

Pediatric Dysphagia

Challenges and Controversies

Julina Ongkasuwan
Eric H. Chiou
Editors

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ISBN 978-3-319-97024-0 ISBN 978-3-319-97025-7 (eBook)
<https://doi.org/10.1007/978-3-319-97025-7>

Library of Congress Control Number: 2018956734

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Preface

Pediatric dysphagia is a clinical problem that crosses disciplines. Children may be seen by numerous medical specialties including pediatric otolaryngology, gastroenterology, pulmonology, speech pathology, occupational therapy, and lactation consultants. The myriad approaches to the diagnosis and management of dysphagia can be confusing for both clinicians and families, resulting in recurrent trips to medical professionals. Feeding is integral to socialization and to bonding between infants and parents. Disruptions in feeding development can be extremely taxing emotionally and economically for families. Children with dysphagia are some of the most challenging patients even for clinicians who specialize in their care.

Given the heterogeneity of causes and manifestations of pediatric dysphagia, this textbook incorporates the perspectives of multiple types of clinicians that care for these patients including otolaryngologists, gastroenterologists, pulmonologists, speech pathologists, occupational therapist, and lactation consultants, which are important to consider according to the individual features and needs of each patient. We also present the advantages as well as potential limitations of various diagnostic modalities. Finally, we highlight current clinical challenges and controversies in the management of pediatric dysphagia. We hope that this book will encourage cross-specialty pollination of ideas and knowledge as well as stimulate further research in the field.

We would like to thank our chapter authors for their time, effort, and erudite contributions. We would also like to thank the Springer editorial staff for their invaluable assistance. Most of all, we would like to thank our spouses, Shirley Chiou and John Anguay, and our children, Sophie, Elyse, Nathan, Dominic, and Christopher, for their continuing love and support.

Houston, TX, USA

Eric H. Chiou
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Part I
Diagnosis and Treatment of Pediatric
Dysphagia

Chapter 1

Embryology and Anatomy



Annie K. Ahn and Mary Frances Musso

Abbreviations

CN	Cranial nerve
CPG	Central pattern generator
LAR	Laryngeal adductor response
LCR	Laryngeal cough reflex
LES	Lower esophageal sphincter
UES	Upper esophageal sphincter

Introduction

The average individual swallows about 500 times per day [1]. Deglutition or swallowing is an essential function for ingestion of nutrition as well as clearance of secretions from the upper aerodigestive tract. This complex process requires the precise coordination of more than 30 muscles located within the oral cavity, pharynx, larynx, and esophagus [2]. The swallowing apparatus is made up of three upper aerodigestive structures: the oral cavity, pharynx, and larynx. These structures function as a hydrodynamic pump with valves that allows food and liquid to be transferred into the stomach without entering the respiratory tract [3]. The act of swallowing is divided into four phases: oral preparatory phase, oral transport phase, pharyngeal phase, and esophageal phase. Dysphagia, or difficulty swallowing, can be secondary to congenital errors or acquired neurologic or anatomic problems. Dysphagia can lead to many negative consequences including malnutrition, dehydration, pneumonia, and reduced quality of life [2]. To effectively treat dysphagia, a comprehensive understanding of deglutition is essential.

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Embryology

The neurovascular and musculoskeletal structures of the oral and pharyngeal apparatus of deglutition are formed from branchial arches and pharyngeal pouches. The four pairs of branchial arches are derived from ectodermal and mesodermal tissues and form on the lateral side of the head as outgrowths around 5 weeks of gestation. The mesodermal tissue within each arch remodels to form muscle, connective tissue, cartilage, and bone within the head and neck. The arches derive their motor and sensory innervation from adjacent cranial nerves during development, namely, the trigeminal, facial, vagus, and accessory nerves [3].

The frontonasal prominence leads to the formation of the forehead and nose, and its proper development along with the maxillary and mandibular prominences is necessary for normal craniofacial structures such as the nose, choanae, lips, tongue, palate, mandible, maxilla, and cheeks, which are involved in deglutition and are crucial for an intact swallow [3, 4]. Improper development of these structures can result in problems such as cleft lip and/or palate and velopharyngeal insufficiency.

