Managing Cervical Aortic Arches in the Adult Population: a Meta-analysis of Case Reports

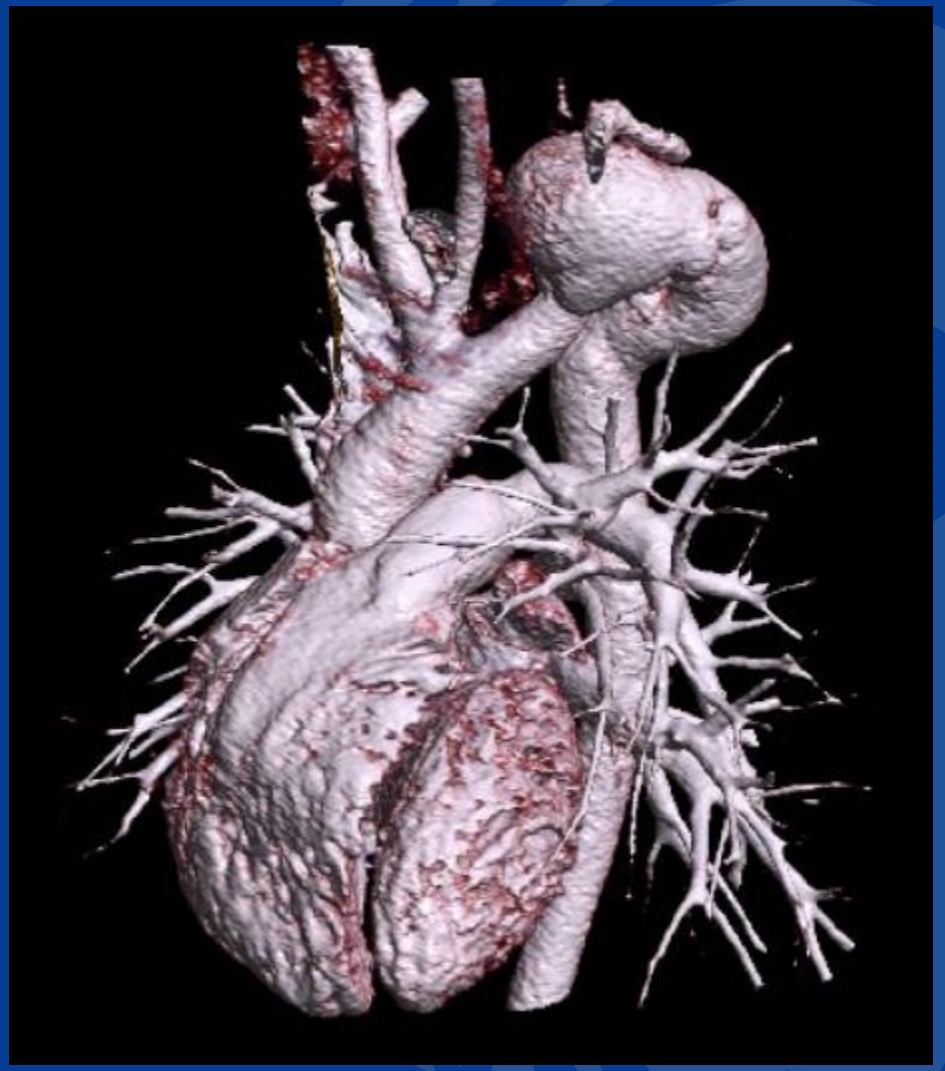
Massimo Baudo, MD[1,2]; Serge Sicouri, MD[1]; Yoshiyuki Yamashita, MD, PhD[1,2]; Basel Ramlawi, MD[1,2]

1. Department of Cardiac Surgery Research, Lankenau Institute for Medical Research, Main Line Health, Wynnewood, PA, USA

2. Department of Cardiac Surgery, Lankenau Heart Institute, Main Line Health, Wynnewood, PA, USA

The cervical aortic arch (CAA) is a very rare congenital anomaly of the aorta development in which the aortic arch is located above the superior aspect of the clavicle, sometimes protruding high into the neck.

