ORIGINAL ARTICLE

WILEY Congenital Heart Disease

Right aortic arch with situs solitus frequently heralds a vascular ring

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Abstract

Objective: We hypothesized that a right aortic arch in situs solitus, with or without an associated cardiovascular malformation, is often associated with a vascular ring.

Methods: From those born in Southern Nevada between March 2012 and March 2017, we identified 50 (3.6 per 10,000 live births) with a right aortic arch and situs solitus. From the 50 patients, 6 did not meet inclusion criteria for further analysis.

Results: Of the 44 remaining, 33 (75%) had a vascular ring. Of the 33 with a vascular ring, 26 (79%) occurred with an isolated right aortic arch, and 7 (21%) had an associated cardiovascular malformation. Of the total 44 patients with a right aortic arch in situs solitus, 34 (79%) were diagnosed prenatally.

Conclusions: In conclusion, we found a right aortic arch in situs solitus was often associated with a vascular ring. Further, to the best of our knowledge, no previous general population study has demonstrated an equal or higher right aortic arch, prenatal detection rate of 79%.

KEYWORDS
prenatal diagnosis, right aortic arch, vascular ring

1 | INTRODUCTION

Previously, we reported changing trends in vascular ring (VR) management over time.[1] Specifically, we found that the most common VR, whether asymptomatic or requiring surgical repair, consists of an isolated right aortic arch with an aberrant left subclavian artery arising from a Kommerell diverticulum and accompanied by left sided ligamentum or patent ductus arteriosus (RAA-ALSCA VR). Others have reported similar findings.[2] Nevertheless, our previous report noted that then the two major textbooks of pediatric cardiology listed a double aortic arch as the most common VR. Currently, the latest 2016 edition of one of these textbooks still lists a double aortic arch as the most common VR.[3] This report updates our data on situs solitus with a right aortic arch (RAA).

2 | METHODS

This study received approval from the local Institutional Review Board. We obtained data for this observational, nonrandomized, retrospective report by inquiring our research database (Epi-InfoTM) and electronic health records. We analyzed data from all patients, born in Southern Nevada between March 2012 and March 2017, with a RAA, levocardia, and situs solitus that underwent diagnosis and management at the Children's Heart Center Nevada. For the same period and for purposes of prevalence comparison, we separately identified those born with a double aortic arch. We do not consider a double aortic arch a RAA or a left aortic arch variant. We excluded patients with dextrocardia or with heterotaxic situs, including left and right atrial isomerism and situs inversus.

We defined an additional cardiovascular malformation (ACM) as both intracardiac and extracardiac malformations that included those with an isolated, discontinuous pulmonary artery supplied by a patent ductus arteriosus. We defined a VR as the presence of vascular and or ligamentous structures encircling the trachea and esophagus, with or without symptoms. Despite previous reports,[4] we do not consider a left aortic arch with an aberrant right subclavian artery a vascular ring or a "partial" vascular ring, and no such patients were included in this analysis. We undertook fetal cardiovascular evaluations as previously reported.[5] We defined a prenatal detection rate as the number of patients detected prenatally divided by the total number of patients identified that were born during the study period. For Southern Nevada birth numbers, we inquired United States census information.[6] The Children's Heart Center Nevada is the sole provider of congenital heart care in the state, and our database and EHR contain information on all patients diagnosed with a RAA in Southern Nevada. We used SPSS version 13.0 (SPSS Inc., Chicago, Illinois, United States of America) for data analysis. We used non-parametric testing for statistical analysis. We set a P value of < .05 as significant.

