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Service de Cardiologie

"Aortic Valve Dysfunction and Aortic Dilation in Adults with Coarctation of the Aorta"

THESE

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par

Mathieu CLAIR

Médecin diplômé de la Confédération Suisse
Originaire de Bofflens (Vaud)

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Directrice de l'Ecole doctorale	*Madame le Professeur* **Stephanie Clarke**

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Aortic Valve Dysfunction and Aortic Dilation in Adults with Coarctation of the Aorta

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Madame le Professeur Stephanie Clarke
Directrice de l'Ecole doctorale

Résumé:
Dysfonction de la valve aortique et dilatation de l'aorte chez les adultes avec une coarctation de l'aorte

Objectif: Déterminer la prévalence de la dysfonction de la valve aortique, de la dilatation de l'aorte proximale et des interventions au niveau de la valve aortique et de l'aorte ascendante chez les adultes avec une coarctation de l'aorte.

Contexte: La dysfonction de la valve aortique et la dilatation de l'aorte proximale sont rares chez les enfants et les adolescents avec une coarctation de l'aorte. A long terme, les adultes pourraient être plus à risque de développer ce type de pathologie.

Méthode: Nous avons rétrospectivement passé en revue tous les adultes avec une coarctation de l'aorte corrigée ou pas suivis au « Boston Children's Hospital » entre 2004 et 2010.

Résultats: 216 adultes (56 % d'hommes) avec une coarctation ont été identifiés. L'âge médian à la dernière évaluation était de 28 (de 18 à 75) ans. Une bicuspidie aortique était présente dans 66% des cas. Au dernier contrôle, 3% avaient une sténose aortique modérée ou sévère et 4% avaient une insuffisance aortique modérée à sévère. Une dilatation de la racine de l'aorte ou de l'aorte ascendante était présente dans 28%, respectivement 42% des patients. Une dilatation au moins modérée de la racine de l'aorte ou de l'aorte ascendante (score $Z > 4$) était présente dans 8%, respectivement 14%. Les patients avec une bicuspidie aortique étaient plus sujets à avoir une dilatation au moins modérée de la racine de l'aorte ou de l'aorte ascendante comparés à ceux sans bicuspidie (20% contre 0%; $p<0.001$). L'âge était associé à une dilatation de l'aorte ascendante ($p=0.04$). Au dernier suivi, 6% avait nécessité une intervention au niveau de la valve aortique et 3% un remplacement de la racine de l'aorte ou de l'aorte ascendante.

Conclusion: Chez les adultes avec une coarctation de l'aorte, une dysfonction significative de la valve aortique et des interventions sur la valve ou l'aorte proximale étaient peu fréquentes. Cependant, la dilatation de l'aorte était prévalente, en particulier au niveau de l'aorte ascendante chez les patients avec une bicuspidie aortique.

ORIGINAL ARTICLE

Aortic Valve Dysfunction and Aortic Dilation in Adults with Coarctation of the Aorta

Mathieu Clair, MD,* Susan M. Fernandes, LPD, PA-C,* Paul Khairy, PhD, MD,* Dionne A. Graham, PhD,* Eric V. Krieger, MD,* Alexander R. Opotowsky, MD, MPH,*† Michael N. Singh, MD,*† Steven D. Colan, MD,* Erik J. Meijboom, PhD, MD,‡ and Michael J. Landzberg, MD*†

*Department of Cardiology, Boston Children's Hospital and †Cardiovascular Division, Department of Medicine, Brigham and Women's Hospital, Harvard Medical School, Boston, Mass, USA; ‡Pediatric Cardiology Unit, Department of Pediatrics, University Hospital, Lausanne, Switzerland

ABSTRACT

Objectives. To determine the prevalence of aortic valve dysfunction, aortic dilation, and aortic valve and ascending aortic intervention in adults with coarctation of the aorta (CoA).

Background. Aortic valve dysfunction and aortic dilation are rare among children and adolescents with CoA. With longer follow-up, adults may be more likely to have progressive disease.

Methods. We retrospectively reviewed all adults with CoA, repaired or unrepaired, seen at our center between 2004 and 2010.

