

TRANSPLEURAL CARDIOPLASTY IN ACHALASIA

OPERATION RESULTS AND SEQUELAE

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The clinical syndrome characterized by dysphagia, retrosternal or epigastric pain, and, in advanced stages, by regurgitation of undigested food and total obstruction of the oesophagus, has long been known and has been called by many different names.

The Englishman, Thomas Willis, gave a detailed description in 1672, and after 1882, when v. Mikulicz, who believed it to be caused by a spasm in the region of the cardia, published his studies on the subject the syndrome was known as "cardio-spasm"; this name is still in current use in many countries. When later investigations disproved the occurrence of a spasm in the cardiac region Hurst (1914) introduced the name "achalasia," or lack of relaxation of the cardiac sphincter. But in the light of our experience and knowledge to-day it seems that the condition is due to a combination of factors and not solely to failure of relaxation, so that Hurst's suggestion is no longer satisfactory.

Imbalance in the nerve supply to the cardiac sphincter may be responsible for the onset; if so, the sphincter does not respond by relaxing to waves of peristalsis from the oesophagus but remains closed and thereby arrests the descent of the food to the stomach until the cardiac orifice has been opened by dilatation or surgical intervention or until later on it spontaneously relaxes.

Our knowledge of the aetiology is still obscure, but causal factors do not seem to be uniform. Some authors ascribe the disease to a disorder in Auerbach's plexus (Rake, 1927; Cameron, 1927; Mosher and McGregor, 1928) or to lesions of the central nervous system. Others envisage the possibility of vagotonia (Held and Gross, 1916), degenerative changes in the vagus (Kraus, 1902; Heyrovsky, 1913; Pollitzer, 1913) or a sympathetic nerve derangement (Carlson and Luckhardt, 1921; Kuré, 1929; Knight, 1934, and others). Sauerbruch and v. Haecker (1906) suggested that the diaphragm was concerned, and Jackson, who shared this view, proposed the name of "phrenospasm" for the syndrome.

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Recent experience from transpleural or trans-abdominal gastric vagotomy for gastric and duodenal ulceration has contributed to the elucidation of the role the vagus plays in the occurrence of achalasia. Judging by the many reports in the literature no clinical or radiological evidence of any spastic sequelae of vagotomy has ever been seen in spite of due attention being directed towards this point. Neither have we made such an observation in the course of our experience, which covers some 100 cases of vagotomy. It is, therefore, evident that the oesophageal disorders frequently seen in vagotomized dogs have no counterpart in man.*

Psychogenic factors have also been suspected of playing a contributory role in causing the condition, and judging by the results of investigations hitherto made, the suspicion does not seem to be entirely unfounded. It ought perhaps to be mentioned that a psychiatrist is at present making a personality study of our patients, and the results will be the subject matter of a later paper.

Some years ago very few cases of achalasia were admitted to hospitals, but this is no longer the case. Gray and Skinner (1940), of the Mayo Clinic, published a report of 1,200 cases, and in a large oesophagoscopic series Moersch (1933), for example, found the condition in 17.4% of the cases. Guisez (1935) found a similar percentage in his material, and many cases have been reported from Sweden (Gjertz, 1948).

The symptoms vary from occasional difficulty in swallowing, with a temporary feeling of obstruction behind the sternum or in the epigastrium, to complete obstruction in the lower section of the oesophagus, marked oesophageal dilatation above the functional obstruction, and pulmonary complications in the form of gangrene or abscess of the lungs caused by nocturnal regurgitation and aspiration. As a rule medical aid is not sought until the disease is advanced.

*Several surgeons in Great Britain have had isolated cases in which vagotomy has been followed by the typical radiological and clinical manifestations of achalasia. These symptoms have been of temporary duration.—EDITOR.

Owing to the many aetiological factors concerned in this disease numerous treatments have been suggested and tried with varying success. Relief has sometimes been achieved by the application of antispasmodics, and sometimes the symptoms pass off spontaneously. In more advanced cases dilatation therapy has been applied. In a series of 805 patients treated by dilatation Moersch (1933) reported that 71% were discharged from hospital without symptoms; Wachs (1940) reported dilatation to be successful in 70% of his patients, and in a Swedish series reported by Gjertz in 1948 the results were just as favourable.

