The cause of achalasia is unknown (idiopathic). There is a loss of the esophageal wall and in the lower esophageal sphincter which occurs at any age between childhood and the ninth decade having an equal frequency between males and females. Achalasia is a disorder characterized by a loss of peristalsis (propulsive muscle contractions or movement) in the esophagus and failure of the lower esophageal sphincter to relax and allow emptying of the esophagus.

The term achalasia means to loosen or relax. This condition is seen in newborns, especially premature infants who regurgitate after most feedings for the first several months of life. At about four months of age the lower esophageal sphincter (valve) begins to close normally between swallows and the regurgitation from the stomach is prevented. The opposite is true for achalasia which occurs at any age between childhood and the ninth decade. Regurgitation (evacuation of esophageal contents into the throat and mouth) occurs as a consequence of both overflow and abnormal esophageal contractions. Failure of emptying into the stomach, regurgitation and, in some, a fear of eating, all may lead to weight loss. Patients learn to improve esophageal emptying by eating slowly and swallowing extra fluids. The problems with esophageal emptying and regurgitation can cause considerable embarrassment in social settings. Regurgitation, especially at night, can result in aspiration of fluid into the lungs causing bronchitis and pneumonia. Another aggravating factor is sialorrhea, which is the production of large amounts of saliva in response to the esophageal obstruction. Patients notice the sialorrhea as a large amount of "foamy mucus" requiring frequent swallows to keep the hypopharynx free from this bothersome fluid.

The diagnosis of achalasia is usually first suspected after the physician takes a complete history. A barium swallow (esophagram) reveals a dilated esophagus with a large amount of retained barium. The esophagus at the lower sphincter is tapered to show a narrow column of barium likened to the shape of a bird beak.

Endoscopy reveals a large esophagus with retained food and fluid. However, if the patient is properly prepared before this exam by two days of clear fluids, only a small amount of liquid content is found. A clean esophagus is essential at the time any treatment is performed. The endoscope passes into the stomach through the closed lower esophageal sphincter with only slight resistance.

The diagnosis of achalasia is best confirmed by performing an esophageal manometry (motility) study which confirms the loss of peristalsis in response to swallows and a high pressure in the lower esophageal sphincter with incomplete or no relaxation.

Once the diagnosis is established a decision regarding therapy is in order. Some patients with early achalasia and mild symptoms may elect to delay treatment or try medical (drug) treatment. Such treatment with calcium channel blocker drugs may help by transiently relaxing the closed sphincter but this response is usually short-lived.

Ultimately most patients opt for some form of treatment. None of the treatments available will substantially improve the peristalsis in the esophagus. All are designed to weaken the lower esophageal sphincter. The treatment options have been primarily dilation of the tight sphincter with balloon dilators, available in diameters of 30, 35 and 40 mm. These instruments are intended to over-stretch the sphincter to the point that the muscle fibers lose their ability to contract and thereby allow the esophagus to empty more efficiently. This form of therapy provides good to excellent improvement in from 70 to 90% of patients depending on the size of balloon used. The main risk of this treatment is perforation or full-thickness tear of the esophagus that occurs in 2 to 5% of patients. If this occurs a surgical operation usually is needed to close the tear. At the same operation a myotomy or muscle splitting operation to treat the achalasia may be performed to provide definitive treatment.

In past years, when balloon dilations failed on two occasions, an operation called a Heller myotomy (surgical division of the sphincter muscle) was recommended. This surgical procedure gives good or excellent results when performed by an experienced surgeon. Currently a modification of the Heller myotomy, called a laparoscopic myotomy, can be performed using the laparoscope and accessory instruments through several small incisions. As more experience is gained this procedure likely will be preferred when the surgical approach is needed. If the sphincter muscle is rendered too weak by myotomy, there is a risk for acid in the stomach to reflux into the esophagus and cause esophagitis or a stricture. Surgeons attempt to reduce this risk by performing an antireflux operation at the time of the myotomy.

The latest treatment for achalasia currently under evaluation is the injection of a bacterial toxin, clostridium botulinum toxin A (Botox). The botulinum toxin prevents the release of a chemical (acetylcholine) from the nerves in the lower esophageal sphincter resulting in relaxation and reduction of the high pressure. This effect prevents the muscle from contracting and often leads to increased emptying and improvement of the patient’s swallowing and regurgitation. The Botox is injected through a small needle passed through the endoscope. The early results of this treatment are encouraging. It appears to have few if any complications and appears safer to perform compared to other treatments.

The long-term results are not yet known but most of the patients treated beginning about 18 months ago by physicians at Johns Hopkins University have continued to have improved swallowing as of the latest report in May, 1994. The results in our patients treated with Botox since December, 1993 likewise are encouraging.
Aspiration, or unwanted entry of food, liquid, or saliva into the lungs following an attempt at normal swallowing, is the single most serious problem that can occur as a result of swallowing disorders (dysphagia). Aspiration places the person at risk for development of pulmonary infection and pneumonia. As a result, this complication ranks as the fourth (4th) most frequent cause of death in the elderly and the fifth (5th) leading cause of mortality in the United States.

