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Contemporary Outcomes of Vascular Rings in Children: A Single-Center Retrospective Study

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Abstract

Background

Vascular rings encircle the trachea and esophagus, potentially compressing them and leading to respiratory and gastrointestinal symptoms in children.

Objective

This study aimed to describe the clinical and anatomical features as well as the surgical outcomes of children with vascular rings.

Material and methods

All children referred for cardiac evaluation were screened for vascular rings using echocardiography. Between January 2016 and January 2023, 29 patients diagnosed with vascular rings at Prince Sultan Cardiac Center, Qassim, Saudi Arabia, were enrolled in the study. This included vascular ring patients under 18 years of age with normal or abnormal intracardiac anatomy and with or without syndromic features. Patients diagnosed with a left aortic arch with an aberrant right subclavian artery were excluded. Patient records have been reviewed for demographic, clinical, echocardiographic, and surgical outcomes.

Results

Twenty-eight patients were diagnosed with vascular ring. The mean age at diagnosis was 3.8 years (± 4.4), ranging from 0 to 15 years. Males were 12 (42.8%), and females were 16 (57.1%). Eleven (39.2%) had associated intracardiac lesions. The right aortic arch with aberrant left subclavian artery and left ligamentum was the most frequently found vascular ring. The double aortic arch was the second most common. Twelve patients (42.8%) were diagnosed while asymptomatic and 16 (57.1%) experiencing respiratory and gastrointestinal symptoms. Twenty-one patients (75%) had surgical management of the vascular ring. Vascular ring division was performed concurrently with intracardiac repair in 5 patients (23.8%). The mean follow-up for the patients was 2.23(± 2.49) years. Complete improvement was noted in 10 (66.6%) patients. Two mortalities occurred due to non-VR-related causes.

Conclusion

Careful echocardiographic assessment of the aortic arch for children referred for cardiac evaluation using echocardiography enables early detection of vascular rings and facilitates planned follow-up and intervention. Early surgical intervention is recommended for symptomatic patients with relatively low operative risks. Some patients may still experience postoperative respiratory or digestive issues despite improvement in symptoms after ring division.

Clinical trial not applicable

Introduction

Vascular rings (VR) are congenital aortic arch anomalies that compress the trachea or esophagus. Respiratory symptoms are common in infants, while older children frequently experience swallowing difficulties. [1] The symptoms that often occur are noisy breathing, barking cough, recurrent upper respiratory infections, wheezing, exertional dyspnea, and dysphagia. Patients with severe compression experience respiratory distress that requires intubation. [2] Some infants experience acute life-threatening events (ALTE) or apnea. Dysphagia is uncommon until the child can eat solid foods. [3] Survival of VR pediatric patients requires early diagnosis and treatment, and accurate VR evaluation is a prerequisite for surgical repair. VR assessment is traditionally done using multiple technologies, including echocardiography, computed tomography (CT), and magnetic resonance imaging (MRI). [4] Noninvasive imaging, such as CT and MR angiography, allows for comprehensive evaluation of the aortic arch and branch vessels to surrounding structures. [5] Dividing the ligamentum can be safely and effectively done in patients with a right aortic arch (RAA) through video-assisted thoracoscopy; however, open surgical techniques deal with all vascular compression locations, not only the ligamentum. [6]

This study aims to describe the clinical and anatomical features and surgical outcomes of children with VR at our center.

Material and methods

All children referred for cardiac evaluation were screened for VR using echocardiography. Between January 2016 and January 2023, 29 patients diagnosed with VRs at Prince Sultan Cardiac Center, Qassim, Saudi Arabia, were enrolled in the study. The Regional Research Ethics Committee approved the retrospective observational study (No. 21-1109, 23/5/2023), clinical trial not applicable. The VR patients under 18 years with normal or abnormal intracardiac anatomy and with or without syndromic features were included. Patients diagnosed with a left aortic arch with an aberrant right subclavian artery were excluded as this congenital arch anomaly does not become symptomatic in children or form a VR. Patient health records have been reviewed for demographic, clinical, echocardiographic, and surgical outcomes. Follow-up was conducted using electronic health records and telephone communications. Symptomatic patients were defined as those with the presence of respiratory symptoms (stridor, recurrent wheezing, prolonged cough) or gastrointestinal symptoms (dysphagia) attributable to the VR.

