The Longitudinal Muscle in Esophageal Disease

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Dedication

To my darling Do.

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Although the conclusions recorded here evolved over many years, they were not reduced to writing until I served as a consultant with the Tomah, Wisconsin VAMC. Needless to say, the opinions expressed are entirely my own.

Abbreviations

CB Captive bolus

GE Gastroesophageal

GER Gastroesophageal reflux

CDR Cannon-Dougherty reflex

CM Circular muscle

CMC Circular muscle contraction

dC de Carvalho test

DES Diffuse esophageal spasm

EMD Esophageal motor disturbance

EPD Epiphrenic diverticulum

HH Hiatal transtract ("Hiatus hernia")

LER Lower esophageal (Schatzki) ring

LES Lower esophageal sphincter

LESP Lower esophageal sphincter pressure

LM Longitudinal muscle

LMC Longitudinal muscle contraction

LMT Longitudinal muscle tone (or tension)

p-wave Peristaltic wave

PEL Phreno-esophageal ligament

PVS Plummer-Vinson syndrome

SHH Sliding hiatus hernia

TC Tertiary contractions

UES Upper esophageal sphincter

UESP Upper esophageal sphincter pressure

Preface

"Pythagoras," said Socrates, "wrote that the ancient Phoenicians cooked eggs by placing them in slings and whirling them about their heads. Nevertheless, we observe that today, although we have eggs and slings and strong men to whirl them, the eggs do not cook. On the contrary, if the eggs start out hot, whirling quickly cools them. Obviously this is because we are not ancient Phoenicians!"

The literature of the esophagus is vast, but a large amount of it is devoted to shoring up the misconceptions of internationally recognized authorities - as though Socrates' students had missed his irony. This book is different. My intention is to refute the errors and place the field on a logical, not authoritarian, foundation.

Its substance evolved from a single insight: a lower esophageal ring and the dysphagia it caused cleared after the patient's stomach was pulled down and anchored to the abdominal wall. It seemed that the cure could only be explained by postulating that the ring was an accordion pleat of redundant mucosa formed when contraction of the longitudinal muscle shortened the esophagus. Further observations bore out this assumption in elaborate detail and aroused my interest in the function of the longitudinal muscle itself.

The LM could interact with the other muscular components of the esophagus in many ways to accomplish its several functions. To create some order in the bewildering array of possibilities, a Boolean logical approach was used. The states of the organ could be related to the vertices of a 3-cube according to the joint contraction or relaxation of its three components. This had the advantage of forcing my thinking into systematic channels and eventually proved a veritable secret weapon for producing hypotheses for subsequent verification or, more commonly, rejection. With few exceptions, my testing facility was the daily observation of patients in a radiology practice.

With two tools - a logical model and a reliable method of deciding when the LM was contracted - the rest of the work became the task of integrating the LM into esophageal physiology. That this had not been done earlier in any systematic way is due to the 2-dimensional outlook of the instruments of the physiologist: they see only the circular muscle and the sphincter. Of 500+ references cited here, I could find only 2% that were concerned with longitudinal muscle function.

There in an incalculable gain in insight when an extra dimension is embraced. Boyle would have faced an impossible task in formulating his gas law if, despite sophisticated instrumentation for measuring temperature and volume, he was deprived of any measure of pressure. Even worse, if he could not comprehend the idea of pressure. Imagine a Flatlander imprisoned by a circumscribed line because he could not conceive of the vertical dimension. What a liberating revelation it would be if he were given that ability. If we were given the ability to

function in 5 dimensions, who knows what understanding and power might be ours.

Unlike statistics, Boolean methods can spell out the answers. Because they employ digital logic to test for truth or falsity, a wrong hypothesis can be rejected on the basis of one counter example. On the other hand, one can often learn more from a single case report than from a multi-university population study. My approach, therefore, has been logical and phenomenological.

I have tried to tell the story twice: the logic in the text and the lore in the illustrations. The latter are the shortest route to understanding the concepts. The reader may note that I have not presented statistical studies. I have, of course, relied on those of others and sought to rationalize them with each other. However, confronted with a group of similar but not identical cases, my penchant was to analyze the differences rather than tabulate the similarities.

Flashes of insight can also be blinding. I fear that, with considerable territory to cover, I may have overlooked the obvious or gone overboard. I have no hope that all of the concepts put forth here will survive the scrutiny, not only my radiological peers, but of interested parties in the fields of gastroenterology, physiology, surgery and laboratory investigation. I will be content if the work is successful in attracting their interest to the third dimension of esophageal physiology.

A Boolean model of the esophagus

[Please refer to hard copy for diagrams.]

The purpose of a model is to reduce the real thing to something simple enough to be studied free of obscuring detail. This model of the esophagus, as an extreme simplification, reduces it to three elements, the circular, sphincter and longitudinal muscles. The superior constrictor will be ignored. Subsequently I will show that both longitudinal and circular muscle contraction can be either peristaltic or en masse, however this too will be ignored in the model.

Each element can be in one of two states, "OFF" or "ON"- relaxed or contracted. These states can be symbolized by the letters C, L, and S for the contracted state of the circular, longitudinal and sphincter muscles and ~C,~L,~S and for their corresponding relaxed states.

The status of the esophagus at any instant in time can be specified by giving the state of each muscle. These specifications are achieved by "and-ing" the three symbols or their negations. Thus, the normal condition of the esophagus is ~C&~L&S - that is, the longitudinal and circular muscles are relaxed and the sphincter is contracted.

Detecting the formula for the resting esophagus is easy because it stands still while we are doing so. We can tell that the sphincter is closed because there is no reflux, that the LM is relaxed because there is no shortening, hiatal herniation or tenting of the diaphragm or PEL. The circular muscle state is less obvious, but with the fluoroscope we can see that there is no peristalsis going on, a swallow of barium initially meets no resistance when discharged from the hypopharynx. Manometrically the pressure is zero or negative.

Even at this point, the insight provided by a model enables one to ask some questions, the answers to which, if they could be found, would not be trivial. During deglutition the state formula must change from the resting formula. Knowing the exact sequence in which the state formulas changed during this function of the esophagus would be interesting. Obviously, there is no reason to exclude L, the state of the longitudinal muscle, from consideration.

Before attempting to trace the changes in the state formula, a further simplification will be employed as it will give direction to the search. We can assume they are there, drop the ampersands and write the resting formula as ~C~LS. In addition, because its order in the formula already identifies the muscle, we can use the binary numbers 0 and 1 to stand for the relaxed and contracted states of the components. Thus simplified, the resting state formula becomes 001.

If these binary numbers are then treated as the x, y and z coordinates of a point in space,(1) they define the 8 vertices of a unit cube. This produces a major simplification at once. We can require that only one of the three muscles changes its state at a time. This is equivalent to a rule restricting the "state paths" to those from one vertex to an adjacent vertex. Thus, instead of having 7 possible ways things could change from one state to another, the possibilities are reduced three. Because the only normal state node of the organ is 001, a second rule is that all paths on the 3-D cube must be closed.

This rules are useful if one is going to trace the state path with a fluoroscope. For example, if we want to learn which is the first change from the resting state there are only three possibilities: the relaxed LM can contract, the contracted sphincter can relax or the relaxed circular muscle can contract. Only one muscle changes at a time. The other muscle components continue as before.

The problem is now reduced to deciding what happens first. That done, one is ready to look for the next change, one component at a time. Tracing the entire path in a given subject is not even necessary. It is enough to work out one transition. The next can be worked out at liesure. A directed observation is far easier and more likely to be accurate than sizing up the esophageal gestalt.

In addition, the longitudinal muscle (L) now becomes an essential part of the picture at least as important as the other components. Simply because it does not affect a manometer or a balloon is no reason for ignoring it.

Another consequence of the model is to emphasize that the esophagus is not a single-purpose organ designed only for swallowing. This is because there are a great many paths from vertex to vertex that can be traced on a 3-dimensional cube. Here is a clue to the ability of the organ to swallow liquids via a different path than it uses for solids or when swallowing against resistance or in the upside down position. We can now see that esophageal speech might have a still different path. Deploying its resources in varied sequences is the means by which the esophagus carries out its multiple functions - swallowing liquids and solids, belching, gagging, vomiting.

In principal, it would be also be anticipated that various malfunctions could cause interruptions of the smooth transitions along normal pathways or that arrests could occur - possibly at different vertex of the cube. When these very specific questions have been answered, we should have a much more detailed knowledge of the diverse functions of the organ.

Fluoroscopic observation is generally sufficient to map the paths of changing state patterns with the aid of this search algorithm. In principle, however, it should be possible to develop instrumentation to detect all three types of muscle contraction simultaneously. A computer could constantly monitor the changing patterns with a high degree of accuracy.

I will trace the state pattern in several modes of esophageal activity. By treating the 3-bit binary coordinates of the vertices as octal numbers and using them as labels for the vertices they can be referred to more conveniently. Thus, 111 becomes 7, 001 translates to 1, etc.

The "resting" node, from which all evolutions of the esophagus start and end, is 1 (001). That is, the sphincter is closed preventing reflux and the circular and longitudinal muscles are relaxed. In swallowing liquids, we note that the circular muscle - initially relaxed - does not contract as a whole or by peristalsis (except on final cleanup). The initial change, then, must be in either L or S. We can note a slight but consistent upward movement of the diaphragm preceding each spurt of barium into the stomach.

The contraction of L changes the formula to ~CLS or 011 = 3 and the subsequent sphincter relaxation to ~L (010) = 2. A rapid alternation among nodes 1, 2 and 3 then occurs with each subsequent swallow. The graph never gets off the not-C plane.

Swallowing against resistance, whether it be with solid food, in the upside down position or during a Valsalva effort causes the path to leave the left (not-C) plane and involve the circular muscle. The first event is active peristalsis (001 to 101). Peristalsis gradually "latches" the longitudinal muscle (101 to 111) causing the sphincter to yield (110). Just before the peristaltic wave reaches the sphincter, the longitudinal muscle relaxes (110 to 100), the ring of circular muscle contraction becomes the sphincter (100 to 101) and so vanishes (101 to 001). The path traced is 1, 5, 7, 6, 4, 5, 1.

As there are three possible transitions from a given vertex, and swallowing involves 6 transitions, there are 33 or 729 possible sequences of which only one is appropriate to swallowing against resistance.

Comparison of the graphs of the two swallowing modes shows strikingly different pathways. Note that the state formula changes one bit at a time, and that it is difficult to leave the designated path without the swallow aborting. Both activities are highly mechanical acts that the model represents quite faithfully. One might say that LMC is essentially concurrent with the peristaltic wave and ought to be represented as a path on the diagonal from vertex 1 to vertex 7. However, it seems more helpful to think of paths confined to the edges of the cube between vertices even if close temporal relationships or overlapping activity warp the picture.

For belching the following schema is easily detected:

Formula

Node

Esophageal state

~C&~L&S

001

Resting state

~C&L&S

011

Longitudinal muscle contracts

~C&L&~S

010

Sphincter opens, gas escapes

~C&~L&~S

000

Longitudinal muscle relaxes

~C&~L&S

001

Sphincter closes

C&&S

101

Circular muscle evacuates gas

&&S

001

CM relaxes to resting state

It is evident from inspection of the various state-path diagrams that they are not restricted to a single plane but move in all 3 "dimensions." This highlights a remarkable aspect of esophageal physiology to date: it has been largely restricted to the CS plane. That is, it is fundamentally 2-dimensional.

If this handicap is accepted, we are as limited in our comprehension as were the Flatlanders of Edward Abbott,(2) who could be imprisoned by circumscribing them with a line because they could not conceive of a third dimension. The available balloons, manometers and transducers are Flatlander instrumentsas they see only the circular and sphincter muscles.

This has serious consequences even for careful laboratory research. No matter how meticulously LES pressure and peristaltic wave pressure are measured or sequenced under various experimental conditions, the interactions between stimulus and response may be happening in a plane that is invisible to the experimenter who is not also tracking LM function. Unfortunately, instrumentation has not been developed to measure LM contraction conveniently in the intact subject.

Another aspect of esophageal physiology that can be read from the model is the fact that, with one exception, the organ normally never remains indefinitely at a node. It occupies them only in transit. The exception, of course, is the resting state - (001). Permanent occupation of another vertex is pathologic. Myotonia dystrophica, for example, seems to arrest the organ at 010 as does scleroderma.

The various formulas that occur can be listed and "or-ed" together and so manipulated with operations of the sentential calculus of mathematical logic(3) to yield a result that I will subsequently show to be supported by radiological and clinical observations. It might be called the Fundamental Law of the Esophagus:

~S <-->L

That is, the sphincter is open if and only if the LM is contracted.

SUMMARY

Because it ignores the function of the longitudinal muscle, the prevailing esophageal paradigm is 2-dimensional. A 3-D model reveals several rules of esophageal physiology, the most important of which is the rule that the longitudinal muscle and the sphincter are never contracted simultaneously. This is equivalent to the assertion that the LM opens the sphincter. The vastly increased number of possible state sequences together with alternative peristaltic and en masse modes of contraction account for the ability of the esophagus to carry out a variety of functions.

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The mechanism of mucosal fold formation

Although I am primarily concerned with the muscle of the esophagus, some of the things to be proved will be derived from the fold pattern of the organ. A correct understanding of fold formation and its significance is, therefore, essential.

Historical background

Since the early years of radiology, the "autoplastic" theory of mucosal folds formation has had the status of dogma. This theory was enunciated by Forssell,(1) the founder and for many years editor of Acta Radiologica, and given currency in the 1940s in Templeton's(2)

influential textbook. Forssell was impressed that mucosal folds about a cube of meat in a cadaver stomach seemed to grip the food as though to manipulate it and theorized that mucosal folds came about by contraction of the muscularis mucosae.

Forsell's theory was widely accepted and it would be fair to say that it is still unchallenged. As recently as 1983, Eastwood(3)

undoubtedly stated the conventional wisdom when he wrote: "The longitudinal folds in the lower esophagus are due to contraction of the muscularis mucosae." On the other hand, Levine and Laufer(4) (1992) use these fibers to explain transverse folds in the esophagus. Olmstad(5),(6) echos these views in 1994.

Unfortunately, the autoplastic theory is a hypothesis that leads nowhere. If the theory is correct, no conclusions could be drawn from the fold patterns of the alimentary tract. Fold thickening or thinning could only be interpreted as variations in the activity of the m. mucosae. An increase or decrease in the number of folds would have no particular significance other than suggesting that a different type of m. mucosal activity was present. Fold direction becomes meaningless.(7)

To be sure, radiologists pay little more than lip service to the theory, drawing diagnostic inferences from fold patterns as their collective experience indicates. Nevertheless, the discrepancy between theory and practice creates difficulties.

These difficulties are not ameliorated by endoscopists whose methods do not refute the autoplastic theory. With the organ distended, the enlarged folds seen by radiologists often were not confirmed by the endoscopist. This could not help but be embarrassing and inevitably led radiologists to distrust their own findings and even refuse to draw inferences from fold size or numbers.

This reticence was not lost on the radiological audience. A professor of medicine(8), speaking as one of a "think tank" of experts on the esophagogastric junction could say without contradiction by the other members of the symposium that ". . . . neither [gastritis or esophagitis] is diagnosable radiologically."

Endoscopy also underdiagnoses. A recent study(9) (the endoscopists were unaware it was being done) showed that although endoscopy missed only 17% of histologically verified instances of duodenitis, 68% of the antral gastritis was not diagnosed by inspection. The fact that the stomach is distended for the procedure while the duodenum, presenting no distal barrier to the escape of gas, is less easily distended, explains the different percentages: when an organ is distended, the folds disappear.

Flaws in the autoplastic theory

If the m. mucosae caused folds, it could only produce them by contracting. If it contracted en masse, this could conceivably cause an arching of the mucosa above and at right angles to the long axis of the contracting cells just as tightening a bowstring arches the bow. As it turns out, in the esophagus the cells of the muscularis mucosae are aligned exclusively in the longitudinal direction.(10),(11)

The autoplastic theory, therefore, predicts that esophageal folds will be exclusively transverse! In fact, of course, esophageal folds are longitudinal - exactly the opposite of the prediction. If, in unusual circumstances, a transverse fold does form, we are unable to explain why the usual constraints are in abeyance.

When the stomachs of cats were surgically removed and immediately frozen in liquid nitrogen, sections of the folds showed that the smooth muscle layer of the m. mucosae simply followed the fold parallel to its surface instead of extending directly across the base of the fold bowstring fashion as would be predicted by the autoplastic theory.

One would also have to consider the possibility that folds could form in a way analogous to the mechanism by which an inchworm folds its body by muscular contraction. However, this analogy will not hold: the inchworm has muscle layers on opposite sides of a body 4-6 mm thick. Because the m. mucosae cells are separated from each other by microns, there is no lever arm that could exert a folding force.

Finally, it is hardly reasonable to attribute fold formation to a microscopic muscle layer when there are macroscopic muscle layers that have not been taken into account.

A demonstration that mucosal folds are caused, not by the muscularis mucosae, but by the muscularis propria has immediate practical consequences. It brings theory into accord with observation. It makes appearances intelligible. It offers predictions that can be verified. It aids in the analysis of new observations. It leads to rejection of the ad hoc assumptions that must be used presently to effect some sort of concordance between autoplastic dogma and the way things look - assumptions that tend to take on a life of their own. Best of all, a correct theory makes gastrointestinal radiology less of an enigma to the student.

A new theory of fold formation

The proof that the circular muscle of the gut is the cause of longitudinal folds arises out of the circumstance that when a hollow organ contracts the cross sectional area of its lumen decreases to zero. Because the mucosa is not perfectly elastic, its circumferential length cannot decrease to zero. The excess length is taken up by redundant fold formation.

This inescapable fact allows us to set up equations that can be solved analytically to yield a formula that predicts the number of folds that will form with a lumen-obliterating contraction. The derivation is detailed in the Appendix. For practical purposes, the following simplified form of the result is adequate:

Where T = the thickness of the mucosa, R = the radius of the organ and E = a decimal fraction for elasticity. Both T and R are measured while the circular muscle is relaxed.

From the formula, several conclusions can be drawn:

The need to take up the free mucosal surface is both a necessary and sufficient cause of mucosal folds. Contraction of the muscularis mucosae is a redundant postulate. The folds would form even if the m. mucosa did not exist.

The number of folds is proportional to the resting radius. That is, the larger the organ, ceteris paribus, the more folds it should have.

Fold number is inversely proportional to the thickness of the mucosa.

Fold number is decreased by elasticity. As the elasticity is expressed as the ratio of the mucosal circumference in the contracted state to that in the relaxed state of the circular muscle, it will always be < 1. The function of the m. mucosa, which shortens the mucosa when it contracts, therefore, is to prevent folds from forming - quite the reverse of the postulate of the autoplastic theory. That is why in the esophagus, where transverse folds would inhibit transit, the fibers run longitudinally to prevent them forming.

Pathologic states that affect the thickness and elasticity of the mucosa should reduce the number of folds while increasing their thickness. There is a reciprocal relationship between fold number and fold size - obviously, the fewer the folds, the larger they must be to encompass the width of the organ. In many ways, however, it is desirable to use fold number rather than fold size diagnostically. Although counting folds is not always easy, it is a great deal easier to quantify fold number than absolute size. The great variety of imaging media affects the measured size of the folds, borders are not always definite and magnification by the divergent beam varies a good deal.

Qualitatively, the course of fold formation can be reconstructed as follows: As the gut contracts, no folds need form until the radius of the hollow organ is reduced beyond the ability of the mucosal elasticity to take up the slack. Thereafter, the surface is thrown into folds as the increasingly redundant lining membrane adjusts to the diminishing circumference of the lumen. Finally, when the tone of the contracted organ reduces the area of the lumen to zero, the maximum number of folds will be present.

Due to the fine-structure of the cellular and stromal elements of the mucosa, it does not, except under stress, depart much from a fixed configuration. When partially distended, the fold pattern is still traceable. Folds always form in the same places as the living tissue "takes a set" if the lumen is normally collapsed.

There may well be and probably are, interrelationships among the variables, but this does not affect the validity of the formula. It is likely, for example, that inflammatory disease affects both the thickness and the elasticity of the mucosa.

In deriving the formula, it was assumed that, when the circular muscle contracts, as much mucosa is squeezed into a given cross section as is squeezed out. There are rare instances where this assumption is not valid and a progressively tighter squeeze diminishes the fold size as tissue is extruded from the area of interest. In nature, folds are often compound. No attempt was made to take this into account in the derivation, yet the prediction of fold number is extremely accurate.

Clinical correlation

Having developed a new theory to account for mucosal fold formation, it is appropriate to see how well it does at explaining or predicting the familiar clinical-radiological appearances.

In conditions that increase the thickness of the enteric mucosa the number of folds is markedly decreased - and, by the same token, their size increases. Inflammation, edema, lymphoma, and Menitrier's disease all thicken the mucosa; they are all associated with a reduction in the number and an increase in the size of the folds.

The reverse is also true. Mucosal atrophy - best visualized in the stomach - leads to many "cigarette paper" folds as would be predicted by the formula. Similar folds may occur in ulcerative colitis.

The larger organs have more folds, e.g., the stomach vs. the esophagus. Very small organs, exhibit no folds at all - the elasticity is enough to take up all the slack as the distended state is little different from the contracted state. This is also predicted analytically from the derived formula as follows:

If the thickness of the mucosa equals the radius of the organ, the numerator of the fold formula becomes zero.

therefore

That is, when the radius of the lumen is equal to the mucosal thickness, no folds will form. This is approximately true, of course, for all small tubular organs - the ureters, the eustachian tubes the fallopian tubes, the vas deferens - which, as predicted, have no longitudinal folds.

Even in the vascular system, the appearances are in accord with the prediction of our model: it predicts no folds at all! The difference in diameter between the contracted and distended states is such that elasticity can take up any redundancy that develops in diastole. Unlike the gut, the lumen of a blood vessel is not obliterated on contraction. Hence, any redundancy of the endothelium on contraction is minimal and the elasticity is enough to absorb it.

What has been said applies specifically to folds caused by contraction of the circular layer of the muscularis propria. It will be noted that the folds formed by circular muscle are orthogonal to the plane of the circular muscle fibers, i.e., are longitudinal or parallel to the long axis of the organ.

The corresponding case for the longitudinal muscle of the gut is more intuitively obvious on the one hand and on the other less susceptible to algebraic formulation. In an analogous manner, it is apparent that, as contraction of the longitudinal muscle shortens the gut beyond the ability of mucosal elasticity to take up the slack, redundancy will result. This redundancy will lead to the formation of transverse mucosal folds.

We can generalize and state categorically that mucosal folds are orthogonal to the muscularis propria fibers that produce them.

In the esophagus, our present area of interest, this new theory has immediate application. The fold pattern of the esophagus supports the theory and the theory explains the pattern. Unlike the rest of the gut, the esophagus has the singularity that it normally has no slack. Because it is attached to the skull via the pharynx above and to the diaphragm below, its ability to shorten with LMC is limited.

Consequently, in the normal organ, there are no transverse mucosal folds. Lacking such constraints, the circular muscle can produce longitudinal folds. If, due to a hiatus hernia, rupture of the phreno-esophageal ligament or marked kyphosis, the esophagus can shorten with LMC, then transverse folds - rings or webs - do form.

Looking at this interesting situation teleologically one can get a clue as to the function of the m. mucosae. It is appropriate to the function of the esophagus that it does not have transverse folds. No useful purpose served by the delay such folds would produce in transport through the organ. Folds such as webs and LERs produce dysphagia.

Lengthwise arrangement of submucosal muscle fibers also affects the other parameter of the fold equation, elasticity. The greater the elasticity, the fewer the folds. Smooth muscle fibers in the submucosa increase the elasticity of the mucosa and prevents fold formation. If the m. mucosae could produce transverse folds in the normal esophagus, it would do so. That the esophagus must shorten about 15% before transverse folds appear gives us a rough estimate of .85 for the numerical value of the elasticity, E, of esophageal mucosa.

An understanding of the mechanism of fold formation will be useful in the study of the esophagus because we can work the theory both ways and use the mucosal fold pattern to monitor continuously which component of the muscularis propria is contracted.

A compelling example of the utility of the theory is in the differential diagnosis of mechanical and paralytic ileus. In both conditions the small bowel is markedly dilated. This does not necessarily mean that the circular muscle is paralyzed or otherwise defective. The dilatation is occasioned by mere distention. However, the presence of transverse (Kerckring) folds in the typical "coiled spring" appearance signifies LMC. Correctly, we can infer that if the LM is contracting, the ileus is not paralytic.

Both radiologist and physiologist know when the circular muscle is contracting because it reduces the caliber of the lumen, produces longitudinal folds, compresses a balloon or raises intraluminal pressure. In sharp contrast, none of these indispensable signs is seen with LMC. The ability to identify which component layer of the muscular wall is contracting fluoroscopically is very useful, particularly with the LM of the esophagus.

SUMMARY

When a hollow organ contracts enough to obliterate its lumen, the mucosal lining becomes redundant. This redundancy is accommodated by the formation of folds. It can be shown analytically that the number of folds is a function of the resting size of the organ and the thickness and elasticity of the mucosa. The

direction of the folds is orthogonal to the direction of the muscle fibers of the m. propria that cause them.

Unlike the autoplastic theory, the present analysis generates testable predictions that are in exact correspondence with the radiological findings in hollow tubular organs, particularly the gut.

Once it is realized that transverse fold formation implies LMC, a new range of physiological observations becomes possible.

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The captive bolus

AND

THE PINCHCOCK AT THE DIAPHRAGM

The "captive bolus" phenomenon(1) provides the only opportunity to observe swallowing against resistance. A detailed description of what happens during a sustained Valsalva maneuver allows radiologists to satisfy to themselves that several reported "diseases" are spurious and that pouches described as functional components of the lower esophagus are simply hiatus hernias or a normal response of structures to internal pressures.

In this way, the 2-pouch theory of esophageal competence can be disproved; both prolapse of gastric mucosa into the esophagus and prolapse of esophageal mucosa into the fundus can be eliminated as radiological diagnoses; misinterpretation of easily explained appearances at the GE junction could be avoided. Among the latter are the "Saturn ring," the "wing sign" and the "arum lily" signs.

The test explains why an "empty segment" appears at and above the diaphragm. It shows that the "diaphragmatic pinchcock" is not due to the diaphragm and does not play any part in preventing GE reflux. With this traditional underbrush cleared away, it can be seen that it is the LES and only the LES that prevents reflux of gastric contents into the esophagus.

The phenomena elicited by the test cannot be explained without the phreno-esophageal ligament (PEL). The test thus proves the existence and functional role of a structure that furnishes a key to an understanding of reflux. It clearly shows the sphincer. It reveals the swallowing act in 3-d.

This is how to do it:

In the prone (right anterior oblique) position, the patient is asked to take one normal mouthful of barium, immediately after swallowing, take a deep breath, hold it and make a maximum sustained straining effort, i.e., a Valsalva maneuver.

In a patient with a "positive" test, the barium column is pinched off at and for a variable distance above the diaphragm. This constriction is the so-called "diaphragmatic pinchcock." A peristaltic wave then forms in the proximal esophagus and advances at a uniform rate of about 4 cm/second forcing barium ahead of it until it reaches the physiologic sphincter. Peristalsis continues to the lower margin of the sphincter and stops. The sphincter remains contracted either briefly or as long as the patient can hold his breath. Trapped between the

sphincter above and the "pinchcock" below is a bolus of barium. This is the captive bolus.

Experience shows that the success rate of this demonstration depends largely on the clarity, redundancy and enthusiasm with which one gives the instructions. Getting it right the first time seems important. If the patient starts straining too late and the procedure is repeated, the findings are seldom as clear-cut.

The mechanics of the captive bolus

This is what is happening: When the patient takes a deep breath the phrenoesophageal ligament (PEL) is stretched between its attachment to the diaphragm and the distal esophagus. The PEL forms a "skirt" or tent attached to the esophagus at its apex and with its lower edge flaring out to its attachment on the inferior surface of the hiatal circumference.

As the intrathoracic pressure builds with straining against a closed glottis, the fundus, drawn into the hiatus by its attachment to the esophagus, is collapsed: a.) laterally by intrathoracic pressure and b.) from below by (incompressible) abdominal tissues crowded into the base of the PEL tent by increased abdominal pressure. Because thoracic air is compressible and the tissues in the tent are not, the pressure from below wins, the tent becomes convex or dome shaped and the portion of the fundus within the tent is occluded.

This occlusion sets up the distal barrier that arrests the bolus when it arrives. Note that 1.) If the PEL is not stretched enough to form a tent, this barrier cannot be set up. 2.) If the PEL does not exist, the barrier cannot be formed. Thus we have a means of testing whether the PEL is intact but stretched or ruptured.

The obstruction at the diaphragm is not obvious until peristalsis begins distending the distal esophagus. One reason for telling the patient not to take a very large mouthful is that if the bolus is too large the circular muscle cannot bring the esophageal walls into contact and peristalsis aborts.

Assuming this is not true, a peristaltic wave will form in the cervical esophagus and push the fluid barium ahead of it. As it moves down the esophagus the lumen distends because the same amount of fluid is now contained in a shorter tube. Distention of the lumen (and circular muscle) is the stimulus to LMC,(2) so, as the peristaltic wave moves distally, the circular muscle ahead of it is progressively distended and the LM progressively contracted. A maximal distention produces a maximal LM contraction that pulls the gastric pouch farther through the hiatus.

During the test, the fundus of the stomach rises above the diaphragm synchronously with the advance of the p-wave. Despite the transtraction of the

stomach through the hiatus, no esophageal redundancy appears because LM is shortening it.

Although, judging from manometric tracings, one might think the p-wave was simply an advancing ring of CM contraction, in reality it is a complex advancing cone of CM contraction. Initially, the cone is long and gradually tapering. As it progresses distally, it becomes shorter so that by the time it reaches the sphincter it has shortened to a ring. While the region of CM actively contracting is 8-10 cm or more, a corresponding region of LM can be presumed activated.

At this point there are two almost turnip shaped "pouches" base to base. The ancients thought these were some kind of sphincter mechanism. Distinguished anatomists' names became attached to them furthering the impression that they were anatomical structures with specialized functions. It is now generally accepted, however, that the lower pouch is gastric fundus and not a "fore-stomach." The upper, of course, is the "phrenic ampulla." Unlike the "vormagen," the phrenic ampulla is always considered a separate anatomical element of the esophagus and, as such, teleological thinking presumes it should have a distinct function, presumably sphincteric.

There is a simple reason for the phrenic ampulla: it is at the end of the esophagus. A distal obstruction means that the bolus initially occupying most of the body of the organ must be contained in the distal few centimeters when the p-wave milks it there. Thus the region is passively dilated. The same segment that is now ampullary in shape can be tubular a moment later and trumpet shaped after that. When inflated by double contrast techniques, this region is no more distensible than others. Its upper margin is pointed because it is in the LES. Its opposite end is flat-topped because the plane of the LER defines it.

When the p-wave reaches this point, all activity stops. The obstruction persists, the p-wave has reached the end of its travel and the bolus is trapped between the two. That this condition may persist since the patient can maintain the Valsalva effort tells us something. The now ring-like p-wave, without any auxiliary support, is entirely capable of containing the bolus against the considerable hydraulic pressure of the tissues being forced into the tent of PER like a thumb on a bulb syringe. It has become the sphincter. By now high above the diaphragm, there are no other sphincter candidates in the region.

Lessons of the captive bolus test

These simple mechanics also explain a great deal about the GE junction, sphincters and hiatal hernias. First, it explains why there is sometimes a pinchcock-like mechanism at the diaphragm and why it is sometimes lacking: The subdiaphragmatic soft tissues crowding into the tent-like PEL collapse the tube of gastric fundus drawn into the tent by contraction of the LM. The barium column, therefore, appears "pinched" or occluded.

Some have supposed that it was the diaphragm itself that caused this constriction. However, the pinch effect is not limited to the level of the diaphragm - the diaphragm is only a few millimeters thick - but extends both above and below the diaphragm.

As Hayward has emphasized, the PEL is an exceedingly elastic structure ". . . rich in elastic fibers" that " . . . if divided near the esophagus . . . may be taken right up into the adventitia of the esophagus and vanish." If the PEL is not stretched or elongated, no tent will erect and there is no pinchcock. Therefore, a CB is itself a sign of hiatus hernia (HH) because that is what a sliding HH is - a stretched, elongated PEL. The reason it is a sliding HH, is that an intact, elastic PEL provides the reducing force. LMC stretches the PEL and pulls a tube or pouch of fundus above the hiatus; the elastic recoil of the PEL reduces the HH when LMC stops.

Of course, if the PEL is stretched beyond its elastic limits or actually ruptures, we can predict that a HH will not be self-reducing. And that is exactly what happens. The HH also can become larger because the PEL is no longer a limitation on its size. If the PEL ruptures there is no pinchcock. Abdominal soft tissues can slide into the chest alongside the herniated fundus but, as they are unconstrained, they cannot exert enough pressure to pinch off the fundus.

Is the pinchcock a defense against reflux? No. If a patient with a diaphragmatic pinchcock has a successful HH repair, the pinchcock will disappear. It is illogical, therefore, to postulate as a normal antireflux mechanism a diaphragmatic pinchcock that depends on a pathological set of circumstances and that vanishes when the pathology is removed. This tells us that the pinchcock at the diaphragm can have nothing to do with the prevention of reflux. It is simply an anatomical pattern that occurs when the PEL is stretched but not ruptured.

It is generally supposed that a Valsalva test provokes a sliding HH by increasing intra-abdominal pressure. However, it is not the increased intra-abdominal pressure per se but the maximal LMC provoked by swallowing against the resistance of the pinchcock which draws the fundus above the diaphragm.

LMC, by the shortening esophagus, causes mucosal redundancy. As there is a good deal of elasticity in esophageal mucosa, signs of redundancy do not begin to appear until the esophagus has shortened 3 to 4 cm. Thereafter, the redundant mucosa may form a fold or accordion pleat. This is the "Schatzki" ring.

Even before the fold forms, however, it is often possible to identify the mucosal junction of esophagus and stomach by 1 mm notches, sometimes called the notches of McLean,(3) at the precise location that the ring will form. Once either

is identified, it is easy to see that the esophagus is shortening as the p-wave proceeds distally and that the ring deepens as the esophagus shortens.

Why isn't the "ampulla" obliterated by the advance of the peristaltic wave? Because peristalsis stops at the sphincter.

Three results

From these observations, we can establish three results that are neither common knowledge nor consistent with early speculations as to the function of the phrenic ampulla.

The phrenic ampulla does not have any sphincter function

The ampullary dilatation is merely a ballooning of a segment that is incapable of peristalsis.

Most important, however, is this demonstration that the p-wave stops at the lower edge of the sphincter - which is not necessarily the end of the esophagus. It is well known that there is normally no peristalsis in the gastric fundus, but the existence of an aperistaltic esophageal segment, to the best of my knowledge, has not been observed either radiologically or manometrically. This fact will assume importance when we consider "achalasia."

The pinchcock may be forced at times, but usually the sphincter is first to yield so that a retrograde jet of barium squirts up into the relaxed esophagus proximal to the sphincter. The ease and rapidity (the jet may reach the height of the aortic arch in .25 seconds) with which the proximal esophagus then fills demonstrates the lack of tone in the body of the esophagus in the wake of the p-wave - as we also know from the fall in the manometric pressure tracing behind the p-wave.

Also illustrated is a unique aspect of LM contraction: unlike CM contraction, it does not relax in the wake of the p-wave. Instead, the p-wave appears to latch the LM as it passes through it. Only when the p-wave reaches the sphincter does the LM relax.

Within 1.5 seconds after the sphincter yields, the proximal esophagus is refilled from below. This presents an excellent opportunity to view the sphincter, still contracted but forced open in the initial stage of this retrograde filling. It generally measures 1 cm a few mm - far less that the 3-4.5 cm that manometry has indicated.

The bulb syringe effect of the PEL tent

The yielding of the sphincter is not due to a further advance of the p-wave, but to further encroachment on the space within the PE tent by abdominal tissues crowding into it from below. These act as a piston to create the necessary hydrostatic pressure to force the bolus back through the sphincter.

If the observer's attention is on the body of the esophagus, he may have the impression of reverse peristalsis, however, this is not so. Cine-films of this phenomenon centered on the gastric pouch(4) show that it collapses in its axial direction before its transverse dimension decreases appreciably - as though a piston were moving up from below ejecting barium ahead of it.

When tracings of successive frames of a cine strip of the process are superimposed, it can be seen that, whereas the outline of upper pouch and sphincter remains nearly constant, the outline of the gastric pouch moves proximally. Calling this a piston-like action is scarcely accurate. It is an extremely complex event with a torus of abdominal soft tissues rolling up into the PEL tent. The net effect, however, is a piston or bulb syringe-like action.

It is doubtful that this process has any significant role in, say, causing GE reflux. If it occurred during swallowing, no acid would be retrojected. It is also noteworthy that the pinchcock zone is much wider than both the diaphragm and the physiologic sphincter. 1, 2 or 3 prominent gastric folds are usually seen in the constricted area. If only one is seen, the resemblance to the bird beak, described as characteristic of achalasia, is perfect.

Locating the sphincter

The lower esophagus must exhibit either McLean notches or a LER to demonstrate that the p-wave does not extend to the end of the esophagus but stops at the lower sphincter margin. About 3 cm is the maximum distance between the ring and the lower edge of the sphincter I have encountered. Usually this distance is .5 to 1.5 cm.

One cannot help wondering why the sphincter is not at the very end of the esophagus. It is a common observation that, except in most unusual circumstances, there is no peristalsis in the fundus of the stomach. The gastric peristaltic wave starts high on the greater curvature of the stomach. That the esophageal wave stops short of the stomach would have been difficult to anticipate. There is no manometric evidence of an aperistaltic esophageal zone. The aperistaltic region may act as the opening wedge of the sphincter during belching, vomiting and reflux when it flares to its trumpet shape.

If the patient is still able to sustain the Valsalva effort, a new peristaltic wave will form and again force the regurgitated barium into the double compartment between sphincter and pinchcock. Repeated cycles of this nature may follow without so much as a drop of barium escaping through the pinchcock.

Antireflux significance

If one studies the captive bolus during its arrested phase, it is easy to see that:

There is no angle of His in sight.

The right crus of the diaphragm is well below the area of interest.

There is no subphrenic esophagus.

The area of muscular thickening at the GE junction described by Liebermann-Meffert et al.(5),(6) is 9-21 mm below the squamo-columnar junction - which, as we know, is exactly at the LER. This would place the thickening in the upper portion of the "herniated" (actually retracted) fundus where it clearly is not playing any part in the obstructing mechanism or even influencing the shape of the fundus.

The sling fibers of the stomach are also completely overpowered by other forces and nowhere in evidence.

Yet, despite the absence of any of the classical configurations invoked as antireflux mechanisms, the unaided sphincter is doing a magnificent job of remaining competent. Even the considerable pressure built up by upper abdominal tissues crowding piston-like into the confining tent of PEL does not force it.

The illusion of prolapse

Turning attention again to the incursion of a torus of abdominal soft tissues into the base of the PEL tent, we can see that as this occurs, mucosal folds appear to be moving proximally into the gastric pouch, but this is simply because its walls are coming into contact. If only the retrograde mucosal margin is noted, it will be mistaken for "retrograde prolapse of gastric mucosa"(7),(8),(9) into the esophagus unless morphological details are carefully analyzed.

Of course, all of this is happening in a relatively few seconds and, without the aid of cinefluoroscopy or a sequence camera, analysis of the process is difficult. Prolapse of esophageal mucosa into the fundus has been reported(10),(11) repeatedly, but it is very unlikely that this occurs. If it did, there would be no reason for a ring to form. For some reason, the enteric mucosa, that is so loosely attached to the wall in the esophagus, is firmly attached in the fundus. Instead, the motion of the torus into the tent simulates sliding or prolapsing mucosa - gastric when it is rising and esophageal when it is reducing.

Strange formations are seen at the end of the CB test when the pouch of fundus above the diaphragm begins to return to its normal infradiaphragmatic position.

As mentioned, the PEL provides the restoring force. It is applied at the apex, rather than the base of the retracted pouch of fundus. Therefore the soft tissues - both the fundic pouch and supporting omentum, etc. - are pushed into the abdomen. Like a crowd of people escaping a theater fire, these tissues tend to overrun the exit. The result is the interesting shapes aptly described as "arum lily," "Jack-in-the-Pulpit," "Saturn ring" or "wing sign" as the fundic pouch telescopes into itself or overhangs the diaphragm. These signs do not suggest any special disease states.

Finally, we are able to explain why a Valsalva maneuver demonstrates sliding hiatus hernias. It does so by occluding the alimentary tube thus provoking maximal peristalsis and maximal LM contraction. With normal swallowing in the upright position, LMC is minimal. With swallowing against an obstruction, however, peristalsis becomes more forcible and LM contraction acts to pull the esophagogastric tube, stocking fashion, over the bolus pulling the gastric fundus into the chest in the process. In a subsequent chapter a more extensive proof of the cause of hiatus hernias will be elaborated.

SUMMARY

A detailed study of the minutiae of the captive bolus phenomenon yields results that we can use in the proof of many propositions throughout this presentation. Among them the following:

The LM shortens as peristalsis proceeds down the esophagus.

There is an aperistaltic segment that corresponds to the phrenic ampulla and accounts for its formation.

The elasticity of the phrenoesophageal ligament is the restoring force in sliding HH. The size of the sliding HH is a measure of the stretch of which the PEL is capable.

The 2-pouch appearance is explained.

The "Saturn ring," "arum lily" and "wing signs" are stages in the reduction of a sliding HH.

Retrograde prolapse of gastric mucosa and orthograde prolapse of esophageal mucosa are misinterpretations.

The pinchcock at the diaphragm is not due to the diaphragm, is not a part of the anti-reflux mechanism and could actually cause reflux.

The physiologic sphincter is less than a third the length it is judged to be manometrically.

The LES is not only highly competent in the face of extreme retrograde pressure, but is the sole anti-reflux protection of the esophagus. It is present and functioning when there is no evidence of an angle of His, subphrenic esophageal segment, pouting of the gastric or esophageal mucosa, valve of Guberoff, etc. In the next chapter I will discuss some more fundamental misconceptions about the sphincter.

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Where is the sphincter?

Fundamental misconceptions about the lower esophageal sphincter (LES) pervade the literature of manometry.

It is widely believed there is a subphrenic esophageal segment that contains the sphincter.

It is also believed that intraabdominal pressure "backs up" the physiologic sphincter rendering it competent(1) Such statements imply that their authors believe the sphincter is in this subphrenic segment.

It is believed to be 2.5 cm or more in length.

Unfortunately, these beliefs provide the rationale for antireflux surgical procedures. It is equally unfortunate that the actual sphincter described and named by Lerche(2), that is plainly visible radiologically, has been misinterpreted(3) and even described as an obstructive lesion similar to the LER.(4) Radiologically, there is no difficulty in locating the normal sphincter: it is at the end of the peristaltic wave. The peristaltic wave does not pass through the diaphragm. Hence there could hardly be a subphrenic segment of esophagus.

As the PEL in inserted at the sphincter, the sphincter will be above the diaphragm in a patient with normal LM tone. In a sense the sphincter is the stationary end of the peristaltic wave. There is no peristalsis in the fundus of the stomach or distal to the sphincter in the .3-3 cm aperistaltic esophageal segment interposed between the sphincter and the fundus.

The manometric data are detailed, pervasive and hazardous to challenge as they are backed by immense authority. It has been done, however, although the work has been successfully ignored. In 1985 Clark(5) resected the sphincter area and replaced it with a tube of jejunum or colon in 12 Rhesus monkeys. Remarkably, a high pressure zone (HPZ) was recorded on 89.6% of the manometer tracings after jejunal interposition! Even the pattern of "receptive relaxation" during the pharyngeal phase of swallowing was recorded in many. Moreover, some of the reported pharmacological effects on the sphincter were reproduced in these animals. Clark concluded that ". . . the HPZ resulted mainly from compression of the bulky jejunum as it passed through the oblique right crus of the diaphragm."

Clark's criticism of the HPZ location of the LES is well taken and may explain the belief in a subphrenic sphincter. The manometer must be recording a squeeze of the fundus in the hiatus. This was demonstrated unintentionally by a study in which the GE junction area was simultaneously recorded by both cineradiography and manometry in 13 patients with lower esophageal rings (LER).(6) The HPZ was found to lie below the LER. In rigid conformity with manometric doctrine, the authors were forced to conclude that, "The lower

esophageal ring was found at the proximal margin of the lower esophageal sphincter in all [13] patients."

As the number of pathological examinations of LER's increases, it becomes impossible to maintain that the LER is not exactly at the junction of squamous and glandular mucosa. The above finding can only have one interpretation: the HPZ the manometer was recording was in the stomach! Examination of the reproduced cine frames shows this to be the case. Metal tipped catheters are 2.5-3 cm into a hiatus hernia and hence subject to a hiatal squeeze.

If manometry places the HPZ about 2.5 cm distal to the true GE junction, i.e. in the stomach, 100% of the time in 13 patients, we have an explanation for the manometric inference of a subdiaphragmatic esophagus: the low resolving power of the manometer could easily misplace it.

It is interesting to note in Clark's report that all of the familiar "sphincter" phenomena - reduced pressure with deglutition, pharmacologic effect of propantheline, etc. - were present at the HPZ after extirpation of the sphincter! Clark advances this as evidence that there is no LES, but it also demonstrates that something will lessen the "squeeze" pressure being measured at the HPZ whether or not it is acting on a sphincter. That something, it will be shown, is the LM. The pharmacological effects, etc. could be equally well be due to action of the drug on the LM.

Another splendid piece of evidence that manometers are not measuring the sphincter is provided by manometry itself. Using multiple catheters - as many as 8 - it has been shown that the "sphincter" pressure is asymmetrical about the radius of the lumen.(7) ,(8) It is mechanically impossible, however, for a contracting ring of muscle to exert anything but radially invariant pressure on the lumen. The multiple catheters must be measuring extrinsic pressure on the lumen, i.e., hiatal squeeze which can vary depending on proximity to the hiatus.

If , for example, one were to loop a cord around the a hollow organ as an external constraint, traction on the cord would press on only one side of the lumen. The organ would move laterally until constrained by equal pressure from the opposite side. On the other two sides the pressure would be intermediate. Intraluminal readings would vary depending on where pressure was being measured. What is this external constraint? It could only be the hiatal ring.

These conclusions introduce a perplexing question: if the HPZ is not the LES, why isn't the true sphincter being demonstrated on manometric tracings? We can see from the captive bolus phenomenon that it is a very effective sphincter and undoubtedly is capable of preventing reflux. Yet there seems to have been no manometric evidence of it! It is quite possible, however, that mere introduction of a foreign object is enough to obliterate evidence of the true sphincter. A barium

tablet arrested in the distal esophagus, for example, will provoke repeated LM contractions. Like air contrast esophograms, manometry is unphysiologic.

Liebermann-Meffert and her collaborators,(9),(10) perhaps because of their exacting technical methods, have had the last word as far as anatomical sphincters in the region. They described a gastroesophageal ring (GER) of circular smooth muscle 16.4 mm' below the descending limb of the PEL. The ring was 4.2 mm thick on the greater curvature side and a mm less on the lesser curvature side. It tapered up into the normal esophageal CM thickness of 2.1 mm over a distance of about 5 cm but tapered more abruptly distally. It was asymmetrically higher on the greater curvature side. [See their Figure 12]

As can be seen from their figure, the fibers do not run circularly but consist of short clasp-like fibers on the lesser curvature side and oblique, almost longitudinal, fibers on the greater curvature side. The axial length of the thickening was determined to be 2.3 cm on the lesser curvature side and 3.1 cm on the greater curvature, but because of the tapering, it is difficult to define the end points.

It is difficult to see what significance should be attached to this GER but the suggestion that it represents the physiologic LES must be refuted. It is 9-21 mm below the ora serrata. [We must recall that the mucosal LER is here.] It is even farther below the PEL. If all of its fibers were to contract simultaneously, they would produce a distortion of the GE junction, but not the annular constriction required for sphincter function.

It has, nevertheless, been widely accepted as the anatomical counterpart of the physiologic sphincter as defined by manometry. The acceptance derives from the fact that manometry also assigns a 3-5 cm length to the physiologic sphincter and locates it below the diaphragm, well below the PEL. This places it below the aperistaltic segment that can be demonstrated radiologically between the sphincter and the ora serrata. As this has a length of .3 to 2.5 cm, the Libermann-Meffert muscle thickening is 1.4 to 4.6 cm below the lower edge of the radiological LES - that is, well below the endpoint of the peristaltic wave and clearly in the stomach.

I cannot find any justification for supposing that the physiologic sphincter of the esophagus is in the stomach and in a region quite devoid of peristaltic activity. It is in an area that, if above the diaphragm, would be a HH!

The Libermann-Meffert GER does not make sense in physiological terms. It is physiologically and logically imperative that the p-wave pass uninterruptedly to the sphincter - passing the baton, so to speak. If there were a gap, the bolus would be lost and reflux would occur back into the body of the esophagus as soon as the advancing ring of circular muscle contraction stopped and died out.

The p-wave does stop. It then either dies out or it does not. If it dies out, it loses control of the bolus. If it does not, then it is, by definition - a stationary ring of contracted CM - a sphincter. The point where the p-wave stops is where we have to look for the sphincter. Yet Ott(11) probably expresses the consensus of current opinion when he states that the sphincter ". . . . is not a distinct muscular entity."

Actually, it has been found, accurately described, located and illustrated as a "muscular ring" in several articles and texts but misidentified as ". . . . a distinct radiologic and clinical entity." although "relatively rare." and mostly occurring in children.(12) It is Wolf's(13) "A ring." It is listed as a cause of dysphagia for which bougienage and even surgery may be appropriate therapy.(14) One shudders to think that it may even have been resected!

No one recognizes it as the sphincter(15),(16),(17) because innumerable manometric reports have conditioned even radiologists to be looking for a 3 to 5 cm constriction below the diaphragm, not the actual 9-11 mm sphincter above the diaphragm. Wu states that ". . . . some authors even dispute their [muscular rings] existence."

The captive bolus phenomenon reveals the sphincter: its location relative to the mucosal junction (.2-3 cm above), its length (8-11 mm) and its competency. It becomes apparent at the point where the p-wave stops. There it resists the considerable back-pressure from the bulb- syringe effect of the Valsalva effort. When it is forced from below by that pressure, its proximal margin is displayed in sharp contrast to the relaxed CM of the esophageal body. It then disappears as the esophagus is dilated by refluxing fluid.

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Reflex control of the sphincter- the Cannon-Dougherty reflex

Normally we are quite unconscious of the nauseating odor and the highly disagreeable taste of the gastric contents, and for this pleasant security the closed cardia is responsible.

---- Walter B. Cannon(1) 1908.

Actually, in spite of many interesting theories we are still not certain why the trapeze artist is safe in the upside down position.

---- Richard Schatzki(2) 1965

These quotations, reflecting the importance of the subject on the one hand and the unsatisfactory state of knowledge on the other, aptly epitomize the "problem" of the lower esophageal sphincter.

The problem, as commonly formulated, requires an answer to two questions:(3)

Why does the closure mechanism yield to a pressure as low as 5 mm Hg from above?

Why will intragastric pressures as high as 80 mm Hg fail to force it?(4)

Some of the solutions proposed are: a.) The angle of His, b. the valve of Guberoff, c. the sling fibers of the right crus of the diaphragm, d. the pinchcock at the diaphragm, e. the sphincter of Lerche,(5) f. the bracket bundles,(6) g. the esophageal vestibule as variously defined by Lushka,(7) Arnold,(8) and Ingelfinger, et al.(9) Lerche,(10) Hayward,(11) and others, h. pouting of the gastric mucosa by the muscularis mucosae,(12) i. differential pressure in the abdomen and thorax, j. the submerged segment or "empty segment."(13) Singly or in combination, a case can be made for many of these proposals.

The multiplicity of solutions should suggest that the basic problem has been under defined. When two linear equations in three unknowns are specified, an infinite number of solutions can be found. In the same way, the present problem is so lacking in stringency that conjecturing solutions is too easy. These are neither apt nor unique and fail when measured against the test of clinical and radiological experience. It is not difficult to show that we must feed in more conditions to be satisfied before we can test the various solutions.

