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Clinical Presentation and Surgical Outcomes in Adolescents and Pre-Adolescents with Vascular Rings

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Abstract

***Background*:** Vascular rings (VRs) produce multiple respiratory and swallowing symptoms, which may vary by age. This study characterized symptoms for pre-adolescent and adolescent VR patients and evaluated whether surgical intervention is associated with clinical improvement.

Methods: Single-institution, retrospective review of patients aged 9–21 years with complete and incomplete VRs. Patients with complex cardiovascular comorbidities, esophageal atresia/tracheoesophageal fistula, or prior VR repair were excluded. Symptoms were compared pre and postoperatively.

Results: 49 patients (median age 13.1 years, IQR 11.2–16.1) were included. Diagnoses consisted of right aortic arch with aberrant left subclavian artery (n=25), double aortic arch (n=7), left aortic arch with aberrant right subclavian artery (n=16), and right aortic arch with mirror-image branching and a left ligamentum (n=1). At baseline, specific individual symptoms and overall symptom counts did not differ significantly between the surgical (n=33) and non-surgical (n=16) cohorts. Significant symptom reduction among the surgical cohort was observed for adaptive eating behaviors (pre-op 76% vs. post-op 9%, $p<0.001$), dysphagia with solids (pre-op 49% vs. post-op 9%, $p<0.001$), sensation of food getting stuck (pre-op 39% vs. post-op 15%, $p=0.039$), dyspnea on exertion (pre-op 76% vs. post-op 15%, $p<0.001$), chest pain (pre-op 46% vs. post-op 9%, $p<0.001$), increased URIs (pre-op 36% vs. post-op 3%, $p<0.001$), and chronic cough (pre-op 30% vs. post-op 0%, $p=0.002$).

Conclusion: Pre-adolescent and adolescent vascular ring patients present with specific symptoms, such as dyspnea on exertion, chest pain, and adaptive eating behaviors. Surgical

repair is associated with symptom reduction. Improved symptom recognition can optimize VR management.

Introduction

Vascular rings (VR)—which consist of complete rings such as right aortic arch with aberrant subclavian artery (RAA ALSA), variants of double aortic arch (DAA), right aortic arch with mirror-image branching and left ligamentum arteriosum (RAA MIB); and incomplete rings, notably left aortic arch and aberrant right subclavian artery (LAA ARSA)—are associated with respiratory and swallowing symptoms of varying onset and severity. Commonly identified symptoms include noisy breathing or stridor (32%–91.8%), persistent or chronic cough (13.7%–63.5%), frequent upper respiratory infections (4.7%–47.0%), dysphagia (15.2%–50%), and dyspnea on exertion (11.5%–34.1%); these symptoms are primarily documented in infants and toddlers with a median operative age frequently less than 2 years old [2–11]. Older children and adolescents, while less visible in the literature, also require comprehensive VR treatment and surgery. A review from Texas Children's Hospital found that 10.8% of patients who underwent VR repair were over 5 years old, while at Royal Children's Hospital, 4.1% of operated patients were older than 10 years [9,10]. At Boston Children's Hospital, 31.4% of patients with complete VRs were over 10 years old at time of operation, suggesting the possibility of a late diagnosis, persistent symptom burden, or new onset of symptoms in those with known VR [7].

Currently, sparse age-specific guidance is available regarding symptoms for pre-adolescents and adolescents navigating a VR diagnosis. Prior studies of patients undergoing VR reoperations, who tend to be older on average, suggest that these patients present with more

subtle residual symptoms, including adaptive eating behaviors and exercise limitation [7,12]. Patients with LAA ARSA have also been shown to present at older ages, with dysphagia being a primary symptom [1,3,13–15]. However, there is limited research to support older children, their families, and clinicians in monitoring aerodigestive symptoms associated with vascular rings and determining indications for surgery. Moreover, the relatively lower operative frequency has limited a comprehensive evaluation of surgical outcomes within this age cohort.

The present study aimed to characterize symptoms and postoperative outcomes among pre-adolescent and adolescent patients diagnosed with VRs. Addressing this gap will provide clinical guidance for patients, families, and providers navigating uncertainty in VR diagnosis and treatment to facilitate timely and effective symptom management, diagnostic testing, and referral to specialized surgical care.

Methods

We completed a retrospective chart review of patients aged 9–21 years with VRs (i.e., DAA, RAA ALSA, RAA MIB and left ligamentum, LAA ARSA) who underwent structured preoperative and postoperative symptom monitoring through the Boston Children's Vascular Ring and Airway Program and/or the Esophageal and Airway Treatment (EAT) Clinic. The Vascular Ring and Airway Program brings together specialists in cardiac surgery, general surgery, cardiology, gastroenterology, pulmonology and otolaryngology to collaboratively review VR patient symptoms and imaging. This multidisciplinary team develops individualized recommendations for ongoing monitoring, diagnostic evaluation, or surgical intervention.

Surgical recommendations are guided primarily by symptom prevalence and severity. Operative planning is individualized to patient anatomy and associated airway and esophageal findings.