It is evident, both from the literature and from our own experience, that dilatation by expanding metal instruments or the introduction of a hydrostatic or air-rubber-bag often gives but temporary relief and is undoubtedly sometimes followed by complications such as rupture of the oesophagus or mediastinitis. We know from our own experience that such ruptures and mediastinitis are more common in cases treated previously by dilatation than might be inferred from statistics. Moersch reported nine cases of death from oesophageal rupture, two from cachexia, and 12 from other post-operative complications. In 64 cases, treated by dilatation, Gjertz reported rupture in three, but there were no deaths. Sometimes the oesophagus is distended to such a degree as to contraindicate dilatation therapy.

In some of these cases surgical intervention must therefore be carried out, but it is not always easy to decide when and in which patients it is indicated. Conservative measures sometimes give relief even in cases in which at first sight there seems to be an obstinate constriction, but on the other hand it is not always wise to delay radical treatment. Each case must be judged on its own merits, due regard being paid to the circumstances as a whole and not solely to the degree of oesophageal dilatation. For instance, when relief can only be obtained by frequently repeated dilatation, or when the oesophagus is grossly distended and sigmoid in shape so that digestion is disturbed and the patient is cachectic, or when there is evidence of pulmonary complications or mediastinitis, immediate radical treatment is indicated.

We would emphasize that delay in remedial treatment is likely to make an anatomical *restitutio ad integrum* of the oesophagus impossible, because prolonged and extreme distension renders it less capable of recovering its original form: the muscle fibres separate and degenerate into connective tissue with the result that the oesophagus

remains dilated, thin-walled, and non-elastic even after the obstruction has been overcome. Similar changes are seen, for example, in hydronephrosis in which plastic surgery is also necessary to give the renal pelvis a more or less normal shape after the removal of the obstruction.

Several aetiological theories have been put forward and almost as many operations have been suggested to cure the condition.

The most popular of these operations are: (1) "extramucous myotomy," i.e., longitudinal incision of the muscle coats of the oesophagus carrying the incision down to the mucous membrane (Heller, 1913; Röpke, 1914); (2) division of the fasciae of the diaphragm in relation to the oesophagus and incision of the hiatus, "phrenicotomy" (Jackson, 1922); (3) establishment of an anastomosis between the oesophagus and fornix ventriculi (the cardiac orifice) *ad modum* Heyrovsky (1913); (4) cardioplasty with a longitudinal incision through all layers from the oesophagus down to the stomach with suture at right angles to the incision (Marwedel, 1903; Wendel, 1910); and (5) operations upon the autonomic nervous system (Meyer, 1911; Sauerbruch, 1925; Knight, 1934).

Concerning the first four methods, there has been much discussion as to whether the abdominal or the transthoracic approach should be given preference. At first, when the operative mortality from transpleural intervention was relatively high, the abdominal approach was naturally the one of choice, but the favourable results obtained with the transpleural approach during the last decade have changed the situation. The transpleural methods, which provide a better approach and a better survey of the field of operation, are now held to involve but small risks, and our experience supports this view.

MATERIAL

From May, 1944, until October, 1948, 21 patients suffering from achalasia have been treated in our department of thoracic surgery. In the meantime three other cases of achalasia have undergone cardioplasty with success, but the operations are too recent to be included in this paper. One patient was operated upon twice. The youngest was 6 years of age, the oldest 63, and the highest incidence of the disease lay between the ages of 18 and 37 years. In other words, the frequency is greatest in that age period when the struggle for life is hardest, a fact which should be borne in mind, because psychogenic factors are also concerned in the onset of the affection.

All the cases, except two, had previously been treated medically or by dilatation by otolaryngologists. The patients were not transferred to our department until endoscopic treatments had failed to give relief or had been abandoned on account of discomfort or the risk of rupture. In general the affection was advanced and in some instances the shape of the distended oesophagus was grotesque.

Two patients had not undergone pre-operative dilatation. One of these was an 11-year-old boy whose history was typical—namely, loss of weight, markedly distended oesophagus, and occasional aspiration of oesophageal contents into the lungs. The other case was that of a 36-year-old man, in whom the radiographs revealed an oesophagus as thick as an arm. He presented himself complaining of a lung infection, which was found to be a large pulmonary abscess secondary to aspiration; and he had stagnant food in the dilated oesophagus. Both patients were operated on without pre-operative dilatation because continuation of the conservative treatment would in our opinion have incurred undue risks and even threatened the patient's life.

