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

Fleur M. M. Meijer^a, Anastasia D. Egorova ^a, Monique R. M. Jongbloed^{a,b}, Claire Koppel ^a, Gracia Habib^a, Mark G. Hazekamp^c, Hubert W. Vliegen ^a and Philippine Kies^{a,*}

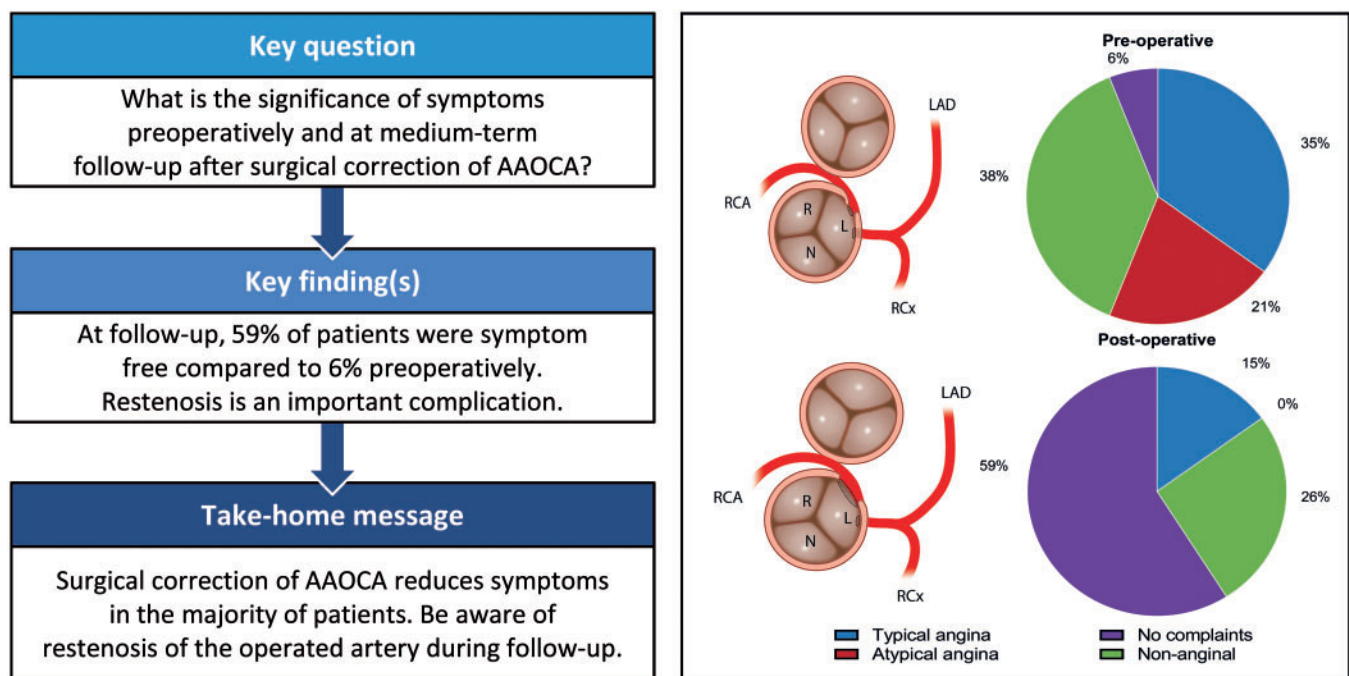
^a Department of Cardiology, CAHAL, Center for Congenital Heart Disease Amsterdam Leiden, Leiden University Medical Center, Leiden, Netherlands

^b Department of Anatomy and Embryology, Leiden University Medical Center, Leiden, Netherlands

^c Department of Cardiothoracic Surgery, CAHAL, Center for Congenital Heart Disease Amsterdam Leiden, Leiden University Medical Center, Leiden, Netherlands

* Corresponding author. Department of Cardiology, Leiden University Medical Center, Box 9600, 2300 RC Leiden, Netherlands. Tel: +31-71-715261793; e-mail: p.kies@lumc.nl (P. Kies).

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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 anomalous aortic origin of a coronary artery (AAOCA).