Our study included both non-surgical and surgical patients who presented to our institution with a VR diagnosis. Given this study's focus on pediatric patients, 21 years was established as the upper age bound. The minimum cutoff of 9 years of age was established based on internal database review, corresponding to the oldest quartile of operated pediatric patients at our institution. Patients with major cardiovascular comorbidities such as Tetralogy of Fallot, Double Outlet Right Ventricle, Hypoplastic Left Heart Syndrome, or heterotaxy were excluded, as were patients with esophageal atresia/tracheoesophageal fistula. This study excluded patients with a prior VR operation before the age of 9. Patients who underwent VR operations at other institutions were excluded regardless of age.

Respiratory and swallowing symptoms were characterized and identified through provider documentation in the electronic medical record. Surgical and non-surgical groups were compared with respect to initial individual clinical presentation using Fisher's exact test, and differences between these independent cohorts in the baseline number of symptoms were evaluated using the Mann-Whitney U-test. Postoperative symptoms in the surgical group were recorded at the patient's most recent visit with the multidisciplinary Vascular Ring and Airway or EAT team. Pre-to-postoperative symptom changes within the surgical cohort were evaluated using the McNemar test for binary matched pairs, and the Wilcoxon signed-ranks test was utilized to assess differences in the median number of total symptoms. Statistical testing was conducted using IBM SPSS Statistics version 32 (IBM Corporation, Armonk, NY). Two-sided $p<0.05$ were considered statistically significant. A formal waiver of review was obtained by the Institutional Review Board at Boston Children's Hospital (IRB-P00042409).

Results

Patient Characteristics and Symptoms

We identified 49 patients (32 female) who were first evaluated for a vascular ring consultation between 2016 and 2025. Of these, 48 patients were first evaluated between 2021 and 2025, while one patient had been followed longitudinally since 2016. The median age at the first preoperative encounter was 13.1 years [IQR 11.2–16.1, range 9.0–20.4]. Primary diagnoses included RAA ALSA (n=25), LAA ARSA (n=16), DAA (n=7), and RAA MIB (n=1). The most common baseline respiratory symptoms noted across the total cohort were dyspnea on exertion (67.3%), chest pain or tightness at rest (40.8%), and increased or prolonged URIs (36.7%). The most common preoperative swallowing symptoms were adaptive eating behaviors (71.4%), solid food dysphagia (51.0%), and a sensation of food getting stuck (42.9%).

Out of 49 patients within the total cohort, 33 underwent vascular ring surgical intervention, while 16 were managed non-surgically. No significant differences in individual preoperative symptom frequencies were found between the surgical and non-surgical patients (Table 1). Furthermore, the baseline median number of swallowing symptoms (Surgical: median 2 [IQR 1–3, range 0–5] vs. Non-Surgical: median 2 [IQR 2–4, range 0–5]; $p = 0.172$) and respiratory symptoms (Surgical: median 2 [IQR 1–3, range 0–5] vs. Non-Surgical: median 1 [IQR 0–3, range 0–4]; $p = 0.112$) did not differ significantly between cohorts when analyzed by Mann-Whitney U-testing. Similarly, Kruskal-Wallis testing demonstrated no significant variations in the baseline number of preoperative respiratory or swallowing symptoms when

analyzed across the structural diagnostic categories (DAA, RAA ALSA, RAA MIB, or LAA ARSA). Figures 1 and 2 show preoperative symptom frequency by type of VR diagnosis.

Initial Workup

A total of 33 patients (20 female) had operations to address RAA ALSA (n=17), LAA ARSA (n=10), DAA (n=5), and RAA MIB (n=1). Pre-existing genetic conditions were identified in 6 patients (18.2%), including Trisomy 9 (n=1), Trisomy 21 (n=1), Rubenstein Taybi Syndrome (n=1), congenital disorder of glycosylation type II (n=1). Kawasaki Disease was present in 2 patients. The majority of patients underwent preoperative triple aerodigestive endoscopy, consisting of bronchoscopy, esophagoduodenoscopy (EGD), and direct laryngoscopy and bronchoscopy (DLB). Preoperative bronchoscopy was conducted in 29 patients (87.9%), with 8 patients (24.2%) observed to have severe tracheobronchomalacia (TBM), defined as dynamic airway collapse on expiration greater than 80%. Preoperative EGD was conducted in 30 patients, with 24 patients (72.7%) found having greater than 80% extrinsic compression of the esophagus by visual estimation. Preoperative DLB was conducted in 27 patients (81.8%), with 6 patients (18.2%) observed to have a type I laryngeal cleft.