Incomplete fusion of the posterior cricoid lamina and formation of the tracheoesophageal septum can lead to a laryngeal or laryngotracheoesophageal cleft or a bifid epiglottis. Incomplete separation of the trachea and alimentary tract will lead to a tracheoesophageal fistula, which can present as aspiration [5].

Supporting Structures

Supporting structures including the bones, cartilage, teeth, spaces, salivary glands, and muscles are found within the oral cavity, pharynx, and esophagus that help carry out a normal swallow. The mandible, maxilla, hard palate, hyoid bone, cervical vertebrae, styloid process, and mastoid process of the temporal bone support and stabilize the involved muscles and aid in mastication [2]. Various cartilages including the thyroid cartilage, cricoid cartilage, arytenoids, and epiglottis provide support for several muscles of mastication and help with transferring the lingual and pharyngeal bolus. The teeth are vital to bolus preparation. Two sets of teeth develop in humans, deciduous teeth and permanent teeth. The deciduous teeth erupt between 6 months and 2 years of age [6]. Premolars and third molars are absent in children. The progression of deciduous teeth to 32 permanent teeth begins at about 6 years of age, optimizing mastication and swallowing. Prior to molars erupting, children are able to bite off pieces of food with their incisors but unable to grind it adequately in preparation for swallowing, making them vulnerable to choking with particular food such as nuts, popcorn, grapes, and hotdogs.

The upper aerodigestive tract is divided into four main areas or spaces: the oral cavity, nasopharynx, oropharynx, and hypopharynx. These main spaces are further subdivided into smaller spaces including the piriform sinuses and vallecula, through which a bolus and liquids pass during a normal swallow. This is in comparison to

the lateral and anterior sulci, laryngeal vestibule, and laryngeal ventricle which are spaces that normally do not come in contact with the ingested bolus [2]. When residue of liquids or solids is noted in any of these spaces at the conclusion of a swallow, this is indicative of dysphagia. The major salivary glands including the parotid, submandibular, and sublingual glands found in the oral cavity produce 95% of saliva [7]. Minor salivary glands that line the oral mucosa produce additional saliva. Saliva aids with mastication and bolus preparation and transport. Saliva is mostly composed of water; however, the enzymes found within the saliva initiate the digestive process [7].

Neuroanatomy of Swallowing

Swallowing pathways involve a complex neuronal network including portions of the supratentorium (cortical and subcortical), infratentorium (brain stem), and peripheral nervous system (motor and sensory) [2]. Cortical regions including the primary and secondary sensorimotor cortices are active during the voluntary oral preparatory and oral transport phases of swallowing. Several cortical and subcortical sites that are active during the pharyngeal phase of swallowing include the primary and secondary cortices, insula, anterior and posterior cingulate cortices, basal ganglia, amygdala, hypothalamus, and substantia nigra. The medulla oblongata housed within the brain stem is especially active during the involuntary pharyngeal and esophageal phases of swallowing. The regulation of these two phases is aided by a central pattern generator (CPG) found within the medulla oblongata [8]. CPGs are neuronal networks that can produce rhythmic patterned outputs such as respiration and deglutition [8]. Motor neurons that are involved in the swallowing CPG are localized in the brain stem. These motor neurons include the trigeminal, facial, hypoglossal, and motor nuclei, the nucleus ambiguus, and the dorsal motor nucleus of the vagus nerve and two cervical spinal neurons (C1 and C3) [8]. Sensory neurons that regulate the pharyngeal and esophageal phases of swallowing are housed within the brain stem and include the nucleus of the solitary tract and the neighboring reticular formation [2]. Both motor and sensory neurons are found bilaterally within the medulla oblongata and form what is known as the swallowing center (swallowing CPG).

