Preoperative work-up

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Despite several advances in the understanding of the pathophysiology of achalasia, treatment remains palliative as the neuronal defect of the disease seems to be irreversible. Currently, the most effective treatment options are graded endoscopic pneumatic dilation and laparoscopic Heller myotomy with partial fundoplication. Although both treatments seem to have similar efficacy in the short-term, the durability of surgical myotomy makes it the favored approach in young patients and in those who want to avoid frequent repeated interventions. Predictors of treatment response have been well defined and should be considered when one therapeutic option is chosen over another. In addition, patient preferences and local are the major are major factors that determine treatment choice. A complete preoperative work up, evaluating patient and disease characteristics, is a key element of a successful treatment.

KEY WORDS: Achalasia, Achalasia score, Preoperative work-up

Introduction

Achalasia is characterized by a loss of peristalsis in the distal esophagus (whose musculature is comprised predominantly of smooth muscle) and a failure of the lower esophageal sphincter (LES) relaxation. Although both of these abnormalities impair esophageal emptying, the symptoms of achalasia (eg, dysphagia and regurgitation) are primarily due to the defect in LES relaxation. The relentless LES contraction in achalasia causes functional obstruction of the esophagus that persists until the hydrostatic pressure of the retained material exceeds the pressure generated by the sphincter muscle.
Since the exact pathogenesis of the disease remains unclear and the underlying defect cannot be reversed, treatment of achalasia remains palliative with the goal to improve clinical symptoms and to restore quality of life and facilitate the passage of the food bolus. Given the low incidence of the disease, only a few randomized controlled trials are available, therefore, cohort studies and expert opinions have been used to develop guidelines for treatment. Currently, pharmacologic treatments offer modest and transient improvements at best, while endoscopic and surgical treatment options remain the best therapeutic choises for management of achalasia.
Our goal is to draw, based on literature data and practice guidelines, a pre-operative work up taking in account patient and disease characteristics.

Preoperative work-up

The best strategy to manage achalasia is to achieve a correct diagnosis and staging of the disease. The symptoms of achalasia often are insidious in onset and gradual in progression. As a result the diagnosis of achalasia is typically delayed 2-3 years from the beginning of symptoms. For many years, the diagnosis of esophageal achalasia was based almost exclusively on the presence

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TABLE I - *Findings and clinical relevance of diagnostic tools for achalasia*

	Esophageal manometry	Barium swallow	Endoscopy	pH monitoring
Findings	Esophageal aperistalsis	Smooth tapering s distal esophagu	Dilated esophagus with barium column	Abnormal reflux due to stasis
	Poorly relaxing LES			
	Hypertensive LES	Dilated esophagus with food retention	Absent peristalsis	
Clinical relevance	Needed for diagnosis	Supports diagnosis	Excludes pseudoachalasia	Limited utility

LES: lower esophageal sphincter

of a hypertensive LES which was considered essential for the diagnosis and therapeutic choice. However, some investigators have shown that the LES pressure can be normal in up to 45% of patients with untreated achalasia[1]. The key finding for the diagnosis is the complete absence of esophageal peristalsis detected by esophageal manometry.

Currently, the diagnosis of achalasia is based on findings from barium swallow and esophageal manometry. Endoscopic evaluation and prolonged ambulatory pH monitoring may also reveal important features of the disease even if they do not allow to achieve the diagnosis (Table I). Other imaging studies, such as MRI, CT or endoscopic ultrasound can be used as adjunct tools to rule out neoplastic or infiltrative processes that can be the cause of pseudoachalasia[2,3].

