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ORIGINAL ARTICLE

WILEY Congenital Heart Disease

Right aortic arch with situs solitus

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Abstract

Patients: We reviewed all patients evaluated at our center with situs solitus, levocardia, and a right aortic arch that were born between January 2000 and January 2018. **Results**: From our databases, we identified 204 patients. We excluded patients with a double aortic arch from analysis. Of the 204 patients, 103 (50%) were male. Of the 204 patients, 95 (47%) had an isolated right aortic arch. Of the 95 with an isolated right aortic arch, 4 (4%) had chromosome 22q11.2 deletion syndrome, and 89 (94%) had a vascular ring. Of the 204 patients, 109 (53%) had a surgical intracardiac malformation. Of the 109, with an associated surgical intracardiac malformation, 38 (35%) had chromosome 22q11.2 deletion syndrome, and 28 (26%) had a vascular ring. Overall, of the 204 patients, 200 (98%) had a vascular ring, an associated intracardiac malformation, or both. Prenatal detection was 85% (40/47) for the subset of patients born in Southern Nevada between January 2015 and January 2018. **Conclusion**: this review, to best of our knowledge, reports one of largest series of right aortic arches in the literature. Data from our cohort demonstrates that a right aortic arch in situs solitus is almost always associated with pathology. Further, our center's right aortic arch prenatal detection rate exceeds previous reports.

KEYWORDS
congenital heart disease, levocardia, prenatal detection, right aortic arch, situs solitus

1 | INTRODUCTION

At the time of initial cardiac evaluation, a right aortic arch (RAA) is most often first imaged during transthoracic or fetal echocardiography. A right aortic arch is expected in situs inversus with dextrocardia: however, in the presence of situs solitus with levocardia, a RAA is often associated with a vascular ring (VR), an associated surgical intracardiac malformation (AICM), or both.[1,2] Although our opinion differs, some previous RAA reports have also incorporated double aortic arches.[3,4] This study reports our center's wider experience with RAA in situs solitus than our previous reports, and we detail a continuing improvement in prenatal diagnosis.

2 | METHODS

This investigation 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. 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. We analyzed data from all patients born between January 2000 and January 2018 with situs solitus, levocardia, and a RAA. Additionally, we analyzed the period, January 2015 to January 2018, for patients exclusively born in Southern Nevada to estimate a prevalence figure for RAA in the Southern Nevada population and to determine the prenatal detection rate. We did not include patients with a double aortic arch, as we do not consider a double aortic arch as a right aortic arch or a left aortic arch variant. We excluded patients with dextrocardia and heterotaxic situs, including left and right atrial isomerism and situs inversus. We tabulated only surgical AICM. We defined a VR as the presence of vascular and or ligamentous structures encircling the trachea and esophagus, with or without symptoms. Right aortic arch, VR, presence and position or absence of the ductus arteriosus were all

TABLE 1 Data summary for total cohort

January 2000–January 2018 Right aortic arch (RAA) n = 204	
Isolated RAA, n (%)	**95/204 (47)**
VR, n (%)	89/95 (94)
22q11.2DS, n (%)	4/95 (4)
With ICM, n (%)	**109/204 (53)**
VR, n (%)	28/109 (26)
22q11.2DS, n (%)	38/109 (35)

Abbreviations: 22q11.2DS, chromosome 22q11.2 deletion syndrome; ICM, intracardiac malformation; RAA, right aortic arch; VR, vascular ring.

identified by either fetal echocardiography, neonatal echocardiography if the ductus arteriosus were patent, computed tomography, magnetic resonance imaging, or cardiac catheterization and angiography. We undertook fetal cardiovascular evaluations, as we 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 evaluation period. For Southern Nevada birth numbers, we inquired United States census information.[6] Fluorescent in situ hybridization (FISH) testing confirmed chromosome 22q11.2 deletion syndrome (22q11.2DS).

3 | RESULTS

For the period January 2000 to January 2018, we identified 204 patients with a RAA, situs solitus, and levocardia. Of the 204, 103 (50%) were male. Table 1 details data on those with an isolated RAA versus with an AICM, comparing percentages of vascular rings and those with 22q11.2DS,

For the period January 2015 and January 2018, we identified 47 patients born specifically in Southern Nevada. During this period, there were approximately 81,000 live births, and 40 of 47 (85%) patients were diagnosed prenatally, exceeding our previous report.[2] The 40 prenatal diagnoses were made from 4998 pregnant women that were primarily referred for fetal cardiac evaluation because of risk factors for fetal congenital heart disease or a suspected fetal cardiac malformation.

TABLE 2 Data summary for selected Southern Nevada births

January 2015–January 2018 Right aortic arch (RAA) n = 47	
Isolated RAA, n (%)	**28/47 (60)**
VR, n (%)	25/28 (89)
22q11.2DS, n (%)	0
With ICM, n (%)	**19/47 (40)**
VR, n (%)	6/19 (32)
22q11.2DS, n (%)	6/19 (32)

Abbreviations: 22q11.2DS, chromosome 22q11.2 deletion syndrome; ICM, intracardiac malformation; RAA, right aortic arch; VR, vascular ring.

No patient, with a prenatal diagnosis of a RAA, was postnatally found to have a double aortic arch. For the general population of Southern Nevada, 47 patients identified out of 81,000 live births resulted in a RAA prevalence of 5.8 per 10,000 live births. Table 2 details data on those with an isolated RAA versus with an AICM, comparing percentages of vascular rings and those with 22q11.2DS, for the period January 2015 to January 2018.

