Pneumatic Dilatation in Patients with Symptomatic Diffuse Esophageal Spasm and Lower Esophageal Sphincter Dysfunction

E.C. EBERT, MD, A. OUYANG, MD, S.H. WRIGHT, MD, S. COHEN, MD, and W.H. LIPSHUTZ, MD

Nine patients with severe symptoms of diffuse esophageal spasm and lower esophageal sphincter dysfunction who were unresponsive to medical therapy and bougienage dilatation were treated by forceful pneumatic dilatation. Treatment with pneumatic dilatation in eight of the nine patients produced a marked improvement in dysphagia and regurgitation (average follow-up of 37.4 months). Esophageal motility performed up to three years (average 12.4 months) after clinically successful pneumatic dilatation revealed a decrease in lower esophageal sphincter pressure from 34.0 ± 4.0 mm Hg (mean ± standard error) to 19.2 ± 2.7 mm Hg ($P < 0.01$). There were no significant changes in either the percentage of lower esophageal sphincter relaxation or the type of esophageal motor pattern. We conclude from this study that pneumatic dilatation is an effective form of therapy for a select group of patients with severe symptomatic diffuse esophageal spasm with lower esophageal sphincter dysfunction who are unresponsive to conventional medical therapy.

Symptomatic diffuse esophageal spasm is a motor disorder of unknown etiology that is manifested clinically by chest pain, regurgitation, and dysphagia. Radiographic studies of patients with diffuse esophageal spasm demonstrate esophageal configurations that range from normal to bizarre patterns referred to by various authors as cork-screw, rosary bead, or pseudodiverticuli. Esophageal manometry reveals both normal peristalsis as well as high-amplitude simultaneous contractions, spontaneous repetitive contractions, and elevation of the baseline esophageal pressure. Lower esophageal sphincter pressure is normal in most affected patients, but in one third of patients there is impaired lower esophageal sphincter relaxation with or without an increase in pressure (1). The conventional treatment of diffuse esophageal spasm includes reassurance of the patient that his symptoms are not cardiac in origin and the use of drugs that relax smooth muscle such as nitrates and anticholinergics. Medical therapy, however, has not been overwhelmingly successful. Several case reports mention benefit from bougienage or pneumatic dilatation (2–7).

Diffuse esophageal spasm and achalasia may be part of a spectrum of esophageal motility disorders rather than two distinct diseases. There is consider-

Manuscript received August 5, 1982; revised manuscript received October 22, 1982; accepted December 10, 1982.

From the Department of Gastroenterology, The Hospital of the University of Pennsylvania, and Pennsylvania Hospital, Philadelphia, Pennsylvania.

Dr. Ebert is a recipient of the Dr. Hershel Smith Research Fellowship Award.

Address for reprint requests: Dr. William H. Lipshutz, Pennsylvania Hospital, 8 and Spruce Streets, Philadelphia, Pennsylvania 19107.

0163-2116/83/0600-0481$03.00/1

able evidence for this conclusion. A positive mecholyl test may occur in both conditions. A case report demonstrates a transition of the manometric pattern from diffuse esophageal spasm to achalasia (4), and a significant fraction of patients with motility disorders has the clinical and manometric features of both disorders. Vantrappen et al (8) reported that 24% of 156 patients presenting with esophageal motility disorders were intermediate between classic achalasia and diffuse esophageal spasm.

The major factor that contributes to the dysphagia in achalasia is the increase in lower esophageal sphincter pressure and incomplete relaxation upon swallowing. The beneficial effect of pneumatic dilatation in achalasia is related to the decrease in lower esophageal sphincter pressure produced by this procedure (9). The purpose of this study is to report on a select group of patients with diffuse esophageal spasm with lower esophageal sphincter dysfunction refractory to medical therapy, who were treated by forceful pneumatic dilatation.

