



NEWS

SWALLOWING

VOLUME 15

DECEMBER 2003

NUMBER 2

DIRECTOR'S FORUM

CONGENITAL ESOPHAGEAL STENOSIS A.K.A. "RINGED" ESOPHAGUS

H. Worth Boyce, M.D.

Professor of Medicine and Director

True congenital esophageal stenosis has been considered one of the least common congenital esophagotracheal anomalies. Its occurrence has been estimated at less than 1 in 25,000 births. It is considered a *forme fruste* (a mild or incomplete form) of esophageal atresia (absence or closure of a tubular structure) that is present at birth and the diagnosis is often missed by physicians. Case reports in the literature describe the diagnosis in infants and children, often at the time of introduction of solid foods into their diet, but rarely in adolescence and adults. The typical clinical, radiographic and endoscopic features permit an accurate diagnosis. Barium x-rays of the esophagus typically reveal multiple rings along the area of stricture, usually in the mid-esophagus. However, in some patients multiple areas of the entire esophagus may be involved. Gradual dilation under fluoroscopic control usually provides safe and effective therapy.

Over the past two decades, the confirmation of this diagnosis has become less clear as many cases with similar esophageal abnormalities are being recognized with symptom onset in adulthood. Another descriptive name, "ringed" esophagus, has been introduced to describe its most obvious characteristic, i.e. multiple rings producing a mid-esophageal stricture. This condition has been shown to also be related to food allergies, eosinophilic esophagitis, and in some cases abnormal degrees of gastroesophageal acid reflux. These multiple potential etiologies support the notion that this is an entity that can be acquired. The current challenge for those interested in this disorder is to study these multiple factors and determine the pathogenesis (origin, tissue abnormalities and natural history) of this fascinating condition. Even more challenging is the need to determine why its prevalence has been clearly increasing over the past two decades.

Our experience over the last 17 years includes 48 patients (33 males, 15 females) with this condition. The patients' average age at the onset of symptoms was in the latter part of the second decade, while the age at diagnosis was over 5 years later. All patients experienced symptoms of dysphagia to solids and most had some degree of esophageal solid bolus impaction, several requiring endoscopic removal. No patients experienced dysphagia to liquids as a primary complaint. Heartburn is reported by almost one-fourth of the patients.

Most patients report prolonged eating times and typically are the last family member to complete a meal. Weight loss is rare since most are able to compensate on caloric intake by gradually reducing diet consistency from solid, to soft, to pudding consistency plus liquids.

The past medical histories of our patients were significant for an increased frequency of asthma and suspected or documented food allergies.

Proper initial evaluation of these patients should include a complete history and physical examination, a barium esophagram, with a bolus challenge if needed, and endoscopy with biopsy at the stricture. The typical appearances of concentric and/or incomplete rings and luminal narrowing are shown in Figure 1. The characteristic endoscopic appearance of the rings is shown in Figure 2.



Figure 1. The typical appearances of concentric and/or incomplete rings and luminal narrowing.

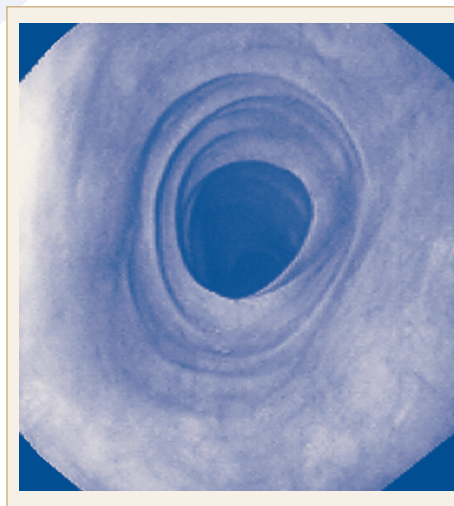


Figure 2. The characteristic endoscopic appearance of the rings.

