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Pharyngeal mis-sequencing in dysphagia: Characteristics, rehabilitative response, and etiological speculation

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ABSTRACT

Objective: Clinical data are submitted as documentation of a pathophysiologic feature of dysphagia termed pharyngeal mis-sequencing and to encourage clinicians and researchers to adopt more critical approaches to diagnosis and treatment planning.
Background: Recent clinical experience has identified a cohort of patients who present with an atypical dysphagia not specifically described in the literature: mis-sequenced constriction of the pharynx when swallowing. As a result, they are unable to coordinate streamlined bolus transfer from the pharynx into the esophagus. This mis-sequencing contributes to nasal redirection, aspiration, and, for some, the inability to safely tolerate an oral diet.
Method: Sixteen patients (8 females, 8 males), with a mean age of 44 years (range = 25–78), had an average time post-onset of 23 months (range = 2–72) at initiation of intensive rehabilitation. A 3-channel manometric catheter was used to measure pharyngeal pressure.
Results: The average peak-to-peak latency between nadir pressures at sensor-1 and sensor-2 was 15 ms (95% CI, −2 to 33 ms), compared to normative mean latency of 239 ms (95% CI, 215 to 263 ms). Rehabilitative responses are summarized, along with a single detailed case report.
Conclusion: It is unclear from these data if pharyngeal mis-sequencing is (i) a pathological feature of impaired motor planning from brainstem damage or (ii) a maladaptive compensation developed in response to chronic dysphagia. Future investigation is needed to provide a full report of pharyngeal mis-sequencing, and the implications on our understanding of underlying neural control of swallowing.

1. Introduction

Swallowing disorders are a common consequence of neurologic impairment. Up to 70% of patients following stroke [1], 31% following acoustic neuroma surgery [2], and 35% of patients with Parkinson's disease [3] present with dysphagia. Additionally, it has been reported that 27% of stroke patients develop a respiratory infection due to their swallowing impairment [4]. Such consequences have been shown to increase risk of death and also increase the cost of hospitalization by approximately $27,633 USD per patient [5].

Safe and efficient swallowing is reliant on an accurate and timely cascade of sensorimotor responses [6]. Once initiated, this complex, but stereotyped, swallowing response is thought to be an irreversible motor sequence, controlled by functional interconnectivity between bilateral neurons in the nucleus tractus solitarius (NTS) and the nucleus ambiguous (NA) [7]. This medullary central pattern generator (CPG) produces the 'primitive' swallowing response in the absence of afferent sensory input [7–9]. Specific to this discussion, there is sequenced but overlapping contraction of the superior, middle, and then inferior pharyngeal constrictors for bolus propulsion [10]. The more complex ingestive swallowing response is thought to incorporate central modulation of the primitive swallowing pattern through cortical and sensory input to the NTS, altering strength and duration of pharyngeal events [7,9]. However, the overall motor sequence of pharyngeal swallowing should remain constant, even in the presence of cortical modulation [11–13].

Historically, behavioral rehabilitation approaches for dysphagia have capitalized on this central ability to modulate strength and duration of the swallowing response [14]. For example, a technique called *effortful swallowing*, whereby patients are instructed to 'swallow hard,'

Abbreviations: NTS, nucleus tractus solitarius; NA, nucleus ambiguus; CPG, central pattern generator; fMRI, functional magnetic resonance imaging; VFSS, videofluoroscopic swallowing study; PEG, percutaneous endoscopic gastrostomy; UES, upper esophageal sphincter; CPA, cerebellopontine angle; CN, cranial nerve.

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was initially developed to compensate for reduced base of tongue to posterior pharyngeal wall contact and improve bolus transit [15]. Treatments such as effortful swallowing are predicated on an underlying presumption that the neurologic damage has led to a weak, or flaccid-type, dysphagia, despite normal swallowing being reliant on submaximal muscular force [16].

Our diagnostic tools likely exacerbate this bias toward weakness as an etiology. For example, the videofluoroscopic swallowing study (VFSS) is considered by many to be the current "gold standard" for swallowing assessment. VFSS provides a two-dimensional dynamic image of swallowing, allowing frame-by-frame analysis of ingestive biomechanics. However, this study is subjectively interpreted, has limited temporal resolution – minimally 25–30 frames per second [17] – and does not provide information on the underlying nature of impairment, such as weakness, spasticity, apraxia, or other neuromuscular change. Thus, rehabilitation approaches that are predicated on weakness as the primary etiology may be fundamentally flawed. Treating a disorder of spasticity, ataxia, or apraxia with muscle strengthening is unlikely to produce substantial positive outcomes and may indeed be contraindicated. Diagnostic specificity is a prerequisite for rehabilitative specificity.

Pharyngeal manometry provides more temporally-relevant, objective measures of swallowing features than videofluoroscopy. This technique enables collection of quantitative information regarding the timing and duration of pharyngeal pressure [18], thus issues of subjectivity and reduced temporal resolution are minimized. Although diminished pressure measured with manometry cannot be definitively linked to neuromuscular pathophysiology, this technique can provide important and quantifiable insights into temporal characteristics of pharyngeal dysmotility that are inadequately captured with VFSS.