METHODS: Consecutive patients who underwent surgery for AAOCA in our tertiary referral centre between 2001 and 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 >3 months after surgery were available in 34 patients with a median follow-up of 3 years (interquartile range 1.0–5.3). Preoperatively, only 35% patients had typical anginal complaints. After surgical correction of AAOCA, 59% of the patients were free of

symptoms, compared to 6% preoperatively ($P < 0.001$). A total of 3 (9%) patients needed a reoperation/reintervention related to the operated AAOCA. All 3 patients presented postoperatively with novel typical anginal complaints.

CONCLUSIONS: 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 postoperatively.

Keywords: Coronary anomalies • Medium-term follow-up • Adult congenital surgery • Complications • Symptoms

ABBREVIATIONS

AAOCA	Anomalous aortic origin of a coronary artery
AAOLCA	Anomalous aortic origin of a left coronary artery
AAORCA	Anomalous aortic origin of a right coronary artery
CABG	Coronary artery bypass grafting
CT	Computed tomography
IQR	Interquartile range
MRI	Magnetic resonance imaging
PCI	Percutaneous coronary intervention
RCA	Right coronary artery
SCD	Sudden cardiac death

INTRODUCTION

Anomalous aortic origin of the coronary arteries (AAOCAs) 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 (SCD), especially in athletes and active young adults (Fig. 1) [1]. Presenting symptoms differ largely amongst patients [2, 4, 5]. 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 SCD and alleviate ischaemia. The decision to operate on a patient is based on the ostial anatomy and course of the anomalous coronary artery and demonstrated ischaemia. The role of symptoms in decision-making with regard to the

surgical intervention and postoperative outcomes is ambiguous. Several surgical techniques for correcting AAOCA have been used, most commonly unroofing of the intramural segment (Fig. 2), coronary reimplantation and coronary artery bypass grafting (CABG) [10, 11]. A few studies have reported persistent symptoms, restenosis of the operated anomaly after surgery, ischaemia 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.

MATERIALS AND METHODS

Study population and data collection

The Leiden University Medical Center serves as a national referral centre 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 centre between 2001 and 2018 were included in this study (Fig. 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®, Leiden University Medical Center, Leiden, the Netherlands) 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

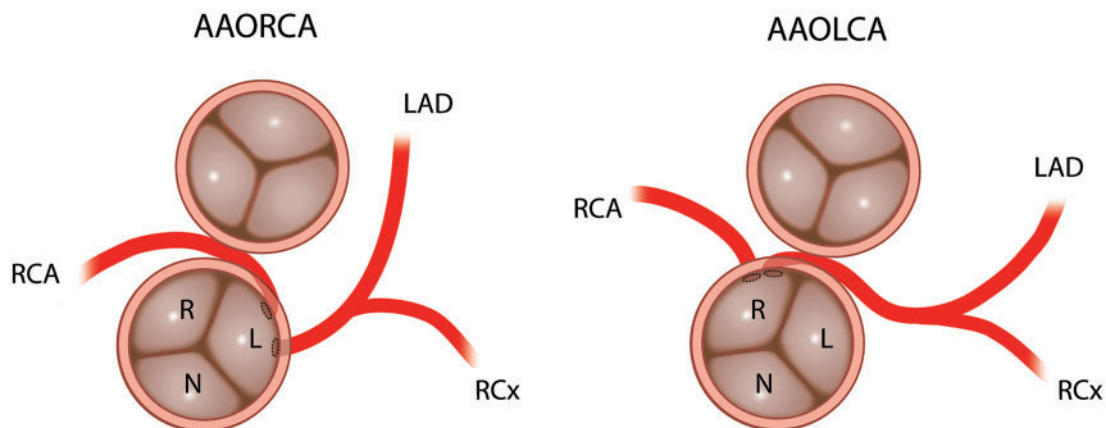


Figure 1: Schematic representation of AAORCA and AAOLCA anatomy with an interarterial and intramural course of the anomalous artery (imaging view). AAOLCA: anomalous aortic origin of a left coronary; AAORCA: anomalous aortic origin of a right coronary; L: left coronary cusp; LAD: left anterior descending artery; N: non-coronary cusp; R: right coronary cusp; RCA: right coronary artery; RCx: ramus circumflex artery.

ventricular tachycardia or ventricular fibrillation, reoperation 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 postoperative setting (<1 month) and patients with <3 months follow-up were excluded.

