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RISK OF CARDIAC ANOMALIES IN ABERRANT RIGHT SUBCLAVIAN ARTERY RELATIVE AORTIC ARCH ANOMALIES FOR PEDIATRICS: A CROSS-SECTIONAL STUDY

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Abstract

Introduction: *An aberrant right subclavian artery with a normal-sided left aortic arch is the most common congenital anomaly of the aorta.*

This study aims to determine the risk of cardiac anomalies associated with aberrant right subclavian artery in relation to aortic arch anomalies among pediatric patients.

Material and methods: *This prospective cross-sectional study was performed between 2008 and 2011. Patients with either clinical or echocardiographic findings of congenital heart disease were referred for complementary evaluation with computed tomography angiography.*

Results: *Finally, 203 patients were included in this study. The prevalence of an aberrant right subclavian artery was presented in 24 patients (11.8%). Moreover, the most common aortic arch anomaly was coarctation 40 (19.7%). The only double outlet right ventricle cardiac abnormality was associated with a higher risk of an aberrant right subclavian artery (relative risk, 1.192; 95% CI, 0.179 – 7.95) relative to other aortic arch anomalies.*

Discussion: *In conclusion, this study showed that the incidence of aberrant right subclavian artery in an extensive series of Aortic Arch Anomalies had been performed (11.8%). Furthermore, pulmonary atresia, total anomalous pulmonary venous return, aortic coarctation, cervical arch, aortic interruption, tubular hypoplasia, and pulmonary sling were significantly higher in the aberrant right subclavian artery group in contrast with Other Aortic Arch Anomalies. However, just double outlet right ventricle was associated with a higher risk of aberrant right subclavian artery.*

Keywords: *aortic arch anomalies, subclavian artery, aberrant right subclavian artery, cardiac anomalies.*

Introduction

Aortic arch anomalies are seen in 1.5% of the population [*Ad N, Vidne B, 1999; Bae S et al., 2022*]. The most prevalent aortic arch anomaly is a left-sided arch with an aberrant right subclavian artery (ARSA) or arteria lusoria, with a prevalence of 0.5% to 2% of the population [*Konstantinou N et al., 2022*]. In these patients, ARSA originated as the last branch of the aortic arch from the Kommerell diverticulum, which is the result of the persistence of the right aortic embryonic arch [*van Rosendael P et al., 2021*]. Aberrant right subclavian artery, in most cases, crosses the mediastinum

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from left to right, passing behind the esophagus, between trachea and esophagus (18%), and from in front of the trachea (4%), inducing related disease symptoms [*Ka-Tak W et al., 2007; Haaga J et al., 2009; Türkvatan A et al., 2009a*].

According to embryologic double arch theory, interruption of the right aortic arch between the right subclavian artery and descending aorta causes fusion of the right subclavian and carotid arteries to form an innominate artery [*Pleş L et al., 2022*]. If this interruption occurs between the right common carotid artery and the right subclavian artery, it leads to an aberrant right subclavian artery [*Türkvatan A et al., 2009a*].

Most patients are asymptomatic and may be symptomatic in two ends of age spectrum, childhood, and adulthood. Clinical manifestations may appear when a complete vascular ring encircles the trachea or the esophagus [*Umapathi K, Bokowski J, 2022*]. Pediatrics manifest by stridor due to tracheal stenosis, and adults present with dysphagia secondary to extrinsic esophageal compression from ARSA. Furthermore, the growing Kommerell diverticulum leads to dysphagia, named Dysphasia Lusoria. Less common symptoms of aberrant right subclavian artery are superior vena cava syndrome and gastrointestinal tract bleeding and, some case reports of distal embolism of ARSA thrombosis [*Carrizo G, Marjani M, 2004; Haaga J et al., 2009*]. This study aims to find the risk of cardiac anomalies in ARSA relative aortic arch anomalies for pediatric patients.

Material and methods

This study was written by The Strengthening the Reporting of Observational Studies in Epidemiology Statement [*von Elm E et al., 2008*].