3 | RESULTS

We identified 50 patients with a RAA, situs solitus, and levocardia. During the study period, there were approximately 140 000 births in Southern Nevada with an estimated prevalence of RAA in situs solitus at approximately 3.6 per 10 000 live births. During the study period, we also identified 5 patients with a double aortic arch for an approximate prevalence of 0.36 per 10 000 live births; however, for this report, we performed no further analysis on these 5 patients. Of the 50 identified RAA patients, we eliminated 3 that lacked postnatal VR confirmatory testing, and 3 that were lost to follow-up and lacked contemporary medical historical information, leaving 44 for analysis. Of the 44, 33 (75) had a VR or 2.4 per 10 000 live birth (P < .0001 when compared to VR prevalence from a double aortic arch). During the study period, we undertook 7083 individual fetal cardiovascular evaluations, from which we diagnosed 34 (0.52%) with a RAA. No patient with a prenatal diagnosis of a RAA was found to have a double aortic arch by postnatal confirmatory testing. Table 1 summarizes patient characteristics for all 44 patients including prenatal diagnosis rates, percentages that underwent VR surgical repair with or without surgery for an ACM, percentages of those with ongoing follow-up to date with an asymptomatic VR, and frequency of identified syndromes. Genetic studies, inclusive of microarray analysis, were not obtained on all patients. Table 2 lists the postnatal VR confirmatory method for those either prenatally or postnatally diagnosed for all 44 patients. Table 3 breaks down the 33 confirmed vascular rings by type, whether isolated RAA or with an ACM. Table 4 lists the RAA branching pattern anatomy in the 11 patients confirmed without a VR, 10 of which had an ACM and only 1 of which had an isolated RAA. Table 4 includes a RAA anatomical variation without a VR but with an ALSA and PDA from the base of the left common carotid that supplies an isolated, discontinuous left pulmonary artery (Figure 1); an anatomical variant that, to be

best of our knowledge, has not been previously published. Table 5 tabulates the types of associated cardiovascular malformations divided into those with and without at VR.

4 | DISCUSSION

Some consider that a RAA is often a normal anatomical variant;[7] however, apart from situs inversus with dextrocardia in which a RAA is expected, an isolated RAA with situs solitus and a mirror image arterial branching pattern is exceedingly uncommon.[8] We encountered only one such patient in our cohort. Rather, a RAA with situs solitus and a mirror image branching pattern occurs most often associated with an ACM.[9,10]

Prevalence figures for a RAA differ between studies.[11,12] Our general population prevalence figure of approximately 3.6/10 000 live births includes those with a RAA, situs solitus, and levocardia, with or without an ACM. Multiple factors affect the variance of reported RAA prevalence figures from one study to next. Such factors may include diagnostic methods, populations studied, and inclusion and exclusion criteria. Further, when an RAA with situs solitus is found through an

TABLE 1 Patient characteristics

Right aortic arch, n = 44		
n (%)	Isolated RAA n = 27 (61)	RAA with ACM n = 17 (39)
Male, n (%)	13 (48)	10 (59)
Current age in years, median	3	3
Prenatal DX, n (%)	23 (85)	11 (65)
Syndrome, n (%)	4 (15)	4 (24)
22 deletion	1	3
21 trisomy	1	0
Dysmorphic	2	0
FAS	0	1
Vascular ring, n (%)	26 (96)	7 (41)
Repaired, n	11	5
Following, n	15	2

Abbreviations: ACM, associated cardiovascular malformation; DX, diagnosis; FAS, fetal alcohol syndrome; RAA, right aortic arch.

TABLE 2 Postnatal vascular ring confirmatory studies

Postnatal vascular ring confirmatory studies		
	Isolated RAA n = 27	RAA with ACM n = 17
Neonatal echo showing a L or R PDA, n	10	2
CT, n	11	4
MRI, n	6	0
Cath, n	O	11

Abbreviations: ACM, associated cardiovascular malformation; Cath, cardiac catheterization and angiography; CT, computed tomography; MRI, magnetic resonance imaging; RAA, right aortic arch.