Results. Two hundred sixteen adults (56.0% male) with CoA were identified. Median age at last evaluation was 28.3 (range 18.0 to 75.3) years. Bicuspid aortic valve (BAV) was present in 65.7%. At last follow-up, 3.2% had moderate or severe aortic stenosis, and 3.7% had moderate or severe aortic regurgitation. Dilation of the aortic root or ascending aorta was present in 28.0% and 41.6% of patients, respectively. Moderate or severe aortic root or ascending aortic dilation (z-score > 4) was present in 8.2% and 13.7%, respectively. Patients with BAV were more likely to have moderate or severe ascending aortic dilation compared with those without BAV (19.5% vs. 0%; $P < 0.001$). Age was associated with ascending aortic dilation ($P = 0.04$). At most recent follow-up, 5.6% had undergone aortic valve intervention, and 3.2% had aortic root or ascending aortic replacement.

Conclusion. In adults with CoA, significant aortic valve dysfunction and interventions during early adulthood were uncommon. However, aortic dilation was prevalent, especially of the ascending aorta, in patients with BAV.

Key Words. Coarctation of the Aorta; Bicuspid Aortic Valve; Aortic Dilation; Aortic Valve Dysfunction

Introduction

Coarctation of the aorta (CoA) is among the most common congenital cardiovascular defects, affecting 1 of 3000 live births.[1,2] Long-term survivors are at risk for aortic and mitral valve dysfunction, aortic aneurysm, aortic dissection, endocarditis, systemic hypertension, heart failure, and sudden death.[3,4] Bicuspid aortic valve (BAV) is present in approximately two thirds of patients with CoA[5,6] and is associated with increased risk for valvular dysfunction and aortic dilation.[3,7,8] Prior reports, however, suggest that children and adolescents with BAV and CoA are less likely to have aortic valve dysfunction and aortic dilation compared with those with isolated BAV[6,9] and consequently are less likely to require valvular intervention or ascending aortic replacement.[10] As patients age, risks for developing valvular dysfunction and aortic dilation accrue. Therefore, we sought to determine the prevalence of aortic valve dysfunction, aortic dilation, and occurrence of valvular intervention or ascending aortic intervention in a population of adults with CoA.

Methods

Clinical databases in the cardiovascular program at Boston Children's Hospital were retrospectively searched to identify adults with repaired or unrepaired CoA ≥18 years of age seen in the outpatient cardiology clinic between 2004 and 2010. We excluded patients with concomitant moderate or complex forms of congenital heart disease,[11] Turner syndrome, 22q11.2 deletion syndrome, Marfan syndrome, and related disorders such as Loeys–Dietz syndrome and Ehlers–Danlos syndrome, given their recognized association with aortic pathology.[12–15] Patients with unicommissural valve, defined as a complete or partial fusion of more than 2 leaflets, were excluded, as were patients with other lesions that could potentially alter the aortic valve, including subaortic stenosis[16] and infundibular, perimembranous, or outlet muscular ventricular septal defects.[17]

Bicuspid aortic valve was defined as partial or complete fusion of 2 aortic valve leaflets, with or without a central raphe, resulting in partial or complete absence of that functional commissure.[18] Systemic hypertension was considered present if the resting systolic blood pressure was >140 mm Hg or the diastolic blood pressure >90 mm Hg at most recent clinic visit or if the patient was being treated with antihypertensive medication. The definition of CoA or recoarctation was based on conventional criteria, including documented repair, upper-to-lower-limb blood pressure gradient >20 mm Hg, and/or evidence of coarctation on imaging.[19,20]

The most recent echocardiogram (or the exam prior to any surgical or catheter intervention or endocarditis) was systematically reviewed. Echocardiographic assessment of aortic stenosis was expressed as the maximum instantaneous Doppler gradient (mm Hg) in the context of normal left ventricular contractile function. Criteria for grading aortic stenosis (AS) and aortic regurgitation (AR) were taken from prior study.[6] Aortic stenosis was considered moderate or severe if the maximum instantaneous Doppler gradient was ≥50 mm Hg. Classification of the severity of AR was based on composite evaluation of proximal jet width, left ventricular end-diastolic volume or dimension, and abdominal aortic Doppler flow pattern. Designation of moderate or severe regurgitation required pandiastolic retrograde flow in the descending aorta and left ventricular end-diastolic volume or dimension >2 standard deviations above the mean, indexed to body surface area. Gradient across the CoA (or arch obstruction) was assessed by blood pressure differential. Aortic measurements were performed in parasternal long axis views using two-dimensional imaging, inner edge to inner edge, at maximum (mid-systolic) excursion. The aortic root was measured at largest dimension of the sinuses of Valsalva, and the ascending aorta was measured at the location of maximum diameter distal to the sinotubular junction. Aortic dimensions were adjusted for body surface area by dividing these dimensions by the square root of body surface area and were expressed as z-scores.[21] Body surface area was calculated using the formula by Haycock et al.[22] All echocardiograms with aortic dimensions missing from formal reports were reviewed by a single investigator (ARO). Mild aortic dilation was defined as z-score >2 and ≤4, and moderate or severe dilation as z-score >4. Additional clinical data regarding catheter and surgical interventions and diagnosis of endocarditis were obtained from medical records and hospital databases. The study protocol was approved by the Institutional Review Board at Boston Children's Hospital.