Whole classes of solutions are ruled out by the circumstance that most instances of reflux are temporary, intermittent and self limiting. The ingestion of raw onions, radishes, martinis, smoked pork sausages, pizza and barbecued spareribs can

hardly alter the morphology! It is not obvious that any of the listed mechanisms can account for this simple fact.

Similarly, in instances of infantile cardioesophageal reflux,(14) when the condition is present, we can find no structural defect. When it clears spontaneously, there is no alteration in architecture. It seems, therefore, that purely morphological devices are excluded, i.e., it is improbable that any of the listed mechanisms are so configured that they could explain the phenomenology.

Perhaps, in designing or searching for the secret of the GE closure mechanism's perfection for its purpose, it has not been sufficiently recognized that we are looking at a moving target. For example, despite the difficulty of forcing the sphincter mechanism from below, it yields gently - even imperceptibly - to whatever stimulus provides for release of gas by belching. Vomiting also releases the sphincter and, in most instances at least, does not destroy it.

Control must be precise

A more subtle, but equally persuasive, objection to most of the listed mechanisms is the fact that, even were they workable, they would be too crude to account for the perfection and nicety of control that is a matter of universal experience. We can observe in ourselves and in patients that, although belching is common, the eructation of acid with the gas is uncommon. The sphincter mechanism operates as though it were a separatory funnel (in reverse) that permitted the flow of gas but not of gastric fluid contents. Observed with a fluoroscope in the standing patient, the air-fluid level in the stomach rises rapidly when the sphincter opens to permit the release of gas. Yet it closes with great precision just in time to prevent even a drop of fluid entering the esophagus.

Moreover, a mass of clinical lore concerning belching, burping (of infants), esophageal speech, vomiting and so on, some of which will be detailed later, cannot be explained by any purely morphologic mechanism proposed to date. These phenomena are so intricate and so exactly executed that one simply has to take as a starting point this postulate: There is a valve-equivalent mechanism between esophagus and stomach that, under reflex control, is open when it should be open and closed when it should be closed.

Writing about 1952, Delmas and Terracol(15) in describing the lower esophagus state unequivocally, "It should be stressed, however, that there is no muscular structure in this region, whether it be in the diaphragm itself or in the tenuous fibers of Juvera and Rouget, which can play the role of a sphincter."

Since then, modern physiologists(16),(17) maintain that there is a zone of sustained high pressure in a short segment of the distal esophagus that they believe is the sphincter. Although the sphincter they believed they were recording

was, as we have seen, illusory, there is a genuine specialized ring of circular which provides this function.

The true sphincter obviously fulfills the function of preventing reflux of gastric contents. It can be forced with difficulty from below, yet mere gravity easily opens it from above. Just as obviously, the sphincter has no morphologic characteristics that would make it a 1-way valve. That such a simple structure can meet the demands upon it is due to the sophisticated reflex system that controls it.(18) An element of this system that can be traced in part to the work of W.B.Cannon but more particularly to the admirable experiments of R.W. Dougherty is the subject of this chapter.

Cannon's experiments

In 1903, Cannon,(19) working with cats, found that when 180-220 cc of a bland solution of potato starch and bismuth subnitrate was introduced into the stomach, it would reflux from stomach to esophagus, be milked back into the stomach by a peristaltic wave and reflux again, repeatedly in a regular back and forth pattern.

If the starch-bismuth preparation was then brought to the normal pH for carnivores (.5%), one final reflux occurred and the back and forth motion ceased. Five years later, he confirmed these fluoroscopic observations on the unanesthetized animal by pressure measurements in the esophagus and gastric fundus of the anesthetized cat.

Section of the splanchnic nerves did not affect the reflex closure. Section of the vagus nerves five and seven days before the experiment and pithing the animal to the brachial region practically eliminated the reflex, but after removing the acid solution from the stomach and immediately reinjecting it, the pressure required to force the sphincter from below increased from 19 cm of water on the first try to 53 cm of water on the forth repetition. As he had eliminated every extrinsic pathway that could be involved in reflex closure, Cannon had to conclude that he was dealing with a locally acting "myogenic" reflex.(20)

Sphincter control in ruminents

After a lapse of two generations, R.W. Dougherty(21),(22),(23),(24),(25) rediscovered the reflex closure of the cardia and, in a beautiful series of experiments, added greatly to our understanding of the mechanism. Dougherty, a veterinary physiologist, did the bulk of his work with ruminants, particularly sheep, because of a peculiarity of this class of animal.

A ruminant can derive nourishment from low quality roughages because of a huge forestomach, the rumen, which serves as a fermentation tank in which bacterial decomposition of forage takes place. The nutrients thus released and

modified, and the bacteria themselves, are then further digested and absorbed in a fashion comparable to the process in man.

Enormous quantities of gas evolve in this bacterial digestion. Hungate et al. calculated that 1.2 liters of gas formed per minute in the rumen and reticulum of a 1000 pound bovine animal. If this gas cannot be eliminated properly, largely by eructation, the animal dies very quickly when the distended rumen forces the diaphragm cephalad and embarrasses respiration. Consequently, the ability to eructate gas, which in man would be but a minor annoyance, is a fatal malfunction in the ruminant. This inhibition of gas release from the reticulo-rumen is known as "bloat." According to some estimates, it is responsible for an annual economic loss of $100,000,000 to cattle raisers.

Dougherty, Habel and Bond, working with decerebrate sheep, found that eructation was inhibited if the area around the cardia were covered with water, ingesta, foam or mineral oil. Eructation was also completely inhibited with the animal on its back even if the intrarumenal pressure were raised to as much as 120 mm Hg by injection of carbon dioxide gas into a rumen fistula.

Although the cardiac sphincter could resist this high pressure, nevertheless, if the rumen were emptied of fluids so that it contained only gas, the animal eructated equally well in the supine position as it did in the prone. One hundred cc of water in the rumen of a small sheep would completely inhibit eructation in dorsal recumbency. When the water was removed and 100 ml of 1% butyn sulphate solution substituted, gas and fluid were eructated when the rumen was insufflated.

In later experiments employing cine-radiography, it was shown that the reticulum contracted in such a way as to empty itself of ingesta. In these remarkable films,(26) a fold can be seen rising out of the floor of the cephalad portion of the reticulum. The fold then becomes more prominent and moves caudally pushing ingesta in front of it away from the mouth of the esophagus and acting as a small dam to keep it away.

In this way, the reticulum empties itself of gas by erecting a barrier over which gas can flow out the esophageal orifice while the fluid content of the rumen is held clear of the esophageal mouth thus preventing contact of ingesta from the region so that reflex inhibition of eructation will not occur. The speed and forcefulness of this activity leave no doubt that it is purposeful and that the purpose is to clear the way for eructation.

Neurologic control of the sphincter

The reflex inhibition of eructation established by Dougherty and his associates was also abolished if a restricted area about the cardia was covered briefly with a 1% solution of butyn sulphate. By perfusion with methylene blue, Hill(27) found

nerve endings in the superficial layers of the epithelium of the reticulo-rumen. Their distribution was to essentially the same areas that, when anesthetized, abolished the inhibitory reflex. This work consolidated the physiologic studies by demonstrating the anatomic structures that could be inferred from them.

Stevens and Seller(28) showed that eructation could be inhibited by procaine hydrochloride block of the dorsal vagal trunk on the one hand and promoted by stimulation of the same nerve trunk.(29)

The conclusion of the Dougherty group, therefore, is that there are receptors in the stomach epithelium immediately next to the mouth of the esophagus that, when stimulated by the presence of ingesta, reflexly inhibit opening of the lower esophageal sphincter. The experimental results also would show that the reflex is mediated through the vagus and, presumably, higher centers. This conclusion corrects the earlier view of Cannon who believed he was dealing with a local "myogenic" reflex.

One can scarcely review the findings of Cannon and Dougherty et al. without being impressed that here is one ingredient for a solution to the classic problem of control of the lower esophageal sphincter in man. If the same reflex can be shown to exist in man, we will have resolved the problem posed by Schatzki's trapeze artiste.

Human applications

Several objections that must be overcome before we can confidently apply these results to man:

1.) Dougherty was dealing with the highly specialized digestive tract of the ruminant. The multi compartment stomach, the largely striated muscle of the esophagus, the fact that there are not separate longitudinal and circular muscle layers (there are two oblique layers instead) all suggest caution.

2.) Although Cannon's experimental animal, the cat, is one from which experimental results can usually be extrapolated to man, the reflex - at least as he interpreted it - seems inapplicable on several counts:

a.) Rhythmic back and forth motion of fluid between the stomach and esophagus is not normally observed in man.

b.) An unphysiologic amount of fluid was given to the intubated animals in the fluoroscopic studies (180-200 cc of fluid is the entire normal daily intake of a large cat.)

c.) In interpreting his experiments, Cannon seems to have neglected the possibility that there might be a reflex that opened the sphincter or decreased sphincter tone.

d.) If the last possibility is admitted, we have to deal with the fact that, in denervating the sphincter, he was sectioning the pathways for both types of reflex and thus simply measuring the baseline tone of the denervated sphincter.

The response to these difficulties is to turn our attention directly to the question of whether an inhibitory reflex, especially of the type showed by Dougherty, exists in man. The reflex can be studied in routine radiologic examinations of the upper GI tract and the highly sophisticated equipment now in common use compensates for the fact that we are not as a rule able to perform surgical experiments. Several lines of evidence converge to show that an inhibitory reflex is also present in humans

The de Carvalho test

When water is administered to a patient in the right posterior oblique (supine) position, in some cases, cardioesophageal reflux will occur. Crummy(30) found this to be true in 10.3% of 650 consecutive cases. Linsman(31) encountered an incidence of 40.5% of 1000 cases. Although there seems to be good correlation between a positive dC test and a history of pyrosis, the question "What does the dC test?" is still an open one.

Yet Cannon's experiments provide an answer. When he administered a large quantity of starch-bismuth solution to cats and observed rhythmic reflux, he was basically performing a de Carvalho test. That is, the stomach was flooded with water and any acid present was washed away or diluted to the point where it could no longer activate the receptor. Released from reflex inhibition, reflux of the solution into the esophagus occurred, distention of the esophagus reflexly caused peristalsis, and so on. The more prolonged the experiment, the less frequent were the episodes of reflux suggesting that accumulating acid /pepsin reactivated the reflex.

If the CD reflex is present in man, then the deCarvalho test can be explained in exactly the same way: the barium meal dilutes any acid/pepsin already in the stomach and the water administered in the RPO position washes over the CD receptor flushing it further. In this way, Cannon's experiment is reproduced. Gastric contents reflux and are milked back into the stomach by peristalsis.

The angle at which the patient is positioned is often quite critical corroborating this interpretation. The best way to demonstrate reflux is to have the patient drink rapidly, as he turns from supine toward the RPO position. Reflux starts at a definite point and usually stops when this point has been passed. This position dependence suggests that the stream of water must be accurately directed at the

receptor area to release the sphincter. It also provides a clue to the location of the receptor.

If one compares the parallel studies of Linsman and Crummy, we find that, although both series are qualitatively similar, reflux was four times as frequent in Linsman's series (40.5%) as in Crummy's (10.3%). Linsman had the patient drink 200 cc of water after the routine barium meal. Crummy, on the other hand, used only 15-30 cc. These results are exactly what would be expected if there were an inhibitory reflex: the more water employed, the more it will dilute gastric acid/pepsin and the more it will flush the receptor area. Consequently, the more patients in whom the CD receptor will be turned off allowing reflux.

A Chi square test for significance shows a positive association between reflux symptoms and a positive deCarvalho test ($p < .005$). It shows that, once the inhibitory reflex is turned off, whatever is operating to cause sphincter release(32) will be uninhibited. The significance of the difference ($p < .001$) between the frequency of reflux in the otherwise parallel series of Crummy (10.3%) and Linsman (40.5%) also supports the above interpretation.

In these patients, as with the sheep in Dougherty's experiments, the receptor must be on the posterior wall of the stomach - the area washed by the stream of water entering the stomach in the deCarvalho maneuver.

Magenblase

The formation of the huge gas bubble in the gastric fundus observed in patients with eventration of the left leaf of the diaphragm is understandable if one postulates an inhibitory reflex. The patient cannot eructate the gas because it is above the submerged receptor area near the mouth of the esophagus.

Other evidence of a CD reflex

There are some pyrosis patients in whom the sphincter remains open for periods of 20 seconds or more. If during this time, they are tilted so that the gastric fluid approaches the mouth of the esophagus, the sphincter will close.

Even more striking is the occasional patient with myotonic dystrophy. In this condition, with esophageal involvement, the sphincter is always widely patent. As a result, the patient swallows air to keep gastric contents out of the esophagus. The air column extends from the cervical esophagus to the gastric sinus. Even in such a patient, the sphincter will close, however ineffectually, when gastric contents are maneuvered into the proximity of the mouth of the esophagus.

If acid/pepsin is required to activate the receptor, it would be expected that removal of the acid secreting portion of the stomach would cause a failure of sphincter inhibition and consequently reflux. This complication has been

described by Lataste and Gonthier.(33) Again, if there is a loss of gastric acid production from atrophic gastritis there is increased reflux. In this condition the pH may be > 5 instead of the normal < 3.(34)

Turning to evidence that is perhaps less "scientific" but nevertheless useful as a check on the conclusions, we recall that mothers burping infants invariably place them in exactly the position that places air rather than gastric fluid in contact with a receptor area on the posterior wall of the fundus. To do otherwise would bathe the receptor area with gastric fluid and activate the inhibitory reflex.

Most people I have questioned have noted subjectively that they automatically lean forward when a premonitory sensation of subxiphoid tension signals an impending eructation. This is most obvious when drinking a carbonated beverage while seated in a reclining chair. One is also conscious of this forward inclination of the body when driving a car as the shoulder strap of the seat belt interferes with the motion and brings it to attention.

Physicians and nurses also note that when bed patients vomit or belch they struggle to turn toward the prone position thus freeing the CD receptor from contact with gastric contents and releasing the sphincter-opening mechanism.

The sensation of "gas," paradoxically, is relieved by the ingestion of bicarbonate of soda although the resulting generation of CO_2 when it contacts gastric acid should, one would think, aggravate it by increasing the amount of gas in the stomach. The immediate effect of ingestion of sodium bicarbonate is to produce an average of 4 cc (!) of gas in the first minute after ingestion. The reason is that, although the reaction is instantaneous, the gas produced remains in solution.(35)(36)

Neutralization of acid in contact with the CD receptor, however, can explain the relief as this permits sphincter release permitting eructation.

SUMMARY

The weight of the evidence derived from these diverse phenomena supports the existence in humans of a reflex essentially identical with that reported by Cannon in cats and elucidated in great detail by Dougherty and coworkers in ruminants. This reflex has a gastric receptor, the physiologic sphincter of the lower esophagus as an effector and a vagal reflex arc. There is evidence that the receptor is on the posterior wall of the fundus.

This reflex explains the phenomena encountered in the deCarvalho "water siphonage test" and the ease of eructating gas in contrast to the difficulty of disgorging fluid gastric contents. It accounts for the "pleasant security" of Cannon and the safety of Schatzki's trapeze artiste.

To be sure, this important reflex is not the only one affecting the sphincter. For example, it has been shown(37),(38),(39) that acid stimulation of the esophageal mucosa causes increased basal production of HCl in the stomach. This seemingly paradoxical vicious circle may actually serve the organism by feeding back a signal that will stimulate the CD receptor and thus close the sphincter.

These experiments do not, however, reveal how inhibition is accomplished. We must recall that the sphincter is normally closed. Therefore, one cannot simply assume that there is reflex tightening of the sphincter. Instead one must look for the mechanism that opens the sphincter, because that is what is actually being inhibited. In a subsequent chapter, I will assemble the evidence for this mechanism.

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Gas/bloat and the physiology of belching

"For the clinician, the bête noir of the symptoms referable to the digestive tract is 'gas'."(1) Patients can assure themselves of instant rejection by complaining, "Everything I eat turns to gas!" Unfortunately, "gas" and the equivalent symptom of "bloat" is the most common of all gastrointestinal complaints.(2)

Efforts to correlate the "gas" symptom with objective evidence of gas have been futile. Although physicians have " . . . been well indoctrinated that swallowed air is the cause of alimentary tract gas.",(3) they often see patients with flat bellies and gasless radiographs who paradoxically complain, "I feel as though I were going to explode!" On the other hand, a patient may have visible abdominal distention without complaining of gas. Patients with ascites, pneumoperitoneum or extreme obesity seem oddly immune to the complaint of bloating. On the other hand, patients with mechanical bowel obstruction and a genuine gas problem, complain of cramps, not gas.

This puzzle of "gas without gas" has motivated many studies. Although these have yielded exact data on the production, composition, absorption, elimination, and clinical correlations of intestinal gas, they only deepen the mystery. Lasser and associates,(4) for example, using a sophisticated isotope washout technique, found that 12 patients with this complaint averaged 23 ml less intestinal gas (177 ml) than 10 normal controls (200 ml).

In extreme cases, the patient may habitually perform alternate Mueller and Valsalva maneuvers to fill and empty the esophagus with gas (as in esophageal speech) to provoke eructation of gas from the stomach in the belief this will afford relief. In an occasional patient, this can get completely out of hand. The false eructation may become a social liability - an outrageous, gross habit that the patient is apparently unable or unwilling to control.

Most such patients are eventually referred for radiologic examination where, aside from demonstrating of aerophagia(5) if it is extreme, the results have given no clue to the cause of this bizarre performance.

Various theories are (6)offered. Nevertheless, none carry any conviction or suggest an experimental test that would either confirm or refute them. Roth(7) relates belching to aerophagia, splenic flexure syndrome, "magenblase syndrome" and neurosis, considering it only in the context of the excessive belching of aerophagia. He lists aerophagia as caused by most of the disorders of the upper GI tract. He speculates that the habit forms because the ". . . patient experienced some relief of a distress with the eructation of air . . . and thus deliberately induces belching by aerophagia to secure that relief again."

Bockus(8) mentions belching as a frequent symptom of hiatus hernia, but does not describe the actual act. Roth does note that ". . . slightly elevating the chin

and extending the neck . . . "(9) is a conscious maneuver to provoke belching, but attributes this act to an effort to swallow air to induce belching.

Earlam(10) lists some quantitative information: the gastric gas bubble is less than 50 ml of air 2-3 ml are swallowed with each bolus; 1,000 swallows/day (70/hr while awake, 7/hr while asleep) would result in total ingestion of 2.0 to 3.5 liters of gas per day. He also reports that the average audible belch contains 20-80 ml and requires simultaneous contraction of the abdominal musculature and relaxation of the sphincter.

One need not consult standard references to become acquainted with the mechanics of belching. Self-observation will go a long way. Thus one finds he unconsciously leans forward, head up, neck extended. The larynx ascends as in swallowing and gas is released. The forward inclination of the thorax is particularly noticeable if one is restrained by the shoulder strap of a seat-belt.(11) Unfortunately, this introspection gives no clue to what is going on at the lower end of the esophagus. It would be much better to study the process fluoroscopically.

Inducing a belch at fluoroscopy

Chance fluoroscopic observation of a belch initially aroused my interest in the problem. Thereafter, I was frustrated by the infrequency with which I could study the process further. Chance observations were infrequent and when they did occur, one seldom had the area of interest in the field of view, good wall coating, cine camera on, etc. One cannot just wait for a belch to happen - fluoro time will run out with the examination uncompleted.

Eventually, as the Cannon-Dougherty (CD) reflex mechanism became clear, I realized that, because the CD receptor was on the posterior wall of the stomach, simply rolling the patient over into the prone (RAO) position after the dC maneuver would remove the inhibitory effect that submersion of the receptor for this reflex has on sphincter opening. This had the desired effect, eliciting a belch in 20-30% of patients.

LMC induction of eructation

There appear to be two factors involved in the success of this maneuver, neither of them the amount of air in the stomach:

1. The gas or bloat symptom. Such patients yield the bulk of the positive responses.

2. The amount of water employed. Using 180-250 cc of water and rocking the patient back and forth to slosh it about the fundus seems to increase the yield.

With this technique, there were many opportunities not only to secure sequence spot and cine films but also to ask patients whether the sensation experienced during longitudinal muscle contraction (LMC) was the same as the sensation they were calling "gas."

The first visible warning of an impending belch is a slight conical tenting of the otherwise hemispherical outline of the fundus or the formation of a hooded appearance of the fundic mucosal folds.(12) This may vanish or become more pronounced. If the latter, a hiatus hernia, if present, becomes drawn upward to the full length of the tethering phrenoesophageal ligament. The whorls of mucosal folds in the fundus then straighten and point to the gastroesophageal junction and that junction assumes the shape of a trumpet bell or, more accurately, an alpenhorn. The diaphragm is tented and, as a result, loses its sharp outline because the central ray of the beam passes through the tent rather than grazing the diaphragmatic dome.

This bell shape is the shape of an elastic membrane under traction. The greater the force applied to the center of the membrane, the more acute the apex angle of the cone of tensed membrane. The perihiatal region of the diaphragm itself may also be tented upward. These changes are manifestations of a LM contraction that, in extreme cases, may shorten the esophagus 36% or more.

The sphincter then opens and stomach gas instantly inflates the esophagus. If the voluntarily controlled superior constrictor opens, gas escapes into the pharynx, is eructated and the fundus collapses. If not, an en masse contraction of the circular muscle and/or a peristaltic wave forces gas back into the stomach.

In this way, a striking event in patients who belched under fluoroscopic study was a preparatory strong contraction of the LM (LMC). Questioning the patients as it occurred established that the gas/bloat sensation was simultaneous with the signs of LMC.

This portion of the act of belching, unlike gagging or vomiting, is quite deliberate and usually occupies several seconds. LMC may be sustained for a considerable time if, for any reason, the superior sphincter does not open. As much as 8-10 seconds is not unusual and in an exceptional instance it may be sustained 30 seconds or longer allowing ample time to question the patient.

Sphincter release does not follow every preparatory LMC. The LMC may simply subside or there may be a partial subsidence followed by contraction leading to an almost rhythmic ebb and flow of tension on the GE junction marked by varying shape of the trumpet bell. The greater the force applied to the center of the membrane, the more acute the angle of the apex of the cone formed by the stretched membrane.

On cine films in such cases, one can see the sphincter margins alternately approaching and receding from each other as though prepared for an instant closure whenever some very delicate balance of forces is destabilized. This process is a clear and convincing demonstration that the LM opens the sphincter: the shorter the LM, the higher the GE junction and the more patent the sphincter.

Gas/bloat is a misinterpretation of LMC

Sometimes, the conscious control of belching can be dramatic as in the following case:

11/4/66 H.D. 45151/M-426 Fluoroscopic note: There was considerable aerophagia and a large amount of gas accumulated in the stomach. A "hiatus hernia" was present that was ideally provoked by having the patient belch, a feat at which he was unusually proficient and could perform on command. This produced shortening and elevation of the esophagus, evoked the HH and was associated with a descent of the diaphragm resulting in a violent jerking motion of the stomach back and forth through the hiatus. Cine films confirmed and showed free cardio-esophageal reflux.

The tension noted during LMC is perceived at the conscious level as gas although the patient's description of the sensation may be quite variable. Most had trouble defining the sensation, but such expressions as "It feels full.", "Kind of pulling.", "Like a pressure.", or "Bloated." were used. When the cardia tents, if a leading question, "Does it feel as though you were going to belch?" is asked, the response is usually an unequivocal "Yes." From there it is a small step to the conclusion that the sensation preceding a belch is what patients mean by "gas" and similar vague fornulations.

Patients often expressed surprise that I knew they were about to belch or that I could see a belch although they had eructated silently and politely! The uniformly positive identification of LMC with the gas sensation left no doubt about the cause of this mysterious symptom. The identification was convincing after relatively few cases because the patient could identify the symptom with the event as it happened. That is, he did not have the sensation throughout the examination, but at the exact time I observed traction on the gastric fundus.

The LM tension has many interpretations other than gas and bloat as in the following case.

LM051846: This 50 year old male truck driver complained almost constantly of the sensation of a mass "about the size of a large potato" beneath the right diaphragm. Occasional heartburn. Ultrasonography revealed a normal gallbladder and minimal evidence of fatty infiltration of liver. Upper GI demonstrated grade ii reflux (asymptomatic), grade ii duodenitis and antral

gastritis. When the fundus tented after the dC maneuver he was surprised I was able to tell exactly when he was having the mass sensation.

He was given a 10 mgm capsule of Nifedipine with instructions to chew and swallow it the next time the symptom was severe. He reported back within the hour, "I didn't take it on the drive home because I wasn't sure what it would do to me, but I had complete relief of the symptom within minutes of swallowing the pill. I feel completely relaxed."

Gas, or better, LM tension is a symptom because there is a condition in which LM tension is hyper - "longitonia" if you will. The LM contracts, increasing its tension on the diaphragm, even when there is no physiologic need for it to do so. It does not require the stimulus of gas distention of the stomach to exert traction on the PEL. A degree of traction great enough to force the sphincter by vector resolution will also tension the diaphragm. For this reason, patients with reflux usually also complain of gas/bloat.

The reason patients misinterpret LM tension on the diaphragm is that LM contraction is an event that frequently precedes eructation of gas. It is independent of whether or not there is actually gas in the stomach. This is why the gas sensation does not correlate with how much gas in the stomach. It is purely a function of LM tension.

As was noted earlier, extension of the neck is a belch-facilitating maneuver. This is true because extending the neck applies tension to the esophagus and may also produce a stretch reflex.

From the point of view of patient care, I believe the identification of the "gas" symptom with LM tension is of value. At a minimum, efforts to decrease gas formation or to adsorb gas on activated charcoal (It has been done!) are futile. Pharmacologic efforts to reduce LM tone would be more rational.

It seems paradoxical that ingestion of bicarbonate of soda - a substance that will generate gas when it contacts gastric HCl - often relieves the sensation of gas. It must do so by lessening the degree of gastric irritation, suggesting that the latter, whether it be by excessive acid, carminative, toxic substance, etc., may be a stimulus to LMC.

There seems at least a lay consensus that certain foods produce gas. Seed catalogs advertize burpless varieties of cucumbers. Is it possible that such foods contain an active ingredient that heightens LM tone?

Belching and mass contraction of the longitudinal muscle

Like the LMC of nausea, pyrosis and vomiting, there is no peristalsis associated with the LMC of belching except for a post-belch cleanup wave. The force of

LMC can be gauged by the size of the conical tent; the higher and thinner the tent, the greater the traction that is being applied to it.

It is noteworthy that, although in peristalsis LM and CM contraction are precisely integrated, in en masse contraction they are can be independent. In pathologic circumstances - particularly diffuse esophageal spasm - simultaneous LM and CM en masse contractions also can occur. In all of these cases, it is striking how the esophagus, to play its many roles, coordinates its several functional elements in sharply different ways.

Because it enables us to correlate pressure relationships in the esophagus with the events just described, it is useful to review the phenomena McNally, Kelly and Ingelfinger(13) recorded from the 2-dimensional viewpoint of the manometer. They found that insufflation of the stomach with air via catheter raised the intragastric pressure to 5-7 mm Hg. Within the 200-1600 cc range, intragastric pressure was independent of how much air was introduced, suggesting that distention rather than pressure is a stimulus to belching.

Manometrically, the escape of air from the stomach into the esophagus was signaled by a sudden equalization of gastric and esophageal pressures [i.e., sphincter release]. In cases where it was possible to record the intrasphincteric pressure, it was found that it could be maintained at values equal to the intragastric pressure for periods of 14-110 seconds before the escape of gastric gas into the esophagus. In two cases ". . . reflux occurred 88 and 96 seconds respectively after intragastric pressures had exceeded those recorded in the sphincter zone." [Emphasis added, i.e., pressure gradients were not sufficient to open sphincter.] The authors were puzzled that ". . . . no. . . . correlation between increased gastroesophageal pressure gradients and the appearance of simple reflux was noted."

In a later study, Sigmund and McNally(14) found a gradual reduction in sphincter pressure preceding a belch induced by essence of peppermint, a carminativet. Perceptively, they drew a significant conclusion: "The gradual decrease in intrasphincteric pressure suggests an active relaxation of the sphincter rather than a passive one, secondary to the increased gastric pressure forcefully distending the sphincter." [Emphasis added.] That active agent, however, is the LM not intragastric pressure.

Both radiologic and manometric techniques demonstrate that release of gas via the superior constrictor may be delayed for many seconds or inhibited entirely. Kahrilas et al.(15) using manometric techniques found that the upper esophageal sphincter (UES) released gas 1-10 seconds after the common cavity effect [gastric pressure = esophageal pressure] that denotes release of the LES. The reflex controlling its relaxation could distinguish between refluxing fluid and gas, probably by sensing the spatial and temporal characteristics of release of gas

into the body of the esophagus. As it was unimpaired by mucosal anesthesia these authors were able to exclude a mucosal pH receptor.

Because the sphincter is closed in these preliminary stages of belching, the state of the circular muscle of the body of the esophagus in not seen directly. However, the instant the sphincter opens, the body of the esophagus balloons to its full diameter. From this, we can be certain that the circular muscle behind the sphincter is relaxed at the time of sphincter release. This establishes that LM contraction is not accompanied by CM contraction in belching.

The esophageal lumen collapses as the gas leaves via the superior constrictor. Fluoroscopically, I find it impossible to decide whether this collapse is due to an en mass contraction of the circular muscle or to a momentary Valsalva maneuver that occurs simultaneously with the belch. The latter could force gas from the esophagus even without circular muscle contraction. There is manometric evidence that, like the LM, the circular muscle is not restricted to one mode of contraction. McNally et al. interpreted a simultaneous spike in the tracing from a catheter placed in the colon as an indication that a brief Valsalva maneuver was occurring. This, however, is subject to another interpretation as will be seen in the next chapter.

Belching and the CD receptor

We have seen that a sustained forceful LM contraction inetiates sphincter opening and allows the escape of gas from the stomach. What terminates the egress of gastric contents? Several lines of evidence indicate that then acid/pepsin contacts a fundic recptor LM contraction is inhibited and the sphincter, released from its dilating force, closes.

Often, a belch is not a single event. It may be one of a sequences of eructations of which the first is usually the largest (noisiest) followed by cleanup secondary or tertiary burps. This is particularly the case in infants. It seems that if they are going to spit up feeding, it occurs on the secondary burp.

Rarely, I have had an opportunity to view a belch in the upright position rather than in the RAO (supine) position in which belching is usually induced. This provided an excellent opportunity to see the exquisite timing of these events. As the fluid level in the fundus of the stomach rose toward the sphincter with escape of the gas, the sphincter snapped shut just in time to prevent the escape of gastric contents. Viewing this left little room for doubt that the sphincter knew the fluid level was coming at it. When a large amount of gas escapes, gastric contents may splash the periesophageal area and end the eructation prematurely. A correcting burp soon follows.

These observations only confirm what everyone knows from personal observation - that normally, even when the stomach is full after a meal, gas can

be released from the stomach very forcefully without regurgitating fluids. Parents burping infants discover this fact several times a day. Mechanistically, the problem is to learn how the sphincter (or better, the LM that is holding it open) knows that in a few milliseconds the esophagus is going to be doused with gastric contents and finds out in time to do something about it.

The experiments of Dougherty et al.(16) demonstrated the elaborate arrangements ruminants have evolved to prevent reflux of gastric fluids while allowing free escape of gas. The reticuloruminal fold acts as a dam to prevent fluid from reaching the esophageal orifice. Gas, of course, can travel over the dam to exit the stomach. Less elaborate, but nonetheless effective measures perform the same function in man.

By the constraints placed upon it, the receptor for this reflex closure should be a chemoreceptor. Yet, if one postulates such a sensor, the rapidity of its transmission to the motor arm of the reflex is difficult to square with the time it takes for a chemical to diffuse to the depth of the sensor and through any mucus coating it. On the other hand, gastric irritation from certain foods may cause increased mucus production that, by coating the receptor, puts the watchdog to sleep with resulting reflux and heartburn. Such a mechanism would account for the latent period between a dietary indiscretion and the onset of heartburn.

The alternative to a chemoreceptor is some system of sensors that continually monitor the shape of the stomach - a possibility, perhaps, as the shape of a stomach partially filled with air would, for the same volume of contents, be different from one filled entirely with fluid.

There are, however, objections to the latter hypothesis. For one thing, the analogy to the CD receptor is too strong. It will be recalled that the nerve network demonstrated by Dougherty and his co-workers is near the esophageal orifice. This is more in keeping with a chemoreceptor activated when contacted by gastric contents than it is with shape sensing. The latter would require a diffusely distributed net of sensors not localized to this specific region at all.

Surgical causes of the gas/bloat symptom

The gas/bloat symptom is inseparable from the act of belching because the LM tension that produces sphincter release of necessity also exerts traction upon the diaphragm. The symptom is produced whether or not sphincter release is achieved. We have seen that the association of the sensation of diaphragmatic tension with belching causes misinterpretation of this tension with its normal result - an eructation of gas.

Without really intending to do so, surgeons will often perform experiments for us on a scale so vast they have great statistical significance. One way of surgically producing tension on the diaphragm is to pull the GE junction down below the

diaphragm and suture the stomach around it so it will not retract to its normal position. By thus "taking a tuck" in the esophagus, a more or less continuous pull on the diaphragm is created as the hypertonic LM tries to pull the plicated fundus through the hiatus. Thus, the gas bloat syndrome which may persist for many years after a Nissen fundoplication. An Angelchik prosthesis often produces the same effect(17) for the same reason.

Infantile colic

Before leaving the subject, there is one aspect of belching that probably deserves discussion - infantile colic. The colicky baby screams, apparently for no reason, as the parent walks the floor wondering what to do to relieve the child's evident misery. The screams seem senseless and different in kind from those of a hungry baby. The infant may be soothed by again feeding it but soon it is having another attack of colic.

Careful mothers learn never to put the baby back in its crib before it has been burped, but this is not always a solution. It is a trial to the parent because the burp may not come until it is nearly time for the next feeding.

Perhaps because of its association with difficulty in burping, the implicit assumption is that colic is due to the excessive air that, failing a satisfactory burp, must be passed through the GI tract. However, radiologists, who see many infant chest and abdomen radiographs, know that non-colicy infants normally have quantities of gas in the small and large bowel. If gas leaves the stomach, there is nothing to stop it being eliminated as flatus. As adults, we are aware that it is no problem to pass any required amount of gas and that it is not painful to do so. Why should it be any different for infants?

When we think of a bowel cramp, we think of a sustained contraction of the circular muscle, because all of the bowel cramps with which we are familiar are circular muscle cramps. Circular muscle spasm can be seen with the fluoroscope, e.g., the cramping pain often associated with barium enemas in a patient with a spastic sigmoid, the pylorospasm noted with delayed gastric emptying - or felt by the examiner as with the broom handle descending colon palpated in patients with spastic colons. We are just not accustomed to seeing LM spasm in the bowel, much less to palpating it.

This produces a typical clinical impasse: a set of symptoms leads the physician to expect a corresponding set of objective findings. The expected findings do not appear. Conclusion: "No disease; over-anxious mother."

Yet colic is too common, too real and too disruptive to the lives of young parents to be dismissed in this fashion. The same parents may alternate children with and without colic. To me it seems far more probable, because of its association with burping problems, that infantile colic may be the simply the pediatric

equivalent of bloat. Unfortunately, infants cannot describe their symptoms, but it would add weight to this supposition if the symptom responded to medication as did my patient's "gas" symptoms.

SUMMARY

The mechanism at the lower esophagus must be able to permit eructation of air while denying egress to gastric fluid contents. Two elements are involved:

1.) The CD reflex, which inhibits LMC when activated, must be turned off.

2.) The sphincter must be opened by vector resolution of the force of LMC. These two mechanisms account for the perfection of control of this physiologic function.

"Gas," a symptom that is the bane of the clinician, is due to LMC. LMC applies tension to the diaphragm via the PEL. Because the resulting sensation normally precedes a belch, it is identified at the conscious level as a sensation of gas, bloating or epigastric fullness. It can be seen radiologically as a tenting of the cardia, especially if a modification of the usual deCarvalho test technique is used to induce belching by turning off the receptor for the CD reflex.

Postoperative gas/bloat is due to surgically tensioning the esophagus by fundoplication or insertion of a prosthesis. The problem of the colicky baby is discussed in the light of the relation of LMC to belching. It seems that "baby bloat" would explain the phenomena better than current rationales.

As would be anticipated, patients who have a slack, elongated esophagus as in achalasia are unable to belch(18) Vagal cooling abolishes transient sphincter relaxation in dogs by abolishing LMC. The deliberate eructations I have described in this chapter, although most easily studied fluoroscopically are not the most common. There is a second method of sphincter release, alone or in cooperation with LMC. This will be discussed in more detail in the following chapter.

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The function of hiccups

It is generally believed that these abrupt diaphragmatic contractions do not serve any useful purpose(1) or have only a nuisance value. After reviewing 192 references, Launois et al.(2) recently concluded "The purpose of hiccup is unknown."- an extraordinary deficiency when one considers that hiccups have engaged the attention of medical practitioners at least since the time of Hippocrates.

Most of our exact knowledge of hiccups comes from Davis(3) who studied the neurophysiology of hiccup in three patients in great detail, measuring the frequency and amplitude of hiccups in relation to phase of respiration, PCO2, integrated electromyogram, etc. He found that a hiccup is essentially an abrupt Mueller maneuver. The glottis closes to prevent inspiration 35 milliseconds after electrical activity rises above the baseline in the diaphragm and external intercostal muscles .

Because of the glottic closure, hiccups had little effect on respiratory exchange (although they did produce hyperventilation in a patient with a tracheostomy). Davis concluded they were not governed by the same centers that controlled inspiration and expiration. This and provocation by gastric distention caused him to conclude that hiccup was ". . . gastrointestinal rather than respiratory in nature." and ". . . more analogous to the vomiting reflex, for example, than to a respiratory reflex such as coughing." Davis also believed hiccups had no useful function in man and the literature echos this belief.

Yet it is hard to believe that a complex, exquisitely coordinated function of the diaphragm, intercostal muscles, glottis, brain stem and somatic and visceral nervous system does not in some way serve the organism. Overeating and ingestion of carbonated beverages are well known causes of hiccups. Parents of small babies are familiar with the hiccups that frequently follow a feeding (and are cured by feeding more!). An association of hiccup and GE reflux is well documented in the literature. One wonders, therefore, if hiccups are an attempt to open the sphincter.

In its effect on the PEL, and thus the sphincter, a sharp downward motion of the diaphragm is the precise mechanical equivalent of a sharp upward contraction of the esophagus. It will tension the PEL and so have the same sphincter-dilating effect. It may even activate an esophageal stretch reflex producing an amplified effect. Perfused catheter studies(4) have shown absence of a detectable LES during attacks. This would indicate reduced hiatal squeeze and as a consequence, hiatal widening.

Although hiccups are always spoken of in the pleural, I first conjectured they might open the sphincter when a solitary hiccup happened as a patient rolled from the supine to the prone position. It provoked gross GER. Attempts to

produce reflux again with the usual maneuvers were unsuccessful. Subsequently I noted than many, perhaps most, belches are initiated by a single hiccup - not the repeated (up to 28,000 times a day(5)) rhythmic ones we usually think of in that regard - but by an isolated event preceding and inseparable from the belch it elicits. One alerted to this association will note a sudden tightness of the belt or out-thrust of the abdomen just before such a belch. A belch initiated by LMC would have a more subtle but reverse effect on the abdomen. A hiccup induced belch is often a cooperative effort with LMC: first the gas sensation of LMC, then the hiccup, then the eructation of gas. Or a LMC type belch may shortly be followed by one or more of the hiccup variety. Elaborate strain gages and strip chart recorders are not required to establish this hitherto unknown phenomenon. The reader will be able to observe it in him/her self. There is just one problem. Glancing downward to observe the abdomen will cause an automatic flexion of the neck. This may inhibit the LMC portion of the process and abort the eructation.

During a hiccup, the glottis either does not close completely or during its delayed closure emits an inspiratory croak as the abdomen expands with a downstroke of the diaphragm. Launois et al.(6) collected the words for hiccup in 23 languages. Many, but not all of them, are onomatopoetic. In English at least, the sound of a hiccup and the burp it produces are considered embarrassing but there is no help for it.(7)

A belch preceded by a premonitory "gas" sensation and gradual LMC can be suppressed.(8) It is due to LMC as described in the previous chapter. A burp initiated by a hiccup, however, may come without warning and be too abrupt and unexpected to be suppressed voluntarily. Recently I witnessed a dozen such affecting a noted economist being interviewed on C-Span. Given the capability, he could have been expected to suppress them on such a public occasion.

Such an isolated hiccups explain the episodes of "inappropriate"(9) transient complete loss of LES pressure(10) that result in reflux both in normal subjects and in esophagitis patients.(11) In another study by the Milwaukee group,(12) 27 % of transient increases in intraabdominal pressure (such as would be caused by a hiccup) were associated with reflux. The glottic closure in singultus is purposeful, therefore - it prevents aspiration on sudden sphincter release.

The concurrent onset and causal relationship of singultus and acid reflux in a patient with protracted and recurrent hiccups have been minutely documented symptomatically and by pH monitoring by Shay, Myers and Johnson.(13),(14) They reasoned that the downward excursion of the diaphragm in hiccup caused reflux by creating a negative intraesophageal pressure. It is not clear, however, how negative pressure per se could open the sphincter - it should merely collapse the lumen as is the case if one tries to suck water through a flaccid straw. It seems more probable that, just as LM tension causes reflux by upward

traction on the PEL, a hiccup causes downward traction of the PEL with the same sphincter-opening effect.

Commenting on this case, Graham(15) alludes to his experience with manometry of hiccups.(16) He found hiccups caused ". . . . A great reduction (or absence) of the lower esophageal sphincter pressure. . . ." and also cessation of peristalsis. He believed these effects were as important as negative intraesophageal pressure in causing reflux.

There is an impression in the literature that complications associated with reflux stimulate vagal afferent nerves and cause singultus. Shay et al. make a good case that it is the other way around - singultus causes the complications. Their patient had no symptoms of reflux until after the onset of hiccups, symptoms were confined to the times the hiccups recurred, and pH monitoring documented that ". . . . acid reflux increased during hiccup episodes and returned to a normal level with their cessation." Gluck & Pope, nevertheless, could provoke hiccups at will in their patient with the Bernstein test. Both points of view may be correct, giving rise to a vicious circle and prolonged bouts of hiccup.

Ataractic drugs(17) such as haloperidol and chlorpromazine(18) as well as atropine(19) also have therapeutic value in otherwise intractable hiccups. Friedgood and Ripstein report an 82% permanent cure rate with 50 mg of chlorpromazine given IV. In one case the hiccups had been present 9 months. Launois, et. al.(20) name baclofen as the drug of choice for chronic hiccup.

We have seen that nausea and vomiting (as well as hyper salivation(21)) are caused by severe degrees of traction on the PEL by LMC. Ataractic drugs must ablate this traction to achieve their effect. Such LM relaxation, if it accounts for the therapeutic effect of these drugs on hiccups would suggest that there is feedback between the esophagus and diaphragmatic control centers or, more likely, that a LMC backs up the diaphragm to effect vector resolution on the sphincter. If the LM were flaccid when the diaphragm contracted, the PEL would be too slack to resolve the force generated.

This in turn suggests that clonic LMC may also be a feature of hiccups. Clonic LM contractions synchronized with hiccup would explain why the latter have persisted even after bilateral phrenic interruption.(22),(23) With the LM jerking on the PEL from above and the diaphragm from below, the sphincter-opening force would be augmented as the pull of one is opposed by that of the other. However, in a single case of hiccups in which I was able to obtain 10/sec 105 mm frames of the cardia, there was no evidence of such. Unfortunately, this patient had a ruptured PEL.

Stimulation of vagal afferents by a sudden influx of air has also been shown to cause a reflex loss of LES pressure, probably via the same mechanism.(24) This reflex is abolished by bilateral cervical vagotomy. The existence of such a reflex

also suggests that LMC is an element of hiccup. Vagal cooling or vagotomy is said to abolish the belch reflex.

Although unstated, it seems implicit in Davis' results that there are not only somatic neuron discharges to the diaphragm and intercostals but visceral discharges to the glottis via the 10th cranial nerve and vagus that control it.(25) As the latter also supplies the esophagus, specifically the LM, it is tempting to postulate that this end organ too is neurologically activated in a hiccup.

A common denominator exists among the various maneuvers used to break up the hiccup cycle: most affect the esophagus. Many involve performing a Valsalva maneuver that, as we have seen, can cause a forceful, sustained LMC. The celebrated Hippocratic(26)

.Kellogg, Edward L. and Meyer, William, Hiccough. Medical Record 142:441-4, 1935.(27) maneuver is said to cause gagging (a single forceful LMC) as well as sneezing. The same may be said of depressing the tongue or pulling out the tongue or inducing vomiting.

Startling the hiccup sufferer, commonly with a loud and sudden sound, is a favorite and effective home remedy for hiccups. Such sounds, if in the 70-125 dBA sound level, uniformly produced tertiary contractions(28) in subjects exposed to 1000 Hz acoustic stimuli. TCs, as has been pointed out,(29) are markers for simultaneous CM and LM contraction. The production of LMC is the common thread. Perhaps inducing a different mode of LMC inhibits a mode of LMC associated with hiccup.

No one seems to have a good idea why hiccups are so often a cyclical phenomenon. Davis concludes, ". . . . there is some feature of the hiccup, itself which predisposes toward a further hiccup and thus perpetuates the bout." This could be the sudden impulse it transmits to the esophagus.

SUMMARY

Although hiccups have engaged the attention of philosophers at least since the time of Plato, there was no reason to suspect their physiologic function until the function of the LM was known. The solution to one mystery was the key to another. The abrupt diaphragmatic downstroke of a hiccup generates the same sphincter-opening vector forces as does a contraction of the LM. A hiccup, therefore, rather than being a useless biological quirk at best and a nuisance at worst, is actually a useful physiologic mechanism. It performs the identical sphincter-opening function of LMC in eructation of gas. In addition, the associated glottic closure prevents aspiration should liquid as well as gas escape the stomach.

Hiccups are also useful in another sense - for the purposes of this monograph. Unless the reader has access to a fluoroscope and a ready supply of subjects, it is difficult for him/her to be totally convinced that it is vector resolution of the upward force of LMC that opens the sphincter. With hiccups, however, the reader can be self-convinced if a few days - a week at most - that a mechanically equivalent down stroke of the diaphragm will do the same thing.

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26. 4 The scholarly will doubtless be interested in this bit of research and its quotation from Plato (a fifth century B.C. younger contemporary of Hippocrates) attributed to a Dr. Gibson by Kellogg and Meyer. "When Pausainis came to pause . . . Aristodemus said that the turn of Aristophanes was next, but either he had eaten too much or from some other cause he had the hiccough, and was obliged to change with Eryximachus, the physician, who was reclining on the couch below him. 'Eryximachus,' he said, 'you ought either to stop my hiccough or to speak in my turn until I am better.' 'I will do both,' said Eryximachus, 'I will speak in your place and do you speak in mine; and while I am speaking, let me recommend that you hold your breath, and if this fails, gargle with a little water; and if the hiccough still continues, tickle your nose with something and sneeze,

and if you sneeze once or twice, even the most violent hiccough is sure to go. In the meantime, I will take your turn and you shall take mine.'"

27.

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Longitudinal muscle contraction

It is important to recognize that the longitudinal muscle (LM) is completely invisible to intraluminal manometers, transducers and balloons. Unlike contraction of the circular muscle or sphincter, LM contraction does not affect intraluminal pressure. For that reason, the LM is the "lost muscle" of the esophagus.

This circumstance explains the paucity of information about the function of the LM in the vast literature of the esophagus. It is not even in the index of Bockus. Castell(1) does not mention it after page 11.Standard reference works, textbooks and even monographs(2),(3),(4),(5) restrict their discussion of the LM to a description of its anatomy. Otherwise minutely detailed physiologic and radiologic descriptions of the process of deglutition,(6) ignore the LM. A monograph on disorders of esophageal motility(7) does not mention the longitudinal muscle in that connection. In his review of the recent literature, Diamant found no studies attempting to distinguish between the physiological characteristics of longitudinal and circular muscle fibers. An extended computer search of the medical literature for the years 1966-94 retrieved no references to the longitudinal muscle.

This creates unique problems and opportunities for the investigator. There is little infrastructure on which to build and the reports that do shed light on the action of the LM must often be reinterpreted.

LM anatomy

A surprising variation in LM anatomy exists among mammalian species. The dog is considered a poor model because its muscle, instead of being arranged in a separate inner circular layer and outer longitudinal layer, consists of two helical sets of fibers. A similar arrangement is found in the cow, sheep, camel and other species. Like humans, the cat and opossum have inner circular and outer longitudinal layers and are so considered more suitable species for physiological research. In teleosts (fish with boney skeletons) it is surprising to learn that the order is reversed with the LM inside the circular.

The relative proportion of the total muscle mass in each layer is also variable. In the cat, the LM is well developed. In the rat it is tenuous. (8)In humans, more than 50% of the muscle mass of the esophagus is LM - the reverse of the situation elsewhere in the gut.(9),(10)

Type(11)	% CM	% LM
All striated	4.2 1.5	5.7 .33
Mixed	34.5 2.4	41.1 3.3
All smooth	62.2 2.9	54.1 3.4

The preceding table, after Meyer & Castell,(12) tabulates the percent of the total esophageal length the indicated muscle type was present. Average length of 11 autopsy specimens of the esophagus was 22 2.3 cm. The table cannot be taken literally. I have reviewed sections taken from the upper, middle and lower esophagus in 12 autopsy specimens. On any given section, cells are cut transversely, longitudinally and obliquely. It would take an elaborate statistical analysis to approximate the number of fibers in each direction. The table ignores the fact that many fibers are cut obliquely and are neither longitudinal nor circular. Such fibers must be spiral.

Since the work of Kaufmann et al.,(13) the conventional view that the human esophagus is composed of an inner circular and an outer longitudinal layer of muscle may require revision. These authors describe spiral fibers running in all directions: clockwise up, clockwise down, counterclockwise up and down. They appear to start from the adventitia and end at the submucosa.

Diamant(14) was unable to find confirming studies but these may yet be forthcoming. The spiral configuration seen in "corkscrew esophagus" and the "curling" phenomonon would seem to agree with their description. Contraction of spiral fibers should both shorten and constrict the organ. As will be seen, esophageal shortening can occur without either peristalsis or en masse contraction of the CM.

In dogs and most rodents, the esophagus contains chiefly or only striated muscle. In cats, striated muscle makes up all but the distal 1/4th.(15) Pelot(16) as well as Netter and Mitchell(17) state that the LM is thicker than the circular muscle in man, a relation which is unique to the esophagus and the reverse of the rest of the gut where the circular muscle predominates. The proportion of LM to CM varies not only longitudinally but axially. Near its origin from the cricoid cartilage the LM is mainly massed in two thick bundles posterolaterally. The average thickness of the esophageal wall as measured by ultrasound is 2.6 mm.(18)

Earlam(19) states that smooth muscle cells form a syncytium. They transmit electrical signals via a low resistance connection called a nexus so small it can only be seen with the electron microscope. There is no difference between circular and long smooth muscle in this respect.

The median thickness of the mucosa is .2mm(20) The organ functions exclusively as a conduit. Unlike the rest of the gut, no digestive enzymes are secreted here. Its only glands are the lubricating, mucus-producing glands. The stratified squamous epithelium best resists erosion.

The phrenoesophageal ligament (PEL), which attaches the esophagus to the diaphragm is extraordinarily elastic and seems to have been designed to buffer the force of LMC. Hayward(21) noted that when the PEL was cut, its proximal

portion retracted into the adventitia. Groszek and Matysiak(22) state that it does not have the histologic structure of a true ligament (thick collagenic fibers) but is composed of collagenic fibers with a rich admixture of elastic fibers which begin to appear antenatally and show increasing prominence into young adult life. They diminish in number with advancing age. A maximal sliding HH is 8 cm on films. Allowing for 30% enlargement by the divergent beam, this is an esophageal shortening of 5.6 cm or 25% of its length without rupture of the PEL. This remarkable elasticity serves an essential function in deglutition.

The vagus trunk in the thorax is mainly sensory (afferent fibers) and not motor. It is estimated that there are only 3000 efferent fibers in the vagus (for 5,000,000 ganglion cells in the gut.)

