



## **Upper Esophageal Sphincter Dysfunction**

Upper esophageal sphincter (UES) dysfunction occurs when the muscles of the upper esophagus, specifically the cricopharyngeus, develop impaired relaxation. Symptoms include progressive dysphagia from pills to solids and then liquids. Diagnosis is made from a thorough history and Modified Barium Swallow Study (MBSS). Other testing such as manometry and manofluoroscopy can be obtained as well. Treatments for upper esophageal sphincter dysfunction include botulinum toxin injection into cricopharyngeal muscle, UES dilation, endoscopic cricopharyngeal myotomy or transcervical cricopharyngeal myotomy.

### **Anatomy and Physiology:**

- The upper esophageal sphincter (UES) is a 2-4 cm high pressure zone inferior to the laryngeal opening. It is bound by muscles, cartilage and aponeuroses. Posteriorly the inferior pharyngeal constrictor muscle starts at the median raphe and inserts laterally into the thyroid cartilage. The cricopharyngeus muscle attaches laterally to the cricoid cartilage. The uppermost cervical esophageal fibers are involved as well. Anteriorly the UES is made up of cricoid and arytenoid cartilages.
  - Lim KG. The mouth of the esophagus. *Laryngoscope* 1907;17:421-428.
  - Singh S, Hamdy S. The upper oesophageal sphincter. *Neurogastroenterol Motil* 2005;17(1):3-12.
  - Sivarao DV, Goyal RK. Functional anatomy and physiology of the upper esophageal sphincter. *Am J Med* 2000;108:27s-37s.
- Opening of the upper esophageal sphincter occurs with relaxation of the cricopharyngeus and inferior pharyngeal constrictor, elevation of the larynx and pressure exerted on the bolus of food by hypopharyngeal musculature and the tongue base.
  - Cook I, Dobbs W, Dantas R et al. Opening mechanisms of the human upper esophageal sphincter. *Am J Physiol* 1989; 257:G748-G759.
- The cricopharyngeus and inferior pharyngeal constrictor muscles are made up of slow-twitch type II myosin heavy chain muscle fibers as the inner layer. These are active in sustained muscle contraction. The cricopharyngeus is felt to be the more dominant muscle in the resting UES pressure.
  - Mu L, Sanders I. Muscle fiber-type distribution pattern in the human cricopharyngeus muscle. *Dysphagia* 2002;17:87-96.
  - Mu L, Sanders I. Neuromuscular organization of the human upper esophageal sphincter. *Ann Otol Rhinol Laryngol* 1998;107:370-377.

### **Assessment:**

- A complete patient history regarding their dysphagia symptoms is an important aspect of assessment. Additional studies for diagnosis including videofluoroscopy and manometry.
- On videofluoroscopy a cricopharyngeal bar on lateral view indicates cricopharyngeal dysfunction. This is classified as non-obstructing, obstructing and Zenker's diverticulum. Cricopharyngeal bar that is obstructing is based on the opening relative to age-appropriate normative data.



- Belafsky PC, Rees CJ, Allen J, Leonard RJ. Pharyngeal dilation in cricopharyngeus muscle dysfunction and Zenker diverticulum. *Laryngoscope* 2010;120:889-894.
  - Leonard R, Kendall K, McKenzie S. UES opening and cricopharyngeal bar in nondysphagic elderly and nonelderly adults. *Dysphagia* 2004;19:182-191.
- High-resolution manometry has 36 circumferential sensors spaced at approx. 1 cm apart that allow for improved measurement of pressure of the esophageal segments. The upper esophageal sphincter elevates when swallowing and these more concentrated sensors can capture a generalized pressure in the area. This technique is used to measure pressures in a normal upper esophageal sphincter as well as in upper esophageal sphincter dysfunction.
- Ghosh SK, Pandolfino JE, Zhang Q, Jarosz A et al. Deglutitive upper esophageal sphincter relaxation: a study of 75 volunteer subjects using solid-state high-resolution manometry. *Am J Physiol Gastrointest Liver Physiol* 2006;291:G525-G531.

