Canadian Journal of Cardiology 35 (2019) 438–445

Clinical Research

The Natural and Unnatural History of Congenital Aortic Arch Abnormalities Evaluated in an Adult Survival Cohort

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See editorial by Guron and Oechslin, pages 373–375 of this issue.

ABSTRACT

Background: This study describes the different types of congenital vascular rings according to their anatomy, symptoms, and age at clinical onset and reports the surgical outcomes.

Methods: A retrospective observational database study was conducted, reviewing the medical charts of 69 adult survivors with a history of a vascular ring, identified from the Dutch Congenital Cor vitia database.

Results: Median age at presentation was 8.5 years (0-53.0 years). Thirty patients (43.5%) had a "left aortic arch with aberrant right subclavian artery," 21 patients (30.4%) a "double aortic arch," and 16 patients (23.2%) a "right aortic arch with aberrant left subclavian artery." The main symptomatology at presentation comprised respiratory symptoms (82.9%). Almost three-quarters of patients were also diagnosed with asthma/bronchial hyperreactivity. Patients with a

RÉSUMÉ

Contexte : Cette étude décrit les différents types d'anneaux vasculaires congénitaux selon leur anatomie, leurs symptômes et l'âge à leur apparition clinique ainsi que les issues de l'intervention chirurgicale.

Méthodologie : Une étude rétrospective observationnelle a été réalisée à partir d'une base de données, dans laquelle ont été analysés les dossiers médicaux de 69 adultes survivants ayant des antécédents d'anneaux vasculaires repérés dans la base de données des anomalies congénitales *CONgenital COR vitia* des Pays-Bas.

Résultats : L'âge médian à la survenue était de 8,5 ans (0-53,0 ans). Trente patients (43,5 %) présentaient un « arc aortique gauche avec artère sous-clavière droite aberrante », 21 patients (30,4 %) avaient un « double arc aortique » et 16 patients (23,2 %), un « arc aortique droit avec artère sous-clavière gauche aberrante ». Les problèmes

The term "vascular ring" indicates a rare congenital vascular anomaly.[1] It represents less than 1% of all congenital heart defects,[2,3] due to an abnormal development of the branches of the aortic arch. This causes vascular encirclement of the trachea and/or esophagus, leading to variable degrees of compression and giving rise to a wide range of respiratory and gastrointestinal symptoms that vary in severity and age at presentation.[3,4] Anatomically, different types of vascular rings exist, which may be divided into 2 groups (complete and incomplete rings).[5,6] Because of the low incidence of vascular rings, their anatomical heterogeneity, and their nonspecific symptomatology, this pathology is often hardly recognized, leading to mis- or undiagnosed cases having lifelong complaints.[7,8] The aim of this study was to describe the different types of vascular rings, in a cohort of 69 patients, according to

Received for publication June 23, 2018. Accepted December 4, 2018.

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See page 444 for disclosure information.

https://doi.org/10.1016/j.cjca.2018.12.004

double aortic arch had more symptoms than patients with a left aortic arch with aberrant right subclavian artery and right aortic arch with aberrant left subclavian artery ($P < 0.001$), requiring surgery most often ($P < 0.001$). In patients with childhood onset of symptoms, preoperative spirometry (ie, peak expiratory flows) was more often abnormal as compared with adult patients ($P = 0.007$). Surgery was performed in 42.0% of all patients at a median age of 17 years (0-63.0 years). Twenty-four (92.3%) of the operated patients showed improvement or complete relief of symptoms shortly after surgery. Of 26 asymptomatic nonoperated patients, 3 patients (11.5%) eventually developed symptoms.

Conclusions: The low incidence of vascular rings, their anatomic heterogeneity, and a wide range of common symptoms often lead to misdiagnosis. Clinical awareness is warranted as a large subset of patients could benefit from surgery, even at an adult age.

respiratoires étaient les principaux symptômes initiaux (82,9 %). Près des trois quarts des patients avaient également reçu un diagnostic d'asthme ou d'hyperréactivité bronchique. Les patients présentant un double arc aortique avaient plus de symptômes que ceux qui présentaient un arc aortique gauche avec artère sous-clavière droite aberrante ou un arc aortique droit avec artère sous-clavière gauche aberrante ($p < 0,001$) et avaient plus souvent besoin d'une chirurgie ($p < 0,001$). Chez les patients dont les symptômes étaient apparus pendant l'enfance, la spirométrie préopératoire (c.-à-d. la mesure du débit expiratoire de pointe) était plus souvent anormale que chez les patients adultes ($p = 0,007$). Une intervention chirurgicale avait été pratiquée chez 42,0 % de tous les patients à un âge médian de 17 ans (0-63,0 ans). Une atténuation ou une disparition complète des symptômes a été observée chez vingt-quatre (92.3%) des patients opérés, peu de temps après la chirurgie. Sur 26 patients asymptomatiques et non opérés, 3 patients (11,5 %) ont fini par présenter des symptômes.