It is important to remember that the esophagus is merely a specialized segment of gut. The most significant difference between the esophagus and the rest of the gut is its relatively fixed length. Firmly secured to the cricoid cartilage above and, via the phrenoesophageal ligament (PEL), to a less mobile portion of the diaphragm below, its ability to shorten is limited to the elasticity of those attachments and the position of the diaphragm. As respiration is inhibited during deglutition, the diaphragm is often stationary when the LM is active.

LM physiology

Animal studies, chiefly on the opossum, have yielded findings that are of clinical interest as they often support what one can establish from fluoroscopic observation of patients. Both the longitudinal and circular muscle are supplied with cholinergic neurons. The m. mucosae is similarly supplied. As would be expected, swallowing begins with myelohyoid activity (an index of the upward migration of the larynx that initiates swallowing). LM contraction precedes circular muscle contraction (CMC) and is of longer duration - 5.5 and 6.3 sec in the opossum.(23) Latency is shorter for the LM. Unlike the circular muscle, the LM does not hyperpolarize. Vagal stimulation causes LM and CM responses that are qualitatively similar to those elicited by swallowing.(24)

In lower animals at least, peristalsis can occur even though the organ is denervated. This suggests myenteric or myogenic transmission and control of peristalsis. Yet the experiments of Janssens(25) show that a central program controls peristalsis. A peristaltic wave crossed the transected esophagus whether or not it was re-anastomosed. Balloon distention of the distal segment after transection would incite peristalsis in the proximal segment.

Yet without vagal input the intramural plexus can produce peristalsis independently - an apparent example of distributed processing. In this the esophagus is similar to the jejunum, a free graft of which to replace a resected cervical esophagus continued to show independent migrating muscular contractions.(26)

Stimulating the cut end of the vagus will reduce lower esophageal sphincter pressure (LESP) but this effect may not be primarily on the sphincter as such stimulation also causes LM contraction. Stimulation of the dorsal motor nucleus of the vagus or nucleus ambiguus will also lower sphincter pressure if the vagus is intact.(27) Oxygen dependence of the lower esophageal sphincter (LES) muscle of the opossum has suggested a clue to reflux in anemias.

In the opossum,(28) the LM contracts throughout the duration of electrical stimulation. The CM, on the other hand, begins contracting after the stimulus - either electrical or stretching - is turned off.

Normal physiologic pressure measurements

Level	Pressure
Pharynx	+ 0 mm Hg
UE sphincter	+100 mm Hg
Body	- 5 mm Hg
LE sphincter	+ 20 mm Hg
Stomach	+ 5 mm Hg

LM power

The LM has a contractile power so great it can tear loose from its moorings at the hypopharynx and at the diaphragm.(29) LMC is the only conceivable explanation for tearing the stitches out of a fundoplication. It may even pull the plicated stomach through itself and/or through the hiatus in failed Nissen fundoplications.(30) Either by attrition or brute force, it can rupture the phrenoesophageal ligament. Its power is such that it can pull the stomach through a constricting Angelchik prosthesis.(31),(32),(33)

Although the force of LM contraction (LMC) has not been reported in man, an estimate can be derived from measurements of intraesophageal pressure. Presumably, a LM fiber can exert at least the contractile tension of a fiber of circular muscle. The LM mass, however, is greater. Given that the CM can produce manometric pressures of 200 mm Hg on a catheter of, say, 4 mm radius we can apply Laplace's law(34) to arrive at the wall tension.

Laplace's law states that p = t/r, where t = wall tension, p = intraluminal pressure and r = radius. Inserting these values in the formula gives:

200 mm Hg = t/ 4 mm, or t = 800 mm2Hg and dividing by the CM thickness of 1 mm gives 800 mm Hg. But 1 mm Hg = .01934 lb/inch2. Substituing in the above formula yields the considerable tension of 15.7 lb per square inch for the CM. Maximum LM tension should be at least this great - enough to create havoc at its attachments. In abnormal cases ("nutcracker esophagus") 350 mm Hg has been recorded - equivalent to 27.6 lb/in2.

In studies of cats, Torrance(35) found that ". . . . maximal stimulus of the vagus resulted in [esophageal] shortening to at least 50 percent of the original length, the cut end retracting under the aortic arch."(36) To produce this shortening the LM tension had to be great enough to stretch the PEL.

Torrance could produce vomiting by stimulating the central end of the transacted vagus. Apomorphine potentiated this stimulus. A strong LMC was the penultimate event of vomiting. He also found that vagal stimulation produced gastroesophageal reflux from a water distended stomach. Reflux was accompanied by minimal increase in intragastric pressure.

Torrance concluded reflux was ". . . . due to a mechanical effect on the oesophago-gastric junction and not merely due to inhibition of the circular smooth muscle fibers of the distal esophagus." He based this conclusion on the following:

Curare(37) completely eliminated reflux after vagal stimulation,

Mechanical traction on the esophagus produced immediate regurgitation comparable to LMC contraction.

Sympathectomy 6 weeks prior to vagal stimulation had no effect.

Unfortunately, these excellent studies were interpreted in terms of the then prevailing theories according to which the angle of His and its mucosal analog, the valve of Guberoff, were the defense against reflux. With the demise of these mechanisms, they are now seldom cited.

The effect of vagal stimulation on LMC has been studied quantitatively in dogs by Edwards(38) who measured an elevation of the GE junction of 2.2 cm [~ 25% shortening] on stimulation of one vagus nerve and 2.4 cm on stimulation of both. The contraction was described as "violent" with a nearly vertical kymograph tracing. Displacement was so abrupt that simultaneous measurement of LES pressure was impossible. This velocity of contraction is seen in human subjects during vomiting and must be unique for an organ that, unlike the dog esophagus, is largely smooth muscle.

LMC was abolished by d-tubocararine but not by l-hyoscine and, in agreement with Torrance and Johnson, Edwards believed the striated muscle was responsible for the shortening. In the dog esophagus, which is entirely striated muscle, abolition of contraction by curare is to be expected. The result should not be extrapolated to man.

Edwards could abolish most LM activity by stripping one or both vagi from the esophageal musculature. Tacitly recognizing that LMC could cause HH, he suggested that selective vagotomy might be employed as an adjunct to anatomical HH repairs to prevent recurrence.

Matthews, Thorpe and Little(39) stimulated a single vagus at thoracotomy in several groups of patients. The control group, that had no mediastinal disease, all showed a "brisk" response - defined as 1.5 cm or more of shortening in 1 to 2 seconds - to stimulation of either nerve. On the average, patients with HH showed a diminished response to vagal stimulation [perhaps because the esophagus was already shortened]. Those with achalasia, as might be expected, showed no response to vagal stimulation. The less profound contractions seen may well be attributable to the fact that these patients were under general anesthesia.

Pharmacology

Neurohumoral agents affecting LESP

(After Ouyang-Cohen)

Increases LESP	Decreases LESP
Acetylcholine	Beta-adrenergic agonist
Alpha-adrenergic agonist	Dopamine
Histamine (H1-receptor)	Histamine (H2-receptor)
Serotonin (neutral receptor)	Serotonin (neutral and muscle receptor)
Gastrin	Vasoactive intestinal peptide (VIP)
Substance P	Progesterone
Methionine enkephalin	Secretin
Bombesin	CCK-OP
Pancreatic polypeptide	Glucagon
Motilin	Gastric inhibitory polypeptide (GIP)
	Bradykinin
	Cyclic AMP

Of course, these effects are simply gleaned from the literature. In most cases it is not known whether they affect the sphincter directly or by way of LMC or both.

LES pressure is lowered by: 1.) Fat ingestion. 2.) The secretin "family" (VIP, gastric inhibitory peptide, glucagon) - especially VIP (vasoactive intestinal peptide) 3.) Progesterone - the probable cause of heartburn of pregnancy.

Adrenalin (epinephrine) has two types of activity - alpha and beta. The main beta activity of epinephrine is vasoconstriction. Propranolol blocks the beta activity of adrenalin. Phentolamine is the alpha blocker.

Carminatives apparently lower the LES pressure. This class of products includes peppermint, spearmint, onion, garlic, anise, caraway, cinnamon, cloves, dill, fenner, rosemary and turpentine. Are all products of steam distillation. They are said to promote easy belching.

Fluoroscopic recognition of LMC

Despite its lack of recognition, LM activity is apparent to the questing eye and can, on occasion, be dramatic. Because good landmarks define the lower end of the esophagus, fluoroscopic observation can establish that the LM contracts in at least four distinct ways. Moreover, all of these modes are purposeful, reproducible and fully integrated with the other muscular elements of the organ.

There are many signs of LM contraction (LMC).

1.) Orad displacement of lower esophageal landmarks including

The lower esophageal ring.

The notches of McLean

The sphincter itself.

The mucosal transition

2.) Tenting of the phreno-esophageal ligament, fundus or diaphragm which causes a loss of sharpness of the latter.

3.) Retraction of the fundus through the diaphragm (HH).

4.) A trumpet shaped flaring of the mouth of the esophagus.

5.) "Tertiary contractions"

6.) On occasion, the esophagus, presumably because of left atrial enlargement, can occupy two stable positions. In one of these the LM is relaxed and in the other contracted. This so-called "wandering esophagus" provides an excellent, although rarely encountered, opportunity to study the effects of LMC.

7.) With familiarity, one becomes constantly conscious of the state of the LM merely by its gestalt - whether it meanders loosely among its neighboring organs or is taut.

A cine camera is an invaluable aid in studying LM contraction because exposures can be made at a low frame rate (typically 7.5/sec) and subsequently viewed at a rapid rate (24 frames/sec). When LM contraction is accelerated, it becomes much more obvious, just as time lapse photography reveals the motion of an opening flower. Analysis of slow motion studies is less useful except for analysis of anatomical details. Exposure technique for spot films should be chosen to yield the broadest possible latitude as the usual "black and white" GI films will not show the PEL or the outer wall of the esophagus.

Although useful for some mucosal details and for demonstrating the LM response to gas distention, I have avoided double contrast studies as unphysiologic.

A conceptual model of the esophagus is a helpful guide to analysis of all components of esophageal muscular activity because it focuses attention where the action can be expected. Most esophageal functions are reproducible, so that the boolean model gives a lead to what to be looking for next: from one vertex of the cube there are only three things that can happen - one of the three components must either contract if it is relaxed or relax if it is contracted.

The following descriptions were compiled from such directed observations. In over 600 cases, cine strips of the phenomena under study were made for confirmation or analysis.

Peristaltic LM contraction

Peristaltic LM contraction occurs when swallowing against resistance and as a terminal cleanup during the ingestion of liquids. It is integral with the peristaltic wave of the circular muscle. The LM is tensioned initially by the voluntary act of swallowing. If one observes a marker - and a small Zenker's diverticulum is occasionally a convenient natural landmark - he will see that the initial act of swallowing is an upward excursion of the larynx and the mouth of the esophagus to which it is attached. The excursion amounts to about the height of one vertebral body and one interspace.

This motion is transmitted to the diaphragm as an upward impulse of the PEL. When a normal patient swallows liquids in the upright position, a slight upward impulse with each swallow. may be the only evidence of LM activity.

More vigorous peristaltic LM contraction occurs during swallowing against resistance as with ingestion of particulate food or during a Valsalva maneuver. As the peristaltic wave moves aborally, the LM shortens in proportion to the distance the cone of circular muscle contraction has moved toward the sphincter. Unlike the circular muscle, that relaxes behind the peristaltic wave, the LM remains contracted until the peristaltic wave is completed - as though circular muscle activated LM in its path and "latched" it en passant.(40)(41)

From the characteristic manometer tracing, it is natural to think of a narrow ring of CM contraction advancing toward the sphincter. Serial radiography, however, shows that there are at least 5-7 cm of CM contracting at a time. Peristalsis is not a migrating ring, but an advancing cone of CM contraction. By the same token, a corresponding length of LM is excited. As the mouth of the cone leads, wall contact is achieved only at the lagging apex of the cone. When the leading edge of the cone of contraction approaches the sphincter region, the cone becomes progressively shorter as its apex approaches its base and finally vanishes leaving, at most, a nipple behind. Propulsion of the bolus, therefore, is mediated

by three things: 1.) Aboral migration of the cone of CM contraction, 2.) A decrease in the length of the cone and 3.) Shortening of the organ by LMC.

As the peristaltic contraction cone forces a barium bolus ahead of it, the simultaneous contraction of the LM, by shortening the esophagus, pulls the bolus into the cone and opens and effaces the sphincter. The sphincter effacement and orad excursion were also noted by Dodds, et al.(42) in cats but the sphincter opening was attributed to passive stretching by the bolus. Sphincter effacement is maximal when LM contraction produces maximal shortening of the esophagus.

Once the sphincter opens, the esophagus and stomach form a common cavity thus allowing the bolus to enter the stomach. Two events then happen so nearly simultaneously that I have been unable to determine precedence with certainty:

1.) The LM relaxes.

2.) The peristaltic wave flows into the sphincter area and closes it.

Perhaps because of the structural differences of the sphincter region, the p-wave effectively becomes the sphincter when its closed end reaches the sphincter area. Thus, peristalsis stops at the sphincter and not at the end of the esophagus, i.e., the mucosal junction. This is easily observed and will account for some otherwise inexplicable findings in achalasia and its look-alikes. \

The aperistaltic segment

The peristaltic wave does not continue into or through the stomach. The stomach has an independent pacemaker in the distal corpus discharging 3-4 times/minute.(43) These discharges are not synchronized with deglutition, nor are they under voluntary control as is deglutition. With the aid of the lower esophageal ring, or even more frequently the notches of McLean, one can verify that the peristaltic wave does not progress into or through a short esophageal zone .5 to 2.5 cm in length extending from the lower margin of the sphincter above to the ring or notches marking the mucosal transition below. This zone, which might be called the aperistaltic segment, because it has neither sphincter tone nor peristaltic ability, acts as an opening wedge for eructation of gas or gastric contents when elevated by LM contraction.

The synchronized activities of LM, CM and sphincter during swallowing against resistance are remarkably efficient in performing indispensable functions in the simplest possible way. As the peristaltic wave advances, LM contraction progresses concurrently with opening of the sphincter. Neither gas nor fluid can reflux through the open sphincter because the advancing p-wave - in effect a moving sphincter - prevents it. Reflux can not occur because the peristaltic wave persists until it becomes the sphincter.

Without this mechanism, it would be impossible to drink liquids in the upside-down position, to swallow air or to swallow against resistance. Because the LM comes into play gradually, the sphincter opens only when the peristaltic wave is tight enough to prevent reflux. At the same time, the LMC pulls the wall of the esophagus over the bolus as one would pull a stocking on a foot.

Conceptually, one can think of the sphincter of having two components of tone: a.) its baseline tone that even chemical denervation will not abolish and b.) a supplementary component that it receives when the p-wave "merges" with it. This would explain the sphincter closure even in esophagitis patients who have lost their p-wave.

Christensen and Lund(44) found that circular muscle stimulation (by distention) caused a much wider but undefined LM contraction. This is consistent with the appearance of "latching" of the LM by the same neuronal discharge that activates the CM. Apparently, when the LM is in peristaltic mode, neural connections activate the LM adjacent to the advancing cone of CM contraction. The results (also in the opossum) of Sugarbaker, et al. suggest that LM contracts first and stays contracted longer.

One can also infer an inhibitory reflex originating in the sphincter area that, when activated by the arrival of the peristaltic wave, causes "unlatching" of the LM. Here, then, is a significant difference - one that would not have been expected a priori - between peristalsis in the two muscle layers:

In the CM, contraction is segmental with immediate relaxation both in front of it and behind it.

In the LM, peristalsis causes incremental contraction involving the entire length of the esophagus. Relaxation occurs only after LM contraction reaches its maximum and peristaltic CM contraction reaches the sphincter.

The effect of LMC on the lower esophageal sphincter

The close temporal relation of sphincter closure and LM relaxation and vice versa argues for a neurologic mechanism: arrival of the peristaltic wave at the sphincter must signal a reflex relaxation of the LM. The relation of sphincter opening and closing to LM contraction and relaxation is of the utmost importance because this relationship explains many of the ills that afflict the esophagus. The rule is:

LM contraction opens the sphincter.

All evidence points to sphincter release as the raison d'etre of the LM. Just as closing the sphincter appears to reflexly deactivate the LM, LM contraction reflexly deactivates the sphincter and also mechanically effaces it. Timing of

sphincter release high on the up-slope of the curve of LM contraction is further evidence for LM release of the sphincter.

Elsewhere, I will show that whenever the sphincter is open in belching, reflux, vomiting, etc., the LM is contracted. The contrapositive is also true: all conditions that tension the esophagus - LM contraction, TEF repairs, cervical hyperextension, myotonia, scleroderma, etc. - are associated with reflux.

Vector resolution of LMC force dilates the sphincter.

Turning off the sphincter reflexly does not open it. Baseline tone must be overpowered before the sphincter will dilate. Mechanically, a sphincter is incapable of opening itself. A distending force is required. Although an advancing bolus may force the sphincter, in general, LM contraction contributes the distending force, especially in the absence of peristalsis.

The mystery of sphincter release has been that, anatomically, there are no radial muscle fibers to be found that, like the ciliary body, would dilate the lower esophageal sphincter. The muscles of Juvara and Rouget to which Jutras(45) attributes this function, have never convinced their critics. They are only mentioned in this connection to be disparaged. They may have been some artifact of the dissection process. In any event we can, should they exist, rule them out of this role because of the wide variability in the relationship of the sphincter to the hiatus.

Yet, we know from mechanics that a force need not act only in line with the contracting muscle. Forces can undergo vector resolution into separate components. In the case of the LM, one component of the contractile force is resolved in such a way as to open the sphincter. In this resolution, the PEL plays the essential role.

PEL anatomy

The PEL inserts on the distal esophagus at the sphincter. According to Zaino et al.(46) some fibers insert above and some below the sphincter. This confirms the descriptions by surgeons who have been interested in investigating the hiatal area.(47),(48) Bombeck et al.(49) examined a large number of autopsy specimens using a method designed to demonstrate the details of PEL insertion on the esophagus as they were interested in evaluating the hypothesis that contraction of the diaphragm opened the sphincter.

These authors found that the PEL originates from the inferior margins of the esophageal hiatus as a continuation of the endoabdominal fascia. It then divides into 2 layers. In most individuals, the lower division is little more than a layer of loose areolar tissue easily fractured with a finger. It may be absent entirely. It inserts at or below the level of the ora serrata - 1.4 cm below it on the average.

The other, more substantial layer, is invariably present, often receiving an additional contribution of fibers from the endothoracic fascia. It inserts, on the average, 3.35 cm. above the ora serrata. The insertion is not linear, but occupies an appreciable longitudinal extent - up to 1 cm, although generally less than that. In addition, ". . . . a diffuse fibroelastic network of fibers passes from the main, membranous body of the ligament to the sphincter area of the esophagus in all cases." The point of insertion was taken as the distance from the ora serrata to the fibers obviously taking the load when the PEL was stretched. These details are important because they show that the PEL is exactly designed for its sphincter opening function.

Contraction of the muscle fibers of the sphincter only closes it more tightly. If it is to open at all, a bolus must force it or an external influence must pull it apart. This force must be applied radially and equally in all directions or the result would only be a lateral displacement of the esophagus.

Vector resolution and thePEL

In the case of the LES, this is accomplished with great finesse by employing a longitudinal force, that of LM contraction. It is the vector resolution of that force in all directions that opens the sphincter. The other vector component merely stretches the PEL.(50)

The PEL is essential to this vector resolution of force. If it did not exist, the force would simply pull the stomach through the hiatus. Because of the this restraint, however, the force of LM contraction is resolved into radial vectors that open the sphincter.

The normal mechanism of sphincter opening, therefore, has three elements: 1.) The circular muscle of the sphincter is "turned off" reflexly and 2.) The sphincter is mechanically dilated by vector resolution in the horizontal plane of the force of LM contraction. 3.) An advancing bolus has a wedge-like opening effect.

The second of these elements is most difficult to come to grips with analytically because the geometry changes as the LM contracts and the PEL stretches with an unknown elasticity. Several mathemeticians I consulted found the problem intractable. Attempts to model the process by an aeronautical engineer using an advanced finite element computer package(51) were unsuccessful.

This precise integration of the functions of the longitudinal, circular and sphincter muscles of the organ is necessary to carry out the esophageal function in all positions and under a great variety of circumstances, some of which make unusual demands.(52)

Non-peristaltic LMC

The LMC just discussed is but one mode of recognizable LM activity. The others are en masse contractions rather than the progressive, relatively slowly migrating contraction associated with peristalsis. They include:

Swallowing in the absence of resistance.

LM contraction during tertiary contractions.

Belching.

Gagging.

Vomiting.

In all these activities, the esophagus shortens and tents the PEL and even the perihiatal region of the diaphragm itself. When the PEL is elongated to any extent, it will assume the typical trumpet-bell shape of a membrane under central tension. The retracted gastric fundus also becomes conical as it assumes the same shape as the PEL tent. If the sphincter opens, it does so at the point of maximal LM contraction.

The force of LM contraction can be gauged by the size of the conical tent; the higher and thinner the tent, the greater the traction being applied. There seems to be no basis for forming an opinion whether all of the muscle cells are partly contracted or some of the cells are completely contracted in this shortening. The speed of contraction seems to be greater the more forceful the contraction.

Swallowing liquids

During normal deglutition of barium in the upright position, LM contraction is best seen if the patient is swallowing rapidly. As each bolus leaves the esophagus, it does do on a slight up-stroke of the diaphragm. This upstroke is easily mistaken for respiratory motion. However, it is detectable because 1.) Respiration is suspended during deglutition and 2.) The rate of diaphragmatic upstrokes is faster than the respiratory rate.

The upstroke is synchronized precisely with the spurt of barium from the esophagus into the stomach. Thus, even with the assistance of gravity and with liquids of low viscosity, LMC does come into play.

LMC and the manometric "plateau wave"

The upstroke of the diaphragm is caused either by an en masse contraction of the LM, by the initial upward motion of the larynx to which it is attached or, more

likely, by both. Just as a sharp tap on its tendon may provoke a reflex contraction of a skeletal muscle, the upward tug on the esophagus that initiates swallowing may provoke a stretch reflex stimulating LMC.

Phase 1 of the manometer tracing - a negative pressure wave - is believed to be due to stretching of the esophagus by the upward hyoid/laryngeal impulse.(53) Phase 2 - a positive, plateau type wave - may well be a stretch reflex of the LM.

As respiration is suspended during swallowing, this upward motion of the diaphragm about the hiatus translates into increased intrathoracic pressure. This raises the internal pressure of the esophagus throughout the organ so that manometers at multiple levels record a simultaneous pressure increase. The simultaneous pressure increase at all levels, its positive sign and its relation to the peak caused by the peristaltic wave passing the same level all identify this pressure increase as the "plateau" or "phase 2" portion of the deglutition wave.

In this indirect fashion the LM is able to affect a manometer. It may be the single exception to my earlier statement that the LM is invisible to the manometer.

LMC in belching

The LM contraction that precedes belching is not an all or none evnt. The PEL tent may be observed rising and falling for several seconds as escape of gas from the fundus approaches. The incipient belch may be suppressed entirely, in which case the tent is lowered and vanishes. If air does erupt, however, it does so at maximal elevation of the tent, i.e., when LM contraction is sufficient to open the sphincter. This is at once the most clear-cut and easily reproducible way of demonstrating both the existence of a sphincter and that LM contraction opens it. These phenomona are discussed in more detail in the chapter on gas/bloat.

Examination of the cine footage made available to me by Dr. William Dougherty showed that there is also LMC preparatory to eructation of gas by sheep. As the rumen is inflated with gas via a rumen fistula, repeated, forceful LM contractions occur as the animal attempts to belch. This was most striking in the cervical esophagus - a phenomenon Dougherty referred to as "fluttering." The activity slows or stops after a belch and resumes on reinflation of the rumen.

LMC in gagging

A type of contraction perceived by patients - and examiners for that matter - as a gag also has esophageal manifestations. We are familiar with spasm of the pharynx when the gag reflex is elicited, but this is only the oral aspect. Abrupt LM contraction is the esophageal component of a gag. Although I subsequently verified this effect repeatedly by having an assistant induce a gag with a tongue-blade while I watched the cardia, I first became aware of this while examining the following patient.

J.O. 6541 age 43. This man had a long history of "indigestion," "acid stomach," nocturnal acid regurgitation, pyrosis. "Food gets caught in that tube." and he can't swallow. He regurgitates unchanged food.

Fluoroscopic note: The patient swallowed barium with great difficulty due to a hypersensitive gag reflex. He was, nevertheless, very cooperative and swallowed despite the difficulties, thus providing an extraordinary opportunity to view the act of gagging. This proved to be an instantaneous longitudinal contraction of the esophagus, jerking all the landmarks cephalad. The amplitude of the motion was considerable because a rupture of the phrenoesophageal attachments allowed gross hiatal herniation of the untethered fundus when the esophagus contracted.

LMC in nausea

Whereas tonic LMC during belching has an appearance of delicate control - power applied with finesse - the LM contraction seen in nausea is a much more powerful application of force to the lower esophageal attachments. In a fraction of a second, an abrupt jerk elevates the PEL tent. It may not release the sphincter even though the elevation is obviously more powerful than the force necessary to release a belch. The contraction may partially subside before again increasing.

Because it is precisely at this preliminary stage of nausea and vomiting that hypersalivation occurs, it suggests that stretching the PEL is the stimulus to the hypersalivation that precedes vomiting. This has been observed in studies of farm animals. Reid and Phillipson(54) showed that distention of the rumen provoked increased salivary secretion. Clark and Weiss(55) reported reflex salivation in sheep and goats when an area about the cardia was stimulated mechanically as did Comline and Kay.(56)

The LMC component of nausea, therefore, provides an explanation of the familiar hypersalivation that precedes vomiting. A powerful LMC applies traction to the PEL, stretching it beyond the limits normal for belching. This reflexly stimulates salivation by traction on the cardia. This stretching is perceived as nausea. Because of the alkaline pH of saliva, the hypersalivation has the effect of immediately neutralizing the acid pH of the esophagus after emesis.

The patient studied by Shay et al.(57) tends to confirm the mechanism. He had copious salivation caused by singultus. As is noted elsewhere, hiccups have the same mechanical effect on the cardia as LMC.

Vomiting

Finally, the most severe degree of LM is perceived as the pain and gagging sensations of vomiting. Because the sphincter must be open before the stomach can be evacuated, emesis calls forth a powerful LMC that rides roughshod over sphincter resistance. There is nothing subtle or tentative about this form of LM

contraction. The stomach is yanked into the hiatus to the full extent of the PEL's ability to stretch by a powerful, almost instantaneous LM contraction as the stomach contents are discharged.

To observe the process fluoroscopically the radiologist's instinct for keeping barium out of his shoes must be overcome. Generally one steps back and shouts for the emesis basin at the first sign of gagging and spasmodic abdominal muscle contraction. Perhaps this is the reason standard references fail to mention the LM in connection with vomiting.

This powerful contraction is the reason Daintree Johnson(58) was able to produce HHs in dogs by inducing vomiting with apomorphine.(59) Forceful LM contraction is also the reason patients note subxiphoid soreness for some time after emesis. It is a painful contraction, because it over-stretches the PEL. The discomfort is a part of the reason we struggle to avoid vomiting. Infants trying to burp often scream from the pain of LM contraction stretching the esophageal attachments to the diaphragm in an attempt to open the sphincter.(60)

From time to time reports of retrograde prolapse of the gastric mucosa or gastroesophageal intussusception are encountered.(61) These can generally be shown to be examples of retching or variants of the captive bolus phenomenon. The LMC draws the stomach through the hiatus as wire is drawn through a die. Through an endoscope the gastric mucosa may be seen reaching 8-10 cm above the gastroesophageal junction.(62)

Again, the salient feature of contraction of the LM is that in every instance in which it is seen - belching, nausea, vomiting, cardio-esophageal reflux - the sphincter is effaced.

LMC causes both Mallory-Weiss and Boerhaave syndromes

Retching, an act indistinguishable from gagging, is another manifestation of LMC. The LMC may not succeed in emptying the stomach on its first attempt or the stomach may be empty. The contractions are the same and are accompanied by nausea and hypersalivation. Retching is essentially an aborted emesis.

A cause of about 10% of upper gastrointestinal bleeding, the Mallory-Weiss syndrome, starts with retching or non-bloody vomiting followed by hematemesis. This pattern has always suggested that the initial retching itself caused the bleeding. About 10% of the cases are due to retching during endoscopy (63),(64) providing ample opportunity to confirm the etiology as the endoscopist observes intact mucosa on inserting the instrument, then retching, and subsequently sees the linear tear(s) as he withdraws it.

Knauer(65) observed 58 cases noting that 72% had HHs. There was a striking radial asymmetry in the location of the tears with 52% occurring on the right vs.

only 7% anteriorly. In other series,(66) the incidence of HH has been as high as 100%. The friability of the gastric mucosa within the "hernia" is cited as a factor in the ease of mucosal rupture.

Barring Boerhaave's initial case in which the esophagus was completely avulsed from the stomach, the only thing that distinguishes Boerhaave's syndrome, from Mallory-Weiss is the depth of the laceration. In both the tears are parallel to the long axis of the esophagus.

Like effacement of the sphincter, these syndromes present the paradox of a longitudinal force producing, not the expected transverse tear, but a longitudinal one. Although they are attributed to overdistention of the esophagus or herniated cardia by sudden ejection of gastric contents, this can scarcely be the case as they are seen after retching without emesis and after endoscopy that, of course, is performed on an empty stomach. The wedge shape of the tears(67) observed after endoscopy-induced retching is a clue that the force is applied at the PEL. If overdistention caused them, they would tend to be elliptical.

It is, perhaps, puzzling that most of the tears (78%) are in the stomach just below the mucosal junction. Two circumstances may account for this.

1.) 72% to 100% [Knauer] of the patients have hiatus hernias. The increased friability of the mucosa in the herniated portion of the stomach may account for this localization.

2.) LMC, when resolved by the PEL, causes a trumpet-like flaring of the GE junction. The further down the trumpet, the more the mucosa is stretched. This accounts for the endoscopic observations that wide end of the wedge-shaped tear is aboral and that virtually all of the tears are below the ora serrata.

LMC in myotonia dystrophica and scleroderma

The esophagus of myotonic dystrophy provides an elegant confirmation of the proposition that the LM opens the sphincter. In this disease, characterized by a deficiency in the ability of muscle to relax, if the LM is affected, it may be constantly contracted. For that reason, the p-wave cannot latch the sphincter. Constant LMC keeps the sphincter constantly open. This results in an appearance that, like scleroderma, can be mistaken for achalasia because of the striking air esophagram it produces.

This identifies another important sphincter function: it keeps gas out of the esophagus as well as releasing it from the stomach. If the stomach and esophagus are in constant communication, the circular muscle cannot collapse the lumen. No matter how often a peristaltic wave milks gas into the stomach, if the sphincter does not latch, air rushes back to again distend the body of the esophagus. But distention is the stimulus to peristalsis ("The esophagus abhors

distention." [Dodds]), so the process repeats to the exhaustion of the circular muscle.

When these patients are upright, a continuous air column extends from the superior constrictor of the esophagus through a widely patent sphincter nearly to the gastric antrum. The only way the patient can prevent reflux in the standing position is to swallow so much air that the fluid level never reaches the esophagus. As in achalasia, the circular muscle can never rest and the esophagus ends as a dilated, aperistaltic tube.

The situation in scleroderma is similar. The LM is constantly short - nearly all such patients have tubular HHs, many of which are unrecognized in the published cases. The mechanism is not clear, but these patients end up with atrophy of the CM while the LM is preserved.

Longitonia

Belching, retching, gagging and vomiting, are isolated events that occur to everyone. We also have to deal with a pathologic state of the LM in which its tone or irritability are increased. In its purest form, LMC causes a symptom complex for which esophageal longitonia might be an approprate nane. The salient symptom of this abnormality, reflux, requires a separate chapter.

Clonic LM contraction and pseudo-palpitations

There may also be a clonic type of LM contraction that passes for "cardiac palpitation." From personal experience over many years, I can describe the sensation produced as a jerking, thumping, palpitating sensation in the mid substernal region. These palpitations are so irregular that they remind one of the erratic thrashings of a recently caught fish flopping about on the bank of the stream. So alarming were these "palpitations" that, as a pre-medical student I once called the Student Health Service for assistance.

Although I had never been able to palpate a pulse irregularity, I was satisfied with a medical opinion I was having premature ventricular contractions until I experienced such an episode while I was in the radiology department of a small hospital. The EKG room was next door and the same nun was both x-ray and EKG technician, so, to clear up the mystery, I was able to schedule an EKG instantly.

As I watched the tracing emerge from the machine, I was totally unprepared for the normal rhythm it charted. Ruminating on this over the years, by a process of elimination, I formed the suspicion that the sensation could be due to clonic contraction of the LM. If it were not the heart, barring the internal thoracic muscle, the only other muscular organ in the vicinity was the esophagus.

I then recalled that patients with auricular fibrillation and a totally irregular heart beat seldom complained of palpitation. With this in mind, "Have you ever had palpitations or a sensation like a fish flopping around in your chest?" became a part of my routine questionnaire.

Surprisingly, many patients with esophageal disorders did have palpitations. Among them were several physicians. The latter were curious enough, as I had been, to check their pulses for the compensatory pause of this common arrhythmia. Like me, some failed to detect it, but tended to attribute that to the difficulty of reading one's own pulse. Actually, anyone with some medical training who has had PVCs has no difficulty detecting the irregularity in the radial pulse.

Convincing proof of my hypothesis was eventually forthcoming from an unexpected source. A patient was seen for a followup examination a year or so after a "pull-down" operation for hiatus hernia. In this, the original Nissen operation, the lesser curvature of the stomach is sutured to the posterior surface of the left rectus sheath. To my routine question she responded that she had had cardiac palpations before her operation. Did she still have them? "No," she said, (pointing to the left rectus area), "but now I get this tugging sensation in my abdomen."

It is worth noting that each mode of LM contraction is perceived by the patient as a distinctly different sensation. LM contraction is the basis for explaining a number of very common but misunderstood symptoms.

Nausea

A more severe degree of tonic LM contraction, like hypersalivation, is perceived as a part of the nausea syndrome. It would be interest to determine if ataractic drugs act by decreasing LM tone and irritability. The difference between nausea, "gas" and reflux-inducing LM tone is one of degree. The relation between LM contraction and the sphincter will be discussed in detail when gastro-esophageal reflux is described.

Substernal pain and the LM

With substernal pain, the main effort is to distinguish between esophageal pain and cardiac ischemia. Bennett and Atkinson(68) tabulated the details of location, radiation, relation to effort and posture, aggravating and ameliorating influences in 200 consecutive admissions for precordial pain. A variable overlap of every symptom nuance was found with sizable variations in the percentages for ischemic heart disease and esophagitis - enough to suggest that a Bayesian analysis might be a practical (but expensive) means of differentiating the two. One of the more accurate predictors was simply asking the patient whether he though he had heart trouble or indigestion. The patient's diagnosis was correct in 61% of the cases with ischemic heart disease and 85% of the cases with

indigestion - a distinct improvement on the accuracy of the admission diagnosis of 64% overall.

In contrast to the generally clear-cut radiologic and EKG findings in chest pain of cardiac origin, the correlation of symptoms with objective manometric findings in non-cardiac chest pain is very poor. Patients may show no increase in intraesophageal pressure at all during an attack.

Clause et al.(69) carefully selected patients with daily substernal pain unrelated to exertion and without any identifiable cause other than presumed esophageal spasms. They compared the manometric recordings of 9 such patients with control periods in which no pain was present. The tracings were studied blind so that the interpreter was unaware of the presence or absence of pain.

There were no significant differences between recordings during or preceding an episode of pain and those made in pain-free periods. There were no significant differences in baseline esophageal pressure or peak pressures. The amplitude, duration and percent of abnormal peristalsis were well within the limits established in the control periods. In 3 subjects, no waves at all were seen during pain periods. Thus, despite the widespread suspicion that substernal pain is esophageal in origin, none of the patients had a recorded change in usual motility pattern that correlated with the occurrence of the reported pain episodes. They concluded that abnormal contractions are not the direct cause of pain.

Cold food can also be a cause of esophageal pain, but ice cream induced esophageal pain results in aperistalsis(70) - another result inexplicable in terms of abnormal sphincter or peristaltic activity. Clause et al. suggested that perhaps these peristaltic abnormalities may be markers for some unrecognized cause of chest pain because motility aberrations are so common in patients with substernal pain and no apparent cardiac disease.

Clinical manometry may demonstrate intraluminal pressure of 300 mm Hg or more in patients with pain of esophageal origin, but there may be no pain at the time the high pressures are being recorded. Ott et al.(71) reported a high incidence of tertiary activity in their series of 20 cases labeled nutcracker esophagus(72) but stated that the significance of the finding was unknown. The incidence of HH (70%), reflux (15%), diverticula (15%) and TCs (50%) was dismissed as nonspecific incidental findings.(73) In a typical study,(74) 24-hour ambulatory pH/motility monitoring showed that only 21% of chest pain episodes correlated with motility abnormalities.

Because of these discrepancies, it is reasonable to suspect that the spasm causing the pain is not in the CM or sphincter, but in the longitudinal muscle as this will escape detection in the studies listed. There are several clues:

1. LMC applies traction to the diaphragm and so can produce pain.

2. Vomiting, which entails a severe contraction of the LM, is painful.

3. In DES, there is powerful LMC as well as increased peristaltic activity.

4. The pain may disappear after rupture of the phrenoesophageal attachments.

A patient, age 43, gave a history of left subchondral pain diagnosed as nervous stomach while in the army 7 years previously. It was associated with heartburn and "doubled me up with pain" persisting 30-90 minutes before subsiding. Eventually, although the heartburn persisted, the painful attacks ceased as suddenly as they had begun. On examination, he was found to have a 9 cm HH with rupture of the PEL.

A possible explanation is that rupture of the PEL terminates the ability of the LM to apply painful traction to the diaphragm. This would be consistent with the finding of Dalton et al. that the natural history of nutcracker esophagus is one of spontaneous remission.(75) Radiologic followup of a number of such cases should prove interesting. A radiologic search for signs of LM tension during a typical attack should be even more revealing.

There is no doubt that sub-xiphoid (wishbone) pain and/or tenderness is the most common localization of discomfort in patients referred for an examination of the upper GI tract. For many years I have asked each upper GI patient to identify the site of pain by pointing to it. For at least 10 of these years, I was mystified that in most cases I could find no radiological explanation for the symptom. It is not the site one thinks of as either duodenal or gastric reference and may be present in the absence of pyrosis.

Eventually, it became clear that this is but one of the manifestations of LMC. Although frequently associated with pyrosis, it is not due to pyrosis per se, but to the stretching or tensioning of the esophageal attachment to the diaphragm. Less frequent are other references of diaphragmatic pain - subscapular, left arm and to the neck or even the ear. Anterior flexion of the cervical spine, which lessens esophageal tension may provide some relief.

SUMMARY

The longitudinal muscle of the esophagus plays a dominant role in most functions of the organ. Its most important function is opening the LES. In peristalsis it undergoes a latching type of contraction. It also exhibits several nonperistaltic modes of contraction. These are most commonly the en masse contractions that are associated with nausea, vomiting and belching.

Severe tonic contraction is perceived as subxiphoid pain or as nausea. An abrupt en masse LM contraction of marked degree is a gag. The most severe en masse contractions cause nausea and vomiting. The mechanical stimulation of the cardiac region by LMC causes hypersalivation. LM contraction alone without associated diffuse spasm or nutcracker contractions can produce severe pain.

Clonic LM contraction may well be the cause of sensations perceived as cardiac palpitation.

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40. Judging from animal experiments, it may be the other way around. The integration of LM and CM contraction is more complex than suggested by the fluoroscopic appearance. This is really the resultant of LMC and LM relaxation both of which may be taking place at once. Using miniature strain gages to measure LM and CM contraction, Sugarbaker, et al. found that in laboratory preparations both LM and CM contracted sequentially during peristalsis but the propagation was faster in the LM than in the CM and the contraction lasted longer. The duration of contraction was

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50. Although it seems paradoxical that a force applied longitudinally has a transverse effect, there is no question of its effectiveness. The same force resolved in the same way can tear the mucosa (Mallory-Weiss syndrome) or rupture the wall (Boerhaave's syndrome) and in each case, the tears are almost always longitudinal. See appendix B.

51. Pro-Engineer, a product of Parametric Technology Corp.

52. Some years ago, the Milwaukee Journal published a Metro Group photograph of a group of teammates doing handstands over the following caption:

"Lucki Hofmaier was so good at doing handstands and drinking beer that he decided to form a team in Regensburg, West Germany, to encourage competition between other West German cities. Hofmaier can suck a glassful of beer through a straw in about 5 seconds while doing a handstand."

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59. In this connection, Johnson unearthed interesting supportive anatomical evidence: rats, which cannot vomit, have a very thin LM - only a few cells thick. Cats, on the other hand, vomit with great facility and have a thick LM.

60. These two examples of LMC induced pain add weight to the presumption that noncardiac chest pain has a similar cause.

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The lower esophageal ring

Although it was illustrated as a normal landmark by Templeton(1) and first described in detail by Ingelfinger and Kraemer,(2) the lower esophageal ring has become "Schatzki's ring" colloquially because of a series of studies in which Schatzki and collaborators Gary(3)(4) and MacMahon(5) described it in further detail, reported the pathologic appearance at autopsy and correlated the symptoms with the size of the ring.

These papers established that the ring occurs at the junction of stratified squamous esophageal epithelium and gastric mucosa. It is invariably associated with HH -- usually a sliding HH. It becomes symptomatic when the lumen is reduced to about 1.9 cm. It varies in position from 4 to 5.5 cm above the diaphragm. It is 2-4 mm in thickness. The lumen was reduced in one of their cases to as little as 3 mm.

Schatzki and associates also noted that "If the esophagus distal to the narrowing balloons out, as for instance during the Valsalva test, the ring seems to climb away from the diaphragm and, conversely, it seems to migrate toward the diaphragm when the distal esophagus collapses." (6)

This description would seem to show that they once believed that the fundic pouch was "esophagus distal to the narrowing" yet in the same paper, they reported that in 4 cases a change in the character of the mucosal pattern suggested " . . . that the ring was at the herniated esophagogastric junction." Of course, before the pathologic appearance had been described, it would be hard to be certain that the ring was at the mucosal transition.

Brombart(7) soon ventured a respectful correction, pointing out (incorrectly) that the deep inspiration of the Valsalva test accounts for the withdrawing of the ring from the diaphragm and (correctly) that it is not correct to speak of the segment below the ring as "esophagus" as it is the intrathoracic portion of the stomach. Like Brombart and Schatzki, Templeton(8) stated that the ring " . . . is brought out by distending the esophagus." - also focusing on the distention while overlooking the maximal shortening that then occurs at the end of the Valsalva maneuver.

Goldstein(9) has listed the various ideas concerning the origin of the ring culled from his review of the literature as:

1.) Hypertrophied or dislocated constrictor cardia

2.) Overactive inferior esophageal sphincter

3.) Inflammatory stricture

4.) Cardiospasm

5.) Congenital malformation

6.) Trauma.

7.) In his 1970 review, Earlam(10) added mucosal damage from reflux.

Various sphincters have also been proposed to explain the appearance. These include:

8.) The "bracket bundles" of Ziano et al.,(11)

9.) The sphincter of Lerche(12) and

10.) The lower esophageal sphincter.

At present, he most popular rational for LERs is an undefined "inflammation."

It was inevitable that simultaneous manometric and cineradiographic examinations would be performed to relate the ring to the sphincter. Surprisingly, 2 of the 4 placed the sphincter below the LER!(13),(14),(15),(16) Such high-tech efforts have a flavor of overkill as simply looking at the radiographs should convince one that the sphincter is above the ring.

With MacMahon, Schatzki and Gary were the first to report that the mucosal transition occurred on the lower surface of the ring. The histologic findings they described, however, are not those of inflammation or fibrosis, ruling out Nrs. 3 and 7; the ring proved to be 2 layers of esophageal stratified squamous epithelium. Even the submucosa was not a part of the ring. It split into 3 layers without extending to the free edge of the ring. Subsequently, numerous biopsies, at both thoracotomy and endoscopy, have confirmed the initial pathologic description in several significant details. For example, in 14 biopsies, all obtained at thoracotomy, Postlewait and Musser(17) never found any annular muscle in edge of the web, thus disposing of the idea that it was a contractile ring (Nrs. 4, 8, 9 and 10). It is well established that:

The upper surface is squamous epithelium,

The lower surface is gastric epithelium, sometimes with "dribbles" of squamous epithelium running over the edge.(18)

The muscularis mucosae is split into 3 layers,

The core of the web is usually filled with fibrous connective tissue although the amount varies.

There are several additional features of these rings that are significant and must be explained by any proposed theory of their pathogenesis:

Even a marked ring may be invisible at esophagoscopy

On followup, the ring may have diminished in size.

A single ring may break up into multiple rings or vice versa.

The ring may grow from mere notches to a definite ring as the esophagogastric junction distends.

The ring may vanish when the sphincter fails at the end of a P-wave and regurgitation from the HH to the distal esophagus occurs.

They are seldom seen with the huge herniations that occur with rupture of the PEL.

They usually, but not invariably, disappear after a HH repair.(19)

In 1963, impressed by the latter fact, I suggested(20) that the ring was redundant mucosa milked to the lower end of the esophagus by the peristaltic wave. Schatzki did not agree.(21) According to Ott et al.(22) this mechanism(23) is now the main competing theory with inflammation. As was demonstrated in Chapter 2, however, mucosal redundancy is a consequence of LM contraction. The esophagus is only an exception to the general rule because its length is fixed by its upper and lower attachments. If the PEL is stretched - that is, when there is a hiatal transtract - the elasticity of the mucosa and contraction of the m. mucosae cannot take up the slack and a fold must form. The invariable association of gastric transtraction and LERs is because, in order to create enough mucosal redundancy to form a ring, the esophagus must shorten enough to pull the fundus through the hiatus.

Like a mucosal fold, a LER can come and go and accordion-like pleats can form in various configurations. One can sometimes see this happen in rapid sequence or cine frames. This is not the case with congenital malformations or strictures. The redundancy may form multiple fine folds or be gathered up into a large, deep fold. One patient had a single deep ring, yet presented with 3 shallow rings a year later.

Mucosal folds do, however, tend to display a marked constancy. The same physical circumstances that caused a fold to form in one way the first time operate, ceteris paribus, to repeat that configuration the next time. Moreover, living tissues tend to take a set, as though they were graven in stone ultimately, due to the microscopic fine structure that changes but slowly over time.

Thus, fold formation explains not only the histologic appearance but the observations mentioned above. Esophagoscopy, if done under anesthesia, causes a relaxation of the esophagus and the redundancy disappears. Even the friction of the scope is sufficient to elongate the esophagus and remove the redundancy. It would be necessary to demonstrate the HH with a Valsalva maneuver while the patient was swallowing liquid to redemonstrate the ring - obviously impossible with an esophagoscope in place. On the other hand, if anesthesia does not obliterate esophageal motility, distention with gas will cause LM contraction and should enhance a LER.

The growth of the ring with distention of the phrenic ampulla is due, not to distention per se, but to the shortening of the esophagus that occurs concurrently with that distention. If the ring consisted of an annular band of fibrosis, for example, distention would then produce a ring where none existed in the collapsed organ. However, in that case the band would be in the free edge of the ring on histological section. But this is not the case. Although there is fibrotic tissue in the core, the amount varies from slight to striking and is distributed in a wedge, not in a core at the free edge.

It is not intuitively obvious, but consider that a fold has formed that is prominent enough to narrow the lumen to 1 cm. If the esophagus in the region of the fold then dilates another millimeter, it will take 2 mm of slack to keep the ring just as narrow as it was before. Allowing for, say, 15% elasticity the esophagus must shorten more than 2 mm to produce the required mucosal redundancy. This is the reason that as the ring forms, it does not seem to get deeper.

Of course, the "climb away from the diaphragm" during a Valsalva test noted but not explained by Schatzki and Gary is due to LMC with esophageal shortening.

These 2 factors - a relaxed PEL and a shortened esophagus - are the factors that produce HH. The association of rings with HH occurs because they are both caused by the same thing - LMC. After a repair, especially a pulldown procedure, this ability to shorten unduly is overcome and the redundancy cannot form. This results in disappearance of the ring.

In the intervening 32 years, the suggested mechanism has held up well when tested against day to day experience with one still puzzling feature: further experience showed that not all rings were cured by a HH repair. One has to assume that, in such cases at least, the two surfaces of the ring became adherent so that elongating the esophagus will no longer obliterate the fold. In an occasional case,(24) submucosal fibrosis is a striking finding. It would seem that such rings must be of the type that can survive an effective HH repair.

On the other hand, several additional observations have added weight to the postulated mechanism. The fact that LER's are seldom seen in patients with non sliding HH's is a further point in favor of the plication hypothesis. Once the

restoring force of the elastic PEL is lost, the epithelial layer of the esophagus can accommodate itself permanently to the shortened esophageal length. If the esophagus is continually shortening and lengthening like a camera bellows, then fold formation is necessary for storing the mucosal length needed in the extended phase while the organ is contracted.

LERs are a splendid example of the general rule formulated in Chapter 2: folds are orthogonal to the muscular fibers that cause them. Thus, the rule is as useful in finding the cause of a fold as it is in predicting how it will be arranged. Here, a transverse fold implies LMC as, for that matter, do reflux and HH - the usual concomitants of LER's.

Perhaps because it first called my own attention to the LM, the circumstance that most LER's vanish after a gastropexy of the Boerema type, seems to me the most conclusive evidence for the plication mechanism. How else can one explain the clearing of a pathologic finding when there has been no local attack on it? Elongating the esophagus by a pulldown procedure restores esophageal length, removes the mucosal redundancy and the pleat can no longer form.

The appearance and disappearance of the rings during a radiological examination has been explained by postulating that distention of the ampullary region is necessary to demonstrate them.(25) But this distention is normally achieved by the Valsalva maneuver. Obstruction in the PEL tent forces distention when the bolus cannot pass the sphincter. The region can be distended in other ways e.g., by performing a double contrast esophagogram or by rapid swallowing. If these do not produce shortening, a ring will not appear.

The symptoms of LER are intermittent and episodic. The attacks are often regarded as due to carelessness in eating. Typically, the patient leaves the table, tries to wash the obstruction out with water and, failing that, provokes vomiting. Patients may have only 1 or 2 episodes a year. Rapid eating or eating under conditions of excitement or stress are typical provocations. Symptoms are intermittent for the same reason the ring is not always visible radiologically - it is not always there. This intermittentcy is one of its most characteristic features. Obstruction by tumor or by stricture is a constant disability that progresses in severity. The reason for this is subtle.

Both manometry and radiography by custom only display the deglutition of fluids, for which peristalsis is virtually redundant. Eating, however, is concerned with the ingestion of semi-solid, particulate masses of varying size and consistency. A 2.5 cm marshmallow, for example, can be swallowed whole. Imperfectly masticated particles of meat of this size are sometimes ingested accidentally. For such to clear the esophagus, peristalsis is imperative.

For this reason, patients learn to eat slowly and chew their food well to avoid obstructive episodes, not by large food particles themselves but by the maximal

LMC they provoke. If esophageal transit is mainly by gravity flow, the LM contracts only slightly. If swallowing is against resistance, LMC and esophageal shortening is maximal provoking the ring.

The fact some rings of great chronicity persist after a HH repair suggests that local inflammation and fibrosis eventually fuse the two surfaces of the ring. Perforating veins will be ruptured when the ring first forms. The m. mucosae is split, as MacMahon et al. noted, a portion of it following the superior surface of the ring, a portion extending into the core of the ring and a portion continuing to form the m. mucosae of the stomach. Slight hemorrhage easily explains the usual residual of a few lymphocytes and fibrosis.

I am not aware that proponents of other etiologies have explained this splitting of the m. mucosae. In the case of fold formation, the reason for the split is obvious: once the lamina propria folds, it no longer follows the muscular wall of the organ and must sever its connection at some point. The cleavage is in the m. mucosae with some fibers adhering to the wall, some to the lamina propria and others simply ruptured.

The location of the ring at precisely the squamo-columnar junction can scarcely be fortuitous. The milking function of the peristaltic wave ends at the sphincter so the mucosal redundancy cannot be milked down into the stomach and lost. On the other hand, the p-wave doess not stop short of the sphincter. This is why these rings are not located proximally. The location of the ring below the sphincter is further proof of the pathogenesis and simultaneously a verification of the extent of the p-wave.

