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An Evaluation of the Modified Heller Operation in the Treatment of Achalasia of the Esophagus *

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THE LARGE number of surgical procedures that have been advocated in the treatment of achalasia is a reflection of how little is known about the true nature of the disease. In a superficial review of the literature, we have been able to find 18 different operations that have been proposed at one time or onother. One of the earliest recorded surgical attempts consisting of a resection of the redundant esophageal wall (Jaffe 1897). Mikulicz performed the first direct procedure on the narrowed segment, a finger dilatation done through a gastrotomy incision (1904). During the period 1910 to 1916, the possibilities of vagectomy, phrenicectomy, and esophageal plication were explored, esophageal resection was tried, and a number of plastic procedures (Wendel, Heyrovsky, Gröndahl) were devised.

In 1913, Hurst defined the clinical picture and coined the word achalasia. In the same year, Heller described his operation of esophagocardiomyotomy, in which anterior and posterior longitudinal incisions were made through the muscular layers down to the mucosa. This procedure did not become immediately popular, however, and in the interval between the World Wars, esophagogastric resection, or cardioplasty of the Heyrovsky-Gröndahl type were the most commonly employed forms of surgical treatment. Both of these operations gave adequate relief of obstruction, but later follow ups revealed a high incidence of secondary peptic esophagitis.[1, 3] In 1948, Maingot presented a series of 39 patients treated by a modified (single myotomy) Heller operation, with good early results in 37.[6] Other favorable reports began to appear, and it was soon agreed that the Heller operation, performed either transabdominally or transthoracically, gave results superior to other forms of surgical therapy.[4, 8] Recent reports suggest, however, that the Heller operation alone may not give a permanently good result. Hawthorne, Frobese and Nemir, reported an incidence of 21 per cent poor results in a series of 35 cases, and now advocate the routine addition of a pyloroplasty or esophagomyotomy in an effort to control postoperative esophagitis.[5a] Wangansteen expressed frank disappointment with the standard Heller procedure, and described a balloon-expanded myotomy combined with an outlet pyloroplasty of the Heinecke-

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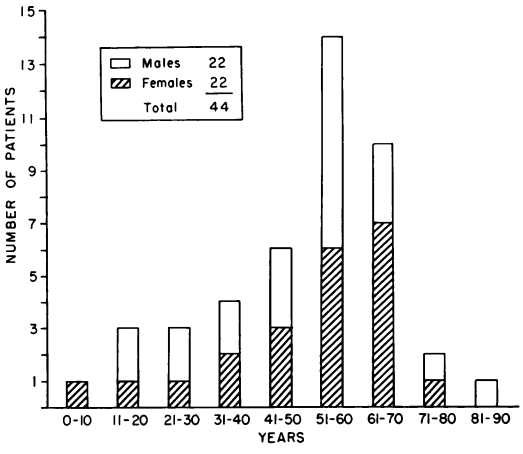

FIG. 1. Achalasia patients. Age and sex distribution 1948 to 1957.

Mikulicz type.[11] Sweet advises the Heller operation only for concentric esophageal enlargements, preferring a cardioplasty for the sigmoid type of dilatation.[9]

Since 1948, all anatomic types of medically refractory achalasia seen at Barnes Hospital have been treated in the same way, by a modified Heller esophagomyotomy performed via the thoracic approach (left posterolateral thoracotomy). A critical review of these cases has been done to obtain information on the long term results, and if possible to direct some light on the reasons for the difference of opinion concerning the value of this procedure.

Case Material

A total of 56 cases have been operated upon from 1948 to the present. For purposes of follow up, however, only those cases seen from 1948 through 1957 are considered. There are thus 44 cases that have been followed a maximum of ten years and a minimum of one year. The distribution of these cases again demonstrate the vagaries of this disease, in that it can afflict patients of either sex, all age and racial groups, and with widely varying economic status. There were 22 men and 22 women (Fig. 1). The oldest patient was 86, and the youngest seven years of age, and within the last year we have operated on a typical case at nine months of age. The sex distribution is about the same for each decade of life. The median age at the time of operation was 55 years. Three of the patients were Negro. Eleven cases, one quarter of the group, were service cases.

