

Achalasia at the Crossroads Between Specialties

Florin Bobircă^{1,2}, Horia Doran^{1,2}, Dan Dumitrescu¹, Anca Bobircă^{2,3}, Lidia Belega-Mursoi¹, Maria-Cristina Alexandru³, Cristina Jauca¹, Ioan Ancuța^{2,3}, Cristina Mocanu⁴, Bianca Smarandache⁴, Bogdan Busuioc⁴, Traian Pătrașcu^{1,2}

¹General Surgery Clinic I, "Dr I. Cantacuzino" Clinical Hospital, Bucharest, Romania

²"Carol Davila" University of Medicine and Pharmacy, Bucharest, Romania

³Internal Medicine and Rheumatology Clinic, "Dr I. Cantacuzino" Clinical Hospital, Bucharest, Romania

⁴Endoscopy Department, "Dr I. Cantacuzino" Clinical Hospital, Bucharest, Romania

*Corresponding author:

Ioan Ancuta, MD

5-7 Ion Movila Street, District 2

Zip Code 020475, Bucharest, Romania

E-mail: ioan.ancuta@umfcd.ro

Rezumat

Acalazia cardiacă la limita dintre specialități

Introducere: Acalazia este o afecțiune care se poate trata fie prin intervenție chirurgicală de tipul miotomiei Heller asociată cu un procedeu antireflux, fie prin dilatare pneumatică, alegerea tipului de tratament fiind un subiect larg dezbătut în zilele noastre.

Metoda: Am selectat pacienți cu diagnosticul de acalazie, calculând scorul Eckardt la internare iar aceștia au fost tratați prin dilatare pneumatică, respectiv prin miotomie asociată cu fundoplicatura. S-a analizat succesul terapeutic la finalizarea tratamentului, precum și la controalele ulterioare. Deopotrivă, alte aspecte importante în cadrul studiului au fost calitatea vieții și complicațiile apărute.

Rezultate: Au fost incluși 48 de pacienți, 20 în grupul celor tratați prin dilatare pneumatică și 28 tratați prin intervenție chirurgicală. Rezultatele succesului terapeutic au fost în avantajul chirurgiei, atât după finalizarea tratamentului cât și la controalele de la un an și la 2 ani (96.4% vs 90%, respectiv 92.9% vs 85%). Calitatea vieții a fost mai bună în cazul pacienților cu dilatare pneumatică pe parcursul întregii perioade.

Concluzii: Tratamentul chirurgical al acalaziei prezintă o rată de succes mai mare decât cel prin dilatare pneumatică, însă acesta din urmă se asociază cu o calitate a vieții mai bună.

Cuvinte cheie: acalazie, miotomie, dilatare pneumatică, Eckardt

Received: 22.12.2021

Accepted: 10.02.2022

Abstract

Background: Achalasia is a condition that can be treated either by surgery, such as Heller myotomy associated with an antireflux procedure, or by pneumatic dilation, the choice of type of treatment being a widely debated topic nowadays.

Methods: We selected patients with the diagnosis of achalasia, calculating the Eckardt score on admission and they were treated by pneumatic dilation, respectively by myotomy associated with fundoplication. Therapeutic success at the end of treatment was analyzed, as well as that in subsequent follow-ups. At the same time, other important aspects of the study were quality of life and complications.

Results: Forty-eight patients were included, 20 in the group of those treated by pneumatic dilation, and 28 treated by surgery. The results of the therapeutic success were to the advantage of the surgery, both after the completion of the treatment, and in the follow-ups from one year to 2 years (96.4% vs 90%, respectively 92.9% vs. 85%). The quality of life was better in patients with pneumatic dilation throughout the period.

Conclusions: Surgical treatment of achalasia has a higher success rate than pneumatic dilation, but the latter is associated with a better quality of life.