Because many rings are not fixed, this milking action of peristalsis may be essential to their formation. When the conventional full column study with Valsalva test is performed the detection rate according to Ott's group(26) was 97% whereas the double contrast method only detected 58% of the 60 rings they studied. The obvious difference is that there is no milking p-wave when air contrast is used. The 39% difference suggests an order of magnitude for the percentage of unfixed rings, however, the same group, in an earlier study,(27) could find only 17% of proven rings with air contrast, suggesting that most rings are not fixed.

The lower detection rate with endoscopy in the Bowman Grey series (35 of 58) reinforces this argument. The scope also elongates the esophagus rather than shortening it and, of course, there is no peristalsis during the examination.

Does inflammation cause the LER?

Because there is frequently reflux and esophagitis associated with HH and consequently with rings, inflammation has been an attractive hypothesis as an explanation of ring formation. However, this is post hoc ergo propter hoc

reasoning. If one tries to see how inflammation could cause the thin, web-like narrowing, he looks in vain for support from the histologic findings. Typically there are a few lymphocytes and plasma cells but rarely a polymorphonuclear cell.

Surgically, the webs cannot be palpated without opening the stomach. One would expect obvious induration if inflammation were present. Endoscopically, two thirds of the cases will show evidence of mild to moderate esophagitis. This does not explain the genesis of LER's in the third that have no esophagitis. Many patients with LERs have no reflux symptoms.(28)

Nowhere else in the GI tract does inflammation cause webs or rings. On the contrary, it causes long hourglass constrictions. Moreover, the appearance of inflammatory disease in the lower esophagus has already been spoken for by esophagitis. Superficial inflammation causes enlarged longitudinal folds to form. The same mechanism cannot be used to explain both longitudinal and transverse folds.

When inflammation is deep, the appearance is that of an inflammatory stricture. Inflammatory strictures invariably have a longitudinal extent several times the diameter of the organ they involve, whether it be the esophagus, Fallopian tube, urethra, ureter, large or small bowel. This is the exact opposite of the situation seen with LER's. If one advances an inflammatory theory of ring formation one must explain why this rule does not apply; why the ring can divide into two or more rings; why the submucosa is not involved; why a pulldown procedure obliterates them; why they occur where they occur; how the same cause can have two effects and so on.

Of course, inflammation cannot explain the choice of the mucosal junction as the location of the rings. When a HH is repaired by pull-down, the ring - if it disappears at all - vanishes at the operating table. This is not the slow resolution typical of inflammatory disease. A recurrence of the ring is associated with a recurrence of the HH(29) and reflux without recurrence does not reproduce it.

The demise of the two-pouch theory of the esophagus

One consequence of the LER and its definition as the true esophago-gastric mucosal junction is the downfall of the two-pouch theory. According to this there are a variety of dilatations of the lower esophagus. On one of these there is widespread agreement. This is the dilatation proximal to the ring that corresponds with what Templeton(30) called the "phrenic ampulla." Distal to the ring, variously identified as the gastroesophageal vestibule (by Lerche), the Vormagen of Arnold, the cardiac antrum of Luschka, the epiphrenic bell and the abdominal gullet favored by several British authors.

The mere fact that all these dilatations are below the ring means that they are lined with gastric mucosa. Without further evaluation this allows us to dismiss not only these "structures", but the large body of opinion, doctrine and speculation on which much of the treatment of disorders of the area is based. They are all unrecognized sliding transtractions.

That competent investigators accepted these interpretations for so long points up an interesting feature of HH: LMC pulling the fundus through the hiatus can convert it from a hemispherical to a tubular shape. This tube looks enough like the esophagus to mislead several generations of radiologists and anatomists. The distorted part of the fundus can take a set so that its tubular shape persists even after the HH that caused it has reduced. In this state, it was seen post mortem by the anatomists as it can occasionally be seen radiologically.

It is noteworthy that, although rings come and go with LM contraction, they nearly always reform in the same place. The preformed split in the m. mucosae facilitates the reformation in the same site rather than at a new one.

As a tissue is torn in the process and perforating veins ruptured, it is not surprising to find some evidence of round cell infiltration and not the profuse polymorph infiltrate characteristic of inflammation.

SUMMARY

LER's are transverse folds caused, like all folds, by contraction of the orthogonal component of the muscularis propria - in this case the LM of the esophagus. Its contraction causes mucosal redundancy that is milked as far distally as the p-wave can carry it, that is just beyond the LES.

Although HH, reflux and esophagitis are all seen with LER's, they neither cause the ring nor are they caused by it. The entire constellation of findings is caused by LMC.

Elongating the esophagus will cure the ring by removing the redundancy unless the two surfaces become adherent. The details of the histologic structure of the rings are only consistent with a plication mechanism.

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How the LM causes GE reflux

Introduction

Gastro-esophageal reflux (GER) is the preeminent esophageal disorder. It causes or accompanies most other esophageal problems, making a major contribution to the sum of human misery. In the United States alone, the provision of over-the-counter antiacid remedies was a $700 million to $800 million a year industry(1) in 1985 with sales exceeding even the $556,000,000 annual expenditures for cold remedies.(2) Cimetidine has passed Valium as number one in the marketplace.(3) About eight billion times a year, someone in the US is sufficiently distressed to take a Tum, Rolaid or Alka-Seltzer - a staggering statistic that does not even include the (forbidden) usage of bicarbonate of soda.

Although current opinion(4) leans toward the view that reflux is primarily due to hypotension of the LES,(5) I will show that reflux is an effect of LMC. Because an open sphincter will result in reflux, the next proposition to be proved is this:

LM contraction (LMC) is the efficient cause of reflux.

The force produced by longitudinal muscle contraction is resolved by the phrenoesophageal ligament (PEL) into two components: one component stretches the PEL, causing "hiatus hernia"; the other opens the sphincter, causing reflux.

The angle of His and the subdiaphragmatic esophageal segment, structures that form the theoretical underpinnings of surgical intervention, prove to be nonexistent. Once the lower esophageal sphincter is recognized as the sole defense against reflux, a wealth of clinical, surgical and radiological phenomena to be cited will show that this thesis is correct.

The clinical background of reflux

Several inciting factors are known to cause reflux. The most familiar are dietary indiscretions. Of the various foods that promote heartburn, many fall into a class of essential oils known as carminatives, nearly all of which lower LES pressure (LESP). Included are onions, garlic, peppermint, spearmint, cinnamon, dill, fennel, ginger, rosemary, caraway and cloves.(6) Onions are the most common offenders. There are people, particularly those who dine in restaurants, who have a great tolerance for onions and can eat them with impunity. To pyrosis sufferers on the contrary, they are poison. Because heartburn is delayed in onset, one cannot say that direct irritation of the esophagus is responsible. Either time is required for the active principal to take effect, partial digestion releases the inciting factor or a still more indirect action relates cause with effect.

Virtually every food that can cause an "upset stomach" is an inciting agent for heartburn - Tabasco sauce, hot peppers, Italian dressing, barbecue sauce, nutmeg, chocolate, smoky links, alcohol, histamine, fatty foods. It is significant that the "gas" producing foods - especially cucumbers and members of the cabbage family are on the list.

These and other substances known to provoke reflux are generally either stimulants or irritants. It would be more in keeping with the normal physiological pattern if the provocative agent stimulated an end organ than inhibited the sphincter. One searches in vain for an example of an irritant or stimulant that relaxes. All one can say with certainty is that there is an active principal that, introduced into the alimentary tract of susceptible individuals, will cause reflux of gastric contents into the esophagus.

Smoking (nicotine?) and coffee are inciting agents. Pregnancy has a long term provocative effect now considered due to endogenous progesterone. Birth control preparations may have the same effect due to exogenous progesterone.

The intermittent nature of reflux, its provocation by foods or drugs, and its consistency in a given individual set the parameters within which the search for a solution of the reflux problem should be conducted. Anatomy is constant; reflux is intermittent. Clearly, this shows that reflux is a physiological, not an anatomical problem. In some fashion, reflux must be tied to the chemical nature of the provocative agents. Strangely, the search for the cause and cure of reflux has focused on illusory anatomical structures.

Conventional theories of the antireflux mechanism

Of the proposed mechanisms for the prevention of reflux, only three have survived. The physiologic sphincter - that dominates the interest of basic researchers and clinical esophageal physiologists - the "subphrenic esophageal segment" and the slightly acute angle (of His) between the esophagus and the fundus shown in anatomy books. Because it still influences surgical treatment (Nissen fundoplication, Belsey Mark IV, Hill, and others), the latter must be considered in detail.

The angle of His

Johnstone(7) has suggested that the idea that the angle of His prevented reflux may have been founded on the clinical observation that reflux is rare in "paraesophageal" HH's - a type of HH with an acute angle of His.

We pay lip-service to the truism that living anatomy differs from that seen in the cadaver, but often not where it counts. The angle of His, if it existed, should be seen in normal patients on upper GI examination. It is not. Yet highly competent surgeons design anti-reflux operations to create this artifact.

Why is the angle of His seen in the cadaver and in the anesthetized surgical patient but not in the living, awake subject? Because in the dead or anesthetized the LM is atonic. In the living subject, the basal tone of the LM pulls the fundus up against the diaphragm, obliterating the angle seen in the cadaver or at laparotomy. When a surgeon applies traction to the lesser curvature of the stomach to visualize the hiatal area, he creates the angle by opposing LM traction. For much the same reasons - the LM expires with the patient or is overcome by traction and anesthesia - anatomists and surgeons have the impression that there is a subphrenic esophageal segment.

An angle of His is seen radiologically in abnormal situations when the LM tone is reduced or destroyed. If a patient who has had a pulldown procedure is reexamined before his hospital discharge, the trauma of operative stretching will have produced an angle of His that may persist for a week or more. It soon vanishes when LM tone returns. If the esophagus is paralyzed with anticholinergic drugs, it becomes flaccid and an angle of His may appear. When the esophagus recovers from the drug or the trauma, the angle vanishes.

An angle of His of an exaggerated type occurs in some cases of "paraesophageal" hiatus hernia. Here LMC cannot snug the fundus against the diaphragm because the fundus is above the diaphragm. With the stomach in the chest an esophagus of normal tone and length cannot take up the slack created by loss of its inferior attachment. Either an angle of His or a molar tooth shape results, the former if the esophagus slides by the fundus (it is sometimes called a "rolling hiatus hernia") the latter if it telescopes into it.

Radiologically, the only conclusion that can be drawn from an angle of His is that the esophagus is flaccid or a HH is present. These, however, are pathologic states. Paradoxically, we rely on a pathologic anatomical configuration caused by death, ligamentous rupture, surgical trauma or drugs to explain the normal competence of the cardia.

Yet, oddly enough, there is a remarkably good correlation between GE competence and an acute angle of His.

Patients in whom an acute angle of His has been created by fundoplication are usually greatly relieved of their reflux problem. Behar(8) refers to the Nissen fundoplication as ". . . the most effective antireflex procedure." not always

If counter traction is applied to the esophagus via a "pulldown" procedure, it relieves reflux and an creates an angle of His.

If the esophagus is inactivated with anticholinergic drugs (Banthine or Probanthine) the LM becomes flaccid, reflux is inhibited and an angle of His can form.

Finally, as already noted, patients with huge HH's - the kind diagnosed incidentally on chest exams - often have an acute angle of His and seldom complain of reflux.

This perfect correlation explains the confidence a surgeon might well feel in the rightness of his rationale. Explaining sphincter competence on the angle of His theory, however, is a post hoc ergo propter hoc fallacy. Reducing LM tension or its vector resolution by any means corrects reflux and, incidentally, may allow formation of an angle of His. If A and B are caused by C, it does not follow that A causes B. Both gastroesophageal reflux (GER) and obliteration of the angle of His are caused by LMC not by each other. On the contrary, if death, drugs, herniation, countertraction, trauma or anesthesia overcome the sphincter-opening force of LMC, GER is cured and the angle appears.

Other objections to the angle of His hypothesis

These logical fallacies are not the only objections to the angle of His rationale for GE competence. Structurally, it is difficult to conceive of an esophago-fundic angle as an effective valve capable of duplicating all of the sphincter functions, responding to reflex control, etc. The angle should form a flap valve that obturates the mouth of the esophagus with increased intragastric pressured. Radiographs of the region, many of them reproduced here, easily refute this idea. Even on a priori grounds such a valve can be excluded. Such a flap would have to be thin, but the sling fibers of the stomach that cause it when unopposed are not a thin band. They envelope the entire fundus.

The mechanism cannot be brought into conformity with what is known about the sphincter from manometric studies. How is this "angle valve"(9) integrated with the peristaltic wave? How is its action recorded manometrically? Is there an increase in intragastric pressure preceding sphincter closure? Where is the article, "Manometric differentiation of the LES from the Valve of Guberoff?"

Pneumoperitoneum was formerly done for the treatment of pulmonary tuberculosis and is still encountered occasionally with perforated hollow viscera. In such patients, as Johnstone noted,(10) the fundus hangs suspended from its esophageal attachments. It becomes merely a larger tubular continuation of the esophagus with an angle of nearly 180 degrees. Yet GE reflux is not a complication of pneumoperitoneum. Splenomegaly may produce the same configuration without causing reflux.

Again, we have recourse to Occam's razor. The angle of His concept is redundant. Demonstrably, there is a physiologic sphincter and it is not at the angle of His.

I have marshaled the evidence against the angle of His in some detail because, unlike some of the other conjectured closure mechanisms, it influences

treatment. The same cannot be said for the sphincter of Lerche, the Vormagen of Arnold, the cardiac antrum of Lushka, the gastroesophageal vestibule, the epiphrenic bell, etc. Most of these were "hiatus hernias" that the older anatomists did not recognize.(11)

The subdiaphragmatic esophagus

Another postulated antireflux system, the "subphrenic esophageal segment," is frequently invoked at least as a backup mechanism for the sphincter. There are reasons this idea has wide credence. When seen at laparotomy, traction on the stomach to obtain exposure pulls the anesthetized esophagus down to the limit of its tether - the PEL - creating the impression that the esophagus ends well below the diaphragm. The PEL is usually found to be stretched in patients having reflux operations for the same reason the patient has reflux.

Moreover, clinicians are accustomed to viewing radiographs of the GE region made in the RAO position. In a patient with a steeply sloping diaphragm, the hiatus is lower than the dome of the diaphragm and to the uninitiated this gives the appearance of a subdiaphragmatic esophagus. Radiographs are seldom made in the lateral position to show the true relationship.

A subphrenic esophagus is in many respects similar to the angle of His: it cannot exist if there is normal esophageal tone. LM tone would shorten the esophagus and pull the "subphrenic segment" through the hiatus until brought up short by the PEL. The PEL inserts at the mucosal junction sending layers above and below the sphincter(12),(13),(14),(15) so the latter must be above the obturating membrane.(16) Any intraabdominal pressure would not be backing up the sphincter, it would be below the sphincter. The distance from the lower edge of the sphincter to the ora serrata, however, is only .5 to 2.5 cm - scarcely long enough in most patients to make any difference even if it were subdiaphragmatic. But it is not. This short segment is also above the diaphragm.(17)

Most readers will recall having seen the surgical clips used in HH repairs on chest radiographs. Invariably, most of them are above the diaphragm once normal LM tone returns. When a hiatal margin (the distal edge of the PEL) is tagged with a clip at surgery, the investigators(18) have been surprised to find that on subsequent radiographs the clip is projected above the level of the diaphragm. Obviously, the hiatal margin is elevated when LM tone returns postoperatively.

It is difficult to explain the confidence with which manometric identification of a subdiaphragmatic sphincter is made(19),(20) when under the more physiologic conditions of a fluoroscopic examination, it is nonexistent. It must be recalled that manometric measurements have proved misleading in the past although they were universally accepted by experts for years. The technique is not as straightforward or as simple as might be supposed. There are numerous ways of

measuring the pressures under consideration - balloon kymography, intracorporeal strain gages, perfused and non-perfused catheters, transducers - and the measurements are seldom in agreement. The recorded pressure can vary appreciably with the rate of perfusion and even with the orientation of the sidehole. In accord with Laplace's law it also varies with the diameter of the catheter. The perfusion itself causes swallowing making absolutely baseline conditions unattainable.

Swallowing a sizable collection of tubes - as many as 8 and at least 3 are recommended to average out the readings - is a stressful experience initially and continues to be so as the apparatus is withdrawn and reinserted many times or while the patient is in unusual or uncomfortable positions. Moreover, as can be seen in the illustrations, the esophagus senses the presence of a foreign body and reacts by contracting the LM which has the effect of decreasing sphincter pressure and/or hiatal squeeze - whichever is being measured.

The physics of the situation is immensely complicated and it is very difficult, perhaps impossible, to calibrate for and correct all possible sources of error. The expression "in our laboratories" - often meaning "Our measurements don't agree with anybody else's, but we are internally consistent." has been a frequent occurrence in the literature of the subject. Finally, it seems likely that manometry cannot as a rule differentiate hiatal squeeze from sphincter pressure. It is likely, however, that hiatal squeeze pressure is a proxy for LMC and thus for the LES as well.

This caveat is not meant to denigrate a useful methodology that is clearly at a disadvantage in measuring absolute pressures. I do think, however, in the matter of the subphrenic extent of the esophagus, it is appropriate to believe what is plainly visible rather than a strip chart that requires elaborate interpretation.

However, as with the angle of His, if a subdiaphragmatic segment is created surgically - whether it be by a pulldown gastropexy a la Boerema, by a fundoplication, by inserting a silastic appliance a la Angelchik or by simply creating one out of stomach a la Collis - reflux will often be alleviated. This encourages another instance of the post hoc fallacy. Each of these procedures in some way interferes with vector resolution of the force of LMC: by creating a slack PEL (Boerema), by changing the angle of resolution (prosthesis), by extending the esophagus (Collis) or by destroying the PEL. The beneficial effects are not due to the intended rationale.

The physiologic lower esophageal sphincter

A major outcome of the manometric methods pioneered by Fike, Code, Ingelfinger and their schools has been the nearly universal acceptance of the existence of a physiologic sphincter of the lower esophagus.(21) This was a giant step in the direction of understanding reflux even though manometrists misplaced

it and radiologists did not recognize it. Physiological research is now concentrated on understanding the role of the sphincter in preventing reflux. The understanding is presently hampered by a fundamental limitation of the instrumentation employed: it is 2-dimensional due to its inability to "see" the LM.

A 1986 report by Dodds, et al.(22)(23) illustrates the problem. This group measured the mean 12 hour lower esophageal sphincter pressure in patients with clinical and esophagoscopic evidence of esophagitis and in a control group without esophagitis. The mean pressure at the LES in the control group was 29 9 mm of Hg. In the patient group it was less than half as much - 13 9 mm. This suggests that a normal (high) average LES sphincter pressure, thought to be a measure of the tone of the circular sphincter muscle, prevents reflux. Yet, 4 of the patient group had mean 12 hour LES pressures that fell in the normal range.

Also unexplained by the hypothesis that a normal LES tone prevents reflux was the finding that transient complete LES relaxation occurred in both the control and esophagitis patients. Even when the measured LESP was reduced to 4-5 mm Hg, reflux did not always occur. In the controls, only 34% of those with complete sphincter relaxations had reflux; the other 66% with the same transient relaxations did not have reflux. In the patient group the results were almost exactly the opposite: 2/3 of the relaxations were accompanied by reflux. In short, there is a 33% overlap of controls and patients - 1/3 of those with normal mean pressures reflux and 1/3 of those with low sphincter pressures do not.

On the other hand, administering a drug believed to increase LESP (metoclopramide 10 mg q.i.d.) produced no significant correlation between increased LESP and decreased symptoms.(24)

Although there is evidence that 80 mm Hg pressure in the stomach will not force the sphincter, Dodds et al. found that even a minimal LES pressure of 4-5 mm Hg was enough to prevent reflux. The study showed that transient complete sphincter relaxation is the cause of reflux - not low resting LES pressure.

In an earlier study,(25) it was found that reflux episodes tended to be "inappropriate." They might occur without any other esophageal motor activity or with random, non-peristaltic activity. These results were confirmed in dog experiments by Patricos, Martin, Dent et al.(26) who also established that belching was initiated by a single transient complete LES relaxation and that it did not occur as long as a measurable LESP existed.

Euler and Byrne(27) studied 49 infants and children under 9 years with 24-hour pH probe testing. Although the symptomatic and asymptomatic groups were sharply differentiated by the number of reflux episodes and their persistence, there was no significant difference in LESP between the two groups. (21.1 1.7 vs 21.7 1.7) Hillemeier et al.(28) found normal or increased LESP in children under 2 years with severe GER.

Despite the work of the Milwaukee group, clearly indicating that 1.) low LESP per se is not the answer to the problem of reflux, and 2.) reflux is due to transient complete sphincter release,(29) great activity is focused on the physiology and pharmacology of the sphincter. Earlam [1975] probably stated the consensus echoed in current texts(30) when he said, "Since the pathology is most likely an intrinsic defect of the gastro-esophageal sphincter and the lower esophagus, the ideal treatment would be to tone up the sphincter and increase the efficiency of secondary peristalsis . . . " [emphasis added]. Behar(31) after reviewing the literature, concluded the causes of LES incompetence were unknown.

Sweeting(32) notes, "There is no adequate explanation for these seemingly random drops in sphincter pressure. None of the factors studied have been shown to be paramount in determining basal LES pressure."

The LES has been intensively studied both in vivo(33) and in vitro. In his 1982 review, Diamant(34) cites a large number of results. Cholecystokinin octapeptide (CCK-OP), for example will decrease sphicter tone in the cat, but increases it in the opossum. It has the reverse effect on both animals if the LES is chemically denervated with tetrodotoxin (TTX). Progesterone lowers LES pressure during pregnancy or if given as medication.(35)

Stimulation of the cut peripheral end of the greater splanchnic nerve increases LES tone 300% in the cat but reduces it 50% in the opossum. Stimulating the central end of the splanchnic nerve decreases LES pressure in both animals. Significantly, CCK-OP decreases LES pressure in normal humans, but increases LESP both in patients with diffuse muscle spasm and in those with achalasia. CCK-OP has the same paradoxical effect on the cat after TTX(36),(37),(38) However, it is hard to attach great weight to the finding because CCK-OP produces a decrease in LES pressure in the chemically denervated opossum. CCK-OP also causes ". . . forceful LMC" in the opossum.(39)

Clinically, "The poor correlation of LES hypotonia with reflux esophagitis suggests that in many patients the occurrence of reflux is either determined by nonspincteric factors or by features of sphincter activity other than basal LES tone." (40)

It is difficult to interpret such findings, in part because of species variation, in part because some are done on intact animals and others on isolated preparations, but principally because LMC is not taken into account. An observed decrease in LESP, for example, could be due to LMC but undetectable with the instruments used. In studies on intact animals, if a single orifice manometer is used, the sphincter may be drawn up above the catheter orifice by LMC to create a false reading of decreased LES pressure. Without repeating the experiments, it is uncertain whether the LES pressure drop is due to the effect of the drug or stimulus on the sphincter muscle directly or indirectly via the LM.

Edwards(41) was unable to measure sphincter pressure after vagal stimulation because the "violent" contraction of the LM on stimulation of the vagus moved the sphincter off the manometer tip. There is no feasible way of obtaining quantitative measurements of LMC and LESP simultaneously in man. Harrington, et al.(42), using a more elaborate experimental setup in the opossum also found that vagal stimulation caused LM contraction and LES relaxation and that LESP was correlated with LM contraction and relaxation whether spontaneous or pharmacologically induced. The location of the sphincter in this animal is 5 cm below the diaphragm so this effect must have been purely neurologic and independent of the vector resolution of LM force by the PEL.

The demands placed upon the LES make it almost unique among sphincters. With the exception of the cricopharyngeal sphincter, it is the only sphincter that must function in the orad as well as caudad directions. It must pass fluids and solids with a minimum of obstruction and yet be a firm barrier against the incursion of the corrosive, not to say repugnant, contents of the stomach. It must allow discharge of the air that is constantly being swallowed at a rate of a few cc per minute, yet nip off the escaping gas before the swiftly approaching gastric fluid level passes its portal. On occasion, nevertheless, it must yield to massive discharge of gastric contents in vomiting while preserving its structural integrity for immediate resumption of its normal function.

The integration of these functions with the circular and longitudinal muscular components of the esophagus in swallowing fluids, in swallowing against resistance and in belching has been described in the appropriate chapters. Here we are concerned with a malfunction in which the LES is open when it should be closed. It would be very strange if this pathological opening of the LES were not due to the same force that opened it physiologically in swallowing, belching and vomiting. Like people, whose faults are rooted in the same qualities as their virtues, malfunction is of a piece with physiology.

LMC and reflux

Briefly stated, my thesis is that increased LM tone is responsible for GER.

LMC -->

Conceptually, LESP could be zero and reflux still would not occur unless the sphincter were opened. Unless affected by outside influence, LESP is never absent because of the intrinsic tone of the sphincter. This is not overcome even by chemical denervation with terodotoxin.(43),(44) Moreover, the sphincter cannot actively open itself. It requires either a distending bolus(45) or the services of the LM.

Before integrating the LMC mechanism with the Cannon-Dougherty reflex (CDR), it is appropriate to review the evidence that LMC can or does open the sphincter. The main points can be listed:

GER is a component of the tetrology of HH, LER, tertiary contractions and reflux. The mutual associations favor a common cause. I have shown or will show that LMC causes HH, LER and TC.

In scleroderma, a disease in which the esophagus is shortened, there is gross GER and HH.

LMC must occur before sphincter release in belching.

Forceful LMC occurs with sphincter release in vomiting.

Observation of patients swallowing against resistance shows that LMC is an integral part of the peristaltic wave. LM contraction during the phase that the sphincter is open and its relaxation synchronously with sphincter closing are meaningless unless the LM has a sphincter-opening function.

The experiments of Torrance(46) demonstrated that the LMC induced by vagal stimulation produced reflux whether or not the sphincter was denervated. H. Daintree Johnson(47) found similar responses in the rabbit. Like Torrance, he found that simple traction on the esophagus through a neck incision caused a gaping cardia.

Vector resolution of the force of LMC opens the sphincter by pulling the opposing surfaces apart. This is a purely mechanical result independent of the pharmacology or neurophysiology of the sphincter, so that we can say, If the LM contracts and the PEL is intact then sphincter-opening vectors will inevitably be generated.

When PEL rupture converts a sliding HH to a "paraesophageal" HH the force vector cannot be resolved and reflux is relieved. This is why the giant HHs in the elderly are generally asymptomatic.

All operations that alleviate reflux do so by destroying the PEL or changing its direction thus impairing the ability of the PEL to resolve LMC into a sphincter-opening force.

Non-effacement of the sphincter is a frequent consequence of PEL rupture.

Fluoroscopic observations of belching, nausea and vomiting demonstrate that LMC is associated with these sphincter-opening events.

Disabling the LM by drugs or surgical trauma can relieve symptoms caused by reflux.

In myotonia dystrophica, a disease in which the LM cannot relax normally after contraction, the sphincter may be always open with a resulting air esophogram.(48),(49),(50)

Shortening the esophagus causes reflux.

The effects of shortening and lengthening the esophagus provide an independent line of proof that the LM opens the sphincter. It is usual for esophagus-shortening operations [that tense the PEL even without LMC] to produce reflux. After repair of a tracheo-esophageal fistulae (TEF) the most common complications are reflux, dysphagia and recurrent aspiration pneumonia. There is no correlation between the size of the postoperative lumen and the patient's clinical symptoms.

Vanhoutte et al.(51) studied whether the reflux was due to interference with peristalsis. They found that resection of 2 cm of the esophagus in newborn dogs did not result in a loss of the peristaltic wave below the resection site. Their speculation that the postoperative complications were due to a ". . . . coexistent congenital abnormality of the vagus nerve."is tantamont to an admission of ignorance. Jannsens has demonstrated that the peristaltic wave also survives vagotomy.(52)

The reflux that occurs after repair of a TE fistula demonstrates that surgical shortening of the esophagus, even though it does not affect peristalsis, will cause reflux. The effect is quantitative: the longer the gap between the blind esophageal pouches, the more shortening required to make the anastomosis. Greater shortening is likely to be necessary in cases of isolated esophageal atresia and some variants of TEF. Because postoperative reflux is especially common in these variants, they are the remaining indications for a 2-stage procedure.(53) In one large series(54) 40% of the deaths were due to pulmonary complications, i.e., aspiration pneumonitis. Hands and Dudley(55) found the gap length the most important predictor of subsequent mortality and complication rate. There were GE reflux complications in 83% of those with a gap of over 2.5 cm but in only 33% in those with a lesser gap.

The complications of TEF repair are such that the operation is a human experiment in producing the syndrome of LM tension. The traction on the lower esophageal segment necessary to approximate the transected esophagus after removal of the atresia can and does cause the three complications of excessive LM tension - strangulation, hiatus hernia and reflux. Although experimental esophageal transection does not produce motility disorders in the dog,(56) opossum or rhesus monkey,(57) a motility disturbance that resembles diffuse spasm (tertiary contractions) and dysphagia for solid food frequently complicate TEF repair in later life(58) because of the esophageal shortening.

TEF repair does not, of course, directly attack the sphincter or the hiatus. (59)The procedure has its effect remotely because like LMC, it creates tension on the PEL that opens the sphincter.

Indeed, any anomaly that applies traction to the PEL may result in an open sphincter. Vascular rings that elevate the esophagus often cause an air filled esophagus with an open sphincter.

Lengthening the esophagus alleviates reflux.

The most severe cases of reflux are those with esophageal strictures. Resection of a portion of the esophagus, e.g., for stricture, shortens it and leads to recurrence. If, however, the esophagus is lengthened by the Collis(60) procedure, the success rate is reported to be 75% despite what is essentially surgical creation of a Barrett's esophagus.(61) Even these failures can be treated by interposition of a segment of left colon or jejunum.(62) Clearly, just as shortening of the esophagus promotes reflux, lengthening it counteracts reflux by relieving its tension.

Coordination of sphincter, CD reflex and LMC

Given a normal range of LM tone, for the sphincter to open, the Cannon-Dougherty reflex (CDR) must be OFF. This is a necessary, but not sufficient condition. The LM must be ON. Three of the components of GER - LM, sphincter, and the CDR - can be arranged in a Truth Table (Table 1).

A simple "circuit" controls the sphincter when all is well: it is closed unless the CDR is turned OFF and the LM is turned ON. Note that this control scheme does not give the sphincter itself any place as a prime mover. It is completely under the joint control of the LM and the CDR.

This schema is not in conflict with the extensive studies of the pharmacologic control of sphincter tone; reflex control must to be mediated via neurotransmitters in any case. The effect of LMC, however, appears to be primarily mechanical. The sphincter snaps shut as soon as LMC stops.

Sphincter control is not entirely an all or none affair regulated in digital fashion, at least as far as the LM is concerned. Given an intact PEL, the LM can always open the sphincter if only it contracts forcefully enough.

TABLE 1

Truth table for the lower esophageal sphincter

CDR	LMC	SPHINCTER	Effect
OFF	OFF	CLOSED	Baseline tone closes sphinct.
OFF	ON	OPEN	LMC Opens Sphincter
ON	OFF	CLOSED	CDR prevents opening
ON	ON	CLOSED	CDR prevents opening

Diagnosis of reflux

Many studies of reflux are needlessly complex, uncomfortable and expensive. Leasing the equipment for pH monitoring can run several hundred dollars a day. The radiologic diagnosis of reflux can be highly specific and uncomplicated. At the moment the de Carvalho maneuver elicits reflux one simply asks the patient four questions:

1. Do you feel anything unusual?

2. Have you had that sensation before?

3. Is this like the symptom that has been bothering you except in degree?

4. Is it in the same place as that symptom.

Four "yes" responses leave yield more certainty that reflux is the cause of the patient's symptom than any strip chart. As usual, there are caveats. Negative responies do not exclude reflux. If too much water is used to elicit reflux, gastric HCl may be too dilute to elicit the symptom. A few patients with life-long reflux are so accustomed to it they believe it is normal.

The association of hiatus hernia and reflux

"Most physicians have for many years associated . . . gastroesophageal reflux with an anatomical hiatus hernia."(63) In a typical report, Edmunds(64) found radiologic evidence of reflux in 93% of sliding HH and 42% of "rolling" HH's. Wright and Hurwitz(65) compiled a chi-square table of 293 patients calculating that the probability that the association between HH and esophagitis was due to chance was less then 1:10,000.

	HH +	HH -
Esophagitis +	32	6
Esophagitis -	32	223

The notion that a 2 with a low P proves causality dies hard. Because of the association, it was it was natural to assume that one caused the other so this view prevailed. As recently as 1994 Paterson and Kolyn,(66) based on their finding that acid perfusion of the opossum esophagus caused it to shorten, conjectured that esophagitis caused HHs. As a result, HH repair enjoyed a long vogue as the accepted treatment for GER.

As the repairs were often less than satisfactory, surgeons directed their efforts toward designing operations that would deal with reflux per se and not simply correct a HH as before.(67) Nevertheless, the strong association remains - with a difference. It can no longer be attributed to cause and effect, but, if it is not cause and effect, why the association? Dodds, et al., however, suggested, "Perhaps hiatal hernia and GE reflux are related to a common cause rather than being related causally to each other."(68)

The hypothesis we are proving nicely solves the dilemma as it shows that HH and reflux occur together because, as Dodds suspected, they do have a common cause, LMC. Looking at it from the opposite way, this association further proves the validity of the sphincter opening mechanism. If there is long continued excessive LM tension sufficient to cause stretching of the PEL (i.e., gastric transtraction) one would expect that there would also be excessive sphincter opening activity (i.e., reflux).

One cannot completely rationalize the considerable complexity of sphincter control with a one-factor mechanism. It seems that there are at least six and probably more factors involved:

The sphincter-opening force generated by LMC

The ON/OFF status of the Cannon Daughterty reflex.

Whether the contractile mode of the LM is peristaltic, tonic, clonic or anaspartic.

The integrity of the PEL.

The basal tone of the sphincter

The contents of the stomach

It will take a more powerful LMC to open the sphincter if the CDR is ON or if there is a high intrinsic sphincter tone. We can see this in the events that preceded a belch. LMC will tent the PEL, then subside without an actual release of gas then again contract, perhaps with a more pronounced tenting as the cone of gas elongates into the sphincter region. In emesis mode, of course, such is the power of the LM, the sphincter is forced even if the CD receptor is activated.

The role of gastric contents in reflux.

We can belch without regurgitating acid because as soon as the gastric fluid level reaches the CDR receptor, the LM is inhibited and the sphincter closes. Watching this process with the fluoroscope, one has to wonder how a reflex mediated by a chemoreceptor can be so quick.

Production of gastric mucus can explain both the delayed onset and the intermittent nature of heartburn related to ingestion of certain foods. As the CDR receptor must be stimulated chemically, mucus coating the stomach will prevent gastric acid-pepsin from contacting the sensor and activating an inhibitory reflex.

From the appearance of surgical specimens, it might be thought that there is copious mucus coating the stomach wall. Radiologically, this is not true. Barium normally coats every fold and crevice displaying the mucosa in sharp detail. If mucus is present, it prevents barium from adhering to the gastric mucosa giving the stomach a "wet," greasy, appearance and causing barium particles to clump. Most radiologists recognize this as an alert to gastritis or irritation of the gastric mucosa associated with duodenal disease.

If it could be shown that carminatives cause excessive mucus production this would tend to show that they cause heartburn by coating the CD receptor with mucus and so suppress the CD reflex sphincter closure. The delayed (45-60 minutes) effect of these substances in promoting reflux(69) would fit this mechanism. It also is a rationale that accounts for the paradox of an irritant causing sphincter relaxation.

Bickel and Kauffman(70) developed methods of measuring the thickness of the gastric mucus layer. They found that distention of the stomach stimulated release of mucus. "The gel mucus layer overlying the gastric mucus is constantly being produced by the surface epithelium and is constantly being eroded within the lumen by the action of acid, pepsin, and stirring of the luminal fluid." The application of certain substances such as prostaglandin E2 could increase mucus thickness.

In theory, there are other ways of turning off the CDR. Local surface anesthesia produced that effect in ruminants(71) as did section of the dorsal trunk of the vagus. Ingestion of a surface anesthetic may explain the effect of procaine noted by Balfour.(72) The deCarvalho maneuver or water siphonage test turns off the guard reflex by washing it free of acid-pepsin. Released of the reflex inhibition, basal LM tone overrides basal LES tone.

It could also be true that a more complex set of receptors feeds a central program that calculates gastric shape. A sudden loss of distention on release of gas from the stomach would then signal the LM to relax.

The cervical spine and reflux

It will be recalled that an elevation of the mouth of the esophagus initiates a swallow thus applying tension to the entire esophagus and through it to the PEL thus facilitating opening of the sphincter.The upward impulse of the larynx that initiates a swallow may also activate a stretch reflex causing further contraction of the LM. In drinking liquids this and gravity are all that are required. Usually no peristaltic wave results except as a final cleanup.

If increasing LM tension facilitates sphincter opening, it is worth considering whether decreasing LM tension would improve sphincter competence thus decreasing the amount or frequency of reflux. In principal, it would seem that ceteris paribus, shortening the distance between the ends of the esophagus should relieve the tension on its attachments. This can be done very simply by anteflexing the cervical spine.

Although unintended, cervical anteflexion is invariably a part of the standard treatment for reflux. Without exception, authorities advise sleeping with the head elevated. Effectively, this is a prescription for cervical anteflexion. Typically, a patient is told to sleep with his/her head elevated.(73),(74),(75),(76),(77),(78),(79) The physician assumes that the patient will elevate the head of the bed with bed blocks as is done in hospitals. The patient however, thinks he/she has been told to sleep on two pillows, does so and gets relief! They would get as much relief by sleeping on a 28" x 10" foam wedge.(80)

There is no rationale for bed blocks

The only conceivable rationale for suppressing reflux by elevating the head of the bed is to affect hydrostatic pressure gradients. We accept that explanation because we are aware we are doing something right(81) and nothing else comes to mind.

A elementary calculation with similar triangles shows that elevating the head of the bed does not change the hydrostatic pressure differential more than a few cm(82) of water. It takes 80 mm Hg or more of hydrostatic pressure to force the sphincter, so .28 mm Hg is not going to tip the balance in favor of sphincter competence. Moreover, even 6 inches of elevation makes people slide out of bed and is likely to be objectionable to a patient's spouse. The bed-raising may tend to get the CDR receptor out of the pool of gastric secretions, but this should promote reflux. Moreover, the esophagus is bowed posteriorly by the heart so that this slight adjustment makes no difference.

Curiously, people have reflux even when the pressure differential is maximized in the upright position. In a study of 100 patients with and without HH, Johnson(83) found that the number of episodes of reflux per hour was three times as great in the upright position as in recumbency! So elevating the head should promote

reflux! How can one explain such a paradoxical result by hydrostatic pressure? Even if it were just a matter of acid pouring out of the fundus through an open sphincter, it would be paradoxical. The universal medical recommendation to sleep with the head elevated is a remarkable triumph of empiricism over logic.

On the other hand, anteflexion of the cervical spine occurs in recumbency on pillows. This explains the favorable effect of this advice and Johnson's results become predictable. The prescription works because the doctor's instructions are either misunderstood or tried and, after being found intolerable, no nurses being around to enforce them, modified. Cervical anteflexion can introduce nearly the same amount of esophageal slack as a Collis procedure and is effective for the same reason.

An unsolicited testimonial for this rationale is provided by Garretts,(84) who, in reported apthous-like denudations on the buccal surface of the lower lip in three patients with reflux, mentioned that the second of these dated his symptoms to a fall in which he hurt his neck. Thereafter, he "...could not use his usual number of pillows..." but had to sleep lying flat in bed. He was advised to raise the head of the bed!

A useful maneuver

Those readers occasionally afflicted with reflux may test these conclusions on their persons on such occasions by firmly forcing their chins down toward their chests. A measure of relief may be experienced almost at once. While the maneuver is not a miracle cure, it can make the difference between going back to sleep and a trip to the medicine cabinet.

This maneuver is about equally effective - and for the same reasons - in suppressing a wave of nausea or an incipient belch. A patient may be able to ward off an attack of angina-like chest pain with the maneuver.(85) As a rule, the body knows how to adjust its position so as to minimize discomfort. It is surprising, therefore, that this is not a posture that everyone discovers for himself as, for example, most people discover that leaning forward and hyperextending the cervical spine will facilitate a belch.

Cervical dorsiflexion can cause reflux.

Just as anteflexion seems to slack off the LM and alleviate its sphincter-opening effects, there is a remarkable association between dorsiflexion of the cervical spine, contraction of the LM of the esophagus and reflux. As might be expected, it has precisely the opposite effect. It tends to open the sphincter. The following 4 cases (for 2 of which only my notes survive) illustrate the extent to which dorsiflexion causes a striking degree of LMC.

An opportunity to observe LMC in its severest form was furnished by an extraordinary patient, male, age 36 who was seen for dysphagia, choking, laryngospasm, heartburn, etc.

With each swallow, the LM contracted en masse producing a trumpet configuration of its lower segment, herniating the stomach and opening the sphincter. The latter remained wide open for more than 30 seconds at a time. As the hernia started to reduce, the sphincter would begin closing, only to reopen as an increase in the force of LMC again elevated the stomach above the diaphragm. A further finding of interest was the accidental discovery that to provoke this "tetanic" LMC, it was only necessary to hyperextend the cervical spine. This maneuver, of course, put the esophagus on stretch and elicited the LMC response just as an abrupt flexion of the ankle can produce clonus.

On review of the cine film of the case, one is impressed with the strength and elasticity of the PEL that can withstand a force of this magnitude without rupture and without loss of elasticity.

The chief significance of this unusual case, seen in the early 1960s was that it illustrated an extreme degree of LMC and thus made it easier to recognize lesser degrees of the same condition in others.

CD/40585 This 45 year old male exhibited many signs of a hyperactive, hypertonic LM. The presence of a trumpet shaped HH, gross, spontaneous GE reflux and marked reflux in response to the de Carvalho maneuver were all considered manifestations of this primary abnormality. The esophageal sphincter remained open for long periods even in the upright position so as to suggest scleroderma initially. Dorsal flexion of the neck produced reflex contraction of the esophagus bringing on the above effects.

An even more striking example, for which I have been unable to find a comparable report in the literature, was seen a few years later.

The patient was a man in his 20s who was brought to the department in what appeared to be a bizarre type of convulsion. Extreme cervical hyperextention occurred spasmodically as the patient thrashed about on the x-ray table so violently it required several aids to keep him from falling off. He was able to cooperate in swallowing barium, but his motions were so erratic the lower esophagus could not be retained in the 4"x 5" field of the cine camera.

The barium-coated esophagus was alternately air filled and collapsed. The filling occurred when he hyperextended his neck. At this time the esophagus contracted longitudinally opening the sphincter and allowing air to escape into the esophagus. This would be followed immediately by an en mass contraction of the circular muscle or Valsalva effort that forced some air back into the stomach.

These activities were accompanied by loud eructations of gas via the superior constrictor as well.

Such dramatic instances are not necessary to demonstrate the effect. It can be seen in almost any patient with reflux and/or HH.

A 59 year old schizophrenic with severe esophagitis and HH was studied. When an assistant dorsiflexed the cervical spine, the HH could be seen moving up and down in the hiatus. Reflux occurred at maximal dorsiflexion.

In all these cases, one had the impression that there was a "knee jerk" reaction such that stretching the LM by dorsiflexion induced contraction just as a percussion hammer, by stretching the quadriceps tendon, causes reflex contraction of the extensors of the knee.

Sandifer's syndrome

Also noteworthy in this connection is the equally bizarre Sandifer's syndrome seen in children. It is marked by head rolling, hyper-extension of the cervical spine, hiatus hernia and reflux.(86),(87),(88) The children began the movements during eating and seemed to get some relief from them. They were not present during sleep. The preferred posture was supine in bed with the head hyperextended over the edge of the bed like the comic strip character Lou Ann. They learned to watch television upside down in this position. All had HH with reflux and vomited at mealtimes. Radiological examination of 5 such patients including Sandifer's by Sutcliffe established that the fundus of the stomach was elevated and the HHs sharply increased in size with the dorsiflexion movements. "The stomach would bob up and down in time with the neck movements." "The neck contortion would immediately be followed by substantial elevation of the GE junction and temporary entry of a further portion of gastric fundus into the thoracic cavity."

Although the movements were so bizarre they suggested basal ganglia disease, they cleared completely after hiatus hernia repair!(89)

A later series of 13 cases mimicking other neurological syndromes was reported by Bray, et al..(90) Most were infants with torticollis, opisthotonic posturing and seizures whose symptoms cleared with postural treatment of their reflux and HH'S. As all of the children either had dysphagia or vomited at mealtimes, it seemed that there was some obstruction of the esophagus that putting the esophagus on stretch relieved. This would have the same sphincter-opening effect as would LMC.

On followup of 31 patients with mental motor retardation who had been subjected to Nissen fundoplication for severe GE reflux, Williams and associates(91) reported that the only factor of prognostic significance in predicting a poor result

was " . . . chronic opisthotonic posturing . . . " that occurred in 80% of failures. Like Sandifer's syndrome, such a posture forces severe cervical hyperextension causing esophageal traction.

Such cases establish the connection between esophageal tension and both HH and reflux and show how movements of the neck affect the lower esophagus. They provide a convincing demonstration that cervical dorsiflexion can produce HH by esophageal traction and that the same LM tension is associated with reflux.

"Whiplash dysphagia"

Orthopedic surgeons have long been mystified that their whiplash patients have dysphagia. I examined three such patients who complained of heartburn and lump in the throat. Although under 35, they had HH's with ruptured PEL's. The hyperextension of whiplash injuries exerts enough sudden stress on both the proximal and distal attachments to rupture the latter and cause local injury to the cricopharyngeus.

Diaphragmatic paralysis for reflux

The "knee-jerk" analogy may also explain the success reported treating reflux by phrenic nerve crush in patients considered poor risks for major surgery. Relying on conventional doctrine, surgically paralyzing the diaphragm seems so lacking in rationale that Earlam cites it with unconcealed skepticism: " . . . if the results are to be believed - paralysis of the diaphragm on the left side does, for some unknown reason, relieve symptoms."

But paralyzing the left hemidiaphragm also gives the esophagus a few centimeters of slack - perhaps as much or more than cervical anteflexion - and this in turn takes the strain off the PEL preventing a stretch reflex in the same way that extending the knee diminishes a knee jerk. Seen in this light,(92) phrenic nerve section is just as rational as sleeping on 2 pillows or a pulldown procedure or lengthening the esophagus.

Esophageal slack reduces reflux

Traction of a hypertonic LM on the sphincter is alleviated by anything that introduces slack in the esophagus. This could be expected to lessen the ability of LMC to produce reflux by opening the sphincter. In experiments on the surgical production of hiatus hernia, Giuseffi, et al.(93) did exactly this. They created a partial intrathoracic stomach in dogs by cutting the PEL and suturing the stomach to the hiatal margins. They observed less esophagitis in dogs so operated than in others in which the left crus was cut to allow a hernia to occur.

The Collis procedure, an anti-reflux operation in which a tube is fashioned from the stomach to provide what is in effect an esophageal extension, is said to be effective because it " . . . eliminates tension on both the repair and the intrathoracic esophagus."(94) Again, elongating the esophagus has the effect of reducing the tension it can apply to the PEL.

PEL rupture alleviates reflux

Elongating the esophagus, flexion of the cervical spine and raising the stomach all achieve their effect by reducing the LM tension on the PEL. There is, however, still another way of reducing the sphincter-opening potential of the LM. Because vector resolution of the force of LMC by the PEL opens the sphincter, transecting the ligament should destroy this resolution thus alleviating reflux. Then LMC, no matter how forceful, would be ineffectual at producing reflux.

The patient in the following case had a symptomatic remission after a sliding HH converted to a non-slider:

25117 AC, M age 69. On 10/21/63 the patient had a sliding HH, gross reflux and a lower esophageal ring. Then he had genuine dysphagia (i.e., not lump in the throat), pain in the right side of the chest and interscapular region as well as symptoms of regurgitation. On the present occasion, all these symptoms have cleared and the patient states he has been asymptomatic for the last 3 months. 10/18/65

Fluoroscopic note: Barium passed freely through the esophagus. The GE junction was 7.5 cm above the diaphragm. Although there was a pinchcock-like appearance at the diaphragm on straining, this was not tight enough to prevent barium from leaving the herniated portion of the stomach idicating rupture of the PEL. The hernia had increased in size by a factor of 100% and the LER, that formerly had fairly sharp, ledge-like margins, on this examination had blunt, lip-like margins.

Although no operation is limited to simple severance the PEL(95), nature has provided an experiment that verifies the above prediction. Many cases of GER are eventually self-limited. It is common experience that the huge HH's seen on admission chest films of the elderly are often asymptomatic. Barrett(96) does not even list pyrosis among the complications of the "paraesophageal HH." Johnstone's comment on the origin of the angle His theory of GE competence will be recalled. The large "rolling", "paraesophageal" hernias were so asymptomatic that surgeons began to mimic their features surgically.

With age and a loss of elasticity, the PEL can rupture, converting what was a slider to a non-slider, molar tooth, type HH. After this the force of LMC can no longer be resolved in such a way so as to open the sphincter. LMC merely shortens the esophagus, pulling anything attached to it through the hiatus. This is

the reason that so many patients with very large HH's are asymptomatic: rupture of the PEL has pulled the LM's sting.

Many operations that are unsuccessful in their original objective incidentally sever the PEL.(97) We have noted that operative results are far better symptomatically than one would expect with the observed number of recurrent HHs.

Patients with ruptured PELs do not escape entirely unscathed, however. As would be predicted, without the assistance of the PEL they may have a problem effacing the sphincter. Radiologically, the distal few centimeters of the esophagus present as a short tapered segment or ring like narrowing that does not dilate with rapid swallowing of large barium boluses. The result may be a mild to moderate dysphagia. Many cases now labeled "terminal esophagitis" are probably examples of non-effacement of the sphincter.

Finally, as has been noted in the hiccup chapter, a sudden downward motion of the diaphragm will release the sphincter by tensing the PEL. This explains the association of reflux with hiccups.(98)

SUMMARY

Gastro-esophageal reflux is the commonest of all gastrointestinal disorders. Because the physiologic sphincter is the sole defense against reflux, its proximate cause is sphincter incompetence. Its fundamental cause, however, is the excessive tension on the PE ligaments that is commonly produced by increased tone or activity of the LM of the esophagus. The PEL resolves the force of LMC into two components. One component stretches or even tears the PEL, leading to hiatus hernia. The other opens the sphincter causing GER.

Whatever tenses the esophagus promotes reflux. Shortening incident to repair of TEF's results in postoperative aspiration pneumonitis. Contraction of the LM opens the sphincter in swallowing, belching, gagging and emesis. Reflux is associated with HH, tertiary contractions and LER's, not because of these conditions per se, or the way they distort the anatomy of the lower esophagus, but because all four of them are attributable to the same cause - esophageal shortening by LMC.

In the rare disease, myotonia dystrophica, in which the LM cannot relax, the sphincter, although not itself defective, remains constantly open.

It is tension on the PEL, not LMC per se that causes reflux. Thus, hyperextension of the cervical spine or surgical shortening of the esophagus - both of which apply traction to the PEL - also cause reflux.

Whatever relieves esophageal tension alleviates reflux. Disabling the LM by surgical trauma, drugs or anesthesia will counteract reflux because these means decrease LM tension on the PEL. Surgical elongation of the esophagus and anteflexing the cervical spine are effective by the same mechanism.

With advancing age or forceful vomiting, rupture of the PEL may cure reflex spontaneously, but non-effacement of the sphincter may then produce dysphagia and an appearance easily mistaken for terminal esophagitis.

Sphincter opening depends on a balance of forces: the LM tension, the integrity of the PEL, basal LES tone and whether the Cannon-Dougherty reflex is activated or not. Even the 2.2 cm of slack created by anterior flexion of the cervical spine may ameliorate symptoms of reflux, gas and nausea.

The cause of reflux is not intrinsic to the LES. Nor is it likely that an extrinsic substance inhibits it. The details of its pharmacology are most relevant in that they provide a clue to the action of the LM. A major implication of this analysis is that investigation of substances that stimulate the LM is likely to be more fruitful in eliciting a chemical cause of reflux. Compounds that inhibit the LM are most likely to be of therapeutic value.

