

Pediatric Dysphagia

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KEYWORDS

• Swallowing disorders • Feeding disorders • Dysphagia

KEY POINTS

- Feeding and swallowing disorders in the pediatric population are becoming more common, particularly in infants born prematurely and in children with chronic medical conditions.
- The normal swallowing mechanism is divided into 4 stages: the preparatory, the oral, the pharyngeal, and the esophageal phases.
- Feeding disorders have multiple causes; medical, nutritional, behavioral, psychological, and environmental factors can all contribute.
- Pathologic conditions involving any of the anatomic sites associated with the phases of swallowing can negatively impact the coordination of these phases and lead to symptoms of dysphagia and feeding intolerance.
- The most common examinations used to evaluate children with feeding disorders include bedside swallow evaluation, upper gastrointestinal series, videofluoroscopic swallow study, flexible endoscopic evaluation of swallowing, and flexible endoscopic evaluation of swallowing plus sensory testing.
- A multidisciplinary team armed with the knowledge of the complexity of the swallowing mechanism, an awareness of the pathologic conditions that can affect swallowing, and an understanding of the different clinical and instrumental testing options available is imperative when treating children with dysphagia.

INTRODUCTION

Feeding and swallowing disorders in the pediatric population are becoming more common, particularly in infants born prematurely and in children with chronic medical conditions.¹ A multidisciplinary approach to the workup of these patients is imperative to facilitate the early recognition of feeding problems, to identify the underlying conditions that may be contributing to the feeding problem, and to determine the most appropriate intervention necessary to achieve the best outcome for the child and the family. The multispecialty team should include the pediatrician, otolaryngologist, speech and language pathologist, occupational therapist, radiologist, dietician, and social worker, among others.

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The normal swallowing mechanism is divided into 4 stages: the preparatory, the oral, the pharyngeal, and the esophageal phases. The first 2 phases are under voluntary control, except in the newborn period, when the sucking reflex is regulated at the level of the brain stem.² The first phase is the preparatory phase, also known as the oral preparatory phase. In this phase food is taken into the oral cavity, chewed and moistened with saliva, and prepared into a bolus, which is held between the hard palate and oral tongue. This process first becomes evident at approximately 6 months of age. It is at this time that solids can be consumed. Before this age, the preparatory phase is limited to sucking from a nipple.

The second phase is the oral phase, during which food is moved from the mouth to the pharynx. Through highly coordinated movements, the soft palate elevates to prevent regurgitation into the nasopharynx and tongue movement propels food into the oropharynx.

The pharyngeal phase follows the oral phase. It begins as the bolus passes the tonsillar pillars and moves into the hypopharynx en route to the esophagus. During this phase, the palate elevates and approximates the pharyngeal musculature, cessation of breathing occurs, the larynx rises, the vocal folds adduct, and the base of tongue and pharyngeal muscles propel the bolus through a relaxed upper esophageal sphincter. The act of swallowing results in mechanical closure of the airway and transient cessation of breathing. The esophageal phase begins when the bolus enters the esophagus and ends when it passes into the stomach.

The act of swallowing is a complex act involving coordination between neural reflexes and voluntary effort that matures with development. An abnormality in neural or anatomic development can lead to swallowing dysfunction or dysphagia. Congenital or acquired structural or anatomic anomalies may cause airway and swallowing defects. This article provides an overview of the anatomic sites involved with feeding and swallowing, associated pathologic condition, clinical and instrumental evaluation, as well as treatment options.

EPIDEMIOLOGY

The overall incidence of dysphagia in children is increasing. A major contributor to this increase is the improved ability to care for infants born prematurely (<37 weeks' gestation). Survival rates for preterm infants have improved and the percentage of infants born prematurely has increased 20% since 1990.^{3,4} Early gestational age, low birth weight, and especially very low birth weight (<500 g or 1 lb, 2 oz) are strong predictors of infant mortality, morbidity, and cerebral palsy (CP).^{5,6} In premature children the neurologic ability necessary to achieve a coordinated, functional swallow has not developed, hence the high incidence of feeding issues in these patients.

