Diagnostic tools in the follow-up and monitoring of congenital heart disease and pulmonary hypertension

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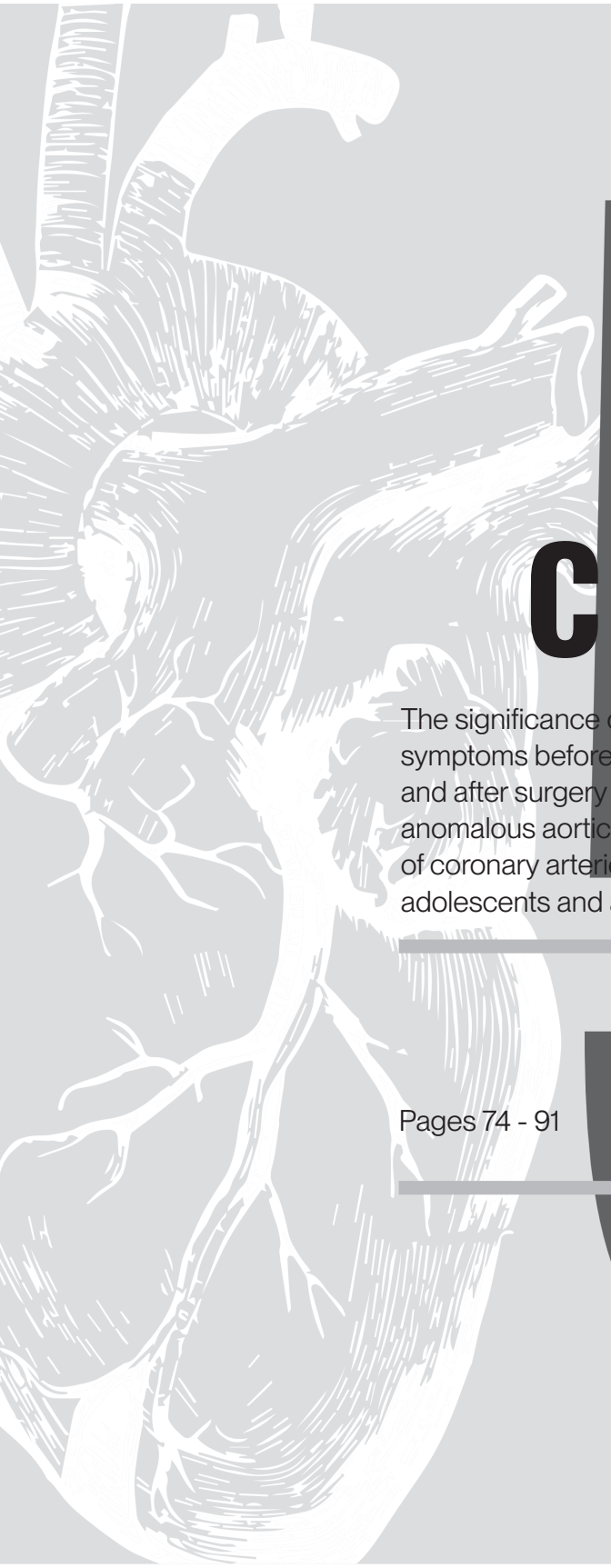

CHAPTER 5

The significance of symptoms before and after surgery for anomalous aortic origin of coronary arteries in adolescents and adults

Pages 74 - 91

THE SIGNIFICANCE OF SYMPTOMS BEFORE AND AFTER SURGERY FOR ANOMALOUS AORTIC ORIGIN OF CORONARY ARTERIES IN ADOLESCENTS AND ADULTS

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ABSTRACT

Objectives

The aim of this study is to describe the significance of symptoms preoperatively and at medium-term follow up in adolescent and adult patients who underwent surgery of AAOCA.

Methods

Consecutive patients who underwent surgery for AAOCA in our tertiary referral center between 2001 to 2018 were included. Clinical characteristics and symptoms were evaluated and medium-term outcomes were recorded. Symptoms were classified according to the '2019 ESC guidelines on chronic coronary syndromes'.