OPERATION METHODS AND TECHNIQUE

Twenty-two operations were done on 21 patients: cardioplasty in 16 and extramucous myotomy in six. All operations were done transpleurally and in the myotomy cases all layers of the oesophagus were incised down to the mucous coat. In these the operation consisted of a single longitudinal incision from the oesophagus down to the cardia and they were done in the early part of the series, but as the results seemed to be unsatisfactory, we adopted once more the method used first in the case operated on in 1944—i.e., transpleural cardioplasty, essentially *ad modum* Marwedel-Wendel.

Technique.—All patients were operated upon in the lateral position and under intubation narcosis with ether-oxygen and nitrous oxide. From 20 to 25 cm. of the ninth rib on the left side were removed subperiosteally. The pleura was incised, and the mediastinal pleura was opened in the triangle between the heart, the aorta, and the diaphragm at the site of the inferior pulmonary ligament. The oesophagus, which was often very dilated, was then easy to free from its attachments. A short incision was made in the oesophageal hiatus so that the lower section of the gullet and the fornix-cardia could be drawn up for inspection. The longitudinal oesophageal incision was made down to the cardiac notch over and across

the ventricular fornix towards the greater curvature. The length of the incision varied between 4 cm. and 7 cm.

In the cardioplastic operations all the layers were cut through, whereas in the myotomies the incision was made down to the mucous membrane only. In the latter operations the cut oesophageal muscles were not sutured. In the cardioplastic cases the longitudinal incision was sutured transversely by means of two rows of stitches, an inner continuous catgut and an outer interrupted silk suture. The cardia and the upper part of the fornix ventriculi were always left supradiaphragmatically, and fixed to the diaphragm by interrupted silk sutures (Fig. 1). Only in one



FIG. 1.—Diagram showing the three main steps in transpleural cardioplasty: (1) longitudinal incision through the lower oesophagus, the cardia, the ventricular fornix, with (2) transverse suture; (3) fixing the cardia and the superior part of the ventricular fornix above the diaphragm.

case (the first) was gastrostomy performed before the radical operation. The pleural cavity was not drained during or after the operation, and no infection of the pleural cavity, the abdominal cavity, or the wall of the thorax occurred in connexion with the operations. In most cases 500,000 units of penicillin were introduced into the pleural cavity and one to three aspirations were adequate to remove the usual post-operative effusion. In the cardioplastic cases which were operated upon between 1944 and 1945 the patients were not allowed to drink for the first four to five days after operation, whereas in the latter part of the series they were given small quantities of liquid on the following day. The patients were generally allowed to leave their beds five to fourteen days after the operation.

IMMEDIATE POST-OPERATIVE RESULTS

In the six cases of extramucous myotomy the primary post-operative course was uneventful, neither did any post-operative complications supervene in 15 of the 16 cardioplastic operations. The sixteenth case was that of a 57-year-old woman inebriate, who was in a very poor condition on admission; her serum albumin was less than 4%.

In spite of about one month's pre-operative treatment no improvement was observed. This resistance to treatment and the severe degree of constriction which was present demanded, in our opinion, immediate surgical intervention, and the operation was carried out in the usual manner. This radical treatment gave complete relief, all the previous symptoms disappeared, and the patient's general condition improved. One morning, about three weeks after the operation and after the patient had been sitting up for some days, she died suddenly from pulmonary embolism without any premonitory elevation of temperature. A normal operation field and unobstructed oesophagus were found post mortem and the fatal embolus was revealed in the pulmonary artery.

On discharge from hospital all the other patients who had undergone cardioplasty were in a good general condition and free from the symptoms of achalasia.

Routine radiological examinations were carried out in all cases three to four weeks after operation, and the films showed a normal passage through the operation area in 12 cases and a slightly retarded passage in three. A comparison of the radiological outline of the oesophagus before and after operation showed a marked reduction in dilatation in three cases, a moderate diminution in nine, and practically no improvement in three.

LATE RESULTS

Cases Treated by Cardioplasty.—The 15 patients who underwent cardioplasty were followed up for periods varying from five months to five years after operation. When last reviewed radiographically and clinically during the latter part of 1948, the films showed a progressive but moderate reduction in the oesophageal dilatation in five, but no further changes were seen in any of the others. We compared the pre-operative oesophageal radiological outlines with those observed some time after operation and found a reduction in size in every case.

As to the subjective symptoms, eight patients were entirely symptom-free, one had very slight dysphagia, and one occasional discomfort from regurgitation. Four patients had slight to moderate symptoms not directly related to the affection, such as nausea, heartburn, and epigastric or oesophageal gas formation. As a rule the symptoms were less troublesome or painful and of shorter duration than before operation.

