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Surgery or Surveillance? Outcomes of Symptomatic and Asymptomatic Patients With Vascular Rings

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ABSTRACT

BACKGROUND AND OBJECTIVES: Although patients with symptomatic vascular rings are typically referred for surgical relief, the management of patients who are asymptomatic remains controversial. We assessed outcomes of symptomatic and asymptomatic vascular rings managed over 14 years at our center, focusing on surgical and conservative approaches.

METHODS: This is a retrospective medical record review of 228 children diagnosed with vascular ring anomaly between January 2011 and December 2023. Data collected included anatomical subtypes, symptoms, diagnostics, management, and long-term outcomes. The primary outcome was symptom-free survival, with subgroup analyses for isolated vascular rings and comparisons by anatomical type.

RESULTS: Of the 228 patients, 177 had isolated vascular rings, most commonly right aortic arch (RAA) with aberrant left subclavian artery and double aortic arch (DAA). A total of 60% (n = 106) were symptomatic, and 83% of patients who were symptomatic underwent surgery, with symptom resolution in 77%. A total of 17% of patients who were symptomatic were managed conservatively, with 61% experiencing symptom improvement without surgery. Surgical outcomes were favorable, with low reintervention rates and no perioperative mortality, although persistent symptoms were observed in 20% of patients with RAA and 28% of patients with DAA. Age at surgery did not significantly affect symptom resolution in patients with RAA. A total of 40% of patients (n = 71) never developed symptoms.

CONCLUSIONS: Although surgical repair is effective, symptoms may persist in some cases. Patients who are asymptomatic can often be safely observed, with surgery reserved for those who develop symptoms. Multidisciplinary aerodigestive evaluations may help in guiding conservative management for select patients who are symptomatic.

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WHAT'S KNOWN ON THIS SUBJECT: The treatment of patients with symptomatic vascular rings is often surgical repair with overall good symptom relief. However, many patients diagnosed with a vascular ring do not have symptoms, especially considering increased prenatal detection.

WHAT THIS STUDY ADDS: We investigated patients with vascular rings who were both symptomatic and asymptomatic to study management strategies and outcomes. Although most patients with symptoms underwent surgery, several patients improved without surgery, and a substantial number of patients never developed symptoms.

Introduction

Vascular rings are congenital abnormalities of the aortic arch and its branching vessels that can encircle and compress the trachea and esophagus. These abnormalities can present with respiratory symptoms like inspiratory stridor as well as gastrointestinal issues such as dysphagia.[1,2] However, the spectrum of vascular ring presentation is broad, varying from patients who are asymptomatic to those with severe respiratory compromise or significant feeding difficulties.[3–5]

Surgical relief of the vascular ring is the standard treatment for patients who are symptomatic and is generally associated with high rates of symptom resolution and low reoperation rates.[2,5–7] However, the approach to patients who are asymptomatic is not uniform, with some clinicians opting for surveillance or conservative management until symptoms develop, whereas others refer for surgical repair at diagnosis.[8] Data on the long-term outcomes of patients who are asymptomatic and the effectiveness of conservative management for mild symptoms are limited.[9] We describe anatomical subtypes and clinical outcomes including symptom-free survival in patients managed with vascular rings over a 14-year period at our center.

Methods

This retrospective medical record review, approved by the institutional review board, included all patients diagnosed with complete or incomplete vascular rings from January 2011 to December 2023. Complete vascular rings included double aortic arch (DAA) and right aortic arch (RAA) variants such as those with aberrant left subclavian artery or circumflex arch, whereas incomplete rings comprised left aortic arch (LAA) with aberrant right subclavian artery (ARSA) and pulmonary artery sling. Patients were identified via echocardiography, radiology, and surgical database searches. Those lost to follow-up were excluded from outcome analyses but included in descriptive statistics.

Data collected included demographics, age and method of diagnosis, symptoms, management (surgical or conservative), postoperative outcomes, and reintervention-free survival. Data were managed using the secure University of Arizona REDCap software.

Statistical Analysis

Patients were categorized by vascular ring type and subdivided based on the presence of major congenital heart disease (CHD), defined as lesions requiring surgical intervention within the first year. Outcome analyses focused on patients with isolated rings excluding those with major CHD to focus on the impact of vascular ring type alone.

