Surgical treatment of aberrant aortic origin of coronary arteries†

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Abstract

OBJECTIVES: Aberrant origin of the coronary arteries is rare but can be life threatening. It is an important cause of sudden death in athletes and other young adults, and may be treated surgically. Consensus exists that interarterial left coronary artery (LCA) should be surgically repaired. For interarterial right coronary artery (RCA), the discussion remains open. The purpose of this study was to analyse our surgical experience.

METHODS: From 2001 until 2014, 31 patients were operated for interarterial RCA, interarterial LCA or intraseptal course of the LCA. Twenty-six patients had interarterial RCA, 4 patients interarterial LCA and 1 patient an intraseptal course of the LCA. Median age at operation was 38 years (range 9–66 years). Twenty-eight patients had previous or current symptoms. The most important were a life-threatening event with resuscitation in 3 and myocardial infarction in 3 others. Surgical repair of interarterial RCA consisted of unroofing of the ostium with or without reimplantation in 25 patients and CABG on the RCA with a venous graft in 1 patient. Reconstruction of interarterial LCA consisted of ostium reconstruction of the LCA with a venous patch in 4 patients. The patient with an intraseptal course had a complete release of the LCA out of the septum and reimplantation in the correct coronary sinus. Follow-up was done by analysis of outpatient records, direct patient contact, echocardiography, electrocardiography, CT-angiography and an exercise test.

RESULTS: Median follow-up was 6 years (range 0–11 years). One patient was lost to follow-up. No early or late mortality occurred. Three patients had ischaemia with ventricular fibrillation or ventricular tachycardia shortly after surgery. Two were immediately reoperated, 1 had a stent implantation 1.5 months after release of intraseptal LCA. Two of these patients show a slight dysfunction of the left ventricle at follow-up. All other patients are asymptomatic.

CONCLUSIONS: Surgery for aberrant origin of coronary arteries is safe. There is a risk of cardiac ischaemia shortly after operation, especially in LCA reconstruction. We strongly believe that a slit-like coronary ostium and an intramural aortic course is an absolute indication for surgical repair, also in asymptomatic aberrant RCA.

Keywords: Aberrant coronary artery • Surgery

INTRODUCTION

Coronary arteries may have an anomalous origin, and this can be life-threatening. Although rare, these anomalies are an important cause of sudden death of athletes and other young adults who are otherwise healthy [1–10]. Sudden death and other problems related to the malformation may be prevented by surgery [11–14]. Prevalence of anomalous coronary arteries arises from the opposite sinus range 0.08–0.61% [15–20]. Two subtypes are interarterial right coronary artery (RCA) and interarterial left coronary artery (LCA).

Interarterial RCA is the most frequently encountered anomaly [15, 20, 21]. The interarterial RCA has its origin in the left coronary sinus with the ostium typically located near the commissure between the right and left coronary aortic cusp, while the interarterial LCA has its origin in the right coronary sinus. The ostium is usually slit-like, and the first part of the coronary artery runs intramurally through the aortic wall [1, 3, 5, 6, 8]. The combination of a slit-like ostium, an intramural course and its position between the two great arteries is thought to be the cause for acute ischaemia, especially in a period of strenuous exercise [22]. Our goal was to analyse our surgical techniques and outcomes of aberrant aortic origin of the coronary arteries.

PATIENTS AND METHODS

From December 2001 until September 2014, 31 patients were operated for interarterial RCA, interarterial LCA or intraseptal LCA (Fig. 1). The median at operation was 38 years (range 9–66 years). Four patients were younger than 18 years. Fourteen were male (45%) (Table 1).
At the time of the diagnosis, 28 patients had previous or current symptoms (90%). The most common symptoms were angina pectoris (14 patients), collapse or syncope (5 patients) and palpitations (5 patients). Five patients had a combination of these symptoms. Three patients had survived a life-threatening event with cardiopulmonary resuscitation. Three patients had a myocardial infarction, 1 was diagnosed with non-ST elevation myocardial infarction (non-STEMI) and 2 patients had an anteroseptal infarction prior to surgery.