MATERIALS AND METHODS

All consecutive patients from the years 1975–1981 from the University of Pennsylvania and Pennsylvania Hospital with diffuse esophageal spasm and lower esophageal sphincter dysfunction unresponsive to medical therapy and treated by pneumatic dilatation were studied retrospectively. These patients were evaluated by clinical history, endoscopy, manometry, and x-ray before and at various intervals following pneumatic dilatation. Patients with diffuse esophageal spasm and lower esophageal sphincter dysfunction were diagnosed as follows: (1) nonexertional chest pain that appeared with eating or spontaneously, dysphagia for liquids and solids, or regurgitation of undigested food; (2) the absence of obstructive intrinsic or extrinsic esophageal lesions by x-ray and endoscopy, and (3) esophageal manometry demonstrating normal peristalsis, abnormal contractions (repetitive, simultaneous, or spontaneous contractions and elevation of the baseline esophageal pressure), and lower esophageal sphincter dysfunction (either an elevated baseline pressure or incomplete relaxation). Patients with heartburn, diabetes, or other disorders known to affect esophageal motility were excluded from the study. The group of patients with diffuse esophageal spasm and lower esophageal sphincter dysfunction consisted of six females and three males, ages 29 to 86 (mean 56.8 years old). Patients were symptomatic from six months to six years (mean 2.6 years) before pneumatic dilatation. All had failed an intensive trial of nitrates and anticholinergics, and six underwent at least one bougienage (size 44 to 54 French) before pneumatic dilatation. All responses to bougienage dilatation were transient, lasting only two to three months in four of these six patients, with a maximum of eight months in one.

Manometric studies were performed on a control group, who had no known esophageal disorder and who were age-matched to the patient group. There were seven females and three males in the control group, ages 25 to 81 (mean 48.4 years old).

Water-filled and continuously perfused polyvinyl catheters, 1.4 mm internal diameter, were used to transmit intraluminal pressure to external transducers (Statham P23BB). The rate of infusion was set at 0.8 ml/min which produced a pressure rise of 200 mm Hg/sec using a pneumohydraulic capillary infusion system (Arndorfer pump). The output from each transducer was graphed on a Beckman curvilinear ink-writing recorder. The three recording catheters were sealed at their distal ends and fused together to form a single assembly. Each catheter recorded pressure through a continuously perfused side orifice of 1.2 mm diameter at 5-cm intervals. Lower esophageal sphincter pressure was expressed as the mean midrespiratory sphincter pressure above the simultaneously measured gastric fundal pressure.

Manometric studies were performed on the fasted patient resting quietly in the supine position. The recording assembly was either passed by mouth or transnasally. Belt pneumographs around the chest and over the larynx were used to record respiration and swallowing, respectively. The recording assembly was positioned with all orifices in the stomach. Following a 20-min rest period, the assembly was moved orad at 1-cm intervals through the full length of the esophagus. Lower esophageal sphincter pressure was taken as the average midrespiratory pressure above gastric fundal pressure recorded as the three separate leads traversed the lower esophageal sphincter during a 1-min interval. The percentage of relaxation of the lower esophageal sphincter on swallowing was obtained from each lead by determining the decrease in lower esophageal sphincter pressure from the midrespiratory level to its nadir during a minimum of five swallows.

Pneumatic dilatation was performed with the Browne-McHardy dilator. All patients were sedated with meperidine, and atropine was administered to decrease secretions. The pharynx was anesthetized with local cetocaine spray. The dilator was passed by mouth as the patient lay in a semirecumbent position on the fluoroscopy table. The balloon was checked by fluoroscopy and positioned so that it straddled the diaphragmatic hiatus, and the dilator was inflated to 8–12 lb/in^2 pressure for 15 sec. This procedure resulted in no complications.

The patients were interviewed on at least two different occasions following the pneumatic dilatation by a physician who was unaware of the manometric or clinical results. They were asked to rate the severity and intensity of their symptoms of dysphagia, chest pain and odynophagia, regurgitation, and weight change, before and after dilatation on a scale from 0 (or no symptoms) to 4 (or very severe symptoms). The responses elicited from each patient at the various interviews were compared to determine consistency and reliability. Statistical analysis of the data was computed using the Student's t-test for two means. This study was approved by the human research committee of the University of Pennsylvania and the Pennsylvania Hospital.

RESULTS

Figure 1 shows the symptom scores before and after pneumatic dilatation. The symptoms were reported with complete consistency from one interview to another. Before dilatation, all patients complained of very severe dysphagia. Six had some degree of spontaneous chest pain and odynophagia, and seven noted regurgitation. Six of nine patients lost weight due to their esophageal spasm. There was no correlation between the type or severity of symptoms and the duration of symptoms before pneumatic dilatation.

After dilatation, the patients were interviewed over a mean follow-up period of 37.4 months. All patients noted immediate improvement following the dilatation. In one patient, just short-term relief lasting several months was obtained, whereas the remaining eight patients had sustained improvement in symptoms for the duration of the follow-up period. Of these responders, none classified their symptoms as "very severe" following dilatation, whereas all but one did before the procedure. The improvement was most marked in six of the eight responders, who had minimal symptoms occurring at most once a week following the dilatation. Four of the eight responders gained weight; all had minimal to no odynophagia, chest pain or regurgitation, and six had minimal to no residual dysphagia.

