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Dysphagia

Food Intake by Maneuver; An Extreme Compensation for Impaired Swallowing

Peter J. Kahrilas, M.D.,[1] Jerilyn A. Logemann, Ph.D.,[2] and Patricia Gibbons, M.S.[2]
[1]Department of Medicine and [2]Departments of Communication Sciences and Disorders, Northwestern University, Chicago, Illinois, USA

Abstract. We report two young patients able to exist on exclusively oral intake despite an absent pharyngeal swallow response. Videofluoroscopic swallowing studies showed that both patients used a sequence of devised maneuvers rather than a coordinated pharyngeal swallow to move the bolus, protect the airway, and open the upper esophageal sphincter during bolus ingestion. We conclude that it is possible for young, highly motivated individuals to maintain oral intake despite ablation of neurologic elements crucial for the normal swallow response.

Key words: Swallowing — Videofluoroscopy — Dysphagia — Swallow maneuvers — Deglutition — Deglutition disorders.

Recent investigations have found that, despite a high level of stereotypy, the swallow response is modifiable. Bolus volume is an unconscious cue that alters the timing and vigor of the motor pattern [1–3]. Volitional control can be used to accentuate the period of upper esophageal sphincter (UES) opening or airway closure at the level of the laryngeal vestibule [4, 5]. Supporting the notion that such compensatory mechanisms may be useful in treating oropharyngeal dysphagia, we describe two patients who, despite neurologic ablation of the normal pharyngeal swallow response, learned to compensate and maintain oral feeding.

Case 1

Patient 1 is an 18-yr-old female college student with type II spinal muscular atrophy (late infantile, intermediate form). Her illness, first manifest at age 18 months, was mainly characterized by progressive skeletal muscle weakness resulting in loss of the ability to walk at age 7 and progressive scoliosis requiring spinal fusion from T3 to the sacrum at age 13. A muscle biopsy done 1 year prior to our evaluation showed changes of chronic denervation. In addition to peripheral muscle weakness the patient had slight nasality of voice and "some difficulty swallowing steak" for the past 3 years. Nevertheless, she consumed normal meals, taking about 1 h to eat. On examination the patient weighed 50 lbs, appeared cachectic, and was confined to a motorized scooter with flexion contractures of the knees and abduction contractures of the hips. She had intact sensation but diffuse muscle weakness and absent deep tendon reflexes. Also noted were bilateral facial nerve weakness and a normal gag reflex. Tongue fasciculations were present along with decreased tongue strength and pooling of oral secretions.

Case 2

Patient 2 is a 34-yr-old male who was well until he underwent surgery for removal of a right carotid body tumor 18 months prior to evaluation. Surgery was done through an incision along the anterior border of the sternocleidomastoid. A carotid body tumor extending cephaloposterior from the carotid bifurcation to the base of the skull, involving the vagus, hypoglossus, and glossopharyngeal nerves, was excised without damage to the carotid artery and without transecting (although clearly manipulating) the involved cranial nerves. Pathologic examination revealed the tumor to be a vagal paraganglioma.

Address offprint requests to: P.J. Kahrilas, M.D., Northwestern University Medical School, GI Section, Department of Medicine, 1526 Wesley Towers, 250 E. Superior St., Chicago, IL 60611, USA