Results

A total of 53 (55% male) patients with mean age of 44 at time of surgery underwent surgical repair of AAOCA. Data on symptoms and events more than 3 months after surgery were available in 34 patients with a median follow up of 3 years (IQR 1.0 – 5.3). Pre-operatively, only 35% patients had typical anginal complaints. After surgical correction of AAOCA, 59% of the patients were free of symptoms, compared to 6% pre-operatively ($p < 0.001$). A total of 3 (9%) patients needed a re- operation/re-intervention related to the operated AAOCA. All 3 patients presented post-operatively with novel typical anginal complaints.

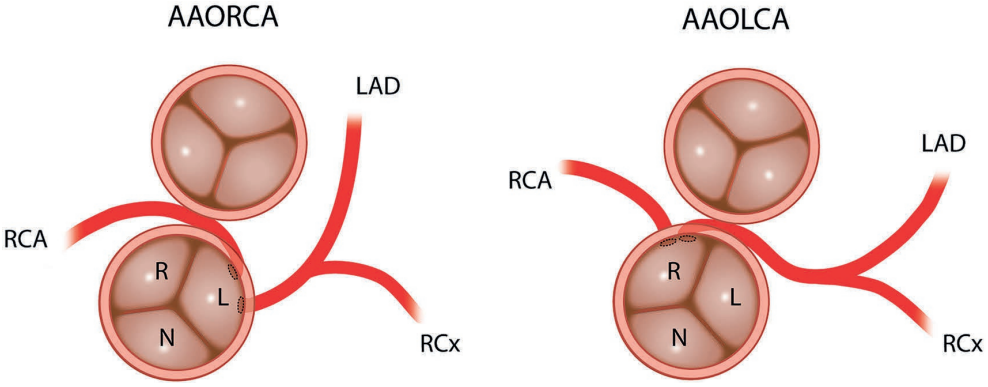
Conclusion

Adolescent and adult patients with AAOCA present with varying symptoms. Only 35% have typical anginal complaints. Surgical correction of AAOCA reduces the symptoms in the vast majority of patients. One should be aware of potential lesions of the operated coronary artery in patients presenting with typical anginal complaints post-operatively.

Introduction

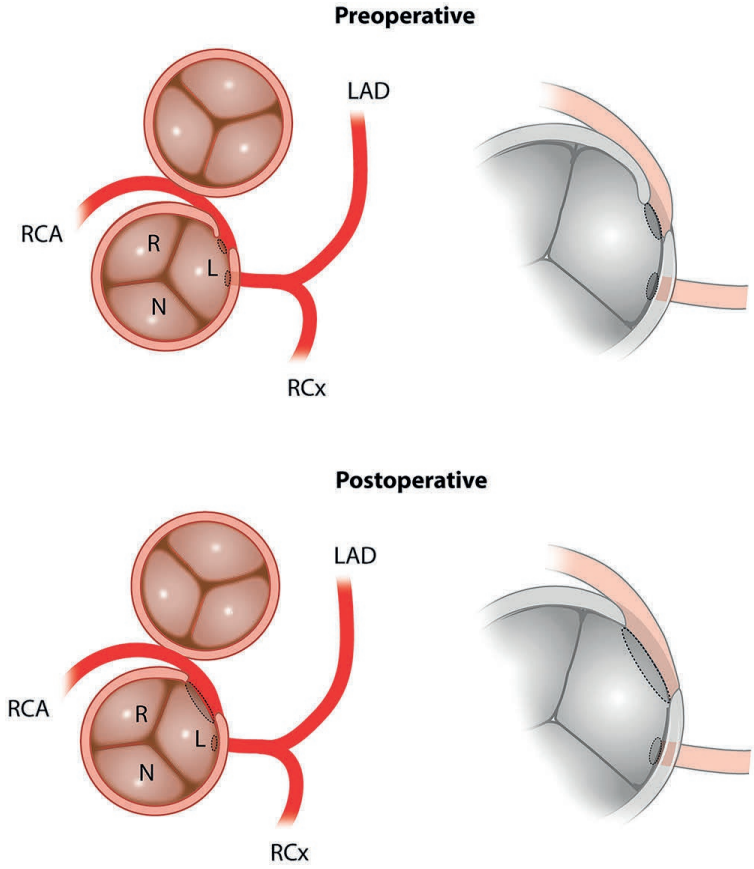
Anomalous aortic origin of the coronary arteries (AAOCA) is a rare congenital condition with a reported incidence between 0.26 and 1.3%, [1-3]. Anomalous coronary arteries which arise from the opposite sinus of Valsalva or contralateral coronary artery are a potential cause of sudden cardiac death, especially in athletes and active young adults (Figure 1) [11]. Presenting symptoms differ largely amongst patients [2,4,52]. To date there is no consensus on indications for surgery versus conservative treatment, especially in middle aged and older patients. Due to lack of long-term follow-up of patients after surgical treatment, indications for surgical treatment are ambiguous, especially in asymptomatic patients [6-9]. The main objective of surgery is to reduce the risk of sudden cardiac death (SCD) and alleviate ischemia. The decision to operate a patient is based on the ostial anatomy and course of the anomalous coronary artery and demonstrated ischemia. The role of symptoms in decision making with regards to the surgical intervention and post-operative outcomes is ambiguous. Several surgical techniques for correcting AAOCA have been used, most commonly unroofing of the intramural segment (Figure 2), coronary reimplantation and

