Dysphagia

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It is a great honour and an even greater pleasure to be able to thank Avery publicly for what he has done for Gastroenterology, and what he has done for me. He stimulated and encouraged my interest in a wide range of subjects, from field natural history to the design of an 'anti-reflux bed', and from burbulence to the problem-orientated record.

In 1950 E.N. Rowlands, A.J. Honour, and I made our own manometric equipment and recorder, and in 1952 we started recording pressure from the oesophagus. Avery was instrumental in the setting up of the M.R.C. Gastroenterology Unit in 1956 at Central Middlesex with Rowlands as Director. In its wisdom the M.R.C. provided radiological equipment with the then revolutionary image intensification for examining the upper gastrointestinal tract. We began to understand something about the mechanism of symptoms by getting detailed accounts of their patterns and relating these to what we observed by radiology and manometry. Avery was a great help because he very generously poured a steady stream of clinical material at us from both N.H.S. and private practice. It was this constant encouragement, with large numbers, feedback, and follow-up, that made possible the achievements of the Unit in understanding more about the mechanisms of the production of symptoms, and in the most effective use of radiology as an instrument for investigation.

Fifty years of investigation

One of Avery's earliest papers (1946) was a refresher article about dysphagia. He wrote 'The investigation of dysphagia should include a detailed history... and barium swallow; oesophagoscopy and barium meal may be necessary.' Nowadays there is a reversal of the sequence; in the U.K. everyone seems to get an endoscopy first and sometimes questions are asked afterwards. In the U.S.A. manometry follows endoscopy. Nevertheless, the major advances in diagnostic ability have come from improvements in the information gained from the history and the barium swallow. It is now feasible for example to determine most mechanisms of

dysphagia, and often the pathology, from the pattern of the symptoms as accurately as they can be determined by instrumental investigation (Edwards, 1976). This enables the clinician to use the instruments more effectively, particularly radiology, where what you observe depends very much on what you do.

Radiology can now provide immeasurably more information than 50 years ago, not only because of better equipment but also because the radiologist can have a much better idea of what he is looking for. The development of cine radiology enabled the recording and analysis of movement. Image intensification, then amplification with television systems brought movement of the barium shadow into the range of retinal cone vision and into daylight (it was rod vision after 30 min dark adaptation, and in the dark, until the late 1950s). Now video tape allows repeated viewing of the moving shadow recorded at low doses of radiation, or freezing of a chosen moment in time. Radiologists can look for evidence of power in the wall of the oesophagus, how it is organized, and roughly quantitate it. They can look for differences in compliance, or resistance to stretch and so detect areas where the tissue has changed. They can measure stricture lumen and length by using a reference grid of known size approximately in the plane of the oesophagus (Edwards, 1983). Every gastrointestinal unit should have its own simple X-ray couch and cheap domestic video tape machine.

Manometry was a useful tool when it appeared in the 1950s. Its use in the U.S.A. multiplied rapidly in the 1960s and 1970s, and in the hands of a few did much to explain and explore the normal and abnormal physiology of the pharynx and oesophagus. It has not become popular in Britain, although in North America it appears to be an essential routine diagnostic procedure. The reasons for the difference in the importance of manometry between the two cultures are complex, partly economic, partly because of differences in the amount of information that radiologists provide and in their approach to radiation hazards, partly perhaps because of different

intellectual and management approaches to diagnostic decision making. My own view is that it has been a very useful research tool, but that there are very strict limitations to its value in making diagnostic or management decisions. We should have grown out of it by now. Indeed we manage very well without it.