CAA was first described by Reid in 1914[1] and currently cases reported in literature are limited.



1. Reid DG. Three Examples of a Right Aortic Arch. *J Anat Physiol* (1914) 48:174–181

Baudo M, Varrica A, Reali M, et al. Cervical aortic arch in the pediatric population: a meta-analysis of individual patient's data. *Front. Cardiovasc. Med* 2023 Sep 28:10:1266956

Historically, Haughton and colleagues proposed in 1975 the first classification of CAA[1] consisting in 5 morphological types (A to E) based on their experience and a review of the available literature.

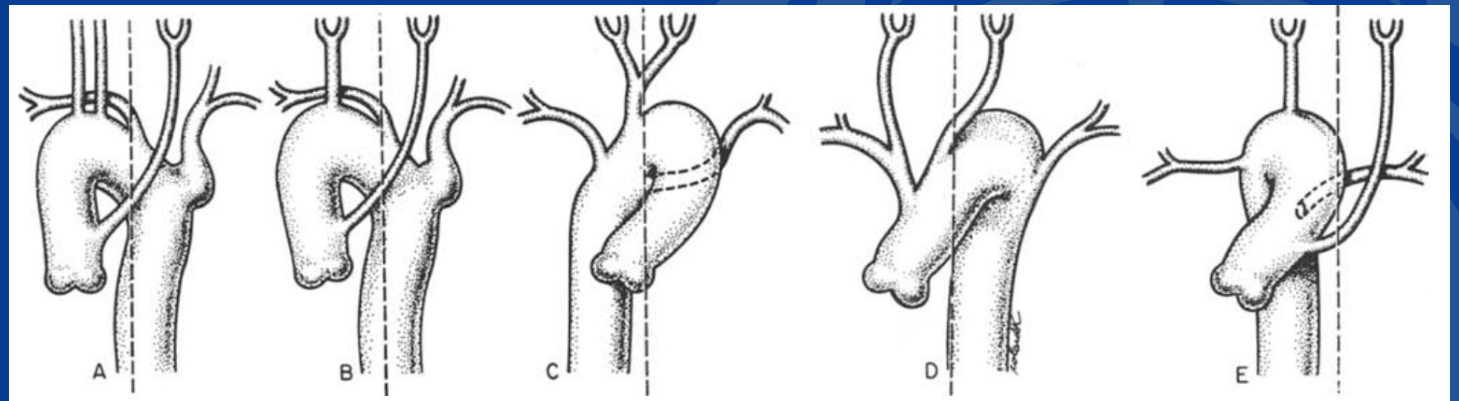
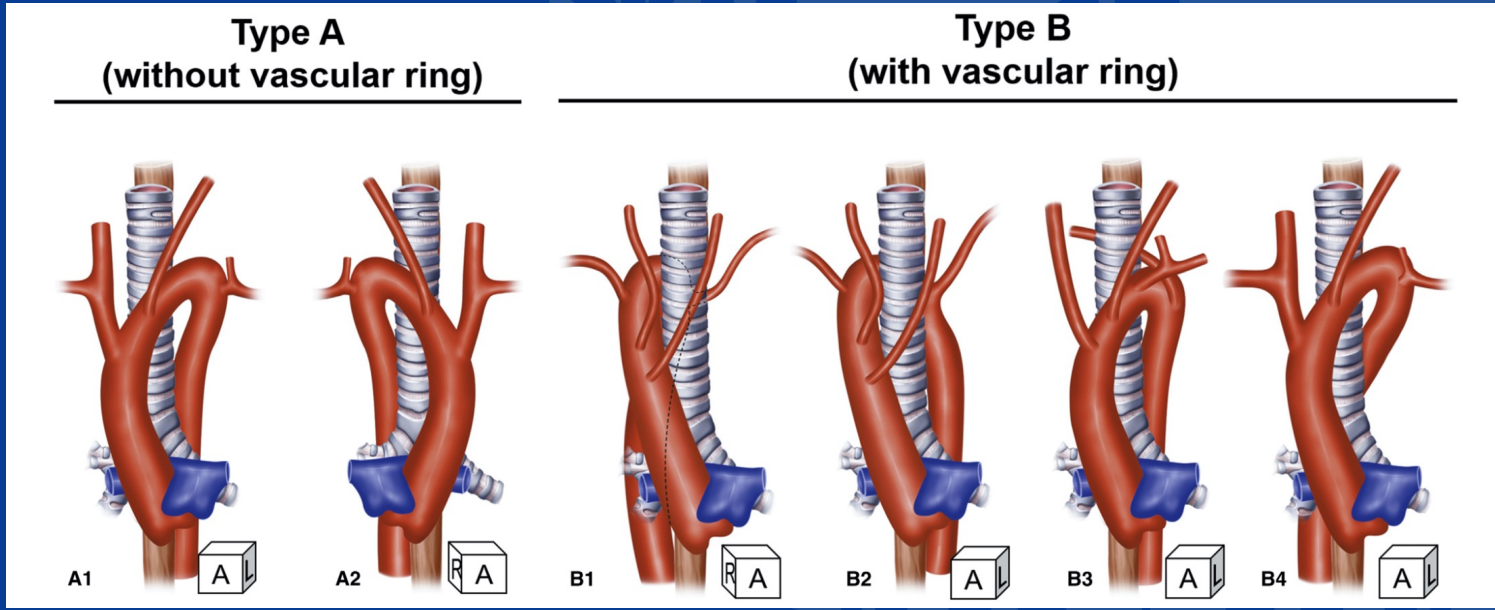


