Treatment of achalasia: a review[1]

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Achalasia has the dual challenge of rarity and incurability. Only a few centres in the world have accumulated a large experience of the condition, and this has hindered critical evaluation of the various techniques used to alleviate patients' dysphagia. Nothing can be done to restore peristalsis to the paralysed and dilated body of the oesophagus, and therapeutic effort has been directed towards relieving the obstruction caused by the poorly-relaxing gastro-oesophageal sphincter. However, a sufficient barrier to the reflux of gastric and duodenal juice must be left, for otherwise oesophagitis, and ultimately a fibrous stricture, will result.

Thomas Willis is traditionally regarded as the first to treat achalasia, following his description in 1679 of his whalebone staff with a button of sponge used to treat 'a certain man of Oxford' who could not swallow. Astley Cooper is said to have recommended bougienage (Purton 1821, Wilks 1866), but only at the end of the nineteenth century were significant advances made. Jaffé (1897) suggested the excision of a longitudinal strip of the lower oesophageal wall, while in 1898 J C Russell of Southport described an inflatable silk and rubber bag with which he had treated 6 patients.

Surgical division of the circular muscle at the cardia – cardiomyotomy – which was proposed, but not performed, by Gottstein (1901), was done by Heller in 1914, but he used two incisions, anteriorly and posteriorly. De Bruine Groeneveldt (1918) used only an anterior incision, an idea popularized by Zaaijer (1923). (It is therefore historically incorrect to refer to today's single incision cardiomyotomy as 'Heller's operation'.)

Surgical efforts were diverted for a time to excision of the gastro-oesophageal junction, oesophagogastrostomy and other bypass procedures, until the severe consequences of inevitable reflux were recognized. Nonsurgical manoeuvres have never been abandoned and, in particular, more effective methods of dilatation have been devised, notably by Starck (1924), Negus (Thomson & Negus 1955), Plummer (1908) and Mosher (1923).

Drugs

The pharmacology of the oesophagus, particularly the lower oesophageal sphincter, has been usefully studied in recent years (Misiewicz *et al*. 1969, Lobis & Fisher 1976) but no effective drug treatment for achalasia has resulted. Nitrites of various sorts are still sometimes recommended, based on Douthwaite's (1943) studies which showed that inhalation of octyl nitrite opened the cardia. The older nitrites' effects were too short-lived to be of more than transitory value, but studies are in progress with longer-acting substances such as isosorbide dinitrate which seem in a few patients to have produced real symptomatic benefit.

Procaine amide, initially promising (Balfour & Wharton 1951), never became useful in practice. Adrenergic blockade (by injection of dibenamine) (Nickerson & Call 1951) appeared to relax the cardia, but no further studies of adrenergic blockade have shown useful therapeutic benefit. Anticholinergics have been tried, with variable results. Lobis & Fisher's (1976) small study suggested benefit, but Yon & Christensen (1975) had only one success in 7 patients. There has been recent interest in nifedipine which lowers the tone of the gastro-oesophageal sphincter (Weiser *et al*. 1978). Two short series report symptomatic benefit from this drug taken over periods of several months (Weiser *et al*. 1978, Hongo *et al*. 1980).

Although the future may hold further developments, drug treatment of achalasia is not at present of practical value for long-term management.

Surgery

For such a rare condition, a considerable variety of surgical procedures has been used in the past century (Steichen *et al.* 1960). The area of the abnormal cardia has been excised (Rumpel 1897, Pribam 1922, Wangensteen 1951), destroyed by cardioplasty (Marwedel 1903, Wendel 1910) or bypassed (Heyrovsky 1913, Grondahl 1916, Lambert 1913), and operations of this type were favoured until about 1949. In that year Allison (1949) and Barrett (Barrett & Franklin 1949) both published papers showing unsatisfactory results because complete abolition or bypass of the valvular cardia led to free gastro-oesophageal reflux, oesophagitis and eventually stricture. Although the severity of this can be diminished by a biliary diversion procedure (Holt & Large 1961), these operations have been universally abandoned in favour of some type of cardiomyotomy. Heller (1914) performed the first cardiomyotomy, and therefore deserves the eponymous credit given to him, though he actually used a double incision (anterior and posterior). Today a single incision is always used, as first practised by de Bruine Groeneveldt (1918) and popularized by Zaaijer (1923).