Figure 1. Schematic representation of AAORCA and AAOLCA anatomy with an interarterial and intramural course of the anomalous artery (imaging view).



AAORCA, anomalous aortic origin of a right coronary; AAOLCA, anomalous aortic origin of a left coronary; RCA, right coronary artery; LAD, left anterior descending artery; RCx, ramus circumflex artery; R, right coronary cusp; L, left coronary cusp; N, non coronary cusp

Figure 2. Schematic representation of AAORCA anatomy prior to and after surgical correction (imaging view)



AAORCA, anomalous aortic origin of a right coronary; RCA, right coronary artery; LAD, left anterior descending artery; RCx, ramus circumflex artery; R, right coronary cusp; L, left coronary cusp; N, non coronary cusp

coronary artery bypass grafting (CABG) [10, 11]. A few studies have reported persistent symptoms, restenosis of the operated anomaly after surgery, ischemia and even cases of SCD [9, 12-15].

The aim of this study is to describe the significance of symptoms preoperatively and at medium-term follow up in adolescent/ adult patients who underwent surgery of AAOCA.

Material and methods

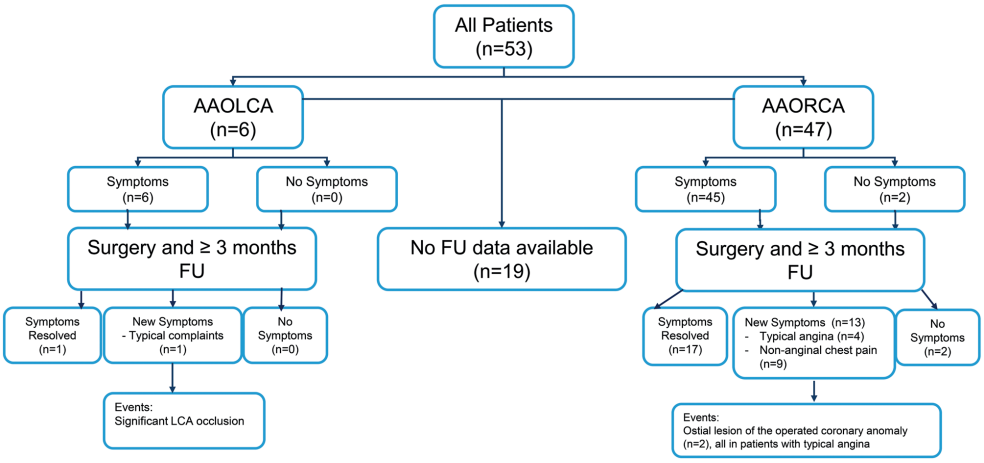
Study population and data collection

The Leiden University Medical Center (LUMC) serves as a national referral center for

