


Dysphagia in infants with single ventricle anatomy following stage 1 palliation: Physiologic correlates and response to treatment

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Abstract

Background: Deficits in swallowing physiology are a leading morbidity for infants with functional single ventricles and systemic outflow tract obstruction following stage 1 palliation. Despite the high prevalence of this condition, the underlying deficits that cause this post-operative impairment remain poorly understood.

Objective: Identify the physiologic correlates of dysphagia in infants with functional single ventricles and systemic outflow tract obstruction following stage 1 palliative surgery.

Methods: Postoperative fiberoptic laryngoscopies and videofluoroscopic swallow studies (VFSS) were conducted sequentially on infants with functional single ventricles following stage 1 palliative surgery. Infants were dichotomized as having normal or impaired laryngeal function based on laryngoscopy findings. VFSS were evaluated frame-by-frame using a scale that quantifies performance within 11 components of swallowing physiology. Physiologic attributes within each component were categorized as *high functioning* or *low functioning* based on their ability to support milk ingestion without bolus airway entry.

Results: Thirty-six infants (25 male) were included in the investigation. Twenty-four underwent the Norwood procedure and twelve underwent the Hybrid procedure. Low function physiologic patterns were observed within multiple swallowing components during the ingestion of thin barium as characterized by ≥ 4 sucks per swallow (36%), initiation of pharyngeal swallow below the level of the valleculae (83%), and incomplete late laryngeal vestibular closure (56%) at the height of the swallow. Swallowing deficits contributed to aspiration in 50% of infants. Although nectar thick liquids reduced the rate of aspiration ($P = .006$), aspiration rates remained high (27%). No differences in rates of penetration or aspiration were observed between infants with normal and impaired laryngeal function.

Conclusions: Deficits in swallowing physiology contribute to penetration and aspiration following stage 1 palliation among infants with normal and impaired laryngeal function. Although thickened liquids may improve airway protection for select infants, they may inhibit their ability to extract the bolus and meet nutritional needs.

KEYWORDS

dysphagia, feeding, hypoplastic left heart syndrome, single ventricle, stage 1 palliation, swallowing

1 | INTRODUCTION

Congenital heart disease is the most prevalent birth defect in the world, affecting over one million infants each year.¹ Among the most severe of these defects are those that include functional single ventricles and systemic outflow tract obstruction.² Survival of infants with functional single ventricles and systemic outflow tract obstruction, such as those with hypoplastic left heart syndrome, requires palliative heart surgery within the initial days of life. Although recent advances in surgical techniques used in the initial palliative procedure have improved infant rates of survival, high rates of postoperative morbidity continue to pose serious health threats once acute cardiac concerns have resolved.^{3,4}

Impairment in the safe obtainment of oral nutrition is a leading morbidity following stage 1 palliation.^{3,5-9} Such deficits, further referenced as feeding deficits, affect as many as 74% of infants following stage 1 palliation,⁹ and are associated with longer hospital stays, increased caregiver distress, and higher rates of interstage mortality.^{8,10-12} Although scientific advances have enabled clinicians to optimize postoperative nutrition among infants with feeding deficits through the use enteral feeding tubes¹³ and associated feeding algorithms,^{14,15} similar advances have not occurred in the development of swallowing interventions that facilitate the safe obtainment of this nutrition by mouth. Instead, clinicians frequently rely on compensatory interventions, such as the use of thickened liquids, with limited evidence to support their beneficial effect in effort to facilitate safe oral intake until swallow function improves.

Despite the appreciated need for targeted, evidence-based, swallowing interventions, the ability to develop such interventions has been stifled by the fact that the pathophysiology underlying these functional impairments remains unclear. Recurrent laryngeal nerve injury acquired during the initial surgical repair has previously been postulated as a potential source for the aforementioned swallowing deficits.¹⁶⁻¹⁸ While it is appreciated that the resulting impairments in laryngeal function place these infants at increased risk for airway compromise, previous reports of high aspiration rates among infants without deficits in laryngeal function suggest other impairments in the feeding-swallowing mechanism may exist.^{3,19} Identification of the underlying source of postoperative feeding and swallowing impairment is critical for the development of targeted interventions that will improve oral intake and postoperative health among these fragile infants.