In one case dysphagia persisted practically unchanged after operation. The patient was a 36-year-old unskilled labourer, who stammered.

Personality studies revealed that he was hypersensitive and neurotic; his intelligence quotient was found to be comparable to that of a 10-year-old child. He was readmitted on three different occasions, and every time the quiet hospital environment and the fact that he could take as much time over his meals as he wished had a very good influence on his condition and resulted in rapid recovery.

Cases Treated by Myotomy.—The results obtained in two of the six cases of myotomy were satisfactory. In one case there was a clear-cut recurrence of symptoms with severe pain so that re-operation was indicated, and this time a cardioplasty was done. The patient is now symptom-free (Fig. 2). Evidence of slight or moderate achalasia was seen in the other three patients after operation.

A comparison of the pre-operative radiological outline of the oesophagus with that seen post-operatively showed that the degree of dilatation was unchanged in four cases; one oesophagus which pre-operatively was moderately distended had returned to normal size, and in another there was a moderate decrease compared with the size shown on the pre-operative films.

The results achieved by myotomy were thus rather discouraging both as to the function and the morphology of the oesophagus, operation having had but little or no effect on the dilatation. This induced us to abandon myotomy and to give preference to cardioplasty.

OPERATION SEQUELAE

Secondary Anaemia.—In 1947, in the course of the follow-up examination of these cases, a young boy, who about four months earlier had been operated on successfully for achalasia, was found to have a severe secondary hypochromic anaemia (Hb 51%; red count, 3.14 millions). As no explanation could be found for the anaemia, and as we thought that the operation, which had been done such a long time before, could hardly be held responsible, relatively large amounts of iron were administered and the patient recovered. Two weeks later another patient, who had undergone the same kind of operation, also appeared with secondary anaemia of the hypochromic type. This made us suspect some connexion between the operation and the anaemia, and we directed our attention to this point, especially as Weber's test was negative and the blood counts in these two patients were normal on discharge from hospital. Perusal of the records of the blood counts in the follow-up examinations of those patients who