Key words: achalasia, myotomy, pneumatic dilation, Eckardt

Introduction

Achalasia is the result of the gradual atrophy of the ganglion cells in the myenteric plexus of the esophageal wall, the result being the insufficient relaxation of the lower esophageal sphincter (1,2). In the foreground, as a clinical manifestation present in all patients is dysphagia, along with chest pain and weight loss. Due to common symptoms of gastro-esophageal reflux disease, patients are often treated in the first phase with proton-pump inhibitors (PPIs), a drug that does not help treat achalasia (3). The causes of achalasia are still unknown, but a number of factors are responsible for the onset of the disease, in addition to the atrophy of myenteric nerve plexuses, such as autoimmune factors, infectious factors, environmental factors, Chagas disease. The Eckardt score is currently used, which is based on the 4 most common symptoms: dysphagia, regurgitation, chest pain, and weight loss. This score is used both in diagnosing the disease and in monitoring the response to treatment. Achalasia may be associated with adrenal insufficiency and lack of tears in patients

with triple A syndrome (Allgrove syndrome). Although epidemiological data are scarce, the frequency of achalasia tends to increase, with an annual incidence between 1.2-2.4 cases / 100,000 people and a prevalence of 11-15.6 / 100,000 people, the risk of both genders being equal (1). The management of achalasia can be divided between Gastroenterology and General Surgery, so the variants are pneumatic dilation with a balloon probe and laparoscopic Heller myotomy (LHM) (4-7). The main objective in the management of achalasia is the early diagnosis, in order not to lose the esophageal function or change its anatomy, with the advancement to the late complication called mega-esophagus, which requires esophagectomy (8,9). The choice of treatment should be personalized for each patient, depending on their needs and the clinical situation, in order to accomplish dilation of the lower esophageal sphincter to allow food to pass from the esophagus to the stomach. After the surgical procedure, about 20-25% of patients will develop gastro-esophageal reflux (4). To avoid this, the fundoplication will be performed, simultaneously with the myotomy.

Material and Method

This study took place between January 2018 and November 2021, and patients from the Gastroenterology and General Surgery I departments of the Dr. I. Cantacuzino Clinical Hospital in Bucharest were enrolled. All authors played roles in interpreting and analyzing the results of the study, and in writing it in its final form. Each patient signed an informed consent prior to enrollment in the study.

Patients

The eligibility criteria for admission to the study were aged between 18 and 90 years old, the presence of achalasia with an Eckardt score greater than 3. The positive diagnosis was based on esophageal manometry, barium transit, and upper digestive endoscopy (10-12).

Patients with advanced cardiovascular or pulmonary disease (which could have contraindicated surgery), those who had previous treatment for achalasia or presence of distal esophagus diverticula, were excluded from the study (13,14).

Interventions

Pneumatic dilation

A balloon is positioned at the esogastric junction that initially expands with a pressure of 5 PSI for one minute, and then increases the pressure to 7 PSI for another minute. The protocol used by gastroenterologists involves an initial dilation with a 30 mm balloon, followed at the time of dysphagia with an intermediate balloon of 35 mm, and in case of recurrence of dysphagia, the 40 mm balloon will be used (15). If the Eckardt Score maintains a value above 3, it will be considered a therapeutic failure, and the patient will have a new series of dilations or will be referred to the General Surgery service.

LHM with Dor's Antireflux Procedure

After sectioning the phrenoesophageal ligament, the distal esophagus is mobilized and a

myotomy is performed extending at least 6 cm above the gastroesophageal junction and at least 1 to 1.5 cm on the stomach. Subsequently, the anterior fundoplication at 180 degrees Dor type is practiced as an anti-reflux mechanism (16,17). If the Eckardt score remains above 3, the patient is considered a therapeutic failure.

Aims

The focus of the study is the result of therapeutic success, defined as a decrease in Eckardt score less than or equal to 3 after treatment, at the time of follow-up at one year and two years. The other results considered the patient's weight gain at 6 months after treatment, and special attention was paid to quality of life (through the SF36 questionnaire). Another important aspect to consider is that of complications in both types of treatments (18).

