in 7%. The most common complications were pulmonary ones (40%)<sup>24</sup>. Postoperative transient dysphonia was observed in 23%<sup>10</sup> and definitive dysphonia in 2,6%<sup>24</sup>. The rate of dysphonia was higher when the laparoscopic transhiatal approach was used compared with open surgery. Presumably, the etiologic factor of dysphonia is the dissection by cervical route of the upper thoracic esophagus and/or removal of a sometimes voluminous surgical specimen through this route, provoking a traumatic inflammatory process in the left recurrent nerve. Incidence of anastomosis leak was 5.9%<sup>25</sup> and 10%<sup>26</sup>. Delayed mediastinal bleeding requiring thoracotomy occurred in 2%<sup>26</sup> and chylothorax in (2%)<sup>26</sup>. Another complication was stenosis of the cervical esophago-gastric anastomosis; nearly 50% required dilatation. The rate reported was 3,6%<sup>27</sup>. Recurrent dysphagia for solid food was observed in 3%<sup>10</sup>, despite the patency of the anastomosis in the radiological and endoscopic study; whereas in the study of Patti et al.,<sup>9</sup> no patient required esophageal resection. One patient experienced dysphagia postoperatively, which resolved after one pneumatic dilatation. Postoperative pH monitoring showed gastroesophageal reflux in one patient. The mortality associated with the procedure was zero. The postoperative course of the group with dilated and sigmoid esophagus was not different from the group with achalasia but without dilated esophagus. In general, the patients were fed on the first postoperative day and left the hospital after approximately two days. Excellent or good results after this operation were obtained in 91% of the patients with dilated esophagus, similar to what has been reported by other studies<sup>28,29,30,31</sup>. We have carried out a conservative procedure in our patient to avoid esophageal resection in a young patient with a successful outcome.

The size or shape of the esophagus could not be implicated in any of the poor results. We believe that the etiology of relapse of the dysphagia in these patients after prior myotomy can be varied: An inadequate myotomy, healing of the myotomy, the development of reflux esophagitis and stricture, obstruction from a concomitant fundoplication, an incorrect diagnosis, the development of carcinoma, or the development of a paraesophageal hernia.

In the opinion of Torquati et al.,<sup>32</sup>, we should study the postoperative LES pressure in patients with failed myotomy.

If the LES pressure is above 20, the patient should undergo redo myotomy. If the pressure is below 20 but above 12, they prefer to do a trial with Botox injection. If the patient responds, he or she could be a good candidate for redo myotomy. Patients with a LES pressure of less than 12 mm Hg are poor candidates for a redo myotomy and therefore for these patients another type of surgical treatment would be suitable.

Was there a relationship between the grade of dilatation of the esophagus, the low LES pressure and the failure of the myotomy? Most of the patients with dilated esophagus have very low LES pressure and usually very high dysphagia score and definitely, those are predictors of a poor outcome after a Heller myotomy. Most of these patients end up in an esophageal resection; however in the opinion of Torquati et al.<sup>32</sup> and Diener et al.<sup>33</sup> we can try to do a Heller myotomy in these patients and see if it works; because the Heller myotomy is a very low morbidity and low mortality operation, and it can be offered in some of these patients before considering an esophagectomy.

We conclude that carefully selected patients with end-stage achalasia benefit from laparoscopic Heller-Dor operation. We believe that safety, reliability, and clinical efficacy of this approach have been documented.

## CORRESPONDENCE TO

Josué Carvajal Balaguera Calle Téllez, 30, Escalera 12, 2<sup>a</sup>, p-3 28007 Madrid, Spain Phone: +34915520026 - +34649078173 Hospital FAX.: + 43915345330 e-mail: josuecarvajal@yahoo.es

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**Author Information**

**J. Carvajal Balaguera**

Surgeon assistant, Service of General and Digestive Surgery, Hospital Central de la Cruz Roja San José y Santa Adela

**J. Camuñas Segovia**

Surgeon assistant, Service of General and Digestive Surgery, Hospital Central de la Cruz Roja San José y Santa Adela

**M. Martín García-Almenta**

Surgeon assistant, Service of General and Digestive Surgery, Hospital Central de la Cruz Roja San José y Santa Adela

**S. Oliart Delgado de Torres**

Surgeon assistant, Service of General and Digestive Surgery, Hospital Central de la Cruz Roja San José y Santa Adela

**L. Albeniz Aguiriano**

Physician assistant, Service of Radiology, Hospital Central de la Cruz Roja San José y Santa Adela

**L. Peña Gamarra**

Surgeon assistant, Service of General and Digestive Surgery, Hospital Central de la Cruz Roja San José y Santa Adela

**C.Ma. Cerquella Hernández**

Chief, Service of General and Digestive Surgery, Hospital Central de la Cruz Roja San José y Santa Adela