patients with congenital heart disease. Consecutive patients who underwent surgical correction of an anomalous aortic origin of a left coronary artery (AAOLCA) or anomalous right coronary artery (AAORCA) arising from the opposite sinus of Valsalva at our center between 2001 and 2018 were included in this study (Figure 3). Patients with concomitant congenital heart defects (e.g. transposition of the great arteries, tetralogy of Fallot, and certain forms of pulmonary atresia), and patients unable or unwilling to communicate with the research team were excluded from analysis. Patient data were collected from the electronic medical file system (EPD-Vision®) and included: patient demographic data, symptoms, sex, indications for surgery, anatomy of the anomalous coronary artery, surgical techniques, imaging modalities, functional tests, clinical course and outpatient visit reports. Major adverse cardiac events included sustained ventricular tachycardia (VT) or fibrillation (VF), re-operation or percutaneous coronary intervention (PCI) on the operated coronary artery, and/or (cardiac) death. The study focused on medium-term outcomes. Therefore in hospital events in post-operative setting (< 1 month) and patients with less than 3 months follow-up were excluded.

All patients were asked about recurrence of chest pain related symptoms and reinterventions. All chest pain (related) complaints were classified according to the '2019 ESC guidelines on chronic coronary syndromes ' [16]: Chest pain is classified as "typical angina", "atypical angina" and "non-anginal chest pain". Typical angina is defined as 1. "constricting discomfort in the front of the chest or in the neck, jaw, shoulder, or arm", 2. "precipitated by physical exertion" and 3. "relieved by rest or nitrates within 5 minutes". Atypical angina meets 2 of these criteria and non-anginal chest pain satisfies one or none of the above-mentioned characteristics. Patients were also categorized into the "typical" group if there were other complaints that were strongly associated with ischemia. All chest

Figure 3. Overview of the 34 patients with follow-up



pain related symptoms were categorized independently into above mentioned groups by 2 experienced cardiologists (HWV and PKI) who were blinded for the results.

Statistical analysis

Analyses were performed with SPSS Statistics (version 23, IBM Corp, Armonk, New York, USA). Descriptive statistics were used for data analysis and were expressed as mean ± standard deviation (SD) and median (IQR; interquartile range). Binary data were expressed in numbers with percentages. All reported p values were two-sided, and values of $p < 0.050$ were considered significant.

Results

Baseline patient characteristics at initial presentation

Baseline patient characteristics are described in Table 1. This study consisted of 53 patients who underwent surgery for correction of AAOCA; 47 (89%) patients had an AAORCA and 6 (11%) patients an AAOLCA. All patients had an intramural course of the anomalous coronary artery. The mean age at surgery was 44 ± 15 years (range 11-68) and 55% were male. Four patients were younger than 16 years old. Fifty-one of 53 patients (96%) had symptoms of some sort at initial presentation. The most common reason for cardiac analysis in these patients was suspicion of ischemia (42 patients, 79%). Three (6%) patients presented with an aborted sudden cardiac death (1 patient with AAOLCA and 2 patients with AAORCA, Table 2). The first patient (patient 2, AAORCA) was 17 years old and playing sports at the time of the cardiac event. No symptoms or cardiac events preceded the cardiac arrest based on ventricular fibrillation. There were no risk factors. The second patient with an AAORCA was 25 years old. This patient was resuscitated due to ventricular fibrillation during exercise, before this event the patient had some non-specific thoracic complaints during exercise and he was a smoker. In 5 (9%) patients AAOCA was an incidental finding and in 3 (6%) patients were identified through familial screening for coronary anomalies. Although no hard evidence exists regarding familial screening in coronary artery anomalies, in these patients screening was performed by the referring center driven by patient desire.

Preoperative testing

Patients were referred to our center with different imaging modalities and functional tests, performed in the referring hospital. Of the 53 patients who were accepted for surgery by the heart-team, 50 (94%) patients underwent computed tomographic angiography (CTA) (Figure 3), 35 (66%) coronary angiography (CAG) and 8 (15%) cardiac magnetic resonance imaging (MRI). Functional ischemia testing was performed in 74% of the patients (Table 1).

In 36 patients (68%) exercise ECG testing was performed, of which 22% had an ischemic response. Ten (19%) patients underwent a nuclear stress test, of which 40% were positive for ischemia.