Clinical Evaluation and Follow-up

The evaluation of the patients before performing the treatments included data from the medical history, the complete physical examination, especially on the predominant symptom, performing the hemogram type tests, biochemistry, etc. The quality of life was taken into account in the form of the SF 36 questionnaire, both physically and mentally, which can have scores between 0-100, the quality of life being better the higher the result. From the point of view of the investigations, the patients underwent barium transit, esophageal manometry, respectively upper digestive endoscopy.

Follow-up after treatment consisted of assessing the quality of life through the SF 36 questionnaire immediately after treatment, one year and two years after it, evaluating the results both physically and mentally. Also, for the one-year check-up, an upper digestive endoscopy was performed, which visualized a possible reflux esophagitis and the degree to which it appeared according to the Los Angeles classification.

Statistical Analysis

The software used for data processing was SPSS v19 (Statistical Package for the Social Sciences Inc., Chicago, IL, USA). Categorical variables are presented as absolute numbers, respectively percentages, and were compared using the Person chi-square test. Continuous variables are reported as average, with 95% confidence intervals. Data distribution was calculated using the Kolmogorov-Smirnov and Shapiro-Wilk tests. Because the groups consisted of a small number of patients and the distribution of numerical values is not normal, a non-parametric test was used for comparative analysis, namely Mann-Whitney U. The analyses were performed on data from the whole group, regardless of treatment. All reported P values were two-tailed, and P values less than 0.05 were considered to indicate statistical significance.

Results

Patients and Enrollment

Initially, 54 patients were enrolled in the study, but 6 of them had pseudoachalasia (esogastric junction tumor) and were later

excluded. There remained 48 patients who were treated either by balloon dilation (20 patients) or by Heller myotomy and Dor fundoplication (28 patients). Patients were followed for an average of 24 months, during which regular check-ups were performed.

Clinical Results

The focal point of the results of this study is the primary success of the treatment, which is defined by a value of Eckardt score less than or equal to 3. As can be seen, the differences between the two groups are minimal, with no statistical significance. It should be mentioned that patients treated by surgery have a higher success rate (100% vs 95%).

Therapeutic success was also quantified at regular check-ups from 1 year and 2 years, respectively. The results have a slight advantage for patients in the surgical group, but this difference is not statistically significant ($p=0.227$ at one year and $p=0.348$ at 2 years) (Fig. 1).

It should be considered that there is a difference in the Eckardt score between the two groups (7.29 vs. 7.00), so we can say that the patients treated surgically were in a