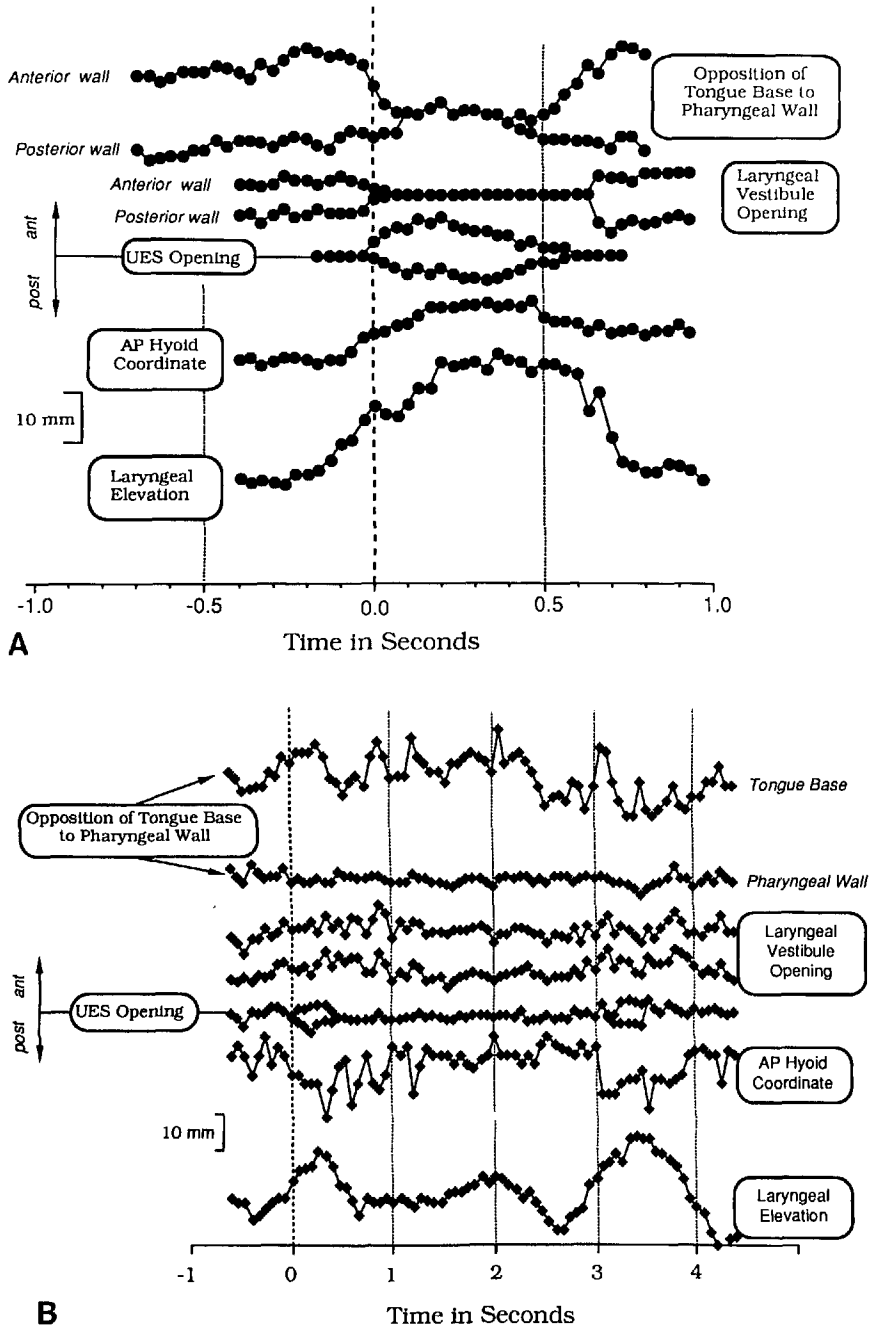

Fig. 1. Composite graphs showing the relationship among biomechanical events in a normal swallow (**A**) and in patient 1 (**B**). In each case, time 0 represents the time of UES opening. In panel A, but not in panel B, laryngeal vestibular closure occurred within 0.1 s of UES opening and was maintained throughout the period of sphincter opening. Similarly, the tongue base contacts the pharyngeal wall propelling the bolus distally in panel A, but not in panel B. Lower two tracings indicate that UES opening is associated with a brisk anterior hyoid tug in panel A but not in panel B, and that all of the key events of the swallow occur during laryngeal elevation. Increased laryngeal elevation during UES opening exhibited in panel B suggests that this motion was being used to help open the sphincter.

