Surgical management of hiatal herniae and oesophageal strictures in systemic sclerosis

R. H. F. Brain

Guy's Hospital, London

Brain, R. H. F. (1973). *Thorax, 28*, 515–520. Surgical management of hiatal herniae and oesophageal strictures in systemic sclerosis. The clinical manifestations of 10 patients with systemic sclerosis whose oesophageal complications were managed by surgery are reported. Ninety per cent had a severe ulcerating type of oesophagitis and all had hiatal herniae demonstrable by contrast radiology. All but two had fibrous strictures.

Their treatment by surgery is reviewed. Earlier diagnosis of reflux oesophagitis should be beneficial in leading to successful control by a simple hiatal hernia repair. The severity of the oesophagitis is explained. Stricture formation, when present, should be treated by excision with replacement by jejunum or colon before the cardiopulmonary manifestations of the disease preclude major surgical measures.

The finding of an impaired vagal neurological supply to the oesophagus in the form of a deficiency in Auerbach's ganglion cells in two patients is of interest. Reflux oesophagitis is classified.

Systemic sclerosis is a bizarre progressive disease responsible for a number of well-recognized clinical syndromes which depend upon its distribution. All are agreed that there is a collagen fault of connective tissue and smooth muscle. The viscera, in particular the oesophagus, the peripheral blood vessels, and lungs, appear to be the areas most often affected.

Involvement of the oesophagus, first reported in 1903 by Ehrmann, is now recognized by its progressive dilatation with poor or absent peristaltic activity and atrophy of the wall. Dilatation was recorded by Rake (1931). An association with gastro-oesophageal reflux was first pointed out in 1943 by Lindsay, Templeton, and Rothman and a high incidence of associated hiatal hernia by Olsen, O'Leary, and Kirklin (1945). Not all writers on this subject are agreed; Somerville, Bargen, and Pugh (1959) found hiatal herniae in only two of their 11 patients while Hale and Schatzki (1944) did not mention such an association.

The objects of this paper are to present 10 personal cases of clinically diagnosed systemic sclerosis (all were females and suffered from dysphagia, an associated reflux oesophagitis, and hiatal hernia) and to discuss the management of these patients by surgery. In terms of the incidence of hiatal herniae the disparity between this and other series may be due to the manner in which the cases present. All patients in this personal group, although subsequently found to be suffering from systemic disease, were originally referred to me solely as cases of dysphagia.

**CLINICAL FEATURES**

The clinical features of the patients in the series are recorded in Tables I and II.

Table I demonstrates the generalized nature of their disease. All had skin and peripheral vascular disease, while six patients in the group had lung involvement causing progressive dyspnoea. One patient had calcification in the lung fields, and in the four patients who died, lung changes appeared to have been the principal cause of death.

Table II summarizes the important oesophageal findings. In most patients the peripheral vascular and skin signs preceded a long history of dysphagia. All had demonstrable hiatal herniae seen by both radiologist and oesophagoscopist. In all but two cases a fibrous stricture was found in association with an ulcerating type of oesophagitis (grades III to IV). An associated congenital short type of oesophagus with a squamous columnar epithelial junction at 32 cm coinciding with a stricture was found in case 9. Diagnoses were made largely on the barium swallow studies; these showed poor or absent primary peristaltic waves.
with dilatation of the oesophageal lumen. Previously reported difficulties arising through confusion between these cases and those showing diffuse oesophageal spasm were not experienced. Oesophagometry studies, recognized as an improved diagnostic procedure by Creamer, Andersen, and Code (1956) and later confirmed by Atkinson and Summerling (1966), were performed in only two patients in the series when they were combined with pH studies confirming reflux from the stomach.