Initial surgical repair

Surgical techniques used included: unroofing (72%), coronary reimplantation (8%), CABG (2%), patch augmentation (10%), or a combination of the above (8%). None of the anomalous LCA patients underwent patch augmentantion of ostium and main stem (Table 1). Concomitant procedures during the surgical repair were performed in 15 cases and consisted predominantly of aortic valve resuspension in order to prevent aortic regurgitation due to manipulation after unroofing or because of pre-existing aortic regurgitation.

Clinical follow-up

One patient (1/53, 1.9%) died one week after LCA ostioplasty due to severe heparin induced thrombocytopenia (HIT) causing disseminated intravascular coagulation (DIC). The central death administration was consulted, and except for the aforementioned patient, every patient (52/53, 98.1%) was still alive at follow-up (median 5 ± 16 years (IQR 2-18)).

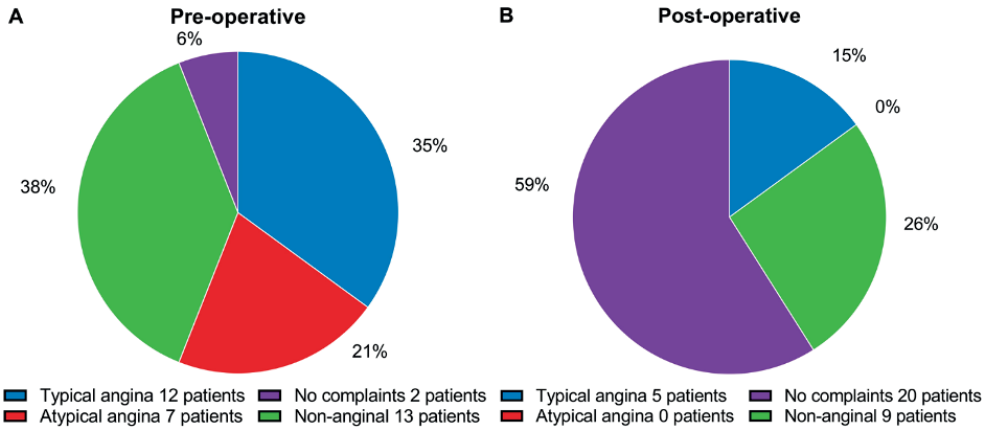
Full follow up data of more than 3 months was available in 34 out of 53 (64%) patients. In 19 (36%) patients full follow-up data could not be obtained due to migration, significant language barrier or inability to contact the patient. In these 34 patients median follow-up of 3.0 years (IQR 1.0 – 5.3, Figure 1) was attained.

The pre- and post-operative symptoms in the 34 patients with more than 3 months follow-up are shown in Figure 4. Pre-operatively, 35% (12/34) of patients presented with typical angina, 21% (7/34) with atypical angina and 38% (13/34) had non-anginal chest pain. Only 6% of the patients (2/34) were asymptomatic before surgery. After surgical correction, 59% (20/34) of the patients reported to be free of symptoms, this is a significant reduction in the total burden ($p<0001$). In 15% (5/34) of the patients, a post-operative catheterisation was performed due to typical complaints after surgery (Table 2). In three of these patients (9%, 3/34), lesions of the operated AAOCA were diagnosed, detailed in Table 2. Patient 6 presented with typical complaints 5 years after surgery, on CAG there was an occlusion of the LAD, thus not associated to the unroofed RCA. Patient 7 (reimplantation of AAOLCA) had a significant left main stenosis for which a successful PCI was performed. Patient 19, also presented with typical complaints, however on catheterisation no stenosis was seen and no additional treatment was performed. Patient 21 (unroofing of AAORCA) had a flattened ostium of the RCA for which a PCI was performed. Patient 30 (unroofing of AAORCA) presented with a near-collapse and angiography revealed ostial stenosis of the RCA for which a CABG was performed (right internal mammary artery graft on the RCA,

clip proximal RCA).

Table 3 is an overview of the remaining 27 patients and their clinical and anatomical characteristics, surgical course and follow-up. All patients with atypical angina at presentation were free of symptoms after surgery. Two patients were asymptomatic prior to surgery and remained asymptomatic postoperatively and during follow-up. In these patients, AAOCA was diagnosed through familial screening and was judged to be a malignant variant and therefore, these patients underwent surgical correction.