Patients who were surgical and conservatively managed were assessed for residual symptoms. Factors for survival free of symptoms and survival free of reintervention were identified. Survival free of symptoms was also compared between those who had surgery during infancy (aged ≤ 1 year) to those who were operated on beyond infancy. Categorical data were compared using nonparametric tests of significance, including χ^2 analysis and Fischer's exact tests. Continuous data were compared using tests such as paired and student's *t* test. A *P* value $< .05$ was considered statistically significant. Data was analyzed using the IBM SPSS Statistics 30.0.

Results

Patient Characteristics

Over the study period of January 2011 to December 2023, a total of 228 patients were identified as having a vascular ring anomaly (Figure 1), of whom 177 (78%) had an isolated ring without any associated major CHD requiring surgical repair in the first year of life.

In the cohort of patients with RAA with aberrant left subclavian artery (ALSA), 26% (n = 44) had major CHD, with the most common being Tetralogy of Fallot/double outlet right ventricle (12%; n = 21) followed by ventricular septal defect (n = 8), dextro-transposition of the great arteries (n = 3), truncus arteriosus (n = 3), discontinuous pulmonary arteries (n = 3), and coarctation (n = 1). Among the 126 patients identified with an isolated RAA type of vascular ring, the majority (n = 110) had an RAA with an ALSA. Less commonly observed anomalies included RAA with a circumflex aortic arch (n = 7) and cervical aortic arch (n = 1).

Isolated Vascular Rings

Diagnostic Evaluation. Multimodal imaging was used for describing the type of vascular ring, aortic arch branching pattern, and presence and size of a Kommerell diverticulum, which is defined as an aneurysmal dilation of the descending aorta at the origin of the right subclavian artery. Figure 2A shows the initial modality of diagnosis in patients with isolated vascular rings. RAA with ALSA was the most identified prenatally at 44% (n = 56), followed by DAA. There was a trend toward increased prenatal diagnosis of vascular rings over the study period (Figure 2C). The median age of postnatal diagnosis was approximately 163 days for patients with RAA, and about one-third (34%; n = 22) were diagnosed in the first month of life. Transthoracic echocardiogram was the most common diagnostic modality for postnatal diagnosis followed by computed tomography angiography.

Following diagnosis, most patients underwent additional imaging to determine the need for intervention. Computed tomography angiography was the most applied advanced imaging modality (Figure 2A). Among the RAA group, 35 patients (28%) had an airway evaluation via a direct laryngoscopy and bronchoscopy, and 8 patients (6%) were evaluated by our aerodigestive and