Fig. 5. Diagrammatic representation of five types of cervical aortic arch.

1. Haughton VM, Fellows KE, Rosenbaum AE. The cervical aoratic arches. *Radiology* (1975) 114:675–68

More recently, Zhong et al. proposed a revised classification of CAA for surgical decision making[2]. The classification consists of 2 types and 6 subtypes based on the presence of vascular ring.

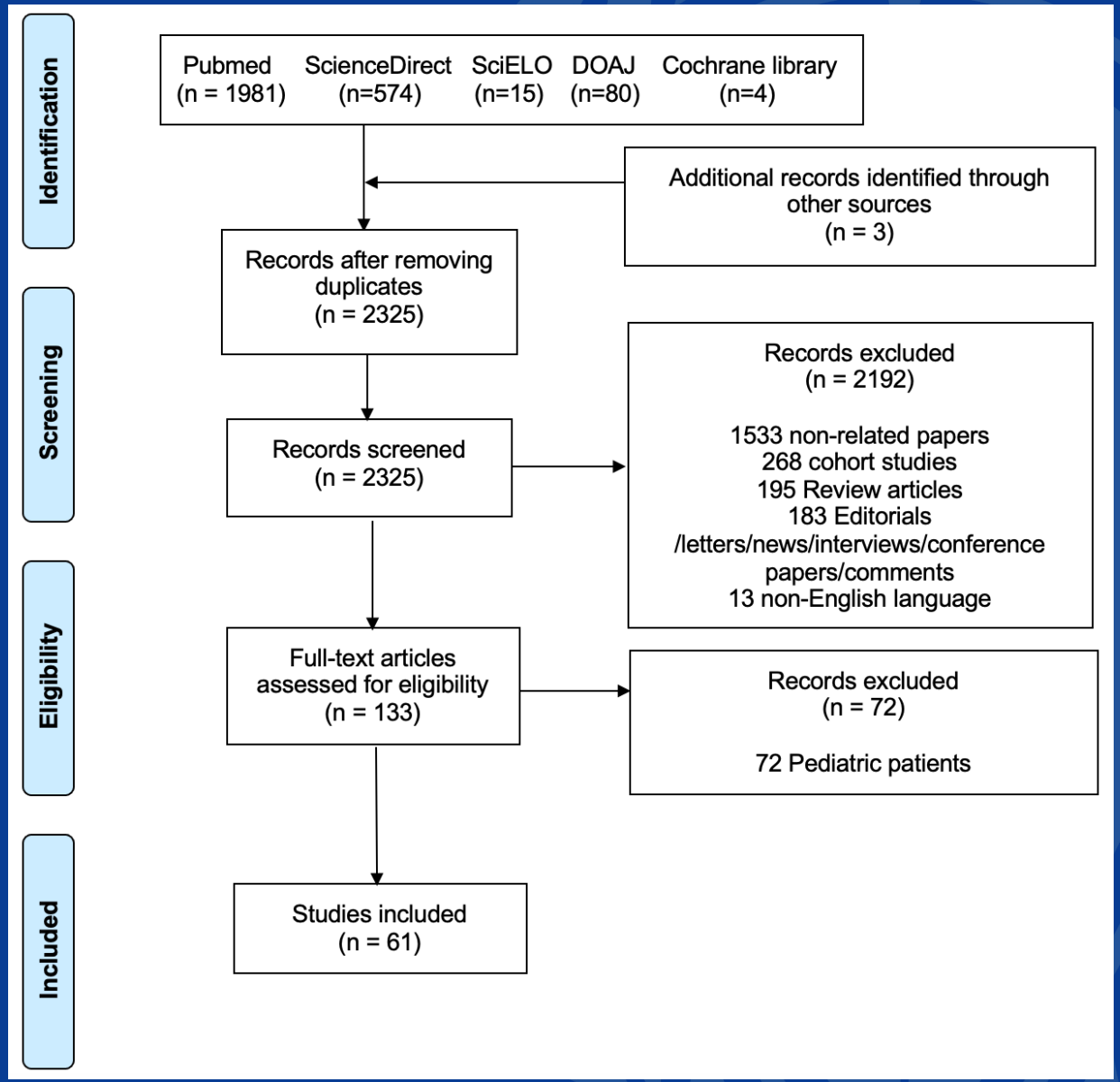


2. Zhong Y-L, Ma W-G, Zhu J-M, et al. Surgical repair of cervical aortic arch: An alternative classification scheme based on experience in 35 patients. *J Thorac Cardiovasc Surg* (2020) 159:2202-2213.e4.

PubMed, ScienceDirect, SciELO, DOAJ and Cochrane Library databases were searched until December 2023 for case reports describing the presence of a cervical aortic arch in the adult age.

Case reports and series were included if the following criteria were met: 1) description of the cervical aortic arch; 2) age ≥18 years 3) English language. Exclusion criteria for analysis were all other forms of papers that did not have individual patient data.