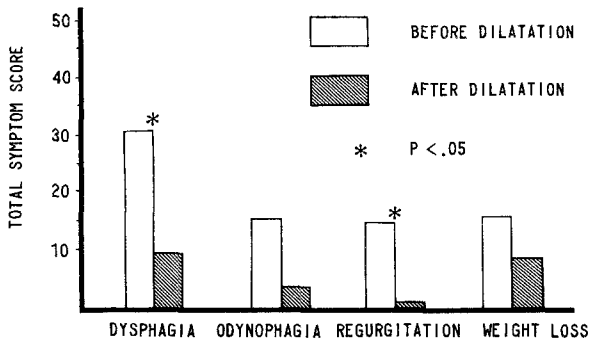

Fig 1. For each symptom, a total symptom score was obtained by adding together the individual symptom scores for the eight responding patients. The individual symptom score was determined as follows: 0 = no symptoms; 1 = mild symptoms occurring ≤1 time per week, or weight loss of 2–9 lb; 2 = moderately severe symptoms occurring >1 time per week, or weight loss of 10–14 lb; 3 = severe symptoms occurring ≧1 time per day, or weight loss of 15–19 lb; and 4 = very severe symptoms occurring >2 times per meal, or weight loss of >20 lb. The mean individual symptoms scores for dysphagia and regurgitation decreased with dilatation ($P < 0.05$). Inclusion of the nonresponder in the calculations did not appreciably alter these findings.

The improvement in total symptom scores as well as the decrease in dysphagia and regurgitation with dilatation were statistically significant ($P < 0.05$).

The nonresponder (L.J.), the youngest patient in this series, had two attempts at pneumatic dilatation. Despite adequate dilatation pressures (9 and 12 psi) and good balloon placement, she had a rebound in symptoms within several months of each procedure. L.J. subsequently underwent a long esophageal myotomy and has been almost totally asymptomatic for the last four months.

Manometric studies were performed on all patients within three months before dilatation (except F.S. whose study was obtained seven months before the procedure). All patients, except F.S., had at least one postdilatation manometry. The mean interval between the procedure and the last manometric study was 14 months. Each tracing was analyzed blindly by two observers to determine: (1) the mean midrespiratory lower esophageal sphincter pressure and the percentage of relaxation with a minimum of five swallows; and (2) the type of esophageal activity occurring either with deglutition or spontaneously. Before pneumatic dilatation, all patients had incomplete lower esophageal sphincter relaxation with swallowing compared to normal volunteers. The lower esophageal sphincter pressure was elevated in seven patients and normal in two.

Figure 2 shows the lower esophageal sphincter pressure in control subjects and in patients with diffuse esophageal spasm who responded to pneumatic dilatation. The mean midrespiratory lower esophageal sphincter pressure in patients with diffuse esophageal spasm before pneumatic dilatation was increased over that in the control group, 34 ± 4.0 mm Hg as compared to 18.0 ± 0.9 mm Hg ($P < 0.005$). Following a clinically successful pneumatic dilatation, resting lower esophageal sphincter pressure was reduced in patients with diffuse esophageal spasm to the level found in the control group, 19.2 ± 2.7 mm Hg ($P > 0.05$). The patient who failed to respond to pneumatic dilatation had a normal lower esophageal sphincter pressure, 16.8 mm Hg, before the procedure, but elevated pressures, 24.6 and 32 mm Hg, following two pneumatic dilatations.

Figure 3 shows the percentage of lower esophageal sphincter relaxation with swallowing in all responding patients and control subjects. These patients had only a partial decrease in lower esophageal sphincter pressure with swallowing, averaging