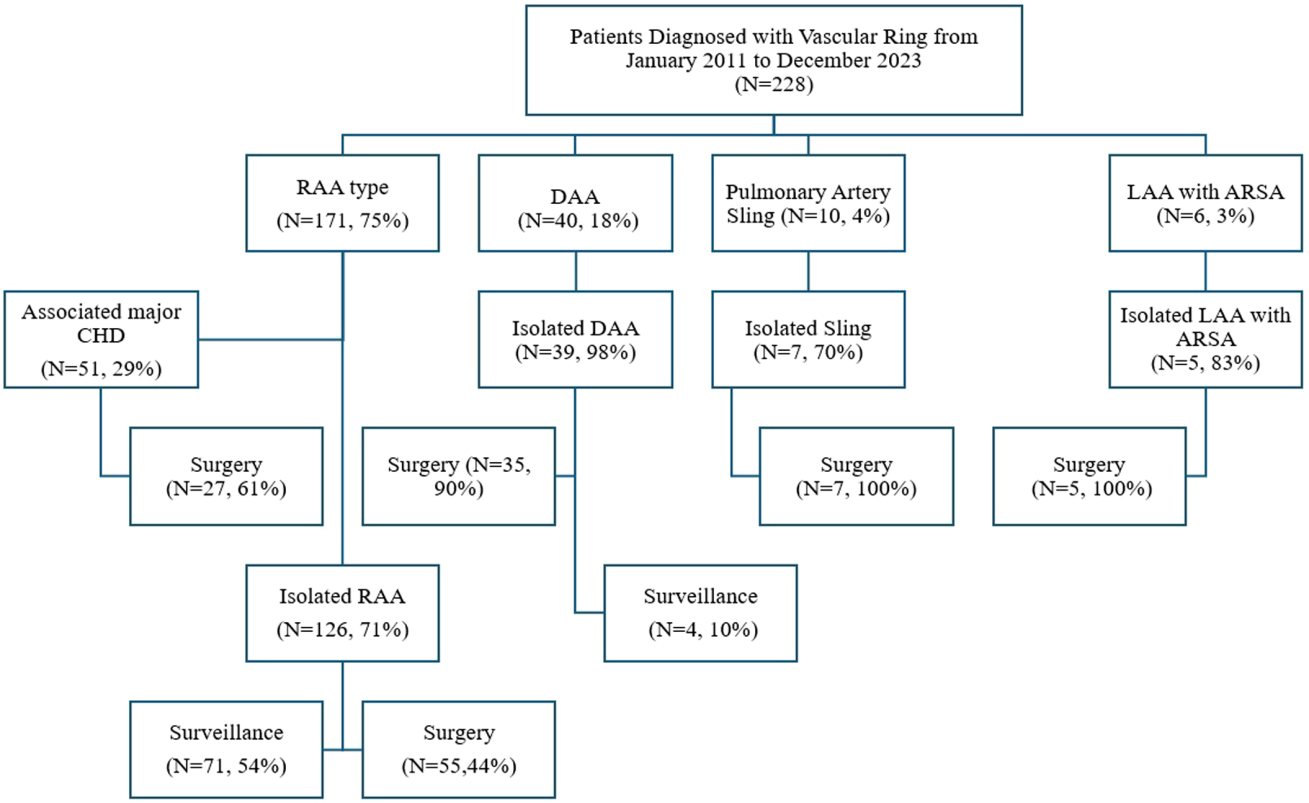

FIGURE 1. Study design and major management approaches in patients diagnosed with a vascular ring.
Abbreviations: ARSA, aberrant right subclavian artery; CHD, congenital heart disease; DAA, double aortic arch; LAA, left aortic arch; RAA, right aortic arch.

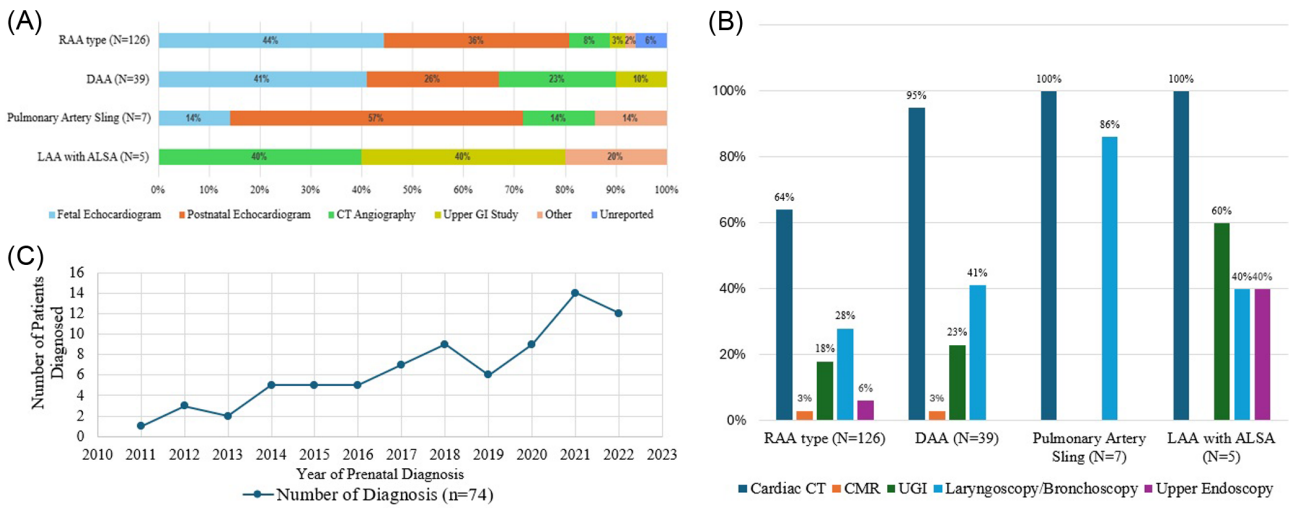

FIGURE 2. (A) Initial modality of diagnosis in patients with isolated vascular rings. (B) Additional imaging modalities for evaluation of vascular rings after initial diagnosis. (C) Prenatal diagnosis of vascular ring by year.
Abbreviations: ALSA, aberrant left subclavian artery; CMR, cardiac magnetic resonance; CT, computed tomography; DAA, double aortic arch; GI, gastrointestinal; LAA, left aortic arch; RAA, right aortic arch; UGI, upper gastrointestinal.

underwent a triple endoscopy, which included a direct laryngoscopy, bronchoscopy, and esophagogastroduodenoscopy.