Study design and Setting: This retrospective study was performed based on the Tooska Medical Imaging Center, Tehran of all pediatric patients suspected of aortic arch anomalies after approval by the Research Council of Shahid Beheshti University of Medical Sciences and the medical ethics committee based on the principles of the Declaration of Helsinki for human research. All patients also were free to participate in the study, and at first, the purpose and method of conducting the study were explained to the parents/guardians of patients under 16 years old with written consent, if participants did not provide informed consent or if this was not required, they were excluded from this study.

Participants: All pediatric patients with congenital heart disease symptoms (a blue tinge to the skin or lips (cyanosis), rapid breathing, rapid heartbeat, swelling in the legs, tummy, and around the eyes, shortness of breath in babies during feeding (making it hard for them to gain weight) and in older children and adults during exercise, extreme tiredness, and fatigue, fainting during exercise, swelling in the hands, ankles or feet) were included by two expert pediatric cardiologists.

Data measurement: Patients with either clinical or echocardiographic findings of congenital heart disease were referred for complementary evaluation with computed tomography angiography (CTA) for three years. Computed tomography angiography was performed by Light Speed QXI four-row detector with collimation: 1.25 *mm* (50 % overlapping), table speed: 3.75 *cm/s*, and pitch: 0.75. All patients' general anesthesia was implemented, then Visipaque (2-2.5*cc/kg*) was injected by a power injector. Injection velocity was 3cc per second proportional to CTA length. Computed tomography angiography sections were from the neck to the iliac crest. Virtual Reality (VR), Maximum Intensity Projection, and Multiplanar Reconstruction imaging were reconstructed from row imaging in workstation 4.1.

Statistical methods: It was used for statistical analysis of SPSS software (SPSS Inc., Chicago, IL, USA) version 26. For quantitative variables to describe the data center from the mean and to describe the data scatter from the standard deviation were used. In qualitative variables, frequency and percentage were used to describe the data. The normality of the data was checked by Kolmogorov-Smirnov test and the Q-Q diagram. Non-parametric Mann-Whitney U test was used to analyze the results. Relative risk (Confidence Interval (CI) 95 %) was used for cardiac Anomalies in ARSA relevant to other aortic arch anomalies, and the level of significance in the tests was considered 0.05.

Results

Finally, 203 patients were included in this study. The average age was 24 ± 1.7 months, and 47.3 % were girls. The prevalence of ARSA was presented in 24 patients (11.8 %). Moreover, the most common aortic arch anomaly was coarctation 40 (19.7 %) (Table 1).

The most common cardiac anomaly in total, ARSA, and other aortic arch anomalies was ventricular septal defect (VSD) (45.3%, 62.5%, and 43.01%, respectively) (Table 2). There was a statistically significant difference between ARSA and other aortic arch anomaly groups for pulmonary atresia (PA), total anomalous pulmonary venous return (TAPVR), aortic coarctation, cervical arch, aortic interruption, tubular hypoplasia, and pulmonary sling (p>0.05) (Table 2).

The only double outlet right ventricle (DORV) cardiac abnormality was associated with a higher risk of ARSA (relative risk, 1.192; 95% CI, 0.179 – 7.95) relative to other aortic arch anomalies (Table 3).

Discussion

In this study, the prevalence of ARSAs were determined in patients with clinical symptoms or echocardiographic findings of congenital heart disease. An ARSA arises from interruption between the right carotid and right subclavian arteries. This is the most common aortic arch anomaly, with a 0.5 to 2% prevalence

Table 1

Baseline demographic factors for patients with aortic arch anomalies **(n = 203)**

Variables		Patients, n (%)
Age, Mean ± SD (Range), (Month)		24 ± 1.7 (1-33)
Sex, Girl, n (%)		96 (47.3)
Aortic Arch Anomalies, n (%)	Coarctation	40 (19.7)
	VAOA	16 (7.9)
	Tubular Hypoplasia	10 (4.9)
	ARSA	24 (11.8)
	Hypoplastic Arch	20 (9.9)
	RAMI	39 (19.2)
	Interruption	8 (4)
	Cervical arch	5 (2.5)
	Bovine arch	2 (1)
	RAALSA	10 (4.9)
	DAA	1 (0.5)
Anomalies occurred Simultaneous with ARSA, n (%)	Coarctation	4 (2)
	Tubular Hypoplasia	1 (0.5)
	Cervical arch	2 (1)
	Interruption	3 (1.5)
	VAOA	1 (0.5)