The literature search identified 2325 potentially eligible articles, 61 of which met our inclusion criteria with 71 patients.



Symptoms	Value
Asymptomatic, % (n)	45.7% (32/70)
Symptoms, % (n)	54.3% (38/70)
Vascular compression, % (n)	60.5% (23/38)
Dyspnea, % (n)	31.6% (12/38)
Dysphagia, % (n)	31.6% (12/38)
Respiratory tract infection, % (n)	5.3% (2/38)
Cough, % (n)	2.6% (1/38)
Pain, % (n)	21.1% (8/38)
Headache, % (n)	15.8% (6/38)
Dizziness, % (n)	15.8% (6/38)
Palpitations, % (n)	13.2% (5/38)
Hoarseness, % (n)	10.5% (4/38)
Fatigue, % (n)	10.5% (4/38)
Paresthesia, % (n)	7.9% (3/38)
Syncope, % (n)	2.6% (1/38)
Diplopia, % (n)	2.6% (1/38)

Signs	Value
Pulsatile mass, % (n)	49.3% (35/71)
Bruit, % (n)	38.0% (27/71)
Limb pressure difference, % (n)	26.8% (19/71)
Thrill, % (n)	18.3% (13/71)
Any Horner’s signs, % (n)	4.2% (3/71)
Cyanosis, % (n)	2.8% (2/71)
Stridor, % (n)	0% (0/71)