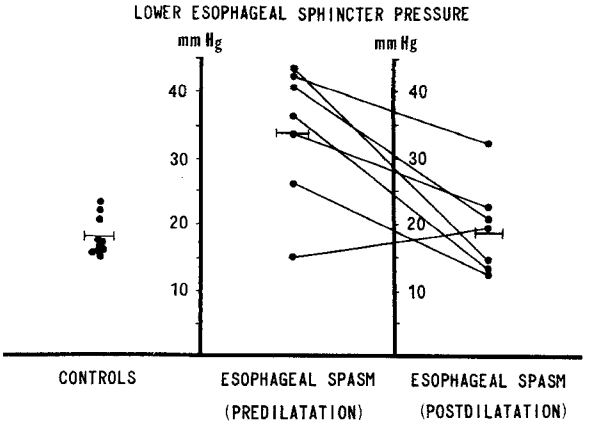

Fig 2. The midrespiratory lower esophageal sphincter pressure above gastric fundal pressure was determined in ten control patients and seven patients with diffuse esophageal spasm and lower esophageal sphincter dysfunction before and following clinically successful pneumatic dilatation (excluding F.S. who refused postdilatation manometry). The mean lower esophageal sphincter pressure as indicated by the horizontal line was 34 ± 4.0 mm Hg (mean ± standard error) for the responding patients before dilatation compared to 18.0 ± 0.9 mm Hg for the control group ($P < 0.005$). Following dilatation, the mean lower esophageal sphincter pressure for the patients (19.2 ± 2.7 mm Hg) did not differ significantly from the control values ($P > 0.05$). If the nonresponder is included in these calculations, the lower esophageal sphincter pressure before dilatation (31.9 ± 4.1 mm Hg) still dropped significantly with dilatation (20.8 ± 2.9 mm Hg) ($P < 0.005$).

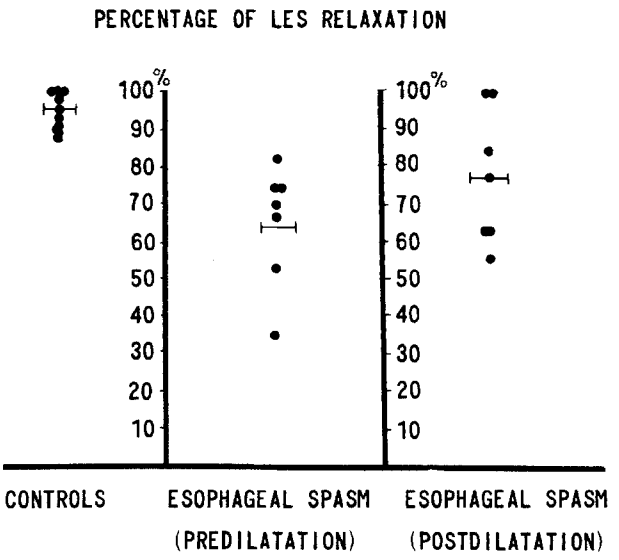

Fig 3. The percentage of lower esophageal sphincter relaxation with swallowing was measured in ten control subjects and seven patients with diffuse esophageal spasm and lower esophageal sphincter dysfunction before and after clinically successful pneumatic dilatation. The mean percentage of lower esophageal sphincter relaxation in the patients, as indicated by the horizontal line, was significantly less than that in the control group, both before and after a clinically successful pneumatic dilatation ($P < 0.05$). These findings were not appreciably altered by including the nonresponder in the calculations.

65.4% ± 6.1% as compared to 94.5% ± 1.6% for the control group ($P < 0.05$). Following pneumatic dilatation, patients with diffuse esophageal spasm continued to show impaired lower esophageal sphincter relaxation with swallowing. The percentage decrease in lower esophageal sphincter pressure with swallowing after dilatation was not significantly different from the percentage of lower esophageal sphincter relaxation before dilatation ($P > 0.05$).

Esophageal activity was categorized as normal peristalsis, simultaneous contractions, spontaneous contractions, absent contractions, double peaking, repetitive contractions, and spasm. The most common type of esophageal abnormality before dilatation was simultaneous contractions. In addition, manometry tracings from eight of the nine patients demonstrated spasm defined as sustained elevation of baseline esophageal pressure lasting at least 15 sec in one or more leads. Spasm was most frequently found in the tracing of the nonresponder. After dilatation, there was no significant change in the type of esophageal activity.

DISCUSSION

The purpose of this study was to determine the response of patients with diffuse esophageal spasm and lower esophageal sphincter dysfunction to pneumatic dilatation. The conclusions of this study are as follows. (1) Pneumatic dilatation in this select group of patients produced clinical improvement in 89% of patients followed for an average of 37.4 months. (2) The improvement in esophageal symptoms is associated with a sustained decrease in lower esophageal sphincter pressure in most of the responders. (3) There is no change in the percentage of lower esophageal sphincter relaxation with swallowing or in the pattern of esophageal motility following clinically successful pneumatic dilatation. The nonresponder was the youngest in the series, and her manometric tracings were distinguished by the frequency of spasm and for a significant rise rather than fall in the lower esophageal sphincter pressure following dilatation.