### Table I

**Table I**

<table>
<thead>
<tr>
<th>Case</th>
<th>Age at Operation (yr)</th>
<th>Total Duration Symptoms (yr)</th>
<th>Duration of Dysphagia (yr)</th>
<th>Presence of Hiatal Hernia</th>
<th>Oesophagitis (grades)</th>
<th>Strictures (cm)</th>
<th>X-ray Sign</th>
<th>Dilatation</th>
<th>Peristalsis</th>
<th>Inactivity</th>
<th>Ulceration</th>
<th>Lung Signs/Symptoms</th>
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<tbody>
<tr>
<td>1</td>
<td>37</td>
<td>20</td>
<td>17</td>
<td>III</td>
<td>IV</td>
<td>+37</td>
<td>+</td>
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<tr>
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<td>54</td>
<td>25</td>
<td>6</td>
<td>+</td>
<td>IV</td>
<td>+35</td>
<td>+</td>
<td>+</td>
<td>+</td>
<td></td>
<td>+</td>
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</tr>
<tr>
<td>3</td>
<td>61</td>
<td>25</td>
<td>9</td>
<td>+</td>
<td>II</td>
<td>+33</td>
<td>+</td>
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<td></td>
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<td></td>
</tr>
<tr>
<td>4</td>
<td>57</td>
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<td>9</td>
<td>+</td>
<td>III</td>
<td>+35</td>
<td>+</td>
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<td></td>
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</tr>
<tr>
<td>5</td>
<td>66</td>
<td>20</td>
<td>19</td>
<td>+</td>
<td>III</td>
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</table>

### Table II

**Table II**

<table>
<thead>
<tr>
<th>Case</th>
<th>Total Duration Symptoms (yr)</th>
<th>Duration of Dysphagia (yr)</th>
<th>Presence of Hiatal Hernia</th>
<th>Oesophagitis (grades)</th>
<th>Strictures (cm)</th>
<th>X-ray Sign</th>
<th>Dilatation</th>
<th>Peristalsis</th>
<th>Inactivity</th>
<th>Ulceration</th>
<th>Histology Oesophageal Wall</th>
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<tbody>
<tr>
<td>1</td>
<td>20</td>
<td>17</td>
<td>+</td>
<td>III</td>
<td>+37</td>
<td>+</td>
<td></td>
<td></td>
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<td>Fibrosis ++ no signs muscle atrophy</td>
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<tr>
<td>2</td>
<td>25</td>
<td>10</td>
<td>+</td>
<td>IV</td>
<td>+35</td>
<td>+</td>
<td>+</td>
<td></td>
<td></td>
<td></td>
<td>Fibrosis ++ reduced ganglion cell count</td>
</tr>
<tr>
<td>3</td>
<td>25</td>
<td>20</td>
<td>+</td>
<td>III</td>
<td>+33</td>
<td>+</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td>Fibrosis/ulceration normal ganglion cell count</td>
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<tr>
<td>4</td>
<td>6</td>
<td>6</td>
<td>+</td>
<td>II</td>
<td>+35</td>
<td>+</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td>Fibrosis: absence of ganglion cells</td>
</tr>
<tr>
<td>5</td>
<td>20</td>
<td>9</td>
<td>+</td>
<td>III</td>
<td>+37</td>
<td>+</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td>Granulation tissue, fibrosis</td>
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<tr>
<td>6</td>
<td>19</td>
<td>14</td>
<td>+</td>
<td>III</td>
<td>+37</td>
<td>+</td>
<td></td>
<td></td>
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</table>

### Surgical Management

Table III presents the surgical experience of this small group of patients. It fairly represents the wide variety of opinion still prevalent among surgeons as to the management of reflux oesophagitis. A gastric resection in case 5 and a vagotomy-gastric drainage operation in case 2 both failed to control progressive stricture formation. Neither operation controls reflux and may aggravate the condition by causing damage to the sensitive cardiac mechanism, possibly with resultant alkaline tryptic reflux from the small intestine, the effect of which on the oesophagus is far worse than that of gastric juice.

Mis-diagnosis resulted in cardiomyotomies being performed.