TABLE 3 Type of vascular ring

Right aortic arch with vascular ring, n = 33		
	Isolated n = 26	with ACM n = 7
Aberrant left subclavian+ left PDA/Lig, n (%)	24 (92)	6 (86)
Mirror image branching+ left PDA/Lig, n (%)	2 (8)	1 (14)

Abbreviations: ACM, associated cardiovascular malformation; Lig, ligamentum; PDA, patent ductus arteriosus.

echocardiogram, either prenatally or postnatally, our data suggests a greater than 70% chance of an associated VR. In our experience, from Table 1, the chance is lower, about 40%, when an ACM is present; however, the possibility may exceed 90% with an isolated RAA. Some previous studies have noted low VR percentages in patients with a RAA.[7,13] However, such reports may have only included symptomatic patients. In one study, more than 80% of the RAA cohort had a left patent ductus arteriosus, yet conclusions noted less than 15% with a symptomatic VR.[7] Regardless of symptoms, however, the vast majority of patients with a RAA and left ductus arteriosus, with or without an aberrant left subclavian artery, have a VR. Analyzing data from our current study, 33 of 37 (89%) of our patients with a RAA and a left patent ductus arteriosus had VR; currently, however, more than 50% remain asymptomatic.

Multiple references, too numerous to cite, continue to list a double aortic arch as the most common VR. A 1993 Mayo Clinic 45-year review of their surgical experience appears to be the key reference supporting the double aortic arch assertion.[14] In the Mayo Clinic review, 18 of 37 (49%) VR operations were for a double aortic. From the same series, 11 of 37 (30%) procedures were for a RAA-ALSA VR. The remaining 21% included various other rare aortic arch and branching vessel variations. Our previous 25-year review showed similar between the years 1990 and 2005 when 66% or 6 of 9 patients operated had a double aortic arch. However, our more current data from our previous report, between the years 2006 and 2015, demonstrated 79% or 34 of 43 patients operated had a RAA-ALSA VR.[1] In our opinion, the principal reason for this shift is secondary to our high rate of

TABLE 4 Right aortic arch without vascular ring

Right aortic arch without vascular ring, n = 11		
	Isolated n = 1	with ACM n = 10
Mirror image branching+ right PDA/Lig, n (%)[a]	1 (100)	4 (44)
Mirror image branching+ PDA from L innominate, n (%)[a]	NA	3 (33)
Mirror image branching+ Absent PDA, n (%)[b]	NA	2 (11)
Aberrant left subclavian+ PDA from L common carotid, n (%)[c] (Figure 1)	NA	1 (11)

[a]By angiography.
[b]By fetal echocardiography.
[c]By CT scan.
Abbreviations: ACM, associated cardiovascular malformation; L, left; Lig, ligamentum; PDA, patient ductus arteriosus.
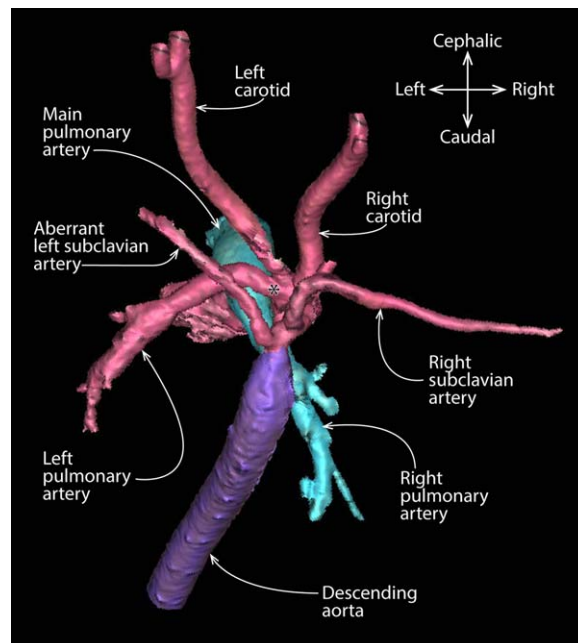

FIGURE 1 Unusual right aortic arch anatomical variation without a vascular ring. Legend: * indicates patent ductus arteriosus

prenatal diagnosis. In our experience, the fetal echocardiography axial, 3-vessel tracheal view more reliably determines the aortic arch morphology, laterality, and branching pattern than an even an immediate postnatal echocardiogram, and especially more reliably than an echocardiogram performed at an older age.