Barium Swallow

The barium swallow is usually used as the first diagnostic test when achalasia is suspected on clinical grounds. The diagnostic accuracy of barium swallow for achalasia is approximately 95%[4]. Patients with achalasia typically show a smooth tapering of the distal esophagus, known as the "birds beak" or "champagne glass" appearance, with proximal dilation of the esophagus and lack of peristalsis during fluoroscopy. While these findings are specific for achalasia, they may be also absent in the early phase of the disease, consequently, a normal barium esophagogram does not rule out achalasia[5]. The timed barium swallow, a modification of the conventional barium swallow, may be preferred, since it has the additional advantage of quantifying esophageal emptying. In consists of the ingestion of 50-100 ml of barium with plain thoracic radiographs taken at 1, 2 and 5 minutes. Measuring the height of the barium column at this time intervals allows to quantify the degree of esophageal bolus retention. This approach before and at regular intervals after treatment can, therefore, be used as a simple and reproducible tool to assess outcomes after surgical or endoscopic treatment and to detect disease recurrence prior to the development of symptoms[6].

Standard and high resolution esophageal manometry

Although clinical and radiographic findings may strongly suggest the diagnosis of achalasia, a manometric examination is needed for confirmation in virtually all cases. Diagnostic certainty is provided by manometry in over 90% of cases[7]. Standard esophageal manometry and, mainly, the high resolution manometry (HRM), remain the gold standard for the diagnosis of achalasia, since they show three cardinal features of the disease: aperistalsis of the smooth muscle portion of the esophagus, incomplete LES relaxation and increased LES pressure. On the basis of conventional esophageal manometry readings, some authors have discussed manometric variants of achalasia, such as high-amplitude contractions (known as vigorous achalasia), the occasional occurrence of peristaltic waves or almost complete LES relaxation in the presence of aperistalsis[8,9]. Another subclassification system for achalasia that may have therapeutic implications was put forward after the introduction of HRM[10]. Compared with conventional esophageal manometry catheters, which typically have three to eight sensors spaced at intervals of 3 to 5 cm, HRM catheters use a much larger number of sensors spaced at closer intervals (36 sensors at 1 cm intervals). The data collected by these multiple sensors are processed using computer algorithms to display the manometric study in the form of color-coded graphs. When the data are displayed with pressure topography plots, the resulting technology is called high-resolution esophageal pressure topography (HREPT). Findings on HREPT correlate with those of standard manometry, but HREPT also provides enhanced details in the characterization of achalasia, esophageal spasm, nutcracker esophagus, and the morphology of the esophageal gastric junction[11,12].

Endoscopic Evaluation

Endoscopic evaluation is an essential part of the initial work-up, as it could display a dilated esophagus with retained food and some increased resistance at the gastro-esophageal junction, as well as it could detect mucos-

al lesions that can cause pseudoachalasia. However, comments on esophageal peristalsis and LES during endoscopy are not very accurate. Reports of the lack of peristalsis and LES being difficult to pass are neither sensitive nor specific. Retention of undigested food in the esophagus can be regarded as a more specific parameter in diagnosing achalasia, but it occurs only in patients with advanced disease and severe transit impairment. The esophageal mucosa usually appears normal, although inflammation and ulceration may result from irritation caused by retained food or pills. In addition, esophageal stasis predisposes to candida infection that may appear as adherent whitish plaques on the mucosal surface. Candida esophagitis in an immune competent patient should raise the suspicion of esophageal retention.

Prolonged ambulatory ph monitoring

Abnormal reflux is quite rare in untreated patients with achalasia. In these patients, heartburn is usually due to stasis and fermentation of food in the distal esophagus. Prolonged pH monitoring should be performed in patients who have undergone previous treatment, to determine if abnormal reflux is present. In patients with a positive score, it is essential to examine the tracings to distinguish between true reflux and false reflux due to stasis and fermentation of food[13].

Achalasia severity scores

Assessing the disease-specific severity scores in achalasia is essential to predict the outcome of the treatment. Several disease-specific score systems in achalasia have been proposed as reliable, valid and cover a wide effective measurement range.