Statistical analysis

Data collection and data management were performed using Microsoft Excel (Microsoft Corporation, Redmond, WA, USA). Statistical analyses were conducted using the Data Analysis ToolPak (Statistical Analysis Tool) integrated within Microsoft Excel. Given the descriptive nature of the study, only summary statistics were performed. Mean/median and standard deviation were used to express continuous variables, while categorical variables were calculated using percentages.

Results

Among 29 patients with VR diagnosis, one with a left aortic arch (LAA) and an aberrant right subclavian artery was excluded. Table 1 displays the demographic characteristics of the patient cohort. The right aortic arch (RAA) with aberrant left subclavian artery (ALSCA) and left ligamentum was the most frequently found VR. The double aortic arch (DAA) was the second most common. Twelve patients (42.8%) were diagnosed while asymptomatic and 16 (57.1%) experiencing respiratory and gastrointestinal symptoms (Table 2). Of the diagnosed 28 (100%) patients, 21 (75%) had surgical management of the VR, while the remaining 7 (25%) patients were on follow-up. Table 3 describes the intensive care and surgical outcomes of the patients. VR division was performed concurrently with intracardiac repair in 5 patients (23.8%). Among these, three patients underwent ventricular septal defect (VSD) patch closure, one had tunneling of VSD in the context of double outlet right ventricle with subaortic VSD, and another underwent tetralogy of Fallot repair. Moreover, two patients were awaiting intracardiac repair and vascular division. Two patients died due to non-VR-related causes. The first mortality occurred in a patient with Ebstein anomaly who had undergone a division of the ligamentum arteriosum for RAA and ALSCA and suffered sudden cardiac arrest on the 4th postoperative day. Another patient underwent intracardiac repair for tetralogy of Fallot and division of the ligamentum arteriosum for RAA and ALSCA. This patient passed away in another hospital seven days after discharge, and the cause of death could not be ascertained. Autopsy was not done in any of the patients. Table 4 describes the clinical features and surgical approach for patients with double aortic arch. All patients with DAA became symptomatic in infancy, except for one neonate referred for cardiac evaluation because of a maternal history of Diabetes. Another neonate presented with apnea at 20 days of life, and further assessment revealed the diagnosis of a double aortic arch.

Table 1
Demographic features of the study population (N = 28)

Variable		**N**	**Mean (SD) / %**
Weight (kg)		28	18.5 (17.16)
Height (cm)		28	96.36 (38.83)
Age at diagnosis (years)		28	3.8 (4.4)
Gender	Male	12	42.9
	Female	16	57.1
Intracardiac anatomy	Normal	17	60.7
	Intracardiac lesions	11	39.3
N, number of patients; SD, standard deviation; %, percent			

Table 2

Clinical and diagnostic features of the study population (N = 28)

Variable		**N**	**%**
Symptoms	Asymptomatic	12	42.8
	Respiratory (apnea/stridor /chronic cough/recurrent chest infection)	11	39.2
	Gastrointestinal (dysphagia)	5	17.8
Type of vascular ring	RAA, ALSCA, and left ligamentum	15	53.6
	Double aortic arch	6	21.4
	LPA sling	1	3.6
	Mirror image RAA, circumflex retro esophageal arch, and left ligamentum	4	14.3
	RAA with circumflex retro esophageal arch, ALSCA, and left ligamentum	2	7.1

GIT, gastrointestinal tract; RAA, right aortic arch; ALSCA, aberrant left subclavian artery; LPA, left pulmonary artery.