Conclusions : La faible incidence des anneaux vasculaires, leur hétérogénéité anatomique et la vaste gamme de leurs symptômes les plus courants conduisent souvent à des erreurs de diagnostic. Une meilleure sensibilisation clinique s'impose, car un important sous-ensemble de patients pourrait tirer profit d'une intervention chirurgicale, même à l'âge adulte.

their anatomical heterogeneity, presenting symptoms, and differences in age at clinical onset, to raise clinical awareness of this complex congenital anomaly. The secondary objective was to report the (long-term) surgical outcomes.

Patients and Methods

Study population

A retrospective observational database study was performed, using the computerized coding system of the Dutch Congenital Cor vitia (CONCOR) registry for adult patients with congenital heart disease. From November 2001, Dutch patients 18 years or older with a congenital heart defect have been recruited in secondary and tertiary medical centres and, after giving informed consent, included in this database. This is a survival cohort because this database includes only patients with a congenital cardiac anomaly who survived (and who are not lost to follow-up) until the age of 18 years or more. In this database, clinical events are coded using the European Pediatric Cardiac Code Short List coding scheme.[9] For this study, a selection of the European Pediatric Cardiac Code Short List coding scheme was made, and all patients coded with "vascular ring," "aortic branch abnormality," "right aortic arch," "double aortic arch (DAA)," "aberrant origin right subclavian artery," and "pulmonary artery sling" as code were selected. A list of 209 adult patients from 7 academic centres in The Netherlands with a possible complete or incomplete vascular ring was obtained.

Patient and clinical variables

The medical charts of the adult patients identified from the CONCOR database were examined. To determine the true existence of a vascular ring, the operative reports and imaging studies were reviewed. If no vascular ring was present, the patient was excluded and no further data were collected. If a vascular ring was present, the following patient and treatment information were obtained: date of birth, sex, type of vascular ring, comorbidities, associated (cardiac) anomalies, date of clinical onset, (presenting) symptoms, date of diagnosis, diagnostic strategies and their results, date and type of surgery, postoperative complications and outcomes, length of hospital stay, need for reoperation, and length of follow-up. The type of vascular ring was classified according to the categories by Backer and Mavroudis,[6] based on the anatomical characteristics of the vascular ring. Of note, according to this established nomenclature of vascular rings, those aortic anomalies that compress a portion of the esophagus or trachea, without encircling it as a complete ring, were also considered a vascular ring.

The date of clinical onset was defined as the patient-reported date of the first occurrence of symptoms, most likely related to the vascular ring. To evaluate the possible differences between patients with an early (ie, below 18 years) or late onset (ie, 18 years or older) of symptoms, patients were divided into 2 subgroups. The postoperative outcome was divided into 3 categories: (1) cured (complete relief of symptoms), (2) improved (decrease in severity, duration, or number of episodes), and (3) no improvement.[9]

Statistical analysis

Data were analyzed using IBM SPSS Statistics version 22.0 for Windows (IBM Corp, Armonk, NY). Descriptive statistics were applied to demonstrate all variables. Continuous data were presented as means and standard deviations when normally distributed or medians and range when nonnormally distributed and categorical data as frequencies and percentages. Comparison of categorical variables was made using the χ^2 test (or Fisher's exact test). Comparison of 2 paired categorical variables was made using the Wilcoxon signed-rank test. Comparison of continuous variables was made using the independent sample t test, Mann-Whitney U test, or Kruskal-Wallis H test, depending on the number of groups

Table 1. Demographic characteristics of a cohort of 69 patients with a vascular ring

Demographics	Value
Sex	
Female	41 (59.4)
Age at diagnosis (y)	11.0 (0-70.0)
Comorbidities (n = 23)	
Asthma/bronchial hyperreactivity	17 (73.9)
Atopy	5 (21.7)
Hypertension	7 (30.4)
Diabetes mellitus	1 (4.3)
Renal failure	1 (4.3)
Associated cardiac anomalies (n = 44)	
Ventricular septal defect	19 (43.2)
Coarctation of the aorta	13 (29.5)
Atrial septal defect	8 (18.1)
Tetralogy of Fallot	8 (18.1)
Transposition of the great arteries	4 (9.1)
Patent ductus arteriosus	3 (6.8)

Values are given as numbers (%) or medians (range).

and distribution of data. Associations between continuous and ordinal variables were assessed using the Kendall Tau-b correlation coefficient. All variables were visually checked for normality using Q-Q plots and histograms. In case of doubt, normality was tested using the Shapiro-Wilk test. A P value of < 0.05 was considered statistically significant.