In our 48 patients, the diagnosis was not made until the adolescent or young adult years in all but one 8-year old male, despite many patients complaining of dysphagia symptoms since early childhood. The characteristic endoscopic finding of a mid to distal esophageal stricture with submucosal rings was seen in all patients. Some patients had skip areas of rings producing several strictures but rarely had evidence of only distal submucosal rings.

The appearance of these rings can be subtle by barium esophagram and esophagoscopy. Adequate lumen distention with barium or air may be necessary to completely visualize the strictured segment. A solid bolus challenge with a barium tablet confirmed the location of stenosis in many patients. In some patients, the narrowing is tubular without apparent ring formation.

In recent years, the “ringed” esophagus has been noted in some patients to be associated with gastroesophageal acid reflux, some with food allergies and in some with both conditions. This multifactorial causation has led to re-evaluation of the initial belief that all cases were congenital. The observation of an increasing number of cases in adults without dysphagia in childhood or adolescence certainly supports the notion that in some this condition is acquired. A strong association, especially in children, with food allergies, asthma, and eosinophilic esophagitis in the absence of documented acid reflux, and their response to oral or topical steroid therapy, supports the likelihood of an allergic etiology in some. In others, the finding of abnormal esophageal acid reflux and symptomatic improvement on acid suppressing drugs suggests a role for acid injury in causation.

Patients are best treated with gradual dilations, under fluoroscopic control, over several sessions. No more than 3 sizes of dilator in increments of 1 mm were used at any one session. The initial dilator size is often less than 10 mm. The end point at each session was either the passage of up to 3 progressive sizes of dilator or moderate resistance in the passage of any dilator. These strictures typically offer mild resistance and as a consequence, there is a tendency for the dilator size to be increased too rapidly, thereby increasing the risk of pain or

perforation. There have been no esophageal perforations and rare post dilation discomfort using this treatment schedule in our patients.

This approach provides good to excellent improvement in most patients. The interval between initial dilation sessions was usually 3-4 weeks. We recommend the following as end-points for each dilation session: patient discomfort (e.g. chest pain), moderate or severe resistance to the passage of any one dilator, or the passage of three dilators of sequential 1 mm size increments. Dilation should be approached with the same caution as pill-induced or caustic ingestion strictures. These ring strictures can be more rigid than the usual peptic stricture, but most often are dilated with little resistance. Rapid dilation may place the patient at greater risk of perforation.

The prevalence of congenital esophageal stenosis or “ringed” esophagus in the adolescent or young adult is more frequent than previously thought and is increasing in recent years. This diagnosis should be considered in all patients with a long history of dysphagia to solids and evidence of mid to distal esophageal stricture of either “ringed” or tubular type. We feel the characteristic concentric, submucosal rings in the mid-esophageal segment and a long history of dysphagia to solids and/or prolonged eating times since childhood strongly support this diagnosis. Gradual dilation affords safe, effective treatment of this fascinating esophageal disorder of uncertain etiology. A complete history for food allergy should be obtained and when eosinophilic esophagitis is confirmed by biopsy, appropriate topical steroid therapy has been reported helpful and should be considered. In some patients, acid reflux is associated and this requires a 24-hour ambulatory pH study for confirmation. Acid suppression therapy is appropriate when acid reflux is documented.

ENDOSCOPIC THERAPY OF GASTROESOPHAGEAL REFLUX DISEASE: BENEFITS, RISKS AND SUCCESS

Milton C. Johnson, M.D.
Associate Professor of Medicine

Unique clinically applicable endoscopic techniques have been developed to treat gastroesophageal reflux (GER). An enormous amount of enthusiasm has been generated in support of these new endoscopic therapies as alternative treatments for patients with symptomatic gastroesophageal reflux disease (GERD). In general, the various endoscopic anti-reflux therapies prevent GER by their “unique” ability to alter the structure of the gastroesophageal junction (GEJ). The anatomy of the GEJ includes the lower esophageal sphincter (LES: 3-4 cm length), the crural diaphragm (muscles for breathing that divide the chest from the abdomen) and the sling fibers of the stomach (portion of stomach that extends around the esophagus or food tube). These three components are the essential elements for an effective anti-reflux barrier. The LES is however the principal barrier to prevent GER. The other two components strengthen the effect of the LES. Endoscopic therapies aim to improve one or more of these components to prevent the results of gastric contents after meals and during various body positions, e.g. bending, stooping and sleeping.