Additional preoperative gastrointestinal evaluation was conducted for 23 patients and included liquid esophagram (n=19; 57.6%), both liquid- and solid-food esophagram (n=8; 24.2%), and modified barium swallow study (n=4; 12.1%). Extrinsic vascular compression of the esophagus was identified in 19 patients (57.6%) and aspiration in 2 patients (6.1%). Additional preoperative respiratory evaluation was conducted in 20 patients and included pulmonary function testing (PFT) (n=13; 39.4%), cardiopulmonary exercise testing (CPET) (n=11; 33.3%), and 6-minute walking test (n=2; 6.1%). Exercise-related abnormalities on CPET were identified in 6 patients, including inspiratory flow volume limitation (n=4) and expiratory flow volume

limitation (n=2), both suggestive of possible airway obstruction; additional abnormalities in these patients included reduced exercise capacity (n=1), and severe bronchial restriction during exercise (n=1). PFTs identified both obstructive (n=3) and restrictive (n=1) lung defects.

The non-surgical cohort in this study consisted of 16 patients, including patients with RAA ALSA (n=8), DAA (n=2), and LAA ARSA (n=6). Pre-existing genetic conditions were identified in 2 patients (12.5%), including 22q11.2 deletion syndrome (DiGeorge Syndrome) (n=1) and Trisomy 21 (n=1). Among the non-surgical cohort, 13 patients received initial respiratory or gastrointestinal evaluation at our institution (e.g. triple endoscopy, swallow studies and/or exercise testing) based on clinical team recommendations. 10 patients within the non-surgical cohort had at least one follow-up appointment with our team at a median of 9 months after intake [IQR 5–14].

Detailed information on pre-operative evaluation for the non-surgical cohort can be found in Supplemental Table 1.

Operative Details and Complications

Table 2 details surgical interventions broken down by VR diagnosis. Median age at operation was 13.1 years [IQR 11.5–16.6, range 9.2–21.9]. A thoracotomy approach (n=32) was preferred to sternotomy (n=1). Kommerell's diverticulum was resected in 15 patients. Out of 27 patients with RAA ALSA and LAA ARSA, 85.2% had aberrant subclavian artery reimplantation (n=23). The most common adjunctive procedures for residual aerodigestive compression were aortopexy (n=10) and tracheopexy (n=5). Recurrent laryngeal nerve monitoring was used in 32 out of 33 patients. Cardiopulmonary bypass was required in 1 case, an aortic uncrossing procedure via median sternotomy. There was no operative mortality.

Median hospital length of stay from the operation was 4 days [IQR 3–6]. Postoperative complications occurred in 9 out of 33 patients (27.3%)–including incomplete Horner's Syndrome (n=2) that resolved by one-year follow up and left vocal cord hypomobility (n=1) that resolved by 30 months postoperatively. Chylous chest tube drainage was observed in 4 patients and was managed with a low-fat diet alone (n=1), chest tube placement with subsequent low-fat diet (n=2), or thoracic duct ligation (n=1). Postoperative pneumothorax (n=2) and hemothorax (n=1) were also observed and managed with chest tube placement.

Change in Postoperative Symptoms

Significant postoperative improvement in symptoms was observed at participants' latest follow-up visit (median 6 months, IQR 1–12). Both the median number of reported swallowing symptoms and respiratory symptoms decreased significantly from 2 [IQR 1–3, range 0–5] preoperatively to 0 [IQR 0–1, range 0–2] postoperatively ($p < 0.001$, Wilcoxon signed-ranks test). Most patients (n=32/33) experienced reduction in total number of symptoms, with 20 patients (60.6%) having no symptoms at follow-up and 12 patients (36.4%) experiencing partial symptom resolution. Out of the 13 patients with at least one residual respiratory or swallowing symptom (swallowing symptoms n=5, respiratory symptoms n=3, both n=5), 10 patients observed a qualitative reduction in symptom severity according to provider documentation. Table 3 highlights the change in symptom prevalence between pre- and postoperative periods, along with respective p-values.

Reduction in the incidence of multiple specific symptoms was observed among the surgical cohort. Improvements in swallowing symptoms included adaptive eating behaviors (pre-op 76% vs. post-op 9%, $p<0.001$), dysphagia with solids (pre-op 49% vs. post-op 9%, $p<0.001$), and sensation of food getting stuck (pre-op 39% vs. post-op 15%, $p=0.039$). Improvement in

respiratory symptoms included dyspnea on exertion (pre-op 76% vs. post-op 15%, $p<0.001$), chest pain (pre-op 46% vs. post-op 9%, $p<0.001$), increased URIs (pre-op 36% vs. post-op 3%, $p<0.001$), and chronic cough (pre-op 30% vs. post-op 0%, $p=0.002$). A type 1 laryngeal cleft was identified in one patient with residual respiratory symptoms. Another patient with both residual respiratory and swallowing symptoms also had preoperative diagnosis of Trisomy 9 and cytochrome p450 abnormal metabolism.

Adjunctive procedures during the VR operation, including tracheopexy (n=3), aortopexy (n=5), bronchopexy (n=1), and subclavian arteriopexy (n=2) were carried out in 8 of the 13 patients with residual aerodigestive symptoms. Postoperative bronchoscopy and postoperative esophagram were carried out in selected symptomatic patients; residual mild TBM was found in 1 patient and mild esophageal compression in 2 patients.