Statistical Analysis

Continuous data are presented as median (range) and were compared between two groups with Wilcoxon rank-sum tests. Categorical data are presented as counts and percentages, and rates were compared with chi-square tests and Fisher's exact tests. Univariate and multivariate logistic regression models were constructed to identify factors associated with aortic valve pathology. Candidate variables included valve morphology, age, sex, and history of hypertension. Logistic regression was used to identify factors associated with aortic root dilation and ascending aortic dilation, with the addition of clinically significant AR, clinically significant AS, and residual arch obstruction to the list of candidate variables. Two-tailed P values < 0.05 were considered statistically significant. Results are expressed as a fraction of available data. Data analysis was performed with SAS software Version 9.2 (SAS Institute, Cary, NC, USA).

Results

A total of 216 adults (56.0% male) with CoA were identified. Characteristics of the study population are provided in Table 1. Median age at last evaluation was 28.3 years (range 18.0 to 75.3). BAV was present in the majority (65.7%). In 20 (14.1%)

Table 1. Baseline Characteristics of Patients with Coarctation of the Aorta

	Entire Cohort		BAV		No BAV		
	Cohort Size n		Cohort Size n		Cohort Size n		*P* value (BAV vs. no BAV)
Demographics	216		142	142	74	74	
Age in years, median (range)		28.3 (18.0–75.3)		27.4 (18.0–71.6)		30.2 (18.0–75.3)	.46
Male gender, n (%)		121 (56.0)		83 (58.5)		38 (51.4)	.32
BAV morphology			142				
R-L fusion				107 (75.4)			
R-N fusion				14 (9.9)			
L-N fusion				1 (0.7)			
Unknown morphology				20 (14.1)			
Intervention, n (%)	216		142	142	74	74	
Any surgical or catheter valve intervention		12 (5.6)		12 (8.5)		0 (0)	.01
Aortic valve catheter intervention		4 (1.9)		4 (2.8)		0 (0)	.30
Aortic valve surgical intervention		11 (5.1)		11 (7.8)		0 (0)	.02
Aortic root or ascending aorta intervention		7 (3.2)		7 (4.9)		0 (0)	.10
Clinical, n (%)							
Hypertension	214	107 (50.0)	140	64 (45.7)	74	43 (58.1)	.08
Prior coarctation repair	216	209 (96.8)	142	138 (97.2)	74	71 (96.0)	.69
Arch obstruction	193	14 (7.3)	125	9 (7.2)	68	5 (7.4)	1.0
On antihypertensive treatment	210	91 (43.3)	138	55 (39.9)	72	36 (50)	.16
ARB/ACEI		57 (27.1)		33 (23.9)		24 (33.3)	.15
β-blockers		53 (25.2)		33 (23.9)		20 (27.8)	.54
Diuretics		13 (6.2)		7 (5.1)		6 (8.3)	.38
Calcium channel blocker		10 (4.8)		8 (5.8)		2 (2.8)	.50
α-blockers		3 (1.4)		1 (0.7)		2 (2.8)	.27

ACEI, angiotensin converting enzyme inhibitors; ARB, angiotensin II receptor blockers; BAV, bicuspid aortic valve; L-N, left and noncoronary leaflet; R-L, right and left coronary leaflet; R-N, right coronary and noncoronary leaflet.

patients with BAV, the subtype could not be ascertained; in all others, fusion of right and left coronary leaflets (R-L commissural fusion) was most common.