The precise length of the longitudinal incision is a matter of individual preference. Most published descriptions agree that it should be extended on to the gastric fundus for two or three centimetres, but the height up the oesophagus is more variable: 6–8 cm is often recommended (e.g. Menzies-Gow *et al.* 1978) but others (e.g. Effler *et al.* 1971) prefer a longer cut, up to the aortic arch. Experimental studies (Geever & Merendino 1953, Ellis *et al.* 1967) suggest that a longer myotomy is more likely to cause gastro-oesophageal reflux. The depth of the incision is not disputed, all authors recommending careful division of all circular muscle fibres. A consequent hazard is cutting through the mucosa into the oesophageal lumen, but this can readily be sutured.

Adequate relief of dysphagia is usually achieved by cardiomyotomy (Table 1). Most recent series report less than 5% residual dysphagia. Cardiomyotomy is a safe operation,, and operative deaths are rare. In a review of 1906 cardiomyotomies from 1950–1967 operative mortality was 1.4% (Ellis & Olsen 1969), though of course there is the expected incidence of postoperative septic and vascular complications.

The main problem with cardiomyotomy is the subsequent occurrence of gastro-oesophageal reflux, and possibly later on an oesophageal stricture. The Mayo Clinic study in dogs suggested

Table 1. Cardiomyotomy in treatment of achalasia

Author	Year	No. of patients	Operative mortality (%)	Poor result (dysphagia) (%)	Reflux (%)	Stricture (%)
Hawthorne & Nemir	1953	22	0	15.0	50	—
Hawthorne *et al.*	1956	35	2.8	5.7	50 (of 21)	—
Acheson & Hadley	1958	35	0	26.0	58	3
Gammie *et al.*	1958	18	0	12.0	17	—
Lawrance & Shoesmith	1959	58	0	8.6	—	—
Douglas & Nicholson	1959	28	0	21.4	50	7.1
Le Roux & Wright	1961	26	3.8	21.0	0	—
Nemir & Frobese	1962	58	0	3.4	40	—
Jekler *et al.*	1964	21	—	—	14.2	—
Ellis & Cole	1965	56	0	0	28.5	7.1
Ellis *et al.*	1967	227	0.37	2.6	4.4	1.7
Tala *et al.*	1968	46	0	11	0	0
Grimes *et al.*	1970	50	0	22	—	—
Bennett & Hendrix	1970	64	0	—	22	11
Rees *et al.*	1970	59	0	1.7	15.2	—
Effler *et al.*	1971	100	0	5	6	0
Barker & Franklin	1971	30	0	0	17	0
Wingfield & Karwowski	1972	27	0	3.7	11.1	3.7
Yon & Christensen	1975	20	0	15	25	0
Black *et al.*	1976	108	0	25	18.5	6.8
Menzies-Gow *et al.*	1978	102	0	0	5.8	4.9

that reflux was more likely if a long myotomy was done (Ellis *et al.* 1967), though Effler *et al.* (1971), who routinely used a long incision, reported an incidence of reflux little higher than that at the Mayo Clinic. It has even been suggested that the gastro-oesophageal junction does not remain functional after cardiomyotomy and that competence of the cardia is maintained by other factors (Ellis *et al.* 1967, Lobello *et al.* 1978). Reflux causing heartburn is a considerable nuisance to a patient after cardiomyotomy, and responds poorly to medical therapy. The occurrence of a peptic stricture is a major problem, for even if it does not become narrow, any stricture produces disproportionate dysphagia because there is no peristaltic 'push' in the gullet above.

Some surgeons (notably Black *et al.* 1976) have advocated some form of anti-reflux procedure at the time of cardiomyotomy in order to minimize reflux, and compared with their overall incidence of stricture of 6.8% – comparable to other series – 11 patients having 'formal hernia repair' at the time of cardiomyotomy showed no evidence of reflux.