The patient experienced severe dysphagia immediately after surgery. He struggled to resume a normal diet without nutritional supplements which he achieved 6 months after surgery. He states that it takes a long time to eat and food often catches in his throat, but he has maintained his weight at 165 pounds. Aside from the dysphagia, the patient also had voice changes postoperatively attributable to unilateral vocal cord paralysis and nasal emission when pronouncing consonants. The patient's speech had improved in the 18 months since surgery. One year after surgery, the patient had a cine-esophagram which suggested bilateral pharyngeal paralysis and dilation. Esophageal motility studies showed normal UES relaxation with swallowing, normal esophageal peristalsis, and normal lower esophageal sphincter relaxation. However, no pharyngeal pressure spike was demonstrated.

Methods

Lateral videofluoroscopic swallowing studies were recorded on each of the patients and compared with a study done on a 22-yr-old normal volunteer (approved by the Northwestern University Institutional Review Board). A dime taped under the chin was used to correct for magnification. Subjects were instructed to attempt to swallow each test bolus (5 ml liquid barium, 5 ml

paste barium, and a barium-impregnated cookie) as a single bolus.

Spatial analysis of the 5-ml liquid barium videofluoroscopic swallowing sequences was done with an interactive computer program written to enable x-y coordinate determination of selected structures [3, 6]. The segment of each patient's "swallow" analyzed was that prior to and immediately following UES opening. The structures marked were: (1) the anterior-superior corner of hyoid bone; (2) the posterior-superior corner of the subglottic air column; (3) the tongue base and the posterior pharyngeal wall at the level of the pit of the valleculae; (4) the arytenoid and base of epiglottis forming the entry to the laryngeal vestibule (identical when the vestibule was closed); and (5) the anterior and posterior walls of the UES (identical when the sphincter was closed). Following the spatial analysis, the patient tapes were analyzed subjectively by slow motion playback to scrutinize the abnormal means by which the patients ingested food.

Results

Normal Swallow vs Food Ingestion by Patients

Figure 1 illustrates the coordination between the fundamental mechanical events that occur during 5-ml swallows in a normal individual (A) and in patient 1 (B). In contrast to the normal coordination shown in Figure 1A, the two patients studied demonstrated very severe, although similar, abnormalities. Note that the fundamental conditions normally achieved in conjunction with UES opening were not achieved in Figure 1B; the laryngeal vestibule was not closed, the tongue base did not contact the pharyngeal wall, and the hyoid did not exhibit the typical anterior tug that is associated with UES opening as in Figure 1A [3].

Figure 2 compares the pattern of UES opening in the normal subject with that of the patients. Each patient required two to five brief, small aperture, short duration "winks" of the UES before most of the bolus had entered the esophagus compared with the single opening required by the normal subject. The mechanism by which UES opening was achieved was not readily apparent in the patients, but clearly, it was not associated with the normal anterior tug of the hyoid. In the example of Figure 1B, each opening event was accompanied by a brief period of increased laryngeal elevation suggesting that this action applied traction to the anterior sphincter wall. In the other subject this pattern was not evident and opening was instead accompanied by neck extension during prolonged laryngeal elevation.

Food Ingestion by Maneuver

As might be expected from the abnormal pattern of UES opening illustrated in Figure 2, the visual pattern of food ingestion in both patients was markedly

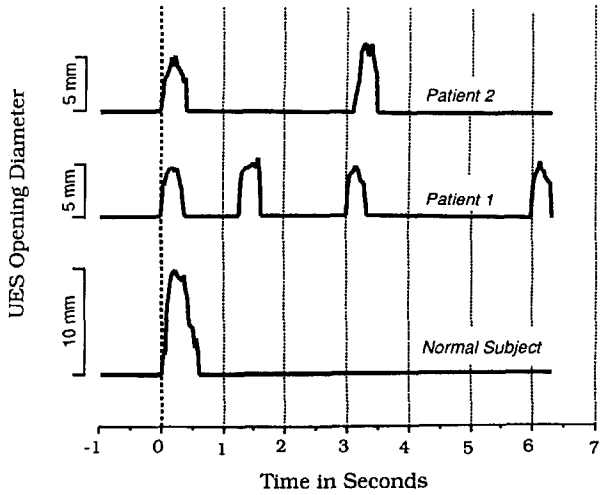

Fig. 2. Opening profiles of the UES in each patient and the normal volunteer. In each case sequence shown is that necessary to clear a single bolus from the hypopharynx. The normal individual accomplished this with a single 9-mm, 0.6-s opening timed to coincide with laryngeal vestibular closure and lingual propulsion as shown in Fig. 1. Patients, on the other hand, exhibited multiple smaller openings of 0.4- to 0.5-s duration.