HRM-based classification

Pandolfino et al. by using HRM findings have provided a new classification of achalasia[10]. They described three distinct variants of achalasia. All the subtypes exhibited incomplete LES relaxation (referred to as esopphagogastric junction relaxation), but they could be distinguished by the swallow-induced pressure response in the body of the esophagus in three types: type I representing classic achalasia with minimal esophageal contractility and low intraesophageal pressure, type II representing absent peristalsis and panesophageal pressure elevations, and type III representing lumen-obliterating esophageal spasm. Responses to all types of achalasia treatment (botulinum toxin injection, pneumatic dilation, surgical myotomy) were best in the type II patients and worst in the type III patients. However, to confirm that high-resolution manometry can be used to predict the response to therapy in achalasia, further studies are needed .

Adams's Stages

Adams classified achalasia in three stages based on the severity of clinical symptoms and the degree of esophageal dilation assessed by radiographic findings[14]. Stage 1, the onset of achalasia, is characterized by episodes of esophageal pain, marked dysphagia, frequent regurgitation after eating small amounts and weight loss. Regarding the barium swallow, only slight morphologic alterations of the esophageal body are obvious in this stage. Stages 2 and 3 express progressively fewer symptoms with advanced radiological dilation of the esophagus. In Stage 2, the compensated stage, the patient may be able to eat normal amounts with no pain, occasional dysphagia and infrequent regurgitation. Weight loss is recovered and few respiratory symptoms might be present. The barium swallow shows marked dilation of the esophagus (30-49 mm). In the final stage of achalasia, Stage 3, with signs of decompensation, patients suffer from substernal oppression and loss of appetite with consecutive gross weight loss. Regurgitation becomes infrequent and chronic pulmonary suppuration is present. At this stage, the barium swallow reveals a sigmoid deformity of the esophagus.

Eckardt Stage

This score is based on a clinical evaluation of symptom frequency and on the degree of weight loss. A score between 0 and 3 is assessed depending on dysphagia, regurgitation, or retrosternal pain (not present, score 0; occasionally, score 1; daily, score 2 and several times a day after each meal, score 3), or on the degree of weight loss (none, score 0; 10 kg, score 3). The highest possible score of the four symptoms is 12 and the lowest score is 0. A symptom score of 0 to 1 corresponds to the clinical Eckardt Stage 0, of 2 to 3 corresponds to Stage I, of 4 to 6 corresponds to Stage II and of >6 corresponds to Stage III[15].

Vantrappen Classification

This clinical score is based on the frequency of dysphagia and retrosternal pain, symptoms that are graded in classes ranging from 1 to 4. The subdivisions are categorized as follows: no symptoms as class 1, short-lasting episodes of dysphagia and retrosternal pain occurring less than once a week as class 2 and dysphagia more than once a week as class 3. assuming that regurgitation and weight loss are only late manifestations of the disease, they are present in class 4 when accompanied by frequent dysphagia[16].

Patient and treatment selection

It is widely accepted that the majority of patients should be treated by the modality that appears to warrant a combination of low initial morbidity, high success rate and good long-term outcome. A complete preoperative evaluation is a key element of a successful treatment. Disease-specific scoring systems developed for achalasia intend not only to objectify the progress of the disease, but also to determine the effect of treatment independently from the patients' or physicians' subjective evaluation. Gockel et al, by evaluating the impact of different disease-specific severity scores on achalasia treatment, found that the Eckardt Score tends toward being the most useful system for clinical practice[17]. Pandolfino et al, by reviewing one thousand clinical HRM studies, showed that type II patients were significantly more likely to respond to any therapy (botulinum toxin injection 71%, pneumatic dilation 91%, Heller myotomy 100%) than type I (56% overall) or type III (29% overall) patients[10]. These findings were recently confirmed in patients with achalasia undergoing surgical treatment with Heller myotomy plus Dor fundoplication[18]. Several authors recently focused on the impact of LES pressure on the outcome of cardiomyotomy: a high pressure (>30 mmHg) consistently emerged as a factor positively associated with a good outcome, suggesting that certain intrinsic features of achalasia might influence the outcome of treatments[19].