Discussion

In this study, we investigated the symptom presentation and clinical outcomes in pre-adolescent and adolescent patients diagnosed with VR. The most common respiratory symptoms were dyspnea on exertion and chest pain, and the most common swallowing symptoms were adaptive eating behaviors and solid food dysphagia. Amongst patients who underwent surgery, the total number of reported symptoms declined postoperatively, with over 60% of our cohort experiencing complete symptom resolution and 36% experiencing partial resolution at latest follow-up. Half of the respiratory and swallowing symptoms (n=7/14) we measured showed significant reduction.

Regarding respiratory symptoms, we found considerably greater frequencies of dyspnea on exertion (67.3%) and lower rates of noisy breathing/stridor (12.2%) compared to the literature in younger children [4–7,16]. Dyspnea on exertion is less commonly documented as a symptom in VR literature. This may be due to later manifestation of this symptom, with pre-adolescent and adolescent children being more active and involved in organized sports, where exertional dyspnea could present. Conversely, noisy breathing/stridor might appear more in younger children as it is a more noticeable symptom for parents to report. Several participants in this study were first diagnosed with VRs after reporting a reduced ability to keep up with peers and self-limiting in physical activities at school or sports practice.

Another possible reason for the observed delay in presentation is confounding exercise-related symptoms of airway compression and asthma, a phenomenon observed in older children with TBM [17]. Functional respiratory tests may help clinicians distinguish between obstructive airway conditions, including those affiliated with VR. A study following children with TBM showed that PFTs can help identify airway obstruction not associated with asthma [18]. Meanwhile, exercise dysfunction on CPET shows significant concordance with CT and bronchoscopy findings for patients with TBM [19]. In our study, 9 surgical patients and 5 non-surgical patients had respiratory abnormalities documented in preoperative CPET and PFT findings. Including CPET and PFT in preoperative workup could elucidate the impact of a VR on respiratory mechanics, with multiple centers utilizing these tests to identify the severity of respiratory burden preoperatively [13,20,21]. While CT and bronchoscopy remain the diagnostic gold standard, there may be a greater role for functional respiratory tests in differentiating and assessing the severity of airway obstruction in children with a suspected or confirmed VR.

Notably, over a third (40.8%) of our patients described experiencing chest pain unrelated to eating or exercise. In our cohort, chest pain had the highest frequency in patients with RAA ALSA, with over half (52.0%) of these patients reporting the symptom. This is a novel finding for clinical diagnosis as few centers document chest pain in their vascular ring reports. A review of LAA ARSA patients identified chest pain in close to 20% of patients [15]. Meanwhile, a study in the Netherlands comparing pediatric versus adult-onset vascular ring diagnoses noted that the adult-onset cohort had significantly greater frequency of chest pain [13]. This study, like ours, cited chest pain as a respiratory symptom, though there remains no identified origin of pain. One possible mechanism is that prolonged compression of the trachea and/or esophagus by a vascular structure contributes to chest pain; however, further research is required to determine symptom etiology and rule out possible VR comorbidities such as gastroesophageal reflux. In our cohort of adolescents, surgery led to significant improvement of chest pain at follow-up, suggesting the pain may be related to vascular compression. Clinicians should assess for chest pain as part of a comprehensive evaluation, particularly among pre-adolescent and adolescent populations suspected of VR.

Swallowing symptoms also appear to manifest distinctly within this older pediatric cohort. Adaptive eating behaviors (e.g., cutting food into small pieces, taking frequent sips of water during meals, over chewing food and meal-related anxiety) were the most common symptom observed preoperatively. Patients with LAA ARSA exhibited these behaviors at a far greater frequency (93.8%) compared to both RAA ALSA (60.0%) and DAA (57.1%). This illustrates how patients with LAA ARSA, often regarded as normal aortic arch variant that does not require further intervention, may present with subtle swallowing symptoms that only become apparent with a thorough and targeted medical history. Another prevailing misconception is that

these patients are simply 'picky eaters,' with eating adaptations attributed to other etiologies such as gastroesophageal reflux or aspiration [1,14]. In our cohort, patients with LAA ARSA presented with adaptive eating behaviors more often than solid food dysphagia (68.8%), suggesting that while patients' swallowing symptoms do not necessarily reach the threshold of traditional dysphagia, there may still be a significant reduction in quality of life. This may contribute to the delay in LAA ARSA diagnosis into older childhood and adolescence. We thus advocate for a thorough assessment of symptoms and complete, tailored diagnostic evaluation of symptomatic patients with LAA ARSA. Interval monitoring for the development or progression of symptoms is also recommended for those with suspected or confirmed LAA ARSA.

Surgery resolved or reduced the frequency of the most common preoperative respiratory symptoms—dyspnea on exertion, chest pain, increasing or more frequent URIs, chronic cough—and swallowing symptoms—adaptive eating behaviors, solid food dysphagia, a sensation of food getting stuck. These findings support existing literature with similar postoperative follow-up on the efficacy of complete vascular ring repair at relieving symptom burden [22,23]. The operative complication rate was 27.3%, which is higher compared to large cohort reviews for pediatric vascular rings [2,3,7,9,10]. All reported complications ultimately resolved with postoperative management, and only one patient required repeat surgical intervention via thoracic duct ligation. Our institution routinely conducts pre and postoperative screening for vocal fold movement impairment (VFMI), which may have increased VFMI rates compared to those who do not routinely screen for VFMI.

