Persistence of Aerodigestive Symptoms after Vascular Ring Repair

Doris Farje BA[1], Ashley Young BS[1], Eli Stein MD[1], Osama M Eltayeb MD[2,3], Saied Ghadersohi MD[4,5], Inbal Hazkani MD[4,5]

[1]Feinberg School of Medicine, Northwestern University, Chicago, Illinois, USA. [2]Division of Cardiovascular-Thoracic Surgery, Ann & Robert H. Lurie Children's Hospital of Chicago, Box 22, 225 E. Chicago Ave, Chicago, Illinois 60611. [3]Department of Surgery, Northwestern University Feinberg School of Medicine, Chicago, Illinois. [4]Division of Pediatric Otolaryngology-Head and Neck Surgery, Ann & Robert H. Lurie Children's Hospital of Chicago, Chicago, Illinois, USA. [5]Department of Otolaryngology-Head and Neck Surgery, Northwestern University Feinberg School of Medicine, Chicago, Illinois, USA.

ABSTRACT

Objective: Vascular rings are often associated with respiratory and swallowing difficulties due to tracheal or esophageal compression. While the results of a vascular ring repair are considered excellent, the long-term effect of tracheal and esophageal remodeling and the persistence of symptoms have scarcely been reported. Our study aims to evaluate the respiratory and swallowing outcomes of vascular ring repair.

Study design and methods: A retrospective cohort study of children who underwent vascular ring repair between 2010-2022 in a tertiary-care children's hospital

Results: There were 108 patients enrolled: sixty-three patients (57.41%) with a right aortic arch, 42 patients (38.89%) with a double aortic arch, and 3 patients (2.78%) with other vascular rings. Forty-three (39.81%) patients were diagnosed prenatally. Of the 65 patients (60.19%) diagnosed postnatally, 35 (53.85%) had either respiratory or swallowing symptoms as the indication for diagnostic workup. Persistent respiratory and swallowing symptoms were noted in 34 (31.48%) and 30 (27.78%) patients, respectively, within a year of surgical repair. Fourteen patients underwent repeated laryngoscopy and bronchoscopy that demonstrated residual tracheomalacia; however, only 2 (1.9%) patients required tracheostomy tube placement, and 6-out-of-7 patients were weaned off positive pressure airway support. Persistent respiratory symptoms were significantly more common in patients with a double aortic arch compared to a right aortic arch. No differences were noted in demographics, comorbidities, and preoperative aerodigestive symptoms between patients with residual symptoms and patients with no residual symptoms.

Conclusions: Persistent respiratory and swallowing symptoms after vascular ring repair are not uncommon. Postoperative evaluation should be pursued by a dedicated team, and treatment considered as appropriate.

CONTACT

Doris Farje, BA or Inbal Hazkani, MD

Division of Pediatric Otolaryngology-Head and Neck Surgery,

Ann & Robert H. Lurie Children's Hospital of Chicago

225 E. Chicago Ave Box 25, Chicago, IL 60611

Email: Doris.farje@northwestern.edu or hazkani@gmail.com

INTRODUCTION

A vascular ring is a rare anomaly, comprising 1-2% of all congenital cardiovascular defects. This anomaly of the aortic arch results in aberrant vessels which compress the trachea and/or esophagus, commonly causing aerodigestive symptoms.
The most common types of vascular rings are the **double aortic arch** (DAA), comprised of two aortic arches that encircle the trachea and esophagus, and the **right aortic arch** (RAA) with an aberrant retroesophageal left subclavian artery and left ligamentum arteriosum.

Children typically present with noisy breathing, barky cough, increased work of breathing, wheezing, and recurrent respiratory infections. Swallowing difficulties are less common and may include feeding difficulties, dysphagia, reflux, and vomiting (5).
Computed tomography angiography (CTA) and Magnetic Resonance Angiography (MRA) are the gold standard for the evaluation and diagnosis of vascular rings (6-8). Upper airway evaluation prior to or at the time of vascular ring repair to evaluate the severity of compression is often done with micro-laryngoscopy and bronchoscopy (MLB) (Figure 1).

The surgical results of vascular ring repair are considered excellent, with significant relief of both respiratory and swallowing symptoms in the majority of children (9-12). However, the long-term effect of tracheal and esophageal remodeling and the persistence of symptoms following a vascular ring repair have scarcely been reported. We sought to evaluate the long-term effect of tracheal and esophageal remodeling and the persistence of symptoms following a vascular ring repair.

METHODS AND MATERIALS

Data were collected from the electronic health records of all patients who underwent a vascular ring repair from 2010 to 2022. Data extracted included demographic data, pre-and postoperative respiratory and swallowing symptoms, pre- and postnatal imaging, and postoperative diagnostic tests up to one year after surgery.
Baseline characteristics were summarized as means and standard deviations (SD) or proportions. Group differences by type of vascular ring for continuous variables were assessed using a t-test. Categorical variables were tested with Chi-square or Fisher's exact tests where appropriate. The relationship between persistent symptoms and variables of interest was modeled with univariant analysis.