The current pilot investigation is the first step toward filling this void by systematically examining the physiologic correlates of dysphagia in infants with functional single ventricles and systemic outflow tract obstruction following stage 1 palliative surgery through the following aims: (1) Identify the physiologic correlates of dysphagia during the ingestion of thin barium contrast; (2) Test the effect of nectar thick barium contrast on rates of low function oropharyngeal swallow attributes; and (3) Test the difference in rates of bolus airway entry between infants with normal and impaired laryngeal function. We hypothesized infants with functional single ventricles and systemic outflow tract obstruction would exhibit high rates of low function oropharyngeal swallowing physiology, penetration, and aspiration regardless of liquid viscosity or laryngeal function.

2 | METHODS

2.1 | Data collection

Postoperative fiberoptic laryngoscopic examinations and videofluoroscopic swallow studies (VFSS) were sequentially conducted on all infants presenting with functional single ventricles following stage 1 palliative surgery for systemic outflow tract obstruction. Infants were excluded from study participation if they were diagnosed with a syndrome, genetic anomaly, noncardiac congenital malformation, or neurologic condition associated with dysphagia. Laryngoscopies were completed by a pediatric otolaryngologist at the patient's bedside. All were performed with a pediatric 2.5 mm laryngoscope and free of topical nasal anesthetics. Laryngeal function was evaluated for vocal fold mobility according to classifications previously described by Skinner et al. (2005).³ Vocal fold paralysis was defined as absent movement within one or both of the vocal folds, and vocal fold paresis as impaired mobility in one or both of the vocal folds. The presence of a glottic gap, defined as a glottic aperture during adduction in the context of normal vocal fold mobility, and laryngomalacia, as defined by prolapsing supraglottic tissue, were also reported as observed by the examining physician. Videofluoroscopic swallowing examinations were conducted using a standardized exam protocol, executed by a speech-language pathologist and radiologist, once infants were deemed hemodynamically stable by their primary medical team. Infants were reclined at 70° and positioned in the lateral viewing plane. The fluoroscopic visualization field was collimated to include the lips anteriorly, nasal cavity superiorly, cervical spinal column posteriorly, and the pharyngoesophageal segment inferiorly. Swallowing was evaluated using continuous fluoroscopy as the infants ingested standardized thin and nectar Vari-bar barium contrast (Bracco Diagnostics Inc., Township, NJ). Alterations to nipple type and feeding method were made at the speech-language pathologist's discretion as clinically indicated. All exams were recorded at 30 frames/second on the KayPENTAX Digital Swallow Workstation (PENTAX Medical, Montvale, NJ).

2.2 | Data analysis

Laryngoscopy results were used to categorize infants exhibiting laryngomalacia, vocal fold paralysis, vocal fold paresis, or glottic gap as having *impaired laryngeal function*, with all others categorized as having *normal laryngeal function*. Charts were reviewed for type of heart defect, surgical approach and feeding history. Videofluoroscopic exams were scored frame-by-frame on a high definition 2560 × 1440-pixel screen by two speech-language pathologists using an ordinal scale that ranks performance within 11 physiologic components of oropharyngeal swallow function found to be of clinical significance to oral intake in the bottle-fed infant (NIH, NIDCD, R01DC011290). Function within each component was scored using an operationally defined rank ordered scale with values ranging from 0, representing the highest level of swallow function, and increasing in number to represent the lowest level of swallow function within each oropharyngeal swallowing component. Bolus airway relationships were categorized as complete airway