Currently, the modified barium swallow study (MBS) is the most widely used and effective technique to detect aspiration. In particular, the MBS is helpful in identifying when aspiration occurs, why it occurs, and how much occurs. The patient is provided with various amounts, types, and consistencies of barium covered food materials by the speech pathologist. The radiologist records the patient's swallowing attempts on a video-recording system with slow-motion capabilities. This allows the physician and members of the multidisciplinary swallowing team to study the patient's swallowing anatomy and physiology in a more detailed manner to more effectively identify a management plan. During the radiographic procedure, the speech pathologist also evaluates the effectiveness of various rehabilitation strategies in reducing or eliminating the occurrence of aspiration.

Unfortunately, the MBS study still is not available to patients in many communities or has limited applicability to others. The reasons are numerous. Most commonly, the MBS is not available in nursing homes or in smaller hospitals. The study may place physical demands on the medically compromised patient who is unable to travel to the radiology suite. Often, sufficiently trained personnel may not be available to perform or interpret the examination. Exposure to radiation prevents frequent repeat exams which may be necessary in the post-operative period or in rapidly evolving swallowing disturbances to make up-to-the minute decisions on case management.

Recently, a modification of current technology has provided an alternative or adjunctive tool to the modified barium swallow study. Since 1968, the flexible laryngoscope has been available to physicians to visualize the structures of the larynx. In 1986, speech pathologist Dr. Susan Langmore initially described a modification of the traditional laryngoscopic examination that focused specifically on the pharyngeal phase of swallowing where aspiration occurs. This examination of dysphagia using the laryngoscope is called flexible endoscopic examination of swallowing (FEES).

FEES is performed by passing a small, flexible scope through the nostril and into the pharynx which provides a clear, direct view of the upper airway and many of the structures involved in swallowing. With this technique, the patient's ability to handle his own secretions or real food substances can be studied without interfering with the function of the structures.

The FEES procedure is not intended to replace the modified barium swallow study as it focuses only on the pharyngeal stage of swallowing and does not demonstrate the complex, sequential interaction between all three stages of swallowing. However, FEES is a valuable dysphagia diagnosis and management tool with many clinical applications. The technology provides useful information when facilities, personnel or patient status prohibit radiographic examination. FEES also is an alternative to the MBS in patients requiring immediate examination or more frequent re-examination thus eliminating the concern of radiation exposure. Most importantly, FEES is an excellent biofeedback tool which can assist the patient and clinician in the retraining of laryngeal and pharyngeal musculature for swallowing. Finally, FEES technology is an excellent educational tool as it assists members of the multidisciplinary dysphagia team, patients and families to better understand the nature, severity and components of the patients swallowing disorder.

Clearly, FEES is an invaluable diagnostic and clinical tool in identifying and managing aspiration in patients with oropharyngeal dysphagia. It also is an excellent adjunct and/or alternative to the modified barium swallow study which remains the "gold standard" method of assessing oropharyngeal dysphagia.

### Bizarre Swallowing (Part I)

Turning to bizarre swallowing, it is quite extraordinary the amazing variety of objects people swallow. There are a number of reasons for this; financial gain, an obsession or an act in the music hall, circus, fairground and, more recently, television.

These have ranged from enormous amounts of food and drink, to all sorts of metal objects—keys, rings, pieces of chain, razor blades; progressing to electric light bulbs, chewing up drinking glasses and finally live creatures such as gold fish, mice, frogs and—the mind boggles—even white rats.

Stone eaters or lithophagous' have been known since the Medieval ages. As recorded in The Book of Wonderful Characters-Memoirs and Anecdotes, 1869, they swallow flints of impressive size, but stones which can be reduced to powder, such as marble, pebbles etc., were made up into a paste providing a most agreeable and wholesome food. The pre-requisites for this diet includes, 'a gullet very large, teeth exceedingly strong, saliva very corrosive and a stomach lower than ordinary'.

---

**New Innovative Technology for the Diagnosis and Management of Swallowing Disorders**

**Paula A. Sullivan, M.S., CCC-SLP**
Manager, Rehabilitative Services, Moffitt Cancer Center
Apart from food and drink, there is in the trade a very definite distinction between the amateur and the professional, the former merely swallowing inanimate things and hoping that his digestion will cope, and the latter, who has learnt the act of re-gurgitation, bringing up the object under strict control.

However, the two most celebrated arts of swallowing are fire eating and sword swallowing.

Fire Eating

Fire eating is an ancient practice, Peruvian priests having used it to cast spells, sorcerers were ironically burnt at the stake in the medieval ages for doing it and to this day witch doctors still indulge in the practice. Apart from plunging a burning taper into the mouth, there are several variations to the routine, with the real experts inserting anything up to four torches simultaneously. Some of the burning petrol can be wiped off the tongue, another torch is then taken and lit from the flames coming out of the mouth. Petrol fumes can be deliberately swallowed, puffed out and then set fire to as they hit the air, an effect called ‘the human candelabra’ with pretty fountains of flames.