LMC, invisible to Flatlanders, is the unseen event that causes "inappropriate" or "transient complete sphincter relaxation" in belching and reflux. This is why low LESP alone does not cause reflux. This is why stimulating the pharynx(99),(100) (provoking LMC via a gag reflex) causes reflux. This is why the unguarded moment(101) or failed peristalsis (LMC without a p-wave) causes reflux; why hiccups (the vector equivalent of LMC) and tertiary contractions (LMC + CMC) cause reflux.

Normal swallowing (p-wave + LMC) would also cause reflux, were it not that the advancing p-wave is a temporary sphincter that prevents backflow while it is in motion and then merges the LES.

Pull-down type operations have a better rationale than procedures that attempt to reconstruct the angle of His or create a subdiaphragmatic esophageal segment - features not normally encountered in the living subject. These procedures apply countertraction to the force of LMC and by relieving tension on the PEL prevent a sphincter-opening vector resolution. Increasing esophageal length, severing or elongating the PEL or changing the direction of vector resolution can all be expected to have a beneficial effect.

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Complications of LM tension: the Plummer-Vinson-Paterson-Brown-Kjelberg Syndrome (PVS)

The etiology of the PV syndrome is unclear. Does the anaemia cause the dysphagia? Does it cause the post-cricoid web? Is the dysphagia hysterical? Does the web cause the dysphagia? Is there an ectodermal defect causing the nail changes and loss of teeth. Are the labial fissures due to staph, monilia or drooling?

Answer: None of the above.

I will discuss the complications of hiatal transtraction and gastroesophageal reflux concurrently with Plummer-Vinson syndrome (PVS) because they are identical. The various features of PVS are merely selections from the extensive menu of pathologic events that are directly or indirectly related to excessive LM tone and/or the reflux it produces.

At present, PVS is widely regarded as due to an iron deficiency,(1) perhaps complicated by a vitamin B complex deficiency.(2) The several findings are, like the anaemia, attributed to iron deficiency or epithelial dysplasia.

PVS is rarely seen as a full-fledged syndrome. Forme fruste occurrences are frequent. Virtually every manifestation of PVS is also seen as an isolated problem. The postcricoid web, once considered pathognomonic of PVS was shown epidemiologically to occur in only 15% of females with dysphagia.(3) The more fully expressed the syndrome, the more orad the location of the manifestations. The classical components of the syndrome include:

Hiatus hernia

Hypochromic microcytic anemia

Sideropenia

Splenomegaly

Gastritis

Achlorhydria

Gastro-esophageal reflux

Esophagitis

Post-cricoid web

Positive vallecular sign

Loss of teeth at an early age

Glossitis

Cheilitis (rhagades, angular stomatitis, perleche)

Koilonychia

Esophagitis

The association of esophagitis with reflux is long established. The reflux occurs as a result, not of low sphincter tone as was initially supposed, but of transient but complete loss of lower esophageal sphincter pressure.(4) Although reflux occurs in normal subjects,(5) there are protective mechanisms that can prevent esophagitis in the face of reflux. These include removal of the bulk of the refluxed material by effective peristalsis (usually primary and provoked by swallowing) and dilution and neutralization of the remaining acid by the more alkaline (pH 6.5 to 7.6) saliva.(6),(7) Any impairment of salivation (e.g., by anticholinergic drugs, Sjörgren's syndrome) or peristalsis (e.g., by esophagitis) favors development of esophagitis by increasing the ACT (acid clearance time) normally 313 21 sec.(8)

A source of corrosive material is also required. This is generally the acid (.1N HCl) of the stomach although bile acids are reputed to be equally or more effective. Alleviating acid hypersecretion with H- antagonists tips the balance between aggravating and relieving factors resulting in subjective and objective improvement.(9)

Diagnosis of esophagitis

Histologically, reflux esophagitis is marked by a.) increased thickness of the basal cell zone and b.) proximitiy of dermal papillae to the epithelial surface. Multiple biopsies correlate poorly with the endoscopic diagnosis. Random biopsies are only 75% positive depending somewhat on the level.

Esophagitis patients display characteristic but non-specific findings on esophageal manometry. The amplitude of contraction is decreased, the transmission rate is delayed a few seconds and and the duration of the contraction is shortened.(10) In severe cases, the peristaltic wave may fail altogether.

The criteria for the diagnosis of esophagitis vary so much among authors, that statistics as to the incidence of esophagitis with PVS, or HH for that matter, are scarcely worth quoting at length. If the diagnosis is made with an esophagoscope, the reported incidence tends to be higher than that reported

radiologically. The reason for this disparity is the reluctance of radiologists to make the diagnosis unless the disease is very severe.

Radiologic diagnosis of esophagitis is not difficult. One need only have an appreciation of the normal size of the longitudinal folds - 1 mm or less - or, simpler still, recognize that the number of folds should be about 5-6. If 3 folds occupy the width of the contracted lumen, the mucosa is abnormal. In severe cases only a single fold may be seen in a given projection.

Radiologists reluctance may stem from an uncertainty about the significance of enlarged folds that is a consequence of the autoplastic theory of fold formation. As shown in earlier, however, fold formation is a function of the circular muscle, not the muscularis mucosae. When we rely on the number of folds for the detection of inflammatory disease, diagnosis becomes less subjective. In the esophagus particularly, the diagnosis becomes very easy: the patient with well marked esophagitis will have only 2 or 3 distinct folds instead of the normal 4-6. A convenient grading system is 5-N = grade, where N = the number of folds. Ulcerations, stricture, wall thickening and other gross changes are not necessary to make the diagnosis.

In the higher grades of esophagitis the primary peristaltic wave, instead of coursing the entire length of the esophagus, dies out at the striated-smooth muscle junction. Secondary p-waves may then partially empty the organ but are not the clean-wiping waves which leave the esophagus empty or outlined only by thin stripes of barium between the longitudinal folds.(11)

Esophageal folds are best seen with the patient supine. In this position the esophagus bows, bowl-like, downward and will retain barium better than when it forms a "hill" in the usual prone position. The same is true of esophageal varices.

There are well known difficulties in pathologic diagnosis of esophagitis. The material obtained at suction biopsy with the flexible scope contains only the lamina propria. Suction biopsies contain the full mucosa but little of the submucosa.(12) In this layer, there is no evidence of proliferative change. An increase in the thickness of the rete layer and of the length of the mucosal "pegs" characterizes esophagitis histologically. Such changes are normal in the distal esophagus that is exposed to "normal" reflux levels. The presence or absence of edema in the submucosa is never described pathologically, nor is it described in the lamina propria for that matter. Such constraints severely limit the possibilities for accurate pathologic diagnosis from biopsies.

The endoscopist is similarly limited because he can only see the surface of things. The mucosal folds are obliterated in the gas-distended organ. There is no means of judging thickness from surface appearance. Erythema, of course, is a sign that one anticipates with inflammation, but edema, which is what the radiologist sees decreasing the fold number, should cause a pale mucosa. It is

not generally appreciated that a barium coating magnifies depth enormously. A crevice a few thousandths of a centimeter deep is easily seen when barium filled because of the great density of the medium . Radiographically, these crevasses define folds.

It has been shown that mucosal permeability is increased in esophagitis. Any resulting edema must involve a layer neither pathologist nor endoscopist can visualize. This layer can only be the submucosa.

There is some distinction made in the literature between "superficial" and "deep" esophagitis. The basis for the distinction is that "deep" esophagitis produces thickening of the esophageal wall. This seems reasonable, as in some cases of reflux the wall of the esophagus does appear grossly thickened. However, using this criterion, I found that at surgery the diagnosis of inflammatory disease involving the muscular wall of the organ was seldom verified.

There are two reasons for this, both geometrical. The esophagus shortens in conditions leading to reflux - as much as a third its length without rupture of the PEL and that much or more after rupture.Contraction converts a long narrow cylinder of esophagus to a shorter, thicker cylinder. The percent of thickening can be roughly calculated from the formula for the volume of a cylinder as this remains constant before and after contraction:

$\pi r_1^2 l_1 = \pi r_2^2 l_2$	Inserting average values for the radius and length of the resting esophagus, it works out that an 8 cm shortening will increase the wall thickness 25% giving a misleading appearance of deep inflamation.

Thickening of the mucosa and submucosa in an organ with a radius as small as that of the esophagus also will increase the apparent diameter of the organ. Using the formulae developed in the section on mucosal folds, a calculation shows that a doubling of the mucosal thickness will double radius of the organ. Both effects act to increase the apparent wall thickness, although the muscle itself is quite normal. One must be careful, therefore, not to mistake these geometrical effects for inflammatory infiltration of the wall or muscular hypertrophy of the wall.

Postcricoid webs

These were evidently first described by Kelly.(13) Walderstrom and Kjellberg(14) documented their association with "sideropenia." Brombart considered the finding of such webs virtually pathognomonic of PVS,(15) however, the more recent literature contains reports of postcricoid webs without other manifestations of the full-blown syndrome. I have seen many postcricoid webs in non-anemic patients ranging from a mere nick of the anterior outline of the esophageal lumen to typical, fairly deep shelves. Like LERs they may be multiple.(16) Also like LERs,

my own cases have been associated with a HH in nearly every instance. Seaman,(17) however, in a retrospective study found only a 6% incidence of HH. Only 5 of his 53 patients with postcricoid webs were classified as PVS and only 4 had gastrointestinal bleeding.

Nosher, et al.(18) reported a 5.5% incidence of webs in 1000 consecutive cineradiographies of the throat. Of these 55 patients, dysphagia was present in only 6 and none of the patients were iron deficient. The association of webs with dysphagia is difficult to evaluate because "dysphagia" is frequently not defined in reports. Certainly webs can occur without either true dysphagia or "lump in the throat" sensation. 17 of 32 webs seen by Chisholm et al. were asymptomatic. Yet there is no doubt that a prominent web can cause true dysphagia and that dysphagia will sometimes be relieved by dilatation or rupture of the web.

The webs can hardly be due to anaemia per se as they also occur in hypothyroidism and in other patients without anaemia. Yet they do occur with increased frequency over the normal population in pernicious anaemia and with postgastrectomy anaemia.

Postcricoid webs resemble lower esophageal rings histologically. Curiously, the reluctance to recognize them as mucosal folds which characterizes authors who have studied LER's is not manifested with upper esophageal webs or rings. Entwhistle and Jacobs,(19) in 49 postcricoid web specimens from 39 patients found that "The [histological] appearance is essentially that of a fold of normal esophageal epithelium with some underlying loose connective tissue." In 6 of their cases there was no evidence of inflammation in the subepithelial tissue. A further 8 showed only a few chronic inflammatory cells. Seven showed plasma cell and lymphocyte infiltration. Half of the 14 cases biopsied by Chisholm et al.(20) were uncomplicated folds. The others showed similar minimal inflammatory changes. These reports tend to show that inflammation, while it may occur secondarily, has no part in the genesis of the web although, as with the LER, inflammation continues to be listed as a possible cause.

Biopsies of webs do not normally include the muscle layers of the pharynx. However, in the 3 specimens obtained at necropsy by Entwhistle and Jacobs, " . . . the main longitudinal muscle, which is normally thin anteriorly, showed degenerative changes most marked in the region of the web." Among these were atrophy and replacement collagenosis.

Although these were the only 3 cases in which the state of the LM could be determined, it is startling to learn that it was abnormal in all 3 in precisely the area of the web. It seems almost too pat a confirmation of what fold theory would predict. That is, if as a result of atrophy or necrosis the LM retracts anteriorly, it could well throw up a fold. Quantitatively, however, this explanation scarcely passes muster. Some of the webs are a centimeter in depth. It would require over 2 cm of shortening to produce that much redundancy.

There is an association between postcricoid webs and HH. One case was reported incidentally(21) and my impression is that they are almost always associated, at least when the HH is over 4 cm. Smiley and associates(22) found 19 of their series with various hypopharyngeal "obstructions" (webs, strictures and carcinoma) also had HH. Two had LERs as well.

The analogy of webs with the LERs is so good that there is a strong probability that it in some fashion they have a similar cause. A web requires a source of redundant mucosa and a reason for fixation. The following case, exceptional in that there was no HH associated with the web, suggested that the etiology may be similar to that of the LER.

SW CN 40987 11/12/65 Female, age 59. Fluoroscopic note: In view of the clinical history of iron deficiency anaemia, cine films were made of several barium swallows. These show a typical postcricoid web. The body of the esophagus was also of interest as it appeared short constantly keeping the fundus of the stomach under tension so that it formed a conical tent at and above the diaphragm. Because the esophagus was under constant tension, there was free reflux spontaneously on inspiration, during quiet respiration and with the de Carvalho test. The sphincter stayed open for several seconds at a time.

The stomach filled to show a high subtotal resection with gastrojejunostomy. There was very little evidence of a mucosal fold pattern in the stump.

On inquiry, the patient reported she also has a sore tongue and fissures at the corners of her mouth both of which she herself had concluded were due to acid reflux. She had lost her teeth at the age of 30 because of "dental caries."

The patient was reexamined a week later after 7 days iron therapy. Repeat cine films showed that the postcricoid web had completely cleared. The esophagus was no longer under tension and some redundancy had reappeared. The stomach was no longer tented into the hiatus and the fundus had lost its conical appearance. Reflux could only be elicited with the de Carvalho test.

The rapid response to Fe suggested that either low serum Fe or anaemia may increase LM tone. If so this would result in a vicious circle.The disappearance of the web when LM tension was relieved would tend to show that webs are also due to mucosal redundancy. Why isn't the mucosal redundancy milked to the distal end of the esophagus to form a LER? It is obvious that there is a need for elasticity of the mucosa to contend with the 2 cm or more upward excursion of the mouth of the esophagus with every swallow. If that elasticity disappears with esophagitis or atrophy, an accordion-pleat can partially substitute. Inflammation could also fix the mucosa to the muscularis layers.

At any rate, postcricoid webs are not pathognomonic of PVS and correlate poorly with the other features of the syndrome. There is some evidence that, like LERs,

they represent mucosal redundancy occasioned by LMC, thus their association with HHs.

Hysterical dysphagia

Vinson(23) named the syndrome "hysterical dysphagia" and, even today, this manifestation is almost the sine qua non in making the diagnosis. As we have seen, those who have given esophageal diseases names with etiological connotations get tagged with their mistake if they failed to assign the correct cause. It is not surprising then to find that, in the usual sense of the words, "hysterical dysphagia" is not dysphagia and that there is ample evidence the symptom has an organic basis.

The "dysphagia" of PVS is sharply distinguished from true dysphagia. In the latter, ingested food does not go down. It piles up producing substernal discomfort. It forces the patient to stop eating. Unchanged food is regurgitated. It may be painful or associated with weight loss.

In PVS the complaint is of "a lump in the throat." That is, the patient has the sensation of something lodged in the throat that cannot be dislodged either by repeated swallows, by washing it down or by regurgitating. Generally the patient will indicate the location of the "lump" by pointing to a definite area at the level of the thyroid cartilage. The sensation may be lateralized in patients routinely sleeping on only one side. Occasionally he/she may report choking, burning or coughing. Unless the patient also has a lower esophageal ring (a common enough associated finding) or another complication, there is no real obstruction to the passage of food.

Schatzki(24) believed the symptom was due to aerophagia because of the repeated dry swallows it occasions but these are a result, not a cause of the symtom. Malcomson,(25) a laryngologist, tabulated the findings in 231 patients with this complaint. Only 20% were entirely negative radiologically. Hiatus hernias (77) were the most frequent positive finding, accounting for two thirds of the lesions that were not local to the neck. Webs (5) were rare. The symptom responded to " . . . medical treatment for hiatus hernia."

I found that if the patient gargles a spoonful of viscous zylocaine it produces local anesthesia of the of the hypopharynx and eliminates the lump in the throat sensation - clear enough proof that the symptom is not hysterical.

Although "lump in the throat" dysphagia is characteristic of PVS, it is not limited to that syndrome. Indeed, one will see many patients with this symptom before encountering a full-blown case of PVS. Hallewell et al.(26) found 22 patients with HH and reflux who exhibited a lump in the throat sensation and/or hoarseness. All found relief on antireflux therapy. Delahunty and Ardran(27) found that of 25 patients with the globus complaint, 22 were suffering from reflux esophagitis. The

globus symptom cleared on an anti-acid regimen. They ascribed the symptom to a motility disturbance (aperistalsis and non-peristaltic contractions) that they were able to provoke after ingestion of "acid barium" (pH 1.7). They also regard the motility disturbance as proof of reflux. There were cine-radiologically demonstrated HHs in 13 patients and reflux in 10.

This work convincingly ties globus to reflux, but it does not necessarily follow that the proximate cause of the symptom is the motility disturbance. Far more profound disturbances of motility in "diffuse esophageal spasm" may be asymptomatic.

The most striking and unequivocal cause of the globus symptom is enlargement of the lingual tonsil. This midline structure forms the base of Waldeyer's ring. It is embedded in the base of the tongue directly anterior to the tip of the epiglottis. The tip of the epiglottis is centered on the tonsil and, in the position of rest, separated from it by an air space of several millimeters. Because of the air space, the two are never in contact except during swallowing. However, when the tonsil is enlarged, the two structures are in constant contact, producing the sensation that "There is something there." that shouldn't be. In some cases of prolonged contact, the tip of the epiglottis may create an umbilication of the tonsil at the point of contact. In other cases more diffuse swelling at the base of the tongue completely obliterates any air space so that the epiglottis is plastered against the tongue base. Hypopharyngeal edema, particularly of the epiglottis and lingual tonsil would appear to be a more direct explanation of the globus symptom. This explains the association of the globus symptom with reflux.

One can actually predict the symptom from the film appearance. Occasionally, on noting an enlarged lingual tonsil on radiographs made primarily for the cervical spine, I have gone to the waiting room and verified that the patient did have a chronic lump in the throat sensation. This finding, easily demonstrated on lateral films of the soft tissues of the neck, may occur in the absence of reflux because of a viral or bacterial tonsillitis.

"Hysterical dysphagia," therefore, is neither hysterical nor dysphagia. It is hypopharyngeal consciousness due to the irritating effect of GER.

The Vallecular Sign

On radiologic examination, the valleculae usually clear of barium so cleanly and rapidly that it may be difficult to obtain a satisfactory spot film. When severe, hypopharyngeal inflammation is manifested radiologically by a positive "vallecular sign"(28) -- pooling and delayed clearing of barium from the valleculae and pyriform sinuses.

The sign is not specific. It is also seen in myasthenia gravis, senility, post nasal drip syndrome, central or peripheral involvement of the 9th and 10th cranial

nerves, nonspecific inflammation or any condition that even slightly impairs the hypopharyngeal swallowing mechanism.

It is frequently caused by a characteristic hypopharyngitis encountered in patients with severe reflux. Otolaryngologists, once they have been made aware of this cause, become quite proficient at predicting that reflux will be demonstrated on radiologic examination. They report a "dusky reddening " of the hypopharyngeal mucus membrane. This appearance may be passed over as normal by the laryngologist or as an insignificant URI. Thus Cherry et al.(29) found GER radiologically in 12 patients with unspecified "pharyngeal symptoms" who were reported normal on pharyngeal and otolaryngological examination. Symptoms were reproduced by perfusion of the esophagus with .1N HCl and cleared on an antiacid regimen.

Delahunty(30) was able to demonstrate that posterior laryngitis was caused by acid reflux, presenting 9 patients with typical symptoms of chronic laryngitis (variable hoarseness) and laryngoscopic findings of " . . . interarytenoid heaping of mucosa with chronic inflammation of the posterior third of the true cords." Five also had the globus symptom. In most cases, however, actual reflux was demonstrated. Significantly, symptoms were relieved and the local lesion healed on antireflux therapy.

In a much larger series(31) Larrain et al. found that 74 of 78 patients with "intrinsic" asthma showed posterior laryngeal white plaques of varying prominence. Nearly all had pH probe-proven reflux although most either had no symptoms of reflux or only admitted to reflux on close questioning.

Thus, a GER based hypopharyngitis should be added to the list of conditions causing a positive vallecular sign. Radiographic diagnosis from the subtile differences in outline of hypopharyngeal structures is difficult except for the laryngeal ventricle which loses its sharp "fish-mouth" appearance or vanishes when edematous..

Pulmonary symptoms of reflux

Although not included in the classical symptom list of PVS, respiratory complications are very frequent associated findings in GER. A history of high reflux is reliable and far more specific for reflux than the vallecular sign. However, it is necessary to make a specific inquiry as it is seldom volunteered. Much of the reflux occurs at night when the patient is sleeping. Virtually pathognomonic of reflux is a history of nocturnal laryngospasm. The patient wakes coughing, choking and unable to get a breath. This signals high reflux with spill into the larynx. It is a very common symptom that I have often suspected may account for some cases of bronchiectasis. Most patients are not even aware of the reflux because it the symptoms of laryngospasm overshadow it.

"Do you ever wake at night coughing, choking or gasping for breath?," is also a good question to include in the routine history of patients with "hysterical" LIT syndromes. It is astonishing that many patients with a lifelong reflux problem never complain of heartburn, the symptom we, as physicians, associate with reflux. They have come to believe it is a normal state of affairs! "Water brash" - the sensation of acid rising up into the throat(32) - occurs even during normal waking hours and correlates well with all these radiologic findings.

These nocturnal episodes of GER-caused laryngospasm may be misdiagnosed for years as bronchial asthma(33),(34) and the pulmonary fibrosis caused by aspiration of acid pepsin is thought to be "idiopathic."(35) Patients with intrinsic asthma have a high incidence of pH probe-proven reflux - 89% in one series(36) of 142 patients - and show objective and subjective response of their pulmonary symptoms after reflux treatment.(37)

Tuchman, Boyle, et al.(38) showed experimentally in cats that introduction of microvolumes (.05 ml) of .1N HCl into the trachea increased lung resistance 4-fold by receptor mediated reflex bronchospasm. This suggests that actual parenchymal aspiration is not necessary for GER to alter pulmonary function.

Infants under 6 months with reflux related symptoms (apnea, choking, recurrent pneumonia, chronic cough, wheezing) had a mean duration of reflux episodes of > 6 minutes during sleep on pH monitoring.(39) Reflex bronchospasm is a possible cause of the sudden infant death syndrome.

Cheilosis

Although they are not quite synonymous, the terms cheilosis, rhagades, angular cheilitis, perleche and lateral stomatitis are used almost interchangeably for characteristic fissure-like erosions at the corners of the mouth.(40) They may be crusted or denuded when fresh, fading to slightly bluish discoloration when healed. They have long been considered a manifestation of vitamin deficiency, particularly of riboflavin(41) or pantothenic acid. Paterson(42) [of Paterson-Kelly] appears to have been the first to add fissures at the corners of the mouth to the syndrome. Their presence in PVS has suggested that the rest of the syndrome might also be a vitamin deficiency. Goldstein(43) states that the injection " . . . of liver extract and the administration of vitamins have appeared to affect favorably some of the epithelial changes, especially the cheilosis." A review of 156 cases by two oral surgeons(44) concludes that a reduced vertical dimension of the lower half of the face was an etiologic factor in 34% of the cases. Moniliasis is frequently incriminated although this is a secondary infection.

The following case, in which cheilosis occurred with several signs and symptoms due to high reflux, is one of many that suggested an explanation of the problem that turned out to be more easily proved.

LK. CN 41221. 11/6/68 Female, age 50. History of "lump in the throat" and heartburn. She has been seeing a dermatologist because of fissures at the corners of her mouth. (One can still see erythematous and atrophic scars from these fissures, which have healed under treatment.)

A preliminary film of the cervical soft tissues shows a marked anterior curl of the epiglottis so that there is no clearance between its tip and the slightly enlarged lingual tonsil.

Fluoroscopic note: Ingested barium passed freely through the hypopharynx and esophagus. There were no signs of stricture or obstruction. There was pooling in the valleculae but not in the pyriform sinuses. There were no signs of a diverticulum.

When the patient was given a swallow or two of water in the supine position, gross cardioesophageal reflux occurred. Some of this reached the hypopharynx and was aspirated causing a typical episode of laryngospasm. In provoking reflux, a sliding HH was also provoked. This was about 5 cm in length. There was a slight LER when it was maximally provoked.

Multiple sequenced spot films show that when the HH is provoked, the slight ring is present and the sphincter is widely patent. When the hernia reduced, the ring disappears and the sphincter closes. They also show a slight mucosal crinkling in the post-cricoid area.

Patients such as this suggested that, as the canthus of the mouth is dependent while a patient is sleeping, nocturnal reflux could well cause acid burns. There is ample evidence that patients reflux in their sleep. Drooling of acid pepsin from the corner of the mouth seemed a reasonable etiologic speculation.

It remained a speculation for some time, but, to my routine history, I did add the question, "Do you ever get cracks or sores at the corners of your mouth that take a long time to heal?" There were enough affirmative answers to demonstrate convincingly that cheilosis was not unique to the PVS.Cheilosis was a common finding in the GE reflux population.

On receiving an affirmative reply to the above question, I routinely turned up the lights to inspect the patient's mouth. The lesions were always bilateral and, even if not ulcerated or crusted, they were often visible long after healing as symmetrical, faintly bluish scars.

After a year or so, I encountered an exception to the rule of bilaterality. The patient I was examining had involvement only on the left side of her mouth. She was standing in the "slot" of the radiographic table, ready to be given the first swallow of barium. I stood up to look more closely at the lesion. Glancing down, I

discovered to my delight that she had a plaster cast enveloping her right arm and shoulder!

Other patients unable to sleep on one side or another because of casts, bursitis, habit, etc. also proved to have lesions only on the contralateral side. As a unilateral avitaminosis is out of the question, the explanation fits.

Although the evidence is less dramatic, on questioning, most patients with cheilosis report finding a wet spot near their cheek on their pillow on waking - the result of nocturnal drooling of refluxed gastric acid pepsin. Both patients and their physicians are inclined to attribute the wet spot and cheilitis to nocturnal drooling of saliva. However, salivation ceases during sleep(45) as does the output of the mucus glands of the head. If further proof is required, the patient, or in the case of children, a parent can be given an indicator solution to test the wet spot's acidity.

This seems to dispose of avitaminosis as a cause of cheilosis per se, but does it rule out an avitaminosis causing LM tension, causing reflux, causing cheilosis? Only to this extent: It presently seems a redundant hypothesis. There are a great many patients with reflux (as the WSJ article suggests) - far more than any reasonable estimate of the number of clinical cases of nutritional deficiency.

Loss of teeth at an early age

I had always assumed that the premature loss of teeth described in PVS was due to poor oral hygiene, lack of dental care or neglect, but the following history, elicited from a young nurse with a severe, full-blown PVS was revealing:

"My parents spent a fortune on my teeth. I would have a lot of fillings and then a few months later there would be another crop of cavities. Finally, the dentist told them there was nothing he could do, that I just had "soft teeth" and when I was 12 years old they were all extracted."

Is premature loss of teeth also due to acid reflux? The patterns of destruction, age of onset and association with other signs and symptoms of reflux answer this question affirmatively. The ability of acid to dissolve calcium compounds, the known destruction of the teeth in situations with obvious exposure of the teeth to gastric content - bulimia, cancer chemotherapy, anorexia nervosa - point to the same conclusion. Even conditions attributed to other causes ("nursing bottle caries") make a telling contribution to the argument.

For the past 25 years, I have routinely queried edentulous patients having upper GI examinations as to their age when their teeth were extracted. Responses leave no doubt that their stories resemble that of my young nurse. Remarkably, most of them have gross reflux even though their teeth were lost decades earlier.

White(46) has documented the loss of tooth structure associated with chronic regurgitation and vomiting. Katherine Byrne,(47) a professional medical writer, observed in her own daughter that the enamel of the upper incisors was first to be involved and caused these teeth to become sensitive to hot and cold. When damaged teeth were crowned, the crowns also became eroded.

The following account, by my dental hygienist, of the teeth of a patient with bulimia is a graphic description of the "soft teeth" syndrome and destruction wrought by recurrent regurgitation of acid gastric contents.

The enamel was porous - easily indented with a pick - and the teeth seemed to be shells. The buccal surfaces had been jacketed by her dentist, the lingual surfaces were eroded.(48)

Crib caries

"Crib caries"(49) are discussed here because the mechanism of the disorder is essentially the same as that which causes loss of the teeth in PVS. Crib caries is a rampant form of dental caries in infants usually attributed to the custom of putting them to bed with a nursing bottle. By the age of 2 or 3, all of the teeth may have been lost except for the lower anterior teeth. Destruction generally begins with the upper incisors and spares the lower teeth. Although almost any tooth may be involved, the lingual sides are more extensively involved.(50) It is believed that alteration of the bacterial flora by sugar in the feeding bottle promotes caries formation. The front teeth may be spared because of their contact with the tongue and because the high pH of the submaxillary gland saliva protects them. It has also been theorized that the swallowing pattern of infants somehow protects the front teeth because in infancy the tongue is thrust forward with sucking and swallowing.

However, the same condition has been reported in infants who were breast fed.(51) In an epidemiological study, Richardson and Cleaton-Jones (52) rejected the "nursing bottle hypothesis" as they found that the incidence rises with age, " . . . being far more common at five than at two, that is long after the age of weaning . . . " Moreover, comparing the infant feeding patterns of blacks and whites in South Africa, they found equal numbers of labial caries among black children who did not receive fruit syrups as in white children who did. This seems to rule out alteration of bacterial flora by excess sugar as cause of the caries unless one makes unprovable, theory-saving assumptions (ectodermal defect, intrinsic susceptibility, weakening of enamel by childhood diseases, etc.

Weyers(53) classified 50 children between the ages of 2 and 6 according to whether they had received "sugar infusions" for prolonged periods. The statistics showed a strong inverse relationship. If anything, drinking sugar-containing liquids from nursing bottles protected against crib caries.(54)

I had asked the hospital dentist(55) to alert me if he encountered an example of acid destruction of teeth. A short time later he found such changes in a patient of 35 with a history of heartburn for years.

On examination, the same pattern of tooth destruction presented as that described for crib caries! The front teeth were relatively spared, but the lingual surfaces of the distal teeth were beveled down to the gingival margin. It was as though they had been ground down obliquely with a peripheral rim of opaque enamel and a center of semi-translucent dentine.

A striking proof of the reflux etiology of the dental abnormality was a severe cheilosis on the left side only. The patient said that the cheilosis occurred "every couple years" and was always on the left side - the side on which he habitually slept when not sleeping on his stomach. The oral mucosa was discolored (xerostomia).

On fluoroscopy, "The esophageal mucosal folds were greatly thickened, 2 of them occupying the entire width of the esophagus. A 5 cm sliding hiatus hernia was demonstrated with the Valsalva maneuver and gross GE reflux occurred in response to the de Carvalho maneuver. There was no postcricoid web."

A similar case was described in a 14 year old boy by Abdulla et al.(56) who had " . . . large numbers of chalky enamel lesions . . . of the facial and lingual surfaces and some encircled the teeth." All of the molars had crowns and some of the restorations had secondary caries. Xerostomia was also present with saliva production reduced to 2 ml/hour (vs. a normal of 60 ml).

The boy had dysphagia since the age of 4. An esophageal stricture was dilated at the age of 8. When seen in the dental clinic he had an angular cheilitis (ascribed to B vitamin deficiency by the authors) completing the picture of chronic reflux.(57)

Noting that patients with esophageal strictures were frequently edentulous, Maxton et al.,(58) computed a chi-squared table for edentulism vs. stricture for a group of 1759 patients undergoing endoscopy at St. Thomas' Hospital and found a $p < .01$ that the association was due to chance. Among a variety of explanations offered for the association (poor nutrition from edentulism, lack of saliva causing both stricture and caries, avoidance of solid, esophagus dilating boluses of food) they did not include the possibility that acid reflux caused both the stricture and the loss of teeth.

These examples, however, leave no doubt that the loss of teeth in PVS and crib caries is not due to some ectodermal defect, vitamin deficiency or change in eating habits but is instead due to the lytic action of hydrochloric on teeth. The pattern of destruction is exactly what would be expected with acid reflux. The

lower anterior teeth are protected by submaxillary saliva (pH 6.5) and the buccal surfaces of the distal teeth by parotid saliva.

We can use Occam's razor to exclude the superfluous speculations as to the cause of crib caries. The pattern is that of acid destruction. The infants may perform a self-administered de Carvalho maneuver by drinking - whether it be fruit juice or mother's milk - while lying on their backs thus insuring acid reflux by turning off the CD receptor.

Destruction of teeth is yet another manifestation of GER that has masqueraded under a variety of misdiagnoses. The appearance of cheilitis in an infant should be a cause for alarm and investigation for the presence of reflux and its sequelae of which the next symptom may be sudden infant death. A finding of many caps and crowns on the teeth of patients having films of the face, skull and sinuses should alert the radiologist to the possibility of acid reflux. Oncology patients should be warned to flush their mouths with an alkaline or buffer solution after vomiting.

Sideropenic anemia

The term "sideropenic anemia" was applied by Kjelberg who also documented the frequency of post-cricoid webs in PVS. It is a somewhat vague term that may be understood in several senses. "Sideropenia" - i.e., low serum iron - can have several causes: 1.) Decreased uptake because of a) dietary deficiency or b) impaired absorption. 2.) Increased utilization of iron. 3.) Increased iron loss. Of these, "sideropenic" evokes the sense of dietary deficiency.

The prompt response to orally administered iron in the overwhelming majority of cases would show that there is no defect in absorption from the gut. Percent absorption of iron may actually be increased 300% in cases of chronic blood loss. Malabsorption of other nutrients was never mentioned in the 183 reported cases I reviewed.

There are local areas where the syndrome is endemic. This has suggested a lack of iron in the soil. However, agricultural production is now international in scope and, except in areas practicing subsistence agriculture, it is hardly likely that a nutritional deficiency of iron could explain the incidence. The adult iron requirement is only 1-1.5 mg/day.(59) There is no evidence of iron deposition in the tissues in cases of PVS or other indication of increased utilization.

Chronic gastrointestinal blood loss, however, depletes the serum iron level by excretion of iron in the lost blood. Additional iron is then required for hemaglobin production. The serum iron will be reduced to low values even before the anemia is apparent. Consequently, a serum iron determination (or better yet ferritin) is advised as a definitive test for confirmation of PVS.(60)

Chronic blood loss also explains the PVS sex ratio as menstrual blood loss (as much as 200-500 cc per month) is additive. Superimposed on blood loss from a HH, this is a probable cause of a severe iron loss and anaemia. In the definitive study of blood loss in HH patients by Holt and coworkers,(61) iron absorption and blood loss were determined with isotope methods and whole body counting. HH patients with anemia lost an average 15 cc of blood per day, the non anemic patients only 3 ml/day. There was no deficiency in the uptake of iron. The anemic patientsabsorbed 39% of administered Fe vs. 8% in the non anemic group.

The problem in sideropenic anemia, therefore, is not nutritional iron deficiency, deficient uptake, inability to metabolize iron or iron sequestration, but chronic blood loss. This is what ties it to hiatal transtraction and to the LMC that causes it. HH per se, by causing chronic blood loss, can account for the anemia of PVS. The mechanism involved will be discussed in detail in the chapter dealing with achalasia. Here it need only be said that there is an increased friability of the mucosa in the supradiaphragmatic portion of the fundus occasioned by impaired venous return. This is why lesions of the fundus are more prone to bleed.

In a series of 200 cases, Edmunds(62) found that up to 55% of patients with "paraesophageal" hernias were anemic. Almost the same percentage of patients with large HHs in the Mayo Clinic series (50% of 109 cases) were anaemic.(63) On endoscopy, a third of such patients have linear erosions on the surface of the rugae at the level of the diaphragm producing a striped appearance on endoscopy which has been called "watermelon stomach." Cameron and Higgins concluded that mechanical trauma to the folds sliding through the hiatus and eroding each other was the proximate cause. Morrissey (64)concurred, suggesting that "The hiatus may be tight enough in some patients to cause intermittent venous stasis." and that mechanical trauma was a factor in the friable, erythematous appearance of the mucosa in sliding HH as well. Identical friability of the mucosa in the herniated portion of the stomach was described by Cohen.(65) Radiographically, fundic folds are visibly enlarged when constricted by a small hiatus in sliding HHs as well. This is a good piece of evidence that hiatal size can produce GE abnormalities.

Thus HH itself can produce a "sideropenic" anemia. Holt's group found that these patients improve on iron therapy, but, because of the chronic blood loss, they begin to go downhill when it is stopped. This creates the clinical impression of a refractory anaemia as the blood loss is undetectable with commonly employed guiac test.

Actually, there is no real defect in iron metabolism, no inability to absorb iron and no lack of a normal iron intake. The low serum iron, in nearly all cases, is due to chronic blood loss with consequent loss of iron and the typical hypochromic, microcytic anemia.

Splenomegaly

There is a 10% incidence of splenomegaly associated with sideropenic anemia. The enlargement is minimal, pathogenesis is said to be unknown(66) and there are no specific pathologic changes to suggest it is a disease sui generis. It seems likely that the reticuloendothelial system of the spleen merely exercises its normal function and filters abnormal microcytic RBCs from the bloodstream.

Hiatus hernia

To complete this chain of evidence, it remains to be shown that HH is a feature of the PVS. This has been done by Smiley, McDowell and Costello.(67) In their series, HHs were demonstrated in nineteen of 27 patients with "pharyngo-esophageal obstruction" (post-cricoid webs, rings, diaphragms) and other classical features of the syndrome. The Smiley group also suspected chronic blood loss from the HH as at least a supplementary factor in the anemia because a tendency toward relapse suggested a continuing blood loss. However, they did not appreciate the role of reflux in causing the buccal, lingual and pharyngeal lesions, attributing them to " . . . faulty replacement of the foregut epithelium caused by iron-deficiency anemia." That is, they believed it likely that an iron deficiency anemia per se caused lingual mucosal atrophy, etc.. For this reason, and because most HH patients do not have webs, they felt the argument for chronic blood loss from HH as a cause of iron deficiency was flawed. The work of the Holt group, however, clarifies this issue.

Achlorhydria

Now I must deal with the extraordinary paradox of PVS that has probably long obscured its cause. Although I have shown that multiple symptoms and signs of the syndrome are due to LMC causing reflux of acid pepsin, HH, etc., one characteristic finding remains to be explained - achlorhydria! On the face of it, this appears to destroy the entire unifying hypothesis.

Of course, one could shore it up by citing the evidence that bile acids and other contents of the duodenum are just as corrosive as acid pepsin, but this would mean that we would have to postulate that there was pyloric incompetence or reverse peristalsis in the duodenum. There seems no reason to make that assumption.

In actuality, no further assumptions are required. Achlorhydria is one of the consequences of the anemia. The mechanism is as follows:

1.) LMC causes reflux and hiatal transtraction

2.) One or both of these result in chronic blood loss.

3.) The resulting anemia per se causes a superficial gastritis that, if untreated, progresses to atrophy of the gastric mucosa.

4.) Atrophy of the gastric mucosa causes achlorhydria.

The atrophic mucosa and achlorhydria of pernicious anemia are classical. Perhaps less well known are the changes that are seen in "iron deficiency" anemia. Davidson and Markson(68) studied 42 patients and 39 age-matched controls with gastric biopsy. They found that the gastric mucosa was abnormal in three quarters of the patients with iron-deficiency anemia. When histamine-fast achlorhydria was also present, the mucosa was abnormal in 95% of cases. The abnormalities ranged from chronic superficial gastritis to atrophic gastritis to gastric atrophy. In most cases the lesion was an atrophic gastritis.

The incidence of achlorhydria was 48% in the anemia group vs. 13% in the controls (who were other hospital patients without anemia). In 5 patients in whom the hemoglobin level returned to normal after iron treatment, free Hcl reappeared in the gastric secretions of the two with superficial gastritis but not in the 2 with atrophic gastritis. The 5th patient had free HCl prior to treatment despite a severe anemia and superficial gastritis.

Leonard(69) found that of 47 cases of hypochromic anemia in military inductees, 13 had achlorhydria of which 6 were reversible on iron therapy. The frequency of achlorhydria was higher in the groups with the lower hemoglobin levels. In a series of 50 patients with hypochromic anemia, Badenoch et al.(70) also found that 43 (86%) had abnormal gastric mucosa. They noted that "There was a good correlation between the severity of the mucosal changes and the incidence of histamine-fast achlorhydria."

Two circumstances influenced Davidson and Markson(71) to resolve the question of which came first - gastritis or achlorhydria - in favor of the gastritis.

Their experience and the experience of others that treatment of the anemia restored free HCl.

In the milder superficial gastritis, 50% of the patients retained the ability to produce gastric HCl.

The alternative hypothesis is that a primary achlorhydria could lead to faulty absorption of iron and so cause an anemia. However, in PVS the blood loss is the cause of the anemia so there is no point in postulating an idiopathic achlorhydria as well.

Badenoch et al. also concluded the achlorhydria was secondary to the gastritis and that gastric mucosal changes, like koilonychia and "angular stomatitis" are a result rather than a cause of iron deficiency.

While concurring in this order of precedence, I would have to demur that, since atrophic gastric mucosa also occurs in pernicious anemia, it is more reasonable to conclude that the mucosa atrophies because of the anemia than because of lack of iron. As has been noted, the cheilosis is secondary to reflux.

Koilonychia

It is not quite as clear that the koilonychia is due to anaemia rather than low iron levels in the blood serum. Like the anemia, it clears when massive doses of iron are administered(72). If nail changes were seen in pernicious anaemia with normal iron levels, it would suggest that iron was not the cause of the altered nail growth. An the other hand, nail changes are common in conditions with low O2 saturation levels - cyanotic heart disease and chronic pulmonary disease - suggesting that anemia per se is the cause of the nail disorder.

SUMMARY

I have presented the evidence that the LM of the esophagus causes each of the features of PVS. It does this by producing both hiatal transtraction and reflux. The former leads to chronic blood loss, anemia, sideropenia, gastric atrophy and achlorhydria. Intractable reflux leads to esophagitis, hypopharyngitis, laryngitis, glossitis, dental destruction and cheilosis, i.e., chemical trauma to the esophagus, pharynx, larynx, tongue, teeth and skin. LERs and, possibly postcricoid webs, are other manifestations of the LMC that causes the reflux. "Hysterical dysphagia" is neither hysterical nor dysphagia but is due to chemical hypopharyngitis and/or lingual tonsillitis. The high incidence of cancer of the hypopharynx and esophagus is referable to the chemical insult of long duration.

Anemia, whether sideropenic or pernicious, leads to atrophy of the gastric mucosa and, eventually, to achlorhydria. Other pathologic entities attributable to reflux are identified. These include posterior laryngitis, a simulated "bronchial asthma" and pulmonary fibrosis with some cases of bronchiectasis suspect.

It is clear that some agent or agents can cause relentless hyperfunction of the LM of the esophagus leading, in this syndrome at least, to life-threatening consequences. The fundamental problem for esophageal research is to discover the nature of these agents and means of counteracting them.

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"Hiatus hernia" and rupture of the PE ligament

Like "achalasia," "hiatus hernia" is an example of a wrong name paralyzing thinking about a disorder. Because they are called hernias, "hiatal hernias" are lumped in with inguinal, femoral and ventral hernias. We tend to assume that our instructors gave us the correct names for things! Standard references(1)(2)(3)(4)(5)(6) do not even discuss their pathogenesis. It is simply taken for granted. After reviewing 636 references, Postlethwait(7) concludes they are due to increased intra-abdominal pressure in combination with weakness of the supporting structures.(8) Even a group(9) that reported experimental production of hiatus hernia by vagal stimulation concluded that most were due to increased intra-abdominal pressure. A recent review(10) lists 17 possible causes, except for increased intra-abdominal pressure, most of them nonspecific.

The central problem of "hiatus hernias" (HH), therefore, is to prove that they are not hernias. Instead, I must show that the condition is a traction phenomenon - that the fundus is drawn above the diaphragm by the tractive force of longitudinal muscle contraction (LMC).

Support for this position is many-sided:

The function and power of the longitudinal muscle are appropriate to the task.

Vagal stimulation of the longitudinal muscle will produce hiatal "herniation."

The morphology of the various types of HH is inconsistent with their supposed origin by pressure from below the diaphragm; it is exactly consistent with a traction mechanism.

The frequency distribution (90% sliders -- 10% others) is only consistent with a traction pathogenesis.

Extrinsic traction, such as that produced by cervical hyperextention (whiplash injuries, Sandifer's syndrome), also causes HHs.

The near 100% association of Zenker's diverticulum with HH is a further line of proof for a traction mechanism.

Because ideas are embedded in words, it is appropriate to start with a definition: A hernia is "a protrusion of an organ or part . . . through the wall of a cavity in which it is normally inclosed."(11) Further protrude is defined "L, protrudarei, to thrust forward, to cause to project or stick out." and, finally, "project, to throw out." Again, Dorland(12) attributes to Celsus the definition: "The protrusion of a loop or knuckle of an organ or tissue through an abnormal opening." [Emphasis added.]

The fundamental idea here is that the force that causes the "throwing" and "causing to project or stick out" is behind the thing thrown. The gunpowder is behind the projectile. The rocket thrust is from behind.

And so it is with most hernias. The force that causes the organ to "protrude or stick out" is behind the organ and inside the space from which it is protruding. This is an entirely correct concept whether we are speaking of an inguinal, a ventral or an umbilical hernia; whether we are describing a mediastinal hernia, an intercostal hernia, a herniation of the cerebellum or of the nucleus pulposus of an intervertebral disk. It may even be true of protrusions through the diaphragm at the foramina of Bochdalek and Morgagni. The same mechanism (increased intra-abdominal pressure) is assumed to be etiologic for "hiatus hernias," but is not. (I will call them "HHs" from now on to avoid the awkward but necessary quotes.)

This unfortunate choice of a name and our innate feeling for the meaning of words has virtually closed the door to an understanding of the cause, effects and treatment of HH. The semantic disability is difficult to cure because there does not exist, in English at any rate, a word meaning "External traction on an organ or part pulling it out of the cavity that normally contains it." Perhaps this is not surprising; there would be only two situations to which it could apply.(13)

HHs differ from abdominal hernias

HHs do not fit the definition of hernia and are not analogous with hernias.

Hernias occur through an abnormal weak spot in the wall of a body cavity. HHs occur through a preformed, normal opening.

In HH, the protruding organ is a continuation of an organ, the esophagus, in another body cavity. No true hernia is so constituted.

Unlike abdominal hernias, transients aside, the basic hydrostatic pressure differential across the wall of the containing body cavity that favors protrusion is lacking in HH.

No true hernia is ever drawn from its proper body cavity by traction from without. HHs are.

True hernias can be repaired by reinforcing or occluding the weak area in the wall of a body cavity. Without severing the esophagus, the esophageal hiatus cannot be closed.

Laboratory studies

As early as 1932, Von Bergman and Goldner(14) had suggested that HH might be due to traction due to esophageal shortening in response to vagal stimulation.

They quoted earlier experiments of Kuckuck showing that stimulation of the vagal trunk produced hiatal herniation in rabbits. Sir Arthur Hurst, of achalasia fame, subscribed to a similar hypothesis.(15)

In 1945 Dey, Gilbert, Trump, Roskelly and Rall(16)(17) experimentally produced HHs in dogs by stimulating the proximal end of the transected vagus nerve, by stimulating the intact vagus and by peritoneal or upper abdominal organ stimulation. Later (1967) Torrance(18) found an identical response in cats and may have been the first to associate both HH and reflux with LMC.

In 1969 Christensen and Lund(19) performed much the same experiment on the opossum (Didelphis virginiana) as this animal has the same distribution of striated and smooth muscle as is present in humans and, conveniently, has a 4-cm intra-abdominal esophageal segment. It enters the stomach just proximal to the pylorus instead of inserting in the fundus. They found that stimulation of the esophagus in vivo by distending an intraluminal balloon produced ". . . visible shortening of the intra abdominal segment with rostral sliding of the esophagus into the diaphragmatic hiatus."

Similarly, balloon distention of the isolated esophagus in a Krebs solution bath caused longitudinal muscle contraction both proximal and distal to the distending balloon. This contraction persisted as long as the balloon was distended.

Transducers connected to the distal esophagus of the cat, opossum and monkey by Dodds, et al.(20)(21) demonstrated that ". . . a forceful longitudinal tug is generated during esophageal peristalsis." These authors also suggested LMC as a possible factor in the genesis of HH. Daintree Johnson(22) (1966) produced hiatal transtraction in dogs by stimulating LMC with apomorphine.

As these studies have not made much of an impression or perhaps are regarded as tentative or as laboratory curiosities, I will consider at length and from every conceivable angle the etiology of this common disorder. I wish to show that LMC causes not just the occasional HH, but all of them.

A radiological misconception

The usual method of eliciting abdominal hernias is with the Valsalva maneuver - forced expiration against a closed glottis, but this also elicits HHs and, perhaps because of this, HHs are presumed to be etiologically identical with other hernias. Despite the superficial resemblance, however, there is a fundamental difference. A Valsalva maneuver elicits a sliding HH only when a bolus is being swallowed during the maneuver. The distention of a bolus causes enough LMC to erect the PEL tent, after which increased intrathoracic and intra-abdominal pressure occlude the lumen of the portion of the fundus in the tented PEL. Thereafter, swallowing must occur against resistance. The near-maximal LMC provokes the HH via the captive bolus effect.(23)

Thus, although straining against a closed glottis also produces HH, it does so not because it increases intra-abdominal pressure relative to the thorax, but because it provokes LMC. It is easy to see why an observer could have the impression that the gastric segment is being extruded upward. If this were the case, however, the esophagus would become redundant and either telescope into the fundus or be pushed aside by the extruding stomach (as does happen with non-sliding hernias). Instead, the esophagus is short and taut as a bowstring.

A gedanken experiment

To refute the increased intra-abdominal pressure pathogenesis, it is useful to perform a "gedanken experiment" such as those used by physicists in thinking about situations it would be difficult or impossible to set up practically.

As, hydrostatically, the abdomen behaves like a bag of water,(24) we start by imagining a muscular cylinder divided into two compartments by a flexible, diaphragm-like partition. The lower compartment is lined by a thin elastic membrane (peritoneum) and is filled with water. The upper compartment is filled with air at normal atmospheric pressure.

If holes or weak spots are then created in the cylinder wall, the elastic membrane, driven by the force of hydrostatic pressure, will bulge through the holes in typical hernia fashion. Those that are lowermost will bulge the most because there is a greater head of hydrostatic pressure extruding them.

Next, without perforating the lining membrane on its inferior surface, we make holes in the "diaphragm." How much will the membrane bulge through these holes? Not at all. There is zero hydrostatic pressure at the top of the fluid filled cavity. In fact, if the "diaphragm" were inflexible, the lining membrane would bulge downward, because the volume would remain constant and any extrusion below would be matched by intrusions of equal volume on top.

We conclude that, given the constant hydrostatic pressure relationships, ventral and inguinal hernias will occur simply from hydrostatic pressure on a locus minoris resistentia, but diaphragmatic hernias would never occur. Indeed, the very opposite would be the case.

Of course, it is possible to increase intra-abdominal pressure by performing a Valsalva maneuver. In the gedanken experiment, this would be simulated by a contraction of the entire muscular cylinder. It is apparent that a manometer connected to the fluid-filled chamber would register an increase. The membrane would bulge farther out of the "hernias."

Would there now be an upward bulge through the holes in the diaphragm? No, because the air pressure above the diaphragm is now increased by an amount that is exactly equal to the elevation caused by the muscular contraction in the

lower compartment. The pressure gradient across the diaphragm remains unchanged.

In the experiment, as in the body, it is impossible to contract only the wall about the air chamber or only about the water chamber. Any transient differential is immediately compensated by an upward or downward motion of the diaphragm. The only way an upward protrusion through the holes in the diaphragm could occur - in man or in the experiment - is if the diaphragm could move downward without expanding the abdomen.

These pressure relationships are, of course complicated by transient effects, blows to the abdomen, etc. However, they explain why hernias seldom occur at the other superior openings in the diaphragm, e.g., those for the aorta and inferior vena cava or the foramina of Bochdalek and Morgagni or via the transdiaphragmatic lymphatics or at the fat-filled openings in the diaphragm seen on 6% of CT examinations.

An eventrated diaphragm may be so thin as to be little more than its membranous investments, yet organs do not herniate through it. Such a thinning of the abdominal wall would lead to gross herniation.