Self-bougienage
The passage of bougies to maintain the ability to swallow was popularized by Hurst (1913, 1927), whose mercury-weighted bougie is still available in this country. Any dilatation of the cardia in achalasia improves flow temporarily, and if the patient is prepared to swallow a bougie daily he can preserve reasonable swallowing. Such a regimen is not particularly pleasant for the patient, and is unsuitable for an oesophagus which is dilated and tortuous.

Benedict (1964) reported on 40 patients who had been treated by mercury bougies: 32 (80%) are said to have produced a good result, but detail is lacking. Yon & Christensen (1975) had 32 treated with mercury bougies, 10 of whom were instructed in self-bougienage. There were two perforations, and the authors considered that none of the results were satisfactory. Self-bougienage must be regarded as a poor substitute for more effective and lasting treatment.

Forceful dilatation
Instruments intended to dilate the cardia forcefully (rather than the gentle stretch applied by a simple bougie or oesophagoscope) have been mechanical, hydrostatic and pneumatic.

Mechanical
Max Einhorn (1888) described an expanding metal dilator, but that of Starck (1924) has been more widely used. Its jointed metal arms are opened by a hand-operated lever. The force applied is abrupt and cannot be measured. To most people this would seem a disadvantage, though Schindler (1956), in the most recent published account, regards it as a virtue. Schindler treated 84 patients and claims to have cured 95% (criteria not given). Four patients (5%) were not cured, and in 5 of those cured there were 'febrile complications', two going to laparotomy.

Hydrostatic instruments
Negus dilator (Thomson & Negus 1955): This instrument carries an inflatable bag on a rigid stem, intended to be passed through a rigid oesophagoscope. It is distended by water. It is this instrument which, until recently, was best known in Britain.

Lawrance & Shoesmith (1959) failed to relieve 41% of their 100 patients, while le Roux & Wright (1961) found that 50% became normal, and 42% were improved. Others have recorded poor results, anecdotally. The likely reason for dissatisfaction is that the instrument is positioned by eye down the oesophagoscope, usually under general anaesthesia, making it difficult to place it accurately over the cardia.

Plummer dilator (Plummer 1908): This is a similar instrument used in North America. The only large series have been reported from the Mayo Clinic (Olsen *et al* 1951, Sanderson *et al.* 1970). From 1935 to 1946, 555 patients were so treated – a uniquely large experience; 452 were followed up. Sixty per cent were considered satisfactory results, and in those who were unsatisfactory, second and third dilatations produced good results in 38% and 19%. There was a perforation rate of 1.8%, with a mortality of 0.36% (in pre-antibiotic days). By 1970 the incidence of perforation (or clinical incidents suggestive of leakage) was 5%; 65% of results were considered good.

Pneumatic dilators
Four dilators operated by pneumatic pressure are currently in use.
Mosher dilator (Mosher 1923): This has a cylindrical bag with maximum diameter of 3.6 cm.
It is mounted on a flexible metal stylet introduced through the rubber tube.
Sippy dilator (van Goidsenhoven *et al.* 1963): This has a series of waisted bags from 3–5 cm
diameter, inflated to 300 mmHg. They are mounted on a metal spiral terminated by an olive
bougie, passed over a previously swallowed string.
Hurst-Tucker (Tucker 1939) *or Brown-McHardy*: These are identical instruments, with
cylindrical balloons 3 cm maximum diameter inflated to a maximum of 15 lb/in². The balloon
is mounted on a Hurst flexible mercury-weighted rubber bougie.
Rider-Moeller: This is a waisted balloon mounted on a flexible metal staff with a terminal olive
which slides over a wire guide which can be introduced endoscopically or under flouroscopic
guidance.

Although the basic mechanism of each of these instruments is the same, the details of
manufacture are of some importance. (1) A cylindrical bag makes it easier to determine that
the cardia is being dilated, as it produces a ring of compression which, when the balloon is
correctly placed, should be in the middle of the bag. (2) The diameter needs to be large enough
to produce adequate dilatation without being so large as to rupture the oesophagus. The Sippy
dilator, as used in Louvain, is perhaps safest, using increasing sizes of bag with the sphincter
pressure measured between dilatations; this does add technical problems to the procedure. (3)
Method of introduction. The weighted flexible bougie of the Hurst-Tucker instrument, which
passes under its own weight, seems safest, but may not get through the cardia if the gullet is
dilated and tortuous. Passage over a wire or string is then more satisfactory.