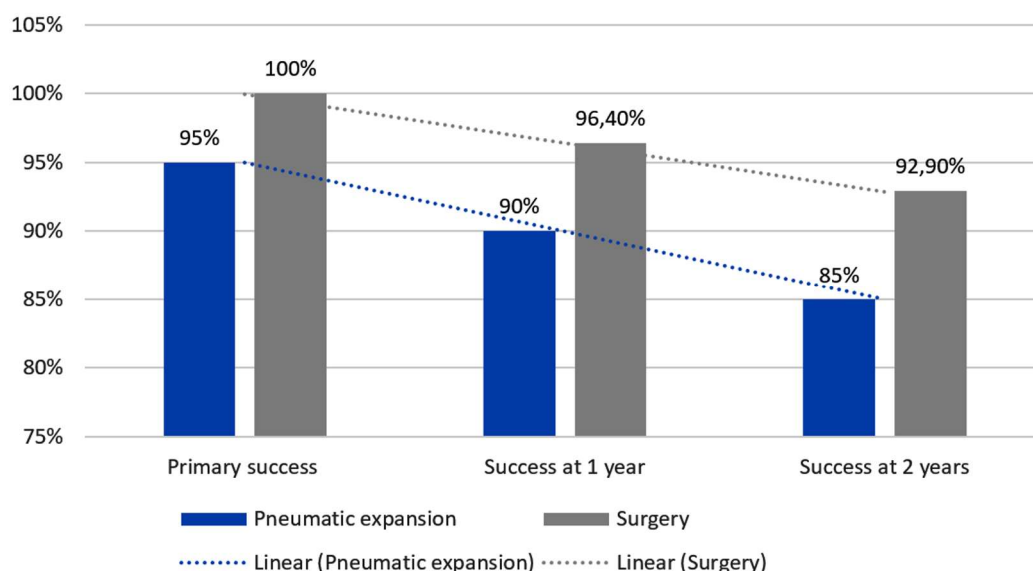


Figure 1. Success rate comparison pneumatic expansion vs. surgery

Table 1. Patient characteristics

Characteristics	Pneumatic Dilation N=20 (41.7%)	Surgery N=28 (58.3%)	P Value
Mean Age	61.50	55.50	.348
CI 95%	48.33-74.67	48.00-63.00	
Sex			.493
Male n (%)	12 (60%)	14 (50%)	
Female n(%)	8 (40%)	14 (50%)	
Eckardt score on admission			.464
Mean	7.00	7.29	
CI 95%	6.17-7.83	6.67-7.90	
Weight on admission			.500
Mean	66.40	64.14	
CI 95%	59.28-73.52	59.13-69.16	
Weight gain			.830
Mean	5.50	5.57	
CI 95%	4.59-6.41	4.90-6.24	

more advanced stage. Patients' weight gain at 6 months after treatment is another important indicator of the effectiveness of therapy. And in this case, there is a small difference favoring the patients treated surgically (5.57 kg vs 5.50 kg), but this fact must be correlated with a lower average weight of the patients in the Gastroenterology department (66.40 kg vs 64.14 kg). Also, considering these values, it should be noted that patients treated with pneumatic dilation have a slightly higher average age compared

to those in the surgical group (61.50 years vs. 55.50 years) (Table 1).

Another important aspect of this study is the quality of life, quantified by the SF-36 questionnaire, which takes into account the physical condition and mental state of patients. Patients in the group treated with pneumatic dilation have an increased quality of life compared to those undergoing surgery, which is valid both for the immediate post-operative / postoperative period, and for follow-ups from 1 year to 2 years (Figs. 2, 3).