A major change in diagnostic procedure came with the development of the flexible fiberoptic endoscope and of suitable sedation for its use. In 1934 endoscopes were rigid and all but the most talented sword swallower needed a general anaesthetic. Morbid and mortal damage to the oesophagus was many times more common and the procedure was generally left to surgeons who could at least try and clear up their own mess. As with most things Avery was up there in front, and wrote about the use of the 'flexible oesophagoscope' in 1958 although this was a rigid tube with a flexible end. The modern truly flexible fiberoptic endoscope was developed by Hirschowitz up to 1963, and was later adapted by many others. It had some of its gestation in Avery's department where Hirschowitz was a regular visitor when he was collaborating with H.H. Hopkins (later F.R.S.), the fiberoptic physicist at Reading. The rate of change in clinical as opposed to technical progress in fiberoptic endoscopy is slowing down. The amount of necessary information which endoscopy can provide as a diagnostic instrument in the oesophagus probably reached a plateau years ago. The great advantages of the fibrescope to the physician are that he can look himself, it is incomparably safer, and does not need a general anaesthetic. Given a good history and good radiology however there is very little extra that a fibrescope can discover in the oesophagus that will modify a management decision. There is a great deal that the endoscope *cannot* tell or frequently misses, that good radiology will define. It *can* help to determine the histological type of a tumour when the biopsy contains adequate tissue, but in good hands it will still fail about 1 in 5 times to produce a positive biopsy. Endoscopy and biopsy can never *exclude* malignancy, they can only prove its presence.

Twenty-four-hour pH monitoring of the oesophagus is a more recent development of modern instrumentation. Swallowable pH electrodes were used in the stomach in the 1950s and in the oesophagus in the early 1960s. Prolonged recording from the oesophagus as a means of detecting reflux was developed in the late 1960s but was a time-consuming technique. Since the late 1970s more stable and smaller glass electrodes have become available commercially, together with portable instrumentation and 24-hr tape recording. This looks a promising research tool and perhaps a recording device for use in regional centres on patients who present with symptoms which might be, but are not clearly defined as being caused by

reflux. The advantage of the new generation of recorders is that they are comfortably worn as a small body pack by the patient while carrying out their daily chores and sleeping, and have a channel for the patient to record when they have symptoms. The equipment records the onset of a reflux episode fairly precisely; it probably cannot quantitate the volume of refluxate; and there is a dispute about how faithfully it records the hydrogen ion concentration of the microclimate of the squamous epithelium.

Fifty years of diseases

Since Avery's 1946 paper some disease processes have almost disappeared as indigent problems, for example diphtheria, syphilitic stenosis, and tuberculosis of the oesophagus. Some age-old clinical entities are now better understood, and some probably age-old disorders have since been recognized. Surgeons with new operations have created several new complications and at least one new iatrogenic disease problem has developed. Some of these are discussed below.

'Cricopharyngeal spasm' was not generally recognized in 1934 and later had many alternative names such as "hypopharyngeal bar" and "cricopharyngeal achalasia". Recently Cruse *et al.* (1979) have shown it to be not a spasm but an increase in the resistance to stretch of the cricopharyngeus muscle as a result of myositis which proceeds to loss of muscle fibres and much local fibrosis. The remaining muscle fibres seem to contract and relax appropriately. Although stretching it with a bougie may help a little, the most satisfactory treatment is to cut the cricopharyngeus muscle.

The cricopharyngeus has been the subject of other misunderstandings. It has been fashionable to attribute the development of a pharyngeal pouch to obstruction caused by inappropriately timed contraction of the muscle, based largely on manometric records which are better interpreted as showing the result of the pouch rather than its cause. Patients with fibrosis of the cricopharyngeus and those with other obstructions distal to Killian's triangle do not develop a pouch more often than those without obstruction, and cutting the muscle does not stop recurrence of pouch.

'Reflex cricopharyngeal spasm' is supposed to be caused by hiatal herniation or the effects of reflux upon the mucosa of the lower oesophagus. The concept purports to explain two sensations, (1) the sensation that a bolus sticks at the cricopharyngeus level while the subject with a peptic stricture at the lower end of the oesophagus is eating, and (2) the sensation of a 'lump in the throat' or that food has stuck or is sitting in the neck, which occurs between meals and usually disappears during eating and

drinking. The first sensation is referred from the site of obstruction. It develops at the moment of impact of the bolus on the stricture and not as the bolus passes the cricopharyngeus, which contracts, relaxes, and stretches normally when viewed radiologically with a solid radiopaque bolus. The mechanism of the second sensation is not understood but is likely to be referred. Again the muscle behaves normally and there is no abnormal sensation during swallowing. Both these misconceptions arise from lack of precision in taking the history.