The rising incidence of pediatric feeding issues can also be attributed to the increased life expectancy of infants with comorbidities, such as chronic lung disease and congenital abnormalities. The treatment of these conditions can involve repeated, persistent, or prolonged orotracheal or oroesophageal instrumentation, which can inhibit the development of a normal swallow.

CAUSES OF DYSPHAGIA

Feeding disorders have multiple causes; medical, nutritional, behavioral, psychological, and environmental factors can all contribute.^{7,8} The medical causes of dysphagia can be organized into diagnostic categories that include neuromuscular disorders, aerodigestive tract anatomic abnormalities, genetic abnormalities, mucosal and esophageal pathologic abnormality, and other conditions affecting suck/swallow/

breathing coordination.⁹ Rommel and colleagues¹⁰ categorized feeding problems as either medical, oral, or behavioral, stating that adequate management of medical and oral feeding disorders is beneficial because the stress induced by the feeding problem burdens the child's mental and psychologic development. The cause of the dysphagia may be due to a combination of causes further complicating the workup.¹¹

Neuromuscular Disorders

Neuromuscular disorders encompass many diseases that impair the functioning of the muscles, either directly, being pathologic conditions of the muscle, or indirectly, being pathologic conditions of nerves or neuromuscular junctions. A disorder of this type can potentially cause low muscle tone and poor coordination of the swallowing mechanism, which can result in respiratory compromise, aspiration, and poor weight gain. As initially proposed by Ramsay and colleagues,¹² the origin of a feeding-skills disorder is more likely to be neurophysiologic than experiential or environmental. This concept was illustrated by a study conducted by Rommel and colleagues that assessed feeding among 700 infants and young children with dysphagia and demonstrated a significant correlation between prematurity and feeding disorders. This study also noted that children with feeding disorders had a significantly lower birth weight for gestational age, which implies that feeding problems could be related to intrauterine growth retardation. More specifically, 38% of infants in this group needing assisted ventilation had an underlying neurologic condition. Preterm infants, who often have associated comorbidities, have been shown to be at risk for neuromuscular disorders, in addition to respiratory and developmental issues with subsequent dysphagia. A gestational age of 34 weeks is considered critical for the development of feeding efficiency and tolerance.^{13,14} The correlation between oral feeding disorders and a history of ventilation, aspiration, and nasogastric tube feeding indicates that medical intervention may influence oral feeding skills. These findings are strengthened by the work of Blaymore-Bier and colleagues,¹⁵ who demonstrated that premature low-birth-weight infants with prolonged intubation had significantly poorer sucking abilities at term and at age 3 months.

Central nervous system conditions are also associated with dysphagia. These conditions include CP, Arnold-Chiari malformations, and cerebral vascular accident. CP, in particular, is the most common neurogenic condition associated with dysphagia in children. Over the past 20 years, the life expectancy of children with CP has been increasing, and some think this is a contributing factor to the rising incidence of pediatric swallowing disorders. CP has been reported in 20% of infants born between 24 and 26 weeks' gestation and in 4% of infants born at 32 weeks' gestation.⁵ In a study by Selley and colleagues¹⁶ approximately 30% of children who had CP and were referred to a feeding program had histories of preterm birth. Children with other developmental abnormalities are also noted to have increased life expectancy as well. Increased awareness and detection have led to an increase in prevalence of pediatric dysphagia among children with neuromuscular abnormalities.

Anatomic Abnormalities of the Aerodigestive Tract

The 4 phases of swallowing are a coordinated effort designed to produce a bolus of food and direct that bolus to the stomach with simultaneous airway protection. The anatomic sites associated with the phases of swallowing include the nasal cavity, nasopharynx, oral cavity, oropharynx, hypopharynx, larynx, and esophagus. Pathologic conditions involving any of these anatomic sites can negatively impact the coordination of the phases of swallowing and lead to symptoms of dysphagia and feeding intolerance.

