After 18 years of instrument development and a similar period of clinical use, the technique of endosonography (ES) has become well established as an accurate diagnostic method. ES was first performed in the Southeastern United States in 1987 by physicians at the USF Center for Swallowing Disorders. We had the good fortune of being one of the first five institutions in the United States to perform ES and have continued to do so for eleven years.

The combination of visual inspection capability for mucosal surface changes plus the capacity to "electronically dissect" the wall of esophagus, stomach, rectum and adjacent organs by ultrasonic imaging provides unique information for diagnosis and planning therapy. Standard endoscopy and ultrasonography capabilities are combined in the endosonography equipment, which incorporates an ultrasonic transducer into the distal tip of a videendoscope. This endoscope permits video observation of the inner mucosal surface of the wall of the GI tract (esophagus, stomach, duodenum, or rectum), as well as the sonographic imaging of the other three layers of submucosa, muscularis propria, and serosa or adventitia of the esophagus (fig 1).

The major role of ES is the staging (determining the extent of local cancer invasion) of cancers of the gastrointestinal tract and pancreas. Staging of a cancer using the complementary method of ES and computed tomography (CT) scans provides physicians with accurate information on the local invasion (by ES) and distant invasion (by CT) that is changing the approach to cancer therapy. One of the most valuable results of ES and CT staging is a reduction in the number of needless "open and close" surgical procedures that occur in patients who have inoperable spread of esophageal cancer. In some patients, ES can detect cancer recurrence before symptoms have developed. ES also is capable of confirming the limitation of cancer to the most superficial (mucosal) layer, thereby permitting nonsurgical therapy in some patients.

ES also is indicated for examination of lesions beneath the surface of the inner lining (mucosa) of the gut. Such nodules, large folds or masses are commonly found incidentally during endoscopy for other reasons and are not accessible to standard biopsy. ES can determine whether the mass is due to large blood vessels (varices), benign tumors of muscle (fig 2), fat or connective tissue, enteric cysts, compression of the gut wall by an adjacent mass, as well as detecting changes suggestive of subsurface malignancy. In many cases, the benign appearing lesions can be safely followed periodically without having to resort to surgical removal. In others, echogenic features otherwise unsuspected may point to malignant changes which indicate prompt removal with a better chance for cure.

The relatively high frequency ultrasound (7.5 – 12MHz) possible with the Olympus GF-UM20 endoscope with a 360 degree sector scanner, provides excellent imaging of the multi-layered wall structure of the esophagus, stomach, duodenum and rectum. It can determine penetration of cancer into the various layers, adjacent organs, and especially periesophageal, gastric, and rectal lymph nodes. The new TNM system (1992) has been accepted as the best staging classification to use. The staging accuracy of ES is greatest for depth of tumor invasion (T), of intermediate accuracy, but superior to CT for lymph node involvement (N), and is inadequate for assessment of distant metastases (M), which are best evaluated by CT. The accuracy of ES for T and N staging is significantly more accurate than CT, while CT is clearly superior for M staging. Consequently, these methods are complementary and when used together have an overall staging accuracy of over 85%. The high frequencies used give excellent definition of the wall layers but the range of effective ultrasound transmission is limited, thereby explaining why the technique is of little value in determination of distant metastases.

(continued)
Endoscopic ultrasonography offers the best delineation of small masses or tumors arising in the wall of the GI tract or pancreas that typically are not clearly visible with CT or other external imaging modalities. Adjacent organ and lymph node involvement can be assessed by virtue of this instrument's ability to transmit ultrasound and receive images through the five-layer gut wall, thus permitting examination of surrounding tissue and vascular structures.