Results

Demographics

Of 209 patients, 140 patients were excluded, because they did not meet the criteria of a vascular ring (eg, some appeared to have a "right arch" only). Also, no patients with a pulmonary sling could be identified. In total, 69 patients with a vascular ring were included in this study. At the time of data extraction, 15,602 patients were included in the CONCOR database, leading to a point prevalence of vascular rings in the adult population with a congenital heart defect of 0.44%. The demographic characteristics are depicted in Table 1. The median age of diagnosis for all patients, including the asymptomatic patients, was 11.0 years (range, 0-70.0 years). Comorbidities were found in 23 patients (33.3%), with asthma/bronchial hyperreactivity being the most frequently diagnosed comorbid disease (73.9%) and being significantly more common in patients with a DAA than in patients with other types of vascular rings (52.9% vs 47.0%, $P = 0.046$). Associated cardiac anomalies were found in 44 (63.8%) patients.

Type of vascular rings

The distribution of vascular rings in the study population and the associated age at clinical onset and the age at surgery are shown in Table 2. Of the 69 patients with a vascular ring, 30 (43.5%) had a "left aortic arch with aberrant right subclavian artery" (LAARSA) (Fig. 1), 21 (30.4%) a DAA (Fig. 1), and 16 (23.2%) a "right aortic arch with aberrant left subclavian artery" (RAALSA). One patient had an "innominate artery compression" and 1 patient a "right cervical aortic arch." A Kommerell's diverticulum was reported in 8 (11.6%) of the 69 patients.

Presenting symptoms

Of all patients, including patients with innominate artery compression and right cervical aortic arch, 37 patients became symptomatic (53.6%). The median age at clinical onset was 8.5 years (range, 0-53.0 years). Twelve patients (32.4%) had symptoms present early after birth. There was no significant difference in age at clinical onset between the different types of vascular rings ($P = 0.738$, Table 2). The median age of diagnosis for the symptomatic patients was 16.0 years with a wide range from 0 to 63.0 years. Three patients (8.1%) were diagnosed before symptoms started. Thirteen patients (35.1%) had a delay in the diagnosis of 1 year or more, with a median delay of 11.0 months (range, 1.0-639.0 months). Patients with a DAA were more often symptomatic than patients with a RAALSA and LAARSA (85.7% vs 50.0% and 30.0%, respectively, $P < 0.001$). Respiratory symptoms were the most common type of symptoms (n = 29, 82.9%), with a significant predominance for patients with a DAA (ie, 81.0% vs 37.5% for RAALSA and 20.0% for LAARSA, $P < 0.001$). Dyspnea was the most common respiratory symptom (n = 25, 86.2%). A significant difference between the different types of vascular rings was found for dyspnea ($P < 0.001$), recurrent respiratory tract infections ($P = 0.009$), stridor ($P = 0.001$), chest pain ($P = 0.037$), and cyanosis ($P = 0.043$), all of them

Table 2. Distribution of vascular rings with ages at clinical onset (CO) and surgery

Type of vascular ring	Patients	Age at CO (y)	Age at surgery (y)
Double aortic arch	21 (30.4)	8.0 (0-53.0)	10.0 (0-55.0)
Balanced arches	2 (9.5)		
Left arch dominant	1 (4.8)		
Right arch atretic	–		
Right arch patent	1 (100.0)		
Right arch dominant	14 (66.7)		
Left arch atretic	6 (42.9)		
Left arch patent	5 (35.7)		
Unknown	4 (19.0)		
Right aortic arch, left ligament, aberrant left subclavian artery	16 (23.2)	3.0 (0-30.0)	11.0 (0-44.0)
Left aortic arch, aberrant right subclavian artery	30 (43.5)	23.0 (0-39.0)	23.0 (0-63.0)
Innominate artery compression	1 (1.4)	26.0 (–)	28.0 (–)
Right cervical aortic arch	1 (1.4)	8.0	–
Total	69 (100.0)	8.5 (0-53.0)	12.0 (0-63.0)

Values are given as numbers (%) or medians (range).