Clinical Characteristics of Isolated Vascular Rings. Table 1 compares the clinical and demographic features of patients with isolated RAA (n = 126) and DAA (n = 39) types of vascular rings. Hispanic and Native American patients represented about 40% of all patients in each group. An association with genetic anomaly was significantly more common in the RAA group compared with the DAA group (30% vs 11%; $P = .01$), with the most common anomalies being 22q11.2 deletion and trisomy 21.

Symptomatic Vascular Rings. Table 1 shows the symptoms that were present for the major subtypes. Gastrointestinal symptoms were common with a higher rate in the DAA group. Stridor was the most common airway compression symptom and was present in about one-third of the DAA group.

Figure 3A shows the overall distribution of patients based on their symptoms. About 60% of patients became symptomatic. Patients with DAA were significantly more likely to be symptomatic compared with patients with RAA with ALSA (79% vs 50%; $P = .001$). Only 49 patients (28%) were symptomatic at the time of diagnosis. Most patients (72%) were either diagnosed prenatally or found incidentally. Of these patients, 57 (45%) developed symptoms after diagnosis.

Most patients who were symptomatic (83%) were operated on for relief of the vascular ring; however, relief from symptoms was not complete, and about 25% still had persistent airway or gastrointestinal residual symptoms at the last follow-up. Figure 3B compares the rate of persistent symptoms at follow-up by vascular ring type. About 20% of patients with RAA and 28% with DAA who underwent surgery for their symptoms continued to have persistent symptoms, and there was no significant difference between the 2 vascular ring types ($P = .2$).

About one-fifth (18%; n = 18) of patients who were symptomatic were managed conservatively without surgery. During follow-up, most of them were able to achieve freedom from symptoms (61%; n = 11), whereas 7 continued to have persistent symptoms. Patients with DAA were more likely to experience persistent symptoms without surgery compared with patients with RAA with ALSA (66% vs 33%; $P = .1$). However, this difference was not statistically significant, likely because of the small sample size (Figure 3B). At their last follow-up appointment, symptoms had resolved in all but 5 patients with RAA with ALSA, with 2 patients reporting residual respiratory symptoms and 3 reporting residual gastrointestinal symptoms.

Multidisciplinary Conservative Management of Patients with Vascular Rings. Among the 18 patients who were symptomatic who were managed conservatively, 5 (28%) were able to receive a comprehensive multidisciplinary aerodigestive evaluation involving specialists in otolaryngology, gastroenterology, and pulmonology. These patients underwent a triple endoscopy and were subsequently followed by the aerodigestive team. At a median follow-up of 3.5 years (IQR, 1.9–5.6 years), 4 of the 5 (80%) were asymptomatic. These 4 patients included 3 who had dysphagia and improved with gastrointestinal interventions including feeding therapy (n = 2) and a medical management of chronic gastritis and esophagitis found on his esophagogastroduodenoscopy (n = 1). One patient with stridor was found to have laryngomalacia and underwent supraglottoplasty.

TABLE 1. Clinical Characteristics of Patients Diagnosed With Isolated RAA and DAA Types of Vascular Ring