Figure 4. Preoperative and postoperative symptoms of 34 patients with follow-up classified according to the "2019 ESC Guidelines on chronic coronary syndromes".



Discussion and conclusion

In this study we report on the medium term outcomes (median of 3 years IQR 1.0 – 5.3) of 34 patients who underwent surgical correction for AAOCA. Our main findings are the following:

1. Of patients who were referred to our center with AAOCA, 94% initially present with symptoms: 35% have typical complaints, 21% atypical complaints, 38% non-anginal complaints, and 6% have no complaints at all.
2. After surgical correction of AAOCA, 59% of the patients are free of symptoms. Compared to 6% pre-operatively ($p < 0.001$).
3. Patients who had significant lesions of the operated coronary artery during medium term follow up (3/34, 9%), all presented with novel typical anginal complaints in the outpatient clinic.

The clinical presentation of adults with an AAOCA varies. In our study 35% of patients presented with typical angina which is comparable to previous reports [4,13,17]. Consequently,

the indication for intervention is based on other clinical factors [18, 19]. Guidelines of the American College of Cardiology, American Heart Association and Thoracic Surgery suggest that surgical intervention may be warranted in younger patients with evidence of ischemia [18]. Palmieri et al reported good clinical outcomes after conservative treatment strategy (exercise restriction) in 23 young athletes [19].

To our knowledge, we are the first group to report specifically on an adolescent and adult group, with mean age of 44 years at time of surgery. Particularly in older patients with AAORCA without signs of ischemia, indication for intervention is currently not clearly defined. In previous studies the risk of SCD appears highest in young patients and particularly in interarterial AAOLCA, therefore the indication for surgical correction in this group is not up for debate [5, 20]. The current guidelines recommend revascularization for interarterial AAOLCA, regardless of ischemia of symptoms [21]. In patients with AAORCA without signs of ischemia the indication for intervention relies on numerous factors to guide management. Clinical presentation, anatomical and functional characteristics of the AAOCA as well as patient specific factors all have to be taken into account [18, 21].

Perioperative mortality in our study was 1.9% (1/53), which is in line with previously reported post-operative mortality rates of AAOCA correction in children and young adults [9, 22-24]. According to literature, in the majority of the patients AAOCA is an incidental finding, probably due to a vast increase of the use of CT and MRI imaging in our current clinical practice. In our study only 9% of the patients were diagnosed with AAOCA as an incidental finding, reflecting the subselection of patients who were operated. In current clinical practice, therefore, numerous anatomical, (patho)physiological factors and the individual operative risk are considered when evaluating an AAOCA patient. Our results show a low discriminative value of the type of complaints as over 60% of all AAOCA patients did not have typical complaints at initial evaluation.

After more than 3 months following the surgical correction of AAOCA, 59% of the patients were free of symptoms. This was a significant improvement compared to the preoperative situation and was unrelated to the type of pre-operative complaints. Interestingly, out of the 5 patients having typical complaints at follow-up, 3 (60%) needed re-intervention due to a significant lesion of the operated artery. This is in line with previous literature [15, 25]. In our series 9% (3/34) of the operated patients needed re-intervention due to a significant lesion in the operated artery. The rate of re-intervention is relatively high in relation to literature which varies between 1.7 and 3.3% [9, 13, 14]. This may be a reflection of the older age of the study population compared to most series reporting on pediatric patients [7, 26].

Mainwaring et al. report on a significantly younger group with 115 AAOCA patients with a follow up of 6 years, the median age at surgery was 16 years [13]. In this study two patients had recurrent symptoms of chest pain and underwent reoperation (one had revision of the initial repair and one had repair of a myocardial bridge) [13]. Nees et al. 14 reported on 2 patients with AAOLCA that needed reoperation due to restenosis of the anomalous coronary artery, 2 months and 6 years after surgery, respectively. One patient, aged 68 had recurrent chest pain, showed an abnormal electrocardiogram and was treated with a bypass graft because of significant stenosis of the operated artery. The other patient, aged 10, survived an aborted SCD 6 years post-operatively and CT and interoperative examination showed ostial narrowing due to fibrous tissue around the left coronary orifice [14]. In the study of Padalino et al. 3 patients needed re-intervention of the operated artery [9]. These cases, together with our data indicate that restenosis of the corrected anomalous artery is a complication that can be observed during medium and long term follow-up of adult patients. It therefore seems justified to perform lifetime follow-up in patients after surgical correction of AAOCA.