Seven patients conducted an exercise test before surgery. For only 2 patients, the exercise test was positive for ischaemia.

Final diagnosis was made by CT-angiography in all patients (Figs 2 and 3).

All interarterial RCAs originated from the left coronary sinus next to the commissure with the right coronary sinus. All interarterial LCAs originated from the right coronary sinus. Twenty-six patients had interarterial RCA, 4 had interarterial LCA and 1 patient had an intraseptal course of the LCA. The opening was slit-like in all patients. An intramural course was observed in 26 patients (84%).

Surgical repair of interarterial RCA consisted of unroofing of the ostium in 17 patients (Fig. 4), unroofing and reimplantation in the correct coronary sinus in 8 patients and coronary artery bypass on the proximal RCA in 1 patient.

Reconstruction of interarterial LCA consisted of ostium reconstruction of the LCA with a patch of the vena saphena magna in 4 patients (Fig. 5). A complete release of the LCA out of the septum and reimplantation in the right coronary sinus was necessary in the patient with intraseptal main LCA.

Unroofing of both RCA and LCA was needed for 1 patient, because both ostia were slit-like.

In 1 patient, resuspension of the aortic valve commissure between the right and left commissure was necessary.

Other atherosclerotic coronary anomalies were found in 2 patients who underwent associated CABG with a single graft on the proximal left anterior descending. Another patient had a Bentall procedure and an aortic aneurysm and aortic insufficiency.

All patients were operated by median sternotomy, normothermic cardiopulmonary bypass and cold crystalloid cardioplegic myocardial arrest.

Reimplantation of an aberrant coronary artery into its ‘correct’ sinus was done in the beginning of our experience. Later on, we focused on ostial reconstruction only.

Figure 1: Examples of anomalies in our group of patients. RCA: right coronary artery; LCA: left coronary artery.
## Table 1: An overview of the type of malformation, the age at surgery, the symptoms the patients experienced, surgical technique, complications and follow-up time

<table>
<thead>
<tr>
<th>Patient</th>
<th>Sex</th>
<th>Type of aberrant coronary artery</th>
<th>Age at surgery</th>
<th>Symptoms</th>
<th>Surgical technique</th>
<th>Postoperative complications</th>
<th>Follow-up time (months)</th>
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<td>Ventricular tachycardia</td>
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M: male; F: female; RCA: right coronary artery; LCA: left coronary artery.
One patient had been previously operated in a different institution with right mammary artery placement on the midpart of the RCA. As the mammary artery had closed down and symptoms were still present, he underwent RCA ostium unroofing.

Postoperative anticoagulation was always with a platelet aggregation inhibitor (carbasalate calcium and/or clopidogrel) for 6 months.

Follow-up was obtained by contact with the patient’s cardiologist and direct contact via telephone. Patients were evaluated with electrocardiography (ECG), echocardiography and an exercise test.

**RESULTS**

The median follow-up of this study is 6 years (range 0–11 years). One patient was lost to follow-up 11 years after repair because of emigration abroad.

No early or late mortality occurred.

Postoperative imaging was performed in all patients. All patients had a postoperative echocardiography, and 9 patients had a CT angiography. All reconstructed coronary arteries were observed to be patent (by echo or by CT scan). All patients had an ECG after surgery, all at rest, and 6 patients had an exercise test.

Three patients suffered from ischaemia with ventricular fibrillation or ventricular tachycardia, all with loss of cardiac output, shortly after surgery. One had interarterial LCA, another had interarterial RCA while the third patient had an intraseptal course of LCA. Dysfunction of the unroofed reimplemented RCA occurred 4 h after surgery, and necessitated urgent CABG with single graft on the RCA. The second patient showed left ventricle (LV) pan-ischaemia 3 h after surgery for correction of interarterial LCA. At reoperation, a thrombus was found in the reconstructed coronary ostium. The coronary ostium was made even wider, and the patient recovered uneventfully. The third patient had ventricular fibrillation on the intensive care unit shortly after release, and reimplantation of intra-septal and aberrant LCA was successfully resuscitated. This patient was not reoperated immediately but suffered from mild septal non-STEMI 2 months after surgery for which a stent was placed.