Sideropenic dysphagia (Plummer-Vinson-Paterson-Brown Kelly syndrome) is now subdivided into:

(a) dysphagia caused by a sore tongue, mouth and pharynx associated with gross, long-standing, iron-deficiency anaemia, but not associated with obstruction. This is what Plummer and Vinson (1922) initially described.

(b) what appears to be submucosal fibrosis, possibly from bleeding from the post-cricoid venous plexus, causing increased resistance to stretch in a ring just distal to the cricopharyngeus, sometimes at a second ring about 12 mm distal, and sometimes with submucosal fibrosis between, producing a 12 to 15 mm long tough but elastic constriction. The two rings of increased resistance to stretch are responsible for the appearance of a web. They are not present as epithelial ridges post-mortem and are visible only when the lumen is distended. Their association with sideropenia seems fortuitous and is inconsistent.

Avery rightly noted in 1946 that the sideropenic dysphagia quickly responded to iron and vitamin B. The obstructive dysphagia does not respond, and usually requires repeated dilatation. In Sweden the obstructive dysphagia is sometimes treated by incising the muscle and fibrous layer down to the mucosa over the zone of constriction with good results.

'Vallecular dysphagia' or 'selective glossopharyngeal palsy'. Avery in 1946 mentions 'vallecular dysphagia...' in which the only abnormality found has been a delay in emptying of the valleculae. This may be the same problem as 'selective glossopharyngeal palsy' (Edwards, 1982a) whose recently described characteristic features are that the back of the tongue is unable to form a bolus and propel it into the pharynx as part of the initiation of a swallowing sequence. Neurological examination does not reveal any defect, the tongue can be rolled and moved rapidly backwards and forwards, there is no dysarthria, but nasal and laryngeal spill commonly occur during eating and drinking. Many patients cannot drink but sip and swallow a few millilitres at a time; most cannot manage solids and have to live on slops. The hypopharyngeal and cricopharyngeal muscles appear to be normal. The motor defect is easily seen by radiology in the lateral view as the subject tries to

swallow bread mixed with barium but the disability may not be noticed if liquid barium is used.

'Tender oesophagus' A pseudo-obstruction with real dysphagia has recently been described (Edwards, 1974). There is no evidence of its previous recognition as an entity and its manifestations were probably attributed to muscle spasm. The characteristic features are that food and drink provoke a sensation that they are being obstructed as they go down the oesophagus. The larger or more viscous the bolus or the faster the patient drinks, the more intense the sensation, which commonly amounts to pain, and may be so unpleasant that the patient voluntarily regurgitates. The sensation may persist for minutes or hours after the bolus has passed. The compliance and the motility of the wall of the oesophagus are normal, radiological and endoscopic examinations yield normal results, but the wall is locally sensitive to distention, demonstrated by inflating a 20 mm diameter balloon at 2 cm intervals up the oesophagus. This sensation may be stimulated by distension to 10 mm, and is always elicited by distension to 20 mm diameter.

Distension to 20 mm diameter does not stimulate a sensation in a normal subject, nor in those parts of the patient's oesophagus which are not affected. The aetiology and treatment are unknown. Many sufferers live on liquids. The condition is not associated with reflux oesophagitis.

One new 'disease' process is gaining importance and recognition, namely corrosive damage to the normal oesophagus by tablets. Tablets greater than 10 mm diameter tend to hesitate or sit above the indentations of the normal oesophagus caused by the arch of the aorta, and less commonly, that caused by the left bronchus. The larger the tablet or capsule the more likely it is to stay until it disintegrates, unless washed down with a large draught of water.

Substances in tablet form known to produce oesophagitis in the normal oesophagus include oxytetracycline, doxycycline, emepronium (Cetiprin) and potassium salts. The two latter are known to cause stricturing in addition to producing a self-perpetuating lesion. I know of three patients in whom emepronium produced such a violent reaction of the germinal layer of the squamous epithelium that the cells were mistaken for those of squamous carcinoma.