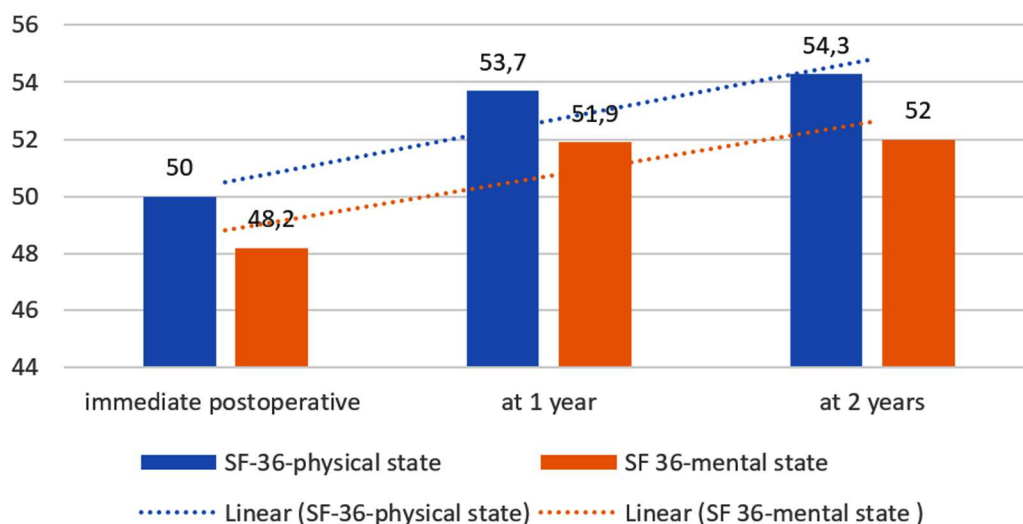


Figure 2. SF-36 score in patients treated by pneumatic dilation

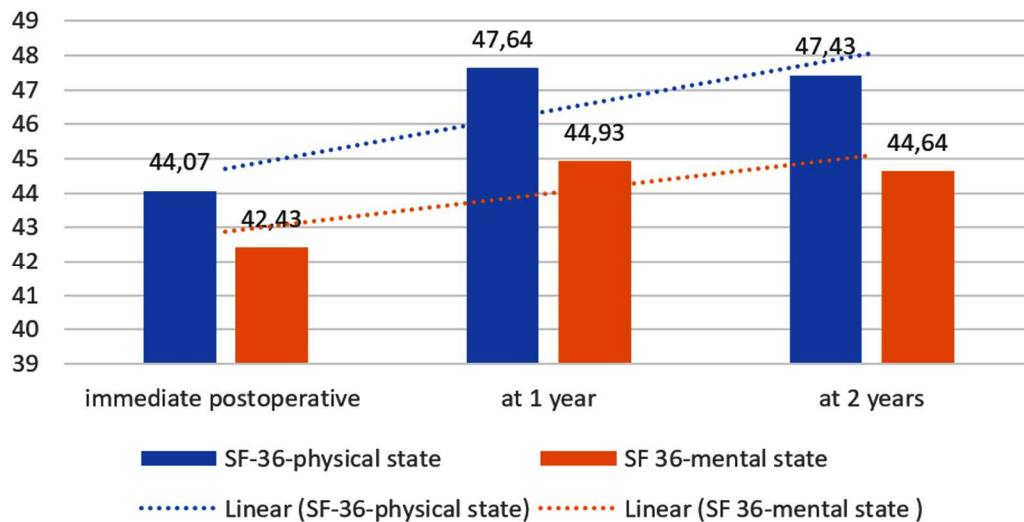


Figure 3. SF-36 score represented in patients with surgery

Complications

In the group of patients with pneumatic dilation, a complication appeared at the time of dilation with the 40 mm balloon, this being addressed by a conservative treatment, consisting of antibiotic treatment and total fasting. On the other hand, the esophageal mucosa was severed during surgery, but this complication was remedied at the same time as the operation (*Table 2*).

At one year after treatment, superior digestive endoscopy was performed in all patients. The occurrence of reflux esophagitis was detected in 21.4% of patients treated surgically (these were divided according to the Los Angeles classification: 10.7% with grade A, 7.1% with grade B, and 3.6% with grade C, respectively), while patients treated by dilation

developed in 25% of cases (15% grade A, 5% grade B, respectively 5% grade C) (*Table 2*).

Discussion

We conducted a study in which we highlighted the benefits of the two treatment options, LHM with Dor fundoplication, respectively pneumatic dilation, the results of primary success, respectively at the follow-up of one year and two years revealing the differences between the two types of treatments. Due to the encouraging results of LHM in various studies (19), laparoscopic surgery is gaining ground in the treatment of the disease. The criterion chosen to highlight the success of the treatment is the Eckardt score less than or equal to 3. Thus, a success rate was recorded after 1 and 2 years of follow-up of

Table 2. Presence of complications and reflux esophagitis - comparison between the two groups

Characteristics	Pneumatic Dilation N=20 (41.7%)	Surgery N=28 (58.3%)	P Value
Complications n (%)	1 (5%)	1 (3.6%)	.807
Reflux Esophagitis n (%)	5 (25%)	6 (21.4%)	.772
Grade A	3 (15%)	3 (10.7%)	.658
Grade B	1 (5%)	2 (7.1%)	.762
Grade C	1 (5%)	1 (3.6%)	.807