Muscle movements are controlled by several cranial and peripheral nerves and are coordinated within the swallowing center of the brain stem. Oral sensation is transmitted in the trigeminal nerve (CN V). Efferent information in the trigeminal nerve goes to the mylohyoid muscle, the anterior belly of the digastric muscle, and the four muscles of mastication: the masseter, temporalis, and pterygoid muscles. The facial nerve (CN VII) mediates taste sensation from the anterior 2/3 of the tongue. The facial nerve is also responsible for efferent control to the salivary glands, the muscles of facial expression, the stylohyoid, the platysma, and the posterior belly of the digastric muscle. The glossopharyngeal nerve (CN IX) carries taste

information from the posterior 1/3 of the tongue. The glossopharyngeal nerve innervates the stylopharyngeal muscle. The most important nerve for swallowing is the vagus nerve (CN X). The pharyngeal and laryngeal mucosae are innervated by the vagus nerve. A branch of the vagus nerve, the recurrent laryngeal nerve, transmits sensation from below the vocal folds and the esophagus. Efferent control in the vagus nerve is facilitated by the ambiguous nucleus (striated muscle) and the posterior nucleus of the vagus (smooth muscles and glands). The intrinsic and some of the extrinsic muscles of the tongue are innervated by the hypoglossal nerve (CN XII).

Muscular Control

Finely tuned coordination of more than 30 muscles located within the oral cavity, pharynx, larynx, and esophagus is necessary for a normal swallow (Table 1.1). The majority of the muscles involved with swallowing are striated, with the exception of the medial and distal esophagus, which have segments that are partially or

Table 1.1 Involved muscles and their innervation and function for the phases of deglutition

Involved muscle	Innervation	Function
<i>Oral preparatory phase</i>		
Orbicularis oris	CN VII	Closes oral fissure; compresses and protrudes lips
Buccinator	CN VII	Presses cheek against teeth
Masseter	CN V ₃	Elevates mandible; protrudes mandible
Temporalis	CN V ₃	Elevates and retracts mandible
Medial pterygoid	CN V ₃	Elevates mandible; protrudes mandible
Lateral pterygoid	CN V ₃	Protracts and depresses mandible
Superior longitudinal	CN XII	Curls tongue upward, elevating the tip and sides of tongue
Palatoglossus	CN X	Elevates posterior tongue; pulls soft palate onto tongue
Genioglossus	CN XII	Depresses central part of tongue to form a central trough; tongue protrusion; tongue deviation with unilateral contraction
<i>Oral transport phase</i>		
Genioglossus	CN XII	Depresses central part of tongue to form a central trough; tongue protrusion; tongue deviation with unilateral contraction
Hyoglossus	CN XII	Depresses tongue; retrudes tongue
Styloglossus	CN XII	Retrudes tongue; curls up sides of tongue
Palatoglossus	CN X	Elevates posterior tongue; pulls soft palate onto tongue
Superior longitudinal	CN XII	Curls tongue upward, elevating the tip and sides of tongue

Table 1.1 (continued)