Nasal cavity and nasopharynx

Nasal and nasopharyngeal obstruction can impair breathing, which in turn negatively impacts the coordination of the oral and the pharyngeal phases of swallowing, leading to dysphagia, especially in newborn infants, who are obligate nasal breathers. Nasal obstruction can be caused by allergic rhinitis, adenoid or turbinate hypertrophy, choanal atresia, pyriform aperture stenosis, or congenital mass (encephalocele, dermoid, glioma). Bilateral nasal obstruction presents early in life, whereas unilateral obstruction may not present until much later with symptoms of nasal congestion and rhinorrhea. Bilateral choanal atresia is a severe form of nasal obstruction and usually presents as cyclical cyanosis in infancy relieved with crying.

Oral cavity and oropharynx

Mouthing and sucking movements have been observed by ultrasonography in fetus at 13 weeks' gestation. In the severely preterm infant, mouthing patterns persist until approximately 32 weeks' gestational age, when these disordered patterns of sucking bursts and pauses are replaced with identifiable rhythmic sucking and swallowing that more closely resemble a normal swallow.¹⁷ At approximately 3 to 4 months of age, the infant develops lateral tongue movements that allow some bolus manipulation, and by 6 months of age, infants can remove soft-textured food from a spoon. At 12 months, the sucking becomes less prominent and children transition to drinking from a cup. The ability to produce and propel a bolus of food continues to improve by 18 to 24 months of age.^{17–21} Development of the oral and pharyngeal phases of swallowing requires normal anatomy. Children with anatomic abnormalities of the oral cavity and oropharynx often present with poor feeding skills in infancy.²² Anatomic abnormalities include cleft lip/palate, genetic conditions such as CHARGE syndrome (coloboma, heart anomalies, choanal atresia, retardation of growth and development, and genital abnormalities), Treacher-Collins syndrome, Stickler syndrome, and Pierre-Robin sequence (retrognathia, glossoptosis, and cleft palate). Macroglossia can be associated with genetic conditions as well, such as Down and Beckwith-Wiedemann syndromes.

Hypopharynx and larynx

The pharyngeal phase of swallowing involves laryngeal elevation, transient cessation of breathing, and airway protection. Anatomic abnormalities that can contribute to dysphagia in infancy include laryngomalacia, vocal fold paralysis, vallecular cysts, posterior laryngeal cleft, and laryngeal webs.

Laryngomalacia is the most common cause of stridor in infants, accounting for approximately 70% of cases. Infants with laryngomalacia present with inspiratory stridor caused by the dynamic collapse of supraglottic tissue that occurs during inspiration. Laryngomalacia can be quite severe and can be accompanied by increased work of breathing, subcostal retraction, cyanosis, feeding difficulties, and subsequent failure to thrive. The respiratory distress can lead to a disorganized and poorly coordinated suck-swallow-breathe sequence. If the child takes a breath too soon in the sequence, they may aspirate before the bolus has had a chance to pass the larynx. Clinical findings are confirmed by flexible fiberoptic laryngoscopy performed by an otolaryngologist. More than 90% of infants with laryngomalacia do not require intervention because the symptoms are mild and typically resolve by age 6 months to 2 years of age. Almost all cases of laryngomalacia have signs of associated gastroesophageal reflux disorder (GERD).²³ GERD has been associated with laryngeal edema and decreased laryngeal sensation, which can lead to laryngeal residue or laryngeal penetration that does not initiate a cough response and may be aspirated.²⁴ Medical and behavioral treatment of GERD can improve the symptoms of

laryngomalacia and the associated dysphagia.²³ Severe or refractory laryngomalacia can be treated surgically by performing a supraglottoplasty, during which the posterior supraglottic tissues are trimmed and the aryepiglottic folds are cut, releasing a posteriorly displaced epiglottis.