Several reports in the literature claim benefit from forceful dilatation in patients with diffuse esophageal spasm. Rider et al (5, 6) treated nine

patients with diffuse esophageal spasm with pneumatic dilatation, but multiple procedures were sometimes required. Craddock et al (10) initially treated 12 patients with a Negus hydrostatic dilator, with temporary improvement. Five subsequently underwent a myotomy. These early reports included in their series patients with a variety of esophageal disorders, such as esophagitis (6), stricture (10), and a repaired paraesophageal hernia (10). In addition, no measurements of the lower esophageal sphincter pressure were made due to the lmitations of the manometry equipment at the time. More recently, forceful dilatation has been mentioned in case histories of patients with diffuse esophageal spasm with lower esophageal sphincter dysfunction, with some improvement noted (2–4, 7). Vantrappen and Hellemans (11) mentioned their experience with pneumatic dilatation in 14 patients and found that symptoms completely disappeared in only three. Details, however, are unavailable. Both Vantrappen et al (8) and Bennett et al (7) found an improvement in the motility pattern with pneumatic dilatation.

This study represents the first detailed description of a uniform group of patients with diffuse esophageal spasm and lower esophageal sphincter dysfunction who were treated by pneumatic dilatation. The results suggest that pneumatic dilatation should be tried in such patients who have failed medical management.

The conclusions from this study, however, are limited by several factors. First, the retrospective approach has inherent biases, which were hopefully minimized by the blinded analyses of the manometry tracings and by patients interviews being run by a physician unaware of clinical and manometric results. Second, the trial was conducted before the availability of calcium-channel blockers, which may have an important role in the therapy of diffuse esophageal spasm (12). Finally, since pneumatic dilatation has potentially serious complications, only those patients with disabling symptoms from diffuse esophageal spasm and lower esophageal sphincter dysfunction unresponsive to other forms of therapy underwent this procedure. The value of pneumatic dilatation for other patients with diffuse esophageal spasm is unknown.

The only symptoms that improved significantly with dilatation in this study group were dysphagia and regurgitation. As in achalasia, this may be due to the decrease in the high lower esophageal sphincter pressure accomplished by the procedure resulting in improved esophageal emptying. The abatement of odynophagia and chest pain occurring in some patients may be due to an improvement in the esophageal motility pattern with pneumatic dilatation noted by some authors (7, 8) but not verified here.

Diffuse esophageal spasm with lower esophageal sphincter dysfunction, then, may represent a motility disorder intermediate between diffuse esophageal spasm and achalasia, and so may be amenable to the therapy used in the two established diseases. This study provides evidence that pneumatic dilatation may have a role in the treatment of this motility disorder.

ACKNOWLEDGMENT

The authors are grateful to Mrs. Barbara J. Scott for excellent secretarial assistance.

REFERENCES

1. DiMarino AJ Jr, Cohen S: Characteristics of lower esophageal sphincter function in symptomatic diffuse esophageal spasm. Gastroenterology 66(1):1–6, 1974
2. Eckardt V, Weigand H: Supersensitivity to pentagastrin in diffuse oesophageal spasm. Gut 15:706–709, 1974
3. Orlando RC, Bozymski EM: Clinical and manometric effects of nitroglycerin in diffuse esophageal spasm. N Engl J Med 289(1):23–25, 1973
4. Kramer P, Harris LD, Donaldson RM Jr: Transition from symptomatic diffuse spasm to cardiospasm. Gut 8:115–119, 1967
5. Rider JA, Moeller HC, Puletti EJ: Diffuse esophageal spasm. Am J Gastroenterol 44:97–106, 1965
6. Rider JA, Moeller HC, Puletti EJ, Desai DC: Diagnosis and treatment of diffuse esophageal spasm. Arch Surg 99:435–440, 1969
7. Bennett JR, Donner MW, Hendrix TR: Diffuse esophageal spasm: Return to normality. Hopkins Med J 126:217–224, 1970
8. Vantrappen G, Janssens J, Hellemans J, Coremans G: Achalasia, diffuse esophageal spasm and related motility disorders. Gastroenterology 76:450–457, 1979
9. Cohen S, Lipshutz W: Lower esophageal sphincter dysfunction in achalasia. Gastroenterology 61(6):814–820, 1971
10. Craddock DR, Logan A, Walbaum PR: Diffuse oesophageal spasm. Thorax 21:511–517, 1966
11. Vantrappen G, Hellemans J: Diffuse muscle spasm of the oesophagus and the hypertensive lower oesophageal sphincter. Clin Gastroenterol 5(1):59–72, 1976
12. Silverstein BD, Kramer CM, Pope CE II: Treatment of esophageal motor disorders with a calcium-blocker, diltiazem. Gastroenterology 82(5):1181, 1982 (abstract)