abnormal and inefficient. Whereas a normal swallow will result in complete clearance of the oropharynx within 1 s, each of these patients took in excess of 10 s to clear most of a single bolus from the pharynx. Figure 3 depicts tracings from still images during a swallow in a normal subject (A) and in patient 1 (B). Tracings from patient 2 were very similar to those of patient 1. Note that patient 1 achieved the fundamental swallow events (bolus propulsion, UES opening, airway protection) sequentially by devised maneuvers. First the bolus was dumped into the valleculae by oral tongue propulsion. Then, by alternately retracting the chin which compresses the valleculae and relaxing the chin which widens the pharynx, the bolus was squeezed out of the valleculae and fell into the pyriform sinuses. Finally, small amounts of the bolus are moved across the UES in increments as the sphincter is opened briefly by a voluntary tug from the larynx or by neck extension.

Discussion

The pharyngeal swallow is a complex, rapidly coordinated activity that facilitates food ingestion while at the same time preventing aspiration or nasopharyngeal regurgitation. The swallow response is attributable to activation of the medullary swallow center which initiates a sequence of excitation and inhibition of medullary motoneuron pools followed by the sequenced activation of oropharyngeal muscles causing: (1) elevation and retraction of the soft

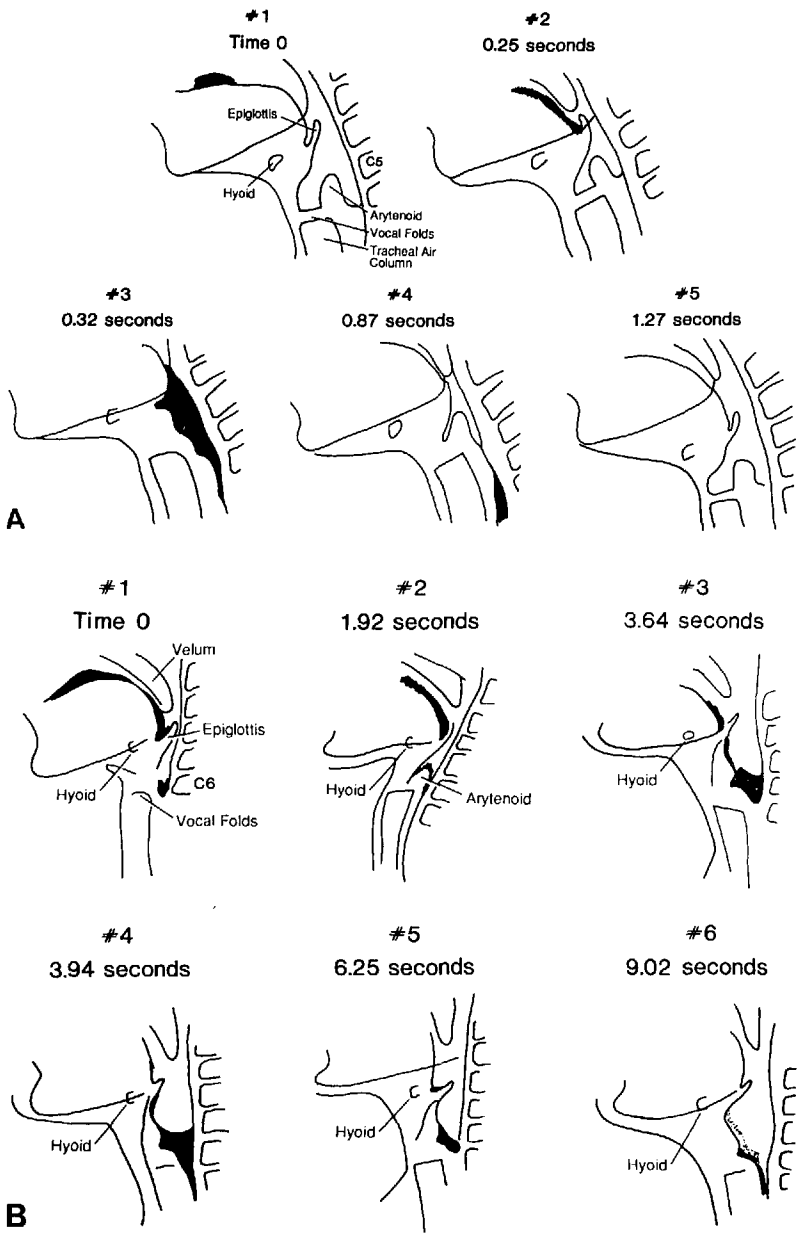

Fig. 3. Videofluoroscopic tracings comparing the timing and mechanism of key events in a normal swallow **(A)** compared with patient 1 **(B)**. Entire sequence in panel A is completed within 1.27 s: in image 1 the bolus is in the oral cavity, the laryngeal vestibule is open, and the UES is closed; in image 2, 0.25 s later, the bolus has been propelled into the valleculae and the larynx has begun to elevate; in image 3, less than 0.1 s later, maximal anatomic alteration occurred with the laryngeal vestibule obliterated, the UES opened, and the tongue base fully retracted against the pharyngeal wall; image 4 shows structures beginning to return to rest position which is fully achieved in image 5. Images in panel B, taken over a span of 9 s, illustrate the process of food ingestion in patient 1. Image 1 depicts the resting condition, albeit with residue from the previous swallow. In image 2, almost 2 s later, the patient retracted her chin to dump the barium into the hypopharynx while narrowing the laryngeal inlet and protecting the airway. The bolus then sits above the closed sphincter until the UES winks open (images 4 and 6). Note that the bolus literally falls through the sphincter rather than being propelled by anything resembling a pharyngeal contraction.