Patient's Characteristics	Value
Age, years (mean ± SD)	38.6 ± 15.4
Female sex, % (n)	67.1% (47/70)
Hypertension, % (n)	15.5% (11/71)
Aortic features	
CAA left laterality, % (n)	67.6% (48/71)
Aneurysm, % (n)	62.0% (44/71)
Kinking, % (n)	11.3% (8/71)
Kommerell diverticulum, % (n)	9.9% (7/71)
Coarctation, % (n)	8.5% (6/71)
Arch Dissection, % (n)	1.4% (1/71)
Type B Dissection, % (n)	1.4% (1/71)
Rupture, % (n)	1.4% (1/71)
Bicuspid aortic valve, % (n)	5.6% (4/71)
Persistent left SVC, % (n)	5.6% (4/71)
Previous Cardiac Surgery	4.2% (3/71)
Previous Neck exploration	2.8% (2/71)

Treatment	Value
Surgical, % (n)	62.7% (42/67)
Open, % (n)	88.1% (37/42)
Sternotomy, % (n)	48.5% (16/33)
Thoracotomy, % (n)	33.3% (11/33)
Cervicotomy, % (n)	18.2% (6/33)
Laparotomy, % (n)	9.1% (3/33)
Clamshell, % (n)	3.0% (1/33)
Endovascular, % (n)	11.9% (5/42)
Conservative, % (n)	37.3% (25/67)
Refused surgery, % (n)	7.5% (5/67)
Inoperable, % (n)	1.5% (1/67)

Among papers reporting follow-up time (n=28), mean follow-up was 12 months [IQR: 6-24].

Only one death was reported occurring in a critical patient presenting with a dissecting aneurysm and considered at inoperable risk.

There is only one clinical study reporting on CAA including 35 young-adult patients.

Characteristics were similar to our analysis:
- higher left-sided CAA prevalence,
- higher female prevalence,
- around 50% asymptomatic patients.

Despite possible publication bias of our analysis, surgical outcomes were similar regarding postoperative and follow-up mortality (0%).

Surgical repair of cervical aortic arch: An alternative classification scheme based on experience in 35 patients

Yong-Liang Zhong, MD, Wei-Guo Ma, MD, Jun-Ming Zhu, MD, Zhi-Yu Qiao, MD, Jun Zheng, MD, Yong-Min Liu, MD, and Li-Zhong Sun, MD

ABSTRACT

Objective: Cervical aortic arch (CAA) is rare and difficult to repair. Clinical experience is limited. We report the surgical techniques and midterm outcomes in 35 patients with CAA based on an alternative classification scheme.

Methods: Of 35 patients with CAA, 30 (85.7%) had left-sided aortic arch and 5 had (14.3%) right-sided aortic arch (all 5 had a vascular ring). Mean age was 34.2 ± 13.1 years, 23 were female (65.7%), and 18 were asymptomatic (51.4%). Surgical access and procedure were chosen according to an alternative classification scheme that is based on the presence or absence of vascular ring and relationship of descending aorta to the side of the aortic arch. In the left-sided aortic arch group, aortic arch reconstruction though median sternotomy was performed in 15 patients, and distal arch and descending thoracic aortic replacement via left thoracotomy in 15 patients. In the right-sided aortic arch group, ascending-to-descending aortic bypass was done via median sternotomy in 2 patients and right thoracotomy in 1, and distal arch and descending thoracic aortic replacement via right thoracotomy in 2 patients.

Results: Neither death nor spinal cord injury occurred. Left recurrent laryngeal nerve injury, prolonged ventilation, and reexploration for bleeding occurred in 1 each. In 11 patients with coarctation, the upper-lower limb gradient decreased significantly postoperatively (from 34.0 ± 12.7 to 10.2 ± 2.7 mm Hg; $P < .01$). The diseased aortic segment was excluded in 34 patients, except 1 with residual aneurysm in the proximal descending thoracic aorta. Follow-up was complete in 100% at mean 4.4 ± 2.0 years. No late death, limb ischemia, or stroke occurred. Endovascular repair was performed in 1 patient, and ascending aortic dilation occurred in 1 patient. The residual aorta remained nondilated in 33 patients. Aortic grafts were patent in 100%, with no anastomotic leak or pseudoaneurysm. At 6 years, the incidences of death, aortic events, and event-free survival were 0%, 6.5%, and 93.5%, respectively.