Randomized, controlled, clinical trials (studies) have not demonstrated the superiority of one of the current Endoscopic Anti-Reflux Therapies (EARTHs) over another. In fact, most of the EARTHs remain under investigation. The current literature pertaining to the effectiveness and safety of EARTHs is not strongly supportive of their use in most individuals with GERD. The majority of individuals enrolled in clinical trials that evaluate EARTHs are classified as having “mild GERD,” i.e., disease that is responsive to

standard proton pump inhibitor therapy (Aciphex, Nexium, Prevacid, Prilosec, Protonix) rather than histamine-2 receptor antagonist therapy (Axid, Pepcid, Tagamet, Zantac). Although the various endoscopic anti-reflux therapies have relieved symptoms in some individuals, other individuals have experienced severe complications. Clinical trials are required that are better designed to inform us of the “true” benefits and risks associated with EARTHs. An intermediate role, at best, may be the only indication found for EARTHs once adequate studies have investigated their use.

I will briefly review the EARTHs currently undergoing investigation. It is important to note that several different companies may market the same EARTH by using a similar principal of therapy, but a different instrument design or technique of applying therapy. The data on the “true” costs of these procedures is not known since this number will involve many cost factors.

One type of endoscopic anti-reflux therapy involves the use of radiofrequency (RF) energy (similar to radio, cellular or microwaves) waves. The most well studied device is the Stretta (Curron Medical, Inc., Sunnyvale, CA). RF energy is delivered into the tissue of the lower esophagus (food tube) by a probe that is capable of delivering thermal (heat) energy below the surface cells. The normal neural (local nervous system) network is altered, thereby decreasing the spontaneous LES relaxations (thought to be important cause of GER in most individuals). A tissue-tightening effect also results from delivery of RF energy to the lower esophagus in the region of the LES.

A second type of endoscopic anti-reflux therapy is referred to as suture plication (gastroplasty). The most well known device is the Endocinch (Bard Interventional Endoscopic Suturing System: Billerica, MA). Simply, the technique involves the placement of suture material or stitches into top of the stomach folds or creases in the region of the GEJ. A unique technique of sewing has made this technique possible during endoscopy. This suture plication method tightens the region of the GEJ and strengthens the LES contractions after meals and change of body positions.

The third type of EARTH requires the implantation (injection) of Plexiglas microspheres (manufactured beads) into the lower esophagus region of the LES. One specific type of microsphere is composed of a substance called polymethylmethacrylate or PMMA. The PMMA microspheres are 10 mm in diameter (less than one-third of an inch). They are mixed into a gelatin solution (3 ml prepared syringe for injection) and injected beneath the lining cells in the top portion of the LES region. A mean volume of 32 ml (range: 24-39 ml or about 1 ounce) is injected per patient per treatment session. The submucosa layer of the esophageal wall is the intended recipient of the Plexiglas microspheres gelatin solution. This implantation method creates a "bulking effect" or "sandbag effect" and reduces spontaneous complete LES relaxation events when properly performed.

The United States Food and Drug Administration (FDA) Center for Devices and Radiological Health has ruled that the two endoscopic anti-reflux therapies involving the delivery of RF energy (Stretta) and suture plication gastroplasty (Endocinch) were safe for use in human subjects. The Agency has not yet ruled on the effectiveness of these two EARTHS for treating GERD. Since the initial marketing of the Stretta 30 French RF device in May 2000, the FDA has been provided information concerning eight esophageal perforations (local rupturing or bursting) and two patient deaths that occurred during or following the use of the RF device. A data summary reveals that 45% to 49% of individuals undergoing EARTH either by the RF energy method or suture plication (gastroplasty) had incomplete relief of GERD symptoms. These individuals required continuation of medical therapy (acid suppression medications) or were referred for anti-reflux surgical therapy.