The intra gastric pressure in a supine patient is about 2.7 inches of water - about the depth of the catheter below the skin surface. It is numerically equal to the intra-abdominal hydrostatic pressure plus the pressure generated by gastric tone. Certainly, this is not high enough to stretch or rupture the PEL. How then can we explain the enormous incidence of "hernias" at the esophageal hiatus?

The explanation, of course, is the special circumstance that a powerful muscle is pulling the stomach through from above. The force of a LM contracting up to 42% of its resting length is what does it.

HH morphology is only consistent with LMC pathogenesis

A further line of proof that traction from LMC causes HH is more extended. It is of considerable radiological interest, however, because it explains the morphology of the several types of HH. The argument is based on the classification and relative frequency of the three classical types of HH. It also explains the relative frequency of each and leads to an understanding of the role of the PEL in HH.

Following Akerlund(25) we can define three types:(26)

Type I - The Axial or sliding HH

This is the most common type. It is seen in younger individuals and its gastric portion is aligned with and centered on the long axis of the esophagus. It generally requires a Valsalva maneuver to demonstrate it. Unless large, it is self

reducing. The captive bolus test is positive. It is very frequently associated with GE reflux.

Type II - The "molar tooth" variety

The "tooth" appearance is due to the distal end of the esophagus telescoping into the fundus of the stomach. It occurs in older patients and its demonstration does not require a Valsalva test. It can be elicited by using a bolster, pressure on the abdomen, bending forward or - most commonly - without any maneuver at all. Gas in the stomach, or merely the buoyancy of the attached omentum, floats the fundus through the hiatus into the chest. It is larger than the type I hernia. It either has a molar tooth shape or a pronounced angle of His is present, but not both. The captive bolus test is negative. It tends to be asymptomatic.

Type III - The "paraesophageal" variety

Although the distinction is seldom made,(27) the name "paraesophageal hiatus hernia" can be understood in 2 ways: a.) as meaning a hernia through the esophageal hiatus alongside of the esophagus or b.) as a hernia through the diaphragm beside the esophageal hiatus. The distinction may be moot as both are so rare their very existence is questionable. The published illustrations appear to be large Type II HHs. Both the fundus and the gastroesophageal junction are above the diaphragm. For either definition to apply, the GE junction would have to be normally situated at the diaphragm.

In these large hernias, the fundus, instead of telescoping over the shortened esophagus, can float up into the chest beside the esophagus in a way that produces an acute "angle of His." This has been a source of confusion. A paraesophageal HH would have a sharp angle of His because, while the esophagus remained securely anchored by the PEL, the fundus of the stomach, having broken through the PEL, would lie in contact with its lateral aspect.

A logical fallacy accounts for the misidentification: because paraesophageal HHs would have an acute angle of His, it does not follow that a HH with an acute angle of His is paraesophageal. In reality, such HHs, while morphologically distinct, are no different from Type II HHs etiologically or functionally. Yet surgical operations to create an angle of His have been based on this fallacy.

The morphological feature that makes a HH truly "paraesophageal" is a firm attachment of the esophagus to the diaphragm to the right of the fundus. The appearance may be simulated by the slope of the diaphragm, but one can always prove unequivocally that the HH is not paraesophageal if the lesser curvature of the stomach in indented by the diaphragm in any projection as this could not occur if the PEL were still intact.

TABLE 1

Comparison of Type I and Type II hiatus hernias

	Radiological sign	Slider	Non-Slider
1	Size	up to 8 cm	larger than 8 cm
2	Self-reducing	yes	no
3	Captive bolus test	positive	negative
4	Axial	yes	variable
5	Diaphragmatic notch	no	yes
6	Reflux	frequent	seldom
7	Angle of His	never	frequent
8	Esophagus	taunt	redundant
9	Associated LER	frequent	seldom
10	Shape	bell, turnip	molar tooth
11	Frequency	90%	10%
12	Age	younger	older
13	Sphincter effacement	complete	incomplete

This diversity of morphology (Table 1) has a unifying principle: the PEL is intact in Type I (sliders) and ruptured in the other(s). Although the PEL is a structure that can be visualized directly only in part, its presence is manifested by the way it affects the fundus and esophagus.

1. Size: The slider remains small - the vast majority of them are 4.5 cm in length and they rarely exceed 7-8 cm -- because the esophagus is tethered to the diaphragm by an intact PEL. Once the PEL ruptures, nearly the entire stomach can rise above the diaphragm because its only restraint is then the gastric attachment to the retroperitoneal portion of the duodenum.

2. Self-reducing: When the LM contracts, it stretches the PEL. The sliding HH reduces spontaneously because there is a restoring force - the elasticity of the PEL. Once the PEL ruptures, there is nothing to pull the fundus back into the abdomen when the LM relaxes. Reduction of sliders is sometimes partial as, to the extent the PEL is permanently elongated, it cannot completely reduce the HH.

3. Captive bolus: The captive bolus phenomenon depends on an intact PEL to constrain abdominal tissues about the fundus and so obstruct it. Thus, it is positive in sliders and fails when the PEL ruptures. Although the Valsalva maneuver may also provoke a Type II HH, it is not necessary as even a gas bubble in the fundus can float it through the hiatus once the PEL is gone.

4. Axial: The slider is axial because it is retracted from above by the LM. The Type II HH is not axial because, once the PEL ruptures, the fundus follow the

path of least resistance, either rolling by ("periesophageal") or telescoping over ("molar tooth") the esophagus. Because the LM is no longer involved in HH production at this stage, the esophagus does not contract and get out of the way of the herniating fundus.

5. Diaphragmatic notch: Once the constraining effect of the PEL is destroyed, the stomach can slide freely through the hiatus. The diaphragm forming the left edge of the hiatus then causes a distinctive notch on the greater curvature that moves up and down the curvature with respiration as the stomach remains stationary while the diaphragm moves.

6. Reflux symptoms: Oddly enough, patients with the larger Type II HHs picked up on admission chest films the patient may be asymptomatic. Earlam,(28) for example, states, ". . . they are not associated with gastroesophageal reflux." Paradoxically, the symptoms are inversely related to size. This tells us that an intact PEL is a factor in reflux. This connection will be discussed in detail in the chapter on gastroesophageal reflux.

7. Angle of His: The angle of His is only a potential angle. Normally, esophageal LM tone keeps the fundus snugly against the under surface of the diaphragm obliterating the angle. Once the PEL ruptures, the angle can form because the entire fundus is above the diaphragm

8. Taunt esophagus: Because it provides the motive force, the LM is taunt when retracting a slider but (usually) passive and relaxed during the occurrence of a Type II HH. This may seem a subtle distinction, but fluoroscopically it is a reliable distinguishing sign.

9. Associated LER: These are more common with sliders because the esophageal mucosa never has an opportunity to adapt to a shortened state. When the esophagus relaxes, the elastic PEL restores its length. Just as a sphincter can close but not open itself, a longitudinal muscle can shorten but not elongate itself. Once rupture destroys the length-restoring force of the PEL, the esophageal mucosa can fit to a shortened organ and so lose the redundancy that is necessary to form the accordion-pleat fold. A LER may then disappear, become less prominent or become shallower and thicker.

10. Shape: Only when the PEL is ruptured can the stomach telescope over the end of the esophagus or roll alongside of it.

11. Frequency: Sliders outnumber other types about 10 to 1(29) because they represent the initial stage of a process. Not every stretched PEL goes on to rupture. However, the tendency is to an increase in size with the passage of time. Of 19 patients followed 6 or more years by Sprafka et al.,(30) 11 (58%) showed progression from small to large HHs

12. Age: The longer the PEL is exposed to the trauma of swallowing many thousands of times daily, sustained hypertonia of the LM, belching, gagging or episodes of vomiting, the more likely it is to rupture. Hence the older age of the patient with the ruptured PEL.

13. Sphincter effacement: A frequent finding in PEL rupture is a thicker ring like narrowing at exactly the location of the physiologic sphincter (1-2 cm above the ora serrata). It is about 1 cm in length instead of web-like. It represents the uneffaced physiological sphincter itself - uneffaced because an essential element of the effacement mechanism, the PEL, has been destroyed.

Basically then, the PEL is what determines the morphology of the GE junction. Although this conclusion was reached by a phenomenological route, it is possible to demonstrate the actual ragged skirt of ruptured membrane radiographically in Type II HHs if one searches for it.

In the early years of my interest in HHs, I diagnosed many paraesophageal HHs. Oddly enough, once theoretical considerations led to the conclusion that they could not exist, with one exception, I never saw another. Despite the resemblance, they all turn out to be ruptured PELs. The single exception was iatrogenic.

One should, perhaps, be diffident in refuting the opinions of surgeons who have had the benefit of exploring these patients and yet have reported many "paraesophageal" hernias. Surely, they would note whether or not the PEL was intact on one side of the hiatus. Evidently, however, they just accept the radiologist's classification without making a point of examining this critical connection.

In type II HHs one can see a sliding constriction in the stomach as it passes through the diaphragm. It slides down the stomach on inspiration and up on expiration. In doing so it rubs the longitudinal rugae against each other promoting erosions and bleeding -- the so-called "watermelon stomach."(31)

I do not wish to give the impression that the differential diagnosis between the two types of HH is always sharply etched. There are stretched, inelastic PELs that can confuse the issue by presenting some signs of each variety. Nevertheless, usually, the differential is obvious.

As an alternative name for Type II and III HHs, "rupture of the PEL" is somewhat of a simplification. There are 5 layers of tissue in the PEL and any combination of them can lose its elasticity allowing the others, e.g., the pleura or peritoneum, to stretch and so conceal the rupture of the elastic connective tissue that forms the ligament proper.

The genesis of HH

It is the absence of paraesophageal HHs that makes a compelling contribution to the proof that LMC causes HHs. Although it has been shown that the steady state hydrostatic pressure at the diaphragm is zero, what about transients - cough, sneeze, blows to the abdomen, etc.? The skirt of PEL obturates the hiatus. If transient elevation of intra-abdominal pressure were the cause, a herniation, if it occurred at all, would first work its way through the weakest part of the PEL. As the entire circumference of the PEL would hardly weaken simultaneously to the same extent, most early, small herniations would be paraesophageal extrusions! This is exactly the reverse of what actually occurs.

Traction from above, on the other hand, stretches the entire PEL without initially rupturing any of it. We know that Type I sliders far outnumber all the rest. The obvious conclusion is that the smaller, sliding HH is an earlier stage of the larger, Type II HH. Rupture of the PEL is the event that converts a Type I to a Type II.

The morphology and frequency distribution of the various types of HH, therefore, are consistent with traction from above and inconsistent with the conventional assumption they are caused by pressure from below.

The only distinction is that the PEL is intact in Type I and ruptured in Type II. It seems appropriate, therefore, to discard the various classifications and place emphasis where it belongs - on the state of the PEL - discarding the inappropriate "hernia" designations. What we have been calling a "sliding hiatus hernia" is more accurately designated "elongation of the phrenoesophageal ligament." What has been called a "paraesophageal hernia" or "Type II hernia," etc. is simply "rupture of the phrenoesophageal ligament."

The analogy with the ureter

The esophagus and the ureter are comparable organs. Their walls are composed of alternating layers of circular and longitudinal muscle. They are both fixed at either end instead of being loosely coiled like the intestine.

Physiologically both organs have one-way characteristics as there is a physiologic need to prevent reflux from the stomach in the one case and the bladder in the other. The esophagus cannot easily tolerate acid and the kidney cannot tolerate ascending infection from the bladder.

Pathologically, the main diseases of both organs result from failure of their one-way characteristic with reflux from the terminating organ. A large literature has grown out of the resulting problem of vesico-ureteral reflux and its treatment. I shall not attempt to analyze or digest it but merely point out that the analogy with the esophagus is not a superficial one.

Just as the esophagus by longitudinal contraction draws the stomach out of the abdomen into the thorax, there is evidence that the LM of the ureter can avulse the ureter from the bladder. The mucosa, of course, remains intact, but the orifice is moved cephalad and the ureters develop bulbous distal extremities that, when extreme, are remarkably faithful miniatures of a HH.

The treatment rationales are identical except instead of ascribing competence to a sphincter, the oblique insertion in the bladder muscle is given credit for ureteral competence against reflux. It seems likely that LM spasm not only avulses the ureters (intravesicle pressure surely does not do it!) but, by the same vector resolution, causes reflux. I have seen one ureter that presented a fair approximation of tertiary contractions.

Sandifer's syndrome and whiplash injuries

Children affected with this condition maintain a posture of extreme dorsiflexion of the cervical spine. This causes sustained and repetitive traction on the esophagus and PEL.(32) All reported cases had HHs.

TABLE 2

Six Cases of Sandifer's syndrome

	Case Number					
Symptom	1	2	3	4	5	6
Hyperextension of neck	X	X	X	X	X	X
Reflux	X	X	X	X	X	X
Hiatus hernia	X	X	X	X	X	X
Vomiting with meals	X	X	X	X	X	X
Post-op relief	X	X	X	X	X	X
Esoptagitis	X	X	X		X	
Aggravated by eating	X		X			
Anemia	X	X		X	X	X
Elevated fundus	X	X			X	
Esophageal stricture			X		X	
Dysphagia			X		X	
Abdominal pain				X		X
Age of onset (months)	20	48	0	`	60	

Orthopedic surgeons who see cases of whiplash injuries of the cervical spine report dysphagia as a component of the post-whiplash syndrome. It would seem that he mechanism of injury is violent dorsiflexion of the cervical spine applying a sudden force to both the superior and inferior attachments of the esophagus. I have seen three patients in their 2nd or third decades with a history of whiplash syndrome who had radiologic signs of rupture of the PEL. This mechanism explains both the dysphagia (trauma to the superior attachments) and the rupture

of the PEL. The mechanism of injury is identical with that of a tear of the trachea or main-stem bronchus, but, because it does not lead to life threatening consequences, it can easily be overlooked.

The power of LMC

It is appropriate to ask, "What kind of force could rupture the PEL? Is it possible that a thin layer of striated and/or smooth muscle could contract with enough force to tear this structure?"

Whatever the cause of increased irritability or contractile power of the LM, there is ample evidence that its tensioning and stretching of the PEL can weaken it. One has only to observe a vomiting patient at the fluoroscope to be convinced the power is there. The esophagus contracts instantly, violently retracting up to a third of the stomach above the diaphragm. Just as quickly, with LM relaxation, the herniation reduces as the PEL literally snaps it back into place. Seeing this, even in a young patient or infant, the wonder is that the PEL is not ruptured in a single vomiting episode.

Earlam(33) cites in detail Herman Boerhaave's graphic description of the patient whose rupture of the esophagus following self-induced vomiting was the first case of Boerhaave's syndrome. At autopsy, the esophagus was found to be completely separated from the stomach!(34)

A more dramatic proof of LM power would be difficult to find.

Although dogs do not naturally develop HHs because of a thick, strong PEL(35) H. Daintree Johnson(36) demonstrated typical HHs with cineradiography in dogs by inducing vomiting with apomorphine.

The process is identical in man. Viewing this instant massive spasm(37) does not engender optimism that a delicate transthoracic Allison repair of the PEL(38)(39) will survive postoperative emesis. Raphael et al. reported only a 25% recurrence rate in 114 Mayo Clinic patients who had postoperative evaluation after HH repair, however, small recurrences were not counted! They were puzzled that the patients experienced symptomatic relief even though the HH recurred. This is not as strange as it might seem. Operative rupture of the PEL, because it destroys the ability of the LM to open the sphincter, may be an effective treatment for GER.

Does the stretching result from such violent, episodic LMC or is it a matter of constant tension wearing away stone? I tend to favor the latter - at least for the stretching seen in the so-called slider. If one hangs a weight in the ear lobe, Ubangi-fashion, it will eventually produce elongation. In the same way, a hypertonic LM exerts a constant, lifelong tension on the PEL that must eventually elongate the ligament. One can be sure that this is the case because patients will

tell you they feel this tension a major fraction of the day. And this is what they say: "Everything I eat turns to gas!"

The significance of the HH concomitants

A further powerful line of proof is a synergistic one: HH is not an isolated disorder. It occurs with LERs, reflux, esophagitis, "gas" symptom, tertiary contractions and non cardiac chest pain unrelated to circular muscle contraction. The association of multiple abnormalities with each other makes it increasingly difficult to use different ad hoc explanations for each of them. It will be shown, for example, that there are separate lines of evidence that LERs are caused by LMC. Thus, every fact that tends to show that LMC causes the rings is a further piece of evidence that LMC causes HH because of the invariable association between LER and HH.

The same is true for the other concomitants of HH mentioned above. If a single mechanism accounts for LER, GER, HH and TTCs, it is more likely to be correct than four unrelated hypotheses each of which can only explain one of the four.

The association between HH and Zenker's diverticula

Although is is not universally accepted, the current view of the pathogenesis of Zenker's diverticulum is that pressures generated during deglutition force an outpocketing through a congenitally weak area, Lanier's fascia. This postulated mechanism does not explain the remarkably strong association between HH, GER and Zenker's diverticulum. Henderson et al.(40) found that of 75 patients with "cricopharyngeal dysphagia," 11 with diverticula had HH. This is probably the lower limit of the association.

Smiley et al.(41) became interested in the association and made a special effort to call patients back after surgical treatment of the diverticulum to reexamine them for HH. Prior to surgery, it was often difficult to demonstrate a HH because the patient could not swallow enough barium for an adequate study. Of 32 patients with Zenker's diverticulum, 30 (94%!) also had HHs. After reviewing the literature and evaluating the various mechanisms proposed to explain Zenker's diverticula, these authors concluded that GER caused a "dysfunction" of the cricopharyngeus muscle that initiated diverticulum formation. They were themselves dissatisfied with that formulation. Reviewing the subject in 1985, Lerut, Leman and Gruwez(42) concluded the origin of these diverticula remains unknown. As of 1994, the cause is still "controversial."(43)

As usually happens when two conditions coexist, speculation has centered on how one causes the other. Seaman(44) states that "Neuromuscular incoordination . . . is held to be responsible . . . but the evidence is conflicting." The conflict is between studies showing that the superior constrictor does not relax normally, that it relaxes normally but closes too soon, and that it relaxes too

late.(45) With the most current techniques (Arndorfer pneumo-hydraulic capillary infusion system) Knuff et al. found normal relaxation of the upper esophageal sphincter (UES) and no evidence of spasm, impaired relaxation ("achalasia") or incoordination in their nine cases.

Yet one strives in vain to conceive how the three abnormalities could be etiologically related if increased intra-abdominal pressure or a weak area of the diaphragm is the cause of HH, low resting sphincter pressure the cause of reflux and neuromuscular incoordination the cause of Zenker's diverticulum.

Yet the concurrence is obvious once one knows that HHs are due to LMC. The attachments of the esophagus are to the diaphragm and to the hypopharynx, when the LM contracts it exerts an equal and opposite tension on either end. If it is powerful enough to avulse the lower end of the esophagus from the diaphragm, it is obviously strong enough to stretch or tear the superior attachments as well. One need not postulate either an area of congenital weakness or "neuromuscular incoordination." Once the attachments to the hypopharynx are stretched or torn, the normal pressures generated by deglutition will do the rest.

Actually, there is little evidence of a weak area at the point of origin of Zenker's diverticula. The hypopharyngeal wall at this point has a double thickness - the cricopharyngeus and the inferior constrictor of the hypopharynx overlap. Wilson(46) states, "I have dissected the posterior wall of the pharynx on many occasions and have always failed to find anything to suggest a natural triangular area of weakness in this position."

Heuristically, I find this association particularly elegant as each disease provides the clue to the cause of the other and for a bonus solves another puzzle - cricopharyngeal dysphagia.

Cricopharyngeal dysphagia

The deep indentation of the barium column by the cricopharyngeus muscle was once thought to be due to spasm. Subsequent manometric investigations disproved this. Mistiming has been incriminated but then disproved.

Cruse et al.(47) investigated the microscopic pathology in a series of 7 patients ranging from age 1 to 70 who had a cricopharyngeus myotomy for treatment of dysphagia using tissue obtained from the myotomy. Twenty post mortem examinations served as controls. The pathologic findings encountered were death of muscle cells, phagocytosis, replacement fibrosis, shrunken myotubes, atrophy, regeneration, etc. None of the controls were so involved. Torres et al.(48) describe only hypertrophy, but their study was keyed to demonstrating the correlation between the size of the impression and the size of the muscle and the sections shown may have been selected to show hypertrophy.

There was no evidence of inflammatory infiltrates or of any systemic muscular disorder, although one patient did have systemic sclerosis. After excluding primary fibrosis and myopathy,

Cruse and his associates were left with no hypothesis to account for the damaged muscle.

The histologic description provided by the Cruse group is typical of the pathology of repeated episodes of injury with repair and replacement fibrosis seen in several stages in the same specimen. As with Zenker's diverticulum, once we are aware of the mischief LMC can create at its nether extremity, we are not at a loss for an explanation of the injury to the cricopharyngeus muscle. Repeated tears from vomiting, gagging or simply from long continued tension not only account for the injury to muscle but for its repeated nature.

Stretching or tearing of the proximal attachments of the esophagus would have a predictable effect on the upper esophageal sphincter. These structures serve to resolve the force of the upward displacement that initiates a swallow. The lateral components of this force open the UES. To the extent this force is late (with ligamentous stretching) or absent entirely, the difficult of a bolus gaining entrance to the mouth of the esophagus increases.

This can best be visualized by imagining what would happen if the pharyngeal attachments of the esophagus were completely severed. Then, when the larynx rose, the transverse slit-like esophageal lumen would remain a slit and the bolus would simply overflow it, pooling in the pyriform sinuses and valleculae, regurgitating into the nasopharynx or being aspirated. In other words, exactly what happens with cricopharyngeal bars.(49)

One can hope that in 40 years, when and if this information has infiltrated the conventional wisdom, cricopharyngear myotomy will no longer be practiced.

Shortcomings of the increased intra-abdominal pressure hypothesis

This hypothesis - it is more of a default judgement - fails to account for the morphology of the various types of HH. It gives a false prediction of relative frequency. It gives no explanation of the association with LERs, reflux and TTCs and does not account for symptoms.

This lack of understanding has given rise to the situation in which clinicians complain that radiologists have become too proficient at demonstrating HHs. The consensus of one symposium was that HHs are only significant (refluxwise) if they are demonstrated without trying too hard! Yet the association of HH and reflux is obvious once it is known that they are both due to the same cause - LMC.

Therapeutic implications

If large hernias are less symptomatic than small ones, is there any point in making little ones out of big ones? A consensus is emerging that there is no point in treating a HH per se but that the emphasis should be on antireflux procedures.(50) Unfortunately, in many minds "antireflux" still means creating an angle of His. As a result a fundoplication is by far the most popular surgical approach. Treatment should be directed at the symptoms of LER, strangulation and reflux. The mere presence of a HH is no indication for treatment.

Given that there is an indication for surgical intervention, does the Allison procedure (plastic repair) make sense in the light of the pathogenesis? This is surely a question that will be debated - radiologists' views on treatment are seldom embraced by surgeons. However, I have fluoroscoped too many vomiting patients to have any confidence that a surgical repair of the PEL will survive even one emesis. Given that the state of the LM is known to be "hyper", the stress that caused the HH in the first place will frequently cause a postoperative recurrence. Years ago, when anesthetists were less expert at preventing postoperative emesis, recurrences were routine. They probably occurred in the recovery room.(51)

On the other hand, the pulldown operations of the original (non-fundoplication) Nissen(52) and Boerema(53) type appear rational. They are far less formidable operations and have a striking benefit. The expedient nature of the operation - a quick fix for patients in poor surgical condition - seems to have tainted it in the eyes of some. Moreover, given the repair philosophy of dealing with hernias in general, pulldown procedures had no rationale. Not surprisingly, they were not considered quite respectable, at least among thoracic surgeons of my acquaintance, and fueled what Earlam calls the trade union debate between thoracic and abdominal surgeons over the preferred approach.

However, if HH is due to the force of LMC, the shoe is on the other foot. A pulldown procedure not only has a rationale, but the rationale is a correct one. If traction caused the HH and its concomitants, countertraction is a reasonable way to treat them. Elongating the esophagus or preventing it from shortening unduly is a rational way to treat reflux. Shortening a redundant PEL should only make it worse. Rationally, rupture of the PEL makes sense! It would destroy the ability of the PEL to open the sphincter. It would prevent Zenker diverticula and cricopharyngeal tears.

One drawback would be the creation of a iatrogenic Type II HH. Barrett was concerned that the "paraesophageal HHs" would strangulate and expressed a willingness to operate on them. Although in my experience clinicians treat such cases with benign neglect, I have yet to see such a patient get into serious trouble.

SUMMARY

All lines of evidence point to the longitudinal muscle as the cause of HHs. They can be produced experimentally by provoking LM contraction, either by stimulating the peripheral end of the transected vagus or by inducing vomiting with apomorphine. They can be elicited clinically by inducing LMC by forcing the esophagus to swallow against resistance.

LMC explains the intimate association of HH with LERs, tertiary contractions, cricopharyngeus spasm, Zenker's diverticulum, non-cardiac chest pain, "gas" and reflux. It causes all of them.

LMC accounts for the morphologic details of the various HHs and explains their relative frequency. Sliding HHs differ from other types because, in them, the PEL is intact whereas it is ruptured or attenuated in the others. LMC cannot only avulse the inferior attachments of the esophagus to the diaphragm, but also weaken its attachments to the hypopharynx thus causing hypopharyngeal diverticula and cricopharyngeal bars.

Like LMC, hyperextension of the cervical spine also exerts traction on the inferior attachments of the esophagus. This accounts for rupture of the PEL in whiplash injuries and for the production of HH in Sandifer's syndrome.

The multiple causes of vomiting, whether it be intestinal flu, food poisoning, anesthesia, drug reactions and the like, provide all of the trauma required to produce the appearances seen by the radiologist and surgeon.

Initially, the PEL undergoes elastic elongation and contraction. The elongated membrane forms a tent-like hood over the retracted fundus. When LMC subsides, the elastic recoil of the PEL restores appearances to normal. The tent vanishes and the fundus returns to the abdomen. The process is perceived as a "sliding HH."

With the passage of time and the repeated insults of life, the PEL loses some of its elasticity and elongates to permit sliding HHs up to about 7-8 cm to form. Beyond that, it will not stretch. The next time the patient vomits, the PEL ruptures. The sliding HH is cured, and a new, generally less annoying syndrome supervenes - rupture of the PEL. The latter is occasionally marked by a mild dysphagia due to non-effacement of the sphincter or anaemia due to the mechanical trauma of rugae rubbing against each other as they pass through the diaphragm with respiration.

The original limitation on size is now removed so the amount of stomach in the chest can be much greater. Portions of the omentum are then seen in the chest. The edge of the diaphragm can be seen notching the greater and lesser curvatures of the stomach, riding up and down with respiration.

Such HHs occur whether or not the esophagus is shortened by LMC. The fundus must either telescope over the esophagus producing the familiar "molar tooth" configuration or push the esophagus to the right. In the latter case, there will be an acute angle between the esophagus and the fundus. This is usually cause for misdiagnosing it as a "paraesophageal HH."

A major significance of hiatus "hernias" is their reliable testimony to an abnormal increased tone of the LM.

A more accurate knowledge of pathogenesis should lead to improved treatment. Although the term "hernia" is sanctioned by long usage, it is not appropriate. Elongation of the PEL and rupture of the PEL are the correct designations. A shorter, etiologic designation would be "esophageal transtraction" or "a gastric transtract" for sliding HH and "rupture of the PEL" for the others.

For present purposes, however, we are now able to use the insight gained from the analysis of H Hs to take a fresh look at "achalasia" and the various brands of "esophageal motor disorders". The result of this application may be unexpected.

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"Achalasia" and related misdiagnoses

It defies comprehension that a disorder causing such profound muscular hypertrophy that it is treated like hypertrophic pyloric stenosis is still universally attributed to a motor neuron deficit . This is equivalent to believing that the denizens of Muscle Beach are paralytics.

On the face of this inconsistency it should be obvious that "achalasia," in the conventional sense, is a misdiagnosis. The related "esophageal motor disorders" (EMDs) that lead up to achalasia (including "idiopathic muscular hypertrophy") must also be misdiagnoses. Our task is to arrive at the correct diagnosis. With an appreciation of the cause of hiatus hernia to build on, we can do so. To preserve for the reader the pleasure of discovery, I will depart from a principle of exposition and not reveal the correct diagnosis up front.

In the discussion, a familiar awkwardness crops up, for the condition under scrutiny is no more "achalasia" than a "hiatus hernia" is a hernia. Let it be understood, therefore, that in what follows achalasia has reference to the constellation of signs and symptoms that is called "achalasia" and that no acceptance of the implied pathogenesis is meant. Once the pathology of the condition is understood, the present name should go away.

Historical review

Early observers of achalasia took it for granted there would be increased tone in the lower esophagus because a standing column of barium in the lumen well above the diaphragm seemed to indicate a measure of resistance to outflow. Once transducers and manometry became available, this simple concept was no longer tenable: measurements with non-perfused catheters did not show the expected elevation of pressure.

But the appearances persisted and had to be explained. Beginning in the 1940s, those who followed Hurst and Rake(1) preferred to refer to the condition previously known as "cardiospasm" as "achalasia."(2) According to the new concept embodied in the name, the condition was not a spasm of the sphincter but simply a failure of the sphincter to relax. "Failure of relaxation" papered over the radiographic appearance and the manometric findings. What to a radiologist looked like spasm of the sphincter, to the manometrist, did not. Although a barium swallow was arrested at the diaphragm, a bougie or esophagoscope passed through with "gentle pressure."

The new term was still unsatisfactory. While stating the obvious, like the term it replaced, it implied that there was some intrinsic malfunction of the sphincter. Although it inspired many studies of the pharmacology and neurology of the sphincter, the implication was never established. The attention thus directed to

the sphincter reinforced the idea that the disease was an aganglionic destruction of normal reflex control.

The idea of an aganglionosis arose from the finding, again by Hurst and Rake, of a 90% reduction in the number of ganglion cells per low power field in sections taken from the body of the esophagus. A 90% loss of ganglion cells could be expected to interfere with the normal function of the delicate and complex submucosal and myenteric plexuses - whatever that function might be. Unfortunately, once the problem is pushed down to the cellular level, it becomes a problem for the neurophysiologist.(3)

The aganglionic theory got additional support from the finding of Kramer and Ingelfinger that the achalasic esophagus was hypersensitive to Mecholyl. According to Cannon's law,(4) this also implied a denervated end organ.

The aganglionic hypothesis found ready acceptance because of two analogs or "models" - aganglionic megacolon and Chagas' disease.(5) The strong radiologic resemblance between achalasia and aganglionic megacolon - a narrow segment of gut with a grossly dilated gut behind it - supported the hypothesis. Chagas' disease, an aganglionosis due to destruction of ganglion cells by a trypanosome, produced an esophageal appearance identical with achalasia. Apparently clinching the proof was the finding of degeneration of cells in the dorsal motor nuclei of the vagus and the nucleus ambiguus, the centers for neurological control of the esophagus.(6)

Thus, by 1969 Misiewicz et al.(7) could accurately state, "It is generally agreed that achalasia of the cardia is caused by degeneration of the myenteric nervous plexus." In a more recent review (1983), Wong and Johnson(8) found that the focus of current research is still on the neurologic changes but concluded that the underlying cause of these changes was unknown. A 1986 surgical monograph(9) includes achalasia in the chapter headed "Neurogenic Disturbances." Castell(10)

(1986,1992) indorses this view. Investigating the complex neuropharmacology(11) remains a problem of great interest, however, because of the widespread conviction that achalasia is caused by an aganglionosis.

In the last twenty years, there have been nascent changes in the outlook on achalasia that, without refuting the aganglionosis theory, have begun to put the disease in a different light. Suspicion that diffuse esophageal spasm (DES) was intimately associated with achalasia was well documented in a significant single case report in 1967 by Kramer, Harris and Donaldson.(12) They followed a patient for eight years during which the clinical, radiological and manometric picture evolved from that of DES to achalasia. There is a still earlier report by Schroeder, et al.(13) recording the same transition and Barrett(14) also reported observing a transition from what appeared to be diffuse spasm to cardiospasm.

The significance of these cases was long in being recognized. In 1979 Vantrappen's Belgian research group,(15) aware of the association, attempted to define four classes of esophageal motility disorder (EMD) according to the presence or absence of peristalsis and sphincter relaxation. These were further subclassified as vigorous or non-vigorous, making eight groups in all.(16)

As a result of study of a large series, and perhaps as a way of sidestepping the cardiospasm-achalasia-DES tangle of nomenclature, the all-inclusive term "esophageal motor disorder" (EMD)(17) is now employed for them all. This implies a belief they share a common, unknown, cause.(18) Thus, there is reason to hope that, if we find a cause or cure for one, we have found it for all.

It is probably fair to say, however, that the aganglionosis theory of achalasia is still regarded as essentially correct(19) or correct but incomplete. That it remains so entrenched after more than two generations and an enormous number of clinical, surgical, radiological and physiological studies, is a splendid exemplification of Kuhn's(20) thesis that scientific research will always be interpreted in terms of the current theory whether or not the theory is correct. Many camels have been swallowed in the process.

Flaws in the aganglionic theory

On examination, the evidence for an aganglionic origin of achalasia proves flawed. The lack of any difference in response to a broad range of sympathetic drugs between muscle strips taken from normal and achalasic patients(21) is unexpected and argues against an aganglionosis. Patients with achalasia do not have autonomic nerve deficits elsewhere in the gut.(22)

Moreover, the analogy with aganglionic megacolon is patently false. "In Hirschprung's disease there is usually a narrow segment with no ganglion cells, a transitional zone with few cells, and proximal gut with normal neurons, dilated because of the distal obstruction."(23),(24),(25) This is precisely the reverse of the situation with achalasia in which the dilated region is supposed to be aganglionic, and the sphincter area to have ganglion cells.

Nor does the analogy with Chagas' disease withstand scrutiny. Padovan, Godoy et al.(26) studied 24 patients with Chagas' disease and found that the average resting LESP(27) was three standard deviations below normal. Moreover, these patients were less sensitive to pentagastrin than normal subjects. These results are the exact opposites of the findings in achalasia. Such patients are reported to have high resting LESP and are supersensitive to pentagastrin.(28) Holloway, Dodds (29) most recently report resting LESPs in achalasia five standard deviations above normal (41 mm Hg). Both groups found average values of 20 mm Hg in their normal controls. The Holloway, Dodds group found untreated achalasia patients to be supersensitive to pentagastrin as did Orlando and

Bozymski.(30) Animal studies in Chagas' disease(31) suggest that aperistalsis precedes incomplete LES relaxation.

Therefore, both "models," instead of supporting the aganglionic hypothesis, clearly refute it. Moreover, on analysis the cell "reduction" turns out to be a geometrical artifact. The reduction is expressed in cells per low power field.(32) A microscopist is accustomed to making judgements of relative frequency of cells by counting their number per microscopic field. Although this method is valid as a rule, it grossly underestimates the cell count when applied to an inflated and stretched organ.

As the esophagus dilates, ganglion cells get farther apart so that there are fewer per microscopic field. Like dots on the surface of a balloon, as the balloon inflates, there are fewer dots per square cm although the total number of dots is unchanged. So accurate is the variance of the cell count with radius that, given one, we can calculate the other with great accuracy. As this fact is generally overlooked, the idea that achalasia is an aganglionic dysplasia has survived for two generations to obfuscate any serious analysis before it starts.

The geometry also explains why investigators who took sections from the undilated sphincter region found no significant reduction in ganglion cells although, in the opossum, for example, ganglion cells reach a nadir in the sphincter region.(33)

Under the conditions prevailing in esophageal dilatation, many muscle cells are effectively denervated. Stretching neurons and their processes in two directions to many times their normal length may exceed their elastic limit or even avulse them from motor end organs. In addition, stasis and infection can more easily reach the myenteric plexus to cause degeneration through the thinned esophageal wall. These effects can account for the hyperreactivity to Mecholyl.

The hypersensitivity of the achalasic esophagus to cholinergic drugs is undeniable, but the conclusion drawn from this - that the cause of the disease is aganglionosis - does not necessarily follow. Many cases of DES also have a positive Mecholyl test, although DES is not an aganglionic disease. Pathologic studies(34),(35) have failed to show degeneration of the myenteric plexus in DES. The sensitivity seems to progress as DES tapers into achalasia. Making use of this fact, graded doses of Mecholyl have been used(36) to differentiate the two or to place a patient in the DES-achalasia spectrum.

While this demonstrates the kinship of the two conditions, it also shows that the seeds of achalasia are already present before the presumed ganglionic degeneration occurs. Indeed, with further progression of the disease, the hypersensitivity disappears. This is attributed to degeneration of the muscle end organ with late stage disease. It is equally valid to attribute degeneration of nerve cells to the same cause.

Well documented complete recovery from clinically, radiologically and manometrically typical achalasia(37) and its forerunner, diffuse esophageal spasm,(38) is inconsistent with the aganglionosis hypothesis.

Finally, the brainstem lesions are not primary. Commenting on the central lesions, Earlam,(39) after reviewing the evidence for trans-synaptic degeneration of postganglionic cells after destruction of their afferent nerve supply, concluded that "there is absolutely no experimental evidence that trans-synaptic degeneration actually occurs." Because it is unlikely that well separated nuclei in the medulla would be affected bilaterally, Earlam concluded it was " . . . far more likely that the central nuclei degenerated after losing the neurons of the esophageal wall as retrograde degeneration is well documented."(40) Dogs, the best studied animal analogue, also have normal ganglion cells early in the disease.(41) The ganglion cell degeneration observed is a fairly late effect, not the cause of the disease.

Why are there no cases of HH and achalasia?

In this connection, the reported incompatibility of hiatus hernia and achalasia becomes significant. Hiatus hernia is an extremely common radiologic finding. It has been demonstrated radiographically in 100 consecutive cases! It would be very surprising if many or most cases of achalasia were not accompanied by hiatus hernia. Yet, in a review of the New Haven experience, Binder (42) found that of 42 recorded cases of achalasia, only one was also reported as having HH. On review of the exception, they concluded that diagnosis had been erroneous.

HHs occur in association with every other esophageal disease -- with lower esophageal rings (LER), tertiary contractions (TC), cardioesophageal reflux, Zenker's diverticulum, scleroderma, carcinoma. It is an arresting circumstance if they do not coexist with achalasia.

This exclusive or relationship has logical significance. If we ask, "How would it be possible for an HH to preclude a diagnosis of achalasia in the same individual?" The obvious answer is that they are the same thing! That is, achalasia is simply a name for an HH with special characteristics that prevent it from being recognized as such. This leads at once to the insight that HHs are a factor in the pathogenesis of achalasia. We need only discover the nature of those special characteristics. (The possibility that HH protects one from achalasia can be dismissed.)

If a hiatal ring is too small to contain the fundus comfortably, some degree of obstruction is predictable when hiatal transtraction occurs. When the esophagus passes through the diaphragm there is ample room for it. When the fundus is drawn through the "die" of the hiatus, it becomes tubular. Because it still has the space formerly occupied by the equally tubular esophagus, it might be supposed that it has adapted to the hiatal constriction.

More detailed examination of the morphology of the GE junction shows this cannot be true. The distal esophagus can exist unobstructed in the hiatus because, except for branches of the left gastric artery, the region derives its blood supply from above the diaphragm and has no mesentery. With hiatal transtraction, the fundus is drawn through the hiatus and its vascular supply with it. Because the fundus has a mesentery, the lesser omentum, the left gastric (coronary) artery and vein, nerve fibers from the coeliac axis, peritoneum, fat, lymphatics, lymph nodes and connective tissue are also crowded into the hiatus.

The gastric wall thickness is greater than that of the esophagus. Finally, the circumference of the fundus is many times that of the esophagus. This mass of tissue, with its greater cross sectional area, is pulled into a hiatus appropriate for a much smaller organ.

Some hiatuses are small

If these supporting tissues are drawn through the hiatus, with some patients there may still be no obstruction because the hiatus is large or easily distended. Reading surgical instructions for making the hiatus smaller, one might suppose that the size of the hiatus is normally more than adequate. Because there is an impression that hiatal size is a factor in HH, the surgeon may "correct" a large hiatus with a few sutures.(43)

Some patients, however, have a hiatus that is small, barely large enough for the esophagus itself, and not easily enlarged manually. Indeed, the hiatus itself may itself be a cause of esophageal obstruction even without HH as in the following patient:

CN SF-519A, male age 68. The patient states, "Meat and pancakes choke me up." He has to interrupt meals to regurgitate food that won't go down. He has nocturnal dyspnea and regurgitation of "foul tasting stuff" especially at night. Had an excellent singing voice but lost it.

At fluoroscopy, barium passed freely through the esophagus. No HH could be demonstrated nor was there any evidence of a LER despite the typical history. The distal esophageal mucosal folds were coarse and reduced to three in number. The deCarvalho test elicited gross reflux and, when it occurred, the patient said that, in all but intensity, it duplicated his symptoms of acid regurgitation.

He managed to swallow two marshmallows each of which held up at the diaphragm but was soon forced through by an effective peristaltic wave. On spot films, the caliber of the lumen measured 1.8 cm across the momentarily impacted marshmallow in the hiatus.

Despite the extraordinarily small hiatus, there was a good, effective peristaltic wave and no esophageal dilatation. This and several similar cases, shows that the fundus must be retracted into a small hiatus to produce the obstruction of achalasia.

Surgeons calibrate hiatal size by the number of fingers that can be introduced alongside the esophagus. Harrington(44) routinely examined the hiatus in 1000 patients during upper abdominal explorations. He found that in 55% of cases " . . . no opening could be felt around the esophagus, 35% admitted one finger, 8% two fingers and 2% three or more fingers." [Emphasis added.] In operating on "short esophagus" HHs, H. Daintree Johnson(45) " . . . was struck with the fact that . . . the hiatus often seemed within normal limits."

When the gastric fundus and its mesentery are drawn through a small hiatus by LMC, they exceed the available space so that the gastric lumen is compressed from the start. Subsequent vascular engorgement from constriction of the vascular and lymphatic return aggravates the obstruction.

This, of course, is the region in which there are venous connections between the portal and systemic systems. The submucus veins of the esophagus are tortuous and run in the 4-5 longitudinal mucosal folds " . . . they connect the submucus veins of the esophagus to those of the stomach, thus forming another set of anastomoses between the portal and systemic venous systems. At or just above the level of the cardia valves may be found in the esophageal veins, but they are inconstant. If present they are so oriented that they direct the blood flow from the esophagus to the stomach." [Butler](46) Either because of valves or tortuosity, they offer most resistance to retrograde injection and are the first to become varicose in portal hypertension. Because of this, blood forced into the constricted portion of the stomach has difficulty draining into thoracic veins. Vascular engorgement and mucosal friability results.

There are many examples of true hernias in which strangulation occurs even without the entire circumference of the organ herniating. In HH the entire circumference is surrounded by a thin ring of potentially constricting diaphragm making strangulation even more likely. There is, therefore, a priori reason to believe that simple strangulation in the hiatus can occlude the lumen and produce the obstructive appearance that is typical of the disease. Is there any clinical evidence this is the actual mechanism?

Three cases

My experience bears out the strangulation mechanism. Casting aside preconceived notions, most radiologists would, I think, concur that the appearance of DES under the fluoroscope is absolutely typical of an obstructing

bowel lesion.(47) Because we have been taught to call it "diffuse spasm," we can totally overlook the correct diagnosis. The following case is probably typical in that I mistakenly believed I had done the job once I had identified a radiologic appearance with a published description:

The patient was seen for severe dysphagia after he had lost 15 pounds in a few weeks and was unable to keep even keep water down. He had a ravenous appetite.

The fluoroscopic appearance of the esophagus was that seen with an acute mechanical obstruction of the bowel. There was violent, rapid, churning esophageal peristalsis that was almost completely ineffective in forcing barium below the diaphragm. The forceful peristaltic waves lost control of the bolus as they approached the diaphragm so that barium refluxed through the advancing ring of CM contraction. The peristaltic wave immediately reformed and the process repeated. There was a typical small tubular HH.

A literature search for a comparable case turned up equivalent descriptions for "pseudo-diverticulosis of the esophagus" (German) and "diffuse muscular hypertrophy of the esophagus" (British) or "diffuse spasm" (American). Fortunately, the attending surgeon, an exponent of the Borema/Nissen pulldown operation, elected to do that procedure on the rationale that repairing the HH might somehow be beneficial.(48) It cured the patient. He had no further dysphagia and gained 17 pounds by the time he left the hospital.

I had described the appearance of an obstructed bowel without making the connection. The esophagus is gut. Even after the successful therapeutic result I was slow to realize that reducing the HH had relieved an obstruction.

One cannot always be operating from first principles. It is a useful shortcut in everyday work to make a diagnosis when satisfied that published criteria are met. There is also a time for proceeding from fundamentals, however, and the earlier case was an alert when the following patient presented.

The patient was a man in his 20s who had the diagnosis of achalasia for several years. A year previously he had a cardiomyotomy (Heller procedure) to treat his dysphagia without obtaining significant improvement. Review of previous films showed esophageal dilatation typical of achalasia but without redundancy.

On examination, a short tubular HH was demonstrated with extreme hyperperistalsis that was ineffective in emptying the esophagus. The level of the obstruction was at the diaphragm. The sphincter region was well above that level, but, because the herniated segment of the stomach was the same diameter as the esophagus, this could have been overlooked except that a slight LER identified the GE junction. In addition there was now a pulsion type

epiphrenic diverticulum in the distal esophagus that had not been present before the myotomy.

With some trepidation, I made a diagnosis of hiatal strangulation but with the caveat that this was a completely unorthodox diagnosis that went against established opinion. Because the procedure dictated by conventional wisdom had already been done, both patient and surgical consultant were willing to act on that diagnosis.

Esophagoscopy on 10/15/68 revealed " . . . patent and voluminous reflux into the esophagus in the supine position. The terminal half of the esophagus was red, edematous and friable, but no distinct strictures were noted. There was a pulsion diverticulum of the lower lateral esophagus presumably at the site of his previous esophagomyotomy."

At a thoracotomy the following day, " . . . a left lateral esophageal diverticulum was obvious and a small hiatus hernia was present. The normal anatomy of the esophagus was reconstructed by involution of the pulsion diverticulum, and an Allison type repair of the esophageal hiatus effected."(49) Two weeks later the surgeon reported " . . . he is free of symptoms of reflux and is able to swallow his food normally."

The third patient had a much longer period of observation. He had been followed more than fifteen years for various GI symptoms with diagnoses of HH, severe duodenitis, post-bulbar duodenal ulcer. There was no obstruction and no dilatation at the last examination.

On reexamination in January of 1975, He had typical findings of achalasia with delayed esophageal emptying, dilatation and redundancy of the organ. The experience with the above two patients was described to the patient, his attending internist and surgical consultants in another city where he elected to have treatment. Preferring the conventional wisdom, he was treated by forceful dilatation and, nevertheless, did reasonably well until his symptoms recurred 17 years later.

These cases illustrate several points: 1.) Simply reducing a hernia can relieve the obstruction, 2.) It can relieve the obstruction even after the classical Heller procedure has failed to do so. 3.) "Forceful dilatation" will also relieve the obstruction. 4.) None of these clinical phenomena are consistent with the postulated aganglionosis.

Cases of strangulated HH have been recognized. There are at least two reported surgical cases(50)(51) but these were seen purely as surgical emergencies and a connection with achalasia was not suspected. Henderson(52) (Figures 16.3 and 16.4) illustrates a case of hiatus hernia with the manometric findings of DES.

Many other unrecognized cases can be found in monographs and the periodical literature.

Boerema(53) reports(54) that his operation, in which he pulls the stomach down and anchors it to the posterior rectus sheath, although designed as a treatment for HH, gives excellent results with both HH and achalasia! This is exactly what would be anticipated if achalasia were a strangulated HH.

The significance of epiphrenic diverticula

There is additional evidence, from another quarter entirely, that HH can lead to strangulation and obstruction. Because they are markers for obstructive disease, it is no coincidence that epiphrenic diverticula (EPD) usually occur with either HH, DES or achalasia. In cases of EPD, an incidence of HH of up to 50% is reported.(55) Longitudinal studies of patients with such diverticula will show that they are acquired and not congenital. They present the appearance of a "blowout" proximal to an obstruction.

Epiphrenic diverticula, although highly uncommon in the general population, occur with extraordinary frequency in patients with either achalasia or diffuse spasm. Significantly, the patient of Kramer, et al. mentioned above also developed an epiphrenic diverticulum as did my patient. Harrington(56) was also aware of an association between esophageal spasm and epiphrenic diverticula. Knuff and Castell(57) found diffuse spasm in 45% of their examples of esophageal diverticula. Effler and his associates found 65% of their cases of epiphrenic diverticula had associated " . . . cardiospasm and/or diffuse esophageal spasm."(58) Goodman and Parnes(59) also reported 65% of patients with epiphrenic diverticula had achalasia. Another 6% had hiatus hernias. Habein, Moersch and Kirklin found only 3% of patients in their series of 149 cases of diverticula had achalasia but there was also a 12% incidence of "diffuse spasm." Allen and Clagett(60) found that 69% of 160 cases of epiphrenic diverticulum encountered at the Mayo Clinic over a 20-year span had either achalasia (16), DES (39) or HH (55). More recently, Henderson(61) reported " . . . either a primary or secondary motor disorder . . . in all patients [with epiphrenic diverticula] studied."

Whatever the exact percentage may be, this association of uncommon diagnoses cannot be dismissed as coincidental. It has prompted the naive conjecture that epiphrenic diverticula may cause achalasia.(62)

Epiphrenic diverticula are typical pulsion diverticula. They consist of a mucosal layer only. They arise because, propelled by the force of peristalsis, esophageal contents find it easier to bulge through the wall of the esophagus than to exit through the hiatal canal. It normally takes only 5 cm of water pressure to force the sphincter from above. Obviously, it takes more force than 5 cm H2O to blow

out the esophageal wall. The mere presence of EPD therefore, is conclusive evidence of obstruction distal to its origin.(63)

As its name implies, the obstruction is at or very near the diaphragm. Fluid under pressure follows the path of least resistance. The reason for this is that early in the progression of a p-wave, the pressurized bolus merely distends the caudad esophagus. As it nears the end of its travel, there is no more esophagus to distend. The bolus must enter the stomach or, failing that because of a distal obstruction, either reflux through the p-wave or be forced through the esophageal wall. If the wall presents less resistance than the esophageal outlet, there must be a high grade obstruction of the esophagus outlet.

A diverticulum is not a totally useless structure to be extirpated at the earliest opportunity. It serves a physiologic function as a buffer to contain a bolus that cannot be forced through the usual channel by peristalsis. Without a buffer, the incompressible liquid content of the esophagus must find an outlet when compressed. The hypertrophied esophagus of DES may resemble a string of beads because, when an en masse contraction of the circular muscle occurs, alternate segments are either compressing the fluid or being blown out as buffers. Such contractions are typical of esophageal obstruction.(64)(65) Manometrically(66) they are distinguished by the simultaneous rise in pressure at catheters placed at different levels.

Because epiphrenic diverticula are prima facie evidence of an obstruction and occur with high frequency in achalasia, we can make a useful inference: at some period in the genesis of achalasia, there was not only an obstruction to esophageal emptying but also peristaltic activity that was forceful enough to blow out the wall. But this is a description of diffuse spasm.

Achalasia is only the final, exhausted, decompensated state of the chronically obstructed gut. At this stage, there is no effective peristaltic activity present to blow out a diverticulum. As DES and achalasia share an association with EPD, it is further evidence they are related to each other and to HH.

From the above, in conformity with the mounting evidence in the literature from the isolated case of Kramer et al. to the huge series of Vantrappen et al., it seems reasonable to conclude that DES is the initial stage of achalasia.

The obstruction is at the diaphragm

In achalasia, obstruction is not at the sphincter but at the level of the hiatus. As the sphincter is 4-9 cm above the hiatus in patients with HH, it might be thought that the observer should have no difficulty in establishing this point. Yet it can be very difficult to detect the sphincter location in HH with obstruction - especially if one is intent on demonstrating the "bird beak." The fundus is drawn out into a tube that is easily mistaken for esophagus. "Tramlines" similar to those seen in

hypertrophic pyloric stenosis may extend through the diaphragm and terminate at the effaced sphincter.

