A REVIEW OF THE PLUMMER-VINSON STRICTURE OF THE CERVICAL OESOPHAGUS

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This note is written because of a belief that though the diagnosis of the Plummer-Vinson stricture is easy, still it is not being diagnosed; that though the treatment is straightforward and efficient, it is not yet thoroughly carried out; that though it can be prevented easily enough, it is not being prevented; that though it is well known to be pre-cancerous, the cancer is not being prevented.

The Plummer-Vinson stricture is a simple, painless, permanent stricture in the cervical oesophagus, not associated with inflammation, swelling or ulceration, which causes obstructive dysphagia. Its anatomical forms, from thin web to long sleeve stricture, are best seen on x-ray examination and are described and illustrated in this supplement. It is usually formed during longstanding iron deficiency, and therefore it is much more common in women. It never shows spontaneous regression. Correction of iron deficiency and anaemia are believed to halt the process, and hinder progression to carcinoma, but such correction does not change the stricture, which remains as it was. The nomenclature has varied much. Until the eighteenth century it was included in the anginas, a name till then reserved for obstructions of breathing and swallowing above the clavicles. Cases with only mucosal atrophy or mucosal inflammation in the foregut, without obstructive dysphagia, should not be included in this stricture syndrome, related though such changes may be.

We exclude stricture due to trauma, radiation, surgical operation, instrumentation and chemical injury as well as congenital webs. Also excluded are similar strictures occurring in benign mucous membrane pemphigoid and epidermolysis bullosa. By definition we exclude high cervical dysphagia due to myopathies, or to neurological disease, or to external pressure, as well as obstructive dysphagia due to isolated cricopharyngeal muscle disease.

Some advance in knowledge and also some confusion has been brought about by the concept of "achalasia" of the cricopharyngeal muscle in persons with no neurological disease to affect the function of the pharynx or oesophagus (Sutherland, 1962; Belsey, 1966; Bishop, 1974; Kilman and Goyal, 1976; Nicks, 1976; Cruse *et al*, 1977). Clinically the dysphagia is obstructive and similar to that of the Plummer-Vinson stricture. Radiologically it is shown by the prominent rounded projection of a large cricopharyngeal muscle bulging anteriorly into the oesophageal lumen from behind. Such an appearance is often seen as a phase of swallowing in people with no dysphagia. Before such an observation can be taken as evidence of achalasia, it has to be associated with obstructive non-neurological dysphagia, clinical and radiological, and it has to be constant at the same phase in each swallow. It is likely, however, that, valid though the concept seems to be, cases of Plummer-Vinson stricture of sleeve type are being erroneously diagnosed

as cricopharyngeal achalasia. Vertical cricopharyngeal myotomy (often with extension of the incision into muscle above and below the cricopharyngeus) is being practised for cricopharyngeal achalasia, and seems to have been done for cases of the Plummer-Vinson stricture passing under that diagnosis. The myotomy seems to have improved swallowing in some Plummer-Vinson stricture cases. This would be in accord with the evidence that muscle as well as mucous membrane is involved in the Plummer-Vinson stricture.

It is probable that the confusion arises from the surgeons concerned restricting the diagnosis of Plummer-Vinson stricture to simple webs, and not accepting that the Plummer-Vinson stricture includes both webs and longer sleeve or cuff strictures. This review includes both types of stricture in the Plummer-Vinson group because both occur predominantly in women, and in anaemic, iron deficient women, and because some patients have a mixture of web and sleeve stricture. The question will have to be settled by more histological examination, more study of the progression to carcinoma, and if possible identification of the constitutional background, in each type. It will not be settled by showing that cricopharyngeal myotomy helps, because it may help both the Plummer-Vinson stricture of sleeve type and cricopharyngeal achalasia.

The stricture and its obstructive dysphagia cause nutritional impairment, more of meat than of total calories as a rule, because soft and liquid foods pass through until a very late stage. Eating becomes a task and loses its tranquillising effect. The social disability lies in the patient not being able to eat in company, or with the family, in the usual way. There is much social and psychological and even religious importance in eating together. A companion is one with whom, by etymology, one eats bread. There are the unpleasantness and danger of the impactive attacks, and impaction of a food fragment in the stricture often enough needs emergency endoscopy. Death from starvation in this country occurs only very rarely, in neglected or mentally disordered patients. It occurred here in the past, because Home (1795) was able to follow three cases over some years until they died, and then to make a postmortem examination in each. Death from starvation must still be a frequent result in underdeveloped countries, even when no carcinoma supervenes. Clifford (1970) considers this disease may be related to disabling oropharyngeal fibrosis in young Hindu women. The stricture may occasionally be the cause of what appears to be anorexia nervosa. In such cases it should always be excluded. Further, the Plummer-Vinson stricture is a stage in a pathogenic process beginning with, as a rule, menorrhagia, and continuing through chronic haemorrhagic iron deficiency (with or without anaemia) to mucosal change in the alimentary tract from the lips to the stomach, then to the Plummer-Vinson stricture, and in the end to carcinoma in the stricture itself, or in the pharnyx, or in the mouth. All this can be prevented. It is likely too that better understanding of this process would advance knowledge of the pathology of mucous membranes and the underlying tissues. For all these reasons its study is worth while.