Figure 2 shows the symptoms encountered. The characteristic triad of dysphagia, acid-free regurgitation, and weight loss were present in almost all cases. Dysphagia was the presenting symptom in the majority of cases, and in almost all instances the fluctuating nature was evident, with increasing difficulty occurring during periods of nervousness or stress. Regurgitation was also present in every case, and varied in degree from soiling of the pillow at night, to evacuation of the accumulated food of several meals. Weight loss was present in all but four cases, the average loss being 22 pounds. The men as a group lost an average of ten pounds more than the women.

Pain as a major symptom was found in seven cases (21%). It was usually described as a retrosternal cramping, aching or burning, and was frequently relieved by regurgitation or antispasmotics. There

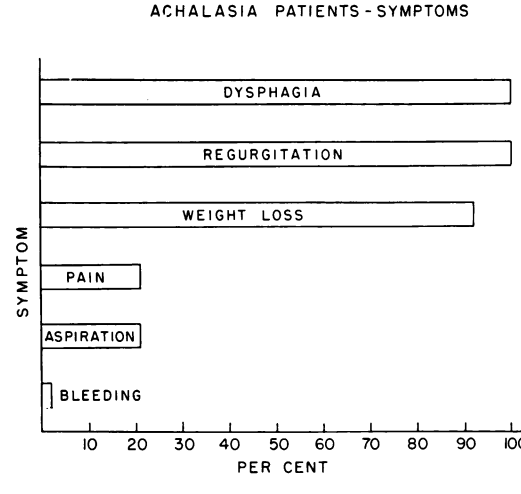

FIG. 2. Achalasia patients. Symptoms.

was no correlation between pain and a specific age, sex, or type of esophageal dilatation, nor did its presence affect adversely the outcome of operation. Seven cases also gave a definite history of aspirative episodes. This complication was not limited to the older age group. Clinical pneumonitis was present in a 13-year-old girl, and a 29-year-old man was seriously ill with pneumonia on two occasions before the underlying cause was determined. Only one patient had recognized bleeding. This was a 38-year-old woman who had symptoms for 15 years, and then developed a massive hematemesis late in the third trimester of a pregnancy. Spontaneous delivery of a normal child occurred during the resuscitation efforts, and right away the bleeding stopped. A successful Heller operation was carried out ten day later.

The duration of symptoms varied greatly (Fig. 3). Two patients had operation after only two months of brief but severe difficulty, while one man smoldered along with trouble for 30 years and finally had operation at age 71 after he almost lost his life from aspirative episodes. It is interesting that the oldest patient in the series, who was operated upon at 86 years of age, had the onset of difficulty only two years before. The men came to operation much sooner after onset of symptoms than the women, the median duration of symptoms for the men being two years and for the women seven years. This fact, along with the significantly greater weight loss in men, suggests in this series that the disease was more severe and more rapidly progressive in men than in women.

X-Ray Features. The esophagrams fell into two general groups of esophageal enlargement, the concentric and the sigmoid varieties. There were 33 cases of the concentric type, ten cases of the sigmoid type (Fig. 4). We do not feel these represent separate and distinct types of achalasia, as suggested by Sweet. We could detect no essential difference in the clinical picture

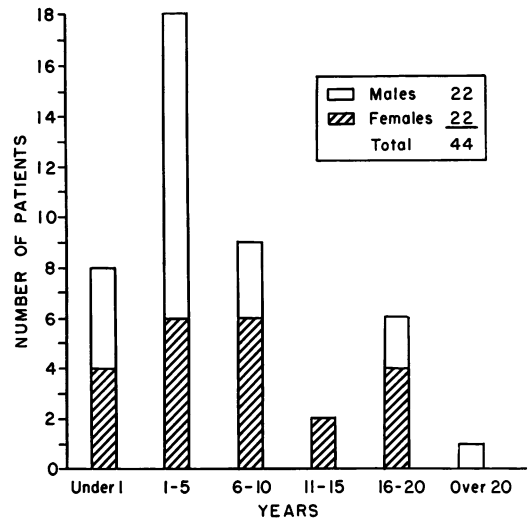

FIG. 3. Achalasia patients. Duration of symptoms 1948 to 1957.

other than the suggestion that elongation and tortuosity are sequels to long-standing disease. The length of symptoms for the sigmoid group averaged 11 years, for the concentric group five and one-half years. Aspiration did not occur with any greater frequency in one group than the other.