The clinical trial data provided to the FDA involving the use of the implantation method of EARTH appeared more optimistic. Sixty to seventy percent of individuals undergoing the Plexiglas microsphere implantation therapy reported discontinuation of all acid suppression medications by seven months following therapy. There remain questions concerning the accurate injection of the PMMA microspheres into the top of the LES region. The recent use of a new microsphere containing ethylene vinyl alcohol polymer (EVAP) has enabled researchers to inject it more accurately by the aid of fluoroscopy (x-ray guidance demonstrating site of injection). Unfortunately, this new method requires more time to perform.

On average the endoscopic anti-reflux therapy methods involving the delivery of RF energy, suture plication (gastroplasty), and injection of EVAP require 60 minutes to 90 minutes to complete. Although all three EARTH methods have been FDA approved for safety in human subjects, the long-term effectiveness of their usage has not been proven. EARTHS are not recommended in individuals with a hiatus hernia. The final outcome of EARTH methods will be decided after more clinical trials have provided stronger evidence of their safety, cost-saving benefit, and control of GERD symptoms in individuals with mild and moderate-severe GERD and esophagitis (inflammation of the food tube).

In summary, the proven, current, best anti-reflux therapy for the majority of individuals with GERD symptoms includes medical (maximal use of proton pump inhibitors) and surgical (laparoscopic) anti-reflux therapies. Individuals with persistent symptoms proven to be GERD-related and who are candidates for surgery have responded well to surgical anti-reflux therapy. Endoscopic anti-reflux therapies may be viewed as alternative therapy for GERD, but they have not proven themselves effective in the majority of individuals undergoing therapy. New EARTHS are being developed each year despite the limited data available to support their use. At present, EARTHS should not be thought of as a therapeutic alternative to standard medical or surgical therapy. Additional randomized, clinical, controlled trials are necessary to increase our understanding of which patients are most likely to benefit and to prove the benefit of endoscopic anti-reflux therapies before they can be recommended. The viewpoint of many experts who have followed the development and marketing of all of the endoscopic anti-reflux therapies is expressed concisely by the following quote from an anonymous expert who states, "We are not yet close to the ideal EARTH, but many are working fast and furious to convince us otherwise!"

5th ANNUAL

UPDATE FOR CLINICIANS

*New Horizons in Management of
Esophageal and Gastric Disorders
December 4-6, 2003*

*Location: Boardwalk Inn, Walt Disney World
Orlando, FL*

For further information contact: University of South Florida
Office of Continuing Professional Education, P.O. Box 550610
Tampa, FL 33655-0610 Or Fax to: (813) 974-3217

Sarah A. Garza, ARNP

The Swallowing Center is pleased to announce the addition of Sarah A. Garza as a member of our medical staff. She received her Bachelor of Science degree and a Master of Science degree in nursing from Vanderbilt University. She also has received national certification as an Advanced Registered Nurse Practitioner. Sarah has worked with a graduate of our Gastroenterology Residency program in private practice in Nashville, Tennessee for 5 years. She will be assisting Drs. Johnson and Boyce in their tertiary level consultation practices and also will manage our Esophageal Physiology Laboratory. Sarah's husband, Austin, currently is an Advanced Subspecialty Resident in Gastroenterology with the University of South Florida College of Medicine Division of Digestive Diseases and Nutrition.

CONTINUING MEDICAL EDUCATION

The Center for Swallowing Disorders has continued active participation in graduate medical education by lectures at regional, national and international meetings and by contributions to the medical literature.