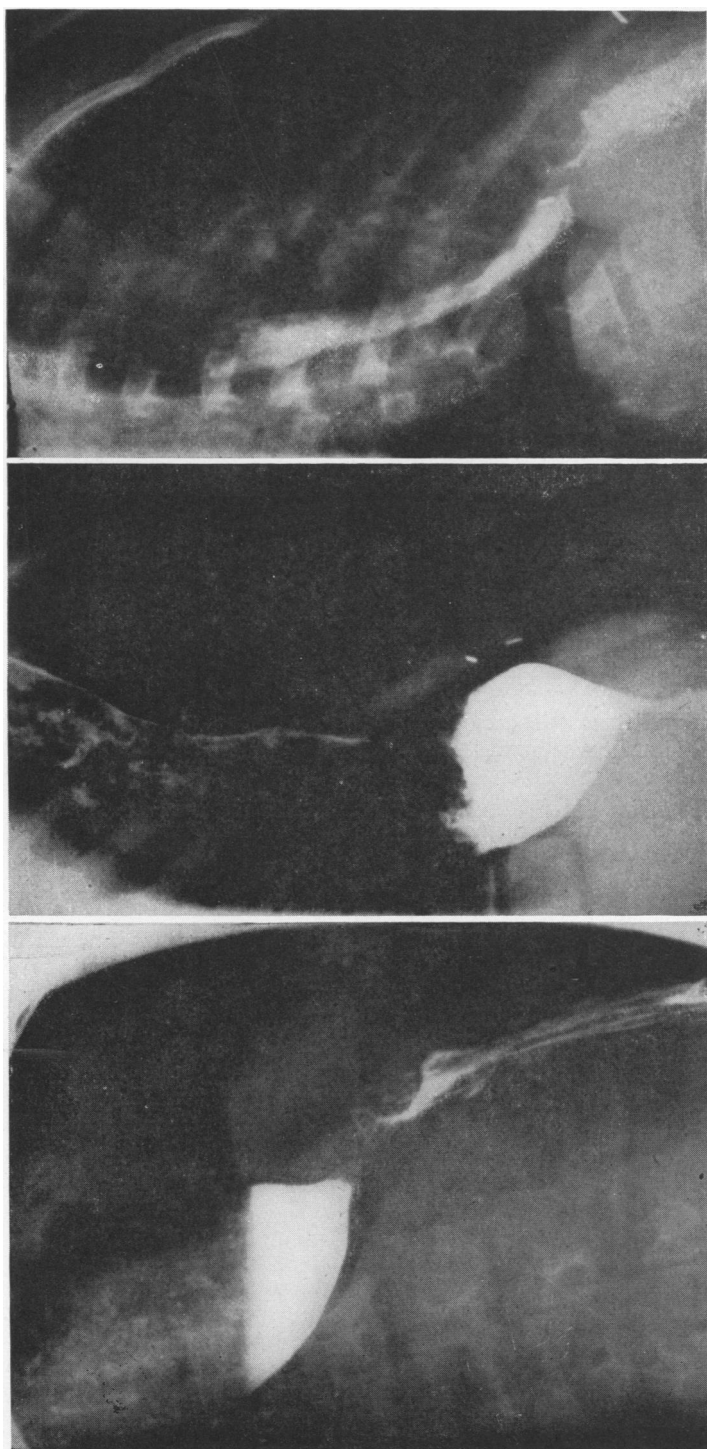


FIG. 2.—Radiographs of chest of 6-year-old boy showing (a) marked achalasia before myotomy; (b) evidence of achalasia 20 months after myotomy and immediately before cardioplasty; (c) diminished dilatation and satisfactory passage 11 months after cardioplasty.

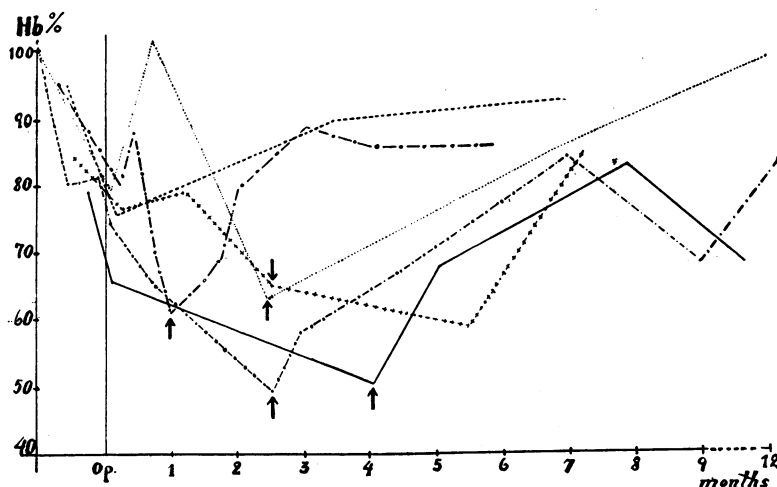


FIG. 3.—Diagram showing degree and time of onset of anaemia in six cases of achalasia treated by transpleural cardioplasty. The anaemia responded to iron medication. Arrows indicate initiation of iron therapy.

had until that time undergone myotomy and cardioplasty showed that the incidence of secondary anaemia was so high in the cardioplastic cases (Fig. 3) that we considered that this operation must in some manner be held responsible for the blood changes.

The secondary anaemia in these two cases and in those described later did not occur immediately post-operatively, but during convalescence (Fig. 3). We found that in all of the cases operated on during the last year, with the exception of the one who died three weeks after operation and another, who 10 days after operation had a severe haemorrhage from a gastric ulcer, there was evidence of moderate or severe anaemia with a haemoglobin value of under 50%.

All the cases were treated successfully by the exhibition of iron (Fig. 3), and except for a certain feeling of tiredness the patients were soon symptom-free. In some the disease recurred as soon as the medication was discontinued.

We must stress that secondary anaemia was observed only after cardioplasty. Judging from our material, myotomy is not followed by anaemia, and we have not seen this complication post-operatively or in the follow-up examinations during the latter part of 1948.

DISCUSSION

In this material secondary anaemia occurred as a late complication of cardioplasty. The incidence was such that the operation was suspected of being responsible for the blood disease. In a personal

communication Barrett (1948) told us that his experience as regards the likelihood of anaemia occurring after cardioplasty was identical with our own. In his cases a high proportion of patients who had undergone oesophago-gastrostomy had also developed a late secondary anaemia whereas those treated by Heller's operation had a normal post-operative blood picture.