Involved muscle	Innervation	Function
Levator veli palatini	CN X	Elevates soft palate
Musculus uvulae	CN X	Shortens and elevates uvula
Superior pharyngeal constrictor	CN X	Constricts pharyngeal walls
Mylohyoid	CN V	Elevates hyoid bone, floor of mouth, and tongue
Stylohyoid	CN VII	Elevates and retracts hyoid bone
Geniohyoid	CN XII; C1–C2	Moves hyoid bone anteriorly and superiorly
Anterior belly of digastric	CN V ₃	Depresses and stabilizes mandible; elevates hyoid bone
Posterior belly of digastric	CN VII	Elevates hyoid bone
Thyrohyoid	CN XII; C1	Depresses hyoid bone; elevates larynx
Stylopharyngeus	CN IX	Elevates pharynx and larynx
Palatopharyngeus	CN X	Tenses soft palate; pulls walls of pharynx superiorly, anteriorly, and medially
Salpingopharyngeus	CN X	Elevates pharynx and larynx
<i>Pharyngeal phase</i>		
Lateral cricoarytenoid	CN X	Adducts true vocal folds
Transverse arytenoid	CN X	Adducts true vocal folds
Thyroarytenoid	CN X	Relaxes vocal ligament; narrows laryngeal inlet
Hyoglossus	CN XII	Depresses tongue; retrudes tongue
Styloglossus	CN XII	Retrudes tongue; curls up sides of tongue
Superior pharyngeal constrictor	CN X	Constricts pharyngeal walls
Middle pharyngeal constrictor	CN X	Constricts pharyngeal walls
Inferior pharyngeal constrictor	CN X	Constricts pharyngeal walls
Mylohyoid	CN V	Elevates hyoid bone, floor of mouth, and tongue
Stylohyoid	CN VII	Elevates and retracts hyoid bone
Geniohyoid	CN XII; C1–C2	Moves hyoid bone anteriorly and superiorly
Anterior belly of digastric	CN V ₃	Depresses and stabilizes mandible; elevates hyoid bone
Posterior belly of digastric	CN VII	Elevates hyoid bone
Thyrohyoid	CN XII; C1	Depresses hyoid bone; elevates larynx
Cricopharyngeus	CN IX, X	Constricts pharynx at pharyngoesophageal junction
Proximal esophagus	CN X	Peristalsis
<i>Esophageal phase</i>		
Esophagus	CN X	Peristalsis

CN cranial nerve, C1 cervical spinal nerve 1, C2 cervical spinal nerve 2

completely smooth muscle [2] (Figs. 1.1, 1.2 and 1.3). Somatic afferent and efferent feedback is provided mainly by cranial and peripheral nerves for striated musculature and an autonomic enteric system for the smooth muscle [2]. The act of swallowing is divided into four phases: oral preparatory phase, oral transport phase, pharyngeal phase, and esophageal phase. The initial oral stages of deglutition are voluntary and trigger the subsequent involuntary pharyngeal and esophageal phases [10].