Table 3
Intensive care and operative characteristics and outcome of the surgical patients (N = 21)

Variable		**N**	**Mean (SD) / %**
Type of surgery (N = 21)	Division of ligamentum arteriosum	9	42.9
	Intra-cardiac repair and division of ligamentum arteriosum	5	23.8
	Ligation and division of the small arch in double aortic arch	6	28.5
	Resection of KD and division of ligamentum arteriosum with re-implantation of ALSCA on left common carotid artery	1	4.8
Surgical approach	Median sternotomy	5	23.8
	Lateral thoracotomy	16	76.1
Cardiopulmonary bypass	On-pump	5	23.8
	Off-pump	16	76.1
Mechanical ventilation time (hours)		21	51(90)
Complication post-surgery		0	0
Duration of hospital stay (days)		21	6.5(3)
Follow-up (years)		28	2.23(2.49)
Symptom improvement after surgery	Yes	14	66.7
	No	1	4.7
	Asymptomatic	6	28.5
Mortality		2	9.5*
Degree of improvement	Complete	10	71.4
	Partial	4	28.6

**Mortality is reported as 2/21 surgical patients (9.5%). KD, Kommerall's diverticulum; ALSCA, aberrant left subclavian artery.*

Table 4
Characteristics of double aortic arch patients (N = 6)

1	Age of presentation (months)	Symptoms	Arch dominance.	Surgical approach
	3	**cough and stridor.**	**right**	**median sternotomy (concomitant repair of VSD)**
2	3	difficult feeding and vomiting	left	right thoracotomy
3	3	cough and stridor	right	left thoracotomy
4	7	stridor and cough	right	left thoracotomy
5	1	apnea	right	left thoracotomy
6	0	asymptomatic	right	left thoracotomy

VSD, ventricular septal defect

Discussion

The branchial arch system provides six pairs of primitive aortic arches that develop into the aortic arch and its branches and the pulmonary system during fetal life. Vascular rings are caused by abnormal regression of the branchial system and may result in encirclement and compression of the trachea, esophagus, or both. Symptomatic VR are often diagnosed early in life. The greater the delay between the first presentation of symptoms and the time of diagnosis and treatment, the higher the risk of tracheomalacia, esophageal dysmotility, and persistence of symptoms. [7, 8]

James Stewart et al., in 1964, divided congenital anomalies of the aortic arch into four major groups: Group I, double aortic arch (DAA); Group II, LAA; Group III, RAA; and Group IV, other and rare malformations of the aortic arch system. [9] The most common variants are DAA and RAA, ALSCA, and left ligamentum, which may represent up to 90% of all VRs. [10] The present study's most common variants were RAA and ALSCA with left ligamentum, followed by DAA.

Echocardiography was used to establish the diagnosis of the VRs for all patients. Our center routinely screens for VRs in all infants and children referred for cardiac evaluation using transthoracic echocardiography, with particular attention to infants who have apnea, chronic or persistent lower respiratory symptoms like cough or wheezing, upper respiratory signs like stridor, or older children with dysphagia. We have previously published a comprehensive echocardiographic evaluation of VRs. The identical protocol was applied for patients with suspected VRs. [11] This regular practice of conducting echocardiographic assessments of the aortic arch at our center may explain the higher incidence of asymptomatic cases in this study group. This practice may be beneficial by detecting asymptomatic cases, allowing for follow-up, early symptom detection, and intervention. The high rate of incidental findings supports a lower threshold for detailed vascular echocardiographic assessment in all pediatric

patients referred for cardiac evaluation. Moreover, patients with an echocardiographic diagnosis of VR underwent CT angiography before the surgery. Axial CT with volume acquisition was performed with sedation using a GE Revolution 16-slice volume CT scanner (GE Healthcare, Waukesha, WI, USA) with a slice thickness of 1.25 mm. Contrast enhancement was achieved using 2 ml/kg Omnipaque 350, and we utilized the Smart Prep technique for automated contrast tracking.

