Anomalous Left Circumflex Artery From the Right Coronary Cusp: A Benign Variant?

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ABSTRACT: Objective. To determine the incidence of angiographically evident coronary artery disease (CAD) of the anomalous left circumflex artery (ALCx). **Background.** Due to the acute angulation of its origin from the aorta and its retroaortic course, some have proposed an increase in CAD in these vessels. **Methods.** We performed a retrospective cohort study of all patients (n = 5205) presenting for left heart catheterization at the St Louis Veterans Affairs hospital from 2006-2012. Quantitative coronary angiography (QCA) was performed by two independent operators for all cases identified as having ALCx (n = 18). **Results.** We identified 18 patients (0.346%) with this anomaly. Sixteen of 18 cases (89%) had atherosclerosis involving the ALCx. QCA performed on the ALCx for all 18 patients resulted in a mean stenosis of 61% (range, 10%-100%). Seven out of 18 patients (39%) had obstructive CAD of the ALCx with the majority (n = 6; 86%) also having concomitant obstructive disease in the left anterior descending (LAD) or right coronary artery (RCA). In the 5 cases where the ALCx underwent intervention, the mean diameter stent was 2.25 mm, with a mean length of 15 mm, indicating that these vessels were small in caliber with focal disease. At a mean follow-up of 18 months for all patients, there was 1 non-cardiac death in the non-intervened group, with the 17 remaining patients (94.7%) free of major adverse cardiovascular events (target lesion revascularization, target vessel revascularization, or death). **Conclusions.** This study suggests that indeed this most common anomaly appears to be benign in that its unique origin and/or retroaortic course do not increase the occurrence of atherosclerosis relative to the LAD and RCA.

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Key words: coronary artery disease, anomalous left circumflex artery

Abbreviations
ALCx = anomalous left circumflex arteries
AP = anterior posterior
Cx = circumflex
DES = drug-eluting stent
LAD = left anterior descending
LAO = left anterior oblique
MACE = major adverse cardiac event
IRB = Internal Review Board
MI = myocardial infarction
PCI = percutaneous coronary intervention
RCA = right coronary artery
TLR = target lesion revascularization
TVR = target vessel revascularization
VA = Veterans Affairs

Anomalous origin of the circumflex coronary artery from the right sinus of Valsalva was first described by Antopol and Kugel in 1933[1] and is the most common congenital coronary variant, with prevalence at coronary angiography of 0.18%-0.67%,[2-6] and is usually considered "benign"[7] since it is not known to predispose individuals to sudden cardiac death. However, due to the acute angulation of its origin from the aorta and its posterior retroaortic course, some have proposed an increase in coronary artery disease (CAD) in these aberrant vessels.[8]

Most studies to date that have proposed a higher incidence of CAD in the anomalous left circumflex (ALCx) have not examined whether CAD was also present in the other coronary arteries concomitantly. Thus, it is unclear if anomalous circumflex arteries are particularly prone to atherosclerosis. In order to answer this question, we sought to characterize and analyze all patients who presented for coronary catheterization and were found to have this anomaly to determine whether these anomalous arteries actually have a higher atherosclerotic burden as compared to the patients' other coronary vessels.

Methods

This study was approved by the St Louis VA Healthcare Internal Review Board (IRB) under the regulations and guidelines set forth by the reviewing body. All coronary angiograms (n = 5205) from January 2006 to January 2012 performed at the St Louis Veterans Affairs Medical Center were reviewed for the presence of an anomalous circumflex artery. Comprehensive chart review to assess the clinical characteristics and outcomes of patients who were identified as having an ALCx was performed using the electronic medical records. The angiograms of these patients were analyzed and quantitative coronary angiography (QCA) of

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Table 1. Demographic and clinical presentation.

Baseline Characteristics	n = 18
Male	17 (94%)
Diabetes mellitus	8 (44%)
Hypertension	16 (89%)
Hyperlipidemia	15 (83%)
Tobacco use (current or past)	11 (61%)
Known coronary artery disease	9 (50%)