Lecture Presentations by CSD Staff

June 5, 2003: *Diagnosis of Dysphagia.* Dothan, AL (Boyce)

June 6, 2003: *Management of Complex Esophageal Strictures.* Valdosta, GA (Boyce)

July 24, 2003: *Grand Rounds Lecture Sun Coast Hospital: Dysphagia – Clinical Clues and Caveats.* Largo, FL (Boyce)

August 6-7, 2003: *Classification of Esophageal Strictures as a Guide to Therapy.* Baton Rouge, LA (Boyce)

August 21-24, 2003: *Medical College of Wisconsin. Esophageal and Aerodigestive Tract Disorders: Real World Challenges: Defiant and Malignant Strictures.* Milwaukee, WI (Boyce)

September 3-5, 2003: *The Obstructed Esophagus: Clinical Clues and Caveats.* Fayetteville, GA (Boyce)

September 20, 2003: *Annual Meeting of Florida SGNA Regional Societies. Adventures in the Esophagus and the Endoscopy Suite.* St. Augustine, FL (Boyce)

September 29, 2003: *Visiting Professor, University of Alabama Birmingham, Lecture: Normal Anatomy and Disorders of the Esophagogastric Junction.* Birmingham, AL (Boyce)

Contributions To Medical Literature

Boyce HW: Barrett esophagus: Endoscopic findings and what to biopsy. *J Clin Gastro* 2003;36(suppl. 1):S6-S18.

Bloomston, M, Fraiji E, Boyce HW, Gonzalvo A, Johnson MC, Rosemurgy A: Preoperative Intervention Does Not Affect Esophageal Muscle Histology or Patient Outcomes in Patients Undergoing Laparoscopic Heller Myotomy. *Jnl Gastrointest Surg* 2003;7(2):181-190.

*This edition of Swallowing News was supported
by an educational grant from Janssen Pharmaceutica.*



Joy McCann Culverhouse
Center for Swallowing Disorders
University of South Florida

University of South Florida Health Sciences Center
12901 Bruce B. Downs Blvd., MDC Box 72
Tampa, FL 33612

MEDICAL STAFF

Director H. Worth Boyce, Jr., M.D.
Professor of Medicine
Hugh F. Culverhouse Chair
in Esophagology

Medical Staff Milton C. Johnson, M.D.
Associate Professor of Medicine

Patient Care Janet L. Jones, B.A., CGC
Coordinator
Instructor of Medicine

Assistant Patient Care Betsy J. Lamoy, R.N.
Coordinator

Nurse Practitioner Sarah A. Garza, ARNP

Office Manager/ Candace K. Harley
Research Coordinator

Administrative Secretary Natalie A. Ralyea

Appointment Secretary Jennifer C. Rust

Speech Pathology Consultants for Oropharyngeal Swallowing Disorders

Speech Pathology Joy E. Gaziano, M.A., CCC/SLP
Linda Stachowiak, M.S., CCC/SLP
Cara Smith, M.A., CCC/SLP

Things To Remember

OFFICE HOURS: 8:00 a.m. 'til 5:00 p.m. Monday through Friday.
Telephone hours: 8:00 a.m. 'til 5:00 p.m. Also, our emergency
telephone number for after hours is (813) 974-2201

BILLING: Payment for services rendered is due at the time of your
visit. Please be prepared to pay any co-payments due at the time of
your visit to the Center. Patients who have problems with their
physician or facility fee bills should contact Gayle Stephens,
Financial Specialist, at the University of South Florida Medical
Clinics at (813) 974-3575 between the hours of 9:00 a.m. and 4:00
p.m. Monday through Thursday. For those patients who are from
out-of-town, a toll-free number has been added for you to call with
billing questions. The number is 1-888-873-3627. This number is
for calls originating in Florida and is only for billing
questions and help with insurance authorizations.

**HAS YOUR INSURANCE COMPANY OR PRIMARY CARE
PHYSICIAN CHANGED?** With an ever changing medical
insurance market (shopping for the best contract, companies
merging, others closing their doors, etc), you may have changed
insurance company. If you changed your insurance company you
may have a new primary care physician. Maybe you have moved
and had to choose a new doctor closer to your home. Regardless
of the circumstances we would very much appreciate your contacting
our office to let us know, (813) 974-3374. This will not only insure
we can obtain the necessary authorizations/pre-certifications and
that your medical bills go to the right insurance company but it will
help us make sure your medical records are forwarded to the right
doctors. Thank you for helping us keep the records straight.

NON-Profit Organization
U.S. Postage
PAID
Tampa, FL
Permit No. 1632