Recognizing that associated cardiac anomalies are not infrequent; therefore, thorough cardiac evaluation is necessary. In the present study, 17.8% of the patients had significant intracardiac lesions that were repaired simultaneously via median sternotomy, a lesser percentage than the previously reported 29%. [12]

There is an increase in the detection of VRs in the current era. This early detection can lead to an earlier age at operation and consequently to improved outcomes. [13] Repair of VR is indicated in the presence of symptoms; however, controversy exists in asymptomatic patients. [14]

Eight patients (28.5%) in this study were found to have VRs incidentally during cardiac assessment despite being asymptomatic. Their referral for cardiac evaluation was prompted by non-VR-related symptoms such as failure to thrive, multiple congenital anomalies, dysmorphic features, joint pain, and cardiomegaly on chest X-ray. The higher incidence of asymptomatic cases discovered in this study may be attributed to the regular screening for VRs in all patients undergoing cardiac evaluation at our center. The advances in diagnostic tools led to the question of when to offer surgical repair for asymptomatic patients. In a study of 11 patients with VRs with mild to no symptoms and followed up to a median period of seven years, there was no mortality, and almost all symptoms resolved by four years of age. [15] However, only follow-up of asymptomatic or mildly symptomatic VRs may not be safe because some VRs are highly likely to develop symptoms later in childhood or even in adulthood. Surgical intervention in these cases later in life is more complicated and may even require a staged approach. Prolonged mechanical compression might affect esophageal motility and result in the persistence of airway symptoms despite the relief of the compression. [16–18] Six asymptomatic patients underwent surgery; four underwent concomitant intracardiac repair, while two demonstrated significant tracheoesophageal compression on CT imaging despite absence of symptoms.

The most common surgical approach in this study was through a left thoracotomy (76.1%). Median sternotomy was used in the presence of concomitant intracardiac defects that needed repair (23.8%). This is similar to previously reported approaches.[18] In this study, division of the VR without Kommerall's diverticulum (KD) resection was the most common surgical technique (47.3%). Only one patient underwent resection of KD with re-implantation of a subclavian artery because our surgical team recently adopted this strategy for patients with the RAA or circumflex aorta with KD and aberrant subclavian artery. A sizable KD, 1.5 times the size of the aberrant subclavian artery, is an indication for intervention.[19] A dissection/rupture rate of 44% was reported with KD as small as 2 cm.[20]

Most of the patients in this study (66.6%) showed improvement in symptoms after surgery. Only one patient (6.6%) showed symptoms that did not improve and required reoperation. Backer et al. reported

that 5–10% will have a recurrence of their symptoms and may need reinterventions. [21] We reported two patient mortalities. Both patients had significant associated congenital heart disease and underwent complex management. No evidence suggested mortality directly attributable to VR division itself. However, we acknowledge that associated cardiac disease may have contributed to overall perioperative risk.

In a recent meta-analysis, the overall pooled mortality was 0% for DAA and RAA surgery. There was a high rate of persistent postoperative symptoms, with symptoms at the last follow-up prevalence of 33% and 40% for DAA and RAA surgery, respectively. The prevalence of reintervention was 3% and 4% for DAA and RAA surgery, respectively[22] The lower symptom recurrence rate in our study may be due to a shorter follow-up period in our patients.

Limitations

This study conducts a retrospective analysis of a small patient sample from a single center. The findings may reflect local referral patterns, institutional screening practices, and surgical management strategies. Therefore, caution should be exercised when generalizing the results to other populations. Given the limited number of patients in several subgroups, comparative statistical analyses would be underpowered and potentially misleading. Future multicenter studies with larger cohorts are needed to permit meaningful subgroup analyses. The midterm outcomes were assessed using a combination of methods, including patient surveys. Moreover, the mean follow-up duration of 2.23 years may not fully capture late recurrence of respiratory or swallowing symptoms. Longer follow-up studies are required to better assess long-term outcomes. There is a possibility of underreporting mild, self-limiting postoperative symptoms during follow-up visits, which may result in an overly optimistic view of immediate outcomes.