Esophagus and trachea

The pharyngeal and esophageal phase of swallowing may be negatively impacted by pathologic conditions involving the esophagus and trachea, more specifically, tracheoesophageal fistula and inflammatory conditions, such as GERD and eosinophilic esophagitis.

Conditions Affecting the Suck-swallow-breathing Coordination

A coordinated suck-swallow-breathe sequence depends on the infant's ability to suck efficiently and swallow rapidly as the bolus is formed to minimize the duration of airflow interruption.²⁵ Preterm infants have a difficult time coordinating these activities and therefore have a high incidence of feeding difficulty. Lau and colleagues prospectively studied a group of full-term and premature infants and examined the relationship between suck-swallow and swallow-breathe. The preterm infants were found to swallow preferentially at different phases of respiration than those of their full-term counterparts. As feeding performance improved, sucking and swallowing frequency, bolus size, and suction amplitude increased. It is speculated that feeding difficulties in preterm infants are more likely to result from inappropriate swallow-respiration interfacing than suck-swallow interaction.

The anatomy of the oral cavity and the mandible changes with development. In the infant, the act of sucking is facilitated by the relatively small mandible and oral cavity with respect to the tongue. Sucking occurs when the infant's lips close around the breast or nipple, and the tongue seals against the pharynx, creating a closed system. The preterm infant generates lower suction pressures, with a smaller amount of milk received per sucking interval.²⁶ Preterm infants and those with conditions such as choanal atresia, laryngomalacia, bronchopulmonary dysplasia, and cardiac disease also may have a difficult time generating sufficient suction pressure and therefore fail to achieve an appropriate suck-swallow-breathe pattern, which leads to poor feeding skills in infancy.²²

EVALUATION TECHNIQUES

The evaluation of a child with a feeding disorder begins with a thorough history and physical examination by a physician, which is supplemented by a clinical swallow assessment performed by a qualified feeding specialist. Detailed information regarding the structure and function of the oral, pharyngeal, laryngeal, and upper esophageal swallow complex is collected. An assessment of the potential benefits of compensatory and treatment strategies is also determined. The most common examinations include bedside swallow evaluation, upper gastrointestinal series (UGI), videofluoroscopic swallow study (VFSS), flexible endoscopic evaluation of swallowing (FEES), and FEES plus sensory testing (FEES-ST).^{2,9} Other diagnostic testing that may be needed in assessing the cause of a swallowing dysfunction include computed tomographic imaging, pulmonary function testing, esophagoscopy, and bronchoscopy.

Bedside Swallow Examination

A bedside swallow evaluation is performed by a speech pathologist to gain a preliminary understanding of the clinical signs of dysphagia that a patient has. By introducing oral feeding material, which includes foods of different consistency and color, the

examination may help determine the cause of the dysphagia, the readiness of the patient to accept oral intake, and the ability of the patient to comply with subsequent radiographic studies. This information is invaluable to the rest of the multidisciplinary team with regard to further testing and selection of optimal swallowing instructions.

Upper Gastrointestinal Series

The UGI is a radiologic examination of the upper gastrointestinal tract and consists of a series of radiographic images delineating the esophagus, stomach, and duodenum. In the setting of dysphagia, a UGI can be helpful by noting anatomic and functional abnormalities, obstructions, as well as physiology of the oropharyngeal structures and UGI system.^{2,9}

Videofluoroscopic Swallow Study

The VFSS, also known as a modified barium swallow, is the most commonly used study to examine dysphagia and is particularly useful in assessing oropharyngeal dysphagia. Barium-impregnated liquid and solids are ingested, which images the structures of the oral cavity, pharynx, and cervical esophagus during swallowing. VFSS may also screen structures distal to the cervical esophagus.^{9,11} The main objectives of the VFSS are to provide information about the anatomy and possible structural pathologic abnormality that may be present, to investigate the movement and coordination of these structures with respect to bolus passage, and to define strategies to aid with safe and efficient feeding. It is also important to note that children must be able and willing to participate in this examination for it to be of any benefit.

