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Endoscopic diagnosis and treatment of disorders of upper esophageal sphincter function

Ronak V. Patel, MD, Ikuo Hirano, MD*

Division of Gastroenterology and Hepatology, Northwestern University Feinberg School of Medicine, Chicago, Illinois

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ABSTRACT

The upper esophageal sphincter functions to facilitate antegrade transit during deglutition and restrict retrograde flow of refluxed material from the esophagus. Neuropathic, myopathic, and inflammatory conditions can result in upper esophageal sphincter dysfunction and lead to detrimental consequences of dysphagia and aspiration. Proper evaluation of oropharyngeal swallowing is critical to diagnosing upper esophageal sphincter pathology. The aim of this review is to describe the diagnosis and treatment of disorders of the upper esophageal sphincter.

1. Introduction

The upper esophageal sphincter (UES) serves as the entry way to the gastrointestinal tract. Its location between pharyngeal and esophageal phases of swallowing is critical in maintaining anterograde transit during swallowing and impeding retrograde flow during reflux events. UES impairment has severe consequences including dysphagia and pulmonary aspiration. Knowledge of the diagnosis and treatment of UES disorders has grown with advancing technology and better understanding of specific disease states.

2. Anatomy and physiology

The UES acts as an entry point to the cervical esophagus during food bolus transfer. Following a voluntary phase of mastication and bolus formation, the bolus is transitioned from the oral cavity to the pharynx. Pharyngeal muscles ultimately elevate and pull forward the larynx to allow for UES opening and bolus transfer [1]. The UES is identified manometrically as a 2-3 cm focus of elevated pressure between the hypopharynx and the esophagus. It is primarily comprised of the transversely oriented striated muscle fibers of the cricopharyngeus with contributions from the inferior pharyngeal constrictor and thyropharyngeous muscles. The cricopharyngeus is a C-shaped muscle that has attachments to the lateral aspects of the cricoid cartilage at the level of the C5-6 vertebral interspace [1]. For a bolus to pass to the esophagus, the larynx is elevated and pulled forward to facilitate UES opening. Tongue pulsion then propels the bolus through the UES, followed by a peristaltic contraction that clears residue from the pharynx and through the esophagus. Properties of the UES have been shown to change with aging, which may highlight why certain diseases of the UES are found in elderly patients [2]. Innervation of the UES is via the vagus nerve which assists in managing resting pressure and relaxation. Diseases affecting the cerebral cortex, brainstem, or vagal innervation can impair UES function. Constriction of the UES helps to prevent anterograde ingestion of air and retrograde aspiration and reflux of retained esophageal or gastric contents.

3. Diagnostic evaluation of the upper esophageal sphincter

Disorders of UES function are largely related to baseline pressure and relaxation properties. Patients with UES dysfunction often present with symptoms of oropharyngeal dysphagia as well as additional symptoms of coughing, choking, frank aspiration, globus, and nasal regurgitation. Localization of dysphagia to the neck combined with a history of aspiration events increases the likelihood of a pharyngeal rather than esophageal etiology. Once oropharyngeal dysphagia is suspected, a careful review of systems and examination should be obtained with particular attention to concurrent neurologic symptoms and signs. Initial evaluation of oropharyngeal dysphagia can be assessed with VFSS to diagnose functional deficits and/or a barium esophagram to identify structural abnormalities. A variety of newer modalities can help identify pathology resulting from dysfunction of the UES.

3.1. Esophagogastroduodenoscopy

Upper endoscopy, or esophagogastroduodenoscopy (EGD), is generally considered the initial diagnostic test for the evaluation of

Abbreviations: BTX, botulinum toxin; CP, cricopharyngeal; FLIP, functional lumen imaging probe; HRM, high resolution manometry; OPMD, oculopharyngeal muscular dysphagia; UES, upper esophageal sphincter; VFSS, videofluoroscopic swallow study

Conflicts of interest: None.

* Corresponding author.
E-mail address: i-hirano@northwestern.edu (I. Hirano).

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dysphagia. While the UES must be passed in order to enter the proximal esophagus, the region of the esophageal inlet is generally very poorly visualized. This is in part due to stimulation of the gag reflex during initial intubation of the esophagus as well as limited ability to distend the lumen at the level of the constricted UES. Difficulty passing a scope can be a sign of a cricopharyngeal (CP) bar or Zenker's diverticulum. The anatomic recesses of the piriform sinuses as well as muscular constriction of the UES create physiologic obstacles to esophageal intubations even in the absence of overt pathology. In one study examining complications during EGD, the esophagus was the most common site to develop perforations, with 23% of esophageal perforations occurring in the cervical region [3]. On the other hand, 3/39 cases of esophageal perforations were in patients with Zenker's diverticula [3]. Although EGD is a widely utilized and valuable diagnostic and therapeutic tool for patients with dysphagia, clinical suspicion for oropharyngeal dysphagia should prompt initial radiologic testing prior to endoscopy. In some cases, clear oropharyngeal dysphagia on radiologic imaging may obviate the need for EGD. In other cases, identification of structural pathology on imaging such as a cervical web or cricopharyngeal bar may lead to plans for an esophageal dilation during the EGD even in the absence of endoscopically visualized pathology.