The web or diaphragm type of stricture consists of a thin fold of mucous membrane, with scanty fibrous tissue between the two layers. In case 855278 (see

below) "There was a small number of histiocytes and plasma cells, together with islands of lymphocytes and congested blood vessels." Similar histology is reported by Shamma'a and Benedict (1958) and Hoover (1935). Watts (1961) says "This infiltration (with inflammatory cells) often extends into poorly developed muscle coats which are often partly replaced by fibrous tissue." Entwistle and Jacobs (1965) report that in 8 out of 26 webs the subepithelial tissues contained "a few chronic inflammatory cells and polymorphs" and in a further 7 "there were considerable numbers of lymphocytes and plasma cells." They report three necropsies on patients with webs. Besides lymphocytes and plasma cells in the lamina propria, the main longitudinal muscle showed degenerative changes most marked in the region of the web. In one a large portion of the longitudinal muscle anteriorly was necrotic and elsewhere there was atrophy. In the only biopsy of a long stricture which contained deeper tissues, there was degenerative change in the striped muscle, but carcinomatous cells were already present in that case. Siegel *et al* (1961) reported a granulomatous myositis of the cricopharyngeus causing obstructive dysphagia. The x-rays reproduced would be consistent with the Plummer-Vinson stricture. The patient was relieved by a cricopharyngeal myotomy.

It seems, therefore, that the stricturing process extends to the muscle coats. This may explain the success, in varying degree, of some operations of vertical myotomy done in cases classified, perhaps erroneously, as achalasia (Belsey, 1966).

The histology of the cuff or sleeve stricture was reported by Owen (1950). "At the site of the stricture there was marked infiltration with inflammatory cells, principally lymphocytes and plasma cells, but also a few polymorphonuclears. At one place the inflammation extended through the muscular coat, completely destroying the latter. Nearby, when the lesion was less severe, the muscle fibres were thin and there was fibrosis and inflammatory infiltration. Another section showed the inflammatory infiltrations more confined to the submucosa, and the muscle layer showing little infiltration and less atrophy."

Autopsy reports are few. The best macroscopic accounts are those of Baillie (1793) and Home (1795) where the patients died of starvation (though the starvation must have produced atrophy in the pharynx and oesophagus). More modern reports are usually obtained when the patient has died of a perforated oesophagus and then sepsis makes interpretation unreliable. Owen (1950) says that Brown-Kelly showed a postmortem specimen (unperforated) from a man of 35 where the lumen was only 4 mm in diameter. Owen's own cases (unperforated and quoted above) had two constricted areas "3.5 cm and 9 cm below the laryngeal opening".

In structure the Plummer-Vinson web stricture resembles hourglass stricture of the stomach (Logan, 1966), and the diaphragm sometimes found in the antrum of the stomach (Irwin *et al*, 1971) as well as the mid-oesophageal web, the web in the lower oesophagus known as the Schatzki ring, and the rare duodenal web (Johnston and Stevenson, 1966). The Plummer-Vinson stricture and the stricture of hourglass stomach may have a similar causation. The relation of the other strictures is not known.

The onset is very gradual. The difficulty in swallowing is obstructive. The patient localises the obstruction in the neck at or above the suprasternal notch.

Liquids and soft foods are easily enough swallowed, but meat, bread, carrots, peas, apples and potatoes begin to stick. The patient manages for a long time by making all food soft, or by cutting it very fine, and by chewing it very well. Sometimes they chew meat for some time, and then spit it out. All this makes eating slow, and a meal takes a long time. Meals away from home are avoided, or soft food is specially selected. Later on the patient is aware that the swallow is getting even smaller, and there may occasionally be total obstruction by food impaction, for instance, by a piece of meat or by a pea. These are referred to as "bad chokes". The word choke is used by people to refer both to a sudden impactive difficulty in swallowing and also to a sudden obstruction of the breathing. At this stage the patient, who is usually the mother, may be eating alone, because the family find the scenes at meals intolerable.