A perusal of the relevant literature revealed that secondary anaemia had seldom been observed before and had not been believed to be related to the operation. Grimson and others (1946) published two cases

and Bell (1946) one. In Bell's case the secondary anaemia appeared 11 months after oesophago-gastrostomy; it was severe and was diagnosed as a symptom of bleeding ulcer, though no clear-cut evidence was available to support the diagnosis.

Many explanations may be proffered to explain the onset of secondary anaemia in these cases. One might be inclined to ascribe the condition to iron deficiency due to achlorhydria, but in most of our cases histamine and insulin tests of the gastric juice showed normal values so that at least in these the root of the evil must be sought elsewhere.

One theory is that granulation tissue forms during the healing process at the site of the anastomosis or of the cardioplasty, and that the numerous traumata caused by the passage of the food might result in small oesophageal lesions and haemorrhages in this region. We know from experience elsewhere—e.g., rectal haemorrhages—that small repeated haemorrhages will not produce symptoms until after a certain latent period. This theory is supported by observations made in two cases in the present series in which post-operation oesophagoscopy revealed a very brittle and readily bleeding granulation tissue at the site of the suture line. On the other hand, the secondary symptom did not as a rule occur until two to four months after the operation when haemorrhage from granulation tissue was hardly to be expected, and this argues against this belief. Moreover, in four of our cases with secondary anaemia both Weber

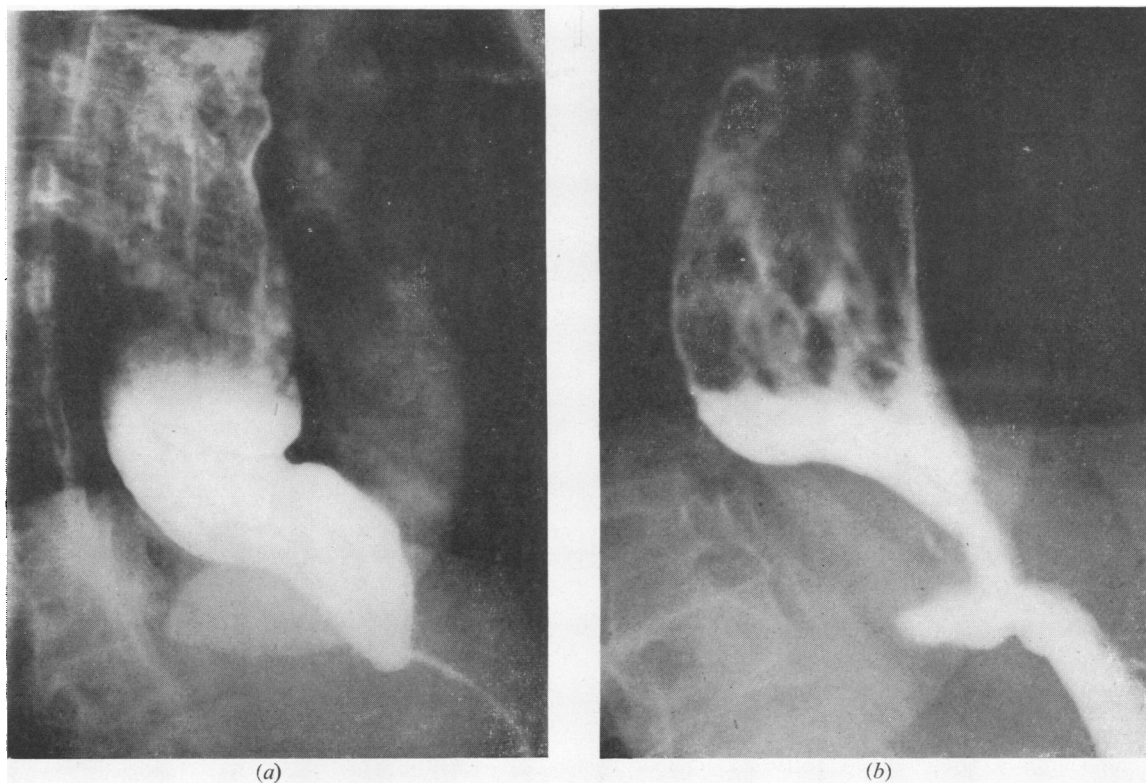


FIG. 4.—Radiograph of chest of 59-year-old woman (a) before cardioplasty; (b) eight months after cardioplasty showing no diminution of dilatation but good passage.