Characteristics	RAA (n = 126)	DAA (n = 39)	*P* Value
Male, n (%)	66 (52)	24 (65)	NS
Race, n (%)[a]			
Caucasian	55 (44)	22 (51)	NS
African American	14 (11)	3 (8)	
Hispanic	44 (35)	12 (35)	
Asian/Native American	7 (5)	2 (5)	
Distance from the tertiary center, n (%)			
Living within the main city	113 (89%)	36 (92%)	NS
Living remotely	10 (8%)	3 (8%)	
Insurance status, n (%)			
Private insurance	44 (35)	10 (26)	NS
Public insurance	74 (57)	23 (59)	
Unreported	8 (6)	5 (13)	
Prenatal diagnosis, n (%)	56 (44)	17 (44)	NS
Birth weight, mean (±SD), g	3080 (±794)	3146 (±667)	NS
Known genetic syndrome, n (%)	36 (29)	5 (13)	.01
22q11.2 deletion	18 (14)	4 (10)	NS
Trisomy 21	5 (4)	0 (0)	NS
Any GI or airway symptom, n (%)	62 (49)	30 (76)	.001
GI symptoms, n (%)	51 (40)	22 (56)	.04
Dysphagia	44 (35)	16 (41)	NS
Reflux	10 (8)	2 (5)	NS
Failure to thrive	3 (2)	1 (2)	NS
Food avoidance	1 (0.8)	2 (5)	NS
Respiratory symptoms, n (%)	40 (32)	14 (36)	NS
Stridor	20 (16)	13 (33)	.01
Dyspnea	8 (6)	1 (2)	NS
Cough	9 (7)	5 (13)	NS
Recurrent URTI	7 (5)	2 (5)	NS

Abbreviations: DAA, double aortic arch; GI, gastrointestinal; NS, not significant; RAA, right aortic arch; URTI, upper respiratory tract infection.
[a] Race identified at birth by family.

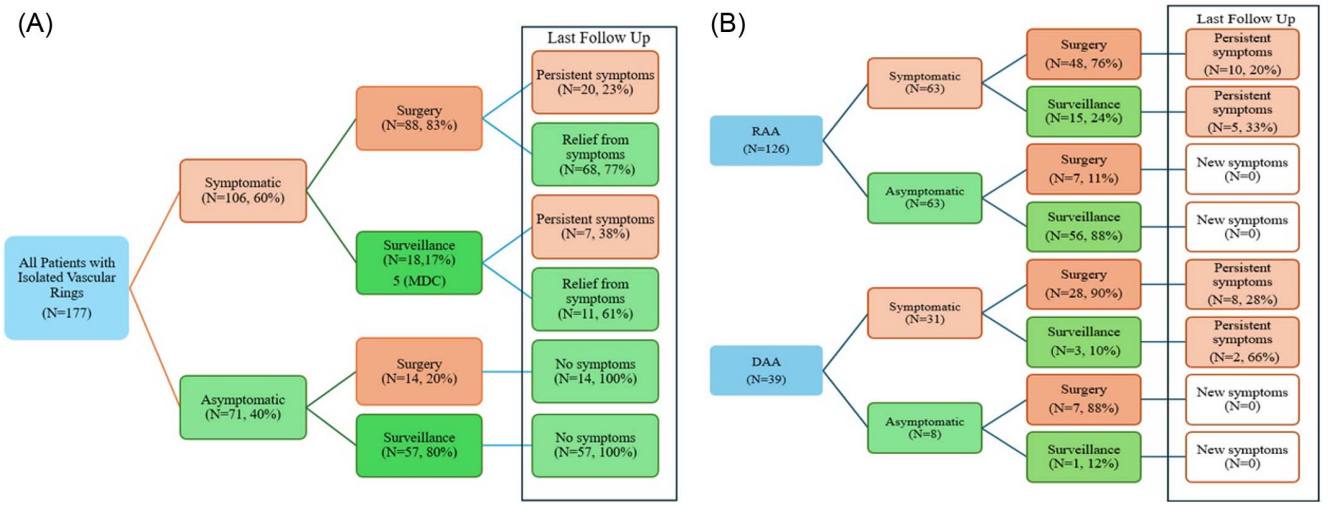

FIGURE 3. (A) Freedom from symptoms in all patients with vascular ring based on management approach. (B) Freedom from symptoms in patients with vascular ring based on anatomic type.
Abbreviations: DAA, double aortic arch; MDC, multidisciplinary clinic; RAA, right aortic arch.