When the stricture makes the lumen very narrow, the dysphagia becomes more disabling, and chokes will be more frequent by impaction of food on the stricture. The alarm and distress when food impacts is due to the feeling of impending blockage of respiration being added to the sudden feeling of total dysphagia. The difficulty is in moving food from the hypopharynx into the oesophagus. The bolus retained in the pharnyx sits on the verge of the glottis, and saliva pools in the hypopharynx. The patient has a feeling of being about to suffocate and breathing may be croupy. Much alarmed, she may make repeated and ineffective attempts to swallow, though indeed if the patient could make an effective swallow, it might drive the food into the larynx. Drinking water does not help the dysphagia. The water itself is blocked and is spluttered up again, if it does not spill over into the trachea. All these features differentiate this dysphagia from lower oesophageal dysphagia. Eventually the patient, with much unpleasant hawking and spitting, brings up the food mouthful. Sometimes she may be admitted for emergency endoscopic extraction of meat or a pea. This kind of attack was included by the old physicians in the "convulsive anginas".

The anxiety, the violent hawking up, the laryngeal crowing, the complaint of choking, the unwillingness of the patient to swallow effectively enough to satisfy the onlookers, lend themselves to a diagnosis of "hysteria", sometimes made by the doctor as well as by the family. But this catastrophic reaction to the impactive attack is not hysteria. It is attention seeking which is highly justified and necessary. The crisis requires quick efficient help to disimpact the impacted bolus.

When the patient complains of this kind of dysphagia, the doctor must recognise the organic nature of the disease, its probable site in the cervical oesophagus, and the necessity for barium x-ray examination and oesophagoscopy. Nothing less will do. Except for emergency endoscopic extraction of an impacted bolus, oesophagoscopy should follow a barium examination and not precede it.

The radiological appearances, and difficulties in x-ray examination and in diagnosis generally, are described later in this supplement to the Journal. Endoscopy ought to be conclusive and almost always is. Yet sometimes when there has been undoubted stricture, the endoscopist does not identify it. Sometimes endoscopy reports mention "difficulty passing the cricopharyngeus" or "cricopharyngeal spasm" or "tightness". Sometimes it seems that a web gives

before the advancing endoscope and is not identified. This probably happened in the eighteenth century when blind bouginage was practised. If the whalebone bougie passed with no or minimal resistance (as it might with a moderate or elastic web) it was taken to support a diagnosis of "spasmodic stricture" or "hysteria". On the other hand, sometimes a narrowing is mentioned by the endoscopist which radiology shows is not a web or a localised stricture. Radiology is then relied on to show whether it is due to external pressure or to a generalised narrowing, as occasionally happens.

The treatment of the stricture is surgical, by dilatation. It is noticeable that where the stricture has been dilated to such a size that the oesophagoscope can be passed through it, swallowing is very good and recurrence is much less likely. Where incomplete dilatation with bougies introduced through the oesophagoscope is all that is possible, swallowing, while improved, is not normal. Recurrence may occur quite soon, and repeated dilatation may be necessary.

In aged women with frail tissues, cervical spinal osteophytes and mucosal atrophy, great experience and judgment are necessary in dilatation, because rupture of the oesophagus, perioesophageal cellulitis and pneumothorax have occurred in the course of dilatation. After such an episode some recover readily enough, but there have been deaths.

The stricture having been dilated, the patient can swallow better, and sometimes very well. But this is the least demanding part of the management. The medical attendant now has to restore to normal the serum iron (and therefore the total body iron), the serum vitamin B12 and the serum folic acid, as well as the haemoglobin and the red cell count. And again, that in premenopausal women is less demanding than the next task, which is to keep the serum iron and other indices normal until the menopause occurs, **no matter how long that may be.** It may be hoped that in the postmenopausal woman, once correction is made, there will be no recurrence of iron and other deficiencies. It is not difficult. A liquid preparation of iron can be taken day and daily all the year round each year till the menopause occurs. Agreeable preparations are available. Iron tablets and capsules are not suitable, not being easily swallowed and being themselves erosive to the gastric mucous membrane and capable of causing chronic haemorrhage. If absorption of iron from the alimentary tract is inadequate, as it often is, then the patient should receive the injection of iron sorbitol B.P., 2 ml intramuscularly once daily in ten day courses, the courses to be repeated so often as the haemoglobin and serum iron indicate the need. Once repletion of iron has taken place, if it proves necessary for maintenance, the injection can be given weekly over many weeks. There is no risk of overdose if the serum iron is estimated from time to time. Absorption of folic acid is usually good and the tablet is easily crushed, but, if needed, the injection of folic acid or of calcium folinate can be given. If vitamin B12 is deficient, and it often is, the injection of vitamin B12 or hydroxocobalamin makes restoration of B12 stores easy. Of course, if menorrhagia could be controlled, the task would be easier. It seems, however, that it usually cannot, short of hysterectomy. Hysterectomy can often be avoided, if it is only a question of a low haemoglobin and iron state, by resolute use of iron,

That means a **prolonged** use of iron, in **sufficient quantity** to keep the serum iron and haemoglobin normal. That implies that the serum iron and haemoglobin must be estimated regularly, not more often than once a month, and not less often than each six months, until the menopause occurs.