Compared to pediatric vascular ring patients, operative mortality in adult cohorts is higher due to a greater incidence of concomitant medical conditions, more complex operations, and a greater risk of rupture of Kommerell's diverticulum [24,25]. At our institution, it is

standard to resect the Kommerell's diverticulum during VR operations based on evidence that resection can substantially reduce residual aerodigestive symptoms and prevent more complex operations in the future [16,26,27]. The risk of diverticular rupture and added surgical complications underscores the importance of early detection and treatment of VRs using targeted, age-appropriate symptom monitoring. Furthermore, the duration of aerodigestive compression may influence recovery following surgical correction. Longstanding compression, such as in older adults, could contribute to persistent airway or esophageal dysfunction and established compensatory eating behaviors that may not fully resolve after anatomic decompression. Earlier recognition and treatment of clinically significant compression may therefore offer an opportunity to limit these secondary effects. However, this study does not establish whether earlier repair reduces persistent symptoms; this question warrants further longitudinal evaluation that incorporate symptom duration, anatomy, and severity.

One limitation of this study is the length of follow-up, which occurred at a median of 6 months. Several studies with a longer postoperative follow-up periods have found higher rates of symptom persistence [5,6,22,28–31]. While long-term follow-up is vital to ascertain lasting symptom improvement, certain operative strategies have shown effectiveness at reducing the likelihood of recurrent symptoms. For example, residual airway compression and TBM, which are associated with higher postoperative respiratory symptom burden [23,29,31–33], can be assessed through intraoperative bronchoscopy and, if indicated, adjunctive procedures such as tracheopexy, aortopexy, innominate arteriopexy, and/or subclavian arteriopexy can be performed to limit the need for possible reoperation [1,7,12,33]. These adjunctive procedures can contribute to higher rates of chylothorax due to added mobilization of soft tissues near the thoracic duct [14,34]. In our study, 4 patients had postoperative chylous drainage—1 resolved with low-fat

diet, 2 resolved following chest tube placement and subsequent low-fat diet, and 1 required thoracic duct ligation. Deciding on the appropriate operation and adjunctive procedures in VR patients, therefore, requires carefully balancing the potential benefits and risks while considering which procedures may provide enduring symptom relief.

Assessing symptom prevalence via retrospective review represents another limitation, as symptoms may be incompletely documented in the medical record, potentially leading to underestimation of their true prevalence. The Vascular Ring and Airway Team at Boston Children's Hospital assesses symptoms through standardized patient-reported symptom surveys to longitudinally monitor participants' subjective respiratory and swallowing symptom burden. However, our institution's symptom surveys have not yet been validated for research in VR populations [14]. Not including patient-reported questionnaire data limits our ability to fully characterize symptom burden and severity within this study. This is particularly relevant given that, although we found no significant difference in the type or number of preoperative symptoms between surgical and non-surgical patients, symptom frequency and severity may have contributed to clinical decision-making regarding surgical intervention but was not consistently captured through retrospective chart review. More comprehensive assessment of symptom frequency and severity in future studies may provide greater insight into factors influencing surgical decision-making and guide quality improvement efforts.

Interpretation of surgical outcomes using single institution data is another limitation to this study's applicability. Inter-institutional variation in symptom monitoring can pose a significant barrier to cross-comparison and standardization of clinical and quality guidelines for congenital heart disease (CHD) including VRs. The expansion of patient-reported outcome measures (PROMs) into pediatric rare disease and adult CHD cohorts represents a valuable

opportunity to establish a global standard for capturing vascular ring symptoms and quality-of-life measures, centered in patient and caretaker perspectives [35,36]. Designing validated, age-appropriate symptom questionnaires based in PROMs would help create an evidence-backed pathway for referral, additional workup, and surgery. Validated, age-appropriate PROMs could improve the timeliness and precision of VR diagnosis from infancy to early adulthood.

Conclusion

This study was the first of its kind examining symptoms and surgical outcomes in a cohort of pre-adolescent and adolescent patients with VR diagnoses. We identified several previously underrecognized symptoms in older children, including dyspnea on exertion, chest pain, and adaptive eating behaviors. For patients who had surgery, 32 out of 33 patients reported fewer symptoms, with complete resolution seen in 20 patients. More research and follow-up are needed to identify the durability of symptom resolution. However, these findings highlight the importance of recognizing the evolving clinical presentation of VRs across the lifespan, particularly in pre-adolescent and adolescent patients. Increased awareness of these symptoms and their role in clinical VR assessment can help providers give more comprehensive care in pediatric populations across all ages.