### Table III

**Table III**

<table>
<thead>
<tr>
<th>For relief of Oesophagitis/dysphagia</th>
<th>Patients</th>
<th>Cases</th>
</tr>
</thead>
<tbody>
<tr>
<td>Repair hiatal hernia</td>
<td>6</td>
<td>2, 4, 6, 7, 8, 9, 10</td>
</tr>
<tr>
<td>Replacement operations Jejunum</td>
<td>3</td>
<td>2, 3, 6</td>
</tr>
<tr>
<td>Colon</td>
<td>1</td>
<td>10</td>
</tr>
<tr>
<td>Cardiomyotomy</td>
<td>2</td>
<td>3, 10</td>
</tr>
<tr>
<td>Partial gastrectomy</td>
<td>1</td>
<td>5</td>
</tr>
<tr>
<td>Vagotomy/pyloroplasty</td>
<td>1</td>
<td>2</td>
</tr>
<tr>
<td>Bougiage</td>
<td>3</td>
<td>1, 5, 6</td>
</tr>
<tr>
<td>Gastrostomy</td>
<td>1</td>
<td>5</td>
</tr>
<tr>
<td>Vascular</td>
<td>Sympathectomy</td>
<td>4</td>
</tr>
<tr>
<td>Others</td>
<td>Partial thyroidectomy (Hashimoto)</td>
<td>1</td>
</tr>
<tr>
<td></td>
<td>Radical mastectomy</td>
<td>1</td>
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</tbody>
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## TABLE IV
RESULTS OF SURGERY

<table>
<thead>
<tr>
<th>Case</th>
<th>Cortisone</th>
<th>Procedure</th>
<th>Outcome</th>
<th>Duration</th>
<th>Complications</th>
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</thead>
<tbody>
<tr>
<td>1</td>
<td>+</td>
<td>Dilatations</td>
<td>Moderate symptomatic relief</td>
<td>Died</td>
<td>20 yr</td>
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<tr>
<td>2</td>
<td>+</td>
<td>'Jejunal' transplant 1961</td>
<td>Complete and permanent relief</td>
<td>Died</td>
<td>9</td>
</tr>
<tr>
<td>3</td>
<td>-</td>
<td>'Jejunal' transplant 1961</td>
<td>Fluids only preoperation</td>
<td>Alive and well</td>
<td>10</td>
</tr>
<tr>
<td>4</td>
<td>+</td>
<td>Repair hiatal hernia 1963</td>
<td>Good result 5 yr, deterioration moderate dysphagia (normal food)</td>
<td>Died</td>
<td>6</td>
</tr>
<tr>
<td>5</td>
<td>+</td>
<td>Dilatations (3)</td>
<td>Complete relief oesophagitis but moderate dysphagia on normal food</td>
<td>Died</td>
<td>2</td>
</tr>
<tr>
<td>6</td>
<td>-</td>
<td>'Jejunal' transplant 1966</td>
<td>Complete relief all symptoms plus 1st weight; fluids only preoperation</td>
<td>Alive and well</td>
<td>6</td>
</tr>
<tr>
<td>7</td>
<td>+</td>
<td>Repair hiatal hernia 1967</td>
<td>Incomplete relief, normal food slowly +1st weight, fluids only preoperation</td>
<td>Alive and well</td>
<td>5</td>
</tr>
<tr>
<td>8</td>
<td>-</td>
<td>Repair hiatal hernia 1968</td>
<td>Incomplete relief—normal food slowly</td>
<td>Alive and well</td>
<td>4</td>
</tr>
<tr>
<td>9</td>
<td>-</td>
<td>Repair hiatal hernia 1969</td>
<td>Incomplete relief of symptoms*</td>
<td>Alive and well</td>
<td>2</td>
</tr>
<tr>
<td>10</td>
<td>-</td>
<td>'Colon' transplant 1970</td>
<td>Complete relief all symptoms, fluids only preoperation</td>
<td>Alive and well</td>
<td>1</td>
</tr>
</tbody>
</table>

*Advised to have transplant but refused—satisfied with improvement

Transplant advised but unfit—cardiopulmonary reasons

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**FIG. 1.** Case 8 (a). Barium study 1961 shows reflux but no definite hiatal hernia. (b) Six years later—an obvious hiatal hernia and stricture. (c) Two years after hiatal hernia repair—improvement only.
performed on two patients of the series, their radiological pictures being interpreted as showing achalasia at a time when other signs of a systemic sclerosis were minimal. These operations led to a rapid worsening of the oesophagitis, stricture formation, and the necessity for an excision and replacement operation soon afterwards.