Without prenatal diagnosis, the rarer double aortic arch is often associated with significant symptoms that result in more frequent early presentations than the more common RAA-ALSA VR, which may be asymptomatic, subtly symptomatic, or have persistent symptoms ascribed to other causes. For example, individuals of any age may simply have dysphagia or other chronic, long-lasting gastrointestinal complaints.[15–20] To expand, we have noted some with unusual eating

TABLE 5 Right aortic arch associated cardiovascular malformation

Malformation	with VR n = 7	without VR n = 10
Tetralogy of Fallot	3	3
Tetralogy of Fallot absent pulmonary valve	1	1
Perimembranous VSD	3	0
D-TGA	0	2 (1 with VSD PS)
Pulmonary atresia with VSD	0	1
Univentricular heart, DORV	0	1
Isolated LPA from left common carotid PDA	0	1 (with VSD)
Isolated LPA from left innominate PDA	0	1

Abbreviations: ACM, associated cardiovascular malformation; DORV, double outlet right ventricle; D-TGA, D-transposition of the great arteries; LPA, left pulmonary artery; PDA, patent ductus arteriosus; RAA, right aortic arch; VSD, ventricular septal defect; VR, vascular ring.

habits such as preferences for soups and other soft or liquefied foods over solids (what we have termed the "soup sign"). Such a history can go unrecognized, as individuals may grow up unrecognized by themselves or by their family members that such habits are symptoms to report, unless specifically asked for in a medical history. Also, patients may be diagnosed with asthma, or "exercise induced asthma," or other respiratory disorders only later, after considerable inpatient and outpatient therapy and workup, to have an undiagnosed RAA-ALSA VR.[21–26] Data from our previous VR report reveals for patients lacking a prenatal diagnosis and presenting after 3 years of age (average of 12 years), 17 underwent surgical VR repair, and 16 of 17 (94%) had a RAA-ALSA VR. For the 16 patients, 9 had primarily chronic respiratory complaints, 4 had a combination of respiratory and gastrointestinal symptoms, and 3 had primarily long-term gastrointestinal issues. The chronic respiratory and gastrointestinal symptoms led to multiple pre-VR diagnoses, failed medical treatments and hospitalizations, resulting in considerable patient and family distress, morbidity, and cost. Further, additional case reports and patient series studies also describe the lethality and potential lethality of an undiagnosed RAA-ALSA VR, such as a death and a near death secondary to exsanguination from esophageal erosion caused by a retained foreign body[27,28] and Kommerell diverticulum dissection and rupture from associated vessel wall cystic medical necrosis.[29–32]

This report's limitations include its retrospective nature and the limited number of patients for analysis. Further, we cannot account for asymptomatic, or symptomatic undiagnosed individuals. Without genetic testing on all patients, our incidence of syndromes may be under-reported. Strengths include our high prenatal detection rates, robust database management, electronic health records on all diagnosed patients from a geographic region, and avoidance of relying on third-party sources.

In conclusion, a VR is common in patients with situs solitus, levocardia, and a RAA and especially common in those with an isolated RAA. During the same 5-year period in Southern Nevada, the incidence of a RAA associated VR was approximately 6 times more common than the incidence of VR from a double aortic arch, a highly statistically significant finding. Based on our general population data and that of others,[2] future textbook editions should cease listing a double aortic arch as the most common vascular ring, which appears to be based on reports of surgical experience from more than 2 decades ago. Symptoms from a RAA-ALSA VR, however, may be subtle, and such patients may be inappropriately deemed asymptomatic. Our previous report outlines our center's VR management, however, for even asymptomatic patients with situs solitus and an echocardiographically identified RAA, we would perform an MRI or CT scan in those at least 2 years of age to rule a VR in or out. Without careful medical history questioning of both the patient and family members, symptoms may be subtle and go unrecognized. Finally, prenatal VR diagnosis allows for a coordinated postnatal management plan.

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CONFLICT OF INTERESTS

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