That a majority of these women are, or have been, iron-deficient and anaemic at some time is the common clinical experience and cannot be controverted. The association has seemed so strong that most physicians have assumed that iron deficiency is one element in the causation. Proof for or against is not, or not easily, to be had. Many of these strictures come to light after the menopause, when, by the cessation of uterine haemorrhage, both serum iron and haemoglobin are likely to have returned to normal (although that cannot happen unless enough iron is ingested). It is common, therefore, to see a case of the Plummer-Vinson stricture, in whom the mouth, tongue, nails, serum iron and haemoglobin have become normal, but the stricture (which once formed will not go away, no matter how normal the iron state) is still causing obstructive dysphagia. The time to study the serum iron and haemoglobin is before and at the time the stricture is forming. It seems impossible that such a study should ever be carried out. However, it is not necessary from an individual or public health point of view to establish the point, because, stricture or no stricture, we should be preventing iron deficiency and anaemia on another and even more important ground. That is, the maintaining of all women in the childbearing years, from 15 to 50, in a normal state of health. A safe non-surgical, non-radiation prevention of menorrhagia is much to be wished for. It may be in times past when lives were short, and most women were married, and during the childbearing years, either pregnant or lactating, that menorrhagia, iron deficiency, anaemia, Plummer-Vinson stricture and post-cricoid carcinoma were less frequent. But that is not the case today. Moreover, with the change from iron to aluminium cooking pots, a useful source of iron has been lost (Witts, 1969).

Individual susceptibility must be important in the causation of the stricture, because, though many women have chronic iron deficiency, only a limited number develop the stricture. It is not known in what this susceptibility lies. No disproportionate incidence of any HLA type has been found (Middleton *et al*, 1978). Presumably the severity, continuity and duration of the iron deficiency play a part. Chisholm *et al* (1971) have assembled evidence that a special immunological state is involved. The infiltration with lymphocytes and plasma cells noted above may support that. The association with thyroid disease recorded so often may also be an indication of an autoimmune process. The thyroid cells embryologically are derived from the foregut, and might be expected to share in such a process affecting the mouth, pharynx and upper oesophagus.

While some women complain promptly of the dysphagia, consult their doctor early, and are even importunate in demanding a diagnosis and treatment, it is striking how many endure the dysphagia with little or no complaint. They are even reluctant to discuss or admit it. Those who complain insistently are apt to be called "hysterical", all the more because their complaints become strident if the doctor does not understand the diagnosis. But many say nothing about it, or

having complained timidly once, and perhaps been told it is "nervousness", never mention it again. This happens the world over. In West Virginia, McGee and Goodwin (1938) remark "the symptoms often were tolerated for years before a physician was sought." In Sweden, Ahlbom (1936) says " we have often observed that even patients who have rather a marked degree of difficulty in swallowing for twenty or thirty years before admission, have not spontaneously mentioned this fact to the examiner. They regard their inability to take more than 'small swallows' more as an individual peculiarity than as a disease symptom." Owen (1950) in Wales says "These women through the years have schooled themselves to put up with this persistent painless pushing down of food. Some become so accustomed to it that they regard it as normal." When the post-cricoid carcinoma begins, such patients only waken up to the new phase after an interval when the dysphagia is much worse, and the growth advanced. Early diagnosis of the carcinoma, post-cricoid, pharyngeal or mouth, can only happen where the patient is being followed up regularly, say each three months.

The incidence and prevalence of the stricture are unknown. Ascertainment is incomplete. There is no separate indexing of the stricture in the hospital diagnostic index, so it is hard even to trace all the cases attending the hospital. A limited survey of death certificates showed that no useful information could be obtained from them about the number of post-cricoid carcinomas in Northern Ireland. Part of the reason is the difficulty in siting the primary growth. McCrea and Dickie (1968), discussing growths of the hypopharynx and cervical oesophagus, after excluding growths of the pyriform sinus, say "in practice it is found that many growths involve the post-cricoid site, posterior pharyngeal wall and cervical oesophagus, and the allocation of the growth to one particular site of origin is rather artificial." The Northern Ireland Cancer Registration Scheme divides malignant neoplasm of the hypopharynx into four sub-groups—post-cricoid, pyriform fossa, other specified parts and parts unspecified—but malignant neoplasm of the oesophagus is undivided. Similarly the Registrar-General classifies the growths as of the oropharynx or hypopharynx or of the pharynx in general, and of the oesophagus only in general. Until the oesophageal growths are classified more particularly, the statistics will not help in investigation of their cause. Jones (1961) points out that at the Christie Hospital, from 1945 to 1956, of carcinomas of the oesophagus 72.6 per cent were in men and 27.4 per cent in women. But of 322 post-cricoid carcinomas 85 per cent were in women and 15 per cent in men.