### **Pathophysiology:**

- Etiologies of upper esophageal sphincter dysfunction can be classified into anatomic (cricopharyngeal bar), neuromuscular (central, peripheral, or myogenic), iatrogenic, inflammatory, neoplastic or idiopathic.
- Halvorson DJ. The treatment of cricopharyngeal dysmotility with a transmucosal cricopharyngeal myotomy using the potassium-titanly-phosphate (KTP) laser. *Endoscopy* 1998;30:46-50.
- Age related changes occur at the upper esophageal sphincter. There is delayed relaxation and diameter of the opening is smaller compared to younger patients.
- Fulp SR, Dalton CB, Castell JA et al. Aging-related alterations in human upper esophageal sphincter function. *Am J Gastroenterology* 1990;85:1569-72.
  - Shaker R, Kern M, Bardan E et al. Augmentation of deglutitive upper esophageal sphincter opening in the elderly by exercise. *Am J Physiol* 1997;272(6 Pt 1):G1518-22.
- It is thought that a non-obstructing cricopharyngeal bar develops into an obstructing one with time and that a Zenker's diverticulum develops from an obstructing cricopharyngeal bar.
- Belafsky PC, Rees CJ, Allen J, Leonard RJ. Pharyngeal dilation in cricopharyngeus muscle dysfunction and Zenker diverticulum. *Laryngoscope* 2010;120:889-894.

### **Treatment:**

- Traditionally the treatment for upper esophageal sphincter dysfunction has been transcervical cricopharyngeal myotomy. The first procedure was done in 1951 on a patient with bulbar poliomyelitis by Kaplan.
- Kaplan S. Paralysis of deglutition, a post-poliomyelitis complication treated by section of the cricopharyngeus muscle. *Ann Surg* 1951;133:572-573.
- The endoscopic approach using a KTP laser was introduced in 1994 by Halvorson and Kuhn. Pitman et al described the technique of CO2 laser cricopharyngeal myotomy in detail. Endoscopic cricopharyngeal myotomy (ECPM) is a safe and effective procedure. Morbidity of



this procedure is thought to be less compared to transcervical cricopharyngeal myotomy (TCPM).

- Halvorson DJ, Kuhn FA. Transmucosal cricopharyngeal myotomy with the potassium-titanyl-phosphate laser in the treatment of cricopharyngeal dysmotility. *Ann Otol Rhinol Laryngol* 1994;103(3):173-7.
  - Pitman M, Weissbrod P. Endoscopic CO2 Laser Cricopharyngeal Myotomy. *The Laryngoscope*. 2009 Jan; 119:45-53.
- Dauer et al. compared the endoscopic and transcervical approaches to myotomy and found that the endoscopic approach had a shorter hospital stay and operative time. The postoperative complications were also much more severe in the transcervical myotomy group. Overall, multiple studies have shown ECPM to have fewer complications.
- Dauer E, Salassa J, Iuga L et al. Endoscopic laser vs open approach for cricopharyngeal myotomy. *Otolaryngol Head Neck Surg* 2006;134:830-835.
- Multiple authors including Mason et al and Kos et al. reported better outcomes after myotomy in patients who had dysphagia from an idiopathic etiology compared to patients with neuropathic or myopathic disease.
- Mason RJ, Bremner CG, DeMeester TR. Pharyngeal swallowing disorders selection for and outcome after myotomy. *Ann Surg* 1998;228:598-608.
  - Kos MP, David EF, Klinkenberg-Knol EC et al. Long-term results of external upper esophageal sphincter myotomy for oropharyngeal dysphagia. *Dysphagia* 2010;25:169-176.
- Alternatives to myotomy include endoscopic dilation of the upper esophageal sphincter. Most of these studies are limited to small case series and have reported success in this technique. Hatlebakk et al. reported improvement in 9/10 of their patients using Savary dilators up to 18-20 mm. Solt et al. reported 100 % success rate with use of balloon dilation up to 20 mm.
- Hatlebakk JG, Castell JA, Spiegel J et al. Dilatation therapy for dysphagia in patients with upper esophageal sphincter dysfunction—manometric and symptomatic response. *Dis Esophagus* 1998;11:254-59.
  - Solt J, Bajor J, Moizis M et al. Primary cricopharyngeal dysfunction: treatment with balloon catheter dilatation. *Gastrointest Endosc* 2001;54:767-771.
- Schneider et al. first proposed the use of Botox injections into the cricopharyngeus muscle. Since then many have supported this technique since it can be done in the office setting with EMG guidance with minimal invasiveness. It is also done through an endoscopic approach. Doses range from 5-50 units per injection, up to 100 units. Botox usually lasts from 3-4 months in duration and then has to be repeated.
- Schneider I, Potatschnig C, Thomfort WF et al. Treatment of dysfunction of the CPM with botulinum A toxin: introduction of a new, noninvasive method. *Ann Otol Rhinol Laryngol* 1994;103:31-35.
  - Blitzer A, Brin MF. Use of botulinum toxin for diagnosis and management of cricopharyngeal achalasia. *Otolaryngol Head Neck Surg* 1997;116:328-330.