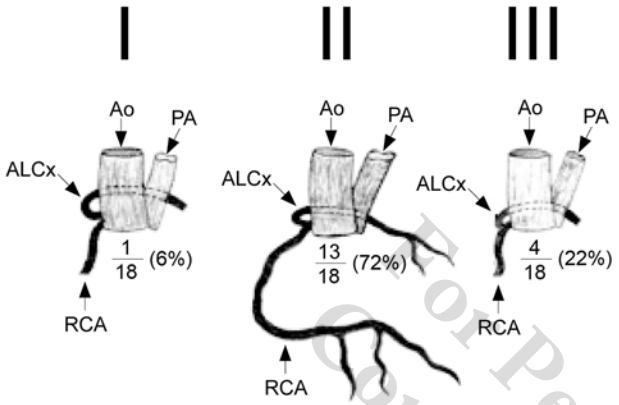

Figure 1. *Graphical 3-dimensional depiction of the 3 variants of ostia anatomy of the anomalous left circumflex artery (ALCX) with respect to the ascending aorta (Ao) and pulmonary artery (PA). There are three recognized variants of the origin of these anomalous vessels.*[8] *Type I is defined by a separate ostia of the circumflex and the right coronary artery (RCA), which we found in 1 patient. Type II is a common or adjacent ostia in the right coronary sinus, which represents the most common variant and was present in 68% of our cohort. Type III refers to the ALCX arising from the proximal aspect of the RCA and was present in 21% of our patients.*

the ALCx and any stenosis using validated software was performed by two independent operators. The course of the ALCx was specifically reviewed per the classification of Page et al[2] type (I-III). For those patients who underwent PCI, procedural details were also examined. Continuous variables were analyzed and reported as mean, median, range, and standard deviation. Student's t-test for continuous variables was used in comparison of data. A *P*-value of <.05 was considered significant.

Results

A total 18 patients with this anomaly were identified, with most patients with this variant being elderly (mean age, 63 years), male (94%), diabetic (44%), and hypertensive (89%) (Table 1). The classification of the ALCx in this cohort was determined according to anatomical variants of the ostium of the anomalous circumflex coronary artery: separate ostia for right coronary artery (RCA) and anomalous circumflex within the right sinus of Valsalva (type I), common/adjacent ostia in the right sinus (type II), and the anomalous circumflex arising as a branch of the proximal RCA (type III) (Figure 1). Within the studied cohort, the number of patients with one of the three variants was determined to be

Table 2. Clinical presentation when undergoing cardiac catheterization.

Indication for Angiography	n = 18
Abnormal stress test, asymptomatic	3 (17%)
Stable angina	6 (33%)
Acute coronary syndrome	9 (50%)
Known coronary artery disease	9 (50%)

Table 3. Characterization of percutaneous coronary intervention to anomalous left circumflex artery.

Guide Catheter	Lesion Length	Stent Diameter
6 Fr, IM	10 mm	2.25 mm
8 Fr, JR4*	5 mm; 5 mm	2.25 mm
6 Fr, AR1	22 mm	2.25 mm
8 Fr, AL0.75	30 mm	2.25 mm
8 Fr, MP	18 mm	2.25 mm

** Two drug-eluting stents placed at 2 discrete lesions.*

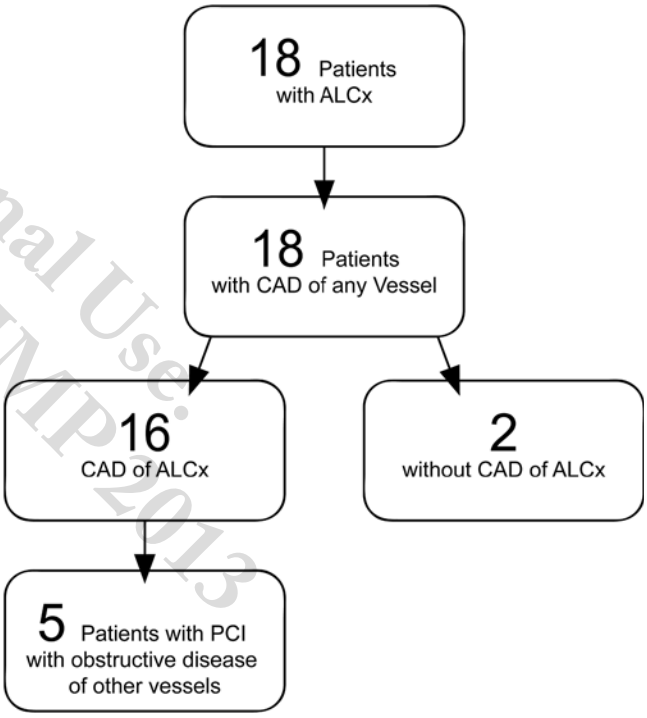

Figure 2. *Chart depiction of patient cohort. All patients had coronary artery disease, with 5 patients with an anomalous left circumflex undergoing percutaneous coronary intervention.*

1/18 (6%) for type I, 13/18 for type II (72%), and 4/18 for type III (22%). All anomalous vessels took a posterior retro-aortic course to the great vessels (aorta and pulmonary artery). Of the 5205 angiograms performed, there were no other anomalous coronary arteries besides the ALCx that took a retro-aortic course. The indication for angiography was positive functional study in 3/18 (17%) (with only 1 in the circumflex territory), stable angina in 6/18 (33%), or acute coronary syndrome (ACS) in 9/18 (50%) (Table