It is unsatisfactory trying to discover how many cases of post-cricoid carcinoma previously had a Plummer-Vinson stricture. McCrea and Dickie (1968) describe thirty-five cases of carcinoma of the post-cricoid area, posterior pharyngeal wall, and cervical oesophagus. Twenty-six were female and twelve of the twenty-six gave a history consistent with having had previously a Plummer-Vinson stricture for between three and thirty years. But the history, while weighty evidence, is not conclusive. At that stage neither x-ray nor endoscopy can say if there was a stricture before the carcinoma. The incidence of carcinoma supervening in the cases of Plummer-Vinson stricture will only be established when a large number of patients with the stricture are followed throughout their lives,

and the number of cancers and their sites recorded, whether in mouth, oesophagus or stomach. It is plain, however, from existing evidence that carcinoma is a serious risk. Once the stricture is diagnosed the patient should be put under surveillance for life. The serum iron should be kept normal and dilatation undertaken when swallowing is difficult. McCrea and Dickie (1968) record that of twenty-three post-cricoid carcinomas, treated by surgery, radiotherapy or both, there were only three five-year survivals; of six posterior hypopharyngeal wall carcinomas there were no five-year survivors; and of six cervical oesophagus cancers there was one. Hope for the future must lie in prevention.

Male cases of the Plummer-Vinson stricture are uncommon. Seven cases were traced in the hospital records.

Case 10168. *Date of birth* 26 *January* 1925. *Single. Farmer.*

He first came to notice in 1953 at age 28 because of haemorrhage from oesophageal varices, when a lieno-renal shunt was done. He had had ill health in childhood, the only detail available being that he had had diarrhoea. The dysphagia dated from the age of six. The liver was the seat of a kind of nodular hyperplasia with no liver cell failure at any time. He was recurrently anaemic, and iron and folate deficient. A web was seen on x-ray in 1963. In 1973 a squamous carcinoma had appeared in the pharynx. The web was found at the same time at oesophagoscopy at 15 cms and dilated. The carcinoma was treated with radiation and he has remained well since except for occasional limited variceal haemorrhage.

Case 756874. *Date of Birth* 20 *November* 1912. *Single. Farm Labourer.*

He first attended in 1966 when the haemoglobin was 19 per cent. For fifteen years he had had increasing difficulty swallowing solids, especially meat. He was almost achlorhydric. Serum iron 14 micrograms per cent. Oesophagoscopy showed "a narrowing at 18 cms". He swallowed barium so poorly that the cervical oesophagus could not be demonstrated. He attended again in 1971 when a stricture was well shown on x-ray (Figures 27 and 28 of Atlas). It was found at oesophagoscopy at 19 cms and dilated so that the oesophagoscope passed it. He now had farmer's lung and instead of being anaemic had erythrocytosis. Red cells 6.6 million per cubic millimeter, haemoglobin 19 grams per cent, packed cell volume 58 per cent. Serum globulin 4.1 grams per cent. White cell counts ranged from 3,000 to 6,600. Platelets in 1971 averaged 81,000. A brother died of carcinoma of the stomach.

Case 632860. *Date of Birth* 17 *September* 1917. *Single. Labourer.*

He first came aged 34 saying that he had had difficulty swallowing so long as he could remember, and now a piece of sausage had impacted in his throat. A marked post-cricoid fibrous stricture was found at endoscopy and seen on x-ray as a cuff stricture. The haemoglobin was 73 per cent and the red cells 5 million. He returned only in 1971 when he had erythrocytosis. The red cell count ranged from 6.1 to 7 million. The highest haemoglobin was 20.9 grams per cent. The serum iron was now 120 micrograms per cent. There was still some dysphagia but it was not gross. No abnormal haemoglobin was found and the oxygen dissociation curve was normal. He had a retinal artery thrombosis and later a cardiac and a cerebral infarction. The change to erythrocytosis and a high haemoglobin level could not be explained.

Case 855278. *Date of Birth* 7 *July* 1912. *Married. Shoemaker.*

In 1969 at age 57 he came to the hospital because of a cardiac infarction and was found to be anaemic. Haemoglobin 11.5 grams per cent. He had had a food impaction many years before. In 1974 he was admitted because of meat impaction on a stricture at 19 cms from the upper alveolus, the meat was removed and the stricture dilated.

A biopsy of the margin of the stricture was reported to show "a portion of non-keratinising, stratified squamous oesophageal epithelium, covering loose connective tissue in which are a small number of histiocytes and plasma cells, together with islands of lymphocytes and congested blood vessels."