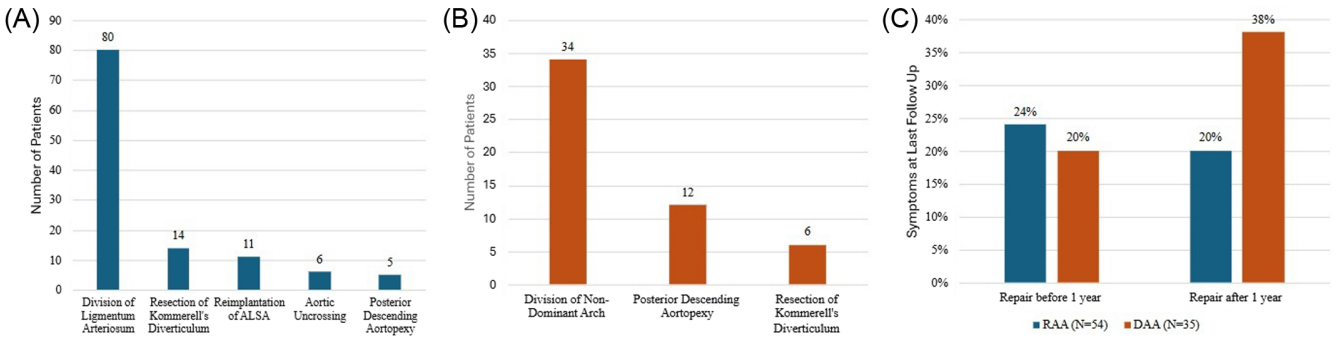

FIGURE 4. (A) Surgical techniques applied for repair of RAA vascular ring. (B) Surgical techniques applied for repair of DAA vascular ring. (C) The presence of persistent symptoms by age of repair.
Abbreviations: ALSA, aberrant left subclavian artery; DAA, double aortic arch; RAA, right aortic arch.

Notably, 3 other patients were evaluated by the aerodigestive team who ultimately underwent vascular ring surgery. These patients continued to follow-up with their aerodigestive team after surgery, and at a median follow-up of 5 years, only 1 of 3 patients remained symptomatic.

Asymptomatic Vascular Rings. A total of 71 of the 177 patients (40%) diagnosed with an isolated vascular ring did not develop symptoms (Figures 3A and 3B). Most of the patients who were asymptomatic were RAA with ALSA (n = 63), followed by DAA (n = 8).

Fourteen patients underwent surgery for their vascular ring, all of whom remained asymptomatic at their last follow-up. The remaining 57 patients (74%) did not report any new airway or gastrointestinal symptoms at the last follow-up, with a median follow-up duration of 2.6 years (IQR, 1.2–5.1 years).

Surgical Outcomes of Individual Vascular Ring Types

Of the 228 patients identified, 148 patients (65%) underwent surgical repair of the vascular ring.

RAA Type of Vascular Rings

Surgical Techniques. A total of 82 patients with the RAA type of vascular ring underwent surgical repair. This included 55 patients with isolated RAA and 27 with associated major CHD (Figure 1). The median age of repair was about 2 years (IQR, 8 months to 6.2 years).

The surgical approach was thoracotomy in 35 patients and sternotomy in 45 patients (Figure 3C). Figure 4A outlines the surgical techniques used. The median postoperative hospital length of stay was 4 days (IQR, 3–5 days).

Surgical Outcomes. There were no perioperative mortalities. Eight patients (10%) developed vocal cord dysfunction

postoperatively. Of these, 5 had undergone division of the Kommerell diverticulum, and 2 had undergone an aortic uncrossing procedure. Of those 8 patients, 1 required lateralization of the vocal cord because of paralysis. Most patients tolerated the lesion and were asymptomatic at the last follow-up, except for 1 patient who continued to have dysphagia and remained gastrostomy tube dependent. Four patients developed chylous output, one of whom was referred for thoracic duct ligation. Nonchylous pericardial effusions developed in 3 patients. One patient had an injury to the phrenic nerve, causing a hemidiaphragm that required a diaphragm plication. Reoperation rates were very low. One patient had their initial vascular ring surgery 20 years prior, who underwent repeat surgery with resection of the Kommerell diverticulum and reimplantation of the ALSA because of reoccurring symptoms. One patient had extensive lysis of adhesions formed by scar tissue, which caused external compression. Every patient followed up at 2 weeks with the surgical team, and 43 patients followed up for at least a year.