When we measured the diameter of the lumen to a solid bolus of 400 consecutive benign strictures, about half were 6 mm or less. Very few tablets are as small as this, and when a tablet is obstructed by a stricture it is uncommon for the swallower to be aware of the obstruction. Tablets which release irritant material, such as ferrous sulphate or succinate, and diuretics containing potassium supplements, are a common cause of the perpetuation or the

sudden worsening of the dysphagia from a peptic stricture in spite of anti-reflux and anti-acid measures. Alprenolol and propranolol may burn if they sit on a stricture.

Hiatal-hernia-reflux syndrome. It may seem curious that such a common condition as peptic oesophagitis was not described as a pathological entity until 1934 and as a clinical problem until 1935, although both gross ulceration and stenosis had been recognized as rarities, usually at autopsy. Hiatal herniation was described in 1932 by von Bergmann. It was thought to be uncommon and one of several kinds of diaphragmatic hernia. In 1946 Avery wrote a correct concise clinical description of the syndrome of reflux oesophagitis, but at that time it was still erroneously thought to be a consequence of a 'congenitally short oesophagus', rather than an acquired condition.

He commented (1946). 'It (peptic oesophagitis) is less uncommon than has previously been thought, and it is invariably misdiagnosed. It must be in the clinician's mind, and a special request given to the radiologist, for it is easily missed unless a special technique is employed'. These words are as true now as then and although special techniques have been developed which allow easy demonstration of the capacity to herniate stomach and the breakdown of the anti-reflux mechanism, they are still not well understood and not widely used. Although peptic oesophagitis is one of the commonest causes of dyspepsia there is still much confusion about its diagnosis and management (Edwards, 1982b). This may be partly because:

- (a) symptoms arise only when both the refluxate is sufficiently stimulating and the squamous mucosa is sufficiently sensitive to be stimulated;
- (b) the presence and signs of damage to the mucosa depend not only on the corrosive quality of the refluxate as well as the frequency and quantity of reflux, but also upon the sensitivity of the mucosa to damage (which is not the same as the sensitivity to producing a sensation);
- (c) some reflux is normal, that is, physiological and occurs in all of us after meals; it may but does not usually cause symptoms or damage to the mucosa;
- (d) the capacity to herniate stomach, and the capacity for abnormal reflux, are common but do not result in symptoms or damage to all people, nor all of the time, because criteria (a) and (b) above are not always met;
- (e) physiological reflux cannot be demonstrated by radiology; the capacity for abnormal reflux is not always demonstrable by radiology; and it is the size of the enlarged oesophageal hiatus which determines how easily abnormal reflux occurs.

It is therefore possible to have reflux with or without symptoms and with or without signs; and to

have herniation with or without abnormal reflux. Herniation by itself does not cause symptoms.

Dysphagia as a complication of oesophagitis is still a relatively rare event and the incidence must be much less than one per hundred sufferers from oesophagitis. It is not yet possible to predict which patient will develop a stricture, and many strictures develop without much warning, particularly in the elderly. A policy of operative repair of all hiatal herniae larger than a certain size would be likely to kill more patients than the number of strictures it prevented.

The outlook for the patient with stricture has changed dramatically in the last 5 to 10 years. Radiology is now capable of distinguishing benign peptic stricture from carcinoma with considerable accuracy. H_2 receptor blockade together with frequent small doses of antacid seems capable of slowing down, but not reversing, the corrosive process leading to stricture. The Celestin bougie and the new fixed diameter inflatable dilators seem less likely to damage the struggling epithelium as it tries to re-epithelialize the circumferential ulceration. The Angelchik ring prosthesis reduces the amount of hiatal flow and reflux by a worthwhile amount considering the very low morbidity currently associated with its insertion. If it is safe in the long term it could be a valuable alternative to the established methods of hiatal and hernial repair, whose morbidity and average success rate, in the hands of those performing the operations in Britain, leave much to be desired.

Achalasia is a disorder which illustrates well the effects of ideas, concepts, diagnostic methods and changes in anaesthetic and post-operative care on the outcome for the patient. When Avery qualified in 1934, Hurst and Rake (1930) had just suggested that the fault in achalasia was loss of the post-ganglionic neurone in Auerbach's plexus, causing failure of relaxation of the cardiac sphincter. In spite of much experimental work we have not materially advanced beyond this concept. Hurst advocated daily stretching of the sphincter by a swallowed, mercury weighted, flexible rubber bougie, up to about 12 mm diameter. Others realised that this had a very limited useful effect but that brusque dilatation to about 40 mm diameter was more effective. Avery favoured the Starck dilator and had reasonable success with it for more than 20 years. In the early 1940s Avery took up the newly developed octyl nitrite capsule and inhaler developed by British Drug Houses and worked hard to keep it on the market for many years after the drug company felt they were not selling enough to justify its continued production. It would still be useful.

