Journal of Geriatric Cardiology (2015) 12: 66–75

Review **· Open Access ·**

Congenital coronary artery anomalies silent until geriatric age: non-invasive assessment, angiography tips, and treatment

Gianluca Rigatelli[1], Fabio Dell'Avvocata[1], Nguyen Van Tan[2], Rames Daggubati[3], Aravinda Nanijundappa[4]

[1]*Section of Adult Congenital and Adult Heart Disease, Cardiovascular Diagnosis and Endoluminal Interventions, Rovigo General Hospital, Rovigo, Italy*
[2]*Department of Cardiology, Thong Nhat Hospital, Ho Chi Minh City, Vietnam*
[3]*Brody School of Medicine at East Carolina University, Greenville, North Carolina, USA*
[4]*CAMC Vascular Center of Excellence, West Virginia University, Charleston, West Virginia, USA*

Abstract

Coronary artery anomalies (CAAs) may be discovered more often as incidental findings during the normal diagnostic process for other cardiac diseases or less frequently on the basis of manifestations of myocardial ischemia. The cardiovascular professional may be involved in their angiographic diagnosis, functional assessment and eventual endovascular treatment. A complete angiographic definition is mandatory in order to understand the functional effects and plan any intervention in CAAs: computed tomography and magnetic resonance imaging are useful non-invasive tools to detect three-dimensional morphology of the anomalies and its relationships with contiguous cardiac structures, whereas coronary arteriography remains the gold standard for a definitive anatomic picture. A practical idea of the possible functional significance is mandatory for deciding how to manage CAAs: non-invasive stress tests and in particular the invasive pharmacological stress tests with or without intravascular ultrasound monitoring can assess correctly the functional significance of the most CAAs. Finally, the knowledge of the particular endovascular techniques and material is of paramount importance for achieving technical and clinical success. CAAs represent a complex issue, which rarely involve the cardiovascular professional at different levels. A timely practical knowledge of the main issues regarding CAAs is important in the management of such entities.

J Geriatr Cardiol 2015; 12: 66–75. doi:10.11909/j.issn.1671-5411.2015.01.008

Keywords: Coronary artery anomaly; Congenital heart disease; Coronary artery angiography; Percutaneous coronary interventions

1 Introduction

Coronary artery anomalies (CAAs) occur in 0.64% to 5.6% of patients undergoing coronary angiography (Table 1).[1,2] Anatomic details and possible pathophysiologic patterns of most CAAs are sufficiently known today,[3,4] but they often continue to represent a challenge for cardiologists and in general for cardiovascular professionals. In this brief review, the authors have focused their attention on the management of various CAA subtypes focusing on their angiographic diagnosis, functional assessment and treatment.

Correspondence to: Gianluca Rigatelli, MD, PhD, Cardiovascular Diagnosis and Endoluminal Interventions Unit, Rovigo general Hospital, Viale Tre Martiri, 45100 Rovigo, Italy. E-mail: jackyheart71@yahoo.it
Telephone: +39-347-1912016 **Fax:** +39-425-394513
Received: September 15, 2014 **Revised:** October 27, 2014
Accepted: November 7, 2014 **Published online:** December 30, 2014

2 Noninvasive anatomical evaluation

Most CAAs are usually incidentally discovered on coronary artery angiography, but a significant number of patients may have clinical symptoms such as thoracic pain, angina pectoris, syncope, supraventricular or suspected ventricular arrhythmias, and even cardiac arrest, depending on the anatomo-morphologic abnormalities correlated with the pathophysiology underlying the specific anomaly. Some non-invasive technique would be helpful in rule out the diagnosis. In particular, Color-flow Doppler echocardiography can be useful to detect some specific subtypes of CAA, especially in the case of ectopic origin of the left main or right coronary artery and in case of coronary artery fistula.[5] MRI and CT imaging have recently been found to be very useful, non-invasive, and effective imaging techniques for detecting single coronary artery and its course,[6] and anomalous vessels originating from the opposite sinus.[7]

Table 1. Incidence of most common CAAs in the authors' experience and their possible clinical manifestations.

Coronary anomaly	Incidence, %	Possible clinical manifestations
Separated origin of LAD and LCx	0.31	-
Ectopic origin of the LCx from the RCA	0.25	-
Ectopic origin of the LCx from the right sinus	0.13	-
Myocardial bridge	0.11	UA, AMI, MA, SD
Ectopic origin of the LCA from right sinus	0.098	UA, AMI, MA, SD
Single coronary artery	0.098	SA, UA, SD
Atresic coronary artery	0.039	SI
Dual LAD IV type	0.039	-
Ectopic origin of the RCA from the left sinus	0.039	UA, AMI, MA, SD
Coronary artery fistula	0.039	HF, UA, AMI, SYC
Ectopic origin of RCA from the pulmonary artery	0.020	SD
Ectopic origin of LCA from the pulmonary artery	0.020	HF, SA,UA
Total	1.21	

AMI: Acute myocardial infarction, CAA: coronary artery anomaly; CAD: coronary artery disease; HF: heart failure; LAD: left anterior descending coronary artery; LCA: left coronary artery; LCx: left circumflex coronary artery; MA: malignant arrhythmias; RCA: right coronary artery; SA: stable angina; SD: sudden death; SYC: syncope; UA: unstable angina.