Of the 55 patients who underwent intervention, most patients (82%; n = 45) had no symptoms with a median follow-up duration of 3.5 years (IQR, 13.7 months to 7.2 years). Of the 10 patients who still had symptoms, 6 had dysphagia, and 2 were gastrostomy tube dependent. Two patients had persistent coughs who were eventually diagnosed with asthma. Figure 3B shows the persistent symptoms in the RAA group following surgery at their last follow-up.

Of the 55 patients with an isolated RAA with ALSA, 21 patients (38%) were repaired before being aged 1 year, and 33 patients were repaired after being aged 1 year. In patients who were symptomatic prior to surgical repair, there was no significant difference in the relief of symptoms by their last follow-up appointment (Figure 4B).

DAA

Surgical Technique. A total of 35 patients underwent surgical management (Figure 3B). The median age at repair was 8.8 months (IQR, 6.4 months to 2 years). The surgical approach was thoracotomy in 31 patients, sternotomy in 2 patients, and video-assisted thoracotomy in 1 patient (Figure 4C). Figure 4B shows the surgical approaches of repair. The median postoperative hospital length of stay was 3 days (IQR, 3–4 days).

Surgical Outcomes. There were no perioperative mortalities. Postoperative complications were noted in 7 patients (17.5%). Two patients had vocal cord dysfunction, 2 patients had chylothorax, 1 patient had a pleural effusion, 1 patient had a paretic left hemidiaphragm, and 1 patient had a superficial dehiscence. Four patients (11%) required reintervention because of recurrent symptoms. Three of these patients underwent resection of the Kommerell diverticulum with subclavian artery reimplantation, whereas 1 patient had an aortic uncrossing procedure. Every patient followed up at 2 weeks with the surgical team, and 24 patients followed up for at least a year.

Of the 35 patients who underwent intervention, most patients (77%; n = 27) had no symptoms with a median follow-up duration of 2.2 years (IQR, 6.5 months to 5.1 years). Of the 8 patients who still had symptoms, 3 patients had dysphagia, 3 patients had stridor, and 2 patients had persistent coughing and were ultimately diagnosed with asthma.

For the 35 patients with DAA who underwent surgical repair, 20 patients (57%) were repaired before being aged 1 year, and 15 patients were repaired after being aged 1 year. In contrast to the patients with RAA with ALSA, there was an overall trend of more patients having persistent symptoms in those repaired after being aged 1 year (Figure 4B).

Outcomes of Other Vascular Ring Anomalies

The other 2 types of vascular ring anomalies identified were LAA with ARSA (n = 6) and pulmonary artery slings (n = 10). Twelve of these patients had isolated defects.

LAA With ARSA

All 5 patients with isolated defects were diagnosed postnatally because of symptoms of dysphagia (n = 5) and stridor (n = 1), with the median age of diagnosis around 1.2 years (IQR, 7.3 months to 4.5 years). Five patients underwent surgical repair with a reimplantation of the ARSA. The median hospital length of stay was 4 days (IQR, 3.5–5 days) with no postoperative complications. Only 1 patient was noted to have dysphagia a year after surgery, with no patients reporting symptoms at their last follow-up appointment.

Pulmonary Artery Sling

Of the 7 patients with isolated defects, 6 were diagnosed postnatally at a median age of 2.6 months (IQR, 2–6.2 months). All patients were symptomatic prior to their repair, with 5 patients having respiratory symptoms and 3 patients having dysphagia. All 7 patients underwent surgical repair at a median age of 7 months (IQR, 3.3 months to 1.9 years). Six of those patients had a tracheal slide procedure done at the time of left pulmonary artery reimplantation. The median hospital length of stay was 18.5 days (IQR, 10–18.5 days), with 2 patients having complications that included difficulty weaning off the ventilator and tracheitis. There was 1 patient death 3 months after surgical repair secondary to respiratory failure. Two patients were noted to have persistent symptoms at their last follow-up, with 1 patient being gastrostomy tube dependent and 1 patient having stridor.

Discussion

With advancements of sophisticated imaging modalities including fetal echocardiography and advanced imaging techniques, the incidental diagnosis of vascular rings has become more

prevalent.[10–12] The treating physicians often face the dilemma of managing these asymptomatic vascular rings. Our data shows that although surgical repair is effective and safe for patients who are symptomatic, those who are asymptomatic can be managed conservatively with favorable outcomes. These findings suggest that routine surgery may not be necessary for all patients, and individualized assessment based on anatomy and clinical presentation is critical in guiding management decisions.