A 5 cm diameter balloon mounted on an endoscope has been described recently (Sakai *et al.*
1979). This enables direct introduction by the standard endoscopic technique, but no
substantial results are yet available.

Results (Table 2)
Results reported in the last 25 years have been analysed, though assessments were not carried
out by uniform criteria. Good results are claimed in from 41% to 98%, most reporting 65% to
80% success. Perforation is a definite risk, most series having about a 5% incidence. These
tears were usually treated conservatively, occasionally surgically, and there were no fatalities.
Subsequent gastro-oesophageal reflux is not always commented upon, but Yon & Christensen
(1975) reported an incidence of 7%, and Bennett & Hendrix (1970), 17%. In neither series did
a stricture develop. Vantrappen *et al.* (1971) suggest that reflux is uncommon after forceful
dilatation as such a procedure is unlikely to damage the oblique muscle fibres at the cardia.

Table 2. Forceful dilatation in treatment of achalasia

Author	Year	Instrument	No. of patients	Perforations (%)	Results (%) Good	Poor
Schindler	1956	Starck	100	5	95	5
Nanson	1962	Mosher	14	0	78	7
Van Goidsenhoven *et al.*	1963	Sippy	57	2	98	2
Kurlander *et al.*	1963	Sippy	92	10.8	84	16
					(of 62)	
Bennett & Hendrix	1970	Hurst-Tucker	57	5	68	23
Grimes *et al.*	1970	Not stated	55	5.5	69	—
Vantrappen *et al.*	1971	Sippy	138	2	87	23
Arvanitakis	1975	Brown-McHardy or Rider-Moeller	33	6	67	33
Yon & Christensen	1975	Pneumatic	48	0	41	19
Vantrappen & Hellemans	1981	Sippy	537	2.6	77	14
					(of 424)	

Comparison of treatments

To quote the title of a *Lancet* (1976) leading article, which is the 'least bad treatment'? The two methods of therapy most likely to give reasonable and prolonged relief of dysphagia are cardiomyotomy and forceful dilatation, though neither restore the oesophagus to normal. A randomized trial, preferably in one centre, might give an answer, but with a rare condition such a study would be difficult to mount.

Four papers have tried to compare results. Bennett & Hendrix (1970) used similar review methods on groups of comparable size treated over the same period – by dilatation in Baltimore and surgically in Liverpool. They found that dilatation gave better results, with less reflux and fewer complications. Arvanitakis (1975) compared groups so treated in the same institution (Wisconsin), and found better results from cardiomyotomy. Yon & Christensen (1975) also compared results of the two treatments in their hospital (Iowa) with possibly a better result from cardiomyotomy (though in fewer patients) but a greater likelihood of reflux oesophagitis. Sanderson *et al.* (1970) compared their reported results of hydrostatic dilatation with the report of Ellis & Olsen (1969) of myotomy results in the Mayo Clinic over the same period; they found that cardiomyotomy had given a greater proportion of good results.

Comment

In experienced hands it should be possible to achieve good and lasting relief of dysphagia in most patients with achalasia by either cardiomyotomy or adequate and accurate forceful dilatation, without mortality and with few serious complications. Destruction of the cardia clearly reduces the barrier to reflux, and there is a hazard – though its size and severity are disputed – of reflux oesophagitis and stricture formation. Such strictures are serious, for they respond poorly to medical therapy and bougienage, and while bile diversion operations may help (Holt & Large 1961), total oesophageal replacement may be necessary. Several series of surgical treatment show a sufficiently greater likelihood of subsequent reflux to render the results unsatisfactory – yet other series report no reflux problem, without any clear reason (though Lobello *et al.* (1978) believe that careful preservation of 'the hiatal mechanism' is the reason). Stricture must be regarded as the most serious long-term consequence of therapy. It has never been reported after forceful dilatation, though it is found in many surgical series, even when particular care has been taken to avoid causing reflux. In favour of forceful dilatation is its relative simplicity. Given a suitable instrument and adequate instruction, any skilled physician can carry it out; no anaesthesia is required, and the patient need be in hospital for only 24 hours. It can be used on virtually any patient, whatever complicating medical problems there may be.