Limitations

Despite our role as a national referral center, the sample size is small, reflecting the rarity of the condition. The nature of the data is largely descriptive, and symptoms may be subjective, particularly when evaluated retrospectively. However, the complaints were judged by 2 independent cardiologist who were blinded for the results. Given our role as a referral center, patients are typically sent back to the referring cardiologist for life-long follow up at the local hospital. This contributed to the high rate of loss to follow up.

Conclusions

Our data shows the varying symptoms at presentation in adolescent and adult patients with AAOCA. Only 35% have typical anginal complaints. Surgical correction of AAOCA reduces the symptoms in the vast majority of patients. One should be aware of potential lesions of the operated coronary artery in patients presenting with typical anginal complaints post-operatively.

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Table 1. Patient characteristics

Patient characteristics	All patients (n=53)
Male, n (%)	29 (55)
Age at surgery, years, mean ± SD	44 (15)
Diabetes mellitus n (%)	3 (6)
Hypertension n (%)	10 (19)
Previous ischemic coronary disease n (%)	0
Hypercholesterolemia n (%) TIA/CVA n (%)	13 (25)
AAOLCA n (%)	6 (11)
AAORCA n (%)	47 (89)
Symptoms present, n (%)	51 (96)
Primary presentation, n (%)	
Suspicion of ischemia	42 (79)
Aborted sudden cardiac death	3 (6)
Familial screening	3 (6)
Incidental finding	5 (9)
Diagnostic imaging techniques, n (%)	
CTA	50 (94)
CAG	35 (66)
MRI	8 (15)
Diagnostic functional test, n (%)	-
Exercise ECG	36 (68)
Positive	8 (22)
Nuclear stress test	10 (19)
Positive	4 (40)
Adenosine stress perfusion CT	4 (8)
Positive	1 (25)

Dobutamine stress MRI	2 (4)
Positive	0
PET-CT	2 (4)
Positive	0
No test	14 (26)
Surgical technique, n (%)	
Unroofing	38 (72)
Reimplantation	4 (8)
Unroofing + reimplantation	3 (6)
Unroofing + CABG	1 (2)
Ostioplasty	5 (10)
Unroofing + ostioplasty	1 (2)
CABG	1 (2)
Concomitant procedure, n (%)	15 (28)
Aortic valve repair	6
Tricuspid valve repair	1
Mitral- and aortic valve repair	1
Epicardial lead placement	1
Excision of cardiac lipoma	1
Pulmonary vein isololation, left atrial resection, aortic valve repair	1
CABG Ao-D-LAD	1

CTA, computed tomographic angiography; CAG, coronary angiography; MRI, magnetic resonance imaging; PET-CT, position emission tomography computed tomography; CABG, coronary artery bypass grafting; AAOLCA, anomalous left coronary artery; AAORCA, anomalous right coronary artery; CABG, coronary artery bypass grafting; Ao, aorta, D, diagonal branch; LAD, left anterior descending artery

Table 2. Consecutive patients with > 3 months follow-up after surgical correction for anomalous aortic origin of a coronary artery with interarterial course and post-operative complaints driven catheterization (n=5)

Pt	Lesion	Age (years)	Clinical presentation	Ischemia detection	Surgical repair	Pre-operative symptoms	Post-operative symptoms	Δt surgery and events (months)	Post-operative events/complications + treatment
6	AAORCA	47	suspected ischemia	positive	unroofing	non-anginal	typical	60	PCI proximal LAD
7	AAOLCA	58	suspected ischemia	positive	reimplantation	typical	typical	1	significant main stem stenosis, PCI main stem
19	AAORCA	49	suspected ischemia	positive	unroofing	typical	typical	15	no stenosis on CAG
21	AAORCA	64	suspected ischemia	positive	unroofing	typical	typical	13	flattening ostium RCA, PCI proximal RCA
30	AAORCA	44	suspected ischemia	negative	unroofing	atypical	typical (near-collapse)	10	stenosis ostium RCA, RIMA-RCA, clip on proximal RCA

Table 3. Consecutive patients with > 3 months follow-up after surgical correction for anomalous aortic origin of a coronary artery with interarterial course and no post-operative events (n=29).