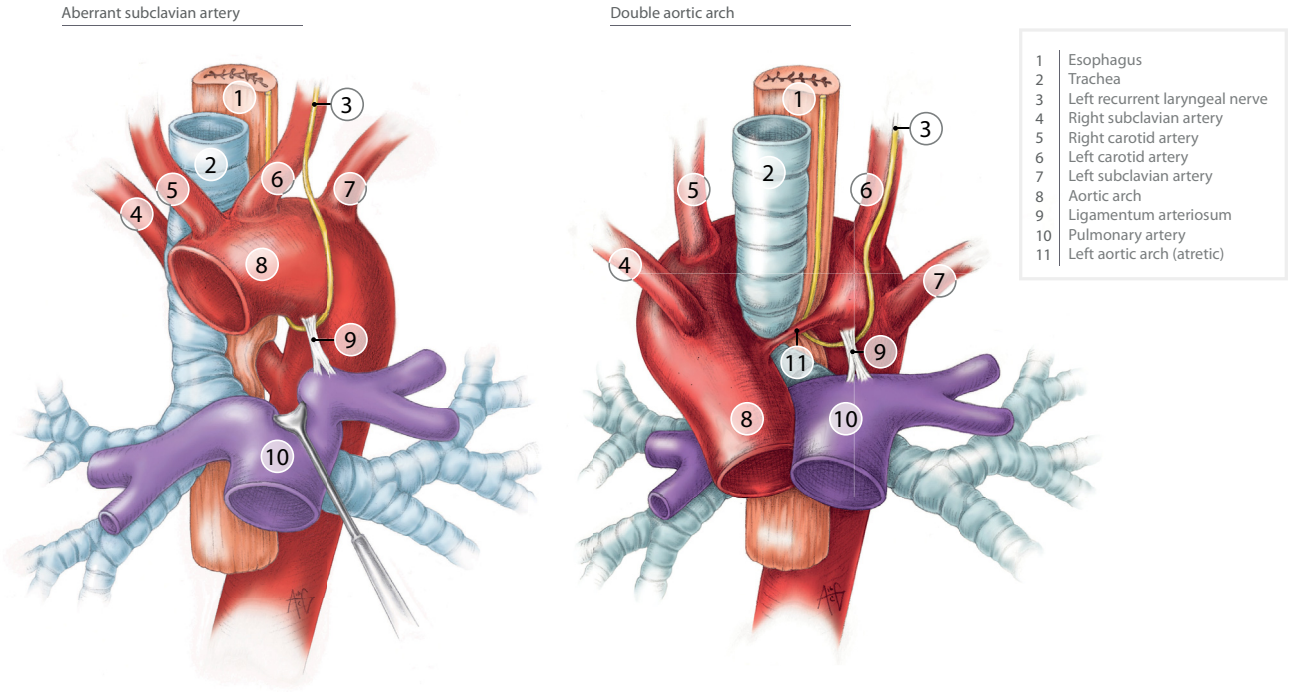

Figure 1. The 2 most common vascular rings: a left aortic arch with aberrant right subclavian artery constituting an incomplete ring and a double aortic arch. Of note, in the latter variant, the left arch is partially atretic, which cannot be visualized easily with current imaging modalities as the atretic parts do not fill with contrast medium.

predominating in patients with a DAA. Gastrointestinal symptoms were found 13 times (35.1%), with dysphagia for solid foods being the most common (n = 9, 69.2%) (Table 3).

Characteristics according to the age of clinical onset

The 2 subgroups of symptomatic patients according to the age of clinical onset (ie, childhood vs adulthood) consisted of 24 (64.9%) and 13 (35.1%) patients, respectively. No significant difference in the types of vascular rings between the adult and the paediatric group existed.

The number of respiratory or gastrointestinal symptoms did not differ significantly between the paediatric and adult groups ($P = 0.643$ and $P = 0.472$, respectively). Only chest pain was significantly more common in the adult group ($P = 0.042$).