protection, penetration, or aspiration. Raters met >80% reliability criteria for their rating of all oropharyngeal swallowing components and bolus airway relationships. Discrepancies between rater scores were resolved by consensus. Thin and nectar thick barium overall impression scores were assigned and represented the worst function within each physiologic component across all visualized swallows of a given viscosity. In the absence of established pediatric oropharyngeal swallowing norms, literature review and expert consensus was used to determine the physiologic attributes within each oropharyngeal swallowing component that were considered *high function*, and those that were considered to be *low function*. High function attributes included component scores that were considered to be variants of normal swallow function given their ability to support safe and efficient milk ingestion. Low function attributes included those component scores most typically associated with feeding-swallowing impairment because of clinical observations that support an associated risk for bolus airway entry or insufficient volume of milk ingestion.

High and low function correlates were summarized using frequencies and proportions. McNemar's test was used to compare the proportion of low function attributes between thin and nectar thick barium contrast among infants who received both viscosities. Fisher's exact test was used to test for associations between laryngeal function impairment status and penetration/aspiration status for thin and nectar bolus. *P*-values <.05 were considered statistically significant. Descriptive measures for continuous variables were median and interquartile range. Descriptive measures for categorical variables were frequencies and percentages. The research protocol was approved by the Institutional Review Board at the Medical University of South Carolina.

3 | RESULTS

3.1 | Sample demographic and surgical characteristics

Between May 2010 and September 2014, 38 infants with functional single ventricles and systemic outflow tract obstruction underwent a routine postoperative laryngoscopy and videofluoroscopic swallow study (VFSS). One of these infants was excluded from study participation due to the presence of a known syndrome (Down syndrome), and another due to the presence of neurologic impairment associated with dysphagia (agenesis of the corpus callosum). A total of 36 infants were included in the investigation. The majority of these infants had hypoplastic left heart syndrome (81%, 29), with other diagnoses including right ventricle dominant atrioventricular septal defect (6%, 2), mitral and aortic stenosis (6%, 2), interrupted aortic arch with ventricular septal defect (3%, 1), double outlet right ventricle with straddling mitral valve (3%, 1), and double inlet left ventricle with interrupted aortic arch (3%, 1). Norwood procedure was the primary stage 1 surgical approach, with the remainder undergoing the Hybrid procedure. Routine postoperative laryngoscopy revealed impaired laryngeal function in 42% (15) of the sample, with the most common laryngeal anomalies being glottic gap and laryngomalacia. Table 1 provides a full description of patient demographics, preoperative diagnosis, surgical approach, and postoperative laryngeal function.

TABLE 1 Sample demographics, surgical, and postoperative evaluation characteristics

Variable	Summary measure ^a
<i>Demographics</i>	
Sex	
Male	25 (69)
Female	11 (31)
Race	
African American	12 (33)
Caucasian	21 (58)
Asian	1 (3)
More than one race	1 (3)
Unknown	1 (3)
<i>Stage I Surgery</i>	
Age at procedure (days)	6 (4.75–9)
Norwood procedure	24 (67)
Hybrid procedure	12 (33)
<i>Laryngeal Endoscopy</i>	
Postoperative day (day)	10 (6–14.5)
Normal laryngeal function	21 (58)
Impaired laryngeal function ^b	15 (42)
Unilateral vocal fold paresis	3 (8)
Unilateral vocal fold paralysis	2 (6)
Glottic gap	6 (17)
Laryngomalacia	5 (14)
<i>Videofluoroscopic Swallow Study</i>	
Age at procedure (days)	28 (20–36)
Postoperative day (day)	19.5 (14–27)
Feeding method at VFSS	
Nil per os	10 (28)
Tastes via pacifier	20 (56)
Restricted partial bottle	6 (17)

^aSummary measures for categorical variables are presented as frequencies (percent). Summary measures for continuous variables are presented as median (interquartile range).

^bThe total of Impaired Laryngeal Function subgroup frequencies exceeds 15 because one subject exhibited both laryngomalacia and glottic gap.