Aortic Valve Dysfunction

Echocardiographic data were available for 189 (87.5%) patients. Aortic valve dysfunction was present in 74/189 (39.2%). AS was present in 39/189 (20.6%), and 6/189 (3.2%) patients had moderate or severe AS. AS was significantly more frequent in patients with BAV (31.5% vs. 0.0%, $P < .0001$) compared with patients with a tricuspid aortic valve. Moderate or severe AS was uncommon and present in 4.8% of patients with BAV and 0.0% of those without BAV ($P = .10$). As depicted in Figure 1A and in Table 2, AS was present in 23.2% of patients with R-L commissural fusion compared with 66.7% with fusion of the commissure between the right coronary and noncoronary leaflets (R-N commissural fusion) ($P = .004$).

Aortic regurgitation was present in 53/189 (28.0%) patients and moderate or severe AR in 7/189 (3.7%) patients. AR was more prevalent in patients with BAV (37.1% vs. 10.8%, $P < .0001$). Moderate or severe AR was uncommon, present in 4.8% of patients with BAV and 1.5% of those without ($P = .43$). The proportion of patients with AR was similar in those with R-L commissural fusion compared with those with R-N commissural fusion (38.4% vs. 33.3%, $P = 1.0$). Male sex was associated with a higher prevalence of aortic regurgitation ($P = .02$).

Older age, arch obstruction, and hypertension were not associated with aortic valve dysfunction.

Aortic Root and Ascending Aortic Dilation

Aortic dilation was present in 82/183 (44.8%) patients. Moderate or severe aortic dilation was present in 31/183 (16.9%) patients. 24.4% of patients with BAV had moderate or severe dilation compared with 3.1% of patients without BAV ($P < .001$).

Aortic root dilation was present in 51/182 (28.0%) patients, 15/182 (8.2%) of whom had moderate or severe dilation. The presence of BAV ($P < .001$) was associated with aortic root dilation. There was no statistically significant difference in prevalence of moderate or severe aortic root dilation between patients with and without BAV ($P = .09$, Table 3). Aortic root dilation was more prevalent in patients with AR ($P = .02$), as well as in patients with R-L commissural fusion,

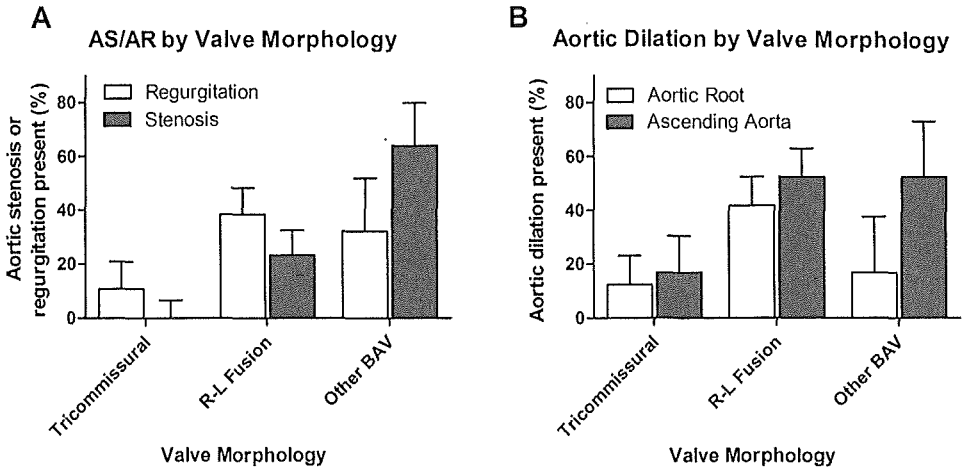

Figure 1. (A) Proportion of patients with aortic stenosis or aortic regurgitation by valve morphology. (B) Proportion of patients with aortic root or ascending aortic dilation by valve morphology. Aortic dilation is defined as a *z*-score of >2. Error bars represent the 95% confidence interval. See Tables 2 and 3 for statistical comparison and multivariable analysis. AR, aortic regurgitation; AS, aortic stenosis; BAV, bicuspid aortic valve; R-L, right and left coronary leaflet.