The point is important, because it exonerates the sphincter as the cause of obstruction in HH. The sphincter may be as much as 8 cm above the hiatal obstruction - a level at which one never sees narrowing in achalasia.

Degrees of hiatal obstruction

Only a few HH patients progress to achalasia, but those that do are the ones with a small hiatus. Obstruction can range from partial to complete - from imperfect clearing of the lumen by a peristaltic wave to life-threatening total occlusion. Radiologically, several degrees of obstruction can be described. The earliest stage is a failure of an effective peristaltic wave to clear the esophagus completely with loss of control of the bolus.

A more pronounced degree of obstruction is the so-called "elevator esophagus."(67),(68) After a barium swallow in the upright position, barium forms a persistent column from the diaphragm to the middle or even upper third bounded above by an air-fluid level. There is no organized peristaltic wave. Instead, en masse contractions of the circular muscle occur as the esophagus attempts to propel barium distally. The fluid level, instead of descending as the circular muscle contracts, rises because the now narrower lumen must hold the same volume that a wider lumen contained before the contraction. This produces an up and down "elevator" effect on the fluid level. Tertiary contractions due to self-buffering may be seen. Neither esophageal contraction nor hydrostatic pressure will clear the organ.

Often such patients will exhibit finely granular filling defects mixed with the first swallow of barium. These represent mucus globules that have accumulated overnight in the fasting patient. The first few swallows do not wash them out of the esophagus because the hiatal squeeze is acting as a separatory funnel to retain them while allowing barium to pass through.

Simultaneous or en masse contractions of the entire circular muscle are characteristic of obstruction. They occur in various grades of achalasia and have been produced experimentally in cats with an implanted inflatable esophageal pneumatic cuff.(69) A Gore-Tex encircling band produced simultaneous contraction in 85% of 17 cats so treated.(70) When the band was removed in several animals, these abnormalities ceased. Kaye(71) found no inconsistency between the manometric patterns and a "functional obstruction" of the esophagus.

The most profound functional changes occur in patients with a severe obstruction. Here a dramatic churning peristalsis of the entire thoracic esophagus is seen. These patients may be in acute distress, unable ingest any food without

vomiting and may have dramatic weight loss. In addition, a radiologist may note gagging, and bizarre tertiary contractions of the extreme degree sometimes described as "pseudo-diverticulosis" or "curling" of the esophagus. Again, one must search carefully to find the sphincter. It may be patulous and thus invisible without the clues mentioned. Such is the typical appearance of DES.(72)

In all three stages and in intermediate stages that could be defined, the obstruction is at the diaphragm - not at the sphincter. When all else fails, finding the sphincter is the key to the correct diagnosis. Perceptually, the TCs or "corkscrew" or "pseudodiverticula" are the attention grabbers. The more subtle, but significant findings locate the sphincter.

To summarize, we conclude that early stage achalasia is no more than the obstruction that occurs with hiatal herniation. Achalasia and HH are never diagnosed simultaneously because the "herniated" fundus is so tubular it is mistaken for esophagus. Diverticula, true and pseudo, are buffers to which incompressible fluid is diverted when the esophagus contracts vigorously to force an obstruction.

Some lessons from diffuse esophageal spasm

Hiatal obstruction of the fundus explains the entire picture of diffuse esophageal spasm:

The churning peristalsis is the normal response of bowel attempting to force an obstruction.

The "string of pearls" or "pseudodiverticulosis" appearance is due to self-buffering of en masse esophageal contractions.

The "curling" and tertiary contractions are due to simultaneous contraction of the LM and CM in an esophagus that is making a maximal effort to overcome an obstruction. (Tertiary contractions are the "valvulae conniventes" of the esophagus and by the same token they imply obstruction .)

Because hypertrophy is the normal response of bowel to chronic obstruction, "idiopathic diffuse hypertrophy" is predictable, not idiopathic.

In the case of Kramer , the transition from DES to "achalasia" was not as rapid as one would expect from the spectacular appearance of the violent peristaltic and non-peristaltic contractions. Their patient was followed for eight years as he developed increasing symptoms of obstruction with fifteen manometric or balloon kymographic studies. They reported that " . . . symptoms, oesophageal radiographs, balloon kymographic and manometric records were diagnostic of diffuse spasm." After forceful dilatation the same studies " . . . were quite

characteristic of cardiospasm." Although this was not a happy result, in other cases,(73) complete restitutio ad integrum occurs.

The hypersensitivity to Mecholyl reported by Kramer and Ingelfinger(74) was present in this patient both when he was considered to have "diffuse spasm" and when he exhibited the picture of cardiospasm. Barrett also noted a transition from DES to achalasia. Kramer et al.(75) had earlier suspected that diffuse spasm and cardiospasm might be related because some but not all patients with DES also have a positive Mecholyl test.(76)(77)

Enlarging the hiatus relieves both DES and achalasia

Because "forceful" (i.e., up to 744 mm Hg) dilatation of the sphincter commonly provides a measure of relief, it is gaining advocates for all of the "esophageal motility disorders" (EMD). The binary classification of Vantrappen et al. allowed patients to be classified before and after treatment. Many changes took place after forceful dilatation. They reported(78) that "vigorous" peristalsis disappeared in half of 44 patients treated. Peristalsis returned in a third of the achalasia patients following treatment with forceful dilatation.

In dilating the sphincter, however, the operator is unwittingly dilating the hiatus because the hourglass shaped dilating bag self-centers, not on the sphincter, but on the hiatus. As another instance of the Law of Compensating Errors, things turn out right for the wrong reason. Relief of the hiatal obstruction follows enlargement of the constricting ring.(79) As with achalasia, dilatation restores the normal peristaltic wave in DES.(80)

It should be noted that the force used to dilate the "sphincter" is completely out of proportion to the delicacy of the structure being dilated. Anatomically, the sphincter is so tenuous that only recently have anatomists been willing to grant its existence. The esophageal hiatus, unequivocally anatomical, is more likely to be offering the resistance to distention.

Such therapeutic results are inconsistent with either an aganglionosis or a "disordered motor function." That an almost brutal stretching procedure could reverse a degenerative process or reorder an incoordination of motor function is inconceivable. On the other hand, dilating an obstruction can be expected to cause a return to a normal peristaltic mode.

Self-limited hiatal strangulations

It could well be objected that if 55% of patients have a hiatus that will not accommodate even one finger, there should be a much higher incidence of strangulation than is encountered, given the very high incidence of HH. This objection is valid, however, it is not really an objection. It merely shows that lesser degrees of strangulation usually go unrecognized.

There are many cases of temporary or intermittent strangulation of the fundus in the hiatus that reduce spontaneously. The portion of the fundus above the diaphragm becomes edematous and engorged. Seen after it has reduced, this engorgement may easily be misinterpreted as a "fundic gastritis" or, with potentially disastrous consequences, as a neoplasm, because the fundus appears separated from the diaphragm. The appearance is so diagnostic - a fundic "mass" concentric with the esophageal orifice - that one can make the diagnosis of "hiatus hernia without herniation."

Mucosal engorgement of the retracted portion of the fundus is so common it is an endoscopic sign of HH. Endoscopists " . . . frequently find friable or hemorrhagic gastric mucosa in the presence of normal esophageal mucosa in patients with symptomatic hiatus hernias."(81) Morrisey(82) notes " . . . relatively little attention has been paid to the gastric mucosa just distal to the mucosal junction in patients with reflux esophagitis. This mucosa often appears erythematous, friable and occasionally frankly eroded." A biopsy of the friable mucosa may be reported as normal because there are no inflammatory cells. This and the finding that some hiatuses hug the scope tightly are significant in elucidating the mechanism of strangulation. The mucosal changes affect only the organ with the constricted blood supply, that is, the portion of the fundus above the hiatal constriction. The esophagus, with an unimpaired blood supply, may appear normal to the endoscopist.

I have occasionally seen hiatal obstruction with spontaneous reduction and remission during a fluoroscopic examination:

DW 46873 M393. Female, age 52 Fluoroscopic note: Ingested barium showed moderate dilatation of the esophagus and a delay in emptying. A fluid level formed at the height of the aortic arch and only slowly descended. Unorganized, irregular contractions of the esophageal outline were noted. They would appear on one side without deforming the opposite side as may be seen with partial LM contractions. They produced no effect as far as emptying the esophagus. The dilatation was so pronounced that the left atrial border indented the esophageal outline sharply enough to at first suggest enlargement.

Although these findings were all typical of achalasia, when she performed a prolonged Valsalva test a typical HH with a lower esophageal ring appeared. As this reduced, all signs of achalasia disappeared, an organized peristaltic wave formed and the esophagus emptied completely.

HHs frequently get stuck above the diaphragm temporarily and then reduce when the stomach distends. This explains their bleeding propensity even without esophagitis. All lesions in the cardia have a greater tendency to bleed than when found elsewhere in the esophagus or stomach - a further indication of the strangulating effect of even a large esophageal hiatus.

The "inflammatory" gastroesophageal polyp: a minimal strangulation

A bulbous enlargement of a single gastric fold in the tubular portion of a sliding hiatus hernia, the "inflammatory esophagogastric fold or polyp," is a frequent finding. It may cross the ora serrata and extend into the esophagus, generally merging with an enlarged esophageal fold.

Although up to 1984 there were reports of only eleven cases,(83) the condition is common enough that many examples can be collected by the interested observer. Its frequent appearance is testimony that in many patients the hiatus is barely large enough to hold the esophagus because the appearance can only be understood in the light of the local blood supply. This is the only region of the esophagus that receives a blood supply from below the diaphragm. Impairment of the venous return produces local vascular engorgement. The existence of the fold is proof that incarceration of the retracted fundus is not momentary or infrequent.

Other hiatal obstructions look like achalasia

The conditions that "mimic" achalasia have great significance. Surgical operations about the hiatus including a too tight HH repair, post-vagotomy periesophagitis(84) and, particularly, a tight fundoplication(85) can simulate the appearance including the "bird-beak," esophageal dilatation and "motor disorders" (repetitive and simultaneous contractions). In one way or another, these procedures all constrict the tissue mass in the hiatus.

Kumar reported a cartilaginous esophageal ring similar to a tracheal cartilage in the esophagus.(86) It was indistinguishable radiologically from achalasia. The abnormality cleared after resection.

Tumors that invade the hiatus provide a more direct, unequivocal reproduction of achalasia. In the following case a tumor of the hiatus, situated half above and half below the diaphragm produced the picture of achalasia.

LL 6872 7/6/64. Female, age 76. At fluoroscopy barium passed freely through the hypopharynx but arrested at the diaphragm. At this point the esophagus showed a long taper to a caliber of a few millimeters. This portion of the esophagus pulsated violently from transmitted cardiac contraction. In the upright position, a fluid level formed above the aortic arch and fluid trickled only slowly through the esophagus. A peristaltic wave would begin to form below the aortic knob, but this never progressed distally. On one or two occasions there was some reflux through the advancing peristaltic wave into the proximal third of the esophagus. The esophagus was only slightly dilated. The cardia was narrow and somewhat separated from the diaphragm.

Multiple films of the area showed a 5-cm mass at the level of the diaphragm and situated posteriorly to the esophagus. The mass was partly above and partly below the diaphragm.

"Secondary achalasia"

It is well known that the radiologic differential diagnosis of achalasia from carcinoma of the fundus of the stomach is impossible. (87)"Idiopathic" and "secondary" achalasia are identical clinically, radiologically, manometrically and endoscopically. Even the Mecholyl test(88) and Seidlitz powder tests do not distinguish between achalasia and these conditions.(89)(90)

This has led to the notion of secondary achalasia - that is, achalasia to which a cause can be assigned. In the three cases of secondary achalasia encountered by Ferguson and Burford,(91) even the endoscopic appearance was the same as in primary or idiopathic achalasia. In the seven cases of regional carcinoma reviewed by Tucker, Snape and Cohen(92) the manometric profile was identical whether the carcinoma was of the stomach, the lung or the pancreas. The endoscopic appearance suggested carcinoma in only two cases. They concluded " . . . the radiographic, endoscopic and manometric studies . . . may not discriminate between the primary and secondary forms of achalasia." Sandler and associates(93) also found clinical criteria unable to distinguish the two.

Such cases pose insuperable problems for the aganglionosis theory of achalasia. To preserve the theory, speculation now must center on how carcinoma invading the myenteric plexus simulates or causes an aganglionosis. When searched for, however, such plexus invasion has not been found.(94),(95) A carcinoma-induced peripheral neuropathy has been suggested. This might preserve the aganglionic theory, but it is hard to conceive of a neuropathy limited to the esophagus. Nor is it reasonable to believe that five different types of carcinoma could all produce an identical, highly selective neurotoxin.

There is a simpler explanation: there is no difference between primary and secondary achalasia. All cases are secondary to hiatal obstruction. Retraction and incarceration of the fundus produce hiatal obstruction just as surely as does carcinoma invading the hiatus.

Again, Occam's razor dictates favoring the explanation that requires the fewer assumptions. A tumor invading a passageway will obstruct it. That is enough to explain the appearances. When the lesion is local, why make the additional assumption of involvement of a plexus that extends the entire length of the esophagus to explain the appearances? Moreover, esophageal carcinoma arising above the hiatus also invades the myenteric plexus but never causes the appearance of achalasia even if it obstructs.

Achalasia clears when hiatal tumors regress.

A single case report of Davis et al.(96) is of great interest because it illustrates both the ability of a hiatal tumor to produce achalasia and that achalasia resolves with tumor regression. This patient had both generalized reticulum cell sarcoma and typical manifestations of achalasia including marked obstruction and uncoordinated contractions. Manometric studies were reported as typical of achalasia. A radioactive gallium scintiscan showed a concentration of activity from D-10 to L-1 in approximately the midline, anterior to the vertebral column [including the hiatal area] and posterior to the liver. His esophagus was restored to normal functionally and radiologically after two weeks treatment with prednisone and vincristine.

The scans before and after treatment prove hiatal obstruction and relief. It could be objected that the hiatal area was never explored surgically for confirmation. This objection does not apply to Kline's(97) case of "vigorous achalasia" diagnosed by manometric findings of increased sphincter pressure (30-40 mm Hg) and simultaneous repetitive contractions. Endoscopy suggested achalasia and the radiological findings were those of dilatation of the body and narrowing at the GE junction. On exploration, an anaplastic gastric lymphoma was found extending from the GE junction to mid-stomach. Manometric and radiologic findings reverted to normal one month following treatment with cytoxin, vincristine, adriomycin and prednisone.

Nelson and Horsley's case(98) of idiopathic retroperitoneal fibrosis produced the typical long, beak-like narrowing centered on the diaphragm typical of achalasia. At exploration, a fibrotic mass arising on the posterior peritoneum enveloped the great vessels, kidneys and both crura of the diaphragm. It extended 2 cm into the mediastinum " . . . enshrouding the esophagus at the GE junction." After it was dissected out, the patient had complete relief of her dysphagia.

In the above report, the authors concluded that " . . . neoplastic compression or infiltration of the esophageal myenteric plexus produced the pseudoachalasia." An aganglionosis, however, should be irreversible - degenerated nerve tissue does not un-degenerate. Regression of nodes or infiltrate obstructing the hiatus, on the other hand, relieves the obstruction. The patient is not wrong ("pseudo achalasia"); the theory is. Nevertheless, pseudo achalasia has persisted as a diagnostic entity.(99)

The case of Menin and Fisher(100) is even more foursquare. Their patient's achalasia (radiologically typical and manometrically "vigorous achalasia") was due to a 2 x 3 cm adenocarcinoma of the fundus extending 2 cm into the esophagus. It " . . . reversed clinically, radiologically and manometrically following surgical resection of the lesion." The reversal included return of a normal progressive peristaltic wave. Involvement of the myenteric plexus did not extend beyond 2 cm into the esophagus.

These tumors and other disorders (amyloidosis(101),(102) and post-vagotomy periesophagitis(103) can be added to the list) do not mimic achalasia. They are achalasia - indistinguishable radiologically, manometrically, pharmacologically and, usually, endoscopically from the more usual cause of hiatal obstruction of the gut.

The esophagus can also be obstructed by tightly wrapping the fundus about it as in a Nissen fundoplication. Such obstruction affected 9 of the 38 fundoplications requiring reoperation by Leonardi and Ellis.(104) They either manifested achalasia (7) or diffuse spasm (2).

Kenney, et al.(105) retrospectively studied five cases of secondary achalasia in their series of 357 admissions for achalasia. Computed tomography was 100% accurate in finding a tumor in these cases whereas none of 11 primary cases was diagnosed as secondary. The reason is if there is an obstruction by tumor, CT scans show it; if the obstruction is a strangulation, there is no abnormal tissue to be seen. The tubular fundus above the diaphragm looks like normal esophagus.

It is more difficult to come to grips with the "nonspecific esophageal motor disorder" concept that now envelopes the DES-achalasia spectrum because its very vagueness protects it. I suppose it to mean that in some fashion, the central or peripheral program controlling the end organ has developed defects that destroy coordinated muscular function. That 4 mg of vincristine, stretching the hiatus or a surgical procedure could reprogram the controller is difficult to conceive.

What all treatments have in common is that they relieve an obstruction. When they do so, the "disordered motor function," that is not disordered at all but merely the normal reaction of gut trying to force its contents through an obstruction - improves as the esophagus reverts to normal function just as would the small bowel after lysis of an adhesion.

The "bird beak" of achalasia has no resemblance to the sphincter.

The radiologic appearance of achalasia is not that of the physiologic sphincter. The typical beak-like termination of achalasia bears no resemblance to the LES. It is not only too aboral, but is also too long - 3 cm or even 4 cm - whereas the LES is scarcely over 1 cm in length.(106) The characteristic feature of the "beak" - its biconcave outline - is due to the torus of mesentery in the phrenoesophageal tent that surrounds and constricts the herniated fundus. Also typical of a squeeze effect is the "tramline" or "twin track" appearance due to barium on either side of an enlarged mucosal fold. It is virtually identical with the tramlines due to the mucosal squeeze of hypertrophic pylorus stenosis.

There is a further proof. We have seen that the Valsalva maneuver collapses that portion of the gut within the tented PEL. This maneuver will also change the contours of the "bird beak" of achalasia making it longer and narrowing or even obliterating the small lumen entirely, producing the "empty segment" appearance. Exaggeration of the squeeze when more omentum is forced into the tent shows what the squeeze is due to in the first instance.

Denervation is a result, not a cause of achalasia.

Under the conditions prevailing in advanced achalasia, many muscle cells are effectively denervated. Stretching neurons and their processes in two directions to many times their normal length may exceed their elastic limit or even avulse them from motor end organs. The terminal varicosities of the axons protrude through ultramicroscopic holes in an enveloping glial sheath to deposit their neurotransmitter in the connective tissue near the smooth muscle cells.(107) Overdistending the esophagus should damage these contacts.

The peristaltic gap

In deglutition, LMC normally tents the distal esophagus and adjacent fundus so that the bolus drops into the stomach when the sphincter opens. When LMC elevates a tubular segment of fundus above the hiatus producing a hiatal squeeze, it causes a fundamental disorder of propulsion. The esophagus cannot hand off the bolus to the stomach in the normal way. As we learned when analyzing the captive bolus, esophageal peristalsis stops at the sphincter.

Despite its resemblance to esophagus, the tubular fundus is incapable of peristalsis. The pacemaker controlling gastric peristalsis(108) virtually ignores the fundus. As a result the bolus cannot be propelled from the sphincter to or through the "herniated" stomach.

An aperistaltic segment forms as surely as if the bowel had infarcted or been stripped of its ganglion cells. As elsewhere, intestinal obstruction is the result of such a peristaltic gap. So, in this sense, the appearance is the same as though it were a true aganglionosis. It is a failure of peristalsis, not an aganglionic segment, that produces the characteristic signs of obstruction. Aperistalsis produces an "obstruction" even without an occlusion - thus accounting for otherwise typical cases of "achalasia" with complete sphincter relaxation.(109)

This situation is unique in the alimentary tract. It may account for the ease with which achalasia has long masqueraded as an aganglionosis. It also explains why the compensatory work hypertrophy that results from hiatal obstruction stops at the sphincter well above the actual obstruction at the diaphragm.

As a result, there are two reasons for obstruction: the hiatal squeeze and the aperistaltic segment. In practice, the hiatal squeeze need not even be very tight: the peristaltic gap alone is enough to account for the appearances.

Strictly speaking, the sphincter has nothing do with the case. Its tone and its ability to relax are unimpaired. They do not enter the equation as a third cause of obstruction. In that sense the emphasis placed on sphincter tone by conventional wisdom is misplaced. In another sense, however, the sphincter is all-important.

Sphincter latching

It will be recalled from the description of the captive bolus phenomenon that the advancing p-wave "latches" the LM. The more distal the wave, the shorter the esophagus. From this it was concluded that the LM, like the CM, is activated by the p-wave passing through it but, unlike the CM, the LM does not relax in the wake of the p-wave.

Although it contracts incrementally, the LM relaxes all at once. Progression of the p-wave into the sphincter is the signal for LM relaxation. Then roles are reversed. Now it is the CM of the sphincter that does not relax in the wake of the p-wave. The p-wave has stopped. Having fulfilled its function of opening the sphincter in the earlier stages of deglutition, the LM can now relax. The antireflux role of the advancing p-wave transfers to the sphincter that now latches in the closed position.

The sphincter requires no assistance in closing or in staying closed. Even after chemical denervation, its basal tone persists. Because it is mechanically impossible for a sphincter to open itself, the LM is needed to overcome the basal sphincter tone.

The unphysiologic conditions of fundic strangulation interfere with the ability of the sphincter to latch. In DES, the early stage of the disease, one sees very forceful p-waves that reach the lower esophageal sphincter only to lose their grip on the bolus. The bolus refluxes into the body of the esophagus stimulating another secondary p-wave. This would not happen if the sphincter were latching.

This failure may be due to increased pressure below the sphincter. Normally, the infrasphincter region is exposed to intragastric pressure at the instant of sphincter latching. In fundic strangulation, this pressure - the full pressure generated by the p-wave - is much greater. Mittal and his associates(110) have shown that esophageal clearance of refluxed acid is not a normal stepwise increase in pH if the subject has a HH. Clearance then becomes biphasic because each swallow induces a new episode of reflux. This would not happen if the sphincter stayed latched until the next p-wave formed.

A summary of the evidence

At this point, it would be well to summarize the major items of evidence that hiatal obstruction is the sole cause of diffuse esophageal spasm and achalasia.

Hiatal occlusion by tumor or fibrosis produces a radiologic, endoscopic, pharmacologic and manometric picture indistinguishable from DES or achalasia.

Removing a tumor obstructing the hiatus by surgery or chemotherapy causes regression of EMDs and achalasia.

The frequency of the sequelae of hiatal strangulation - "pseudotumor of the fundus" and "inflammatory gastro-esophageal polyp" shows that strangulation is a common event.

In many patients the hiatus is not easily distended but is just adequate to allow passage of the esophagus .

Remission of DES occurs after surgical reduction of a strangulated hiatus hernia as in the two cases described here. Boerema reports other cases of achalasia relieved by simple gastropexy.

The high incidence of epiphrenic diverticula with HH, DES and achalasia proves distal mechanical obstruction in all three.

The fluoroscopic appearance of DES is that of acute mechanical obstruction of the bowel.

Hypertrophy of smooth muscle is characteristic of intestinal obstruction. An aganglonosis should cause atrophy.

Longitudinal studies show a transition from DES to achalasia.

The radiologic appearance of the distal esophagus is not that of a tight but otherwise normal sphincter. It is the appearance seen in the captive bolus test when the stomach is obstructed in the PEL tent by mesentery crowded into it.

No test will distinguish esophageal obstruction due to tumor from obstruction due to strangulation.

Mechanical dilitation may relieve the symptoms but dilates the hiatus, not the sphincter.

Whereas it is physiologically impossible for degeneration of a neuron network to cause hypertrophy of the muscle it supplies, work hypertrophy is a normal reaction to obstruction of the gut.

This evidence - most of it inconsistent with either the aganglionic hypothesis or the more nebulous EMD formulation - shows that acute and chronic hiatal obstruction, most commonly by fundic strangulation, are the respective causes of DES and achalasia.

The evolution of "diffuse muscular hypertrophy"

The failure of sphincter latching and the peristaltic gap are the keys to understanding why obstruction at the hiatus causes hypertrophy and/or dilatation of the proximal esophagus whereas benign strictures and carcinoma do not. This distinction has been a last refuge of the aganglionosis hypothesis.(111)

The evolution of the full-blown picture of achalasia from the appearance of esophageal strangulation can be reconstructed. The forceful peristalsis of DES may succeed in reducing the strangulation or it may not. In the former and most common case, achalasia does not occur but the engorged mucosa on reduction may present the appearance of a fundic gastritis or pseudotumor.(112) In the latter case, hyperperistalsis and compensatory hypertrophy of the circular muscle may still force the obstruction. This leads to the circular muscle thickening found by the surgeon at cardiomyotomy(113) or, in the extreme case, to the "diffuse muscular hypertrophy" described by Johnstone(114) and others(115),(116),(117),(118)

If one reads the description of these cases carefully and examines the illustrations, in those cases in which a determination can be made, a HH was present. Some are mentioned in the report, but some, although clearly shown in the illustrations, were not reported by the radiologist because the fundic transtract was the same diameter as the esophagus.

A tubular fundus is easily mistaken for esophagus even at thoracotomy. Why else would the operative report say " . . . the lower esophagus was greatly thickened except in the terminal 4 centimeters where it was normal." [Johnstone, Case I - also Sloper's Case 5] or "The cardia and distal 3 cm of the esophagus were normal but immediately above this segment the esophageal wall was thickened beyond 1 cm and felt like a sausage." [Johnstone, Case 3]. (Emphasis added.) Sloper's Case 4 and a case of Rake's also describe the same phenomenon.

The fundus and the esophagus distal to the sphincter do not undergo work hypertrophy because, being aperistaltic, they do negligible work. The p-wave stops at the sphincter. There is no conceivable reason hypertrophy would stop short of the sphincter. We conclude that, even in these surgically explored cases, the fundus, drawn through a small die-like hiatus, was mistaken for esophagus.(119)

In addition, the radiologic picture is in no way different from DES. The preponderance of the hypertrophy, as would be expected in work hypertrophy, is always in the circular muscle layer whenever the two layers are described separately. In Sloper's illustrations, instead of being somewhat thinner than the LM layer, the CM is over twice as thick.

The radiological and manometric signs of hyperperistalsis and en masse contractions, perforation in one case and an epiphrenic diverticulum in another complete the proof that all of Johnstone's examples of "idiopathic diffuse muscle hypertrophy" were due to normal work hypertrophy from hyperperistalsis secondary to strangulated HHs.(120) Again, assuming an "idiopathic" etiology is redundant when obstruction, the usual cause of muscular hypertrophy, is patently present.

A critical point in the evolution of achalasia occurs when the dilatation becomes sufficiently extreme and/or the circular muscle becomes sufficiently exhausted that the peristaltic wave can no longer approximate the esophageal walls sufficiently to obliterate the esophageal lumen.

End-stage exhaustion

Unless the lumen can be obliterated, peristalsis has no propulsive force. Somehow sensing the futility of peristalsis, the organ compensates by a pattern of en masse contraction that does have a propulsive force. Counterproductively, this is often dissipated by self buffering presenting as tertiary contractions. After that, dilatation and elongation are the only responses of the esophageal wall to the ingestion of food as the organ degrades to a passive conduit. Hydrostatic pressure of the fluid column, perhaps with some assist from a Valsalva maneuver, is the only force that can even partially empty the esophagus. This, of course, is the usual presenting appearance of achalasia. Except for the unique position of the esophagus as the intrathoracic gut, the appearance does not differ from chronic intestinal obstruction elsewhere in the alimentary tract.

Qualman et al.(121) after reviewing the pathologic literature noted that "The neuropathologic findings generally reported in [achalasia] include chronic inflammatory infiltrates within the esophageal myenteric plexus and degenerative changes within smooth muscle or nerve fibers." The hypersensitivity to cholinergic drugs is lost when the end organ fails. Ganglion cell degeneration is a result, not a cause of the obstruction.

One does occasionally see indentations of the outline of the decompensated esophagus as though the circular muscle were contracting erratically. These indentations, however, cannot be due to circular muscle contraction because they occur on only one side.

The contribution of manometry to the confusion

It now appears(122),(123) that, given the proper transducer, the term "cardiospasm," although also incorrect, need never have been replaced! Measurements with the old style non-perfused catheters showed no increase in LESP. However, measured with the newer perfused catheters,(124) LESP is twice the normal pressure. Measured with the still newer intracorporeal transducers LESP is sometimes normal.(125)

Paradoxically, Katz, Richter, Cowan and Castell(126) found that 30% of their patients with otherwise typical achalasia had complete relaxation of the sphincter. They conclude, "Apparent complete LES relaxation may be seen during manometry in achalasia and should not exclude its diagnosis." Anachalasic achalasia seems the final reductio ad absurdum of the aganglionic hypothesis. The detailed mechanics of the disorder, as I have shown, make these inconsistent measurements of LESP/hiatal squeeze understandable.

Although manometry was a technical refinement, it started and perpetuated the confusion. Despite the changing physics of pressure sensing equipment that caused embarrassing reversals when absolute pressure measurements were involved, it is beyond doubt that relative pressure measurements and wave patterns are meaningful. However, in DES and achalasia, these patterns do not represent a spectrum of intrinsic neuromuscular disorders, they are the normal motor response of a gut trying to overcome an obstruction that is in part mechanical and in part interrupted peristalsis, Initially, this response is unusually forceful and unusually frequent peristalsis. Later en masse contractions occur. In the end, hypertrophy or exhaustion and decompensation occur.

The hiatal ring can be very resistant to expansion.

Thus far, I have tacitly assumed that the hiatus has enough intrinsic strength that it will not easily stretch to accommodate a herniated fundus. The diaphragm is a thin structure. It contains considerable muscle, the fibers of which are easily stretched. Even if the fundus were retracted into the hiatus, it could conceivably act as its own dilator, expanding the hiatus and so forestalling strangulation or entrapment. It is natural to ask, "Does the hiatal ring have enough strength to be a persistent strangulating constriction?"

Unless one is a surgeon, he has no opportunity to explore the hiatus personally to see how well it can resist stretching. Fortunately, we have exact information about the resistance of the hiatal ring: the current treatment for EMDs is balloon dilatation. Up to 740 mm Hg of pressure is applied to the hiatus (under the impression it is the sphincter) and even this may not be enough to dilate it adequately. Comparing this with the 150 mm Hg used to inflate a blood pressure cuff gives an idea of the force generated by that pressure. In some cases even this force is insufficient to produce enough hiatal dilatation to relieve the

symptoms and the procedure may have to be repeated three or four times to stretch the "sphincter" to a diameter of as much as 5 cm.(127)

Why haven't surgeons found the strangulation?

The wonder is that a strangulation or hiatus hernia is not recognized at the time of the operation. The skeptic is thinking, "Logic is well and good, but surgeons can see what is going on. They have been looking at these cases for a hundred years. They can't all be missing it."

In my original case, the surgeon who performed a Heller procedure was not alerted what to expect and did the operation without comment and without reducing a HH. The second surgeon, with whom the rationale of the patient's problem had been discussed, did see the hiatus hernia, reduced it and cured the patient although in repairing the blowout he was undoing the myotomy. We have seen that in Johnstone's cases the only explanation of hypertrophy stopping short of the diaphragm was a tubular HH not appreciated by the operators.

A Heller procedure is usually done in an advanced stage of the disease. It appears from illustrations that there is a marked hypertrophy of the circular muscle. It may have taken years to develop this "musclebound" condition that it can be constricting. The original strangulation may have long subsided.

Good results are reported no matter what operation is done. This may not be mere surgical self-congratulation. By the time the area of interest is exposed, landmarks identified, etc., the strangulation may be inadvertently reduced. If the procedure is done by the abdominal approach, and this is preferred by many,(128) any HH present will be reduced when pulling on the stomach to get at the esophagus.

Done by the transthoracic approach, a partial fundoplication (that by necessity gets the fundus out of the hiatus thus relieving the strangulation) is regarded as one of the three principles necessary for success.(129) In the case of the Boerema "anterior gastropexy" of course, the good results are due to pulling the stomach down forcefully and so reducing the HH.

Typically the surgeon finds what he expects to find - a thickened segment of the esophagus similar to a hypertrophic pylorus. Doing a myotomy on this hypertrophied muscle, could be effective treatment accidentally. Any incarcerated fundus will be released when the incision is " . . . carried down into the circular fibers of the cardia." which would be impossible without either dividing the hiatal ring or recognizing that the fundus was already above the hiatus.

Review of a large number of surgical reports turns up the reassuring fact that often the hiatus hernia is not missed at surgery. Most Heller procedures are "modified" and the modification almost invariably involves repairing a HH

unsuspected preoperatively or adding an antireflux procedure to forestall a frequent complication of the Heller procedure.(130),(131),(132) This may include extending the myotomy into the stomach. Despite their use of a transthoracic approach, Ferguson and Burford, for example,(133) recommended opening the diaphragm to detect HH after their experience of finding three HHs(134) unsuspected radiographically and at esophagoscopy and three cases of adenocarcinoma of the fundus that produced " . . . symptoms, esophagoscopic, and x-ray changes indistinguishable from achalasia." Significantly, they also report that most of their unsatisfactory results were due to hiatus hernias either missed at the time of myotomy or possibly caused by " . . . surgical manipulations about the GE junction."

Ellis et al.,(135) for example, report that 16% of their cases had already had an esophagomyotomy and another 20% had HH or a "lax hiatus" at the time of exploration. Several others had actually had a HH repair when originally operated for achalasia!

The actual situation at the time of the procedure may not be as straightforward as the neat anatomical drawings suggest. There may be an additional 20% of strangulations missed as that is about the incidence of unsatisfactory results with surgery. Nissen, Belsey, Collis and Boerema procedures may be done concurrently with the myotomy.(136)

Trounce et al.(137) found that when the narrowed segment as determined by cine-radiography [i.e., a tubular fundic transtract] was examined at operation, " . . . its muscular walls appear quite normal, in notable contrast with the hypertrophied and dilated esophagus above." Johnstone's cases 1 and 3 were similar suggesting these observers mistook a tubular section of stomach for esophagus. Peristalsis ends at the sphincter and so does hypertrophy. These are highly experienced experts on the area and they all saw the hiatal transtract and commented on it in their articles but had no reason to believe was anything but a segment of normal esophagus. The resemblance to esophagus must be striking, indeed.

It is also possible that the diagnosis may be overlooked at surgery because it takes very little tissue to produce obstruction in a small hiatus. It will be recalled that Harrington found that 55% of patients have a hiatus too small to admit one finger. When a tag of fundus or fat further occludes it, it may not be obvious to a surgeon whose attention is directed primarily, not to the hiatus, but to the organ he is operating upon. The truism that we only see what we are looking for probably holds true for surgeons as well as others.

Therapeutic implications

Boerema(138) fortuitously discovered that a pulldown procedure intended to correct a HH alleviated cardiospasm. It would be better to do the procedure in the

early stages of strangulation instead of after the organ decompensates. Fundoplication and "snugging" the hiatus during operations for hiatus hernia or "restructuring the cardia" are irrational and should be abandoned. If anything, the hiatus should be enlarged so that a recurrence will not aggravate the original difficulty.

The Heller muscle-splitting procedure seems to make no sense at all. No one would dream of treating compensatory hypertrophy of the small bowel proximal to an adhesion in this fashion.

SUMMARY

Although solidly entrenched for over sixty years, the aganglionic theory of achalasia topples in a welter of contradictions, unverified consequences and unjustified assumptions. The notion that a neurologic deficit can explain muscle hypertrophy is nonsense.

Like most esophageal disorders, both diffuse esophageal spasm and achalasia can be traced to abnormal function of the longitudinal muscle. A forceful contraction of the LM, such as that occurring with pyrosis, nausea, gagging or vomiting, produces a "hiatus hernia" in which the fundus, epiphrenic fat, lesser omentum and the left gastric artery or its branches are drawn up through a small hiatus where they become incarcerated.

The incarceration may subsequently reduce spontaneously, in which case there will be a "fundic gastritis" or a "pseudotumor of the cardia" caused by the vascular engorgement of the formerly incarcerated fundus. If it does not reduce, the appearance of "diffuse spasm" results. This is an unrecognized high-grade intestinal obstruction. The hiatus, while adequate to allow the passage of the esophagus, is too small to contain the fundus and its attachments. Instead of stretching to accommodate the retracted fundus, it constricts and obstructs the lumen.

The radiologic appearance described in "diffuse spasm" is no different from that seen in small bowel obstruction. Rapid, forceful, churning peristalsis and en masse (tertiary) contractions, that, nevertheless, do not succeed in clearing the lumen of its contents are typical.

Both the "hiatal squeeze" and the presence of an aperistaltic segment of gut above the diaphragm prevent esophageal emptying. This tubular segment of fundus is usually mistaken for esophagus because of the LM tension. Eventually obstruction results in hypertrophy (diffuse muscular hypertrophy) or in dilatation, decompensation and elongation of the esophagus.

The diffuse spasm phase may persist for many years. If it does, a blowout of a weak area in the distal esophagus is likely. This is the epiphrenic diverticulum - a buffer for the bolus.

Eventually, the circular muscle decompensates and dilates. The LM elongates. The typical dilated, sigmoid esophagus of "achalasia" does not differ in any significant way from decompensated bowel proximal to an obstruction elsewhere in the gut.

Dilatation per se spreads out the ganglion cells like spots on the surface of an expanding balloon so that there are fewer seen per microscopic field. Microscopically, this gives the impression of an aganglionosis. As distention thins the bowel wall it separates the neurons from their end organs, denervating many of them. The stagnant contents distending the esophageal wall causes first the neurons and eventually smooth muscle cells to degenerate.

Invasion of the hiatus by tumor or retroperitoneal fibrosis obstructs it causing "secondary" achalasia. Extirpation of a tumor or chemotherapeutic lysis reverses typical radiologic and manometric signs of achalasia. Reducing a HH has the same effect in "primary" achalasia. None of these successful forms of treatment are directed at either cure of an aganglionosis or "reordering" a disordered motor function. None of these treatments could conceivably induce regeneration of lost neurons.

EMDs are not esophageal motor disorders. The manometric and radiographic "abnormalities" are normal motor responses to obstruction.

The implications for treatment are obvious. The strangulation should be reduced as soon as the picture of "diffuse spasm" presents. The pulldown operation in its pure form as employed by Boerema (no fundoplication) is a rational way to treat the condition. Even if supplemented by enlargment of the hiatus to prevent strangulation, this should be simpler and safer than the more drastic means now employed.

"Forceful dilatation" with a bag placed at the hiatal level is effective because it stretches the hiatus, not the sphincter, but can result in rupture and mediastinitis. Many other operations with various rationales are effective because exposing the area of interest reduces the strangulation. If normal peristalsis in not restored, the open operation should be considered. Laparoscopic surgery could well prove the best and safest treatment when a procedure is devised.

The Allison procedure - at least as Allison described it(139) - reduces the size of the hiatus with "non-strangulating sutures." It shares with fundoplication the potential for insuring obstruction should the original mechanics cause a recurrence. It is more rational to reduce the strangulated HH and find means to prevent strangulation if it recurs.

A formidable body of medical research has grown to encrust the aganglionosis theory of achalasia. It is now being revised to fit the theory that DES and achalasia are a spectrum of primary muscle disorders. While not without merit, this work is basically ad hoc, its value dissipated by interpretation in the light of incorrect hypotheses. Diffuse spasm, idiopathic diffuse muscular hypertrophy, EMDs and achalasia are but manifestations of the same disorder - undiagnosed intestinal obstruction. I propose "fundic incarceration" as the name for all four.

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Mallory-Weiss syndrome

A cause of about 10% of upper gastrointestinal bleeding, the Mallory-Weiss syndrome, starts with retching or non-bloody vomiting followed by hematemesis. This pattern has always suggested that the initial emesis itself caused the bleeding. The occasional instance of the syndrome caused by endoscopy(1) confirms that etiology as the endoscopist observes intact mucosa on inserting the instrument, then retching, and subsequently sees the linear tear(s) as he withdraws it. They are believed to be due to ". . . a sudden dramatic increase in intraesophageal pressure."(2)

Knauer(3) observed 58 cases noting that 72% had HH's. There was a noteworthy radial asymmetry in the location of the tears with 52% occurring on the right vs. only 7% anteriorly. the only thing which distinguished Boerhaave's syndrome, from Mallory Weiss is the depth of the laceration. The Mallory-Weiss tear is superficial whereas the Boerhaave tear may rupture the wall. In both, barring Boerhaave's initial case in which the esophagus was completely avulsed from the stomach, the tears are parallel to the long axis of the esophagus.

They could not, as might be expected, be due to overdistention of the esophagus or herniated cardia by sudden ejection of gastric contents as they are seen after retching (i.e., LMC without emesis) and after endoscopy which, of course, is performed on an empty stomach. The wedge shape of the tears(4) observed after endoscopy induced retching is a further clue that the force is applied at the PEL. If overdistention caused them, they would tend to be eliptical. Like sphincter opening, these syndromes present the paradox of an axial force producing, not the expected transverse tear, but a longitudinal one.

It is, perhaps, puzzling that most of the tears (78%) occur in the stomach just below the mucosal junction. Two circumstances may account for this. 1.) 82% to 100% [Knauer] of the patients have hiatus hernias. The increased friability of the mucosa in the herniated portion of the stomach may account for this localization. 2.) LMC produces a trumpet-like flaring of the GE junction. The further down the trumpet, the more the mucosa is stretched. Thus the wide end of the wedge-shaped tear is aboral. It would be more characteristic of distention to cause a symetrical distribution of tears instead of that actually seen. The angle of insertion of the PEL on the esophagus - which is a factor in the force resolution - is radially asymmetrical so that the stretch is also radially asymetrical.

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The (dubious) Barrett esophagus

"Barrett esophagus" (BE), an esophagus lined with gastric mucosa, is a popular endoscopic diagnosis. Its incidence appears to be increasing rapidly as its lore proliferates. Both Winters et al.(1) and Schnell, et al.(2) have reported a 12.4% incidence of BE among adult patients with gastroesophageal reflux - a remarkable epidemic for a disease that may be non-existent. A reviewer(3) suggests that radiologists should also be learning to make the diagnosis.

Some will recall that the literature of the 1940s contained many reports of "congenitally short esophagus with intrathoracic stomach," an entity that vanished after it was pointed out that to prove such a diagnosis it would be necessary to show that the blood supply to a thoracic stomach originated in the thorax. Perhaps it is not coincidental that the advent of the Barrett esophagus (1950) was simultaneous with the demise of the "congenitally short esophagus." The two may be opposite sides of the same coin - complementary ways of misdiagnosing hiatus hernia. Proof that a tube of gut lined with gastric mucosa is esophagus would require demonstration that it is supplied by esophageal, not gastric, blood vessels. Thus far, I know of no case in which this criterion is satisfied.

The existence of BE is solidly based on assertion. As Levine states, ". . . it was postulated . . . ". It is hard to account for the extraordinary attractiveness of this conjecture. It seems to have gained universal, if uncritical, acceptance with the lone exception (albeit temporary) of Barrett himself.

Radiologists seldom make the diagnosis of BE (all they see is a tubular hiatus hernia).(4) Endoscopy, although presumed to be the gold standard in the diagnosis of BE, is a fallible method. The presence of gastric mucosa closer to the incisors than normal does not establish the diagnosis. It merely proves that the esophagus is shorter than normal, as it is in many hiatus hernias. Experimental esophagitis by acid perfusion causes esophageal shortening in the opossum.(5),(6)There is evidence that the same is true in human esophagitis,(7) a condition that is invariably present when BE is reported.

To establish that he is biopsying esophageal mucosa, the endoscopist must first determine internally where the esophagogastric junction is located. There are two landmarks: 1.) The sphincter and 2.) The squamo-columnar junction. The first, however is a manometric(8), not an endoscopic landmark(9) and the second is what he must postulate is misplaced. Spechler and Goyal, who have written extensively on the subject, state, ". . . one cannot determine with certainty where the esophagus ends and the stomach begins." If this is the case, how can an endoscopic diagnosis of BE be made? In practice, there is little doubt that the diagnosis is based on the distance from the incisors at which gastric mucosa is encountered.

In the Veterans Affairs Cooperative Study of BE(10), 20% of 93 patients had the diagnosis on only one of two examinations made six weeks apart. The endoscopically determined LES was 3 cm proximal to that determined by manometry. One criterion for diagnosis was the presence of 3 or more cm of specialized columnar epithelium above the manometrically determined LES. The other was biopsy of "specialized columnar epithelium" (SCE) in "tubular esophagus."

Nine percent of the patients had a difference of 4 cm or more in the proximal level of SCE [i.e. intestinal metaplasia] between examinations. 18% of 192 patients had the diagnosis of BE reversed within 6 weeks. In 81 patients a diagnosis of esophagitis was changed to BE; of 82 initially classified as BE with SCE, 11 had the diagnoses changed to esophagitis; 5 of 29 patients initially classified as BE with columnar epithelium had their diagnosis changed to esophagitis.

The outliers in these statistics are most significant. On a second examination, the most proximal level of Barret's epithelium changed from as much as 7 cm lower to 8 cm higher in patients who had not had surgery in the interim. Kim et al. concluded ". . . approximately 10% of patients had a change 4 cm on endoscopy and manometry between examinations. This led to an apparent change in diagnosis in 18% of patients with Barrett's esophagus."

It is clearly impossible that 4 cm or more of "metaplastic" gastric mucosa could revert to squamous mucosa in 6 weeks; on the other hand, it is certain that the amount of stomach above the diaphragm will vary, not only from one examination to the next but from moment to moment in the same examination. Greater or lesser inflation of the esophagus will produce more or less LMC. Gastric mucosa above the hiatus is a hiatal transtract - not metaplastic squamous epithelium.

The BE population is a subset of the esophagitis-GER population. Statistically, it could be expected that most of the patients in the VA study would have hiatus hernias. As LM tension both opens the sphincter (producing reflux) and stretches the PEL (producing hiatal transtraction), the two are inseparable. Remarkably, none of the 116 patients identified as having both severe GER disease and BE were reported to have HHs!

A Medline search of the 1990 to date database yielded 205 abstracts for the keywords ("hernia" and ("hiatus" or 'hiatal")), 350 for "Barrett" and 11 for the intersection of the two. Of the 11 several were miscodes. Several were not actual case reports, some were didactic. One of the latter baldly stated that 75% of BEs had HHs. So we have here the same Venn diagram as with achalasia-hiatus hernia, forcing the same conclusion: the two do not occur together because they are the same thing - now diagnosed one way, now the other.

Biopsy "proof" of BE is unconvincing for two reasons: 1.) The pathologist can only describe the mucosa. The muscular layers - although even these would not be unequivocal - are not included in the specimen. To make the diagnosis of BE from a biopsy pathologists must rely on supporting information from the endoscopist. That information is usually the distance below the incisors at which the biopsy was taken or the distance above the manometrically determined sphincter. The endoscopist becomes a self-fulfilling prophet. 2.) The pathologist must be pre- indoctrinated that intestinal metaplasia of gastric mucosa is metaplastic stratified squamous epithelium.

Radiologically, the diagnosis is made when transition from normal or inflamed mucosa is seen below a "stricture." Many of these presumed strictures are due to the less distensible sphincter area when seen in air contrast esophagograms.(11),(12) The gastric mucosa looks like gastric mucosa showing an abrupt change in fold size below the stricture/sphincter as it should.

Even at autopsy a pathologist would have difficulty determining whether a supradiaphragmatic tube of gut lined with gastric mucosa is esophagus or a tubular hiatus hernia. The blood supply is destroyed by the usual Rokatansky autopsy technique that transects the viscera at the diaphragm before removal.

The unproven assumption on which BE rests rivals the audacity of the achalasia assumption that a loss of motor neurons will cause a muscle to hypertrophy. The postulated metaplasia from squamous to highly specialized columnar epithelium(13) is a false analogy - backward in fact. Whether it is the lung, the cervix, the endometrium, the gallbladder, the pancreas, the urinary tract or the bile ducts, metaplasia replaces a specialized glandular, columnar epithelium with less specialized epithelium. Usually this is stratified squamous epithelium although gastric mucosa may convert to the less specialized intestinal mucosa as indeed it does in cases claimed to be BE. I have been unable to find reports of reverse metaplasia elsewhere in the GI tract or in any other organ.(14) The burden of proof of BE, therefore, rests on those who postulate that, in the esophagus, it is the other way about. One might expect to encounter islands of gastric mucosa on the tongue or in the labial fissures if this were a possibility.

Despite study of hundreds, perhaps thousands, of cases no one has offered a clue to explain how squamous epithelium can acquire specialized potentials - so specialized in fact that, like the stomach and Meckel's diverticulum, it picks up technetium pertechnetate.(15) Kweka et al. report that all 8 of their histologically verified cases of BA were imaged with this isotope. Isotope imaging merely proves that the stomach above the diaphragm has not lost its ability to take up the isotope. It is wildly improbable that squamous epithelium should acquire this highly specialized ability - an ability we rely on to identify gastric mucosa formed in the embryo.

A transition from squamous to specialized gastric mucosa would be differentiation in the technical sense. Tissues undergoing neoplastic transition - as it is claimed to be true in BE - dedifferentiate. It is difficult to understand how these contradictory concepts can be maintained in separate watertight compartments.

Biopsy of supposed cases of BE tends to refute the diagnosis. The histology is also more in keeping with a tubular transtract than metaplasia. In addition to the normal squamous lining of the organ, three types of mucosa are encountered. Again, according to Spechler and Goyal, these occur in precisely the following order from above to below:

A." Specialized mucosa." This is still recognizable as gastric mucosa but distorted so as to be similar to intestinal metaplasia of gastric mucosa. It is metaplastic, but metaplastic gastric, not esophageal, mucosal. That is, the gastric mucosa is transformed in the usual way of metaplasia in the direction of the less specialized intestinal mucosa.

B. "Junctional" mucosa. This is another name for the normal mucosa of the gastric cardia.

C. "Fundic" This is the normal or somewhat atrophic mucosa of the gastric fundus.

Only the first of these would be considered abnormal. The three mucosal types could be found consistently in some other order or in random combinations of the 6 possible sequences in different patients. But this does not happen. What is actually encountered is the sequence to be expected when the stomach is drawn upward into a tubular HH by esophageal transtraction, i.e.

1.) Gastric mucosa histologically altered by ischemia due to constriction of its blood supply in a hiatus designed to contain esophagus, not stomach, 2.) Gastric cardiac mucosa, 3.) fundic mucosa.

There is a reason for the rising incidence of BE - the increasing use of air contrast examinations. Christensen and Lund have demonstrated that inflating the opossum esophagus causes reflex contraction of the LM(16) and this is certainly the case in man. When inflation is done at esophagoscopy, it will pull a HH through the hiatus and render a saccular HH tubular. The same is true radiologically if the examiner does an air esophagogram. The popularity of these examinations in recent years probably accounts for the current epidemic of BE. The recent reported cases are invariably illustrated in air contrast.(17)'(18),(19),(20),(21) Some of these show the "distal stricture" that is actually the less distensable sphincter area.

Lower esophageal rings are encountered with great frequency. Johnson et al.(22) found them in 15-18% of 22,368 patients undergoing upper GI fluoroscopy. It is widely accepted that they occur at the junction of gastric and esophageal mucosa. If gastric epithelium grew orad into the esophagus, it would have to coat the LER. If any squamous epithelium at all was transformed into columnar mucosa, the mucosa of the ring would be involved. To my knowledge, no cases have been reported in which a LER was located within or below a region of Barrett mucosa.

The demarcation line between squamous esophageal mucosa and columnar gastric mucosa is sharp - at least as sharp as the ora serrata. Yet the entire esophagus is exposed to acid pepsin in patients with reflux. What additional postulate must be made to account for the sharp demarcation of BE? The interdigitations of squamous and gastric mucosa which seem so convincing are simply what one would expect at the ora serrata. The "dribbles" of squamous mucosa on to the inferior surface of a LER when flattened out would appear to be interdigitations.

Ectopic gastric mucosa in the upper esophagus is not uncommon, occurring in about 10% of the population,(23) usually at the level of the thoracic inlet. It is generally agreed to be heterotopic. I have not found any suggestion that it might be metaplastic. Endoscopically and radiographically it bears no resemblance to BE. It presents as shallow saucer-like depressions with slightly raised margins, not as cylinders of gastric mucosa. Unlike BE, it is surrounded on all sides by squamous mucosa.