3 Angiographic identification: tips and tricks

Selective coronary angiography is the gold standard for the evaluation of CAAs and their anatomic and morphologic definition prior to any judgment. Differential diagnosis should be made with other coronary abnormalities such as atherosclerotic coronary ectasia and aneurysm, coronary lesions associated with Kawasaki and other arteritis, and coronary aneurysms and fistulas following coronary artery surgery.

3.1 Atresic coronary artery

It is normally correlated to congenital syndromes such as Rubeolla syndrome, Hurler's syndrome, Friederich's ataxia syndrome and normally its recognition is missed for an atherosclerotic occlusion.[8] Aortography in left anterior oblique or right anterior oblique is mandatory to confirm the diagnosis. The congenital incomplete development or stenosis of the coronary vessels, more often the left circumflex (LCx) or right coronary artery (RCA) and less frequently the left main (LM), is angiographically recognizable by the filling through the collateral circulation from the controlateral side.

3.2 Ectopic origin of the left circumflex from the right sinus or from right coronary artery

Ectopic origin of the LCx from the right sinus or from RCA represents a relatively frequent CAA (Figure 1). The preferred catheter in the case of ectopic origin from the right sinus may be the modified Judkins right or the right Amplatz. When the LCx arises from the right coronary cusp or the proximal RCA, it invariably follows a retroactive course, with the LCx passing posterior around the aortic root to its normal location. On the left anterior oblique, the LCx is seen originating from the proximal RCA. The left anterior descending coronary artery (LAD) is large without LCx. In a 30° right anterior oblique view, the LCx will be seen curving in the posterior area and is seen head-on, as a dot, posterior to the aorta.[9] When the LCx originates from the proximal RCA, near the ostium, if the catheter tip is engaged too deeply, it can pass the ostium of the anomalous LCx and it can miss to opacify the LCx.

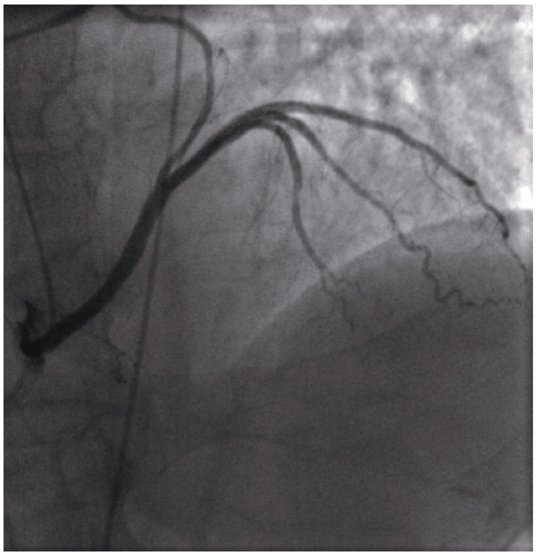

Figure 1. Ectopic origin of the left circumflex coronary artery from the proximal portion of the right coronary artery assessed from the femoral approach in a 78-year old female.

3.3 Ectopic coronary origin from the ascending aorta

Ectopic coronary origin from the ascending aorta is suspected when the angiographer is unable to locate a coronary artery within the sinuses of Valsalva and is normally benign, its presence being important only during cardiac surgery due to the danger of accidental cross-clamping or transecting.[10] Angiography of the aorta in left anterior oblique or right anterior oblique may define the diagnosis. Preferred catheters should be multipurpose, hook or cobra diagnostic catheter. In particular cases left or right saphen vein bypass catheter may be considered.

3.4 Ectopic origin of left coronary artery or right coronary artery from the opposite sinus

The left coronary artery (LCA) originates from the opposite sinus, there are four pathways: the interarterial course (rare), septal course (common), the retroaortic and the anterior courses. The course of an anomalous coronary artery is confirmed by the 30° right anterior oblique view. In this projection, a dot represents the artery seen on end. The interarterial course is diagnosed by the position of the "dot" anterior to the aorta; the retroaortic, by the "dot" behind the aorta; the septal is recognized by the fish hook picture in the right anterior oblique view, because the LM goes down to the septum then comes up to the epicardium, like a fish hook (Figure 2). Then the LCx would curve backward and from the "eye" with the LCx as the upper border.[11] In the anterior (pathway) the LM is in front of the pulmonary artery. This pathway is recognized by the "eye" with the LM as the upper border and the LCx as the inferior border. Selective coronary angiography with previous Swan-Ganz catheter placement in the main pulmonary artery does not really help to recognize interarterial passage. When the RCA arises from the left sinus or from the proximal LM (Figure 3), in the right anterior oblique view, the RCA will be seen head-on, as a dot anterior to the aorta. Eventual intramural passage of the anomalous vessel within the aortic wall is visualized in cranial left anterior oblique for RCA and in cranial right anterior oblique for LCA by left Judkins catheter or right Amplatz catheter, respectively.