Table 3, which we organized by the presence and position or absence of the ductus arteriosus or ligamentum and the aortic arch branching pattern, lists the characteristics of all 201 patients. The table details vascular ring data, the number with syndromes and percentage of the syndromes that were 22q11.2DS, the number with an AICM, and the percentage of the AICMs that were conotruncal abnormalities.

4 | DISCUSSION

A RAA occurs in approximately 5 per 10,000 or 0.05% of the population.[7] However, prevalence figures are subject to a report's diagnostic methods and populations studied.[3,8] Our data, for the period January 2015 to January 2018, generated a Southern Nevada RAA prevalence figure of 5.8 per 10,000 live births. During this period, our prenatal detection rate for a RAA was 85%. We previously reported the results of how our regionalized fetal cardiology program, which over time has improved the detection of significant congenital heart disease.[5] Briefly, our approach consists of ongoing community-wide educational programs for general obstetric and specialized perinatal sonographers. Plus, we provide on-site fetal cardiologists to supervise all diagnostic fetal echocardiograms at each of the region's perinatology clinics five days a week, complemented with 24/7 on-call services. Along with an improvement in detecting significant congenital heart disease, a secondary effect is an increase in detection of conditions such as RAAs and vascular rings. The fetal echocardiography three-vessel view is essential to prenatal RAA diagnosis[9]; further, prenatal diagnosis is fundamental to establishing a population's prevalence for conditions, such as a RAA, which, despite the presence of a vascular ring, may be asymptomatic in the newborn and infant period.[1,2]

Congenital heart disease is common in 22q11.2DS, with malformations occurring in more than 70% of cases. Conotruncal defects, frequently accompanied by a right aortic arch, or interrupted aortic arch, or aberrant subclavian arteries, occur most often.[10,11] For all 204 RAA in our series, 62 (30%) had a syndrome and 42 (68%) of the 62 had 22q11.2DS. The incidence of 22q11.2DS was disproportionate, occurring in 100% of those with a RAA, conotruncal malformation, and an absent PDA versus only 4% in those with an isolated RAA and a left ductus or ligamentum.

This report's limitations include its retrospective nature; however, our cohort is reasonably sized for analysis. Nevertheless, we cannot account for asymptomatic, or symptomatic undiagnosed individuals. Without genetic testing on all patients, our incidence of 22q11.2DS may be under-reported; however, we only included data on 22q11.2DS if FISH documentation was present in the medical record. Additional strengths include our high prenatal detection rate, robust

TABLE 3 Data breakdown for total cohort

Arch branching	Right aortic arch n = 204 Ductus/ligamentum arteriosus										
	Right		Left					Absent		Bilateral	
	Normal	ALSCA	Normal	Normal	Normal	ALSCA	ALSCA	Normal	ALSCA	ALSCA	
Ductus/ligamentum connection	RPA-DAo		LPA-DAo	LPA-V	Iso LPA-V	LPA-DK	LPA-V	Iso LPA-V	N/A	N/A	Right: RPA-DAo Left: LPA-V
Vascular ring	No	No	Yes	No	No	Yes	No	No	No	No	No
n (%)	31/204 (15)	1/204 (<1)	21/204 (10)	10/204 (5)	5/204 (2)	96/204 (47)	3/204 (1)	1/204 (<1)	29/204 (14)	6/204 (3)	1/204 (<1)
Syndrome, n (%)	13/31 (41)	0	6/21 (29)	4/10 (40)	2/5 (40)	16/96 (17)	1/3 (33)	0	13/29 (45)	6/6 (100)	1/1 (100)
22q11.2DS n (% of syndromes)	11/13 (85)	0	3/6 (50)	2/4 (50)	0	6/16 (38)	1/1 (100)	0	12/13 (92)	6/6 (100)	1/1 (100)
ICM n (%)	27/31 (87)	0	6/21 (29%)	10/10 100	3/5 (60)	22/96 (23)	1/3 (33)	0	28/29 (97)	6/6 (100)	0
Conotruncal n (% of ICM)	22/27 (81)	0	3/6 (50)	10/10 (100)	2/3 (66)	8/22 (36)	1/1 (100)	0	28/28 (100)	6/6 (100)	0

Abbreviations: 22q11.2DS, chromosome 22q11.2 deletion syndrome; ALSCA, aberrant left subclavian artery; ICM, intracardiac malformation; Iso LPA-V, isolated left pulmonary artery to an arch vessel; LPA-DK, left pulmonary artery to a diverticulum of Kommerell; LPA-DAo, left pulmonary artery to the descending aorta; LPA-V, left pulmonary artery to an arch vessel or to an isolated left subclavian or isolated left common carotid; RPA-DAo, right pulmonary artery to the descending aorta.

database management with the avoidance of relying on information from third-party sources, and available electronic health records on all diagnosed patients evaluated in Nevada.

In conclusion, this review, to best of our knowledge, analyzes one of the largest series of both prenatally and postnatally diagnosed right aortic arches. Our data demonstrates that a right aortic arch with situs solitus is almost always associated with pathology. Further, our center's right aortic arch prenatal detection rate continues to improve, currently standing at 85%.

CONFLICT OF INTEREST

The author(s) declared no conflicts of interest with respect to the research, authorship, and/or publication of this article.

AUTHOR CONTRIBUTIONS

William N. Evans: Concept/Design, Data analysis/interpretation, Drafting article, Critical revision of article, Approval final version.

Ruben J. Acherman, Dean Berthoty, Gary A. Mayman, Michael L. Ciccolo, Sergio A. Carrillo, Humberto Restrepo: Data analysis/interpretation, Critical revision of article, Approval final version.

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