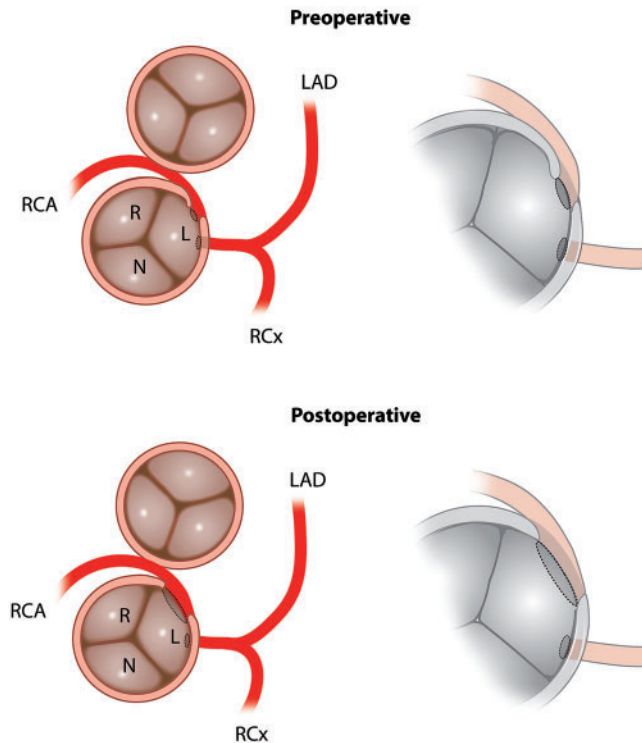


Figure 2: Schematic representation of AAORCA anatomy prior to and after surgical correction (imaging view). AAORCA: anomalous aortic origin of a right coronary; L: left coronary cusp; LAD: left anterior descending artery; N: non-coronary cusp; R: right coronary cusp; RCA: right coronary artery; RCx: ramus circumflex artery.

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 (i) 'constricting discomfort in the front of the chest or in the neck, jaw, shoulder or arm', (ii) 'precipitated by physical exertion' and (iii) 'relieved by rest or nitrates within 5 min'. Atypical angina meets 2 of these criteria and non-anginal chest pain satisfies 1 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 ischaemia. All chest pain-related symptoms were categorized independently into above-mentioned groups by 2 experienced cardiologists (H.W.V. and P.K.) who were blinded to the results.

Statistical analysis

Analyses were performed with SPSS Statistics (version 23, IBM Corp, Armonk, NY, USA). Descriptive statistics were used for data analysis and were expressed as mean \pm standard deviation and median [interquartile range (IQR)]. Binary data were expressed in numbers with percentages. All reported *P*-values were 2-sided, and *P*-values <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