Preoperative spirometry was performed in 16 of 37 symptomatic patients (43.2%). Of them, 9 patients (56.3%) had paediatric onset and 7 patients (43.7%) adult onset

Table 3. Distribution of presenting symptoms according to the type of vascular ring

Symptoms	DAA (n = 21)	RAALSA (n = 16)	LAARSA (n = 30)	Total	*P* value
Respiratory	17 (81.0)	6 (37.5)	6 (20.0)	29 (82.9)	< **0.001***
Dyspnea	17 (81.0)	5 (31.3)	3 (10.0)	25 (86.2)	< **0.001***
Recurrent RTI	7 (33.3)	2 (12.5)	1 (3.3)	10 (34.5)	**0.009**
Stridor	6 (28.6)	—	—	6 (20.7)	**0.001**
Cough	4 (19.0)	3 (18.8)	1 (3.3)	8 (27.6)	0.088
Wheezing	4 (19.0)	1 (6.3)	—	5 (17.2)	0.092
Chest pain	3 (14.3)	—	3 (10.0)	6 (20.7)	**0.037**
Cyanosis	4 (19.0)	1 (6.3)	—	5 (17.2)	**0.043**
Dysphonia	1 (4.8)	—	—	1 (3.4)	0.552
Gastrointestinal	3 (14.3)	4 (25.0)	6 (20.0)	13 (37.1)	0.622
Dysphagia for solid foods	3 (14.3)	2 (12.5)	4 (13.3)	9 (69.2)	0.470
Weight loss	—	—	4 (13.3)	4 (30.8)	0.068
Emesis	—	1 (6.3)	2 (6.7)	3 (23.1)	0.353
Gastroesophageal reflux	2 (9.5)	1 (6.3)	—	3 (23.1)	0.112
Nausea	—	1 (6.3)	—	1 (7.7)	0.096
Total	18 (85.7)	8 (50.0)	9 (30.0)	35 (100.0)	< **0.001**

Values are given as numbers (%). *P* values refer to Fisher's exact test. Because "innominate artery compression" and "right cervical aortic arch" represented only 1 patient per group, these types of vascular rings were excluded from statistical analysis in this table. The bold *P* values are significant.

DAA, double aortic arch; LAARSA, left aortic arch, aberrant right subclavian artery; RAALSA, right aortic arch, aberrant left subclavian artery; RTI, respiratory tract infection.

*χ^2 test.

Table 4. Preoperative spirometry results according to the age of clinical onset (CO)

Spirometry (n = 16)	Patients CO < 18 (n = 9)	Value	Patients CO ≥ 18 (n = 7)	Value	*P* value
FVC (L)	6	4.17 ± 1.17	6	4.02 ± 1.13	0.821
FVC% of predicted	6	101.8 ± 17.8	6	103.3 ± 21.5	0.898
FEV_1 (L)	7	2.82 ± 0.84	7	2.87 ± 0.96	0.931
FEV_1% of predicted	8	81.9 ± 19.9	7	96.0 ± 20.1	0.196
PEF (L/s)	5	3.81 ± 0.83	5	6.98 ± 1.80	**0.007**
PEF% of predicted	5	58.4 ± 4.3	5	90.2 ± 19.5	**0.007**
FEF_{25} (L/s)	4	3.35 ± 0.83	3	5.42 ± 1.00	**0.030**
FEF_{25}% of predicted	4	57.3 ± 8.2	2	88.0 ± 18.4	**0.038**
FEF_{50} (L/s)	4	2.64 ± 0.49	5	4.22 ± 0.60	**0.004**
FEF_{50}% of predicted	4	65.3 ± 3.3	5	94.4 ± 19.3	**0.026**
FEF_{75} (L/s)	4	1.81 ± 0.55	3	1.63 ± 0.59	0.695
FEF_{75}% of predicted	4	84.3 ± 17.0	2	83.0 ± 29.7	0.948
Plateau expiratory flow-volume curve	5		5		0.462*
Plateau inspiratory flow-volume curve	2		–		0.303*

Values are given as numbers. *P* values in the table refer to the independent sample *t* test. The bold *P* values are significant.

FEF_{25}, forced expiratory flow at 25% of the forced vital capacity; FEF_{50}, forced expiratory flow at 50% of the forced vital capacity; FEF_{75}, forced expiratory flow at 75% of the forced vital capacity; FEV_1, forced expiratory volume in 1 s; FVC, forced vital capacity; PEF, peak expiratory flow.

*Fisher's exact test.

(Table 4). Preoperative forced vital capacity (FVC), forced expiratory volume in 1 second, and forced expiratory flow after 75% of FVC expired values appeared to be normal in both groups, with mean percentages of predicted above 80%. Preoperative peak expiratory flow (PEF), forced expiratory flow after 25% of FVC expired, and forced expiratory flow after 50% of FVC expired values were found to be significantly lower and below normal in patients with paediatric onset, with mean percentages of predicted under 80% (independent sample *t* test, $P = 0.007$, $P = 0.038$, and $P = 0.026$, respectively). A plateau in the expiratory flow-volume curve was found in 55.6% and 71.4% of patients with paediatric and adult onset, respectively. Figure 2 shows a flow-volume curve of a representative patient from the study population.