96.4% and 92.9% with LHM, respectively, compared to 90% and 85% with pneumatic expansion. The results of our study did not show any significant difference in the success rates of the two treatment options (20). We also found a statistically significant difference in quality of life (both physically and mentally assessed by the SF-36 questionnaire) in favor of pneumatic dilation, which is maintained both immediately after treatment and subsequent follow-ups, similar to other studies (21). Based on our study, it is also necessary to note the differences between the characteristics of the patients, especially on the Eckardt score on admission (7.29 vs. 7.00) suggesting that the surgical group had on average a more advanced disease compared to those admitted to the Gastroenterology ward. It is also necessary to take into account the differences in age, weight on admission, and the distribution by sex between the two groups. Comparing the two treatment options, pneumatic dilation has a lower success rate than surgery, and the use of repeated dilations is a generally accepted method in medical practice, which leads to a satisfactory control of symptoms and a good quality of life. Each expansion series comprises two or three expansion procedures. It should be noted that in the group treated by dilation only one case had complications, which were resolved by conservative treatment. If a 35 mm balloon had been used in the first phase of pneumatic dilation, the complication rate would have been 30-35%, (22) therefore it advocates a protocol of pneumatic distension starting with the 30 mm balloon; this variant brought a remarkable decrease in perforations. In the group of patients treated surgically, only one had complications, namely ruptures of the mucosa, which were immediately remedied during surgery.

Common to both treatments is gastroesophageal reflux, with a percentage of 25% for pneumatic dilation and 21.4% for patients treated with LHM, respectively. These percentages make us think of the following questions: Are screening tests or any anti-

secretory treatments necessary? Of course, all these questions are to reduce the risk of long-term complications such as Barrett's esophagus, stenosis and we do not rule out esophageal carcinoma.

Conclusions

Although the results of the study show a higher average in the case of the Eckardt score for surgically treated patients, the success rate is higher than in the case of dilation surgery and may be considered to be more severe. Pneumatic dilation also means a better quality of life both immediately after treatment and at long-term follow-ups, which is suitable for both lower Eckardt scores and elderly patients who have a contraindication or refuse surgery.

Therefore, we can conclude that laparoscopic Heller myotomy has higher success rates than pneumatic dilation technique, but graded pneumatic dilation ensures a better quality of life in the long run.

From the point of view of the associated complications, it is very important that the dilation protocol starts with the 30 mm balloon (not the 35 mm balloon), in order to have a low rate of esophageal perforation, and if it appears to be small size and can be treated conservatively. In the case of surgical complications, such as the esophageal mucosal sectioning, the problem must be identified quickly and the mucosa sutured.

Conflict of Interest

The authors declare no conflicts of interests.

References

1. Sadowski DC, Ackah F, Jiang B, Svenson LW. Achalasia: incidence, prevalence and survival. A population-based study. *Neurogastroenterol Motil.* 2010;22(9):e256-61.
2. Richter JE, Boeckstaens GE. Management of achalasia: surgery or pneumatic dilation. *Gut.* 2011;60(6):869-76.
3. Richter JE. The diagnosis and misdiagnosis of achalasia: it does not have to be so difficult. *Clin Gastroenterol Hepatol.* 2011;9(12):1010-1.
4. Lynch KL, Pandolfino JE, Howden CW, Kahrilas PJ. Major complications of pneumatic dilation and Heller myotomy for achalasia: single-center experience and systematic review of the literature. *Am J Gastroenterol.* 2012; 107(12):1817-25.

