Pediatr Cardiol 14:159–161, 1993

Pediatric Cardiology

Aberrant Right Subclavian Artery with Left Aortic Arch: Associated Cardiac Anomalies

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SUMMARY. **Anomalous origin of the right subclavian artery (ARSA) from the aorta distal to the normally positioned left subclavian artery is a relatively frequent congenital anomaly in subjects with left aortic arch. The purpose of this study was to determine the relative frequency of associated cardiovascular anomalies in individuals with this anomaly. From the records of approximately 11,000 pathologic specimens in the Registry of Cardiovascular Disease of United Hospital (St. Paul, MN, USA), we found 128 (1.2%) with ARSA.**

Of the 128 ARSA, 117 (2.9%) occurred among 4102 instances of congenital heart disease.

The 117 cases with congenital heart disease and ARSA were conotruncal anomalies in 38%, septal defects in 28%, obstructive anomalies of the left side of the heart in 21%, right heart anomalies in 5%, and miscellaneous conditions in the other 8%. Down syndrome existed in 14 (12%) of the 117 specimens with ARSA and some congenital cardiac anomaly; nine of the latter had an atrioventricular canal (AVC) malformation.

KEY WORDS: **Aberrant right subclavian artery — Congenital heart disease — Down syndrome — Persistent left superior vena cava**

Aberrant (anomalous) origin of the right subclavian artery (ARSA) from the aorta distal to the normally positioned left subclavian artery is one of the more frequent congenital anomalies in subjects with left aortic arch (LAA). The ARSA crosses the midline from left to right posteriorly to the esophagus. In this study we sought to determine the relative frequencies of cardiovascular anomalies associated with ARSA.

Materials and Methods

Among approximately 11,000 accessions of pathological specimens from patients of all ages in the Registry of Cardiovascular Disease of United Hospital, St. Paul, Minnesota, there are 4102 cases with a diagnosis of one or another congenital cardiovascular anomaly. This study sought the types of associated cardiovascular anomalies in those cases in which there was ARSA with LAA.

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Results

ARSA with LAA was identified in 128 patients, among whom were 117 patients with congenital heart disease. Of these 117 patients, the gender was known in 101 cases. Among these, 49 (49%) were male and 52 (51%) were female. The 117 cases with congenital heart disease were found among 4012 cases with congenital heart disease (2.9%). Most patients studied were younger than 1 year of age, reflecting the seriousness of the associated cardiac anomalies. The 117 patients with congenital heart disease could be classified according to cyanosis—present in 72 (62%) cases; no cyanosis—present in 40 (34%) cases; and variable cyanosis in five (4%) cases.

The 117 cardiac anomalies were grouped into five general categories (Table 1). These were conotruncal anomalies (45 cases, 38%), septal defect (33 cases, 28%), left heart obstructions (24 cases, 21%), right heart anomalies (six cases, 5%), and other (nine cases, 8%).

Among the 72 primarily cyanotic conditions

Table 1. ARSA with cardiac anomalies

Condition	Frequency among 4102 cases		ARSA (n = 117)	
	Group	Specific condition	Group	Specific condition
Conotruncal	987	—	45 (4.6%)	—
Truncus arteriosus	—	98	—	7 (7%)
DORV	—	184	—	11 (6%)
TOF	—	355	—	19 (5%)
TGA	—	350	—	8 (2%)
Septal defects	1587	—	33 (2.1%)	—
AVC (complete or partial)	—	390	—	19 (5%)
VSD (isolated)	—	728	—	10 (1.4%)
ASD (isolated)	—	469	—	4 (0.9%)
Left heart obstructions	1168	—	24 (2.1%)	—
Aortic/mitral atresia	—	72	—	4 (6%)
Mitral stenosis (all forms)	—	64	—	3 (4.7%)
Mitral atresia	—	71	—	3 (4%)
Aortic atresia	—	155	—	5 (3%)
IAA	—	67	—	2 (3%)
Coarctation	—	538	—	6 (1%)
Aortic stenosis	—	201	—	1 (0.5%)
Right heart anomalies	260	—	6 (2.3%)	—
Tricuspid atresia	—	99	—	4 (4%)
Pulmonary stenosis/atresia/hypoplasia	—	161	—	2(1.2%)
Other[a]	100	—	9 (9%)	—

DORV, double outlet right ventricle: TOF, tetralogy of Fallot; TGA, transposition of great arteries; AVC, atrioventricular canal; VSD, ventricular septal defect; ASD, atrial septal defect; IAA, interruption of aortic arch.

[a] Single examples of congenital aortic insufficiency, rhabdomyoma, endocardial fibroelastosis, right pulmonary artery from aorta, total anomalous pulmonary venous connection, single ventricle, conjoined hearts of twins, pulmonary venous stenosis, and patent ductus arteriosus.