Pt	Lesion	Age	Clinical pre-sentation	Ischemiade-tection	Surgical repair	Pre-operative symptoms	Post-oper-ative symp-toms
1	AAORCA	25	screening	negative	unroofing and reimplantation	typical	no complaints
2	AAORCA	17	aborted SCD	not conclusive	reimplantation	typical	no complaints
3	AAORCA	53	suspected ischemia	negative	reimplantation	atypical	no complaints
4	AAORCA	46	suspected ischemia	negative	unroofing and reimplantation	typical	no complaints
5	AAORCA	34	screening	positive	reimplantation	no complaints	no complaints
8	AAORCA	66	suspected ischemia	negative	unroofing and CABG non anomalous vessel	atypical	non-anginal: chest discom-fort
9	AAORCA	66	suspected ischemia	negative	CABG of anomalous vessel	typical	no complaints
10	AAORCA	25	suspected ischemia	negative	unroofing	non-anginal	no complaints
11	AAORCA	45	suspected ischemia	positive	unroofing	atypical	no complaints

Pt	Lesion	Age	Clinical pre-sentation	Ischemiade-tection	Surgical repair	Pre-operative symptoms	Post-oper-ative symp-toms
12	AAORCA	56	suspected ischemia	negative	unroofing	atypical	non-anginal: tiredness/ loss of condition
13	AAORCA	20	suspected ischemia	positive	unroofing	atypical	no complaints
14	AAORCA	50	suspected ischemia	positive	unroofing	non-anginal	no complaints
15	AAORCA	46	suspected ischemia	negative	unroofing	typical	non-anginal: tiredness/ loss of condition
16	AAORCA	13	suspected ischemia	negative	unroofing	atypical	non-anginal: tiredness/ loss of condition
17	AAOLCA	15	aborted SCD	negative	ostiumplasty	typical	no complaints
18	AAORCA	29	screening	positive	unroofing	non-anginal	non-anginal: palpitations
20	AAORCA	51	screening	positive	unroofing	atypical	non-anginal: sharp chest pain
22	AAORCA	53	suspected ischemia	positive	unroofing	non-anginal	non-anginal: tiredness/ loss of condition

23	AAORCA	48	suspected ischemia	positive	unroofing	non-anginal	non-anginal: sharp chest pain
24	AAORCA	59	suspected ischemia	positive	unroofing	non-anginal	non-anginal: tiredness/ loss of condition
25	AAORCA	67	suspected ischemia	positive	unroofing	non-anginal	no complaints
26	AAORCA	42	suspected ischemia	positive	unroofing	non-anginal	no complaints
27	AAORCA	15	screening	positive	unroofing	no complaints	no complaints
28	AAORCA	63	suspected ischemia	positive	unroofing	non-anginal	no complaints
29	AAORCA	11	suspected ischemia	negative	unroofing	non-anginal	no complaints
31	AAORCA	66	suspected ischemia	positive	unroofing	non-anginal	no complaints
32	AAORCA	52	screening	negative	unroofing	typical	no complaints
33	AAORCA	43	suspected ischemia	negative	unroofing	non-anginal	no complaints
34	AAORCA	47	suspected ischemia	not conclusive	unroofing	atypical	no complaints

Pt: consecutive patient number; Ischemic detection: outcome of ischemic detection pre-operatively; AAORCA: anomalous right coronary artery; AAOLCA: anomalous left coronary artery; FU: follow up; Δt time between surgery and event in months; aSCD: aborted sudden cardiac death; CABG: coronary artery bypass graft; PCI: percutaneous coronary intervention; RDA: right descending artery; RIMA: right internal mammary artery; VF: ventricular fibrillation