Table 2. Factors Associated with Valve Dysfunction

	n	Valve Dysfunction, n (%)	Univariate		Multivariate	
			Odds Ratio (95% CI)	*P*	Odds Ratio (95% CI)	*P*
Aortic stenosis						
Valve morphology						
Tricommissural	65	0 (0)	Reference	<.0001	Reference	<.0001
R-L fusion	99	23 (23.2)	27.3 (4.6, ∞)*		24.3 (4.1, ∞)*	
Other BAV	25	16 (64.0)	142.1 (21.6, ∞)*		115.6 (17.6, ∞)*	
Age (per 5 years increase)			1.02 (0.9, 1.2)	.76		
Gender						
Male	103	21 (20.4)	Reference	.93		
Female	86	18 (20.9)	1.03 (0.5, 2.1)			
Systemic hypertension						
No	97	20 (20.6)	Reference	.96		
Yes	91	19 (20.9)	1.02 (0.5, 2.1)			
Arch obstruction						
No	160	27 (16.9)	Reference	.89		
Yes	13	2 (15.4)	0.9 (0.2, 4.3)			
Aortic regurgitation						
Valve morphology						
Tricommissural	65	7 (10.8)	Reference	.001	Reference	.002
R-L fusion	99	38 (38.4)	5.2 (2.1, 12.5)		5.1 (2.1, 12.4)	
Other BAV	25	8 (32.0)	3.9 (1.2, 12.3)		3.7 (1.2, 11.8)	
Age (per 5 years increase)			1.0 (0.9, 1.1)	.97		
Gender						
Male	103	36 (35.0)	Reference	.02	Reference	.03
Female	86	17 (19.8)	0.5 (0.2, 0.9)		0.5 (0.2, 0.9)	
Systemic hypertension						
No	97	25 (25.8)	Reference	.55		
Yes	91	27 (29.7)	1.2 (0.64, 2.3)			
Arch obstruction						
No	160	43 (26.9)	Reference	.15		
Yes	13	6 (46.2)	2.3 (0.7, 7.3)			

*Estimated using exact logistic regression.
BAV, bicuspid aortic valve; R-L fusion, right and left coronary leaflet fusion; ∞, infinity.

compared with those with other types of BAV ($P < .001$, Figure 1B).

Ascending aortic dilation was present in 67/161 (41.6%) patients, with 22/161 (13.7%) having moderate or severe dilation. Older age ($P = .001$) and presence of BAV ($P < .0001$) were significantly associated with ascending aortic dilation (Table 4). The presence of BAV was also associated with at

Table 3. Factors Associated with Aortic Root Dilation (*z*-score > 2)

	n	Dilated n (%)	Univariate Odds Ratio (95% CI)	*P*	Multivariate Odds Ratio (95% CI)	*P*
Valve morphology						
Tricommissural	64	8 (12.5)	Reference	<.001	Reference	<.001
R-L fusion	94	39 (41.5)	5.0 (2.1, 11.6)		5.0 (2.1, 11.6)	
Other BAV	24	4 (16.7)	1.4 (0.4, 5.2)		1.4 (0.4, 5.2)	
Age (per 5 years increase)			1.1 (1.0, 1.3)	.08		
Gender						
Male	99	31 (31.3)	Reference	.28		
Female	83	20 (24.1)	0.70 (0.4, 1.3)			
Aortic stenosis						
No	145	40 (27.6)	Reference	.80		
Yes	37	11 (29.7)	1.1 (0.5, 2.5)			
None or mild	176	49 (27.8)	Reference	1.00		
Moderate or severe	6	2 (33.3)	1.3 (0.1, 9.4)*			
Aortic regurgitation						
No	131	30 (22.9)	Reference	.02		
Yes	51	21 (41.2)	2.4 (1.2, 4.7)			
None or mild	175	47 (26.9)	Reference	.10		
Moderate or severe	7	4 (57.1)	3.6 (0.8, 16.8)			
Systemic hypertension						
No	95	21 (22.1)	Reference	.06		
Yes	86	30 (34.9)	1.9 (1.0, 3.6)			
Arch obstruction						
No	155	41 (26.5)	Reference	.26		
Yes	12	5 (41.7)	2.0 (0.6, 6.6)			

*Estimated using exact logistic regression.
BAV, bicuspid aortic valve; R-L fusion, right and left coronary leaflet fusion.

