Achalasia: Dilation, injection or surgery?

Alberto Peracchia MD FACS, Luigi Bonavina MD FACS

A Peracchia, L Bonavina. Achalasia: Dilation, injection or surgery? Can J Gastroenterol 2000;14(5):441-443. Achalasia results from irreversible alterations of the esophageal myenteric plexus. The target of treatment in this setting is to reduce lower esophageal sphincter resistance to passage of the bolus. Definitive treatment of the disease requires pneumatic dilation or Heller myotomy. Although no controlled studies comparing modern endoscopic and surgical techniques are available, laparoscopic surgery is emerging as the initial intervention of choice.

Key Words: *Achalasia; Botulinum toxin; Fundoplication; Lower esophageal sphincter; Myotomy; Pneumatic dilation*

L'achalasie : dilatation, injection ou chirurgie ?

RÉSUMÉ : L'achalasie résulte de modifications irréversibles du plexus myentérique œsophagien. Dans ce contexte, le traitement a pour but de réduire la résistance du sphincter œsophagien inférieur au passage du bol alimentaire. Le traitement définitif de la maladie nécessite une dilatation pneumatique ou une cardiomyotomie extramuqueuse de Heller. Bien qu'il n'existe aucune étude comparative portant sur les nouvelles techniques chirurgicales et endoscopiques, la chirurgie par laparoscopie émerge comme la première intervention d'élection.

Idiopathic achalasia is a motility disorder characterized by incomplete relaxation of the lower esophageal sphincter and impaired peristalsis of the esophageal body. Defective esophageal emptying leads to progressive dilation and tortuosity of the esophagus. Anatomic and physiological studies suggest a dysfunction of the myenteric plexus in these patients (1,2), and an autoimmune pathogenesis has been hypothesized (3). The estimated annual incidence of the disease is 1/100,000 persons.

Dysphagia and regurgitation are the two major symptoms of the disease. Nocturnal regurgitation can lead to aspiration pneumonia and pulmonary abscess. Inability to swallow leads to weight loss in more than half of the patients. The incidence of squamous cell carcinoma of the esophagus is greater in patients with long standing achalasia than in the control population (4,5).

DIAGNOSIS

Diagnosis of achalasia requires the use of endoscopy, barium swallow study and esophageal manometry. Endoscopy must be performed in all patients to rule out other causes of dysphagia. Malignancy-induced secondary achalasia, often referred to as pseudoachalasia, should carefully be excluded. Adenocarcinoma of the cardia is the most common tumour mimicking achalasia. Clinical features suggesting the possibility of a tumour are a short duration of dysphagia, a significant weight loss and an elderly patient. Because adenocarcinoma of the cardia may present as an infiltrating lesion with apparently normal mucosa, a computed tomographic scan and/or endoscopic ultrasonography should be used in selected cases (6,7).

PRINCIPLES OF THERAPY

Current treatment modalities for achalasia are palliative and aim at improving esophageal emptying by reducing lower esophageal sphincter resistance to passage of the bolus. This effect can be achieved endoscopically by means of either pneumatic dilation (8-10) or botulinum toxin injection (11-13), and surgically by extramucosal myotomy (14-16). No firm consensus has been reached yet regarding the choice of the initial treatment. Retrospective studies have shown better results with myotomy performed by an experienced surgeon (17), and in the only prospective randomized trial myotomy gave better long term results compared with pneumatic dilation (18). Uncontrolled studies show that both procedures have equal success rates if skilled operators are

This mini-review was prepared from a presention made at the World Congress of Gastroenterology, September 6 to 11, 1998, Vienna, Austria
Department of Surgery, University of Milan, Ospedale Maggiore Policlinico IRCCS, Milano, Italy
Correspondence and reprints: Dr Alberto Peracchia, Istituto di Chirurgia Generale e Oncologia Chirurgica, Ospedale Maggiore Policlinico, Via F Sforza 35, 20122 Milano, Italy. Telephone +39-02-55035822, fax +39-02-55035810, e-mail bonavina@polic.cilea.it
Received for publication December 2, 1998. Accepted December 9, 1998

available, and, therefore, the patient should be allowed to make his or her own decision (19). In the only controlled trial of botulinum toxin injection versus pneumatic dilation, both procedures were effective at one-year follow-up (13).

Two randomized, double-blind, placebo controlled trials have shown that chronic treatment with calcium-channel blockers, such as nifedipine or verapamil, does not significantly improve symptoms despite a marked decrease of lower esophageal sphincter pressure in up to 50% of the patients (20,21). This form of therapy may be considered for short term management in individuals with relatively mild symptoms or as a temporary measure when more invasive procedures are contraindicated.

DILATION

Rigiflex balloon dilation of the esophagus is effective in more than two-thirds of patients. In up to 50% of the cases additional dilations are required to maintain symptomatic remission (10); the risk of perforation is estimated to be 1% to 6% in expert hands (22). Twenty-four hour esophageal pH monitoring shows gastroesophageal reflux in approximately one-third of the patients after dilation (23).