Over the study period, we observed an increasing trend in the use of advanced imaging methods, particularly computed tomography angiography and fetal echocardiography. This is reflective of current trends in vascular ring management.[13] This trend was especially evident in patients with RAA with ALSA, who were increasingly diagnosed prenatally through fetal imaging.

Among those with RAA with ALSA, most patients were diagnosed prenatally or incidentally, with half of the patients remaining asymptomatic through their diagnosis to their last follow-up. This highlights that not all patients may require surgical intervention, especially in the absence of symptoms. Additionally, most patients who were symptomatic with RAA with ALSA managed conservatively without surgery and achieved a resolution of symptoms over time with supportive therapy alone. This suggests that symptoms in these patients can be multifactorial, and other factors such as underlying asthma or gastrointestinal reflux may contribute to the symptoms. It is worth noting that relief from symptoms was higher in patients who were symptomatic and referred for surgery. However, 20% of patients had residual symptoms, highlighting the multifactorial nature of the patient's symptoms and the need for individualized treatment plans.[14–16]

In contrast to RAA with ALSA, patients with a DAA were more likely to develop symptoms. Nearly all patients with DAA, whether symptomatic or asymptomatic, underwent surgical intervention. Of the 3 patients who did not undergo surgery, 2 remained symptomatic, suggesting that without surgical intervention, symptoms will likely persist. In this group, it appears that a surgical approach may be a preferred approach compared with only surveillance.

The role of multidisciplinary evaluation through an aerodigestive team emerged as a valuable component of care in conservatively managed patients who are symptomatic. Among those who underwent triple endoscopy and aerodigestive follow-up, most patients experienced the resolution of symptoms, emphasizing the potential of a coordinated, nonsurgical approach in select patients.[5]

Surgical outcomes were favorable across all vascular ring types, with low morbidity and excellent symptom improvement, which is a consistent finding in multiple other studies.[3,13,17] Interestingly, age at surgery did not significantly affect long-term symptom resolution in patients with RAA with ALSA. Patients repaired during infancy had similar outcomes to those operated after being aged 1 year, suggesting that symptom severity and anatomical considerations, rather than age, should guide surgical timing.[9] In contrast, patients with DAA were more likely to have persistent symptoms if they had surgery later in life, which may propose that earlier surgery could be protective against long-term symptoms.[16]

Outcomes for less common vascular ring types, such as LAA with ARSA and pulmonary artery sling, were also favorable. All patients in these groups underwent surgical repair, with patients with a pulmonary artery sling often requiring complex airway reconstructions. These patients had the longest hospital stays and highest morbidity, reflecting the severity of their airway involvement.[18] Conversely, patients with LAA and ARSA had shorter hospitalizations, and all achieved complete symptom resolution by their last follow-up.

Our study has some important limitations, including potential biases from incomplete or missing data and variability in clinical documentation as well as variation in patient reporting symptoms. Some patients were lost to follow-up, which may have led to an underreporting of symptoms. Additionally, although the number of patients included in the study is robust, certain subsets of patients, such as the patients who were symptomatic and managed conservatively, remained small, which limits the strength of the conclusions. Despite these limitations, our study provides insight from one of the largest series of patients with RAA with ARSA managed conservatively, highlighting the variable course of this condition. Future research should focus on identifying which patients are most likely to benefit from multidisciplinary conservative management and further evaluating the role of aerodigestive interventions in larger cohorts to strengthen evidence-based treatment approaches.

Conclusion

Surgical repair for patients with symptomatic vascular rings remains highly effective, with high rates of symptom resolution and low reoperation rates. However, many patients who were asymptomatic can be successfully managed conservatively. The timing of surgery did not significantly affect outcomes, and select patients with mild symptoms may benefit from conservative management as well as a multidisciplinary approach from an aerodigestive team. The findings highlight the importance of a personalized approach, with further research needed to optimize management strategies and evaluate conservative care approaches.

Abbreviations

ALSA: aberrant left subclavian artery
ARSA: aberrant right subclavian artery
CHD: congenital heart disease
DAA: double aortic arch
LAA: left aortic arch
RAA: right aortic arch

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