Late follow-up with echocardiography and ECG showed mild but permanent damage in 2 of these 3 patients. The patient with ischaemia based on a thrombus had a lateral infarction with mild LV dysfunction with an ejection fraction of 38%. The patient with non-STEMI had a septal infarction with a slight decline in LV function. This patient had a normal ejection fraction, but hypokinesia anteroseptal. The patient with ischaemia based on dysfunction of the implanted RCA had no signs of dysfunction on ECG and echocardiography.

All other patients recovered uneventfully. All are in NYHA class I without any symptoms of angina pectoris.

**DISCUSSION**

Some aberrant coronary arteries are potentially life-threatening. A slit-like ostium, an intramural aortic course and the course between aorta and pulmonary artery or right ventricular outflow tract may all contribute to the development of ischaemia, especially at exercise. Modern CT angiography can perfectly distinguish an intramural course, and is the preferred diagnostic tool [23]. In our protocol, CT angiography is the diagnostic method of choice. Slide sections need to be 0.5 mm to distinguish accurately...
an intramural course of the aberrant coronary artery. Surgery for interarterial RCA can be carried out in several ways: simply unroofing the ostium or by a combination of unroofing and reimplantation into a more orthotopic position. When reimplantation is necessary, the aortic defect is repaired by xenopericardial patch implantation. The mechanism of flow obstruction is mainly caused by the presence of a slit-like ostium and an intramural course. We therefore now feel that ostial reconstruction is the most important measure, and in the majority of cases there is no further need to address the interarterial course of the RCA. Unroofing the slit-like ostium is sufficient: this technique both widens the ostium itself and makes the intramural course of the proximal RCA to disappear. If the surgeon can look into the proximal RCA, there is no further risk of compression or obstructed blood flow. Unroofing is performed by excision of the triangular septum that runs between the lumen of the proximal RCA and the lumen of the aorta (Fig. 6). Care should be taken not to perforate the aortic wall in the top of this triangle. This is easily avoided by identifying the course of the RCA on the outside of the aorta. Surgical repair of an interarterial LCA is best done by ostium patch augmentation. A saphenous vein patch is well-suited for reconstruction of the proximal LCA. For patch

![Figure 3: Reconstructed multislice cardiac CT. The RCA arises from the LCA cusp and has an interarterial course between the aorta and pulmonary artery (A and B). An aberrant course of the LCA. The LCA arises from the RCA cusp, and has an interarterial course between the aorta and pulmonary artery (C and D) arrows pointing at the anomaly.](image)

![Figure 4: Photograph of an aberrant RCA before an unroofing procedure. The ostium is next to the commissure and slit-like, and there is an intramural course (A). An aberrant RCA after an unroofing procedure. The intramural course is cut open inside the aortic wall, and there is no more slit-like ostium (B).](image)
augmentation of an aberrant LCA, the aorta is opened transversely at the level of the sinotubular junction. Perpendicular to this incision, another incision is made that proceeds into the LCA until the proximal LCA is well opened. With a rectangular patch of vena saphena magna, the LCA is then augmented and the aortotomy is closed (Fig. 7).

Antiaggregation therapy is prescribed for the first six postoperative months (carbasalate calcium for RCA unroofing and carbasalate calcium/clopidogrel for LCA patch augmentation). When deemed necessary or when postoperative coronary artery ischaemia occurs, a venous or arterial bypass graft may be placed on the proximal coronary artery. In this case, ligation...
of the coronary artery proximal to the bypass graft may be considered.

A recent article suggests that patients with asymptomatic aberrant RCA should not be operated. The authors state ‘The incidence of sudden death in asymptomatic patients with aberrant RCA is extremely low. Over the course of the past 25 years, clinical reports describe 10 patients who died suddenly with RCA from the LCA sinus who were not known to be symptomatic.’ [24]. It furthermore suggests that the risk of sudden cardiac death is elevated. The conclusion is that the risk of surgery outweighs the risk of sudden cardiac death, which means surgery would be riskier than no surgery. We do not agree with the assumptions made in this article. Based on our experience, we can say that surgical correction of an aberrant RCA is a safe treatment. No early or late mortality occurred, and there were no longer symptoms after surgery. Complications may occur shortly after surgery but can be solved successfully.