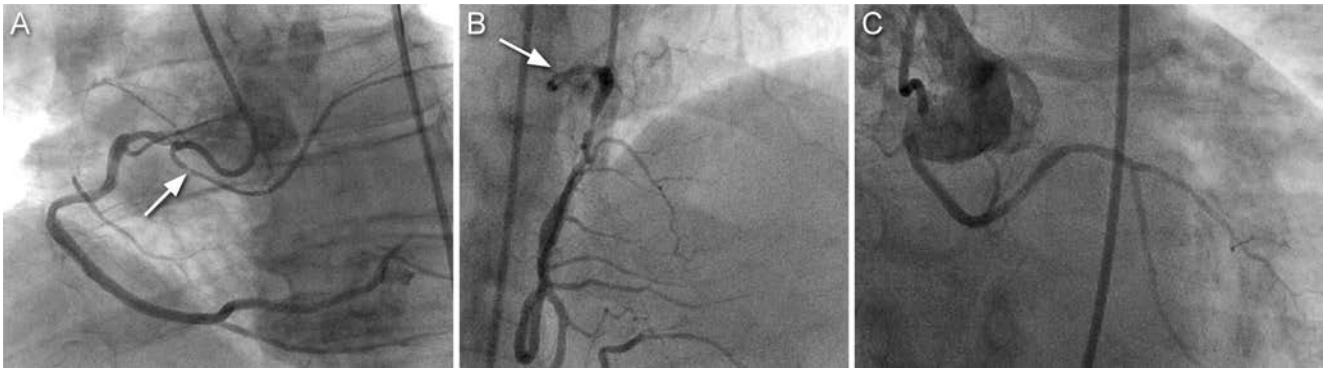

Figure 3. *Angiography of 3 different patients with anomolous left circumflex arteries (ALCX).* **(A)** *Angiography in a shallow left anterior oblique angulation depicting a type II ALX with the right coronary (RCA) and the ALCX sharing a common ostium. There is a discrete stenosis in the proximal aspect of the vessel (arrow), which did not resolve with the administration of intracoronary nitroglycerin.* **(B)** *Angiography from anterior-posterior cranial projection of the RCA with visualization of the retroaortic course of the ALCX with stenosis in the proximal aspect of the vessel (arrow).* **(C)** *Angiography from the left anterior oblique projection depicting a chronically occluded ALCx, which is filled via right ventricular marginal collaterals.*

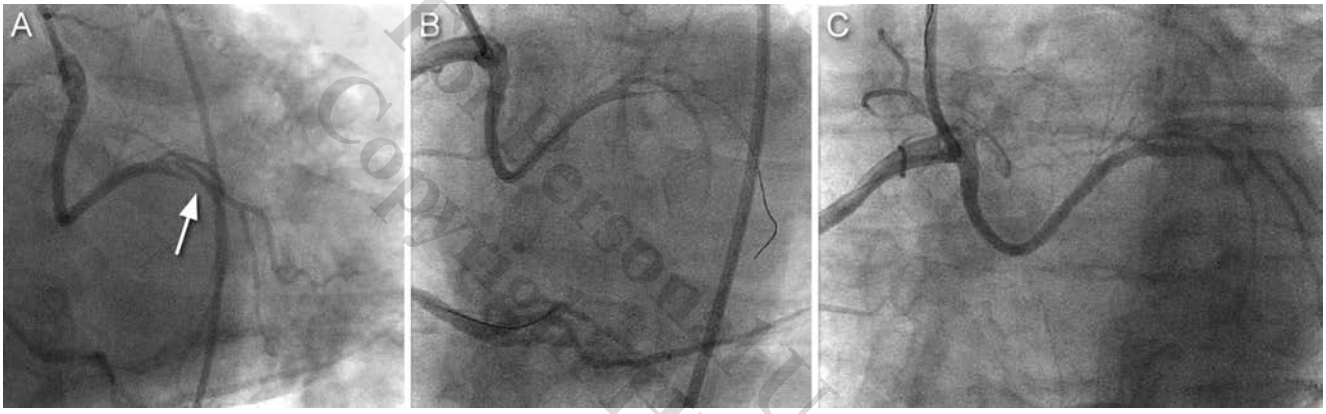

Figure 4. *Percutaneous coronary intervention of the anomalous left circumflex (ALCX).* **(A)** *Diagnostic angiography in the left anterior oblique projection revealing a severe stenosis of the ALCX (arrow).* **(B)** *Double-wire technique involving placement of a separate guidewire into the right coronary artery (RCA) to anchor the guide followed by wiring of the ALCx with the wire being obstructive within the lesion.* **(C)** *After placement of a 2.25 x 18 mm drug-eluting stent in the mid to distal vessel with the wire left in the RCA to permit anchoring and proper opacification of the target vessel.*

2). The circumflex was never the infarct-related artery in the ACS cases.