(Table 1), the anomalies that had the greater frequencies of LAA with ARSA for the specific condition were those with conotruncal anomalies or left heart obstructive anomalies. The conotruncal anomalies were a persistent truncus arteriosus, origin of both great arteries from the right ventricle (double outlet right ventricle), and tetralogy of Fallot (TOF). The left heart obstructive lesions were most often combined aortic and mitral atresia.

Of the 45 primarily acyanotic states (40) or conditions in which cyanosis was uncertain (5) (Table 1), those most frequently associated with ARSA from an LAA were persistent common atrioventricular canal (AVC), including complete and incomplete forms, and congenital mitral stenosis.

Chromosomal and other syndromes were present in 20 of the 117 cases with ARSA and cardiac anomaly (Table 2). Down syndrome occurred in 14 patients, nine of whom had an AVC anomaly.

A persistent left superior vena cava (LSVC) was identified in 20 of the 117 cases. In each, the LSVC connected to the coronary sinus. Of the 20 specimens with LSVC, the cardiac anomaly was TOF in four, AVC in four, VSD in two, aortic/mitral atresia in two, and mitral atresia in two. The other examples of LSVC were single instances of different conditions.

Table 2. ARSA with cardiac anomalies: Associated syndromes

Condition	ARSA
Down syndrome	14[a]
Trisomy 18 syndrome	3[b]
Noonan syndrome	1[c]
Postrubella syndrome	1[d]
Potter syndrome	1[d]

[a] AV canal, 9; ventricular septal defect, 4; tetralogy of Fallot, 1.
[b] Ventricular septal defect, 2; aortic/mitral atresia, 1.
[c] Mitral stenosis due to parachute mitral valve.
[d] Atrial septal defect.

Discussion

The goal of this study was to ascertain the relative frequencies of ARSA as the fourth branch of an LAA in different forms of congenital cardiac anomalies. The overall incidence of ARSA in the general autopsy population of all age groups, or in specific

patient groups such as Down syndrome, was not studied.

The frequency of ARSA of 1.2% (128 of 11,000 cases) in the Registry collection is somewhat higher than the generally reported incidence in the general population of one in 200 individuals [1–4, 13, 14], because our material is heavily weighted with cardiovascular abnormalities. Since the focus of this study was congenital cardiac anomalies, the observed approximately equal gender distribution we found cannot be compared to general studies of the frequency of ARSA in which females predominate [1, 9].

The 117 instances of ARSA were identified from a group of 4102 hearts with congenital cardiac anomalies (approximately 3% of anomalous hearts). Beabout and co-workers [1] observed a congenital cardiac anomaly in 18 (43%) of 42 cases with ARSA studied at autopsy. Molz and Burri [9] reported that about one half of their own cases of ARSA were associated with either or both intracardiac and extracardiac vascular anomalies.

Relatively little information has been reported as to the frequencies of ARSA in specific cardiac anomalies. We found 5% or more frequency of ARSA in truncus arteriosus (7%), DORV (6%), aortic and mitral atresia (6%), and TOF (5%). Blake and Manion [3] emphasized the frequency of ARSA in the TOF. Among conditions without cyanosis or which vary as to arterial desaturation, ARSA was more common in AVC (5%) and mitral stenosis (3%). Becker and co-workers [2] and Molz and Burri [9] reported associations of ARSA with aortic arch anomalies. We found ARSA in 3% of cases of interrupted aortic arch and in 1% of coarctation of the aorta.

In this study, 14 (12%) of the 117 patients with ARSA and congenital heart disease had Down syndrome. Studies of cardiac anomalies in patients with Down syndrome [5–8, 10–12] reported frequencies of ARSA that varied from 2% [10], to 36% [6, 11]. Other reported frequencies [5, 7, 8, 12] were between these extremes. Among our 14 cases of ARSA and Down syndrome, nine (64%) had AVC anomalies.

Twenty patients (17%) with ARSA and a congenital cardiac anomaly had an LSVC. Winter [15] reported two instances of ARSA (7%) among 30 cases of LSVC. Marino and associates [8] found 15 patients with LSVC in their study of 220 patients with AVC defects (7%); eight (53%) of the 15 also had Down syndrome. In our material, TOF (20%), persistent common atrioventricular anomalies (20%), and aortic and mitral atresia (20%) were the congenital cardiac anomalies most often associated with the combination of LSVC with ARSA. Recognition of an LSVC preoperatively may be helpful to the surgeon.

The low frequency of ARSA among our cases without congenital heart disease is to be explained. One explanation is that in the majority of cases without congenital heart disease, only the heart had been submitted to the Registry, leaving a void in answering the question as to whether or not an anomalous right subclavian artery had been present. In contrast, in most of the cases with congenital heart disease, the thoracic aorta was present with the heart-lung block.

Acknowledgment. Supported by the United Hospital Foundation, St. Paul, Minnesota.

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