Conclusions: Open repair of CAA can achieve favorable early and midterm outcomes. Surgical accesses and procedures should be chosen based on type of CAA, anatomic variations and associated anomalies. Our alternative categorization scheme of CAA is intuitive and comprehensive, which may facilitate classification and surgical decision making. (J Thorac Cardiovasc Surg 2020;159:2202-13)

A previous meta-analysis of case reports analyzed CAA characteristics and outcomes in the pediatric population only.

Besides the prevalence of left-sided CAA, distinct cardiac and vascular characteristics (due to congenital heart diseases, CHD) were identified when compared to our analysis on the adult population.
While signs and symptoms were mostly similar.

Mortality rates were higher in the pediatric population, probably due to the concomitant severe CHD affecting pediatric patients.

Cervical aortic arch in the pediatric population: a meta-analysis of individual patient's data

Massimo Baudo[1,2], Alessandro Varrica[1], Matteo Reali[1], Antonio Saracino[3], Mario Carminati[3], Alessandro Frigiola[1], Alessandro Giamberti[1] and Mauro Lo Rito[1]*

[1]Department of Congenital Cardiac Surgery, IRCCS Policlinico San Donato, San Donato Milanese, Italy, [2]Department of Cardiac Surgery, ASST Spedali Civili di Brescia, University of Brescia, Brescia, Italy, [3]Department of Pediatric and Adult Congenital Cardiology, IRCCS Policlinico San Donato, San Donato Milanese, Italy

Background: This is the first meta-analysis to analyze all reports of published pediatric cases of cervical aortic arch (CAA) by highlighting the clinical characteristics and treatment outcomes using the reported individual data of the patients. The aim of the study is to investigate the clinical features and surgical outcomes of such a rare disease in the pediatric population.
Methods: A comprehensive search was conducted in various academic databases, including PubMed, ScienceDirect, SciELO, DOAJ, and Cochrane Library, until June 2022 for case reports describing the presence of cervical aortic arch in the pediatric age. Case reports and series were included if the following criteria were met: (1) description of the cervical aortic arch; (2) patient of pediatric age; and (3) published in the English language. All other types of publications that lacked patient-specific information were excluded from the analysis. This systematic review was conducted in accordance with the PRISMA guidelines. The primary outcome measure of the analysis was early and late mortality.
Results: The literature search identified 2,272 potentially eligible articles, 72 of which met our inclusion criteria with 96 patients including the author's institutional case. At a median of 365 (90–730) days, the overall cohort registered a 7.3% (7/96) mortality rate. In the subset of patients who underwent surgery, the mortality rate was also 7.3% (4/55), and the mortality rate following surgery to treat only CAA was 2.4% (1/42). Dyspnea was identified as an independent determinant of mortality by employing the univariable Firth bias-reduced logistic regression method.
Conclusion: Cervical aortic arch is a rare congenital heart disease that poses treatment challenges due to the high anatomical variability, diverse clinical presentations, and the presence of other concomitant diseases. The surgical treatment appears to be a safe and effective approach for resolving the symptoms, although it needs to be tailored individually for each patient.

Conclusion

- Cervical aortic arch is an uncommon congenital heart condition that presents challenges in diagnosis and treatment due to its high anatomical variability, diverse clinical manifestations, and presence of concomitant diseases.
- Not all adult CAA require surgical intervention. Despite the lack of guidelines addressing this rare condition, possible surgical indications include the presence of symptoms and concomitant conditions requiring surgical attention (i.e., aneurysm, dissection, etc.)
- Surgical intervention appears to be a safe and effective resolution for symptoms, albeit requiring an individualized approach.