Case 820848. *Date of Birth* 1944. *Married. Butcher.*

At 24 he came to the hospital saying that since childhood things were inclined to stick in his throat and in childhood he had transfusions for anaemia. Now meat had stuck in his throat and he could not swallow saliva. At oesophagoscopy the impacted meat was removed and a stricture at 16 cms from the upper alveolus was dilated. However, the oesophagoscope would not pass through. The x-ray after dilatation showed narrowing in the cervical oesophagus and what seemed to be the remains of a web.

Case 961056. *Date of Birth* 5 *July* 1941. *Married. Tyremaker.*

At 32 he came complaining of "something in his throat" without any real difficulty swallowing, or at least with no choking. He had been having diarrhoea for two years and was a large taker of aspirin and salicylamide. There was no anaemia. At oesophagoscopy there was a smooth narrowing "just below the cricopharyngeal sphincter." It was easily dilated by the oesophagoscope although with a little bleeding. This stricture was unlike the rest in not being localised to web or cuff and he was not anaemic.

Case 784121. *Date of Birth* 25 *November* 1921. *Married. Labourer.*

At 45 he began to notice that food stuck in his throat and in 1967 at age 46 barium x-ray showed a very narrow stricture in the cervical oesophagus. The haemoglobin was 11.8 grams per cent, red cells 4 million and the mean cell haemoglobin concentration 30 per cent. At oesophagoscopy there was a very tight pharyngo-oesophageal stricture about a centimetre in length. There was great difficulty introducing the finest bougie but eventually it was dilated and the oesophagoscope passed through it. In 1977 he reported that for ten months the difficulty in swallowing had recurred. At oesophagoscopy a stricture at 15 cms looked malignant and biopsy showed a keratinising squamous cell carcinoma. His brother had died of a carcinoma of throat in 1967.

These cases, with the exceptions of 961056 and 820848, who were not shown to be iron deficient or anaemic, show that long continued anaemia in boys and men may be associated with the stricture in susceptible male subjects just as in susceptible females. Two of the seven developed cancers, one of the pharynx and one in the post-cricoid site. So there is the same progression to cancer in men as in women.

The Plummer-Vinson stricture may occur in childhood. Case 10168 above is clear that his dysphagia began about the age of 6 and by 10 he was having typical impactive attacks and could no longer go to Sunday School parties. Though the Plummer-Vinson stricture therefore may occur in an anaemic child, it has to be distinguished from congenital web in the cervical oesophagus. Gaillard *et al* (1976) describe a stricture indistinguishable from a Plummer-Vinson stricture in a child of 3, but one cannot determine from the report at what age the swallowing difficulty began. Hollinger *et al* (1951) describe a child with a cervical oesophageal web which had caused dysphagia from birth and therefore was not a Plummer-Vinson stricture. Roy *et al* (1975) reproduce a similar x-ray of a web in the cervical oesophagus of an infant which, though anatomically indistinguishable from the Plummer-Vinson web, must be congenital. The case of Bishop (1974)

is certainly not one of Plummer-Vinson stricture and is one of the best established cases of achalasia of the cricopharyngeal muscle.

While there are numerous anecdotes in the older writings of difficulty in swallowing without any inflammation or swelling or ulceration (which the old physicians found puzzling), light only came with postmortem dissections. Good accounts were published at the end of the eighteenth century by Baillie (1793), Home (1795) and Munro (1797 and 1811). Baillie wrote "I once saw a very unusual stricture of the oesophagus. It consisted in its inner membrane being puckered together so as to form a narrowness of the canal at a particular part. The canal at that part was so narrow as hardly to allow a common garden pea to pass. There was no appearance, however, of diseased structure in the inner membrane which was so contracted, and the muscular part of the oesophagus surrounding it was perfectly sound. I know that this disease was very slow in its progress, for the person in whom it took place had been for many years affected with a difficulty swallowing, and could only swallow substances of extremely small size."

Home wrote "Five cases of stricture of the oesophagus have come under my own observation. The patients were all above 30 years of age and only one a man. The four women were in other respects healthy, but had delicate and irritable constitutions. Two of the cases appeared to be principally spasmodic, and by the use of bougies were so far relieved that the patients could swallow with tolerable ease. In the other three cases there was a permanent contraction which did not yield to the pressure of the bougie, but on the contrary had its symptoms increased by using it. The patients lived some years supported by liquids, dragging out a painful existence till overcome by violent attacks of irritation which the disease brought on they died."

"I had afterwards an opportunity of inspecting the parts; and found in each a stricture formed by the contraction of a transverse fold of the internal membrane of the oesophagus, exactly behind the lower edge of the cricoid cartilage. In two of the cases, the orifice of the stricture would admit a crow quill; and in the third a goose quill could be passed through it. The disease was wholly confined to this septum, and scarcely exceeded a tenth of an inch in thickness. The internal membrane both above and below the stricture was more than usually vascular but free from any other appearance of disease." Home's illustration of one of his postmortem specimens is reproduced in the figure.