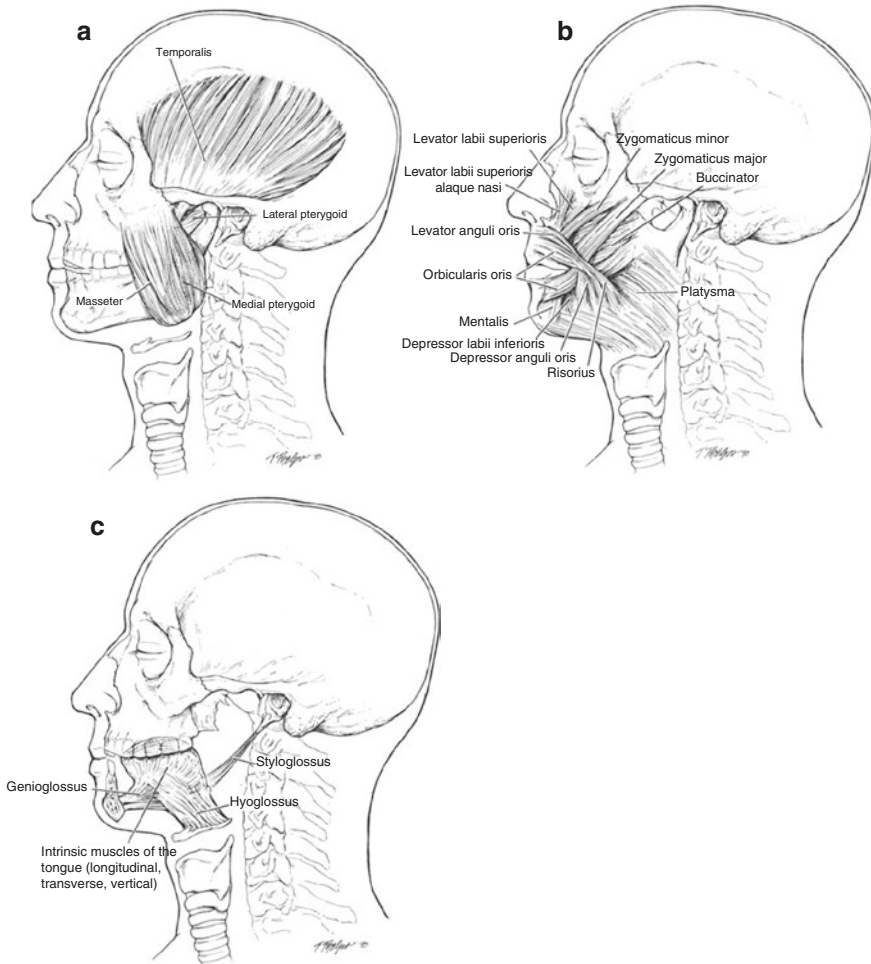


Fig. 1.1 Anatomical relationship of muscles contributing to the oral phase of swallowing. These muscles are controlled by discrete groups of motor neurons in the fifth (a), seventh (b), and twelfth (c) cranial motor nuclei. (From [9]. With permission of Springer)

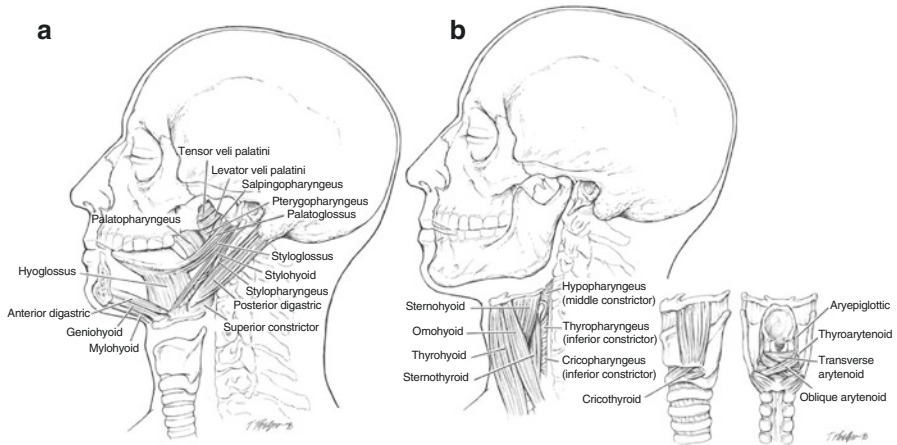


Fig. 1.2 Anatomical relationship of muscles contributing to the pharyngeal phase of swallowing. These muscles are controlled by discrete groups of motor neurons in the fifth, seventh, and twelfth cranial motor nuclei and by motor neurons in the cervical portions of the spinal cord. These muscles are thought of as acting in either the early (a) or late (b) pharyngeal phase of swallowing. The intrinsic and extrinsic laryngeal muscles also are shown (b). (From [9]. With permission of Springer)