Case Report

Case 1. P. N.: A 55-year-old white, single, clerk, was first seen in 1954, with a 6-year history of progressive dysphagia and retrosternal discomfort. Her symptoms were thought due to achalasia, but esophagrams and esophagoscopy were normal. She was admitted one year later with continued dysphagia, some regurgitation, and with increase in the retrosternal pain and burning, not relieved by food or alkali. There was no weight loss. Esophagrams revealed disorganized contractions of the esophagus, with tertiary waves throughout (Fig. 5). Esophagoscopy was again normal. Discharge diagnosis was anxiety neurosis. She continued to have the same complaints for two more years, despite the use of antispasmotics, tranquilizing drugs and sedatives. A third esophagoscopy showed no evidence of stricture, spasm or esophagitis. Despite this a Heller operation was performed on April 4, 1947 (THB), at which time a thickened, hypertrophied muscular coat was found which involved the lower half of the esophagus and required a myotomy incision from the diaphragm to the

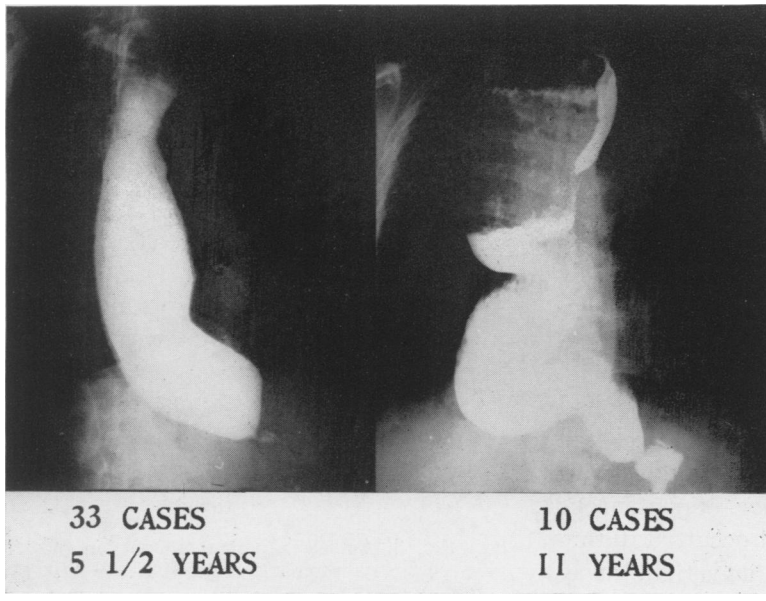

FIG. 4. Examples of the two radiographic types of dilatation. Symptoms were present an average of 5½ years in the concentric type, 11 years in the sigmoid type.

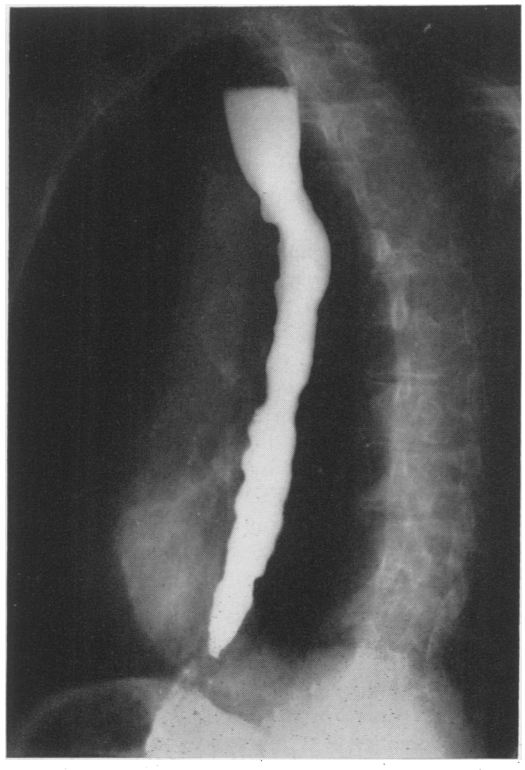

FIG. 5. Esophagram in case of a typical achalasia, resembling giant muscular hypertrophy of the esophagus.

aortic arch. The patient since has been completely free of difficulty, and is grateful that the operation was finally performed. Barium studies show a normal esophagus with normal passage into the stomach. This case is similar to the cases of giant muscular hypertrophy described by Sweet.[9] The condition is rare, and the cause is unknown. Because of similar pathological changes in the involved muscles, it resembles the entity of persistent spasm of the circopharyngeus muscle. We feel it may be a rare variant of achalasia. Certainly the Heller operation gives relief.