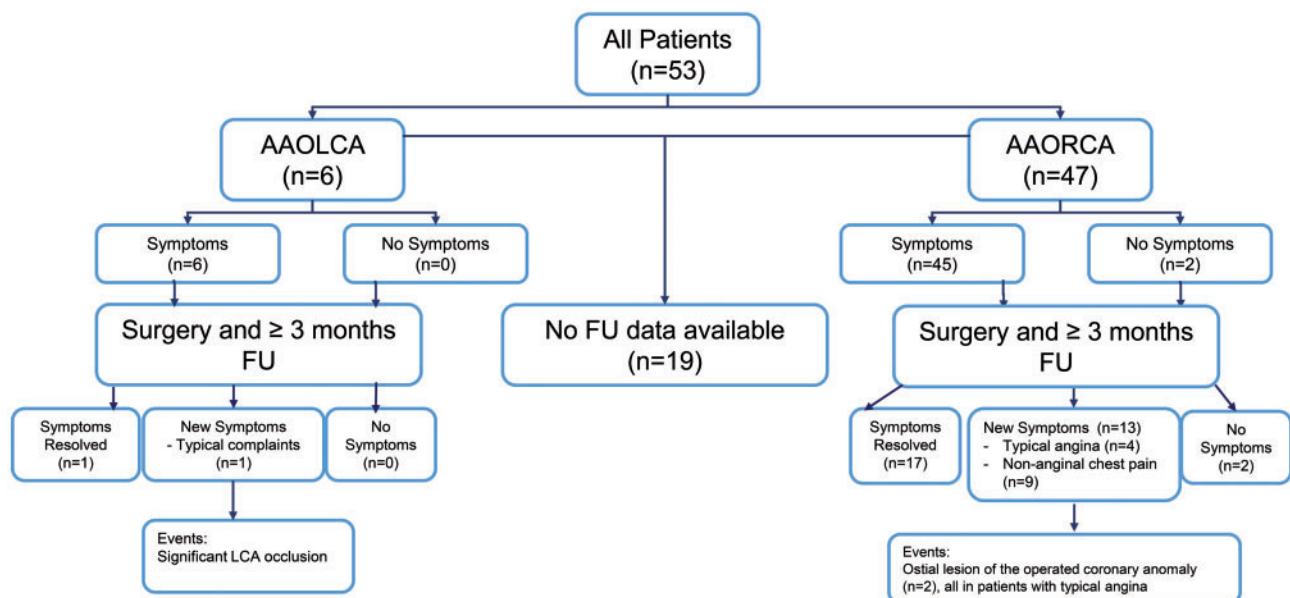


Figure 3: Overview of the 34 patients with follow-up. AAOLCA: anomalous aortic origin of a left coronary; AAORCA: anomalous aortic origin of a right coronary; FU: follow-up.

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 ischaemic coronary disease n (%)	0
Hypercholesterolemia n (%)	13 (25)
TIA/CVA n (%)	
AAOLCA	6 (11)
AAORCA	47 (89)
Symptoms present, n (%)	51 (96)
Primary presentation, n (%)	
Suspicion of ischaemia	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 isolation, left atrial resection, aortic valve repair	1
CABG Ao-D-LAD	1

AAOLCA: anomalous left coronary artery; AAORCA: anomalous right coronary artery; Ao: aorta; CABG: coronary artery bypass grafting; CAG: coronary angiography; CTA: computed tomographic angiography; D: diagonal branch; LAD: left anterior descending artery; MRI: magnetic resonance imaging; PET-CT: position emission tomography computed tomography; SD: standard deviation; TIA: transient ischemic attack; CVA: cerebral vascular accident.

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 ischaemia (42 patients, 79%). Three (6%) patients presented with an aborted SCD (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, it was 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 centre driven by patient desire.

Preoperative testing

Patients were referred to our centre 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 (Fig. 3), 35 (66%) patients coronary angiography and 8 (15%) patients, cardiac magnetic resonance imaging (MRI). Functional ischaemia testing was performed in 74% of the patients (Table 1). In 36 patients (68%), exercise ECG testing was performed, of which 22% had an ischaemic response. Ten (19%) patients underwent a nuclear stress test, of which 40% were positive for ischaemia.

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 augmentation 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 1 week after LCA ostioplasty due to severe heparin-induced thrombocytopenia causing disseminated intravascular coagulation. 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)].