Table 4. Factors Associated with Ascending Aorta Dilation (*z*-score > 2)

	n	Dilated n (%)	Univariate Odds Ratio (95% CI)	*P*	Multivariate Odds Ratio (95% CI)	*P*
Valve morphology						
Tricommissural	48	8 (16.7)	Reference	<.001	Reference	<.0001
R-L fusion	90	47 (52.2)	5.5 (2.3, 13.0)		9.3 (3.4, 25.6)	
Other BAV	23	12 (52.2)	5.5 (1.8, 16.6)		8.7 (2.4, 30.7)	
Age (per 5 years increase)			1.3 (1.1, 1.5)	.001	1.4 (1.2, 1.7)	<.0001
Gender						
Male	90	33 (36.7)	Reference	.15		
Female	71	34 (47.9)	1.6 (0.8, 3.0)			
Aortic stenosis						
No	125	47 (37.6)	Reference	.06		
Yes	36	20 (55.6)	2.1 (1.0, 4.4)			
None or mild	155	63 (40.7)	Reference	.39		
Moderate or severe	6	4 (66.7)	2.9 (0.4, 33.0)*			
Aortic regurgitation						
No	111	43 (38.7)	Reference	.27		
Yes	50	24 (48.0)	1.5 (0.7, 2.9)			
None or mild	154	65 (42.2)	Reference	.48		
Moderate or severe	7	2 (28.6)	0.5 (0.1, 2.9)			
Systemic hypertension						
No	85	36 (42.4)	Reference	.90		
Yes	75	31 (41.3)	1.0 (0.5, 1.8)			
Arch obstruction						
No	136	53 (39.0)	Reference	.46		
Yes	12	6 (50.0)	1.6 (0.5, 5.1)			

*Estimated using exact logistic regression.
BAV, bicuspid aortic valve; R-L fusion, right and left coronary leaflet fusion.

least moderate dilation of the ascending aorta (19.5% vs. 0%; $P < .001$).

Arch obstruction and hypertension were not associated with aortic root and/or ascending aortic dilation.

Interventions

Twelve (5.6%) patients had a total of 22 aortic valve interventions; six underwent more than 2 interventions. Occurrence of aortic valve intervention was associated with BAV ($P = .01$). Bicuspid aortic valve with R-N commissural fusion was more likely to require intervention compared with BAV with R-L commissural fusion ($P = .002$). Four patients had balloon valve dilation as a first intervention, and all but one patient had a surgical valve intervention. The indication for initial valve intervention (catheter-based or surgical) was AS in 8 patients and AR in 4, including 1 with concomitant ascending aorta replacement. Endocarditis precipitated 3 operations. In addition, 7 patients had ascending aorta or root intervention; all presented with aortic dilation at time of surgery. First valve intervention occurred in 2 patients during the first year of life, in early childhood (2.5 and 4.4 years old) in 2, between ages 11 and 15 years in 3, and after age 18 years in the remaining 5.

Discussion

Progressive aortic valve dysfunction and aortic dilation in patients with CoA place patients at increased risk for morbidity and mortality.[3] The results of our study extend understanding of the disease course, with particular application to younger adults, given the population captured in this cohort. The key findings of our study are as follows: (1) mild valve dysfunction is frequent, but significant aortic valve dysfunction is uncommon, in a selected population of young adults with CoA; (2) aortic dilation is prevalent, especially of the ascending aorta, in patients with a BAV; (3) prevalence of valvular or aortic pathology appears to follow recognized pediatric patterns of association with type of BAV commissural fusion; and (4) aortic valve or ascending aortic interventions are uncommon but do occur during adolescence and young adulthood.

Aortic Valve Dysfunction

Most young adults with CoA remain free of significant valvular dysfunction. Although mild AS and AR were prevalent in our patients with BAV, moderate or severe aortic valve dysfunction was rare. In contradistinction, Toro-Salazar et al. reported that in long-term follow up >50 years after CoA repair, valvular and aortic pathologies were common; in their cohort, 10/92 subjects had moderate or severe aortic valve dysfunction.[23] Given the relatively young age of our cohort, that some degree of aortic valve dysfunction was common, and that increased prevalence of aortic valve dysfunction has been associated with aging,[24–27] continued cardiac surveillance appears appropriate in this population. These data, however, cannot determine, but may help refine, recommendations for appropriateness, frequency, and nature of long-term monitoring.

Our findings on valve morphology are consistent with the premise that R-L commissural fusion and R-N commissural fusion represent different disease entities with distinct developmental causes.[6,28–30] We demonstrate that two thirds of patients with CoA have a BAV and that the majority of BAVs are of the R-L commissural fusion type. Moreover, we show that R-N commissural fusion was associated with greater valve dysfunction and was more likely to compel valve intervention compared with R-L commissural fusion. Also, there was a higher prevalence of aortic root dilation in patients with an R-L fusion pattern. These results lend further credence to the concept that different phenotypic patterns are associated with the two common BAV morphologies.