Notes: *ARSA = aberrant right subclavian artery, RAMI = right sided arch with mirror image branching, RAALSA = right sided arch with aberrant left Subclavian Artery, DAA = double aortic arch, VAOA = vertebral artery originating from arch*

Table 2

Cardiac anomalies in aberrant right subclavian artery and other aortic arch anomalies

Cardiac Anomalies	Total (n = 203)	ARSA (n = 24)	OA (n = 179)	p-value *
VSD, **n (%)**	92 (45.3)	15 (62.5)	77 (43.01)	0.072
PA, **n (%)**	66 (32.5)	13 (54.2)	53 (29.6)	**0.016**
PDA, **n (%)**	72 (35.5)	10 (41.7)	62 (34.6)	0.5
ASD, **n (%)**	24 (11.8)	5 (20.8)	19 (10.61)	0.146
TGA, **n (%)**	13 (6.4)	2 (8.3)	11 (6.14)	0.682
Situs inversus, **n (%)**	5 (2.5)	0	5 (2.79)	0.408
DILV, **n (%)**	1 (0.5)	0	1 (0.55)	0.714
TAPVR, **n (%)**	1 (0.5)	1 (4.3)	0	**0.006**
Dextrocardia, **n (%)**	7 (3.4)	1 (4.3)	6 (3.35)	0.838
DORV, **n (%)**	10 (4.9)	1 (4.3)	9 (5.02)	0.855
Hypoplastic left heart, **n (%)**	4 (2)	1 (4.3)	3 (1.67)	0.411
AP window, **n (%)**	2 (1)	1 (4.3)	1 (0.55)	0.094
Aortic coarctation, **n (%)**	4 (2)	4 (16.6)	0	**< 0.001**
Cervical arch, **n (%)**	2 (1)	2 (8.3)	0	**< 0.001**
Aortic interruption, **n (%)**	3 (1.5)	3 (12.5)	0	**< 0.001**
Tubular hypoplasia, **n (%)**	1 (0.5)	1 (4.3)	0	**0.006**
Pulmonary sling, **n (%)**	1 (0.5)	1 (4.3)	0	**0.006**

Notes:

ARSA *- aberrant right subclavian artery,*
OA *- Other Aortic Arch Anomalies*
VSD *- ventricular septal defect,*
PA *- pulmonary atresia,*
PDA *- patent ductus arteriosus,*
ASD *- atrial septal defect,*
TGA *- transposition of great arteries,*
TAPVR *- total anomalous pulmonary venous return,*
DORV *- double outlet right ventricle,*
AP *window - aortopulmonary window,*
DILV *- double Inlet Left Ventricle.*
** - Mann-Whitney U test*

Table 3

Relative risk of cardiac anomalies in aberrant right subclavian artery relative other aortic arch anomalies

Cardiac Anomalies	Relative Risk (with 95% confidence interval)
VSD	0.497 (0.228 – 1.084)
PA	0.408 (0.193 – 0.861)
PDA	0.769 (0.36 – 1.64)
ASD	0.506 (0.21 – 1.238)
TGA	0.753 (0.198 – 2.85)
Dextrocardia	0.821 (0.129 – 5.249)
DORV	**1.192 (0.179 – 7.95)**
Hypoplastic left heart	0.462 (0.081 – 2.63)
AP window	0.229 (0.054 – 0.964)

Notes: *VSD = ventricular septal defect, PA = pulmonary atresia, PDA = patent ductus arteriosus, ASD = atrial septal defect, TGA = transposition of great arteries, TAPVR = total anomalous pulmonary venous return, DORV = double outlet right ventricle, AP window = aortopulmonary window, DILV = double inlet left ventricle*

[*Ka-Tak W et al., 2007; Chaoui R et al., 2008; Türkvatan A et al., 2009*]. This anomaly is usually asymptomatic and an incidental radiological finding, but clinical symptoms are arisen from tracheal or esophageal compression after constituting a vascular ring [*Carrizo G, Marjani M, 2004; Türkvatan A et al., 2009*]; respiratory symptoms and dysphagia are common symptoms in childhood and adults, respectively [*Türkvatan A et al., 2009*].