The effectiveness of dilation does not appear to depend on balloon size, dilation pressure, rapidity of inflation, duration of inflation, number of inflations per session or use of premedication. Patients who do not significantly respond to the first two dilations are unlikely to benefit from subsequent sessions, which may increase the risk of perforation (24).

INJECTION

The results of a double-blind trial of intrasphincteric injection of botulinum toxin compared with placebo showed that 66% of patients remained in remission six months after treatment, and the mean duration of a favourable response was 1.3 years (11). After a median follow-up of 2.4 years, only one-third of the patients were still in remission despite multiple injections. The response rate among patients older than 75 years was 75%, while it was 27% among individuals younger than 50 years (12). The short term safety and effectiveness of the procedure have been confirmed in a French multicentre study (25).

Based on these early results it seems reasonable to consider the use of the toxin in the elderly patients and in those at risk for more invasive procedures (26). It remains to be clarified how long the effects of the injection will last and whether repeated injections will prove to be safe in the long term.

SURGERY

Extramucosal myotomy of the distal esophagus and cardia has been shown to achieve good symptomatic relief in about 95% of patients with idiopathic, previously untreated achalasia. When dissection of the cardia is minimal and an anterior antireflux procedure is added, gastroesophageal reflux is rare (16).

The advent of minimally invasive surgery in the management of benign esophageal disease, by lessening the surgical trauma to the chest and abdominal wall, has made surgery a more attractive option as a primary treatment (27-30). It has been shown that an extramucosal myotomy of the esophagus and cardia combined with a Dor fundoplication can be performed safely and effectively through laparoscopy, with clinical and functional results similar to that obtained with the open approach (31).

The operation is performed through a five-port access. Careful attention to technical details of the procedure is critical for a good surgical outcome. The incision of the lesser omentum is performed taking care to preserve the hepatic branch of the vagus nerve. Dissection is limited to the anterior surface of the esophagus and of the diaphragmatic crura to prevent postoperative reflux by preserving the anatomical relationships of the cardia.

The myotomy is started on the distal esophagus using an L-shaped hook until identification of the submucosal plane, and then continued with the Sugarbaker pericardiotomy scissors. Intraoperative endoscopy helps to identify the submucosal plan, to evaluate the length of the myotomy and to divide residual muscle fibres; additionally, it allows detection of possible mucosal tears that may be safely sutured laparoscopically. The incision is carried out for about 6 cm on the esophagus and 2 cm on the gastric side including the oblique fibres. An incomplete myotomy on the stomach represents the most common reason for a failed operation (32). The cardia is not mobilized except in patients with sigmoid esophagus; in such circumstances, it is preferable to reduce the redundancy in the abdomen and to close the crura posteriorly. The anterior fundic wall is then sutured to both the muscle edges of the myotomy and cranially to the crura. The addition of an anterior antireflux repair sutured to the muscle edges aids in preventing postoperative reflux and healing of the myotomy. After an uneventful Heller myotomy and Dor fundoplication, a gastrographin swallow study is performed on the first postoperative day. The patient is then allowed to drink and to have a soft diet, and is discharged the following day.

EFFECT OF PREVIOUS TREATMENT ON SURGICAL OUTCOME

Patients unsuccessfully treated by endoscopic dilation or intrasphinteric botulinum toxin injection are often referred for surgery. Transient tissue damage in the mucosa-submucosa layer has been documented by high resolution endoscopic ultrasonography (33), but it is unknown whether previous endoscopic treatment may cause histopathological changes leading to periesophageal inflammation, difficult identification of the circular or sling fibres, or difficult dissection of the submucosal plane. Recently, it has been found that patients who previously responded to botulinum toxin show a marked fibrotic reaction at the gastroesophageal junction leading to a higher rate of intraoperative mucosal tears and postoperative dysphagia (34,35).

The increase in technical difficulties encountered at operation after injection therapy can be offset by adequate surgeon's experience; however, these preliminary observations suggest that injection of botulinum toxin should be reserved

to patients who are not candidates for pneumatic dilation or laparoscopic Heller myotomy.

CONCLUSIONS

Pneumatic dilation and Heller myotomy are the two best established therapeutic options in achalasia. Which is the initial approach of choice is still matter of controversy. Although the efficacy of surgery is more predictable, in the absence of a large multicentre, controlled trial, it seems reasonable to state that when expertise in both procedures is available the patient should be clearly informed about the potential risks of each procedure and should make his or her own decision.

The impact of minimally invasive surgery in the treatment of achalasia has been almost as profound as in the treatment of cholelithiasis. The results of laparoscopic Heller myotomy combined with a partial fundoplication show equal efficacy and markedly reduced morbidity compared with the open surgical approach. It is for this reason that, in the near future, laparoscopy could emerge as the initial therapeutic approach of choice in achalasia.

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