It is also important to note the difference between a barium swallow and a modified barium swallow, or VFSS. A barium swallow is performed by a radiologist and radiology technician to evaluate esophageal swallowing from the cricopharyngeus to the gastroesophageal junction, useful in detecting mucosal abnormalities and signs of dysmotility. A modified barium swallow, or VFSS, on the other hand, is performed by a speech pathologist and a radiologist and evaluates oropharyngeal swallowing and all the anatomic structures involved,⁹ which allows the detection of aspiration and also for the administration of controlled volumes of a variety of consistencies containing barium.

In addition to UGI, barium swallow, and VFSS, other radiologic testing includes manometric testing, ultrasound, and nuclear scintigraphy. Manometric testing is beneficial in the assessment of the upper pharynx and esophageal motility. Although technically challenging in the pediatric population, manometric testing can also be used to investigate findings performed on barium swallow further by evaluating pathologic condition, such as GERD and stricture. Ultrasound allows a dynamic view of the movement of the tongue, oral cavity, hyoid, and larynx. Aspiration cannot be directly observed and the field-of-view may be limited because bony structures do not allow transmission of sound waves. Nuclear medicine scintigraphy may be used when assessing gastric emptying and GERD. This test is limited in its ability to determine aspiration amount and tends to underestimate reflux events.^{27,28}

Flexible Endoscopic Evaluation of Swallowing with or Without Sensory Testing

Otolaryngologists, in coordination with a speech pathologist, perform an FEES to evaluate the structure and function of the nasopharynx, oropharynx, and larynx during phonation and deglutition with and without liquids and solids. For pediatric patients, FEES is particularly helpful with patients who display an inability to handle secretions or cooperate with VFSS and have vocal fold dysfunction. FEES also has the advantage of being able to be performed on a baby while they are breast-feeding from their

mother. FEES-ST uses pulses of air that are administered during flexible endoscopy.^{9,27} Intact sensory feedback during a well-coordinated swallowing mechanism is necessary to prevent signs and symptoms of dysphagia. Sensory testing is useful in assessing patients preoperatively for airway reconstruction as well as patients with GERD or neurologic disorders who may have heightened sensory thresholds.

TREATMENT OPTIONS

Improvement of a feeding problem in a pediatric patient is most likely to occur when the underlying cause is determined and corrected. Outcomes data for the treatment of dysphagia are limited,⁹ in part because dysphagia is often secondary to multiple associated comorbidities or syndromes that in and of themselves have no definitive treatment. Premature infants and/or those infants with comorbidities may benefit from interventions that facilitate the development of oral motor skills, lessen the swallowing dysfunction, or provide supplemental nutrition, frequently requiring ongoing intervention and assessments by the speech pathologist and interdisciplinary team as the child matures. Compensatory swallowing therapies have been shown to improve safety and efficiency among infants and children with dysphagia. These strategies may involve positional adaptations, texture variations, bottle or nipple changes, and instructions to the feeder regarding modifications of the feeding technique such as pacing. It is important to communicate and work with the caregiver to ensure appropriate intervention and feeding exercises are followed.²⁹⁻³¹

Despite compensatory therapies, alternative feeding methods such as nasogastric and gastric tube feeding are required on either a short-term or a long-term basis.^{9,27,32-34} Children require tube feedings because they are at increased risk of aspiration, require nutritional supplementation, experience dysphagia for a prolonged duration, have associated comorbidities, or have significant gastroesophageal reflux leading to aspiration of gastric contents.

SUMMARY

The incidence of dysphagia in children continues to increase, largely because of the improved survival of premature infants and children with chronic medical conditions. The ability to obtain objective outcomes data remains difficult because the cause of feeding problems is often multifactorial. A multidisciplinary team armed with the knowledge of the complexity of the swallowing mechanism, an awareness of the pathologic conditions that can affect swallowing, and an understanding of the different clinical and instrumental testing options available is imperative when treating children with dysphagia.

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