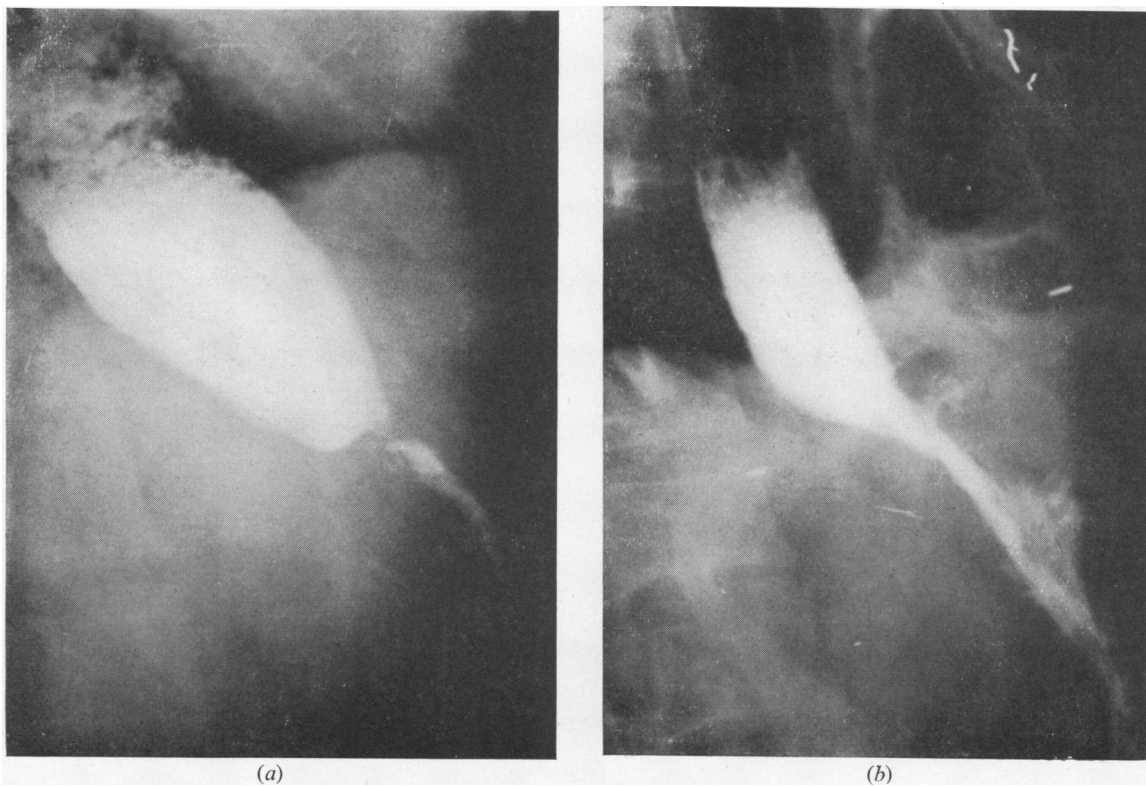


FIG. 5.—Radiograph of chest of 35-year-old man (a) before cardioplasty; (b) seven weeks later showing rapid regression of the dilatation.

tests and Benzidin tests were negative on post-operative review.

Another theory is that in these cardioplastic operations, in which part of the ventricular fornix is drawn up into the thoracic cavity, the blood supply in this part of the stomach is thereby changed, with the result that a varying degree of mucous haemorrhage may be expected. In France clinical evidence seen in ventricular herniation of the diaphragm (Bénard, Rambert, and Canivet, 1947) seems to support the idea of haemorrhages being the underlying cause of anaemia following cardioplasty.

A third theory, which we have embraced as a working hypothesis, is that the underlying mechanism is far more subtle and complicated. In this connexion it should be pointed out that in all of these post-operatively anaemic patients the cardia was no longer able to function as a normal sphincter because food could empty into, and be regurgitated from, the stomach unhindered. In the light of our knowledge of the fact that abnormal variations in the gastric secretion of the mucous membranes of the alimentary tract are liable to affect the blood picture (anaemia in achylia gastrica) this secondary anaemia, encountered after the radical treatment of achalasia, might be ascribed to an abnormal gastric secretion in the oesophagus-cardia-stomach region. If this theory be proved correct it would explain the changes in the blood picture. For the purpose of investigating this possibility we are at present performing a series of experiments on dogs which have been subjected to cardioplasty and other transpleural operations. Judging from preliminary results cardioplasty does not produce secondary anaemia in dogs.