Munro (1811) recapitulated Baillie's and Home's descriptions under the heading of "Stricture caused by a Transverse Fold of the Villous Coat of the Alimentary Canal". He goes on to say "Two strictures have been observed sometimes but much more rarely in the gullet: and I am indebted to Mr. Astley Cooper for a drawing of such a case. The strictures are at a distance of an inch from each other and the uppermost one is situated nearly opposite to the cricoid cartilage of the larynx."

Syme (1837) wrote of "simple organic stricture" that it was "chiefly at the commencement of the oesophagus opposite the cricoid cartilage. The contracted part is usually of small extent, seeming as if it was caused by drawing a thread

round the tube, and exists in various degrees of width from that of a small quill upwards." Syme goes on to say "the only method of positively ascertaining the existence, seat and degree of stricture, is to introduce a series of bougies, gradually decreased in size from that which ought to enter the oesophagus readily, if it were sound, down to that which the constriction is capable of admitting."

Further anatomical examination had to wait for the development of oesophagoscopy and radiology. Latterly some such strictures have been explored ab externo and biopsy done. There is disagreement, as noted before, whether some of these are Plummer-Vinson strictures or cricopharyngeal achalasia.

It is distressing to think that, in spite of this adequate, valid, confirmed information, the stricture through the nineteenth and twentieth centuries has still too often not been diagnosed, and the patient still accused of "nervousness". It is seriously damaging to family relationships if the husband is advised that his wife's dysphagia is imaginary or "nervous", with the implication that "if she pulled herself together" it would disappear. It is just as bad if the patient herself accepts this explanation and resigns herself to the belief that she is an inadequate person.

Even Osler (1898) referred to it as "spasm" and "oesophagismus", and in 1971 a distinguished physician suggested in print that food sticking in the throat was evidence of "globus hystericus".

The greatest difficulty and delay in diagnosis are caused by concepts still lingering in the minds of the profession that localised cervical dysphagia may be due to "hysteria", "nervousness", "spasm" or "globus hystericus". All these notions are erroneous. Spasm implies the irregular intermittent reversible tetanic contraction of a muscle. It may or may not be a proper word to use of the inability to swallow in tetanus, rabies or strychnine poisoning, but it is false in regard to other high dysphagias and particularly to this. The concept of cricopharyngeal muscle achalasia is not based on "spasm". The diagnostician needs to have in his mind a good knowledge of the normal anatomy of the cervical oesophagus, and a correct idea of the stricture, the latter only to be obtained by familiarity with the x-ray appearance. Of course, a dull patient who cannot observe and describe her own symptoms makes diagnosis very difficult, if one does not have the opportunity of seeing her eat. The barium x-rays will identify the stricture and exclude pharyngeal diverticulum. Neurological and myopathic dysphagias are easily distinguished from obstructive dysphagia. Sometimes patients with an obstruction lower down in the oesophagus or in the fundus of the stomach falsely localise the obstruction in the cervical area. One has to recognise and exclude a choking feeling in the throat due to cardiac ischaemia or arrhythmias or acute thyroiditis. Simple goitre rarely, if ever, produces dysphagia. If a patient with an enlarged non-toxic thyroid complains of dysphagia, the cervical oesophagus should be examined for a possible Plummer-Vinson stricture. Hysterical dysphagia should never be diagnosed. The greatest difficulty in management is in securing follow up, and the maintenance for life of a normal iron and swallowing state. The patient needs to be co-operative. The doctor needs an intense understanding that he is responsible for the control of that which without control would be a life-long progression of disease and disability, in some cases to a very unpleasant death from either starvation or carcinoma.