The torch is a piece of wire with a handle at one end and a hook at the other. A strip of rag wadded into a ball is fixed onto the hook, soaked in petrol and when lit produces a flame about two feet in length. As far as technique is concerned, there is no alternative to many weeks of practice. Fire eaters in training have to accept that they will get burnt with considerable blistering of the mouth and lips. These must be wet, the torch held at the correct angle of about 75 degrees, the head pushed right back and it is rather important, as can well be imagined, to breathe out at the correct time.

There are conflicting theories about the physical principles that make fire eating possible. It may be that the extreme heat of the flame coming suddenly into contact with the moisture inside the mouth creates a thin layer of steam which acts as insulation. Interestingly enough, fire eaters never seem to get secondary infection of the mouth, petrol possibly being a good antiseptic.

It is useless coating the inside of the mouth with a chemical that resists heat, it merely poisons the person and anyway, as any good performer will tell you grandly ‘fire eating is an art’, and underhand tricks must not be used.


A Complete History is Essential
Janet Jones, B.A., CGC
Patient Care Coordinator

Your medical history is the most important piece of information you can give your doctor to help diagnose and treat your swallowing problem. The more accurately you can describe your symptoms and how they have affected you, the more efficiently your doctor can begin the appropriate testing and treatment indicated.

Your complete medical history is important as many swallowing disorders are related to other medical conditions. (Scleroderma, for example, is a collagen vascular disease which can affect swallowing. A stroke can affect the muscles that control swallowing).

Many medications can cause or contribute to difficulty swallowing. You should know what medications you take, their strength, how often you take them and what you take them for. You should also mention any non-prescription medications and vitamins you take on a regular basis. It is a good idea to carry an updated list with you at all times and make it available to each new doctor you see.

As part of your swallowing history, the doctor will need to know what type of foods you are currently tolerating. Try to remember when you first had to start altering your diet to accommodate your swallowing difficulty. A diary of food and liquid intake for 2 to 3 days can be helpful in determining if you are taking in enough calories.

The doctor also needs to be made aware of any social situations which may affect your care. For example, do you live alone? Is transportation to appointments a problem? Do you prepare your own meals? Do you have difficulty obtaining necessary medications or medical supplies?

You will be asked to provide all records and x-rays pertaining to your swallowing problem from other doctors. A recent history and physical examination from your primary care physician will provide a baseline of your general health.

When time permits, it is helpful to mail or fax this information in advance of your initial appointment. The actual x-ray films should be hand-carried to your initial appointment.

You should be prepared to ask any questions you may have about why you have been referred to this specialty and realistically discuss expectations with the doctor. Sometimes you are referred by another specialist who has not been able to help you and wants an expert second opinion, confirming that all available avenues of treatment have been explored.

For patients who are winter residents only, it is very important to provide your doctor here with an update on your general health. New medications or changes in medications and reports of diagnostic tests and/or procedures which were performed elsewhere need to be provided. For example, if you were started on an anticoagulant (coumadin or heparin) or take aspirin on a regular basis, these medications commonly need to be stopped prior to many procedures. Your doctor will need to know this in advance of scheduling you for a procedure.

Things To Remember

1. OFFICE HOURS: 8:30 a.m. till 4:30 p.m. Monday through Friday.

   Our office is closed on weekends so it is important to make sure any medication refills are called to us during our regular office hours.

   Also, our emergency telephone number for after hours is (813) 974-2201. Please remember these calls will be responded to by one of our gastroenterology residents who will in turn contact the appropriate attending physician on call.

2. BILLING: Individuals who may have any problems with their accounts should contact the Patient Relations Department of the University of South Florida Medical Clinics at (813) 974-3573 between the hours of 10:00 a.m. till 4:00 p.m. Monday through Friday. For those patients who are from out-of-town, a new toll-free number has been added for you to call with billing questions. The number is 1-800-933-8672. This number is for calls originating in Florida and is only for billing question.

3. DILATIONS: For our patients who receive periodic esophageal dilations: Please try to anticipate and contact our office at least 2 to 3 weeks in advance of your need for dilation if at all possible. We have been having to schedule routine cases 3 to 4 weeks in advance due to our heavy patient load. We do not want any of you to suffer unnecessarily, so please help us with your appointment needs.
During the past six months, members of the Center for Swallowing Disorders staff have continued their active participation in graduate medical education at regional, national, and international meetings. These presentations on topics related to swallowing disorders require considerable research and time to prepare teaching slides and videotapes. Contributions to the medical literature in journals and textbooks also have been significant.

Lecture Presentations by CSD Staff

6. March 17, 1994: Esophageal Cancer - The Role of the Gastroenterologist in Diagnosis and Treatment. Visiting Professor, University of Alabama G.I. Grand Rounds, Birmingham, AL. (Boyce)
7. March 24, 1994: Clinical Esophagology for Internists. USF Internal Medicine Grand Rounds. USF College of Medicine, Tampa, FL. (Boyce)
9. May 6, 1994: Medical Grand Rounds, Barrett Esophagus. Sarasota Memorial Hospital, Sarasota, FL. (Boyce)

Contributions To Medical Literature