SUMMARY

Twenty-one patients were operated on for achalasia (myotomy in six cases, cardioplasty in 16 cases; one patient had two operations—namely, myotomy and cardioplasty).

All cases were operated upon transpleurally. No post-operative complications were observed, except in a single patient, who died from pulmonary embolism three weeks after operation.

In our material myotomy did not produce satisfactory results, with respect both to the function and the morphology of the oesophagus, the operation having had but little or no effect on the dilatation.

Cardioplasty gave functionally satisfactory results. The condition of most patients three

to five years after operation was still satisfactory and the pre-operative degree of dilatation diminished considerably.

Secondary anaemia developed in several cases treated by cardioplasty transpleurally one to four months after operation. In all cases the anaemia responded to iron therapy. In our cases treated by myotomy no such anaemia was observed.

The possible causation of the anaemia is discussed.

REFERENCES

- Barrett, N. R. (1948). (Personal communication.)
 Barrett, N. R., and Franklin, R. H. (1949). *Brit. J. Surg.*, **37**, 194.
 Beattie, W. J. H. M. (1931). *St Bart's Hosp. Rep.*, **64**, 39.
 Bell, H. G. (1946). *Surgery*, **20**, 104.
 Bénard, H., Rambert, P., and Canivet, J. (1947). *Sem. Hôp. Paris*, **23**, 1239.
 Cameron, J. A. M. (1927). *Arch. Dis. Childh.*, **2**, 358.
 Carlson, A. J., and Luckhardt, A. B. (1921). *Amer. J. Physiol.*, **57**, 299.
 Gjert, A. (1948). *Nord. Med.*, **39**, 1495.
 Gray, H. K., and Skinner, I. C. (1940). *J. thorac. Surg.*, **10**, 220.
 Grimson, K. S., Reeves, R. J., Trent, J. C., and Wilson, A. D. (1946). *Surgery*, **20**, 94.
 Guise, J. (1935). *Bull. Mém. Soc. Chirurugiens Paris*, **27**, 331.
 Held, I. W., and Gross, M. H. (1916). *J. Amer. med. Ass.*, **66**, 233.
 Heller, E. (1913). *Mitt. Grenzgeb. Med. Chir.*, **27**, 141.
 Heyrovsky, H. (1913). *Arch. klin. Chir.*, **100**, 703.
 Hurst, A. F. (1914). *Proc. R. Soc. Med.*, **8**, Clin. Sect., 22.
 — and Rake, G. W. (1930). *Quart. J. Med.*, **23**, 491.
 Jackson, Chevalier (1922). *Laryngoscope*, **32**, 139.
 Knight, G. C. (1934). *Brit. J. Surg.*, **22**, 155.
 — (1935). *Ibid.*, **22**, 864.
 Kraus, F. (1902). *Die Erkrankungen d. Speiseröhre*, in Nothnagel, *Spezielle Pathologie und Therapie*, **16**, 129. Wien.
 Kuré, K., et al. (1929). *Pflüg. Arch. ges. Physiol.*, **221**, 367.
 Marwedel, G. (1903). *Zbl. Chir.*, **30**, 938.
 Meyer, W. (1911). *Ann. Surg.*, **53**, 293.
 v. Mikulicz, J. (1882). *Mitth. Ver. Aertzte Nied.-Oesterreich*, **8**, 41.
 — (1904). *Dtsch. med. Wschr.*, **30**, 50.
 Moersch, H. J. (1933). *Ann. Surg.*, **98**, 232.
 Mosher, H. P., and McGregor, G. W. (1928). *Ann. Otol., etc., St Louis*, **37**, 12.
 Pollitzer, H. (1913). *Münch. med. Wschr.*, **60**, 108.
 Rake, G. W. (1927). *Guy's Hosp. Rep.*, **77**, 141.
 Röpke (1914). *Ver. dtsch. Gesellsch. Chir.*, **43**, 121.
 Sauerbruch, F. (1925). In *Chir. Brustorgane* (Berlin) by Julius Springer, **2**, 572.
 — and Haecker, R. (1906). *Dtsch. med. Wschr.*, **32**, 1263.
 Wachs, E. (1940). *Arch. klin. Chir.*, **200**, 259.
 Wendel, W. (1910). *Ibid.*, **23**, 311.
 Willis, T. (1672). *Pharmaceutica Rationalis* (quoted by Hurst, A. F., *J. Amer. med. Ass.*, **102**, 582).