In the ALCx, CAD of any severity was present in the majority of patients, ie, 16/18 patients (89%), with a mean stenosis of 61% (range, 10%-100%). Of these patients with CAD involving the ALCx, the stenosis was >70% in only a minority of them (7/18 patients [39%], with 2 being chronic total occlusions [CTO]). Of those 7 patients with obstructive disease in the ALCx, the majority of them (6/7 cases; 86%) also had obstructive disease in the LAD or RCA requiring treatment. There was only 1 case where obstructive CAD was isolated to the ALCx. In the case wherein the ALCx was chronically totally occluded, there was concomitant non-obstructive disease in the other two coronary vessels. In the other CTO of the ALCx, there was obstructive disease in the LAD/RCA requiring intervention.

In the 5 patients who had obstructive CAD of the ALCx that was not a CTO, PCI was successfully performed without complications (Figure 2). These vessels were small with a mean diameter by QCA of 2.08 mm (range, 1.3-3.9 mm). The disease was relatively focal, with a mean lesion length of 15 mm. In the 5 cases where the ALCx underwent intervention, the obstructive lesion was located in the proximal and mid body of the vessel and never at the ostium. The mean stent diameter was 2.25 mm, with a mean length of 15 mm (Table 3). The other 13 patients, of which 11 had coronary disease of the ALCx, received medical therapy; at a mean follow-up of 18 months, there was 1 non-cardiac death in the medical management group, with the remainder of patients (n = 17) being free of major adverse cardiac events (MACE), defined as target lesion revascularization, target vessel revascularization, or death.

Discussion

Large case series identifying the frequency of anomalous circumflex arteries in a population have suggested that perhaps due to the unique retro-aortic position of this vessel, it is more prone to atherosclerosis. In this paper, we compiled one of the largest cohorts with this anomaly and sought to determine if indeed the ALCx had a higher incidence of

single-vessel CAD or whether CAD in the ALCx simply marked a population with multivessel disease. In this selective, mainly male veteran population with a high number of cardiac risk factors, it was found that atherosclerosis of the ALCx is common. However, in contrast to other studies,[8] we found in the majority of our patients that any atherosclerosis was actually present in the other two coronaries as well. In fact, only a single patient had isolated obstructive disease of the ALCx that happened also to be a CTO. In that isolated case, CAD was also concomitantly present in the other vessels.

Similar to large studies done on the ALCx, the majority of ALCx in this cohort originated from a common/adjacent ostium in the right coronary sinus and took a posterior course to the great vessels before supplying the posterolateral surface of the left ventricle. It is this retro-aortic posterior course that has been proposed to be a contributing factor in the development of atherosclerosis in the ALCx. The hypothesis behind this is that there is increased sheer stress at the ostium and proximal aspect of this vessel given its unique ostial angulation and retro-aortic course. When analyzed by location of disease, the majority of obstructive CAD involving the ALCx appears to be confined to the proximal to mid body of the vessel versus ostial, suggesting that the proposed increased sheer forces are not a factor. Our study suggests that this common anomalous vessel does not have a higher propensity to develop atherosclerosis. What is evident is that these vessels are generally small, with a mean vessel diameter of only 2.20 mm. We only found 3 patients with a vessel size greater than 2.75 mm.

PCI was successfully performed in 5 cases with obstructive disease involving the ALCx, and these stents were small, consistent with the small size of the vessel. The outcome at a mean of 18 months following cardiac catheterization for the 13 patients undergoing medical management and the 5 patients undergoing PCI showed low rates of MACE endpoints with similar survival rates in terms of deaths attributed to cardiac events, and both groups were free of MACEs.

Study limitations. This study is limited by its single-center, retrospective design. It includes only patients with a high pretest probability of diffuse CAD, which may differ from the general population. Our patients are older and systemically have a large atherosclerotic burden and all of our patients were referred for coronary catheterization for either symptoms of angina or had presented with ACS. Hence, a selection bias is present and this could explain why we found more diffuse CAD in our cohort. Therefore, it would be helpful to know more about asymptomatic patients who have ALCx and whether there is increasing incidence of atherosclerosis compared to the normal variant.

Conclusions

In our veteran cohort, we found that the ALCx is in fact a benign variant, with no increased incidence of atherosclerosis as was previously suggested in other studies. The ALCx is generally a very small artery with focal lesions that tend to be in the proximal and mid portions of the vessel, which is no different than the other coronary vessels. These patients did well, with a very low incidence of death or MACE endpoint.

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