3.2. Videofluoroscopic swallowing study

Videofluoroscopic swallowing studies (VFSS) or modified barium swallow examinations are a first line diagnostic tool in the assessment of patients presenting with localization of cervical dysphagia and clinical suspicion for oropharyngeal dysphagia [4]. Exams can be performed by radiologists but are often done in conjunction with speech pathologists trained to assess for functional deficits associated with aspiration risk, modification of swallowing behavior, and specific feeding recommendations. Initially, smaller volumes of barium are swallowed to assess for frank aspiration. As the exam proceeds, increasing volumes and thicker consistencies of barium are administered, typically culminating in ingestion of a barium coated cookie (thus the name, “cookie swallow”) [5]. Evaluation of the esophageal phase of swallowing after the VFSS is an important consideration in order to assess for esophageal pathologies [6,7]. Patients may simultaneously present with both pharyngeal and esophageal disorders. Such a scenario is common in patients with dysphagia following head and neck radiation therapy who are at risk for both oropharyngeal muscular dysfunction including the UES as well as mechanical strictures of the cervical esophagus.

VFSS or pharyngeal imaging during a barium esophagram can provide qualitative assessment on the presence of features such as aspiration and is particularly helpful in identifying anatomic lesions such as a cricopharyngeal bar and Zenker's diverticulum [8]. Quantitative assessment, such as opening diameter of the UES and pharyngeal constriction ratio, has been studied on VFSS [9,10]. However, this is not done routinely at many institutions and there is questionable inter-rater reliability [11].

3.3. High resolution manometry

High resolution manometry (HRM) has traditionally been used for diagnosis of esophageal motility disorders [12]. HRM can provide more quantitative measurements regarding swallowing function compared to VFSS examinations. However, less is known about the utility of HRM for assessment of the UES specifically. Older studies using water perfused manometry assemblies elegantly demonstrated that elevated intrabolus pressures in the hypopharynx were associated with reduced UES opening in patients with both CP bars and Zenker's diverticula [13,14]. HRM with impedance can help to identify patients with oropharyngeal dysphagia. In a recent study in the pediatric population, UES opening and relaxation measurements distinguished patients with and without oropharyngeal dysphagia [15] (Figure 1). Impedance measurements from HRM have also been used to determine nadir impedance values, corresponding to maximal admittance through the UES [2]. Recent studies have described improved inter-rater reliability for impedance measurements as compared to fluoroscopic studies [16]. Nadir impedance varies according to age and disease states, with elderly patients and patients with CP bars and motor neuron diseases having lower maximum admittance [2]. HRM has the capability to give pressure measurements within the pharynx and UES as well as durations of UES relaxation and

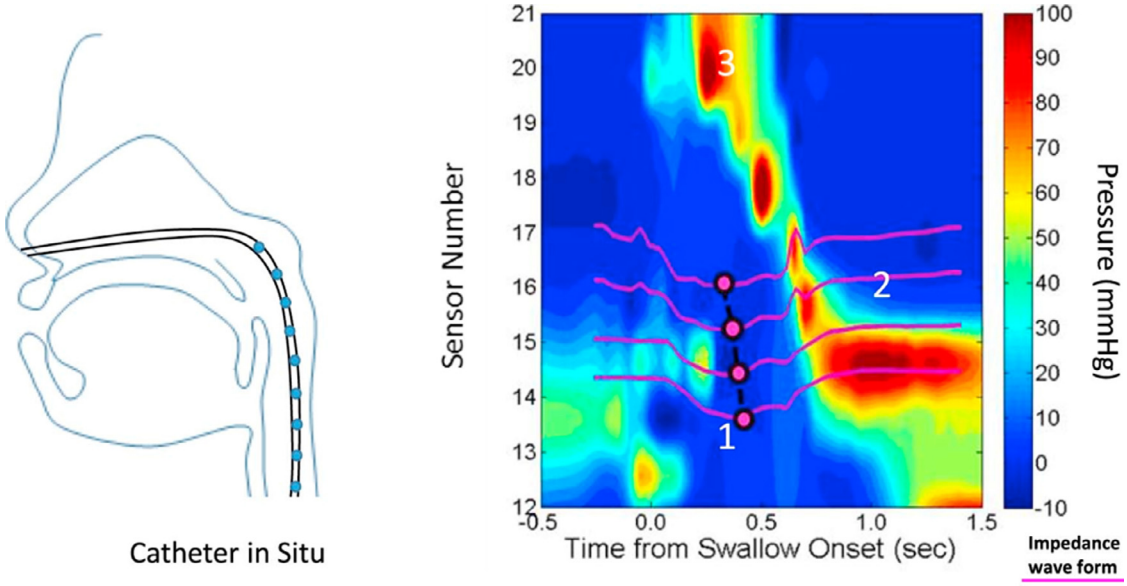

Fig. 1. High resolution manometry with esophageal pressure topography plot of the upper esophageal sphincter (UES) with superimposed impedance tracings (pink lines). The impedance recordings depict bolus transit during the swallow. Locations specified include 1: onset of swallow; 2: proximal margin UES post-swallow; and 3: position of velopharynx. Reprinted from The Journal of Pediatrics, Vol 177, Ferris L, Rommel N, Doeltgen S, Scholten I, Kritas S, Abu-Assi R et al. Pressure-flow analysis for the assessment of pediatric oropharyngeal dysphagia, 279-85, Copyright 2016, with permission from Elsevier. (Color version of figure is available online.)