Aortic Root and Ascending Aortic Dilation

Even after successful repair of CoA, patients may present with evidence of vascular dysfunction[31] and abnormal vascular structure.[32] Our findings of a larger aortic root and ascending aorta in patients with associated BAVs are consistent with prior reports suggesting that BAV is a predictor of aortic wall complications in subjects with CoA.[28,33] The association between systemic arterial hypertension and aortic complications, derived largely from study of older subjects with greater degree of atherosclerosis, remains controversial[28,34] and is not supported by our data, obtained largely from a current and younger adult population. Arch obstruction, associated with increased wall stress in the proximal aorta, was also not found in our analysis to impact aortic size. In part, these results lend support to the paradigm that aortic dilation is not exclusively determined by hemodynamic parameters and that intrinsic anomalies of the aortic wall may be important in pathologic development for patients with CoA and BAV.[35,36] Importantly, despite increasingly characterized

disorders of vascular connective tissue involving loss of elastic tissue in both of these congenital pathologies,[37] clinical consequences appear somewhat discordant. We and others[8] have found that the largest aortas may be present in patients with BAVs alone, and the smallest may be present in patients with solitary CoA. Further studies are required to better elucidate pathophysiological mechanisms associated with aortic dilation and its complications in these settings.

In patients with both tricuspid aortic valve and BAV, age correlates with cystic medial necrosis, fragmentation of elastic fibers, and aortic dilation.[8,34,38] Indeed, in our cohort with CoA, older age was associated with increased ascending aortic size. Given the large number of young patients with CoA and BAV with milder degrees of aortic dilation, the potential for further dilation with advancing age is concerning.

Interventions

Valve interventions were uncommon. Of patients who did have an intervention, just under half had their first valve intervention in adulthood. We have previously reported that congenital severely dysfunctional valves tend to be intervened on in childhood.[10] In adulthood, we were more likely to encounter consequences of degenerative changes. The process may be accelerated by interactions with cardiovascular risk factors that increase the risk of developing worsening of AS and have prompted calls for increased vigilance for and treatment of such risks.[24] Notably, guidelines and indication for intervention on both BAV and CoA in younger and middle-aged adults remain relatively free of data that support optimal timing for such; assessment of comparative outcomes with or without intervention for these patients is welcomed.

While only 7 patients in our young-adult cohort underwent a surgical repair of aortic dilation, intervention was more common in older patients. The need for intervention may be anticipated to increase as this population ages and as recommendations for surgical intervention progress to lower aortic diameter.[28,39–42]

Given the lack of longitudinal data for this population we recognize the need for a prospective study of pathology and outcomes in adults with CoA, with or without BAV. Most recent data suggest that this population remains at increased risk for other comorbidities such as systemic hypertension, heart failure, and coronary artery disease.[43,44] It therefore appears prudent to remain vigilant, promoting lifelong cardiovascular surveillance and optimal health practices in accordance with management guidelines.[43,44] However, our results suggest that in general, patients with mild aortic dilation require less frequent follow-up; this appears particularly applicable to those with concomitant tricuspid aortic valve.

Limitations

Our study represents a single-center retrospective experience in a largely younger adult population. There is a referral bias, as patients with more severe disease or symptoms tend to be referred to regional centers. Aortic dimensions were unavailable in 33 (15.3%) patients; analysis failed to identify differences between baseline characteristics in subjects with and without data on aortic dimensions, suggesting limited impact on our findings.

Conclusion

In this population of young adults with CoA, significant aortic valve dysfunction and aortic valve or ascending aortic interventions were uncommon, though some degree of aortic dilation and valve dysfunction was relatively frequent. As adults with CoA acquire comorbidities with aging, they remain vulnerable to progressive valvular dysfunction and aortic dilation, especially in association with BAV. Recommendation for sustained cardiovascular surveillance guided by specialists in BAV and aortic disease care appears prudent.[43] Longitudinal study and analysis of valve and aortic pathology in adults with CoA are needed to improve guidelines for disease-specific care and follow-up.

Contributorship

Mathieu Clair collected data, interpreted the data, drafted the manuscript, and gave final approval of the version to be published.

Susan M. Fernandes and Michael J. Landzberg conceived and designed the study, interpreted the data, helped draft the manuscript, and gave final approval of the version to be published.

Alexander R. Opotowsky collected data, interpreted the data, revised the manuscript critically for important intellectual content, and gave final approval of the version to be published.

Dionne A. Graham analyzed the data, revised the manuscript critically for important intellectual content, and gave final approval of the version to be published.