If results from the two procedures were identical, dilatation would be preferable because of its simplicity. We lack data to be certain about the relative results, but it does not seem unreasonable to use dilatation in most patients (especially the old and unfit) as the first line of treatment, for failure does not render cardiomyotomy any more difficult. (Although Sanderson *et al.* (1970) suggest that prior dilatation might render gastro-oesophageal reflux more likely after a subsequent cardiomyotomy, there is no published evidence to support this.) This policy is common in the United States and in some European centres, but in only three British cities is forceful dilatation performed with any frequency. It seems to be time for reappraisal.

In the future drug therapy may be sufficiently effective to make it a practical alternative, at least for early cases, or in those without severe dilatation.

References

Acheson E D & Hadley G D (1958) *British Medical Journal* i, 549–553
Allison P R (1949) *Lancet* ii, 91–94
Arvanitakis C (1975) *American Journal of Digestive Diseases* **20**, 841–846
Balfour D C jr & Wharton G K (1951) *Gastroenterology* **18**, 606–608
Barker J R & Franklin R H (1971) *British Journal of Surgery* **58**, 466–468
Barrett N R & Franklin R H (1949) *British Journal of Surgery* **37**, 194–202
Benedict E B (1964) *Journal of the American Medical Association* **188**, 355–357
Bennett J R & Hendrix T R (1970) *Modern Treatment* **7**, 1217–1228
Black J, Vorbach A N & Collis L R (1976) *British Journal of Surgery* **63**, 949–953