3.2 | Physiologic correlates of dysphagia during the ingestion of thin barium contrast

VFSSs were completed a median of 20 days following surgery. At the time of VFSS all infants had enteral feeding tubes in place, with the majority of infants solely receiving tastes of milk via pacifier. Low function physiologic patterns were observed within multiple oropharyngeal swallowing components during the ingestion of thin barium contrast (Table 2). Nutritive sucking exhibited deficits in sucking rhythmicity (28, 78%) and the number of sucks per swallow (13, 36%). These were characterized by the exertion of ≥ 4 sucks prior to the initiation of the pharyngeal swallow, that were frequently arrhythmic in pattern. Once the bolus was formed, the majority of infants did not initiate the pharyngeal swallow until the bolus resided below the valleculae (30, 83%), where it frequently remained for >1 second (28, 78%). Low function physiology was also observed in the physiologic processes facilitating laryngeal closure and pharyngeal bolus clearance. These included reductions in late laryngeal vestibular closure at the height of the swallow (20, 56%), epiglottic inversion (21, 58%), tongue base retraction (11, 31%), pharyngeal stripping wave (26, 72%), and

TABLE 2 Differences in low function oropharyngeal swallow physiology between thin and nectar barium contrast on VFSS (N = 30)

Oropharyngeal swallow component	Low function attributes	Summary measure ^a		
		Thin	Nectar	p ^b
Number of sucks to form bolus	≥4 sucks to form bolus	9 (30)	20 (67)	.003
Sucking rhythmicity	Intermittent or arrhythmic sucking	24 (80)	25 (83)	1.0
Bolus location at initiation of swallow	Bolus below the level of the valleculae prior to swallow initiation	24 (80)	19 (63)	.2
Timing of swallow initiation	Bolus remaining in location of swallow initiation for >1 second	22 (73)	26 (87)	.3
Late laryngeal vestibular closure	Incomplete laryngeal vestibular closure at the point of maximal oropharyngeal swallow contraction	18 (60)	9 (30)	.04
Epiglottic inversion	Incomplete inversion	18 (60)	14 (47)	.4
Tongue base retraction	Narrow or wide column of contrast between tongue base and posterior pharyngeal wall at maximal contraction	7 (23)	9 (30)	.6
Pharyngeal stripping wave	Incomplete pharyngeal stripping wave	22 (73)	24 (80)	.6
Pharyngoesophageal segment opening	Incomplete distension or duration with obstruction to bolus flow	25 (83)	25 (83)	1.0
Penetration	Bolus entry into the laryngeal vestibule without progression below vocal folds	8 (27)	9 (30)	1.0
Aspiration	Bolus entry into the laryngeal vestibule with progression below vocal folds	18 (60)	8 (27)	.006

^aSummary measures are expressed as frequency (percent).^bP-values based on McNemar's test.

pharyngoesophageal segment opening (30, 83%). About 83% (30) of infants exhibited thin barium penetration (12, 33%) or aspiration (18, 50%) during the exam.

3.3 | Effect of nectar barium contrast on rates of low function swallowing attributes

Videofluoroscopic recordings of swallow function were obtained during the intake of thin barium for all 36 infants, however, visualization of nectar barium ingestion was prohibited in 6 infants due to patient rejection of the material. When compared with thin swallows, significantly more infants were found to attain complete laryngeal vestibular closure at the height of the swallow on nectar swallows. While this resulted in significantly lower rates of nectar barium aspiration when compared with thin, nectar aspiration rates remained high. In contrast to the improvements in laryngeal vestibular closure, nectar barium was found to result in significantly more infants requiring ≥4 sucks per swallow when compared with thin barium (Table 2).

3.4 | Effect of impaired laryngeal function on thin and nectar bolus airway entry

No significant associations were found between infants' laryngeal function and penetration/aspiration status for either thin or nectar swallows (Table 3).