Conclusion

Careful echocardiographic assessment of the aortic arch for children referred for cardiac evaluation using echocardiography enables early detection of VRs, even in the absence of symptoms, and can facilitate planned follow-up for symptom detection, CT or MRI angiography, and surgical intervention. The management of asymptomatic patients with VRs is a topic of debate. However, early surgical intervention is recommended for patients presenting with symptoms. The operative risks are relatively low. Despite improvement in symptoms after ring division, some patients may still experience postoperative respiratory or digestive issues.

Abbreviations

ALSCA
Aberrant left subclavian artery
CT

Computed tomography
DAA
Double aortic arch
KD
Kommerall's diverticulum
LAA
Left Aortic arch
MRI
magnetic resonance imaging
RAA
Right aortic arch
VR
Vascular ring
VSD
Ventricular septal defect

Declarations

Ethics approval and consent

The Regional Research Ethics Committee approved the retrospective observational study (No. 21-1109, 23/5/2023). Given the retrospective nature of the study and the use of anonymized patient data, the requirement for obtaining individual informed consent was waived by Ethics Committee. No patient names or personally identifiable information were disclosed.

Availability of data and materials

The datasets generated and/or analysed during the current study are available from the corresponding author upon reasonable request.

Competing interests

The authors declare that they have no competing financial or non-financial interests related to the authorship and publication of this manuscript.

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Authors' Contributions

YAB, AMA, OA, IB, AAA, A Abdullah A, AA, and MAH were responsible for study concept & design. YAB and AMA were involved in data collection. YAB, AMA, OA, IB, AAA, A Abdullah A, AA, and MAH shared in

data analysis & interpretation. AMA wrote the first draft of manuscript, all authors reviewed & permitted the final manuscript.

References

1. Worhunsky DJ, Levy BE, Stephens EH, Backer CL. Vascular rings. Semin Pediatr Surg. 2021;30. https://doi.org/10.1016/J.SEMPEDSURG.2021.151128.
2. Yoshimura N, Fukahara K, Yamashita A, Doi T, Yamashita S, Homma T, et al. Congenital vascular ring. Surg Today. 2020;50:1151–8. https://doi.org/10.1007/S00595-019-01907-5.
3. Backer CL, Mongé MC, Popescu AR, Eltayeb OM, Rastatter JC, Rigsby CK. Vascular rings. Semin Pediatr Surg. 2016;25:165–75. https://doi.org/10.1053/J.SEMPEDSURG.2016.02.009.
4. Xu R, Shi K, Yang Z gang, Diao K, yue, zhao Q, Xu Hyan et al. Quantified evaluation of tracheal compression in pediatric complex congenital vascular ring by computed tomography. Sci Rep. 2018;8. https://doi.org/10.1038/S41598-018-29071-9
5. Hanneman K, Newman B, Chan F. Congenital variants and anomalies of the aortic arch. Radiographics. 2017;37:32–51. https://doi.org/10.1148/rg.2017160033.
6. Backer CL, Rigsby CK. Vascular rings: To scope or not to scope. J Thorac Cardiovasc Surg. 2017;154:616–7. https://doi.org/10.1016/J.JTCVS.2017.03.024.
7. Backer CL, Mavroudis C, Rigsby CK, Holinger LD. Trends in vascular ring surgery. J Thorac Cardiovasc Surg. 2005;129:1339–47. https://doi.org/10.1016/j.jtcvs.2004.10.044.
8. Woods RK, Sharp RJ, Holcomb GW, Snyder CL, Lofland GK, Ashcraft KW, et al. Vascular anomalies and tracheoesophageal compression: a single institution's 25-year experience. Ann Thorac Surg. 2001;72:434–8. https://doi.org/10.1016/S0003-4975(01)02806-5.
9. Li S, Wen H, Liang M, Luo D, Qin Y, Liao Y, et al. Congenital abnormalities of the aortic arch: revisiting the 1964 Stewart classification. Cardiovasc Pathol. 2019;39:38–50. https://doi.org/10.1016/J.CARPATH.2018.11.004.
10. Loomba RS. Natural History of Asymptomatic and Unrepaired Vascular Rings: Is Watchful Waiting a Viable Option? A New Case and Review of Previously Reported Cases. Child (Basel). 2016;3. https://doi.org/10.3390/CHILDREN3040044.
11. Bhat YA, Alhabshan F, Almesned A, Alqwaee A. Can Echocardiography Aid in Diagnosing Vascular. Rings? Cureus. 2023;15:e50899. https://doi.org/10.7759/cureus.50899.
12. Humphrey C, Duncan K, Fletcher S. Decade of experience with vascular rings at a single institution. Pediatrics 2006;117. https://doi.org/10.1542/PEDS.2005-1674
13. Evans WN, Acherman RJ, Ciccolo ML, Carrillo SA, Mayman GA, Luna CF, et al. Vascular Ring Diagnosis and Management: Notable Trends Over 25 Years. World J Pediatr Congenit Heart Surg. 2016;7:717–20. https://doi.org/10.1177/2150135116661279.
14. Pasternack DM, Ludomirsky A, Tan RB, et al. Variations in the Evaluation and Management of Vascular Rings: A Survey of American Clinicians. Pediatr Cardiol. 2024;45:959–66.