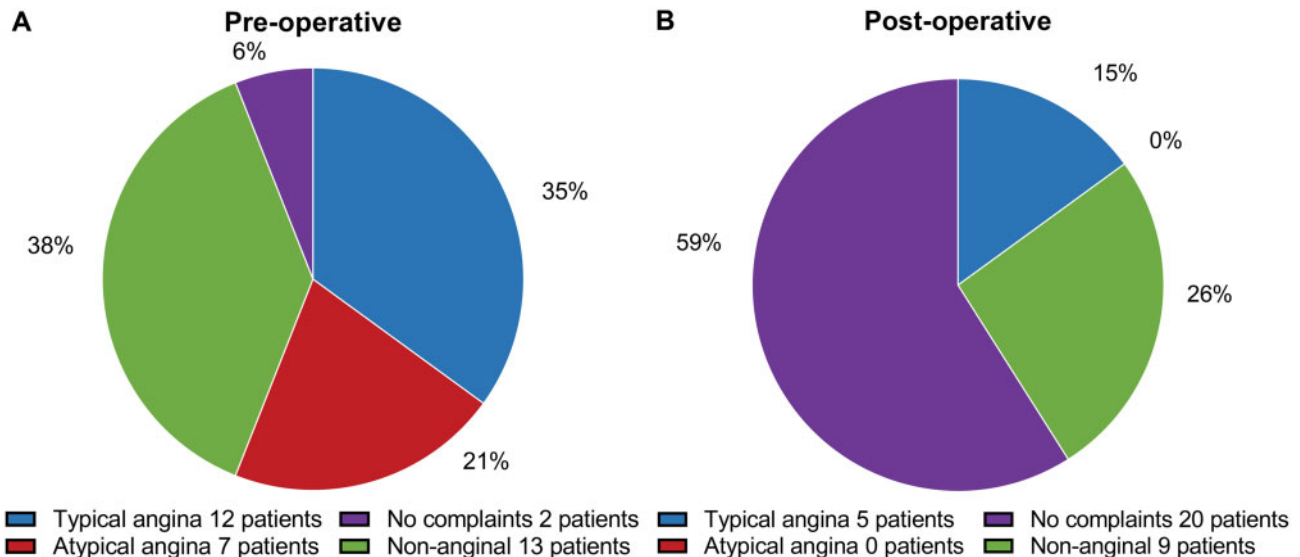
Full follow-up data of >3 months were available in 34 out of 53 (64%) patients. In 19 (36%) patients, full follow-up data could not be obtained due to migration, significant language barrier or inability to contact the patient. In these 34 patients, median follow-up of 3.0 years (IQR 1.0–5.3, Fig. 1)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

The pre- and postoperative symptoms in the 34 patients with >3 months follow-up are shown in Fig. 4. Preoperatively, 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 being a significant reduction in the total burden ($P < 0.001$). In 15% (5/34) of the patients, a postoperative catheterization was performed due to typical

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

Pt	Lesion	Age (years)	Clinical presentation	Ischaemia detection	Surgical repair	Preoperative symptoms	Postoperative symptoms	Δt surgery and events (months)	Postoperative events/ complications + treatment
6	AAORCA	47	Suspected ischaemia	Positive	Unroofing	Non-anginal	Typical	60	PCI proximal LAD
7	AAOLCA	58	Suspected ischaemia	Positive	Reimplantation	Typical	Typical	1	Significant main stem stenosis, PCI main stem
19	AAORCA	49	Suspected ischaemia	Positive	Unroofing	Typical	Typical	15	No stenosis on CAG
21	AAORCA	64	Suspected ischaemia	Positive	Unroofing	Typical	Typical	13	Flattening ostium RCA, PCI proximal RCA
30	AAORCA	44	Suspected ischaemia	Negative	Unroofing	Atypical	Typical (near-collapse)	10	Stenosis ostium RCA, RIMA-RCA, clip on proximal RCA

AAOLCA: anomalous left coronary artery; AAORCA: anomalous right coronary artery; CAG: coronary angiography; LAD: left anterior descending artery; PCI: percutaneous coronary intervention; RCA: right coronary artery; RIMA: right internal mammary artery.

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

complaints after surgery (Table 2). In 3 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 coronary angiography, there was an occlusion of the left anterior descending artery, and thus not associated with the unroofed right coronary artery (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 catheterization, 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 presents 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.

DISCUSSION

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 centre 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% preoperatively ($P < 0.001$).

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 postoperative events (*n* = 29)