Repeated bouginage was needed by three patients, one of whom (case 5) later had to have a gastrostomy.

Simple repair of the hiatal hernia was undertaken in five patients and stricture excision with replacement in four. A sympathectomy for an associated Raynaud's phenomenon had previously been carried out in four patients, while two patients had operations for coincidental Hashimoto's disease and breast carcinoma respectively.

RESULTS OF SURGERY

The detailed results are set out in Table IV. It can be seen that simple repair of the hiatal hernia was rather disappointing. In three of the four patients so treated, despite the patient's initial ability to take normal solids, where only fluids had previously been accepted, a mild dysphagia recurred usually taking the form of slow eating; this tended to deteriorate over the years. Two patients who had repairs now require transplant operations but one (case 8) has refused further surgery on the grounds that she is happy enough with her improvement (Fig. 1 a–c). The other has lung function too poor for safety.

Excision of the stricture with replacement by an interposition operation was carried out in four patients in this series, using a jejunal segment in three and a colonic replacement in the fourth. This type of operation was first undertaken by the author in 1951 (Brain, 1967). It has proved very successful in this series in that three of the four patients have had permanent and complete relief of all their oesophageal symptoms (Fig. 2 a and b; Fig. 3 a–c). One died after nine years from myocardial failure; two...
others are well after six years and two years. The remaining patient 10 years after a jejunal inter-
position operation has deteriorated during the past five years with a mild recurrence of dysphagia in
association with increased dilatation of the oes-
phagus above the transplant; she can swallow
normal food slowly.
There have been no operative deaths. The only
postoperative complication was a large intra-
pleural haematoma in case 10 which became
infected and had to be drained.

DISCUSSION

A feature of systemic sclerosis affecting the oeso-
phagus is its close association with gastro-oeso-
ophageal reflux and hiatal hernia. Which is the
primary change and whether it is possible for the
former to occur without the latter is speculative.
A partial explanation is available for reflux if one
accepts smooth muscle atrophy of the lower oeso-
phagus as an early feature of its involvement by
the disease; this could lead to a loss of the intrinsic
oesophageal sphincter believed by many to play
an important role in the general mechanism
whereby the cardia controls reflux. Once free
reflux is promoted in an oesophagus already
rendered hypersensitive to the effects of intestinal
juice by virtue of its poor ability to contract pro-
pulsively, it seems likely that a severe ulcerative
type of oesophagitis will arise. Such an oesophagitis
is likely to result in progressive fibrosis with
shortening of the oesophagus, early aggravation
of the pyrosis, and later dysphagia.

Support for such a sequence of events can easily
be found in this series; 90% of the patients had an
oesophagitis of the severe ulcerating type—grades
III, IV, and V (author's classification, Table V).
It may be significant that the only patient whose
repair was permanently successful (case 4) had
preoperative oesophagitis of grade II with mini-
mal fibrosis and shortening. If these patients were
considered earlier for a simple repair of their
hiatal herniae the permanent success rate might
be higher.

It seems that a logical solution to the stricture
problem in these patients would be earlier excision,
with refashioning operations using either jejunum
or colon. Delay in such a progressive disease may
produce patients who are too ill for surgery due
to their cardiopulmonary complications (cases 5
and 9).

Considering the likely aetiology of the patho-
logical changes found in the oesophagus, the
Auerbach's ganglion cell counts performed in cases
6, 7, and 8 should be mentioned. One of them was
normal, one showed a complete absence, and the
third a very much reduced count. In contrast,
Atkinson and Summerling (1966) investigated the
histological pattern of their 22 patients and found
in each a normal number of ganglion cells.
TABLE V

GRADE I
Symptoms only—no change at oesophagoscopy

GRADE II
‘Erythema’ the only sign

GRADE III
Ulceration—vertical serpiginous type extending upwards from cardia for a varying distance

GRADE IV
Ulceration—continuous upwards, often with islets of squamous epithelium left behind and encircled by granulation tissue. An irregular junction zone with normal squamous epithelium

GRADE V
Ulceration—additional ‘penetrating’ ulcer of the cardiac columnar epithelium—peri-oesophagitis, adhesions, and enlargement of the focal lymph glands

REFERENCES