palate with closure of the nasopharynx; (2) anterior superior laryngeal displacement; (3) laryngeal closure at the level of the epiglottis, false vocal folds, and true vocal folds; (4) relaxation and opening of the upper esophageal sphincter; and (5) pharyngeal propulsion [7, 8]. Using this motor pattern as the definition of a normal pharyngeal swallow, the two patients described in this report did not have a pharyngeal swallow response. As demonstrated by the data depicted in Figure 1 the coordinated motor pattern constituting the pharyngeal swallow response is simply not there as a consequence of peripheral nerve damage.

Accepting that neither of the patients described in this report could swallow, how did they ingest food? In each case, the patient used a series of self-devised voluntary maneuvers to sequentially achieve bolus propulsion, airway protection, and UES opening. Airway protection was improvised by head posturing that directed the bolus around the laryngeal vestibule. In the absence of normal pharyngeal propulsion mechanism [3, 8, 9] the bolus advances from the valleculae to the pyriform sinuses by gravity. Consequently, only a small bit of the bolus traversed the UES with each opening of the sphincter and the head of the bolus moved with markedly diminished

velocity. Opening of the UES was achieved with a series of laryngeal and neck movements. The obvious cost of this method of food ingestion was reduced efficiency. Each patient was able to ingest only small food boluses and even then it took ten times as long as it did a normal individual using the subconsciously activated pharyngeal swallow response. Nonetheless, both patients were adamant in preferring their present status to the alternative of being sustained by tube feeding.

We conclude that the ability to ingest food orally is not necessarily lost with impairment of the swallow response. Although the normal, subconscious pharyngeal swallow response is clearly the most efficient means of food ingestion, devised maneuvers under voluntary control can be learned to compensate for defective components of the swallow, be it failed propulsion, impaired airway protection, or reduced UES opening. The patients described in this report represent extreme examples of compensation for impaired swallowing, demonstrating that young, highly motivated individuals are capable of ingesting food orally despite complete absence of a swallow response.

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