Preoperative Esophagoscopy: Diagnostic esophagoscopy was done in all but three of the cases and in each instance showed the usual retention in the upper esophagus with a dilatable spasm at the cardia, with the single exception of the case (PN) mentioned above. Nineteen patients (44%) had one to eight preoperative endoscopic dilatations with only temporary relief. No organic strictures were found, although the degree of dilatation that could be obtained with bougienage was variable. Five patients had some redness and edema, probably retention inflammation rather than reflux esophagitis,

since gastric reflux was not seen, and none of the cases had the marked changes associated with acid-pepsin digestion.

Operation: All of the cases in this series were treated through a transthoracic approach, so that there is no opportunity to compare results of transthoracic versus the transabdominal operation. There have been changes in technics which seem to affect the results. Two important modifications have been, (1) the avoidance of any trauma to the esophageal hiatus, and (2) the proper placement of the myotomy incision. From 1948 to 1952, it was frequently the practice to open the diaphragm into the esophageal hiatus and cut the surrounding ligaments. This was done principally to allow the myotomy incision to be carried down several inches onto the stomach wall. Since 1952, with the realization of the important role these supporting ligaments have in preserving the angle of entrance of the esophagus into the stomach, care has been taken not to disturb this area. The myotomy incision is carried down only as far as the true junction of esophagus and stomach and no further. We can see no reason for the gastric extension, since it does not aid in relieving obstruction. The upper limb of the myotomy, however, is extended upward much higher than in the early cases, so that the muscle layers are opened 6 to 8 cm. above the uppermost margin of the narrowed segment. This change was based on a feeling that some of the early cases had a poor result because of inadequate relief of obstruction in this area. All transverse fibers and vessels must be divided so that the mucosa balloons out fully all along the incision. This must be done carefully and completely. If the mucosa is opened by mistake, it can be closed with a few silk sutures without fear of complication. There has been no postoperative morbidity from this cause in the series.

It is our distinct impression that these modifications in technic are more easily carried out through the chest than at laparotomy.

We have believed that the diaphragm should be opened as a routine part of the procedure (through an incision placed well away from the esophageal hiatus), to rule out a co-existing hiatal hernia, or an unsuspected carcinoma of the stomach. In this way three small hernias were found which could not be demonstrated by either barium swallow or esophagoscopy. Repair of such a hiatal hernia is necessary for a successful outcome. In three other cases not included in this series, adenocarcinoma

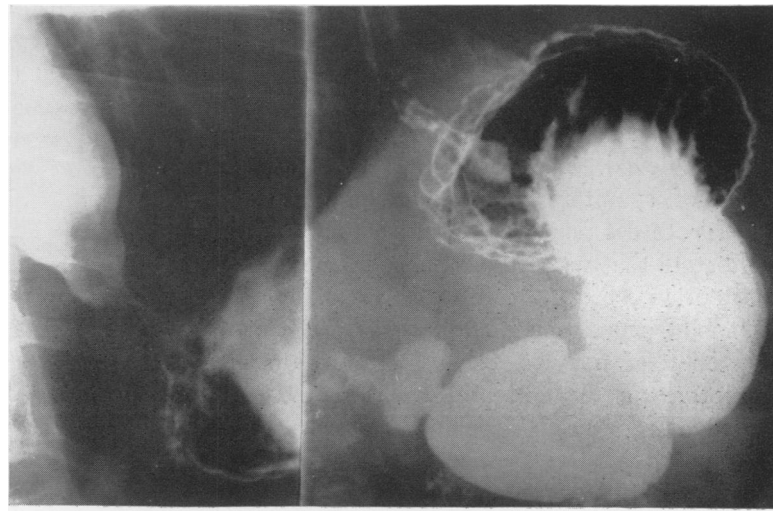

FIG. 6. Esophagram in a 42-year-old man with history and esophagoscopic findings typical of achalasia. At operation a small adenocarcinoma of the cardia was found.