4 | DISCUSSION

Feeding impairments are a leading morbidity in infants with functional single ventricles and systemic outflow tract obstruction following stage 1 palliation.^{6-9,20} The systemic health threats posed by feeding impairments are exacerbated in these fragile infants due to their diminished cardiopulmonary reserve and the importance of obtaining adequate nutrition for future surgical success.²¹ Despite this understanding, little is known about the physiologic deficits in oropharyngeal swallow physiology that contribute to these postoperative feeding impairments in this high-risk population.

In the current investigation, we used a standardized rating metric to systematically evaluate components of oropharyngeal swallow physiology and their relationship to bolus flow among infants with functional single ventricles and systemic outflow tract obstruction following stage 1 palliative repair. One of the primary findings from the current investigation was the presence of high postoperative aspiration rates during the ingestion of thin barium contrast in the presence of normal laryngeal function. This is consistent with previous findings of Skinner et al. (2005), who found aspiration among 13% of infants with normal laryngeal function following stage 1 palliative repair.³ The shared observations of aspiration in the presence of normal laryngeal function in Skinner's study and the current investigation implicate other impairments in swallowing physiology that extend beyond deficits in laryngeal function.

TABLE 3 Associations between laryngeal function impairment status and penetration/aspiration status for thin and nectar thick barium contrast

	Thin		<i>P</i>	Nectar		<i>P</i> ^a
	Normal laryngeal function N = 21	Impaired laryngeal function N = 15		Normal laryngeal function N = 16	Impaired laryngeal function N = 14	
Penetration/aspiration status			.7			.2
None	3 (14)	3 (20)		6 (38)	7 (50)	
Penetration	8 (38)	4 (27)		7 (44)	2 (14)	
Aspiration	10 (48)	8 (53)		3 (19)	5 (36)	

Measures shown are frequency (percent).

^aFisher's exact test.

Successful oral intake requires transformation of the upper aerodigestive tract from a conduit for respiration, to a conduit for bolus flow. The ability to successfully transition between these functions is dependent on the precise, coordinated contraction of 31 paired muscles and 5 cranial nerves.²² These refined movements divert the laryngeal vestibule away from the path of bolus flow before the bolus descends through the pharynx. As a result, deficits in the timing, or in the integrity, of these movements place infants at increased risk for bolus airway entry and incomplete clearance of ingested material through the pharynx. Our findings indicate high postoperative rates of impairment within these critical airway protective and bolus clearance processes. The majority of infants did not initiate movements for airway protection until the bolus had reached the pyriform sinuses; placing them at high risk for aspiration upon pharyngeal contraction during the swallow. Once these movements were initiated, infants were also found to have deficits in their ability to obtain complete late laryngeal vestibular closure. Although the consequences of such deficits are likely exacerbated in infants who have no secondary airway protective mechanisms at the level of the glottis due to impairments in recurrent laryngeal nerve function, these physiologic deficits also pose a high risk for bolus airway entry among infants with intact laryngeal function. While the identification of the underlying source responsible for the observed physiologic impairments is beyond the scope of the current investigation, future work exploring these underlying mechanisms is necessary to identify targets for future dysphagia interventions. Swallowing is a highly dynamic process that is influenced not only by bolus attributes and anatomic integrity, but also by neurologic, muscular, respiratory, and gastrointestinal function.^{23–26} Altered prenatal circulatory patterns, and post-surgical stresses of in parallel circulation, cyanosis, and ventricular volume load can have deleterious effects on these systems^{27–34} and may contribute to the observed swallowing deficits.