Adequate ES imaging requires proper acoustic coupling between the endoscope and the tissue. For this purpose, a balloon attached to the tip of an ultrasonic endoscope is filled with water or emptied as needed to provide optimal acoustic contact and proper focal distance. In this way, the operator can image the target area in closer detail when examining the GI wall. Other techniques used to enhance ES imaging include direct contact of the transducer tip with target organ and immersion of the examination site by the injection of deareated water directly into the lumen around the endoscope. The immersion technique requires about 300 to 500 ml of water for examination of the stomach and somewhat less for the esophagus, duodenum, or rectum. Air between the transducer and the tissue being examined produces imaging artifacts that prevent an adequate examination.

Cancer staging by the TNM system is by far the most important use of ES. Patient outcome is significantly affected if a lesion -- esophageal, gastric, duodenal; ampullary; pancreatic, or rectal - can be accurately determined to be a T1, T2, T3, or T4 lesion (fig 3). Distinguishing between a T2 and T3 lesion can establish whether surgery alone is expected to be curative or whether adjunctive chemotherapy and radiation are necessary as well. Accurate staging is important in planning the type of operative procedure, which may be less extensive for a T1 or T2 lesion, than for a T3 lesion. Surgical removal of mucosal and T1 cancers is currently the standard therapy, but for such early lesions without evidence of spread, the alternative treatments of endoscopic mucosal resection and photodynamic laser therapy (PDT) are under investigation. These non-surgical treatment options are promising for patients who are at high risk for surgery because they have other serious diseases.

The development of small transendoscopic echo probes will facilitate the use of ES through narrow/stenotic GI lumens and in the pancreatic-biliary system. The introduction of improved linear array imaging and aspiration needles, now permits ES-guided cytology sampling of submucosal lesions and lymph nodes outside the esophageal wall. All of these refinements improve diagnostic accuracy and result in fewer unnecessary operations for advanced malignancies.

Non-operative methods offer the best approach to staging malignant lesions of the esophagus and stomach. It must be remembered that the goal of staging is as much to determine inoperability as operability. Because of the known poor prognosis of cancer of the esophagus, stomach, and pancreas, accurate staging will benefit a majority of patients by providing data to support a nonsurgical approach to palliative therapy. It is essential, for sound medical reasons, to avoid needless surgical operations simply to either stage a malignant condition as unresectable for cure or even resectable for palliation, when equally good non-operative palliation may be available. On the other hand, the patients who are found to have cancer limited to the wall, T1, T2, T3 NO, MO lesions can be advised with more confidence regarding the potential benefits of surgery, with or without radiation and chemotherapy, than otherwise would be possible. Accurate staging of lesions favorable for curative surgery allows proper counseling regarding risks and benefits, especially in patients who may have significant concomitant illness that makes the decision on operability difficult.

Early stage disease can be distinguished readily from advanced carcinoma. Tio reports the overall accuracy of ES is about 90% for assessment of esophageal carcinoma. If not adequately dilated beforehand, esophageal stenosis has been a limiting factor for accurate staging, preventing a complete examination in from 16-62% of cases. There is a remarkable discrepancy between ES and CT in the assessment of regional lymph node involvement with an overall accuracy of ES of 80% compared to 51% for CT.

Although there are problems with ES accuracy in some aspects of staging gastrointestinal carcinoma, it is recognized as an essential procedure in the TNM staging of these malignancies. The strengths of ES include its ability to accurately determine the depth of wall infiltration and detection of metastatic involvement of regional lymph nodes. Its weaknesses are due to inability to identify distant metastases and in the differentiation between inflammation and malignant infiltration. Lymph node size is of little help since many reports indicate that histopathology of large nodes often is due to inflammation and small nodes may contain micrometastases. Overstaging by ES may occur due to inflammation and preoperative radiation effects on the tissues in and around cancers. Post therapy restaging is of little value unless a pre-therapy ES has been performed and is available for comparison. Ulcerating carcinomas tend to be associated with the most inflammation, an hence are often overstaged. The most obvious case of understaging is the inability to do a complete examination because the instrument cannot be passed through an esophageal stricture. Proper dilation in one or more sessions prior to the scheduled ES will overcome this hindrance in most patients.