TABLE 1. *Esophagomyotomy—Technical Considerations*

1. Correct diagnosis
2. Thoracic approach
3. Preservation of esophageal hiatus
4. Esophagomyotomy incision
   a. Generous superior limb
   b. Limited inferior limb
   c. Adequate depth
5. Repair of co-existing hiatal hernia

of the junction area of the stomach was found which had given rise to symptoms, esophagoscopic, and x-ray changes indistinguishable from achalasia (Fig. 6). Table 1 summarizes the technical concepts which to our way of thinking are the most important contributors to the success of esophagomyotomy in the treatment of achalasia.

Results

There were no operative deaths and no major complications encountered in the 44 cases. Minor complications were uncommon and included pneumothorax and superficial wound infections. All of the patients have been followed from one to ten years since operation and none lost to follow up. The majority have been seen at intervals since operation. There have been 10 late deaths. Of these, eight were due to unrelated causes, primarily cardiac, and two directly attributable to the disease. Table 2 shows the results. In 36 patients, or 81 per cent, a satisfactory result has been achieved. These patients were relieved of symptoms, gained weight, and are able to eat normally. About a third of them state that when they are under nervous tension, or eat too fast, they may occasionally experience a slight sensation of delayed food passage, otherwise they are asymptomatic. In no case has this "esophageal awareness" developed into true dysphagia with the passage of time. Similarly, the patients who achieved early complete relief of symptoms have remained asymptomatic during the period of follow up. Return of the dilated esophagus to normal size was not considered necessary for a successful result. Less than a third of satisfactory cases have postoperative esophagrams which could be called normal, although most have shown significant reduction in size.

In eight cases, or 19 per cent, the result was not satisfactory, either because of inadequate relief of symptoms (4 cases) or because of the development of reflux esophagitis (4 cases). Inadequate operation was apparent in each instance within a few months, with reappearance of dysphagia and regurgitation of acid-free esophageal contents. Two of the four cases have had periodic esophageal dilatations, but none have had re-operation. One death occurred in this group, a 62-year-old woman who had persistent symptoms for one and one-half years after operation and finally died of aspiration. The other four unsatisfactory results due to reflux esophagitis were also apparent quite soon after operation, with epigastric and esophageal burning severe enough to require hospitalization and investigation occurring one, three, four, and eight months after surgery. It is significant that three of these four cases had an associated hiatus hernia. All three occurred in women. In one case, a 42-year-old woman, a hernia was found and repaired at the time of myotomy, but the repair was inadequate, as she developed severe reflux esophagitis and a recurrent

TABLE 2. *Esophagomyotomy Results—44 Cases 1948 to 1957*

		No.	%
Satisfactory		36	81
Unsatisfactory		8	19
Inadequate	4*		
Reflux esophagitis	4*		

* One death.

hernia was demonstrated. She then had a bilateral vagectomy, hernia repair and Finney pyloroplasty, and a year later required a subtotal gastrectomy. It has now been seven years since her original operation, and she is still having difficulty. In the other two patients no hernia was recognized at the time of myotomy, but were demonstrated when the patients were seen with symptoms of reflux esophagitis several months later. It is not certain whether these hernias were missed at operation, or whether the surgical manipulations about the cardio-esophageal junction predisposed to secondary herniation. One of these patients, a 67-year-old woman, had four separate episodes of moderately severe bleeding over a two-year period following esophagomyotomy, with progressive symptoms of retrosternal pain and burning. Esophagoscopy each time showed a more severe esophagitis, despite vigorous medical therapy. A bilateral vagectomy and Finney pyloroplasty has been done recently. The third patient, a 56-year-old woman seen very early in the series, was improperly managed with low esophagogastric resection to control her reflux esophagitis. She developed even more trouble, only partially helped by a supra-aortic reanastomosis, and finally expired following a gastro-enterostomy. Only one man has been seen with reflux esophagitis following esophagomyotomy. He gave a history of duodenal ulcer which antedated his achalasia symptoms by many years. His obstructive difficulties were completely relieved by operation but he has symptoms of mild esophagitis, and inflammatory changes of moderate degree present on esophagoscopy. He is, however, well controlled on an anti-ulcer regimen, and recent studies nine years after operation show no evidence of progression or stenosis.