Declarations

Author Contributions

All authors contributed to the study conception and design. Material preparation and data collection were performed by Vibhav Nandagiri, Lianne Cole, Carmel Falek, Jennifer Brown, and Linda Karnett. Statistical analysis and visualization were performed by David Zurakowski and Vibhav Nandagiri. The first draft of the manuscript was written by Vibhav Nandagiri and all authors commented on previous versions of the manuscript. All authors read and approved the final manuscript. Benjamin Zendejas and Christopher Baird are co-senior authors for this manuscript.

Ethics approval

This research study was conducted from data obtained for clinical purposes. We consulted with the IRB of Boston Children's Hospital who determined that our study did not need ethical approval. An official waiver of ethical approval was granted from the IRB of Boston Children's Hospital (IRB-P00042409).

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Competing Interests

The authors have no relevant financial or non-financial interests to disclose.

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Table 1. Preoperative Comparison of Baseline Symptoms Between Surgical and Non-Surgical Cohorts

Preoperative Assessment	Surgical Group (n = 33)	Non-Surgical Group (n = 16)	p-value	Clinical Inference
Swallowing Symptoms				
Food gets stuck	13 (39%)	8 (50%)	0.547	No statistical difference between cohorts
Disinterest in eating	1 (3%)	2 (13%)	0.245	No statistical difference between cohorts
Dysphagia with solids[1]	16 (49%)	9 (56%)	0.762	No statistical difference between cohorts
Dysphagia with liquids[1]	3 (9%)	4 (25%)	0.195	No statistical difference between cohorts
Poor weight gain	3 (9%)	3 (19%)	0.377	No statistical difference between cohorts
Pain while swallowing	1 (3%)	2 (13%)	0.245	No statistical difference between cohorts

Adaptive behaviors[2]	25 (76%)	10 (63%)	0.501	No statistical difference between cohorts
Respiratory Symptoms				
Chronic cough	10 (30%)	2 (13%)	0.290	No statistical difference between cohorts
Blue spells	1 (3%)	0 (0%)	1.000	No statistical difference between cohorts
Dyspnea on exertion	25 (76%)	8 (50%)	0.106	No statistical difference between cohorts
Increased URIs	12 (36%)	6 (38%)	1.000	No statistical difference between cohorts
Barking cough	2 (6%)	0 (0%)	1.000	No statistical difference between cohorts
Noisy breathing/stridor	4 (12%)	2 (13%)	1.000	No statistical difference between cohorts
Chest pain	15 (46%)	5 (31%)	0.375	No statistical difference between cohorts

Note: Data are presented as n (%). All p-values were determined by Fisher's exact test.

[1] Dysphagia with solids and liquids correspond to the "Cough/gag" descriptors utilized during

database collection.

[2] Includes clinical behaviors such as cutting food into small pieces, taking frequent sips of water during meals, over-chewing food, and prolonged mealtimes.