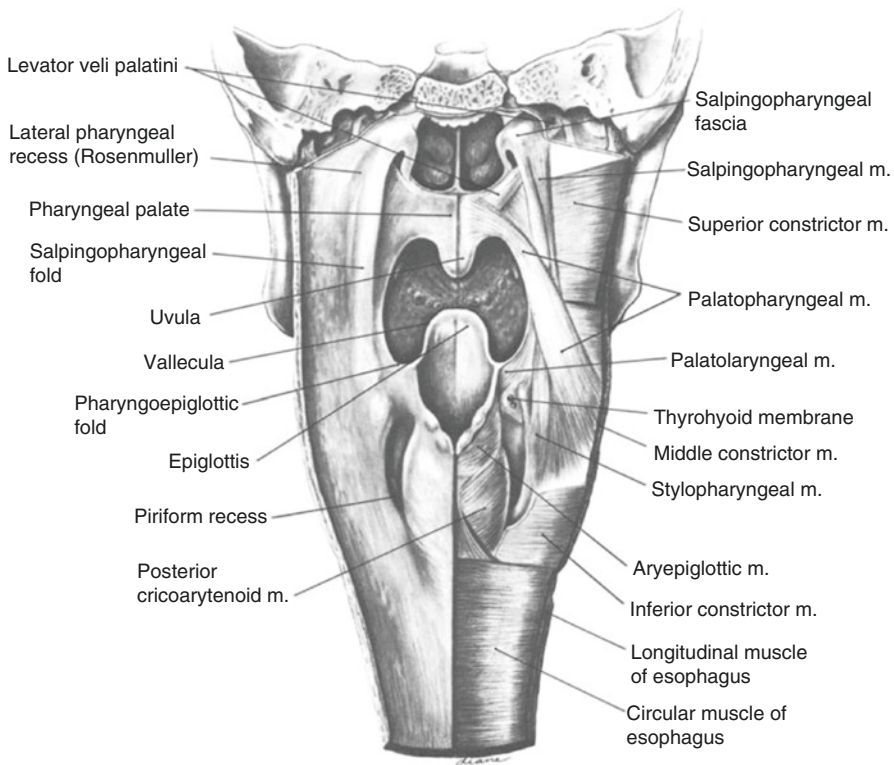


Fig. 1.3 Posterior view of internal pharyngeal musculature and recesses. The mucosa has been stripped from the left half of the preparation to better demonstrate the musculature. (From [9]. With permission of Springer)

Oral Preparatory Phase

The first phase of swallowing, the oral preparatory phase, breaks down food with mastication and forms a bolus in the oral cavity (Fig. 1.4). Bolus formation involves the coordination of lip, buccal, mandibular, and tongue movements. Closure of the upper esophageal sphincter (UES) during this phase is vital to prevent food or liquid from leaving the oral cavity until the individual is ready to initiate swallowing. This phase is under the voluntary control of three cranial nerves. The trigeminal nerve controls the muscles of mastication (temporalis, masseter, medial and lateral pterygoids) that help break down solid food by actively moving the mandible and also relays sensory information. As food particles are broken down, they are softened by saliva to aid with forming the bolus. The facial nerve coordinates the orbicularis oris and buccinator muscles that assist in food position and keep the oral cavity sealed without premature leakage into the oropharynx. Lateral and vertical tongue movements controlled by the hypoglossal nerve help position the food between the teeth. Once the bolus is formed, it is contained between the dorsal surface of the tongue and hard palate. The palatoglossus muscle depresses the soft palate and elevates the posterior tongue, creating a seal against the oropharynx. This prevents premature entry of the bolus into the pharynx. The bolus is captured over the dorsum of the tongue in a spoonlike form, as the genioglossus muscle contracts [2, 12].

Oral Transport Phase

Once a bolus has been formed, it is transitioned into the oropharynx in the oral transport phase. The tongue sits partly in the oral cavity and partly in the oropharynx. It is made up of eight pairs of muscles subdivided into intrinsic and extrinsic muscles. The four intrinsic muscles, vertical, transverse, superior longitudinal, and inferior

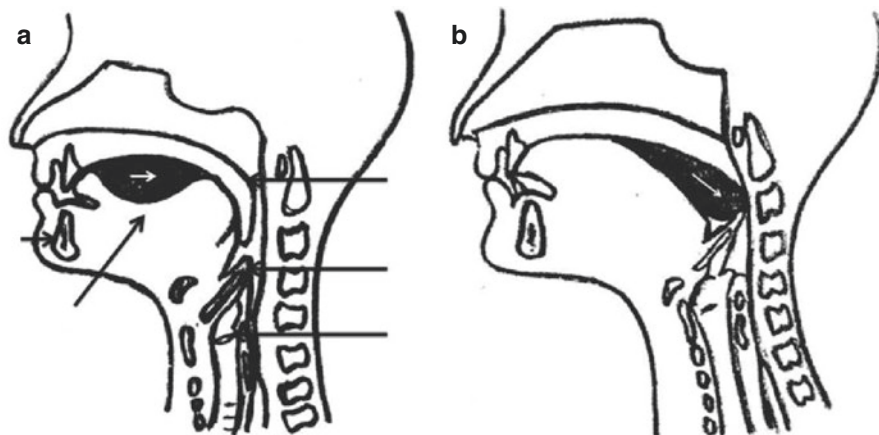


Fig. 1.4 Oral phase of swallowing: (a) the bolus is held between the anterior end of the tongue and the hard palate during the initiation of the oral phase, and (b) the bolus is propelled into the pharynx to trigger the pharyngeal phase. (From [11]. With permission of Springer)