ES has proven to be an accurate method for staging esophageal cancer and should be used in conjunction with CT as a component of all staging protocols. The value of combining ES and CT for staging esophageal cancer has been reported to have a higher accuracy rate of 86% in TNM staging than when either modality is used alone. A recent consensus conference recommended that CT scan be used first to confirm or exclude distant metastases. If no distant metastases are identified, ES should be used for local T and N staging to complete the staging process.
INTRODUCTION

Myasthenia gravis (MG) is a neuromuscular disorder or condition that involves muscles and nerves. It is characterized by severe weakness and easy fatigability of skeletal muscles (muscles involved in voluntary actions). The main underlying defect is a decrease in the number of receptors for a chemical substance called acetylcholine. These receptors are located at nerve endings or neuromuscular junctions, and their deficit is secondary to destruction or inactivation by antibodies from the person’s own autoimmune system.

MG is most common in young women. It may be present in patients age 30 or older who are found to have thymoma (tumor of the thymus gland). This gland is situated in the upper anterior chest cavity and plays a role in immune or gradual and steady onset but may be unmasked by a coincidental recent infection.

MG can be worsened by symptoms of neurologic or oropharyngeal dysphagia (swallowing difficulty). In oropharyngeal dysphagia, the patient recognizes the usual food accumulation in the mouth, but control of the food bolus within the oral cavity can be difficult due to inability or a limited capacity to initiate a swallowing response. Food, especially liquids, can be aspirated into the airway. Other symptoms such as nasal regurgitation, drooling, and weight loss can be present as well.

CLINICAL FEATURES

Patients may present with complaints of double vision, droopy eyelids, and inability to stay alert, difficulty chewing or swallowing, as well as weakness in the extremities, or a combination of these. Symptoms often fluctuate in intensity during the day. Sustained activity of the affected muscles increase the already established weakness, but some improvement is usually experienced after a rest period. In a study from Carpenter and colleagues, 38 of 175 patients with MG suffered from swallowing difficulty as their primary disease manifestation. Swallowing worsens near the end of the day and at the end of every meal. In some cases, the patient may have to close the jaw by hand in order to chew. The movements of the tongue become slow and progressively less efficient with repeated unsuccessful attempts at swallowing, resulting in frustration as well as progressive weight loss.

Swallowing difficulty can be complicated by drooling, choking, and liquid regurgitation with development of nasal speech or hoarseness. With facial weakness, the lips are affected. Tongue weakness slows articulation or the ability to integrate words during a conversation, resulting in poorly understood speech. The laryngeal muscles are involved, and this results in poor function of the vocal cords and shortening of sentences in a progressive fashion.

DIAGNOSIS

The diagnosis can be confirmed by the rapid response to a short acting drug, edrophonium (Tensilon) given intravenously. This drug inhibits the action of an enzyme (acetylcholinesterase) that normally inactivates any residual acetylcholine necessary to allow muscle contraction. As a result of this effect, there is a transient improvement in muscle strength. The detection of antibodies to acetylcholine is a diagnostic of the disease. X-rays or CT scan of the chest are needed to exclude the possibility of a tumor of the thymus gland, which may be associated in some patients. A special x-ray study known as dynamic video esophagram (DVE) performed with assistance of a speech pathologist can be helpful, allowing visualization of difficulty swallowing as well as oropharyngeal food retention and possible aspiration. Weak contractions of the muscles of swallowing also can be documented by a special test, i.e. pharyngoesophageal manometry.

TREATMENT

Swallowing complication related to MG can be improved by treating the underlying disorder. Medication such as pyridostigmine (Mestinon) usually is helpful in providing symptomatic relief. Surgical resection of an enlarged thymus gland can be of benefit as well. The speech pathologist with expertise in swallowing difficulty, can instruct the patient and family in techniques to modify swallowing and control food aspiration. Appropriate postural modification, swallowing maneuvers, oral exercises to strengthen lip and tongue movement as well as diet modification, are imperative in the treatment of patients with swallowing difficulty related to MG.