Figure 1. Preoperative swallowing symptoms by vascular ring diagnosis

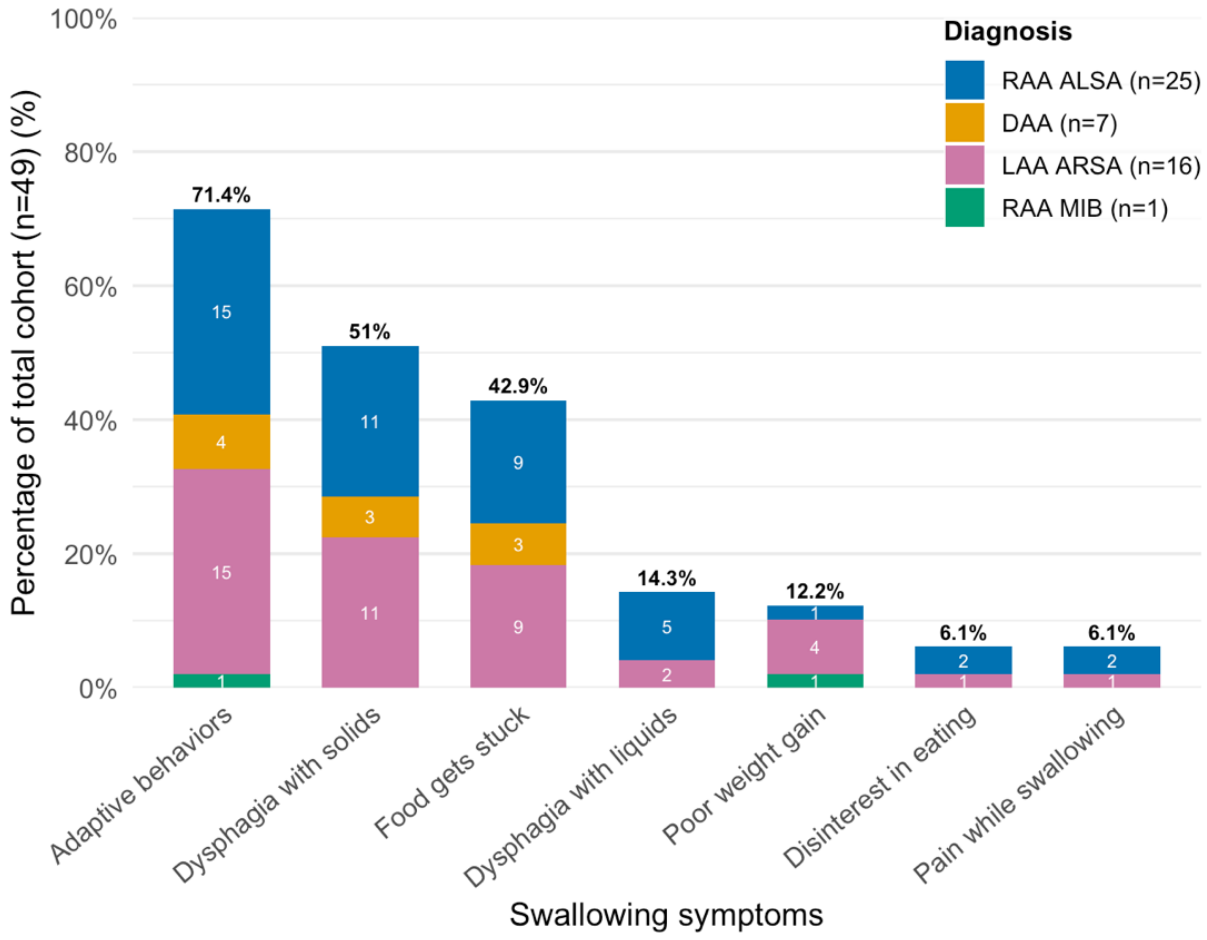


Figure 2. Preoperative respiratory symptoms by vascular ring diagnosis

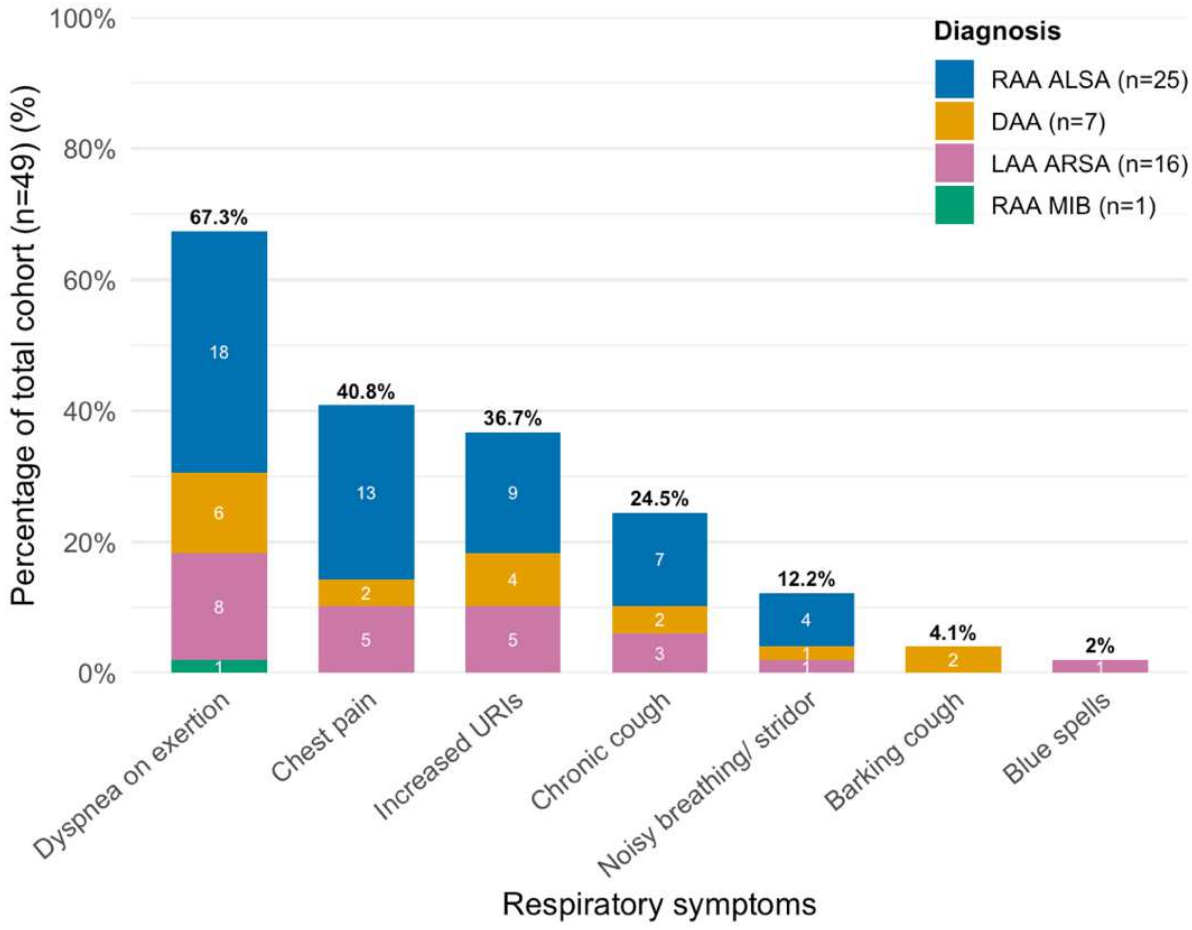


Figure Descriptive Captions

Figure 1. Preoperative Swallowing Symptoms by Vascular Ring Diagnosis.