5. Pratap N, Kalapala R, Darisetty S, Joshi N, Ramchandani M, Banerjee R, et al. Achalasia cardia subtyping by high-resolution manometry predicts the therapeutic outcome of pneumatic balloon dilatation. *J Neurogastroenterol Motil.* 2011;17(1):48-53.
6. Rawlings A, Soper NJ, Oelschläger B, Swanstrom L, Matthews BD, Pellegrini C, et al. Laparoscopic Dor versus Toupet fundoplication following Heller myotomy for achalasia: results of a multicenter, prospective, randomized-controlled trial. *Surg Endosc.* 2012;26(1):18-26. Epub 2011 Jul 26.
7. Boeckxstaens GE, Annese V, des Varannes SB, Chaussade S, Costantini M, Cottitta A, et al. Pneumatic dilation versus laparoscopic Heller's myotomy for idiopathic achalasia. *N Engl J Med.* 2011;364(19):1807-16.
8. Pandolfino JE, Gawron AJ. Achalasia: a systematic review. *JAMA.* 2015; 313(18):1841-52.
9. Sodikoff JB, Lo AA, Shetuni BB, Kahrilas PJ, Yang GY, Pandolfino JE. Histopathologic patterns among achalasia subtypes. *Neurogastroenterol Motil.* 2016;28(1):139-45. Epub 2015 Nov 6.
10. Vaezi MF, Pandolfino JE, Yadlapati RH, Greer KB, Kavitt RT. ACG Clinical Guidelines: Diagnosis and Management of Achalasia. *Am J Gastroenterol.* 2020;115(9):1393-1411.
11. Khashab MA, Vela MF, Thosani N, Agrawal D, Buxbaum JL, Abbas Fehmi SM, et al. ASGE guideline on the management of achalasia. *Gastrointest Endosc.* 2020;91(2):213-227.e6. Epub 2019 Dec 13.
12. Niebisch S, Hadzijusufovic E, Mehdorn M, Müller M, Scheuermann U, Lyros O, et al. Achalasia-an unnecessary long way to diagnosis. *Dis Esophagus.* 2017;30(5):1-6.
13. Salvador L, Voltarel G, Savarino E, Capovilla G, Pesenti E, Perazzolo A, et al. The natural history of achalasia: Evidence of a continuum - "The evolutive pattern theory". *Dig Liver Dis* 2018; 50(4):342-347
14. Rohof WO, Salvador R, Annese V, des Varannes SB, Chaussade S, Costantini M, et al. Outcomes of treatment for achalasia depend on manometric subtype. *Gastroenterology.* 2013;144(4):718-25; quiz e13-4. Epub 2012 Dec 28.
15. Oude Nijhuis RAB, Zaninotto G, Roman S, Boeckxstaens GE, Fockens P, Langendam MW, et al. European guidelines on achalasia: United European Gastroenterology and European Society of Neurogastroenterology and Motility recommendations. *United European Gastroenterol J.* 2020;8(1): 13-33.
16. Torres-Villalobos G, Coss-Adame E, Furuzawa-Carballeda J, Romero-Hernandez F, Blancas-Breña B, Torres-Landa S et al. Dor vs Toupet fundoplication after laparoscopic Heller myotomy: long-term randomized controlled trial evaluated by high-resolution manometry. *J Gastrointest Surg.* 2018;22(1):13-22. Epub 2017 Sep 18.
17. Constantinoiu S. Tratat de patologie si chirurgie esofagiana. In: Constantinoiu S, Constantin A, Achim F, Boieriu M (eds). *Acalazia Cardiei.* Bucuresti: Editura Academiei Române; 2017. p. 382-394.
18. Zurita Macías Valadez LC, Pescarus R, Hsieh T, Wasserman L, Apriasz I, Hong D, et al. Laparoscopic limited Heller myotomy without anti-reflux procedure does not induce significant long-term gastroesophageal reflux. *Surg Endosc.* 2015;29(6):1462-8. Epub 2014 Aug 27.
19. Kostic S, Kjellin A, Ruth M, Lönnroth H, Johnsson E, Andersson M, et al. Pneumatic dilatation or laparoscopic cardiomyotomy in the management of newly diagnosed idiopathic achalasia: results of a randomized controlled trial. *World J Surg.* 2007;31(3):470-8.
20. Vela MF, Richter JE, Khandwala F, Blackstone EH, Wachsberger D, Baker ME, et al. The long-term efficacy of pneumatic dilatation and Heller myotomy for the treatment of achalasia. *Clin Gastroenterol Hepatol.* 2006;4(5):580-7.
21. Hulselmans M, Vanuytsel T, Degreef T, Sifrim D, Coosemans W, Lerut T, et al. Long-term outcome of pneumatic dilation in the treatment of achalasia. *Clin Gastroenterol Hepatol.* 2010;8(1):30-5. Epub 2009 Sep 25.
22. Boeckxstaens GE, Annese V, des Varannes SB, Chaussade S, Costantini M, Cottitta A, et al. Pneumatic dilation versus laparoscopic Heller's myotomy for idiopathic achalasia. *N Engl J Med* 2011;364(19):1807-16.