Several operative procedures had been devised including vagotomy, sympathectomy, instrumental and finger dilatation through a gastrotomy, and

destruction or bypass of the sphincter by excision or by cardioplasty or gastro-oesophagostomy. These resulted in reflux oesophagitis and stricture, worse problems than achalasia. Heller first performed his myotomy in 1914 but although his results were good the operation was not well received until the 1920s. Rodney Maingot, writing in this journal in 1944 and 1949, extolled the advantages of myotomy and drew attention to the disadvantages of the other sphincter-damaging procedures, thoughts echoed by Barrett and Franklin in 1949. The development in the 1940s and 50s of anaesthetic methods which made intra-thoracic operations safe helped to popularize the myotomy. Dilatation is moderately successful. It depends on tearing apart the circular fibres of the sphincter, so it may not succeed, or it may tear the full thickness of the wall of the oesophagus in about 3 to 5% of dilatations. A myotomy in the hands of a few surgeons gives excellent results in the relief of dysphagia with less than a 5% incidence of reflux oesophagitis and stricture. In the hands of many surgeons the incidence of postoperative problems may be much greater. Recent increases in understanding of achalasia and of the anti-reflux mechanism from radiological and manometric studies, suggest that the important points are (1) to cut all the circular fibres of the sphincter, but none on the stomach and not more than 10 mm length of those above the sphincter; and (2) not to disturb the attachments of the oesophagus to the hiatus and to sew back any displacements made. It is not the sphincter which stops the development of reflux oesophagitis, but the hiatal mechanism which should therefore not be disturbed. This also explains why reflux problems tend to be more frequent after a trans-abdominal than after a trans-thoracic myotomy. Adding an anti-reflux procedure to unnecessarily vigorous surgery may reduce the amount or frequency of reflux problems, but introduces a significant morbidity of its own. More information is needed about the important do's and don'ts in an operation which can return eating and drinking to 95% normality.

In his 1946 paper Avery includes 'tertiary contractions with functional diverticula', a condition which had been recently described by the radiologist, Templeton in 1944. This seems likely to have been one of the many variants of achalasia which now seem to include 'symptomatic diffuse oesophageal spasm' and those examples of 'corkscrew oesophagus' or 'pseudodiverticulosis' which have dysphagia. The sinuous, even moniliform outline seen on the X-ray film is but a frozen moment in time; the next would show the contractions in a different place. The incompletely relaxing sphincter is the cause of dysphagia, not the rings of contraction in the body of the oesophagus. The dysphagia is relieved by cardio-

myotomy as in achalasia. Spontaneous pain is common in both achalasia and 'diffuse spasms' and is usually reduced in both severity and frequency by myotomy but not cured. Its cause is still unknown.

Manometry was introduced as a diagnostic tool in the late 1950s. In radiologically well defined examples of achalasia the characteristic features of the manometric record are absence of peristaltic waves and failure of the sphincter to relax. The absence of peristalsis is a technical consequence of recording from several sensors in the same column of liquid in the obstructed, incompletely emptied oesophagus. Commonly some peristaltic activity persists in achalasia but is too inefficient to empty the lumen against the obstruction at the sphincter. Sometimes the sphincter partially relaxes. The distinction between achalasia and symptomatic diffuse oesophageal spasm seems to be man-made on the basis of patterns on the manometric record which are in part artefacts of the technique. When observed by a suitable radiological technique, achalasia and diffuse spasm seem to be part of the spectrum of a single physiological disorder or God-given disease. This is one of many examples where the technique of manometry has dominated a research or diagnostic field, giving results whose interpretation has been incorrect because the observer has not been able to see what was happening at the same time. Radiology is a better tool for investigating motility disorders.

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