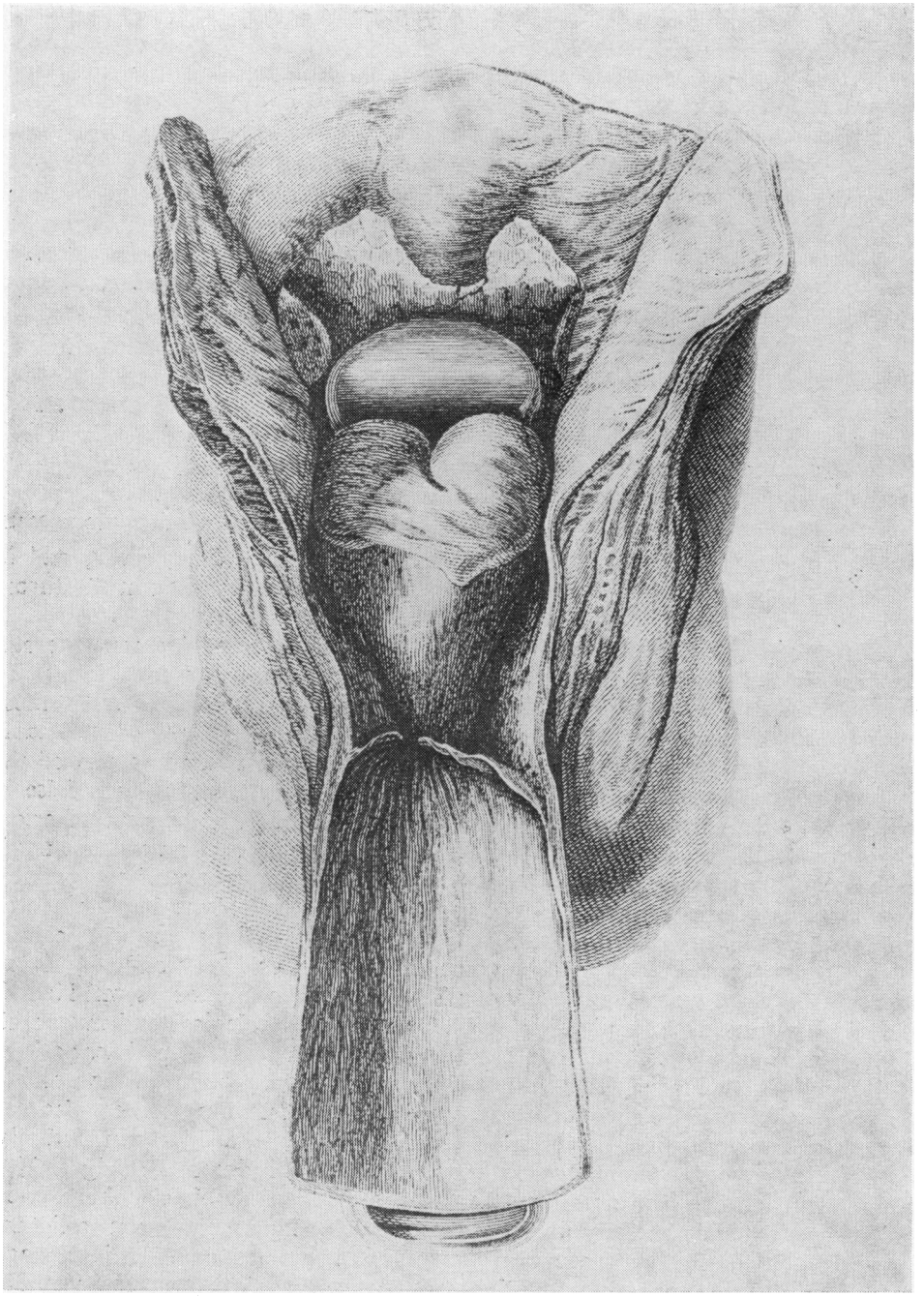

FIGURE 1: *A photograph of Everard Home's illustration of a postmortem specimen of the Plummer-Vinson stricture. "The fauces and oesophagus . . . are exposed by slitting them up from behind . . . and turning the cut edges aside." "The fauces at this part have an infundibular form; the smallest part of the funnel is directly behind the cricoid cartilage, and it is at this part the stricture has taken place, forming a membranous partition across the canal, except at one part to the left side, where there is a narrow passage through it." "The circumstances deserving of notice as they respect the mode of treating the disease, are the small degree of thickening of the coats of the oesophagus surrounding the stricture, and the healthy state of the internal membrance immediately beyond it."* (Home, E., 1803).

SUMMARY

A plea is made for the early diagnosis of the Plummer-Vinson stricture. Early diagnosis requires the diagnostician to be familiar with the morbid anatomy demonstrated by x-rays and oesophagoscopy. It requires him to have enough linguistic ability and empathy to understand what the patient is saying when she describes her symptoms. It requires him to rid his mind of mythical explanations of the symptoms, such as "nervous swallow", "spasm", "globus hystericus" and "the menopause". It requires the examination of the hypopharynx and cervical oesophagus by the barium x-ray and possibly by oesophagoscopy. When the diagnosis is made, it requires in the doctor "a passionate apprehension of process" so that after the stricture is dilated, a life-long programme, of control of the iron and other deficiencies, and of dilatation repeated when necessary, is instituted, to prevent the development of cancer. So soon as methods are available, study is necessary to see if a particular immunological process is in part responsible for the disease. A safe and effective method of preventing menorrhagia is urgently needed. The Plummer-Vinson stricture is not to be confused with cricopharyngeal muscle achalasia, and the latter needs further study to validate the concept.

ACKNOWLEDGMENTS

I am deeply indebted to the medical staff of the Royal Victoria Hospital for permission to study patients in their care and for access to the records of their x-ray and laboratory examinations.

I have received much help from the records department of the hospital and from the Librarian and staff of the Belfast Medical Library.

Mrs. Margaret Clarke of the secretarial staff has given unstinted help in all administrative arrangements and in typing the manuscript.

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