In my experience, the incidence of tubular hiatus hernias approximates the reported incidence of BE. The appearance of a tube of stomach drawn through the "die" of a small hiatus by esophageal shortening is identical with published radiographs. The esophagus can easily shorten one third its length, retracting a long tube of stomach through the hiatus whereapon the less distensable sphincter will appear to be a "smooth stricture." These HHs may be persistent or they may reduce. Science(24) quotes the author of a 10-year study of BE as amazed that occasionally a BE spontaneously reverts to normal. "It's the strangest thing we've ever seen . . . .", he said. It would be strange indeed if a reverse metaplasia again reversed. Not so strange if a HH reduced.

Although proving the diagnosis of BE is difficult, disproving it is easy: esophageal peristalsis stops at the sphincter - there is no peristalsis in the gastric fundus. Therefore, if peristalsis stops on reaching a tube of gut lined with gastric mucosa, one can be certain the wave has encountered stomach lined with gastric mucosa - not esophagus. Unfortunately, this test cannot be performed endoscopically or with air esophagograms.

That said, it must be admitted that there is something unusual and significant about such tubular HHs beside their shape. The persistent shortening of the

esophagus with its attendant reflux and the small hiatus that molds them and constricts their blood supply without strangulating the stomach deserves separate classification and analysis. If the tube of stomach is constantly above the diaphragm, as may well be the case, the LM must be constantly shortened.

I have not attempted to examine the claim that the incidence of carcinoma is greatly increased in such cases although this assertion deserves critical study in view of the wide disparity in reported incidence. A Collis procedure, in which a tube of stomach is formed into an artificial esophageal extension, duplicates most of the characteristics of the postulated BE. It would be worth studying a large series of such cases for the incidence of carcinoma.

Nothing is ever simple. There is some theoretical possibility that a congenitally short esophagus may never have developed a squamous epithelial lining during embryogenesis. Certainly, however, this would be a great rarity and not an affliction of 12% of the GER population.

SUMMARY

Longitudinal shortening of the esophagus can be due to vagal stimulation, to hormonal influences, and to the direct effect of acid pH on the esophageal mucosa. It shortens reflexly from inflation of the organ at radiologic or endoscopic examination. All of these factors are at work in patients with supposed BE. They cause a tubular hiatal transtract that is mistaken by examiners for esophagus lined with gastric mucosa. The supposed metaplasia can vary up to 9 cm over a few weeks or vanish entirely as more or less stomach is pulled through the diaphragm. Metaplasia from unspecialized to more specialized tissue is unknown elsewhere and is not likely here where there is a perfectly reasonable explanation for the appearances. Microscopic diagnosis is illusory as it depends on the distance of the biopsy from the incisors or from a manometrically localized "sphincter" which may be a hiatal squeeze. The epithelium is indistinguishable from intestinal metaplasia of gastric mucosa - which indeed it is as demonstrated by its ability to take up technitium pertechnetate.

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Summing up

The longitudinal muscle is both the leading character of this book and the villain of the piece. It would seem that, on the whole, we might be better off without it. There are no maladies (other than livestock bloat) that can be blamed on a non-functional LM. It is, however, necessary for belching, vomiting and gagging and provides an assist in swallowing, particularly with solid food. Vector resolution of its force by the PEL opens the sphincter. A solitary hiccup is the mechanical equivalent of a sharp contraction of the LM and has the same sphincter-opening effect. Taking the LM into consideration doubles the number of esophageal muscle states, vastly increasing its repertory of sequential operations.

On the debit side, LM hyper function leads to reflux and hiatal transtraction of the fundus. The latter can lead to chronic blood loss, anaemia and achlorhydria as well as rupture of the phrenoesophageal ligament. Reflux of acid/pepsin damages the mucosa of the esophagus, hypopharynx and tongue. It can destroy the teeth and cause angular stomatitis. Together these mechanisms produce all the features of Plummer-Vinson syndrome.

An incarcerated transtraction causes obstruction and leads to "achalasia," EMDs and "idiopathic diffuse muscular hypertrophy." At a minimum LM tension produces the "gas/bloat" symptom in adults - colic in babies.

Longitudinal muscle tension stresses its proximal attachments as much as those to the diaphragm. This results in Zenker's diverticula and/or the hypopharyngeal disruption that masquerades as "cricopharyngeal spasm."

There are serious doubts about the validity of the postulated metaplasia of Barrett's esophagus. The appearances can be better explained by esophageal shortening.

Two questions remain to be pursued: (1.) What makes the LM hyper-function and (2.) What can be done about it.

On the first question I have some clues. One of them is the dramatic postpartum relief of the heartburn, nausea and vomiting of pregnancy - not due to pressure of the gravid uterus on the diaphragm as is usually supposed. Progesterone is surely the cause as it also produces pyrosis in patients on birth control pills. A search of the literature, unfortunately, reveals no studies of the effect of progesterone on the LM.

There are also hormones of intestinal origin that could affect the LM, particularly CCK and secretin. Fats entering the duodenum trigger CCK secretion triggering smooth muscle contraction in the GB to inject emulsifying bile into the duodenum. The systemic release of CCK may have the side effect of stimulating

the LM. This would explain the paradox of soothing oils causing esophageal irritation; why the third helping of turkey dressing causes heartburn.

Patients with pyrosis are often greatly improved by cholecystectomy. The connection warrants looking into. The gallbladder requires CCK to contract. How does make its requirements known to the duodenum? Does it have a neuronal path or does it produce a duodenum stimulating hormone (DSH)? If so a diseased GB could stimulate overproduction of CCK or another intestinal hormone thus overstimulating the LM, opening the sphincter, and so on. Removal of the GB might break this chain.

A related clue is the fact that nearly everyone with reflux has duodenitis. My subjective statistic is 80-90%. We know that the duodenum is the source of intestinal hormones. Are duodenal hormones produced in excess when the duodenal mucosa is inflamed - in the same way that H. pylori infections of the antrum cause hypergastrinemia? If so, then H. pylori may be a culprit. Its treatment is becoming well known and better drugs are around the corner.

The problem of treatment, therefore, seems more endocrinological than surgical. Unfortunately nothing is known about the effects of these hormones on the LM. Investigators have been concentrating their effects on the sphincter.

I believe the rationale of most operations on either end of the esophagus is wrong. It's not the mythical angle of His or a subphrenic esophagus that inhibits reflux. It is the sphincter. The mechanics of vector resolution are such that anything that destroys the PEL destroys one of the two things involved in opening the sphincter. Many operations succeed because they fail, inadvertently destroying the PEL in the process. A direct attack on the PEL might be indicated. There are worse things than a type III "hernia."

The treatment of "achalasia," EMDs and "idiopathic diffuse hypertrophy" should be reduction of the incarcerated fundus and enlargement of the hiatus. This might even be done endoscopically by incising the hiatal ring. Stretching the hiatus helps but the effect may not be permanent. Muscle splitting is irrational and can cause epiphrenic diverticula.

The technical aspects of reconstructing the proximal attachments of the esophagus would be formidable but would make more sense than doing further damage by resecting a "bar."

Appendix A

When a hollow organ is distended, its thin mucosal layer has a maximum circumference. The cross sectional area of the lumen is also maximal. When the organ contracts, the area of the lumen decreases to zero. The circumference, however, cannot approach zero as the elasticity of the mucosa is not infinite. We can get a numerical handle on these circumstances by noting that the area of the mucosal layer remains constant despite distention or collapse of the lumen.

If one assumes that as much mucosa is squeezed into a given cross section as is squeezed out, then the cross sectional area of the mucosal layer will be the same before and after contraction of the gut. Before contraction (Figure 1) the mucosal area is the difference between the areas of the inner and outer circles:

$A_1 = \pi r_1^2 - \pi(r_1 - t_1)^2$	After contraction (Figure 2) the mucosal area is . . .
As the mucosal area is unchanged by contraction, these expressions can be set equal to each other and solved for r2 resulting in the expression . . .	$A_2 = \pi r_2^2$
$r_2 = \sqrt{2r_1t_1 - t_1^2}$	The circumference of the free margin of the mucosa after contraction, C2, is a fraction of its pre-contraction length. The decimal fraction C2/C1 is equal to E, the elasticity of the mucosal tissue. It is the elasticity which determines how much the mucosa can shrink. The fully contracted circumference, in leiu of shrinking to zero, must form folds.

From Figure 3, the mucosal length taken up by one fold is twice the distance from the center of the organ to the depth of the interfold valley or . . .

An expression for Nf, the expected number of post-contraction folds, can now be written.	$2(r_2 - t_2)$
$N_f = \frac{C_2}{2(r_2 - t_2)}$	The dimensions are all post-contraction, however, from the above, we know that C2 = C1E and , from Figure 1, C1 = 2(r1-t1). Moreover, r2 was calculated in equation 3. We can also calculate that t2 C2 = t1C1 or
Performing the indicatedsubstitutions yields a formula for fold numbers in	

terms of the inital values of thickness, organ radius and mucosal elasticity. The subscripts, therefore, can be dropped giving . . .	$t_2 = t_1 \frac{C_1}{C_2} = \frac{t_1}{E}$

$$N_f = \frac{\pi E (r-t)}{(-\frac{t}{E}) + \sqrt{2rt-t^2}}$$

Appendix B

[Please refer to hard copy for diagrams]

If O is a point is space representing the sphincter, a force F, representing the tension applied to it by LM contraction, will be opposed in the plane by the phrenoesophageal ligaments represented by the two vectors b1 and b2 . The projections of b1 and b2 on the vertical are a1 and a2. The vertical components are both in the opposite direction to F and counteract its tendency to elevate point O. In the same way, c1 and c2 are the projections of b1 and b2 in the horizontal direction.

From the geometry we can write:

a1 = b1 cos a2 = b2 cos

c1 = b1 sin c2 = b2 sin

a1 = a2

c1 = -c2

The sum of the a1 and a2 vectors will prevent upward translation of point O. The effect of the c1 and c2 vectors, which are of opposite sign, will be to pull point O in opposite directions.

If O, instead of being a point, is a minute annulus representing the inner surface of a closed sphincter, the effect of the c vectors will be to separate the opposite walls. If the whole diagram is rotated about the vertical axis distributing these vectors in 3 dimensions, all of the periphery of the closed sphincter will be spread open without any lateral translation of the sphincter itself.

The detailed distribution is extremely difficult to model mathematically because the diaphragm, the esophagus and the PEL are all elastic, not rigid structures. Because of this, point O is elevated as the PEL stretches and the angle changes. Nevertheless, it is clear that the pull of the contraction LM will have two effects: 1.) It will open the sphincter and 2.) It will stretch the PEL producing a "sliding hiatus hernia."

If equivalent force is applied at the endpoints of the PEL, D1 and D2 by the diaphragmatic contraction of a hiccup, the resulting distribution of forces will be identical. Thus a hiccup is the mechanical equivalent of a contraction of the LM and has the same effect in releasing the sphincter.The PEL is essential to this force resolution. When it ruptures, reflux is alleviated.

Legends

A GEDANKEN EXPERIMENT

The oval represents the muscular wall of the thorax and abdomen lined, in the case of the latter, with peritoneum. In the static case, the abdominal contents act as a "bag of water" and, with normal abdominal tone, will tend to extrude the peritoneum through the gaps in the wall forming ventral hernias at v and inguinal and femoral hernias at I/f by hydrostatic pressure. There is no hydrostatic pressure on the top of the bag of water. If anything, the membrane closing the hiatal gap h would sag downward.

If the muscular wall contracts, the abdominal pressure increases and the hernias are exaggerated. However, any pressure gradient at the hiatus is equalized by upward or downward motion of the diaphragm.

(2A) Belching: Male age 58. CC "Gas", "Feels like I'm going to explode!", heartburn, nocturnal laryngospasm, cheilosis. Lost teeth at age 19, lump-in-throat symptom.

There was esophagitis grade 2 and grade 2 reflux. Here the patient has been induced to belch. Note how LM traction elevates the PEL above the dome of the diaphragm. Loss of diaphragmatic sharpness is a subtle sign of LMC. Despite a life-long history of LMT symptoms, there is no stretching of the PEL to present as a "hiatus hernia." Such relaxation or even rupture may have a beneficial effect on symptoms.

(2.2) Belching produces a HH. Over the course of a lifetime, belching, gagging, rapid swallowing and vomiting generally stretch the PEL beyond its elastic limits - a limit that is also decreasing with age.

(4-3a and 4-3b) Advanced achalasia: [A] Dilated, redundant esophagus. [B] Narrowing just behind but above the dome of the diaphragm.

(53) Globus due to enlarged lingual tonsil.: CC: "Feels like a peach pit caught in my throat." Nocturnal reflux. On fluoroscopy, grade ii reflux. Captive bolus, duodenitis, 7 cm HH, esophagitis. Normally, there is an air space between epiglottis and the lingual tonsil. An increased anterior curl of the former or enlargement of the latter brings the two in contact and this is perceived symptomatically as "globus." Reflux is the probable cause of the tonsilar in enlargement but lymphosarcoma and other tumors cannot be excluded.

(6A) Enlargement of lingual tonsil causes globus or a "lump in the throat" sensation. The patient states, "It feels like food or a pill is stuck in my throat." Not relieved by drinking water. Reports cheilitis but no wet spot on pillow. On

fluoroscopy, there was a moderately severe esophagitis, copious reflux in response to the dC maneuver, hiatus hernia and a grade iii duodenitis .

Normally, there is an air space between the tip of the epiglottis and the base of the tongue. When reflux causes edema of the epiglottis or, as is shown here, lingual tonsillitis, the air space is lost. In lateral projection [A] the epiglottis is plastered against the lingual tonsil. In the frontal view [B] the contact zone appears as a nearly circular ring above the median raphe of the valleculae. This physical contact between the two structures is perceived symptomatically as a foreign object in the hypopharynx.

(7) Cricopharyngeus "spasm": The force of LM contraction is considerable. Shortening 40% or more of its length, it can tear the lower esophagus from the diaphragm, stretching or avulsing the PEL. A force of equal magnitude and opposite direction is consequently applied to the hypopharyngeal attachment of the organ. The pathological findings in excised specimens of the so-called "cricopharyngeal bar" are those of old hemorrhage and fibrosis - typical of repeated soft-tissue injuries. When the larynx elevates it no longer stretches the upper esophageal sphincter properly giving rise to this appearance. Note that a post-cricoid web is also present causing the marked turbulence (B). This homolog of the "Shatzki ring" further demonstrates the similar mechanics of causation.

The patient had heartburn almost daily. Occasional wet spot on his pillow in AM. Lost his teeth at age 32. At fluoroscopy, grade iii esophagitis, grade iv reflux, tertiary contractions, HH and duodenitis, grade iii, were also noted. The p-wave showed impaired cleanup.

(8A,B) LMC causes hiatal transtraction ("hiatus hernia") (A) Severe reflux esophagitis apparently without HT. (B) A few seconds later LM shortening pulls the fundus through a distensible hiatus. Note the soft tissue investments.

(8A) LM tension causes the gas/bloat symptom: Here the patient is trying to vomit. He habitually induces vomiting to relieve the sensation of left upper abdominal pressure. Severe heartburn. Acid regurgitation but no cheilosis. Frequently a wet spot on pillow in AM. Numerous dental caries, frequent sore tongue. Mild duodenitis.

[A] Severe esophagitis: There are no signs of HH with the LM relaxed. [B] The remarkable force of LMC has stretched the PEL as the patient retches. Spontaneous mass contraction drew the fundus 8.5 cm above the diaphragm. This happened slowly enough over a period of about 10 seconds that the patient could be asked whether he was having the "pressure" sensation that was his chief complaint. He emphatically responded that he was. Questioned a few seconds later, after the LMC had subsided, he reported that the "pressure" sensation was gone.

In vomiting this sequence occurs almost instantly. The LM has contracted 37% of its length (8.5 cm/23 cm). One can imagine what this would do to reconstructive surgery about the hiatus.

(11) The esophagus "sees" foreign bodies. In [A] a barium tablet is arrested by a LER. Although it appears arrested below the diaphragm, it is merely projected below the dome. In [B] LM contraction has been provoked an attempt to dislodge the tablet. Hiatal squeeze is still enough to prevent escape of gas from fundus. The diaphragm is becoming unsharp. At [C], the entire fundus and perihiatal region is elevated by the LM tension that has drawn a gas filled tube of stomach through the hiatus. The PEL tent surrounds this tube. [D] Later a Valsalva maneuver applies external pressure to the tube producing the "empty segment" appearance by inflating the PEL up to its insertion..

It cannot be assumed that the introduction of foreign bodies such as endoscopes, pH meters, catheters, balloons and transducers will be physiologic. Here a small tablet has caused marked LMC and orad sphincter displacement. These and other effects such as sphincter release can invalidate manometric measurements or render them uninterpretable. The air filled gastric tube (arrow) could be mistaken for esophagus lined with gastric mucosa.

(15) Rupture of the phrenoesophageal ligament.: The very elastic PEL provides both the inferior attachment of the esophagus and the force that restores the esophagus to its normal resting length and reduces the sliding HH. In a huge HH such as this, the esophagus is permanently shortened because the elastic PEL is ruptured. Consequently these HH's do not "slide." Resolution of the force of LMC by the PEL also creates the sphincter-opening vectors. When the PEL ruptures, this mechanism is destroyed and the sphincter does not efface well although a bolus will partially distend it. This non-effacement is a not uncommon cause of dysphagia.

By the same token, LMC and hiccups can no longer open the sphincter. These patients usually experience symptomatic remission! This is the explanation for the paradox that the largest HH's are the least symptomatic. Note that the true length of the LES (8 mm corrected for magnification) is much less than it is judged to be by manometric methods. Although one can infer that gastric mesentery herniates along with the fundus in HH, this illustration shows it directly (arrows) proving that it extends to the GE junction.

Without the restoring elasticity of the PEL, the esophagus does not alternate between sort and long. The mucosa no longer needs an accordion pleat, therefore, and LERs are seldom seen after rupture of the PEL.

(18) HH with a wide hiatus. Because of the wide hiatus, there is no hiatal constriction of the gastric blood supply, hence there is no swelling of the gastric mucosal folds above the diaphragm. Such patients do not get achalasia.

(19) "Globus hystericus": [A] The patient had reflux, esophagitis, HH, impaired p-wave and a LER. This deformity of the epiglottis can cause the globus symptom by impinging on the lingual tonsil. [B] There is no laryngeal ventricle "fishmouth" shadow due to swelling of the false or true cords. Reflux can cause a characteristic posterior laryngitis.

(20) With a neutralized CD receptor, LMC causes gross reflux.: The patient has just been placed in the prone RAO position after the dC test has turned off the protective Cannon-Daugherty reflex. A powerful LMC retracts 6 cm of stomach through the hiatus, opens the sphincter and causes gross GE reflux of the water-diluted barium. History of ++ heartburn, nocturnal laryngospasm, "lot of gas" and bloating. Fluoroscopy also showed a captive bolus, esophagitis, duodenitis. Note that the sphincter region is less distensible. This is often misinterpreted as a "smooth stricture" and thought to be a sign of Barrett's esophagus especially when seen in air contrast.

(22) "Gas bloat" with a slipped Nissen 10 years after the operation.: A Nissen fundoplication slips because LM tension pulls the the esophagus through the encircling cuff of stomach, everting the latter in the process and resulting in the complex topology seen here. The esophagus can pass through the hiatus as can a portion of the fundus that is drawn out into a tube [B] The ora serrata (arrow) is 2.5 cm above the diaphragm (arrow) but the stomach bulk must stay behind forming the mass of tissue which now separates the fundus from the diaphragm.

No more dramatic illustration of the power of LMC can be found than its ability to achieve this complication. Because of it, the LM now has a lower purchase on the stomach and need shorten the esophagus correspondingly less to produce the gas/bloat sensation or, from another point of view, the same degree of LMC will produce worse symptoms. Hence the frequency of gas/bloat after this procedure.

Extreme LM tension transmitted to the diaphragm can also produce nausea, a symptom this patient has experienced for 10 years. He also has nocturnal reflux which he believes is affecting his lungs. He has lost 7 teeth in the last 3 years.

(23) Iatrogenic hiatus hernia.: This is the sole example of a "paraesophageal HH" I have been able to collect. Although, it is iatrogenic, this is the way one should look. The esophagus is attached to the diaphragm and the stomach protrudes alongside it. If HH's were due to chronic or intermittent increases in intra abdominal pressure, this should be the most common variety of all.

(24) Chronic esophageal shortening.: The patient has complaints of "gas", nausea and acid regurgitation. Meat sticks substernally. Must sleep in a chair.

[A] The HH is obviously under tension which varies somewhat but never relaxes entirely. It is easy to see how endoscopically this would appear to be a Barrett esophagus - there is gastric mucosa in an esophagus-sized tube 4.5 cm above the hiatus and the sphincter region resembles a stricture. Constant LM traction on the sphincter keeps it open constantly, hence the air esophogram.[B & C] Further LM traction pulls the fundus through the hiatus.

On endoscopy, there was no mass at the gastroesophageal junction (arrow). This probably represents extrinsic pressure by a ruminant of the torn PEL.

(26) Esophageal folds: Normal folds measure 1 mm or less and one can count 5 or 6 of them. They are caused by the circular muscle. Not the m. mucosa that is also longitudinal. There are no transverse folds unless, for one reason or another, the esophagus can shorten with LMC.

(27) Pseudo tumors of the fundus are reduced hiatus hernias: Enlarged, friable mucosal folds in the fundus can be mistaken for a neoplasm. These folds become swollen when the venous return from the fundus is impaired by hiatal constriction. They persist when it reduces.

(28) Rupture of the PEL and the angle of His.: Because of the acute angle of His, it has been assumed that this condition is a paraesophageal hernia. However, it obvious that the esophagus is not attached to the diaphragm at any point due to complete rupture of the PEL. The stomach floats into the chest either alongside the shortened esophagus, producing the acute angle of His seen here, or else telescopes over it to produce the "molar tooth" appearance. Once the PEL ruptures, the patient's reflux may be cured! This is probably responsible for the belief that the angle of His prevents reflux. Note the fatty mesentery along the greater curvature.

(29) LMC and the trumpet GE junction.: The force of LMC is resolved into 2 vector components both of which are well displayed here. a.) One component stretches the PEL, and b.) One opens the sphincter. Because these forces exist in 3 dimensions, are affecting elastic structures and are modified by the oblique PEL insertion, a striking, trumpet-like flaring of the GE junction results.

(30) Tertiary contractions: An esophagus without a functional p-wave will contract en masse. This is the fallback mode when obstruction is encountered. Tertiary contractions are a complex manifestation of a.) LM shortening. [Note the loss of the posterior bowing of the esophagus as the TC's form.] b.) Mass circular muscle contraction. c.) Self buffering - an en masse contraction of circular muscle cannot compress the liquid contents of the organ. The stronger muscle bundles overpower the weaker resulting in alternate constricted and widened

segments. Modern anatomical research has shown that the muscle of the body is in part spirally arranged, a fact that is well shown during such contractions. There appear to be two spirals - one ascending from below, the other descending from above.

The velocity of both LM and CM contraction is remarkable as can be seen on these exposures second apart.

(31) The lower esophageal sphincter. : Here the bulb-syringe action of mesentery crowding into the tented PEL is powerful enough to override the sphincter, forcing the captive bolus back into the body of the esophagus. At this instant, the true length of the sphincter is seen to be only 7-8 mm, far shorter than has been supposed from the evidence of manometry. The sphincter remains closed against this considerable force from below, but when it does yield (after 3 seconds) it then relaxes completely within a second. Gastric mesentery crowded into the PEL tent by the Valsalva maneuver is the driving force.

(31X) Belch: LMC jerks the stomach into the chest to the end of the tethering PEL.

(32) Lower esophageal ring formation:

This sequence illustrates why LER's are at the GE mucosal junction. The advancing p-wave milks any mucosal redundancy to the limit of its travel. It cannot push it farther, because this is where peristalsis stops - in this case about 5 mm above the mucosal junction. In other cases this short aperistaltic segment may be 2.5 or even 3 cm in length. One never sees peristalsis rolling over a LER. Note that peristalsis consists of a shortening cone of contraction.

(33) The De Carvalho test: Frame [A] shows barium in a short tube of stomach above the diaphragm. In frame [B], it is diluted by water being swallowed by the patient in the RPO position (supine, right side down). The sphincter is still closed, however. In frame [C] refluxing barium is diluted by water in the esophagus as the CD receptor is flushed. By frame [D] the entire esophagus is flooded with refluxed barium. This is the time to ask the patient a.) whether he can feel something coming back up and b.) whether it reproduces his symptom in all but degree.

Both cats (Cannon) and ruminants (Daugherty) have a sensory area near the mouth of the esophagus that inhibits LM contraction when stimulated by ingesta. The de Carvalho maneuver, as shown here, washes this area free of ingesta allowing reflux to occur in patients with high LM tension.

(34) Longitudinal muscle contraction causes transverse folds.: The widely distended bowel could be due to paralysis or distention. Folds cannot form unless the lumen is obliterated. Numerous transverse folds, misnamed valvulae

conniventes prove that the LM is contracting and thus distinguish mechanical obstruction from paralytic ileus in which atony of the m. propria prevents any fold formation.

(35) Achalasia can clear in seconds: Frame (A) shows an esophagus emptying slowly by hydrostatic pressure alone as there is no peristalsis. Note the "tram-lines" similar to those seen in hypertrophic pyloric stenosis passing through the hiatus and the "bird beak" termination at the diaphragm. The latter appears to be partly surrounded by a 3 cm soft tissue mass in the gastric air bubble. The beak of the "bird" tends to approach the horizontal because the esophagus is redundant because of the 4.5 cm HT that is not seen until frame [C].

Later frames showed normal peristalsis with excellent peristaltic cleanup despite a grade ii esophagitis. The soft tissue mass has vanished! Frame [D] shows the subtle, yet unmistakable edge of a trumpet which indicates powerful LMC that has released the trapped fundus by reducing hiatal squeeze.

(35X) Transient achalasia: There is overnight retention of mucus globules [A] in the lower esophagus. Emptying was by gravity only. A Valsalva maneuver demonstrated a transtract [C] and when this reduced all signs of achalasia vanished.

(36) Terminal annular constriction [A] is not a carcinoma, a stricture or "terminal esophagitis" of Schatzki but a sphincter that cannot efface because of rupture of the PEL [B].

(37) A slightly more severe stage of achalasia: The nearly horizontal "bird-beak" configuration [frames A &B] and tramline shadows (frames 2-5) are shown to be in the herniated stomach by the fact that the p-wave - which stops at the sphincter - ends well above the diaphragmatic constriction. A good clean-wiping p-wave is still able to force the partial obstruction [C & D]. During this stage, the CM will undergo work hypertrophy.

(38) Failure of sphincter latching: This patient with chronic reflux had a mild impairment of the p-wave. The feeble p-wave cannot latch the sphincter and, on encountering back-pressure because of the temporarily occluded PEL, gives way and allows reflux back into the esophageal body [D]. This may be an important factor in the muscle hypertrophy of achalasia.

(39a) Pseudo tumor of the fundus : The venous return is compromised when a gastric segment is trapped above the diaphragm [A]. The result is engorged and friable mucosa. Clinically, this accounts for the tendency of HH's to bleed. Such mucosa may bleed 1 unit/month without turning stools guaiac positive. When the HH is again normally situated below the diaphragm [B], this engorged mucosa - and gastric wall as well - present as a tumor-like mass in the fundus. Provoking the HH [B] will cause the pseudotumor to vanish as shown here.

(40) LMC opens the sphincter. : In some extreme cases of excessive LMT the sphincter may remain open indefinitely as in this patient. The trumpet shape is the geometrical resultant of resolution of the force of LMC into sphincter-opening and PEL-stretching components. Receptors in the fundus, when stimulated by acid/pepsin, inhibit LMC. The appearance seen here may be reproduced by washing the posterior wall of the stomach with water (the de Carvalho maneuver) thus "turning off" the inhibitory reflex.

(41) The p-wave stops at the sphincter. : The cone of peristalsis has its base on the sphincter. It becomes progressively shorter, but never passes beyond the sphincter. Even without landmarks such as the LER seen here, this fact makes sphincter identification easy. In this patient, as is very common, there is an aperistaltic segment - the region below the sphincter and above the LER. Here it measures 2.4 cm, but may be much shorter or even non-existent.

(42) Non-effacement of the sphincter. : The PEL is ruptured in this patient with 50% of the stomach in the chest destroying the normal mechanism for sphincter effacement. The sphincter may be pylorus-like as in this case, neither opening or closing. This may cause mild dysphagia. The 1 cm sphincer length is far shorter than the 4.5 cm or more derived from manometry.

(44) Disappearing LER: When the HH is demonstrated with the Valsalva maneuver (A), a typical LER forms. When it is evoked by inducing belching (B) there is no trace of a ring. The main difference is that there was no peristaltic wave in (B) to milk mucosal redundancy distally. Also note the long aperistaltic segment between ring and sphincter. The slight hourglass constriction in [B] (arrow) is the less distensible sphincter region. This may be misinterpreted as a stricture in air an distended esophagus and diagnosed Barrett's esophagus..

(45) LMC produces reflux: LMC is evident from the marked tenting of the fundus into an unusually wide hiatus. The lateral stretching of the GE junction explains the fact that, although produced by longitudinal traction, Mallory-Weiss tears are also longitudinal. Barium in the distal esophagus is reflux. The hiatus itself is widened by the lateral resolution of LM force. If a transducer is measuring hiatal squeeze, it will register decreased pressure when the LM contracts!

(46) "Inflammatory" gastro-esophageal polyp: The term is probably a misnomer. Note the tight hiatus which constricts circulation in the portion of the stomach retracted above the diaphragm.

This is a minor degree of pseudotumor of the fundus.

(47) The hiatus itself may be a cause of dysphagia. : A 12.5 mm barium tablet was arrested at the diaphragm [A] Compare tablet with the hiatal size in frame B. The patient also had an apparently normal, but fixed pylorus which neither

contracted or expanded. His symptoms ("Feels like there is a clamp on my stomach.") were completely relieved by endoscopic dilatation of the pylorus.

(48) Severe esophagitis, grade iii. : 2 folds occupy the entire width of the relaxed body.

(49) Corkscrew esophagus: Every book on the esophagus has at least one of these. This elderly gentleman had been followed for many years with frequent GI exams at leading institutions. The spectacular curling allowed the obstruction (arrow) to go unnoticed. It is easy to see spiral muscle bundles in this case. The tracheal aspiration was asymptomatic and did not elicit a cough reflex.

(50) Symptomatic post-cricoid ring with Zenker's diverticulum: CC:"Food sticks in throat." Comparing these landmarks with the first rib or a cervical vertebrae shows they have an upward excursion of 2.8 cm at the outset of deglutition. This exerts an abrupt, forceful tug on the esophagus which, transmitted to the PEL, may supply all the force needed to open the sphincter. The sharp tug may also trigger a stretch reflex causing LMC.

As is invariably the case with Zenker's diverticula, the patient also had a HH. The association of the two is due to the circumstance that powerful LMC stretches or disrupts the esophageal attachment at the hypopharynx as well as at the diaphragm.

(51) Peristalsis stops at the sphincter.: Frames 1,4, 9, 10, 11 of a sequence made at 2 frames/sec. (A) The esophagus is flooded with barium from a large bolus. The thick ring sometimes called the "A" ring or "muscular ring," (arrow) is actually the sphincter. In (B) a mucosal ring comes into view as the esophagus shortens. The sphincter is partially obliterated by increased intraesophageal pressure from the advancing p-wave. Despite mild esophageal varices, the p-wave is clean-wiping to the lower edge of the sphincter. The short segment between the sphincter and the ring is aperistaltic. Note the LM relaxation as soon as the p-wave reaches the sphincter. The patient had gross GE reflux, easily provoked by dorsiflexion of the cervical spine.

(52) Long PEL: Although the PEL is difficult or impossible to see in most cases, it produces effects which are unmistakable. Here a Valsalva effort collapses the stomach in the PEL distal to its attachment because hydrostatic pressure is confined to the (in this case) tubular PEL tent. This extreme example shows the mechanism responsible for the "empty segment." It has no part in preventing reflux. Tissues slowly crowding into the PEL have been mistaken for prolapsing mucosa.

(54) The Cannon-Dougherty reflex: If the posterior wall of the stomach is flushed with water in the supine position [A] it may "turn off" the CD reflex that normally inhibits reflux. A hypertonic LMC then produces gross reflux of either air or

gastric contents when the patient assumes the prone RAO position [B & C]. When LMC occurs at this point one simply asks the patient if he has the gas or bloat symptom at that instant.

The deCarvalho maneuver causes reflux in patients with a hypertonic LM because it neutralizes the CD inhibitory reflex.

Compare the appearance of the gastric segment (a traction cone in [B] & [C]) when a hiatal transtraction is produced by this method with that (captive bolus) produced by the Valsalva test in the same patient [D] The faint outline of the PEL tent can be seen here.

(55) Nocturnal laryngospasm is a frequent symptom of reflux. Aspirated barium defining the pyriform sinuses, true and false cords and laryngeal ventricle. It may cause a posterior laryngitis with intermittent hoarseness.

(56) Post-cricoid ring: In many ways analogous to the LER, it is not restricted to PVS. Oddly enough, unlike LERs, there is no resistance in academia to the idea that these rings are mucosal plications. Note flow disturbance below ring.

(58) Belch: (A) Before, (B) After. Although the release of air from the stomach may be explosive, unless the superior constrictor releases, the esophagus may remain air-distended for 8-24 seconds affording an excellent opportunity to note the traction effects of LMC and their correlation with the opening of the sphincter. Barium is dilute from the dC test which has turned off the CD receptor that acts as a guardian of the gate to prevent reflux when submerged in acid/pepsin. The trumpet shape explains the orad directed wedge shape of Mallory-Weiss tears.

(60) Candida infection: Male, age 26 with HIV-III virus infection. There are actually 3 types of abnormal folds: a.) Thickened longitudinal folds similar to those of reflux esophagitis, b) Mammillary, rice-grain nodularity and 3) Transverse folds. The latter produce the saw-tooth appearance in profile when the stiffened mucosa cannot take up the slack as LMC shortens the organ.

(61) Transverse folds with eventration of the diaphragm: Although the patient did not have an elongated PEL, eventration of the left leaf of the diaphragm provided enough slack for development of transverse folds upon LMC. Longitudinal folds are increased in thickness and decreased in number due to mucosal thickening by esophagitis.

(64A) The pinchcock at the diaphragm: The constriction is actually well above the diaphragm and obviously much thicker than the diaphragm. On this lightly exposed film, one can just make out the bell-like tent of the PEL which, by constraining mesentery protruding through the hiatus, chokes off the fundus attached at the apex.

(64B) Severe grade 3 reflux esophagitis. Grade = 5 minus number of folds.

(68) Achalasia and DES: Work hypertrophy in achalasia/DES is not idiopathic. It is secondary to intestinal obstruction.. The normal response to obstruction is contraction of the LM and en masse contraction of the circular CM. Chronic obstruction of the esophagus causes both "tertiary contractions" and work hypertrophy of the circular muscle. The following table was calculated to determine the cross-sectional area of the esophageal wall at the levels shown on this exposure, a part of a 2-second burst at 10 frames/sec.

OD	ID		AREA	% Increase
24.4	16.2	A	2.62 cm2	0.00
22.9	11.6	B	3.05 cm2	16
22.9	7.5	C	3.65 cm2	39
25.2	9.8	D	4.19 cm2	60
27.1	8.6	E	5.20 cm2	96

Measurements were made with a dial gage and corrected for minification of the 105 mm format camera. A 12.5 mm barium tablet was used to calibrate the latter. Although this hypertrophy is always referred to as idiopathic, work hypertrophy is the obvious cause. Aganglionosis, on the contrary, causes loss of muscle tissue. The reason the hypertrophy increases aborally is that the intra luminal pressure increases as the p-wave moves distally (There is less length of lumen to contain the same volume.) so progressively more work must be done.

The obstruction is in the soft tissue mass between the stomach and the diaphragm [G]. This is a small piece of stomach or omentum trapped in the PEL tent and hiatus.

[B/C/D] The speed of CM contraction can be appreciated from these exposures made at 1/10ths second intervals. Again note greatly increased wall thickness.

68 [E/F] The sphincter (arrow) would be difficult to distinguish from the other constrictions were it not for its usual relation just above a LER (open arrow). Wide open, it is not causing obstruction.

68 [G] In LAO Note faint tramline and a knuckle of HH passing through the diaphragm. Interposed tissues separate the fundus from the under surface of the diaphragm. The obstruction is due to these tissues caught in the small hiatus. This narrowing is generally thought to be the sphincter but is far longer than the sphincter and is, in fact, compressed stomach.

T-1 Reflux into salivary ducts: Note opacified Wharton and Stenson ducts. Apparently this was asymptomatic in this patient with neurogenic dysphagia, although if acid/pepsin were involved gland inflammation would be anticipated. Evidently this is not on the PVS palate.

T-2 "Terminal esophagitis": Shatzki believed this appearance was inflammatory, however, it occurs so frequently in patients with ruptured PELs that it seems more probable it is due to non-effacement of the sphincter. When the esophagus shortens, it becomes thicker for purely geometrical reasons.

T-3 Omentum crowding into the PEL tent. Note the PEL insertion at the sphincter. Although the rest of the PEL is largely invisible, it can be appreciated by the way it constrains the infra diaphragmatic tissues after they protrude through the hiatus.

T-4 An unusual sphincter variant: The sphincter, initially double (A), was demonstrated in the usual way when pressure from the CB forced it. Two frames later [C] it has merged into one.

T-5 Systemic sclerosis look-alike: The circular muscle of the esophagus atrophies in systemic sclerosis while the LM is unaffected. The result is this typical appearance. The esophagus is constantly short. The HH never reduces. The sphincter never closes. As a result, there is a constant air esophogram which can be seen even on chest films. The superior constrictor does not relax which, at least in the upright position, prevents gastric fluids contacting the esophagus. There is no peristalsis below the striated muscle portion. Note the relatively small hiatus and turgid gastric mucosal folds in the transtracted stomach.

The patient is a 71 year old undifferentiated schizophrenic on long-term haloperidol (Haldol) medication. He also had megaduodenum and pseudo-intestinal obstruction but did not exhibit Renaud's phenomenon. Such drugs may mimic systemic sclerosis.

T-6 LMC with tubular HH: The short esophagus with a tubular HH can easily be mistaken for an esophagus lined with gastric mucosa. Note the mucosal transition and compare with published cases of BE. Distention of the esophagus with air causes reflex LMC producing this appearance.

T-7 Multiple LERs: Can each one have a different etiology? Or are they accordion pleats?

T-8 So-called "intramural diverticula:" The name is an oxymoron as if they are intramural, they are, by definition, not diverticula. The only thing they could be due to is barium in ectatic mucus glands, the glands that provide esophageal lubrication.[Case provided by O. Arthur Stiennon, III]

T-9 Fundic peristalsis: The most proximal gastric p-wave I have ever encountered was in this severely nauseated patient. Gastric peristalsis cannot take over when the esophageal p-wave stops.

T-10 Candida infection with AIDS: Note the enlargement of the mucus glands.

T-11 The "Burnoose phenomenon:" Note how a hood appears about the mouth of the esophagus and then vanishes as the LM draws it upward. It has been aptly likened to an Arab headdress by Jutras.

X-10 A toroidal Angelchik prosthesis: closely mimics the toroidal appearance of gastric omentum in the PEL tent and thus the appearance of "achalasia."

Y1 The cause of achalasia: The outlines of the PEL and its contents can be seen here as LMC elevates the latter through the hiatus. Even a tag of fat trapped in a small hiatus when the LM again relaxes can cause obstruction causing the "achalasia" appearance.

Y2 (A) Epiphrenic diverticulum for 34 years. These diverticula form as a buffer because, if there is obstruction at the hiatus, there is no other place for the bolus to go when peristalsis reaches the end of the esophagus. For the same reason, they are epiphrenic. Note the small hiatus. (B)The diverticulum is no longer in contact with the diaphragm because LMC has drawn 7 cm of stomach into a tube in an unsuccessful attempt to clear the obstruction. Shortly after this film was made she was operated on for obstructive symptoms and the diverticulum resected! The result was satisfactory, however, probably because the hiatus was widened incidentally.

Y 3(A) Classical decompensated, redundant esophagus of achalasia. (B) Note the small hiatus.

Y-4 *Achalasia can occur overnight and reduce spontaneously: Fluoroscopic note:"Barium entered the esophagus to mix with a great deal of secretions and food particles. There were spectacular 'tertiary contractions' which were ineffective in emptying the organ. A barium tablet lodged in the distal esophagus. There were 2 narrowings. One was 3 cm above the diaphragm corresponding to the sphincter (frames 1-4) and the other was at the actual diaphragmatic hiatus. The tablet lodged below the sphincter. At this point a longitudinal contraction of the esophagus occurred (frame 5-9) that tented the diaphragm. Simultaneously both areas of constriction disappeared relieving the obstruction."

This sequence illustrates the function of LMC in dislodging obstructing food particles. It not only obliterates the sphincter but widens the hiatus itself. Here a small hiatus is holding up passage of a barium tablet. Note how the GE junction, that appears to be below the diaphragmatic dome in frames 1-4, is abruptly elevated above the dome as the tablet clears the hiatus in frames 7-9.

Y5 Hypertonic longitudinal muscle: Frames 1-17. LER, HH, esophagitis, gr. 2 reflux. For a 19 second exposure, the sphincter alternately opened and closed

depending on strength of LMT. The sliding HH never reduced completely. Note the "alpenhorn" configure of the GE junction when maximally stressed. I have seen a sphincter remain open under LMT for over 28 seconds in a patient with a hypertonic LM and indefinitely in myotonia dystrophia and scleroderma. The LM opens the sphincter. '

Y6 Non effacement of sphincter: Rupture of PEL demonstrated by (A) the dC maneuver, and (B) by inducing belching. The force of LMC is resolved into sphincter-opening vectors by the PEL. When it ruptures this mechanism fails giving the appearance that Schatzki attributed to "terminal esophagitis." It is actually a non-effaced sphincter. A mild dysphagia may result.

Y7 Barrett esophagus: The short esophagus + stricture + tubular HH shown here may appear to the endoscopist to be a Barrett esophagus. Acid reflux can cause esophageal shortening. If films are exposed for low contrast and low density, one can often see the external surfaces of these structures.

Y9 Longitonia: Reflux, esophagitis, vallecular sign, aspiration. For 30 seconds the sphincter remained open, the LM contracted and the diaphragm tented. Despite marked aspiration there was no record of pneumonitis.

Y10 Every Zenker's diverticulum (A) has an associated HH. Noting the damage LMC has caused at the lower esophageal attachments (C= belch), it is surprising there are not more Zenker's diverticula as the force on the upper attachments is the same. The so-called "cricopharyngeus spasm" is also the result of local muscular avulsion. This patient had a persistently taunt LM (B). Only a slight notch at the sphincter identifies the GE junction. These tubular HHs are mistaken for esophagus lined with gastric mucosa and the less distensable sphincter area is mistaken for a stricture.

Esophageal thickening must be interpreted cautiously. A 33% shortening of the length will cause a 50% increase in thickness.

Y11 Non effacement of sphincter with ruptured PEL. The sphincter-opening vectors generated by LMC require resolution by the obliquely inserted PEL. Such cases are often labeled "terminal esophagitis."

Y12 LM tension is the cause of gas/bloat. A sustained forceful LM contraction shortens the esophagus 30% here as it draws the stomach through the hiatus (A), opens the sphincter (B) and causes reflux (B-C). Severe bloat with severe subxiphoid pressure "Making it difficult to breath," ++ pyrosis. A calcium channel blocker (10 mg Nifedipine) relieved the gas/bloat symptoms within minutes. The effect of such drugs on the CM is well known. S = sphincter.

Y13 Hiatal canal obstruction: "Trouble swallowing food." (A) A 12 mm barium tablet was held up at the hiatus. (B) This small hiatal canal was the cause of the obstruction. Note that usual ROA projection gives the false impression that the GE junction is below the diaphragm.

Y14 A typical "bird beak" of achalasia. The "gastroesophageal polyp"(arrow) identifies stomach above the narrowing.

Y15 " Longatonia" Persistent LM contraction keeps the sphincter open after the Cannon-Dougherty receptor has been turned off by the dC maneuver. Typically such patients complain of gas-bloat, reflux and its complications.

Y16 LM power and its effects: This sequence illustrates both the remarkable power of LM contraction and the equally remarkable elasticity of the PEL. In frame 1 there is just a nubbin of stomach above the diaphragm. The diaphragm is sharp. In frames 2-6, made during a belch, the stomach is relentlessly transtracted through the hiatus 7.6 cm as the esophagus shortens 35% of its length. The diaphragm itself is tented and the hiatus stretched to its widest extent (frame 5). LM contraction, as here, widens the hiatus thus affecting measurements of hiatal squeeze. Note the wide open sphincter and alpenhorn sign as the LM relaxes slightly in frame 7. Further relaxation closes the sphincter.

Y17 The captive bolus test: Three things are necessary to demonstrate the captive bolus. 1) A normal p-wave, 2) Enough elongation of the PEL to form a tent, and 3) A Valsalva maneuver to increase intraabdominal pressure, force mesentery into the PEL and so obstruct the fundus. This is the optimal way of studing swallowing against resistance

This patient has a somewhat elongated PEL making a captive bolus possible. Here he is doing a prolonged maximal Valsalva maneuver. It is easy to confirm many of the points made in this book, namely:

The p-wave stops at the sphincter, frames H-L, thus identifying both the sphincter and its location.

Separate crinkles of esophageal mucosa (frames A,B) coalesce into a LER (frame C and onward). This demonstrates why LERs occur where they do - the p-wave cannot force them into the stomach because it goes no farther - and why they form: there is 39 mm of redundant mucosal length that must go somewhere when the esophagus shortens. The two surfaces of the ring account for 19 mm of this leaving 2 cm for the elastisticity of the m. mucosae and slippage.

Mesentery crowding into the tented PEL acts, first as an obstruction inciting a maximal p-wave [C] then as a piston, by crowding into the confined space of the tented PEL. This force may be sufficient, as here, to eject barium back through

the contracted sphincter (frames J-T). Note the smaller size of the CB in the later frames.

The sphincter, which is still activated, then stands in sharp contrast to the CM (frames Q-T) which has relaxed in the wake of the p-wave as proved by its easy distention by the jet of refluxing barium (frames N through R). A dial gage measurement corrected for minification gives a sphincter length of 9.6 mm - far less than the 3.5-4.5 cm squeeze measured by manometry.

It should be noted that in this position - 4 cm above the diaphragm - there is no possibility of the sphincter receiving auxiliary support from the angle of His, pouting of the gastric mucosa, the valve of Guberoff, intraabdominal pressure or any of the myriad of postulated mechanisms upon which surgical treatment of reflux is based. In other cases, despite the considerable force which intraabdominal pressure - acting like a thumb on a bulb syringe - exerts to force the contents of the gastric sphere back through it, the sphincter maintained perfect competence as long as the patient could sustain the Valsalva erffort.

By measuring the relationship of the LER to the diaphragm - stationary throughout the maneuver - we obtain an accurate index of the state of contraction of the LM. This shows that there is a rapid LM contraction while the p-wave is in the proximal esophagus. By the time the p-wave cone is in the distal esophagus, LM shortening is maximal. It peaks just as the p-wave enters the distal 10 cm of the esophagus when it abruptly relaxes.

Maximal LM contraction of 2.8 cm + the height of the CB for a total of 7.7 cm occurs by frame H as the p-wave merges into the sphincter area. Thereafter the LM relaxes. [Relaxation is not as noticeable because the transtracted stomach holds it up.]

In frames G through M a very faint spherical soft tissue density surrounds the CB. This is mesenteric fat inflating the PEL tent. High contrast or over-penetrated films will not show it.

LM contraction causes fundic transtraction ("hiatus hernia") not increased intraabdominal pressure.

The long constriction in the PEL tent (B) is similar to the obstructive appearance in achalasia. Note tram lines.

The reader may note that there is no difficulty in assigning state formulas to each frame in swallowing against resistance.

(Y17-V) The same patient as in Y17A-T. Here the LM "knows" the barium tablet is caught by the LER and contracts in an effort to expand the esophagus and pass the tablet. In doing so it produces a HH that has an entirely different

morphology than when elicited by the Valsalva maneuver. The presence of foreign bodies (tubes and transducers) in the esophagus may invalidate a manometric study.

X1A,B Signs of LMC. Male, 42 with life-long reflux, severe cheilitis, wet spot on pillow in AM. Chronic sore throat, lost his teeth at 19. [A] Grade 2 reflux with de Carvalho test -RPO. [B]Cannon-Dougherty reflex was turned off by dC test and patient turned to RAO provoking belch. Note trumpet configuration of GE junction and loss of diaphragmatic sharpness. L. Gr. 2+ esophagitis. C.) Such patients almost invariably have duodenitis.

X2 Gas-bloat with LMT. Male 56, pyrosis, bloat, nocturnal laryngospasm, lost teeth at 26. An uneffaced sphincter serves for measurement of LMC. Note shortening from A to B. The esophagus was constantly shortened. This causes traction on the diaphragm producing the "gas-bloat" symptom. This patient also has an enlarged lingual tonsil, gr. 2 vallecular sign and the p-wave was ineffectual.

X4 LMC after deCarvalho test. [A] Barium is dilute from the dC test which has turned off the CD reflex.LMC lifts the perihiatal tissues [B] until the sphincter yields [C]. This effect of LMC widens the hiatus, opens the sphincter and translates the latter proximally. It is likely that these effects invalidate many manometric measurements purporting to be of sphincter pressure. LMC is one cause of the unpredictable transient complete loss of LESP. The other is a hiccup.

X5 "Watermelon stomach": Once the PEL ruptures, the stomach remains stationary while the hiatus slides down and up with inspiration [A] and expiration [B]. This can cause stripe-like erosions of the gastric folds as they rub together.

X7 Ruptured PEL: A slight molar tooth shape results when the esophagus invaginates the stomach. Note the poor effacement of the LES.

35xa Transient achalasia: may exhibit all of the classic signs yet have them vanish during the course of the examination.[A] Overnight rentetion of mucus globules with tramlines passing through the diaphragm and narrowed PEL tent are typical of achalasia. [B] Gastric folds above diaphragm. [C] A Valsalva maneuver demonstrates the hiatal transtract that is causing the obstruction. It also dislodged the fundus from the hiatus and thereafter there was normal peristalsis. This and "elevator esophagus" are typical for transient achalasia.

Z1 Post poliomyelitis: The gluteal musculature in this patient with right sided paralysis shows what happens to aganglionic muscle.

Z2 The longitudinal muscle opens the sphincter. Obvious signs of LMC in [B] correlate with the open sphincter. Note gastric mucosa above the diaphragm.

The repeated stretching of the PE attachments over a lifetime will elongate or even rupture them.

ILLUSTRATIONS TO BE COMPUTER GENERATED

CG-1

t1 Thickness of mucosa in distended organ.

t2 Thickness after lumen obliterating contraction

r1 Radius of distended organ - center to muscularis propria.

r2 Radius of contracted organ.

When the m. Propria contracts, it obliterates the lumen. While the x-sectional area of the lumen goes to zero, the circumference of its lining mucosa can not. It is necessarily thrown into folds.

CG-2

If the set of patients with both malady A and malady B is empty, ie., ~(A&B) by the calculus of Boolean algebra equals ~A or ~B. That is, patients are spared either one disorder or the other but not both. This appears to be the case in practice: a patient can have achalasia or HH but not both. This implies that either one disease prevents the other (which is obviously not true) or that one causes the other. The latter can be true if A & B are the same disorder - now diagnosed one way, now the other.

CG-2

In the 1990 to date Medline database, although there were many articles on HH and on BE, there were only 11 in which the abstract mentioned both keywords. After deleting the miscodes, only 4 of this 11 mentioned patients with both disorders. As both the BE and the HH populations are subsets of the GE reflux population, one would expect that at least 70% of the BE articles or 253 would deal with patients who had HHs as well, not the 4 that actually did so. Equally remarkable, 97% of the articles on HH failed to mention BE. The conclusion is that HHs are being diagnosed BE and vice versa or, more likely, they are the same thing.