The ARSA is usually an isolated anomaly, but it sometimes accompanies other cardiovascular anomalies, mainly coarctation, patent ductus arteriosus (PDA), intracardiac defects, anomalous pulmonary artery circulation, and carotid or vertebral artery anomalies [*Türkvatan A et al., 2009b*]. Furthermore, approximately 60% of patients with an aberrant subclavian artery show a Kommerell diverticulum, and 3% to 8% of them present aneurysms of the diverticulum [*Fisher R et al.,* 2005] Aberrant subclavian artery has been prenatally observed to be more common in trisomy 21 fetuses and other chromosomal abnormalities, such as microdeletion 22q11 [*von Elm E et al., 2008*; *Scala C et al., 2015*]. The frequency of this anomaly varies in these disorders because of the targeted examination of this vessel in finding ARSA in prenatal and postmortem studies [*von Elm E et al., 2008; Paladini D et al., 2012; Ayaz R et al., 2020*].

According to the Chahwan et al. (2006) study, the reported anomalies associated with ARSAs are a common origin of the bilateral common carotid arteries, a replaced right or left vertebral artery, a non-recurrent right inferior laryngeal nerve, coarctation, a right-sided thoracic duct, and a right-sided aortic arch. Detection of these anomalies is essential to selecting the most proper method of surgical correction. For more than 250 years, an aortic arch anomaly entitled aberrant subclavian artery has been recognized. Also, different reports have shown its association with disorders such as dysphagia lusoria, Kommerell diverticulum, aneurysms, vascular rings, and Down's syndrome [*Ramaswamy P et al., 2008*].

In our study, an ARSA with coarctation of the aorta (24 cases) and patent ductus arteriosus (PDA) (72 cases) is associated with intracardiac defects. Moreover, the most common cardiac anomalies associated with ARSA include VSD (62.5 %), PA (54.2 %), PDA (41.7 %), and ASD (20.8 %). In the present study, tubular hypoplasia, cervical arch, and interruption were seen in 1, 2, and 3 patients, respectively. Cina C et al. (2008) reported 53% rupture or dissection among 32 patients with aberrant left subclavian artery and right aortic arch, and in the study of Nicholas T et al. (2007), dissection in 2 of their ten patients. In Türkvatan et al. (2009b) study, the ARSA originating from the left aortic arch and left subclavian artery originating from the right-sided arch was identified by multidetector CT angiography in 11 and 6 patients, respectively. In 3 and 2 patients with aberrant right subclavian artery was seen Kommerell diverticulum and aneurismal Kommerell diverticulum, respectively. The aberrant subclavian artery was observed with complex congenital heart diseases such as Intra cardiac defect (1 patient), coarctation of the aorta (2 patients), PDA (2 patients), and vertebral artery (1 patient).

Since multidetector CT angiography has advantages such as shorter imaging time, high spatial resolution, high availability, and being valid and non-invasive, it is the primary imaging technique for evaluating thoracic and vascular anomalies. The aberrant right subclavian artery diagnosis is of clinical importance because it can associate with clinical symptoms of tracheoesophageal compression, aneurysm formation, aortic interruption, and

other anomalies. CT angiography can also show the details of vascular anatomy and their associations with adjacent organs, which is considered the most essential advantage than other techniques in evaluating the right subclavian artery [*Türkvatan A et al., 2009b*].

In conclusion, this study showed that the incidence of ARSA in an extensive series of Aortic Arch Anomalies had been performed (11.8 %). Furthermore, pulmonary atresia, total anomalous pulmonary venous return, aortic coarctation, cervical arch, aortic interruption, tubular hypoplasia, and pulmonary sling were significantly higher in the ARSA group in contrast with other aortic arch anomalies. However, just double outlet right ventricle, was associated with a higher risk of ARSA.

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