longitudinal muscles, control the shape of the tongue [10]. The extrinsic muscles including the genioglossus, hyoglossus, styloglossus, and palatoglossus control the position of the tongue. The hypoglossal nerve innervates all the muscles of the tongue except the extrinsic palatoglossus muscles, which are innervated by the pharyngeal plexus [13]. These intrinsic and extrinsic muscles of the tongue elevate the tongue in an anterior to posterior fashion to push against the hard palate and propel the bolus toward the oropharynx in a wavelike motion [12]. Simultaneously, the soft palate elevates by contraction of the levator veli palatini and musculus uvulae while the base of tongue moves anteriorly and inferiorly to open the path to the oropharynx [2]. The soft palate also seals off the nasopharynx from the oropharynx, along with the contraction of the superior pharyngeal constrictors, which narrow the nasopharynx to aid with closure and prevent nasal regurgitation. The anterior-superior movement of the base of tongue, hyoid bone, and larynx due to the contraction of the suprahyoid muscles (mylohyoid, stylohyoid, geniohyoid, anterior digastric, and posterior digastric) and the thyrohyoid muscle widens the pharynx. The relaxation of the pharyngeal elevators, stylopharyngeus, palatopharyngeus, and salpingopharyngeus, also widens the pharynx transversely. A ramp is created due to the flattening of the posterior tongue, enabling the bolus to slide into the oropharynx [12].

Pharyngeal Phase

As the bolus is transported into the pharynx, the pharyngeal phase ensues (Fig. 1.5). The pharyngeal phase of swallowing is initiated voluntarily as the bolus crosses the anterior tonsillar pillars by sensory information transmitted by the glossopharyngeal and vagus nerves. Once triggered this complex phase is involuntary and generally lasts 1 second [10]. This pharyngeal swallow response can be affected and modified by food properties such as taste, volume, and texture [2]. When the pharyngeal swallow is triggered, respiration pauses to protect the airway by the contraction of the lateral cricoarytenoid,

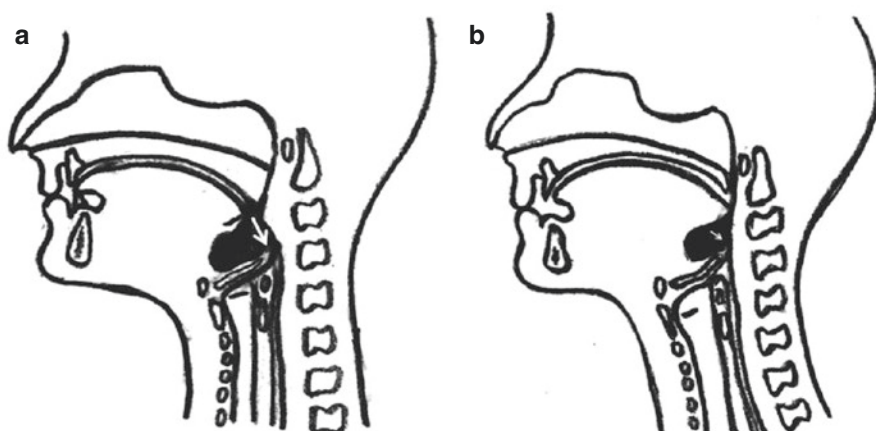


Fig. 1.5 Pharyngeal phase of swallow: the soft palate is elevated and in contact with the pharyngeal wall. The laryngeal inlet is protected by the epiglottis. (a) Bolus in the vallecula and (b) the tongue base retracted posteriorly toward the pharyngeal wall. (From [11]. With permission of Springer)