Pt	Lesion	Age	Clinical presentation	Ischaemia detection	Surgical repair	Preoperative symptoms	Postoperative symptoms
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 ischaemia	Negative	Reimplantation	Atypical	No complaints
4	AAORCA	46	Suspected ischaemia	Negative	Unroofing and reimplantation	Typical	No complaints
5	AAORCA	34	Screening	Positive	Reimplantation	No complaints	No complaints
8	AAORCA	66	Suspected ischaemia	Negative	Unroofing and CABG non-anomalous vessel	Atypical	Non-anginal: chest discomfort
9	AAORCA	66	Suspected ischaemia	Negative	CABG of anomalous vessel	Typical	No complaints
10	AAORCA	25	Suspected ischaemia	Negative	Unroofing	Non-anginal	No complaints
11	AAORCA	45	Suspected ischaemia	Positive	Unroofing	Atypical	No complaints
12	AAORCA	56	Suspected ischaemia	Negative	Unroofing	Atypical	Non-anginal: tiredness/loss of condition
13	AAORCA	20	Suspected ischaemia	Positive	Unroofing	Atypical	No complaints
14	AAORCA	50	Suspected ischaemia	Positive	Unroofing	non-anginal	no complaints
15	AAORCA	46	Suspected ischaemia	Negative	Unroofing	Typical	Non-anginal: tiredness/loss of condition
16	AAORCA	13	Suspected ischaemia	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 ischaemia	Positive	Unroofing	Non-anginal	Non-anginal: tiredness/loss of condition
23	AAORCA	48	Suspected ischaemia	Positive	Unroofing	Non-anginal	Non-anginal: sharp chest pain
24	AAORCA	59	Suspected ischaemia	Positive	Unroofing	Non-anginal	Non-anginal: tiredness/loss of condition
25	AAORCA	67	Suspected ischaemia	Positive	Unroofing	Non-anginal	No complaints
26	AAORCA	42	Suspected ischaemia	Positive	Unroofing	Non-anginal	No complaints
27	AAORCA	15	Screening	Positive	Unroofing	No complaints	No complaints
28	AAORCA	63	Suspected ischaemia	Positive	Unroofing	Non-anginal	No complaints
29	AAORCA	11	Suspected ischaemia	Negative	Unroofing	Non-anginal	No complaints
31	AAORCA	66	Suspected ischaemia	Positive	Unroofing	Non-anginal	No complaints
32	AAORCA	52	Screening	Negative	Unroofing	Typical	No complaints
33	AAORCA	43	Suspected ischaemia	Negative	Unroofing	Non-anginal	No complaints
34	AAORCA	47	Suspected ischaemia	Not conclusive	Unroofing	Atypical	No complaints

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

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 ischaemia [18]. Palmieri *et al.* [19] reported good clinical outcomes after conservative treatment strategy (exercise restriction) in 23 young athletes.

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 ischaemia, 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 ischaemia or symptoms [21]. In patients with AAORCA without signs of

ischaemia, 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 postoperative mortality rates of AAOCA correction in children and young adults [9, 22–24].

According to the literature, in the majority of the patients, AAOCA is an incidental finding, probably due to a vast increase in the use of computed tomography (CT) and MRI 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 >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 preoperative complaints. Interestingly, out of the 5 patients having typical complaints at follow-up, 3 (60%) needed reintervention 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 reintervention due to a significant lesion in the operated artery. The rate of reintervention is relatively high in relation to the 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 paediatric patients [7, 26].

Mainwaring *et al.* [13] report on a significantly younger group with 115 AAOCA patients with a follow-up of 6 years, and the median age at surgery was 16 years. In this study, 2 patients had recurrent symptoms of chest pain and underwent reoperation (1 had revision of the initial repair and 1 had repair of a myocardial bridge) [13]. Nees *et al.* [14] reported on 2 patients with AAOCA 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, and 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 postoperatively, and CT and intraoperative examination showed ostial narrowing due to fibrous tissue around the left coronary orifice [14]. In the study of Padalino *et al.* [9], 3 patients needed reintervention of the operated artery. 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 centre, 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 cardiologists who were blinded to the results. Given our role as a referral centre, patients are typically sent back to the referring cardiologist for lifelong follow-up at the local hospital. This contributed to the high rate of loss to follow-up.

CONCLUSIONS

Our data show 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 postoperatively.

SUPPLEMENTARY MATERIAL

Supplementary material is available at *ICVTS* online.

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Conflict of interest: none declared.

Author contributions

Fleur M.M. Meijer: Conceptualization; Data curation; Formal analysis; Methodology; Writing—original draft. **Anastasia D. Egorova:** Supervision; Validation; Writing—review & editing. **Monique R.M. Jongbloed:** Conceptualization; Writing—review & editing. **Claire Koppel:** Writing—review & editing. **Gracia Habib:** Data curation; Formal analysis; Writing—review & editing. **Mark G. Hazekamp:** Conceptualization; Writing—review & editing. **Hubert W.Vliegen:** Conceptualization; Supervision; Writing—review & editing. **Philippine Kies:** Conceptualization; Project administration; Supervision; Writing—review & editing.

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