Steven D. Colan interpreted the data, revised the manuscript critically for important intellectual content, and gave final approval of the version to be published.

Paul Khairy, Eric V. Krieger, Michael N. Singh, and Erik J. Meijboom revised the manuscript critically for important intellectual content and gave final approval of the version to be published.

Corresponding Author: Mathieu Clair, MD, Cardiology, University Hospital, Bugnon 46, 1011 Lausanne, Switzerland. Tel: (+41) 79 556-59-21; Fax: (+41) 21 314-00-13; E-mail: m.clair@me.com

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Rapport de synthèse:
Dysfonction de la valve aortique et dilatation de l'aorte chez les adultes avec une coarctation de l'aorte

La coarctation de l'aorte est parmi les malformations congénitales cardiovasculaires les plus fréquentes, affectant un nouveau-né sur 3000. Les survivants à long terme sont à risque de dysfonction valvulaire mitrale et aortique, d'anévrisme et de dissection aortique, d'endocardite, d'hypertension artérielle, d'insuffisance cardiaque et de mort subite. La bicuspidie aortique est présente dans deux tiers des patients avec une coarctation de l'aorte et est associée à un risque augmenté de dysfonction valvulaire aortique et dilatation de l'aorte. Cependant des études ont montré que dans l'enfance et l'adolescence les patients porteurs d'une bicuspidie aortique et d'une coarctation de l'aorte étaient moins sujets à une dysfonction de la valve aortique ou une dilatation de l'aorte proximale que ceux porteurs uniquement d'une bicuspidie. En conséquence, ces patients étaient également moins à risque de nécessiter une intervention sur la valve aortique ou un remplacement de l'aorte ascendante. Ce groupe de patients devenant plus âgés, le risque de développer une dysfonction valvulaire et une dilatation de l'aorte augmente. Nous avons donc cherché à déterminer la prévalence de la dysfonction de la valve aortique, de la dilatation de l'aorte proximale et des interventions au niveau de la valve aortique et de l'aorte ascendante chez les adultes avec une coarctation de l'aorte.

Notre étude étend la compréhension de l'évolution de la maladie, en particulier chez les jeunes adultes au vu de la population étudiée dans cette cohorte. Les trois principales conclusions de l'étude sont : 1) Un dysfonction légère de la valve aortique est fréquente mais une dysfonction importante est rare dans cette population sélectionnée de jeunes adultes. 2) La dilatation aortique est prévalente, en particulier au niveau de l'aorte ascendante chez les patients avec une bicuspide. 3) La prévalence de pathologies aortique et valvulaire semble suivre des modèles

reconnus, dans la population pédiatrique, en fonction du type de fusion commissurale. 4) les interventions sont peu fréquentes, durant l'adolescence et l'âge adulte.

Etant donné l'absence de données longitudinales pour cette population, nous reconnaissons le nécessité d'études prospectives sur le devenir des adultes avec une coarctation avec ou sans bicuspidie aortique. Les données le plus récentes suggèrent que cette population est à risque pour d'autres comorbidités telles que l'hypertension artérielle, l'insuffisance cardiaque et la maladie coronarienne. Il apparait prudent de rester vigilant, de promouvoir le suivi cardiovasculaire à vie et d'optimiser la prise en charge en accord avec les recommandations établies par les sociétés savantes. Cependant, nos résultats suggèrent qu'en général, les patients avec une légère dilatation de l'aorte ascendante nécessitent un suivi moins fréquent ; ceci est particulièrement vrai pour ceux porteur d'une valve aortique tricuspide.

En conclusion, dans cette population de jeunes adultes avec une coarctation de l'aorte, une dysfonction significative de la valve aortique et des interventions de la valve aortique ou l'aorte ascendante étaient rare. Cependant, un certain degré de dilatation aortique et de dysfonction valvulaire aortique étaient relativement fréquent. Etant donné que les adultes avec une coarctation de l'aorte vont développer des comorbidités avec l'âge, il reste vulnérable à une progression de la dysfonction valvulaire et à une dilatation de l'aorte, en particulier ceux avec une bicuspidie aortique. Recommander une surveillance cardiovasculaire guidée par un spécialiste dans la prise en charge des cardiopathies congénitales chez ces adultes paraît prudents. Des études longitudinales sont nécessaires pour améliorer les directives de prises en charge et de suivi spécifique à cette pathologie.