Surgery

When looking at surgery details (Table 5), 29 of 69 patients underwent surgery (42%) at a median age of 17 years (range, 0-63 years), with significant predominance for patients with a DAA (16 of 21 patients with a DAA were operated) compared with patients with a RAALSA and LAARSA (76.2% vs 43.8% and 16.7%, $P < 0.001$). Seven of 8 patients with a Kommerell's diverticulum were operated (87.5%). There was no significant difference in age at surgery between the different types of vascular rings (Kruskal-Wallis *H* test, $P = 0.135$, Table 2). Symptoms were present in 26 (89.6%) of these 29 operated patients. Three asymptomatic patients underwent surgery because it was expected that they would develop symptoms. Of the 40 patients who did not undergo surgery, 29 (72.5%) were asymptomatic. The other 11 (symptomatic) patients (27.5%) did have mild symptoms not requiring surgery or were inoperable because of associated cardiac anomalies. The most often performed approach of surgery to divide the arch was a left thoracotomy (n = 16, 55.2%). Complications were registered in 7 patients (24.1%) and included transient vocal cord paresis (n = 4), pneumothorax (n = 2), chylothorax (n = 1), and Horner syndrome (n = 1). A reoperation was required in 5 patients (17.2%) and included aortopexy, tracheopexy, tracheoplasty, tracheal dilatation with a patch, and removal of fibrotic tissue. No patients died during or shortly after surgery.

Postoperative outcomes and follow-up

The median length of hospital stay was 9.0 days (range, 4.0-130.0 days). Figure 3 shows the differences in postoperative outcomes at the first (median 1 month after surgery) and last follow-up (median 5 years after surgery) for the symptomatic patients (n = 26). Of all symptomatic, operated patients, 92.3% showed improvement or complete relief of symptoms at the first follow-up. The outcome score did not

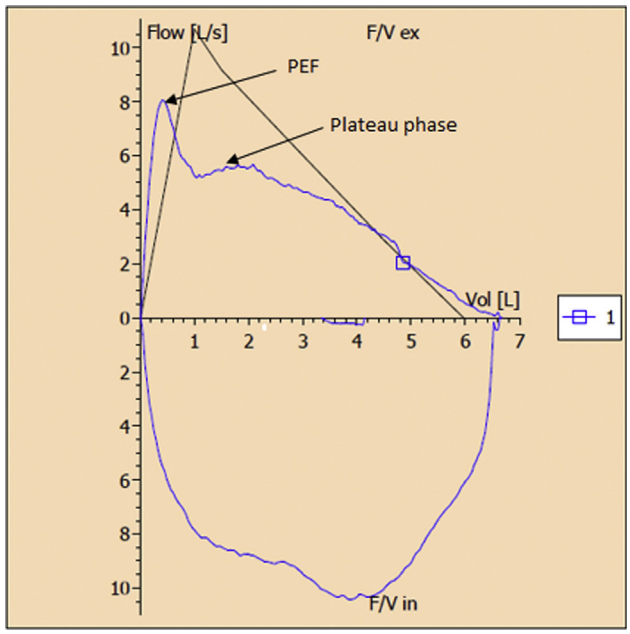

Figure 2. Spirometry performed in a representative patient from the study population with a double aortic arch. The flow-volume curve shows a sudden drop in the expiratory flow after the forced expiratory flow (PEF), followed by a plateau phase, consistent with variable intrathoracic obstruction during forced expiration.

Table 5. Treatment details according to the type of vascular ring

Variables	DAA (n = 21)	RAALSA (n = 16)	LAARSA (n = 30)	INN (n = 1)	RCAA (n = 1)	Total	*P* value
Surgery performed	16 (76.2)	7 (43.8)	5 (16.7)	1 (100.0)	–	29 (42.0)	< **0.001**
Age at surgery (y)	10 (0-55)	11 (0-44)	23 (0-63)	28 (–)	–	17 (0-63)	–
Complications	3 (18.8)	3 (42.9)	1 (20.0)	–	–	7 (24.1)	0.621
Reoperation required	3 (18.8)	1 (14.3)	–	1 (100.0)	–	5 (17.2)	0.221

Values are given as numbers (%) or as medians (range). *P* values in the table refer to Fisher's exact test. The bold *P* values are significant.