Bar graph illustrating the baseline prevalence of specific swallowing and eating-related symptoms among the total pediatric cohort (N = 49), categorized by anatomical vascular ring variant: Right Aortic Arch with Aberrant Left Subclavian Artery (RAA ALSA, n = 25), Double Aortic Arch (DAA, n = 7), Left Aortic Arch with Aberrant Right Subclavian Artery (LAA ARSA, n = 16), and Right Aortic Arch with Mirror-Image Branching and Left Ligamentum (RAA MIB, n = 1). Numerical values inside the bars represent the absolute number of affected patients. Adaptive eating behaviors (71.4%) and dysphagia with solids (51.0%) represent the most prominent clinical symptoms at presentation.

Figure 2. Preoperative Respiratory Symptoms by Vascular Ring Diagnosis.

Bar graph illustrating the baseline distribution of individual respiratory symptoms across the total study cohort (N = 49), stratified by structural vascular ring diagnoses: RAA ALSA (n = 25), DAA (n = 7), LAA ARSA (n = 16), and RAA MIB (n = 1). Absolute patient counts are explicitly annotated within each corresponding bar segment. Dyspnea on exertion (67.3%) and chest pain (40.8%) represent the primary clinical symptoms at presentation.

Table 2. Surgical Interventions Broken Down by Vascular Ring Diagnosis

	Total (n=33)	RAA ALSA (n=17)	DAA (n=5)	LAA ARSA (n=10)	RAA MIB (n=1)
Approach					
Thoracotomy	32 (97.0%)	16 (94.1%)	5 (100.0%)	10 (100.0%)	1 (100.0%)
Vascular					
Arch division	5 (15.2%)	0 (0%)	5 (100.0%)	0 (0%)	0 (0%)
Ligamentum arteriosum division	22 (66.7%)	16 (94.1%)	5 (100.0%)	0 (0%)	1 (100.0%)
Kommerell's Diverticulum resection	15 (45.5%)	12 (70.6%)	2 (40.0%)	0 (0%)	1 (100.0%)
Reimplantation of aberrant subclavian artery	23 (69.7%)	13 (76.5%)	0 (0%)	10 (100.0%)	0 (0%)
Aortic uncrossing	1 (3.0%)	1 (5.9%)	0 (0%)	0 (0%)	0 (0%)
Airway					
Aortopexy	10 (30.0%)	7 (41.2%)	2 (40.0%)	1 (10.0%)	0 (0%)
Tracheopexy	5 (15.2%)	4 (23.5%)	0 (0%)	1 (10.0%)	0 (0%)

Bronchopexy	1 (3.0%)	1 (5.9%)	0 (0%)	0 (0%)	0 (0%)
Subclavian arteriopexy	3 (9.1%)	2 (11.8%)	1 (20.0%)	0 (0%)	0 (0%)
Esophageal					
Esophageal mobilization/lysis of adhesions	32 (97.0%)	17 (100.0%)	5 (100.0%)	9 (90.0%)	1 (100.0%)

Table 3. Postoperative Changes in Swallowing and Respiratory Symptoms Among Surgical Cohort (n = 33)

Symptom Assessment	Preoperative	Postoperative	p-value	Clinical Inference
Swallowing Symptoms				
Food gets stuck	13 (39%)	5 (15%)	0.039*	Significant clinical reduction
Disinterest in eating	1 (3%)	0 (0%)	1.000	No statistical difference
Dysphagia with solids	16 (49%)	3 (9%)	<0.001*	Significant clinical reduction
Dysphagia with liquids	3 (9%)	0 (0%)	0.250	No statistical difference
Poor weight gain	3 (9%)	0 (0%)	0.250	No statistical difference
Pain while swallowing	1 (3%)	0 (0%)	1.000	No statistical difference
Adaptive behaviors	25 (76%)	3 (9%)	<0.001*	Significant clinical reduction
Respiratory Symptoms				
Chronic cough	10 (30%)	0 (0%)	0.002*	Significant clinical reduction
Blue spells	1 (3%)	0 (0%)	1.000	No statistical difference
Dyspnea on exertion	25 (76%)	5 (15%)	<0.001*	Significant clinical reduction
Increased URIs	12 (36%)	1 (3%)	<0.001*	Significant clinical reduction

Barking cough	2 (6%)	1 (3%)	1.000	No statistical difference
Noisy breathing/stridor	4 (12%)	0 (0%)	0.125	No statistical difference
Chest pain	15 (46%)	3 (9%)	<0.001*	Significant clinical reduction

* Statistically significant. All p-values determined by the McNemar test for binary matched pairs.

Supplementary Files

This is a list of supplementary files associated with this preprint. Click to download.

- SupplementaryTableAdolescentVascularRing11426.docx