de Bruine Groeneveldt F R (1918) *Nederlandsch tijdschrift voor geneeskunde* **2**, 1281
Douglas K & Nicholson F (1959) *British Journal of Surgery* **47**, 250–253
Douthwaite A H (1943) *Lancet* ii, 353–354
Effler D B, Loop F D, Groves L F & Favaloro R G (1971) *Surgery, Gynecology and Obstetrics* **132**, 1057–1063
Einhorn M (1888) *Medical Record* **34**, 75
Ellis F & Cole F L (1965) *Gut* **6**, 80–84
Ellis F H, Kiser J, Schlegel J F, Earlam R J, McVey J L & Olsen A M (1967) *Annals of Surgery* **166**, 640–655
Ellis F H jr & Olsen A M (1969) Achalasia of the Esophagus. W B Saunders, Philadelphia
Gammie W F P, Jennings D & Richardson J E (1958) *Lancet* ii, 917–920
Geever E D & Merendino K A (1953) *Surgery* **34**, 742
Gottstein G (1901) *Mitteilungen aus den Grenzgebieten der Medizin und Chirurgie* **8**, 57–60
Grimes O F, Stephens H B & Margulis A R (1970) *American Journal of Surgery* **120**, 198–202
Grondahl N B (1916) *Nord. Ku. Forhandl.* **11**, 236
Hawthorne H R, Frobese A S & Nemir P (1956) *Annals of Surgery* **144**, 653–669
Hawthorne H R & Nemir P (1953) *Gastroenterology* **25**, 349–358
Heller E (1914) *Mitteilungen aus den Grenzgebieten der Medizin und Chirurgie* **27**, 141–145
Heyrovsky H (1913) *Archiv für klinische Chirurgie* **100**, 703
Holt C J & Large A M (1961) *Annals of Surgery* **153**, 555–562
Hongo M, Ishimori A, Kawamura T, Koizumi F, Nakanome C, Sato T & Unoura A (1980) 11th International Congress of Gastroenterology. Hepato-Gastroenterology Suppl. Thieme, Stuttgart; p 45
Hurst A F (1913) *Proceedings of the Royal Society of Medicine* **7**, 150
Hurst A F (1927) *Lancet* i, 618
Jaffé K (1897) *Münchener medizinische Wochenschrift* **44**, 386
Jekler J, Lhotka J & Borek Z (1964) *Annals of Surgery* **160**, 793–800
Kurlander D J, Raskin H F, Kirsner J B & Palmer W L (1963) *Gastroenterology* **45**, 604–612
Lambert A V (1913) *Annals of Surgery* **58**, 415
Lancet (1976) i, 403–404
Lawrance K & Shoesmith J H (1959) *Thorax* **14**, 211–215
le Roux B T & Wright J T (1961) *British Journal of Surgery* **48**, 619–633
Lobello R I, Edwards D A W, Gummer J W P & Stekelman M (1978) *Thorax* **33**, 569–573
Lobis I F & Fisher R S (1976) *Gastroenterology* **70**, 976
Marwedel G (1903) *Zentralblatt für Chirurgie* **30**, 938
Menzies-Gow N, Gummer J W P & Edwards D A W (1978) *British Journal of Surgery* **65**, 483–485
Misiewicz J J, Waller S L, Anthony P P & Gummer J W P (1969) *Quarterly Journal of Medicine* **149**, 17–30
Mosher H P (1923) *Pennsylvania Medical Journal* **26**, 240
Nanson E M (1962) *Canadian Medical Association Journal* **86**, 1107–1111
Nemir P & Frobese A S (1962) *Surgical Clinics of North America* **42**, 1407–1418
Nickerson M & Call L S (1951) *American Journal of Medicine* **11**, 123–127
Olsen A N, Harrington S W, Moersch H J & Anderson H A (1951) *Journal of Thoracic Surgery* **22**, 164–187
Plummer H S (1908) *Journal of the American Medical Association* **51**, 549
Pribam B O (1922) *Archiv für klinische Chirurgie* **120**, 207
Purton T (1821) *Medical and Physiological Journal of London* **46**, 540
Rees J R, Thorbjarnarson B & Barnes W H (1970) *Annals of Surgery* **171**, 195–201
Rumpel T (1897) *Münchener medizinische Wochenschrift* **44**, 383
Russell J C (1898) *British Medical Journal* i, 1450–1453
Sakai P, Ishioka S, Pinotti H W, Ellenbogen G & Raia A (1979) *Endoscopy* **2**, 116–120
Sanderson D R, Ellis F H jr & Olsen A M (1970) *Chest* **58**, 116–121
Schindler R (1956) *Annals of Internal Medicine* **45**, 207–215
Starck H (1924) *Münchener medizinische Wochenschrift* **71**, 334
Steichen F M, Heller E & Ravitch M M (1960) *Surgery* **47**, 846–876
Tala P, Luosto R & Maamies T (1968) *Annales chirurgiae et gynaecologiae Fenniae* **57**, 281–286
Thomson St C & Negus V E (1955) Diseases of the Nose and Throat. Cassel, London
Tucker G (1939) *Annals of Otology* **48**, 808–816
van Goidsenhoven G E, Vantrappen G, Verbeke S & Vandenbroucke J (1963) *Gastroenterology* **45**, 326–334
Vantrappen G & Hellemans J (1981) In: Therapeutic Endoscopy and Radiology of the Gut. Ed. J R Bennett. Chapman & Hall, London (in press)
Vantrappen G, Hellemans J, Deloof W, Valembois P & Vanderbroucke J (1971) *Gut* **12**, 268–275
Wangensteen O H (1951) *Annals of Surgery* **134**, 301
Weiser H F, Lepsien G, Golenhofen K & Siewert R (1978) In: Gastrointestinal Motility in Health and Disease. Ed. H L Duthie. MTP, London; p 565
Wendel W (1910) *Archiv für klinische Chirurgie* **93**, 911
Wilks S (1866) *Transactions of the Pathological Society of London* **17**, 138
Willis T (1679) Pharmaceutice rationale. English edn. Oxford
Wingfield H V & Karwowski A (1972) *British Journal of Surgery* **59**, 281–284
Yon J & Christensen J (1975) *Annals of Surgery* **182**, 672–676
Zaaijer J H (1923) *Annals of Surgery* **77**, 615–618