DAA, double aortic arch; INN, innominate artery compression; LAARSA, left aortic arch, aberrant right subclavian artery; RAALSA, right aortic arch, aberrant left subclavian artery; RCAA, right cervical aortic arch.

significantly change between the first and last time of follow-up (Wilcoxon signed-rank test, $P = 0.109$, with a median outcome score of "2" [improvement] both at the first and last follow-up). There was no significant correlation between the outcome at the first or last postoperative follow-up and the age at surgery ($P = 0.065$ and $P = 0.110$, respectively).

The median length of follow-up of all patients was 18 years (range, 0-63.0 years). Four patients (10.0%), who were not operated, died during the follow-up at 35, 36, 38, and 67 years of age, due to congenital heart disease. Of 10 nonoperated, symptomatic patients, 6 patients (60.0%) had spontaneous relief of symptoms. Of 26 asymptomatic patients who did not undergo surgery, 3 patients (11.5%) eventually developed symptoms.

Discussion

Vascular rings are congenital anomalies characterized by an abnormal development of the aortic arch. This causes complete or partial encirclement of the trachea and/or esophagus and results in a wide range of symptoms, also seen in other more common diseases, which often complicate the diagnosis.[1,2,4] This retrospective observational database study is, to our knowledge, the largest in the world, reporting characteristics of 69 adult patients with a history of vascular rings.

Vascular rings

In this study, a point prevalence of vascular rings in the adult population with a congenital heart defect of 0.44% was found. This is in line with the percentages of less than 1% stated in previous reports.[2,3] A complete vascular ring was present in more than half of the patients and comprised 21 patients (30.4%) with a DAA, with the right arch being dominant in most cases (66.7%), and 16 patients (23.2%) with a RAALSA. The vast majority of patients with an incomplete ring constituted of patients with a LAARSA. These findings are in line with the previous literature,[10-12] reporting DAA as the most common type of a complete vascular ring, with the right arch being dominant most of the times, and LAARSA as the most common aortic arch anomaly overall. However, several reports described a much lower number of patients with a LAARSA.[13-15] This may be explained by the predominantly asymptomatic course of this type of vascular ring, which can let this ring undiagnosed or only found on imaging studies by coincidence.[10,16] In our study, LAARSA was found coincidentally on imaging in 70% of all patients with a LAARSA.

Vascular rings are frequently seen in association with other congenital cardiac anomalies (63.8% in this series, compared with 12% to 63% in previous series[12,14,17,18]), which may dominate the clinical presentation and complicate the diagnosis of a vascular ring.

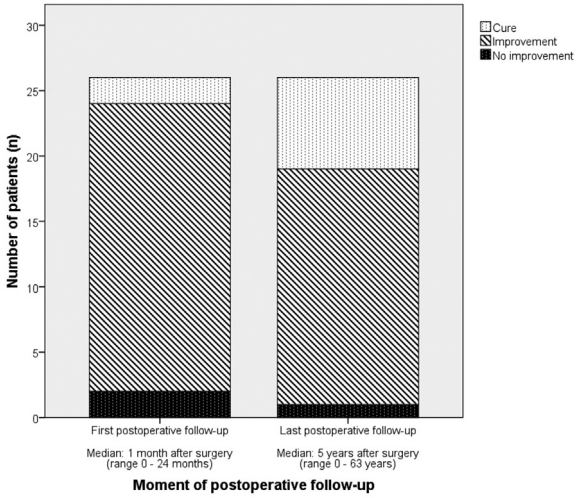

Figure 3. Postoperative outcomes at the first and last time of follow-up.

Presenting symptoms

Symptoms were present in approximately half of the patients, with predominance of patients with a DAA. In one-third of the symptomatic patients, symptoms presented at birth. In line with earlier series,[13,17,19] respiratory symptoms were the most common type of symptoms in this study. Dysphagia for solid foods was the most frequent gastrointestinal symptom and, in line with previous data,[20] most commonly presented in patients with a LAARSA.

In approximately one-third of the patients, comorbidities were found and included mostly asthma/bronchial hyperreactivity, especially in patients with a DAA. However, it is not clear from this data set whether these cases of asthma/bronchial hyperreactivity were true comorbidities or misinterpretations of symptoms of a vascular ring. For example, one of the patients in our study used bronchodilators for over 10 years, which could be terminated after surgery for her vascular ring. Previous reports[7,8] emphasize this risk of misdiagnosing DAA as asthma, which delayed the true diagnosis, delineating the importance of considering vascular rings in the differential diagnosis of patients with respiratory symptoms.

Characteristics according to the age of clinical onset

Previous literature reported respiratory symptoms to be more frequent in infants and children and gastrointestinal

symptoms to be more common in adults.[19] In this study, no significant difference in the number of respiratory and gastrointestinal symptoms between the paediatric and adult groups existed.