However, the indication for myotomy or dilation therapy cannot be set by a specific cut-off point in any system and remains a patient-tailored approach, including patient's age and failed prior options as well as patient's preference and local expertise with these procedures, that should be evaluated in the decision making process. Pneumatic dilatation and surgical myotomy improve dysphagia in the short term in a high percentage of patients, but after 5 to 10 years, only 26 to 49% of patients after pneumatic dilatation versus 33 to 79% after surgical myotomy are free from dysphagia. However, 33 to 38% of patients with recurrent dysphagia manage symptoms without any medical help[20]. According to the only available prospective randomized study, myotomy is more efficient than pneumatic dilation for the treatment of dysphagia at 5-year follow-up[28]. Controversy over the best treatment continues, as both the endoscopic and surgical techniques have evolved. With the advance of surgical techniques, laparoscopic Heller myotomy with partial fundoplication has evolved from a rescue procedure performed when pneumatic dilation fails to a potential primary treatment strategy. Although surgeons and gastroenterologists still argue about the best initial treatment approach, there is now accumulating evidence that a subgroup of patients exists who may be best served by early surgery. Surgical myotomy can also be successfully performed after failed pneumatic dilation or botulinum toxin injection procedures. However, it has been suggested that prior use of botulinum toxin and esophageal dilation, often in sequence, can induce fibrosis of the submucosa, leading to a more-difficult and longer mini-invasive procedure with a higher risk of intraoperative perforation but without a negative impact on long-term outcome[22-25]. These outcomes, however, led to the more frequent choice of surgery as primary treatment. Other predictors of poor surgical outcome have been defined including severe preoperative dysphagia, progressive esophageal enlargement, and low preoperative LES pressures (<30–35 mmHg)[19]. Although advanced age is frequently viewed as an obstacle to surgery, mostly because of the increased frequency of clinically significant comorbidities, a recent study suggests that laparoscopic myotomy can achieve excellent outcomes in patients >60 years of age[26]. At present young age (<40 years), high preoperative LES pressures (>30-35 mmHg) and type II pattern of achalasia on HRM have been confirmed as positive predictors of outcome after surgical miotomy[27-31] (Table II).

Riassunto

Nonostante i progressi fatti nella conoscenza dei processi patofisiologici che sottendono l'acalasia, il trattamen-

TABLE II - *Predictors of surgical outcome in achalasia*

Positive Predictors	Negative Predictors
Age <40 years	Severe preoperative dysphagia
Type II pattern of achalasia on HRM	Low initial LES pressure
Early disease	Prior endoscopic treatments
Postinterventional LES pressure <10 mmHg	Type I or type III patterns of achalasia on HRM
>50% improvement over baseline in barium column height 1 min after initiation of a timed barium swallow	Features of advanced disease (enlarged esophagus)
	Postinterventional LES pressure >10–15 mmHg
	<50% improvement over baseline in barium column height 1 min after initiation of a timed barium swallow

HRM: high resolution manometry; **LES**: lower esophageal sphincter

to di questa patologia rimane palliativo, in quanto il difetto neuronale che ne è alla base sembra essere irreversibile. Attualmente i trattamenti più efficaci risultano essere la dilatazione pneumatica per via endoscopica e la miotomia secondo Heller, associata a fundoplicatio per via laparoscopica. Sebbene entrambi i trattamenti abbiano efficacia simile nel breve termine, l' efficacia nel lungo termine della miotomia chirurgica la rende il trattamento di scelta nei pazienti giovani ed in quelli che vogliono evitare interventi ripetuti. Sono stati ben definiti possibili predittori di risposta al trattamento che devono, pertanto, essere tenuti in considerazione nella scelta del trattamento stesso. Inoltre, la volontà del paziente e le metodologie a disposizione devono essere tenute presenti nella scelta del trattamento. Un completo work up preoperatorio, teso a definire le caratteristiche del paziente e della patologia, è essenziale per il buon esito del trattamento.

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