Although the over all results for the ten-year span can be considered good, 81 per cent of 44 cases with a satisfactory outcome, the poor results are heavily concentrated in the early years of this experience (Table 3). From 1948 to 1952, a total of 14 cases were done, with only 8 (56%) good results, and 6 (43%) poor results, three because of an inadequate Heller and three because of reflux. In contrast to this earlier period, the later experience from 1953 to October 1957, there were more than twice as many cases done—30 in all—with 28 (93%) satisfactory results and only two poor results. This decline in the number of unsatisfactory results parallels the changes in operative technics mentioned above (Table 1).

TABLE 3. *Esophagomyotomy Results—44 Cases*

No. Cases	1948–52 14	1953–57 30
Satisfactory	8 (56%)	28 (93%)
Unsatisfactory	6 (43%)	2 (7%)
Inadequate	3	1
Reflux esophagitis	3	1

Discussion

Several points of interest are brought to light by this study. The over all results agree closely with other published reports on esophagomyotomy for achalasia, that is 80 to 85 per cent good results, a better record than found with any other form of surgical therapy. It seems to us that there are additional reasons for advocating more widespread use of the procedure in earlier phases of the disease. The operation can be applied to patients of all age groups from infancy to old age with no mortality and negligible morbidity. It gives relief equally well in all anatomic types of dilatation. Once a satisfactory result has been achieved, symptoms do not recur. This has been true in patients followed up to ten years. In this series a poor result was apparent in every instance quite soon after operation. No patient classed as a good result a year after operation has had difficulty later.

Another strong argument for a surgical attitude is the missed diagnosis of cancer. Our experience includes three adenocarcinomas of the stomach which arose at the cardio-esophageal junction and extended submucosally a short distance up and around the esophagus. A tumor in this area can create the picture of achalasia in all clinical, roentgenographic and esophagoscopic aspects. Belsey [2] mentions the frequency with which epidermoid esophageal cancer is seen in achalasia, but we had no such cases in this series.

With the establishment of these general facts about the Heller procedure, attention must be directed to a consideration of the one patient in five who does not obtain a good result from the operation. These failures align themselves into two major groups, either inadequate relief of obstruction, or reflux esophagitis. Various theories have been postulated to account for the development of reflux esophagitis, but the fact is that probably any patient who has the mechanical set-up for reflux of gastric juice will develop difficulty. It is not necessary that there be a prior history of duodenal ulcer, or that an increase in tonus of the pylorus be present. The present trend to combine myotomy routinely with pyloroplasty or some other outlet procedure we believe is a poor solution to the problem, for the additional procedure will impose an inevitable increase in morbidity and possibly mortality on the four of five patients who would have been cured by myotomy alone, simply to avert the development of esophagitis in the fifth patient. Our results in the last half of this series show that the incidence of esophagitis can be markedly reduced by the simpler expedient of carefully preserving the normal esophageal-diaphragmatic-gastric relationships, or if these relationships are already altered by a co-existing hiatal hernia, by repairing the hernia at the time of myotomy. The other consideration, inadequate relief of obstruction, is a problem entirely within the control of the surgeon, and he is remiss if he does not make the myotomy incision deep to the mucosa, and equally important, cephalad enough to sever all of the abnormal muscle tissue. This latter point has not received the emphasis it deserves.

Summary

1. Forty-four cases of achalasia treated by transthoracic esophagomyotomy (modified Heller operation) have been evaluated one to 10 years after operation.

2. There were no operative deaths in the series. Eighty-one per cent of the cases for the total 10 year period had a satisfactory result. There have been no late complications or recurrent symptoms in any patient who remained well throughout the first postoperative year.

3. Poor results occurred in eight cases (19%). These failures were equally divided between inadequate relief of obstruction and postoperative reflux esophagitis. There was one death in each group. The poor results became apparent in every instance quite soon after operation.

4. The poor results are largely confined to the early years of the series. Modifications in surgical technic have evolved which are now recognized as being vital to a successful operative outcome. These are: (1) Careful preservation of the normal esophageal hiatal relationships; (2) A properly placed myotomy incision of adequate depth, with a generously high superior limb, and an adequate but limited inferior limb; and (3) Careful search for and repair of a co-existing hiatal hernia.

5. Application of these principles during the last five years of the series has made it possible to obtain lasting satisfactory results in 28 of 30 cases (93%).

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