https://doi.org/10.1007/s00246-024-03442-8.

15. Godtfredsen J, Wennevold A, Efsen F, Lauridsen P. Natural history of vascular ring with clinical manifestations. A follow-up study of eleven unoperated cases. Scand J Thorac Cardiovasc Surg. 1977;11:75–7. https://doi.org/10.3109/14017437709167818.
16. Kouchoukos NT, Masetti P. Aberrant subclavian artery and Kommerell aneurysm: surgical treatment with a standard approach. J Thorac Cardiovasc Surg. 2007;133:888–92. https://doi.org/10.1016/J.JTCVS.2006.12.005.
17. Saran N, Dearani J, Said S, Fatima B, Schaff H, Bower T, et al. Vascular Rings in Adults: Outcome of Surgical Management. Ann Thorac Surg. 2019;108:1217–27. https://doi.org/10.1016/J.ATHORACSUR.2019.04.097.
18. Said SM, Marey G, Knutson S, Rodgers N, Richtsfeld M, Joy B, et al. Outcomes of Surgical Repair of Vascular Rings and Slings in Children: A Word for the Asymptomatic. Semin Thorac Cardiovasc Surg. 2021;33:492–500. https://doi.org/10.1053/J.SEMTCVS.2020.09.013.
19. Backer CL, Russell HM, Wurlitzer KC, Rastatter JC, Rigsby CK. Primary resection of Kommerell diverticulum and left subclavian artery transfer. Ann Thorac Surg. 2012;94:1612–7. https://doi.org/10.1016/J.ATHORACSUR.2012.05.101.
20. Cinà CS, Althani H, Pasenau J, Abouzahr L. Kommerell's diverticulum and right-sided aortic arch: A cohort study and review of the literature. J Vasc Surg. 2004;39:131–9. https://doi.org/10.1016/j.jvs.2003.07.021.
21. Backer CL, Mongé MC, Russell HM, Popescu AR, Rastatter JC, Costello JM. Reoperation after vascular ring repair. Semin Thorac Cardiovasc Surg Pediatr Card Surg Annu. 2014;17:48–55. https://doi.org/10.1053/J.PCSU.2014.01.001.
22. Rato J, Zidere V, François K, Boon M, Depypere A, Simpson JM, et al. Post-operative Outcomes for Vascular Rings: A Systematic Review and Meta-analysis. J Pediatr Surg. 2023;58:1744–53. https://doi.org/10.1016/J.JPEDSURG.2023.02.058.