This disorder requires a combined approach to evaluation and treatment. The internist, neurologist, gastroenterologist, otolaryngologist, and speech therapist are all involved in a successful clinical approach to this medical problem. (See following discussion in this issue by Joy Gaziano, Speech Language Pathologist)
SPEECH AND SWALLOWING DISORDERS RELATED TO MYASTHENIA GRAVIS
Joy Gaziano, M.A., CCC/SLP

Myasthenia gravis (MG) is a neurological disorder that causes impaired nerve conduction at the myoneural junction of strained muscles. Since the oral and pharyngeal stages of swallowing are controlled by striated muscles, swallowing disorders may result. A hallmark of this disease is a tendency of muscles to fatigue after repeated exercise. Therefore, during mealtime, chewing and swallowing may be normal initially, but as the muscles of mastication (chewing) and swallowing fatigue, there may be a complete loss of swallowing ability. Dysphagia (difficulty swallowing) may be an early sign of MG and may be accompanied by ocular (eye) muscle fatigue.

When viewed by an x-ray swallowing study, videofluoroscopy or dynamic video (DVE), certain characteristic findings help with the differential diagnosis of MG. Tongue movements are slow and weak and continue to weaken with additional swallowing attempts. Holding solid foods on the tongue is particularly difficult. Because the tongue fails to move the food briskly into the pharynx, food residue may coat the oropharynx. This residue, if not adequately cleared, may be aspirated into the lungs. Repeated swallowing attempts may result in nasal regurgitation because the soft palate is not sufficiently strong to elevate and close off the nasal passage during the swallow.

About one third of patients with MG have significant swallowing problems due to fatigue when chewing. Because fatigue plays such an important role, patients generally do best at the beginning of meals and tire as the meal progresses. Therefore, it is recommended that patients consume several smaller meals during the day. Since swallowing worsens as the day progresses, it is advised to eat larger meals accounting for the greater part of nutrition in the morning. Additionally, patients should conserve energy and limit physical activity before mealtime. Even excessive talking before meals can result in more rapid fatigue since the speech musculature (the tongue, lips, jaw, soft palate and vocal folds) are also critical for swallowing. Since the tongue has difficulty holding solid food boluses in the mouth, foods that are cohesive like pudding or tuna salad are easier to manage than foods that fall apart like rice and cereal. They require less tongue activity and generally reduce tongue fatigue.

Carpenter, et al. (1979) found that 30 percent of persons with MG experienced oral pharyngeal or laryngeal complaints. Most common complaints were swallowing problems, followed by dysarthria (speech disturbance) and dysphonia (voice disturbance). Speech and voice changes in myasthenia are also characterized by their progression and increase in severity with prolonged speaking. As a person continues speaking, fatigue of the speech muscles may cause increase in hypernasality, deterioration of articulation, reduced loudness and changes in voice quality.

Speech is responsive to the Tensilon test (edrophonium chloride) for MG. After deterioration of speech occurs, injection of the drug produces 30 to 60 seconds of marked improvement in all dimensions of speech production. This drug is for diagnostic purposes only and symptoms return within minutes. Drug therapy with Mestinon (pyridostigmine) however, has been shown to greatly improve muscle function. Such anticholinesterase-producing drugs in proper dosage facilitate speech and swallowing in most patients.

CONTINUING MEDICAL EDUCATION
During the past several months members of the Joy McCann Culverhouse Center for Swallowing Disorders staff have continued their active participation in graduate medical education at regional, national, and international meetings and by contributions to the medical literature.

Lecture Presentations by CSD Staff
4. September 4-11, 1998: Session Moderator: World Congress of Gastroenterology: Vienna, Austria. (Boyce)

Contributions To Medical Literature and Clinical Research Abstracts