Currently, the provision of thickened liquids is a commonly used intervention to facilitate safe milk ingestion in the presence of thin liquid aspiration.^{35,36} This intervention is aimed at slowing the rate of milk flow through the pharynx in effort to promote a timely initiation of swallow and obtain complete airway protection. Although this intervention may have beneficial effects for some, the ability to achieve these effects is limited by the nature and severity of the swallowing impairment. Our findings indicate that while nectar barium contrast

allowed significantly more infants to swallow without aspiration, many infants were resistant to this beneficial treatment effect and continued to aspirate the nectar contrast (Table 2). These findings are hypothesized to reflect the severity of swallowing impairment among select postoperative infants, and highlight the importance of executing an instrumental swallowing assessment prior to clinical implementation. Although the clinical implementation of thickened liquids may provide substantial swallowing benefits for select infants, they have the potential to pose significant health threats to others. In addition to exacerbating swallowing deficits in the presence of select physiologic impairments (i.e., pharyngeal clearance), thickened liquids have also been found to reduce nutrient absorption,^{37,38} and have been attributed as a potential source of necrotizing enterocolitis.³⁹ These findings, in conjunction with our findings that revealed high rates of aspiration and reductions in sucking efficiency during the ingestion of nectar barium contrast, indicate the arbitrary thickening of liquids without objective fluoroscopic assessment, follow-up bedside exam, and consideration of the infant's overall systemic health, has the potential to inflict the opposite of its intended treatment effect.

4.1 | Limitations and future research directions

Although this investigation is the first to the authors knowledge to systematically evaluate characteristics of oropharyngeal swallow physiology in infants with systemic outflow tract obstruction following stage 1 palliation, it is limited by several factors. Due to the ethical implications and regulatory restrictions prohibiting the exposure of healthy infants to radiation during a videofluoroscopic exam, there remains limited understanding of all physiologic attributes that constitute normal swallow function. In the absence of literature based pediatric norms, characteristics of oropharyngeal swallow physiology were dichotomized as high function or low function based on available pediatric norms, normal adult physiology, and theoretical physiologic risk when pediatric norms were not available. Future studies identifying the characteristics of normal oropharyngeal swallow physiology and bolus airway relationships among infants are necessary to further discriminate between normal and impaired swallowing physiology. Another limitation of the current investigation is the use of a small heterogeneous sample. Future investigations are warranted that employ larger sample sizes

and stratify for potential variables that influence oropharyngeal swallow physiology, such as cardiac status, surgical approach, gestational age, and prior oral feeding experience. Lastly, while the current investigation controlled for barium viscosity through the use of standardized thin and nectar barium contrast, it did not standardize the type of bottle nipple that was used for barium presentation. Nipples were chosen based on those that were clinically available at the time the fluoroscopic exam was completed, with flow rates modified real-time based on the clinician's assessment of infant need. Although there have been a paucity of investigations examining the effect of nipple attributes on oropharyngeal swallow physiology among infants with congenital heart defects, past investigations within the preterm infant have demonstrated the ability to improve feeding ventilation by using nipples that reduce the rate of milk flow.⁴⁰ Respiration and swallowing are tightly linked, interdependent processes. Therefore, interventions that improve respiratory stability may concurrently improve swallowing physiology and airway protection. Future investigations examining the effect of clinically available interventions aimed at optimizing respiratory performance, such as reduced flow rate nipples or respiratory enhancing feeding postures, are necessary to guide the clinical treatment of these oropharyngeal swallowing deficits.

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CONFLICT OF INTEREST

The authors declare that they have no conflicts of interest with the contents of this article.

AUTHOR CONTRIBUTIONS

Study design was completed by Dr. McGrattan, Dr. Martin-Harris, Dr. Lefton-Greif, Dr. Zyblewski, and Dr. Bradley. Development of the VFSS evaluation tool was completed by Dr. Martin-Harris and Dr. Lefton-Greif. Videofluoroscopic and endoscopic data collection was completed by Dr. McGrattan, Heather McGhee, and Dr. Halstead. Collection of cardiothoracic and medical data was performed by Dr. Zyblewski. Videofluoroscopic data analysis was completed by Dr. McGrattan and Heather McGhee. Statistical Analysis was completed by Allan DeToma and Dr. Hill. All authors were involved in the preparation of the manuscript and provided final review prior to submission.

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