One such pattern of pharyngeal dysmotility that is best observed through manometry is pharyngeal mis-sequencing. The phenomenon of pharyngeal mis-sequencing is characterized by essentially simultaneous pressure generation at the level of the proximal and distal pharyngeal regions, contributing to aspiration, nasal redirection, and, for some, considerable inability to tolerate a diet safely by mouth. The aims of this paper are to document this previously unclassified presentation of swallowing impairment in a descriptive, clinical report and to provide thoughtful speculation on the underlying etiology of this pathophysiologic feature. Diagnostic categorization will be provided on a series of patients ($n = 16$) who demonstrate this apparent pattern of mis-sequenced pharyngeal motility.

2. Materials and methods

2.1. Participants

Sixteen patients (8 females, 8 males), referred to a specialist rehabilitation research clinic for evaluation and rehabilitation of dysphagia, were found to present with a pattern of pharyngeal mis-sequencing based on manometric assessment. Time post-onset at referral, etiology, and pre- and post-treatment diet level of individual patients are summarized in Table 1. The mean age was 44 years old (range = 25–78) with an average time post-onset of 23 months (range = 2–72) at initiation of intensive rehabilitation. As VFSSs were executed prior to referral to this clinic, specific VFSS results are not included in this report. However, in summary, patients in this cohort presented with radiographic features of decreased pharyngeal motility, diffuse pharyngeal residue, and frequent nasal redirection.

2.2. Pharyngeal manometry

A 3-channel manometric catheter was used to measure pharyngeal pressure; the catheter was 100 cm in length and 2.1 mm in diameter, with three 2 × 5 mm solid-state, unidirectional, posteriorly-oriented sensors (Model CTS3 + EMG, Gaeltec, Hackensack, NJ, USA). Data were collected using the Kay Pentax Digital Swallowing Workstation (Model 7120, Kay Pentax, Lincoln Park, NJ, USA). The catheter was inserted through one nares and was swallowed into the proximal esophagus. Once ingested, the catheter was slowly withdrawn until the lower sensor was positioned at the level of the superior portion of the upper esophageal sphincter (UES), based upon the production of a characteristic "M-wave" in the distal sensor [18,19]. When the posteriorly oriented sensors were correctly positioned, sensor 1 measured pressure at the proximal pharynx approximately at the level of the base of tongue and sensor 2 was 3 cm inferior at the distal pharynx, approximately at the level of the laryngeal additus; sensor 3 was a further 2 cm inferior at the superior aspect of the UES. The catheter was secured to the external nose with tape.

Table 1
Summary of demographic and diagnostic information on patients with pharyngeal mis-sequencing.

	Medical diagnosis	Months post-onset	Pre-treatment diet	Post-treatment diet
1	Demyelinating infectious process in the left side of dorsal medulla and ponto-medullary junction	2	PEG	Normal diet, thin liquids
2	Left posterior fossa meningioma, resected by occipital craniotomy	22	Pureed foods and soups	Normal diet, thin liquids
3	Multiple infarcts on the right cerebellum and brainstem as well as bilateral infarcts in the basal ganglia	3	PEG	PEG
4	Right petrous apex chondrosarcoma, with extension to the clivus and the right cavernous sinus; resected by transotic approach	6	PEG	Normal diet, thin liquids
5	Cerebellopontine angle chordoma; resected by occipital craniotomy	16	Soft diet and Nectar (mildly thickened) fluids	Normal diet, thin liquids
6	Neurilemmoma involving the left jugular foramen and cerebellopontine angle; resected by transotic approach with subsequent radiotherapy	18	PEG	Primarily PEG with oral trials as tolerated with compensatory strategies
7	Right cerebellopontine angle meningioma; resected by occipital craniotomy	28	Pureed, soft diet	Normal diet, thin liquids
8	Choroid plexus papilloma; resected by occipital craniotomy	7	PEG	Normal diet, thin liquids
9	Large infratentorial haemorrhage secondary to a right vertebral artery aneurysm; treated with embolization of dissecting right vertebral artery aneurysm	7	PEG	Normal diet, thin liquids
10	Tumor of the fourth ventricular epidermoid in the cerebellum	21	PEG	Primarily PEG with oral trials as tolerated with compensatory strategies
11	Lateral medullary stroke	48	Pureed diet	Normal diet, thin liquids
12	Arteriovenous malformation rupture	72	PEG	Primarily PEG with oral trials as tolerated with compensatory strategies
13	Brainstem stroke	18	PEG	Normal, thin liquids
14	Bilateral pontine infarcts	29	PEG	Normal diet, thin liquids
15	Multiple brain infarcts secondary to polycythemia rubravera	6	Soft diet	Normal diet, thin liquids
16	Spontaneous vertebral artery dissection	60	PEG	PEG