More than half of patients with a DAA presented in infancy or childhood. This is consistent with the previous literature,[21] reporting DAA to be less common in adults, because of its tendency to cause severe respiratory symptoms, leading to early diagnosis and correction. In LAARSA, symptoms were present in only one-third of patients with a median age at the onset of symptoms of 23.0 years. These findings agree with data previously reported,[10,16] that is, that LAARSA usually produces no symptoms and is commonly discovered coincidentally on imaging. However, when the aberrant subclavian artery becomes tortuous and dilated in the elderly, late onset of symptoms such as dysphagia due to esophageal compression ("dysphagia lusoria") may emerge.[20]

Remarkably, preoperative spirometry was obtained in less than half of symptomatic patients, suggesting that this test is underused for these (respiratory) symptoms. Spirometry showed a plateau in the expiratory flow-volume curve in more than three-quarters of patients in the paediatric group and a decreased value for PEF, forced expiratory flow after 25% of FVC expired, and forced expiratory flow after 50% of FVC expired, indicating a variable intrathoracic obstruction and thereby suggesting a vascular ring.[22,23] In the adult group, all preoperative values were normal. This may indicate a more severe intrathoracic obstruction due to the vascular ring in the paediatric group, leading to an earlier onset of symptoms. Rather than FEV_1 or FVC, the spirometric values most often examined by clinicians, tracheal obstruction is better recognized looking at PEF in spirometry. Although not specific or sensitive, the presence of a variable intrathoracic obstruction pattern in spirometry should raise suspicion for a vascular ring and prompt further investigation.[22]

Surgery, postoperative outcomes, and follow-up

Surgery was performed in 42.0% of patients at a median age of 17 years, with most surgical repairs in patients with DAA. Those who did not undergo surgery were asymptomatic, inoperable, or had mild symptoms not requiring surgery. Only 3 asymptomatic patients who were not operated eventually developed symptoms, supporting a "wait and see" policy for patients with no symptoms.[10,24] In agreement with earlier literature,[10,16,24] a left thoracotomy was the most performed type of surgery in most types of vascular rings. As described in earlier reports,[13,18,24] surgical correction could be achieved without intra- and postoperative mortalities. According to the literature,[13,14] most patients showed (some) improvement or complete relief of symptoms after surgery at the first follow-up. Relief of symptoms may be hampered immediately because of associated tracheomalacia.[16] At the last postoperative follow-up, almost one-third of patients was considered cured and only 1 patient had still no improvement in symptoms. In some patients, surgery may not resolve all airway symptoms when the extrinsic tracheal compression is associated with an intrinsic weakness of the cartilaginous ring (tracheomalacia). It is an important prognostic factor in the management of patients with a vascular ring because it determines postoperative outcome.[25] In contrast to earlier reports describing a relation between postoperative outcome and the age of surgery,[10,15] no significant correlation was found in the current study. This may be attributed to the small size of the study population.

Limitations

The main limitation of this study is its retrospective design and the fact that the CONCOR database is a survival cohort, only including patients with a congenital cardiac anomaly who survived and followed up until the age of 18 years or more. Patients with a history of a vascular ring who were lost to follow-up before the age of 18 years are therefore missed. Secondly, the study population has a large heterogeneity with different physicians and different surgeons from 7 medical centres.

Conclusions

Vascular rings are relatively rare, congenital vascular anomalies. The most common vascular ring is a LAARSA. This is followed by DAA, in which symptoms are present most frequently and most severely, requiring surgery most often. The low incidence of vascular rings, their anatomic heterogeneity, and the wide range of common respiratory and gastrointestinal symptoms often lead to misdiagnosis as asthma/bronchial hyperreactivity or gastroesophageal reflux disease. In patients with suspected asthma who do not respond to guideline therapy, or in whom the diagnosis is doubtful, tracheal pathology (such as subglottic stenosis or a vascular ring) should therefore be considered. In addition, vascular rings are frequently associated with congenital cardiac anomalies, which may dominate the clinical picture and may complicate the diagnosis of a vascular ring as well. Although not specific, additional lung function assessment may reveal signs of a variable intrathoracic obstruction, raising the suspicion of a vascular ring. Surgical repair is required in patients with symptoms and may lead to improvement in most patients, even at an adult age. Therefore, clinical awareness for this condition is warranted.

Acknowledgement

We thank Amanda Gautier, Scientific Illustrator of Figure 1.

Disclosures

The authors declare that they have no relevant conflicts of interest to disclose.

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