RESULTS

There were 108 patients enrolled: sixty-three patients (57.41%) with a right aortic arch, 42 patients (38.89%) with a double aortic arch, and 3 patients (2.78%) with other vascular rings. Forty-three (39.81%) patients were diagnosed prenatally. Of the 65 patients (60.19%) diagnosed postnatally, 35 (53.85%) had either respiratory or swallowing symptoms as the indication for diagnostic workup.

Persistent respiratory and swallowing symptoms were noted in 34 (31.48%) and 30 (27.78%) patients, respectively, within a year of surgical repair. Fourteen patients underwent repeated laryngoscopy and bronchoscopy that demonstrated residual tracheomalacia; however, only 2 (1.9%) patients required tracheostomy tube placement, and 6-out-of-7 patients were weaned off positive pressure airway support.
Six out of eight patients who required enteral feeds prior to surgery remained on enteral feeds one year following repair and 15 patients (13.89%) required texture modification. No differences were noted in demographics, comorbidities, and preoperative aerodigestive symptoms between patients with residual symptoms and patients with no residual symptoms.

Persistent respiratory symptoms were significantly more common in patients with a double aortic arch compared to a right aortic arch (47.62% vs 20.63%, p=0.01).

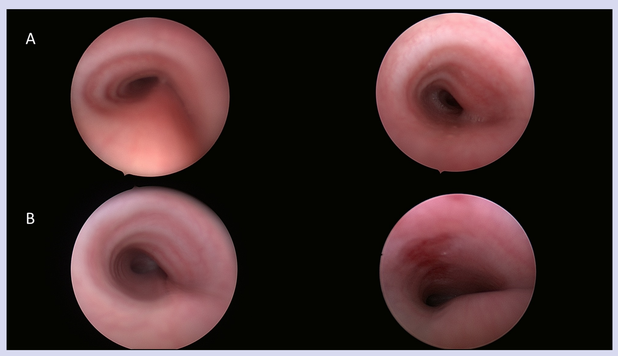
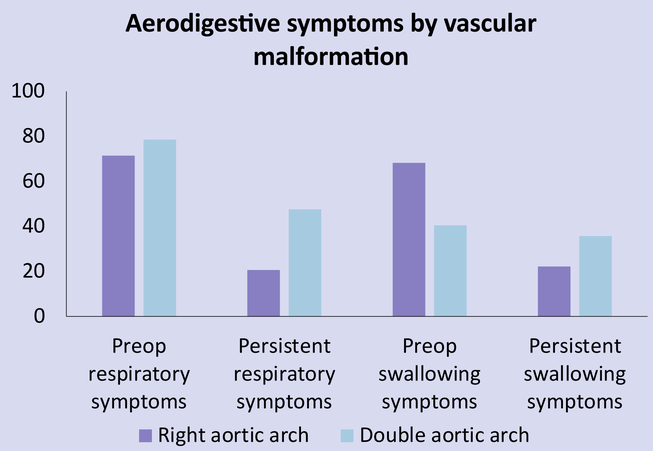


Figure 1 – External tracheal compression associated with vascular ring.
A – Double aortic Arch, B – Right aortic arch



DISCUSSION

Our study demonstrated that despite successful management of the vascular anomaly, residual aerodigestive symptoms were prevalent in 40% of patients. The prevalence of preoperative and postoperative symptoms varied depending on the type of arch. In our study, 48% of patients with DAA and 21% of patients with RAA had persistent respiratory symptoms at 1 year. Specifically, 43% of patients with DAA and 14% of patients with RAA complained of persistent stridor/noisy breathing. These findings were consistent with Callahan et al. that found that 35% of patients with single aortic arch and 57% of patients with DAA had persistent respiratory symptoms at comparable follow-up times (4). Other studies echoed these findings, potentially indicating that those with DAA have a greater respiratory symptom burden (1,5,13,14).

More than half of the patients reported difficulty swallowing prior to surgical repair, and 28% reported persistent dysphagia at 1-year follow-up. These findings correlate with the wide range of persistent dysphagia in the literature (3,4,12). Although many cofactors may attribute to persistent dysphagia, particularly in a diverse patient population with a significant burden of comorbidities, this finding may suggest that vascular ring division may only be partially effective in relieving swallowing difficulties.

This study is subject to the usual retrospective limitations and single-institutional bias. Findings in asymptomatic patients or patients with low clinical burden may have been overlooked due to the lack of a unified protocol for objective evaluation of patients for relevant symptoms. There was a paucity of objective postoperative testing or imaging to evaluate for persistent symptoms and delineate a persistent anatomic cause for those who were symptomatic.

CONCLUSIONS

Persistent respiratory and swallowing symptoms after vascular ring repair are not uncommon, particularly in patients with DAA. Residual tracheal compression was diagnosed post-repair in the majority of patient with residual respiratory symptoms who underwent post-repair MLB, suggesting that some degree of tracheal cartilage remodeling remains despite the successful division of the vascular ring. Careful postoperative evaluation should be pursued by a dedicated team, and treatment considered as appropriate.

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