The other statement made is that the incidence of sudden death in asymptomatic patients with aberrant RCA is extremely low. However, in our group of patients with interarterial RCA, already 2 patients (aged 28 and 38) experienced ventricular fibrillation as first symptom, and had to be resuscitated. Literature shows confusing results concerning the risk of cardiac death in asymptomatic patients with aberrant RCA. In the research that has been done, the risk varies from no increased risk to a risk of 43% [2, 10].

In all our patients with slit-like ostium and an intramural course, there was evident obstruction.

A slit-like ostium was observed in all our patients and an intramural course in the vast majority, this and the occurrence of life-threatening events convince us that interarterial LCA but also interarterial RCA should be treated surgically. The only exception may be formed by elderly patients with an aberrant RCA who have no symptoms and a negative exercise test.

STUDY LIMITATIONS

This is a retrospective follow-up study, and therefore carries the disadvantages of this design.

Both pre- and postoperative exercise testing has not been done uniformly. Postoperative imaging was not always by CT-angiography and for that reason accurate information of coronary artery patency is lacking.

We do not have sufficiently hard data to prove that all aberrant coronary arteries should be considered for surgical treatment.

Conflict of interest: none declared.

REFERENCES


slit-like ostium and intramural course are more important than the interarterial course. And of course, the right ventricle is not as dominant as the left one. We did both of the surgeries and the outcomes are the same, and we think that reimplanting them in the correct sinus only raises the risk of complications, such as a thrombus.

Dr Hörer: Do you think it is still valid to assume that the position of the coronary artery is the risk factor and not rather the origin of the coronary artery?

Dr Kooij: I think the intramural course and the slit-like ostium are way more important than the course, yes.

Dr M. Pozzi (Ancona, Italy): In our experience, which is much smaller than yours, we actually found two patients in whom there was a course of the coronary artery between the aorta and pulmonary artery. We found no intramural course. So translocation of the coronary artery was necessary. But there also was one instance in which we are not quite sure whether it is worth operating or not, and it is a situation where you have a single common origin of the right and left coronary artery. So a single ostium, and from that is the origin of the two coronary arteries with no intramural course. We have used a technique described by Frank Hanley: transecting the main pulmonary artery and reattaching it to the left pulmonary artery, plus a LeCompte manoeuvre. We have done it and the patient is okay but we are not quite sure whether it is such an effective procedure and just wondered if you have had any experience with that type of anatomy, or what would you do if you were faced with such kind of anatomy.

Dr Kooij: I’m sorry I didn’t understand the question properly.

Dr Brown: I think that maybe because of time, sorry, you can discuss that afterwards between you, if you like. Would that be okay? This is a very particular case.

Dr A. Dodge-Khatami (Jackson, MS, USA): Since you operate on asymptomatic patients with an anomalous right coronary, which means that you do it just on the basis of the imaging. Do you work up these patients with a stress test or perfusion imaging scan or anything?

The second question is: do you put these patients postoperatively on aspirin or any other type of medication?

Dr Kooij: Before the surgery some patients have undergone an exercise test. Almost every patient had an echo, and with a CT we could see whether there was an intramural course or a slit-like ostium based on that. So I think based on this we could say if the patient should be operated on or not.

Dr Dodge-Khatami: Did you put them on aspirin postoperatively?

Dr Kooij: We put them on Plavix for six months.

Dr O. Jaber (Leeds, UK): Some people suggest implanting defibrillators for these patients. Did you do any for your patients, defibrillators, ICDs?

Dr Kooij: No.

Dr Jaber: You had two patients who had VF immediately post-op.

Dr Kooij: They were in the ICU at the moment. It happened two and three hours after surgery based on a thrombus.

Dr Jaber: You don’t think they are at risk of VF?

Dr Kooij: No. If your ICU is good then they should call you and you have to repair.