In general, when the inter-arterial course of the anomalous vessel cannot be clearly ruled out, as frequently occurs due to the wide variety of passage of an anomalous vessel and specific cardiac anatomy,[12] coronary CT angiography is the most useful tools to visualized anomalous passage between aorta and pulmonary artery (Figure 3).

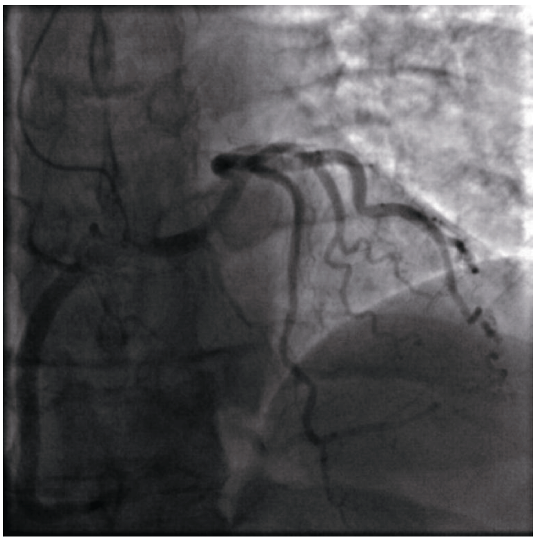

Figure 2. Sub-selective angiography from the radial approach of an ectopic origin of the left coronary artery from the right sinus in a 69-year old male.

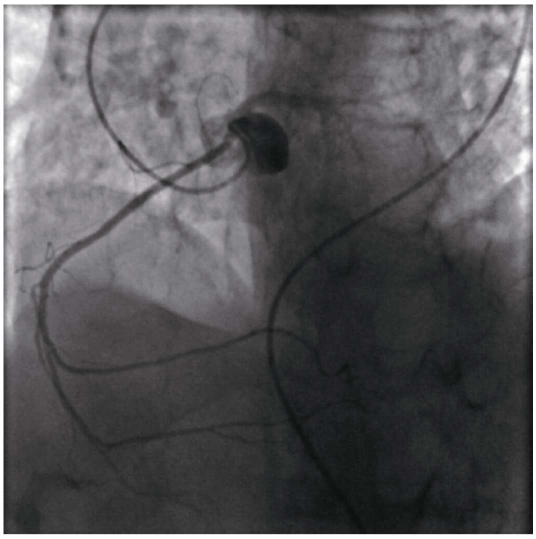

Figure 3. Selective angiography from the femoral approach of an ectopic origin of the right coronary artery from the left sinus in a 70-year old male with atypical chest pain and doubtful stress test.

3.5 Ectopic origin of left coronary artery or right coronary artery from the pulmonary artery

Ectopic origin of LCA or RCA from the pulmonary artery in the adult these anomalies may be suspected during standard examination when cannulation in the standard position is not possible.[13] Left and right pulmonary angiography should be considered in such cases.

3.6 Coronary artery fistula

Most of the fistulas originate from the first segment of the involved coronary artery and those located in the middle or distal segment of a coronary artery usually drain into the atrial or ventricular chambers (Table 2). Right and left ventricular angiography, selective cannulation of the fistula as

Table 2. Origins and drainage sites of coronary artery fistulas.

Artery of origin	Drainage sites
RCA	RV
LAD	PA
LCA	RA
LCx	LV
Diagonal	CS
OM	BV

BV: bronchial vein; CS: coronary sinus; LAD: left anterior descending coronary artery; LCA: left coronary artery; LCx: left circumflex coronary artery; LV: left ventricle; OM: Obtuse marginal artery; PA: pulmonary artery; RA: right atrium; RCA: right coronary artery; RV: right ventricle.

well as the assessment of right and left chambers pressures and Qp/Qs ratio are demanding for deciding an eventual transcatheter closure (Qp/Qs ≥ 1.5 is considered an indication to closure even in the absence of ischemia). Use of guiding catheter instead of diagnostic catheter may be of practical utility in order to properly opacifying large high-flow fistulas.

3.7 CAAs associated with other congenital heart diseases

Various types of CAAs are associated with other cardiac anomalies such as the tetralogy of Fallot,[14] the corrected transposition of the great arteries,[15] pulmonary atresia with intact ventricular septum,[16] criss-cross heart,[17] and many others. Obviously, it is fairly impossible to observe these congenital heart diseases reaching undiagnosed the adulthood. Occasionally, in operated adults coronary angiography might be required to assess eventual atherosclerosis: standard catheter are usually sufficient but CT may be useful in defining complex anatomy involving cardiac chamber, great vessels and coronary arteries and should considered the first choice imaging tools allowing for a global morphological study.

4 Functional assessment

The most frequent CAAs are usually clinically silent,[12] and are unrelated per se to myocardial ischemia and sudden death (Table 3), although they are often observed to have various degrees of atherosclerotic disease. Clinically, angiographic detection of a coronary artery anomaly has not led to any effective recommendations for functional testing. Exercise tests can be performed in order to reproduce symptoms or electrocardiographic signs or nuclear imaging patterns of induced myocardial ischemia, but give confusing results.[12] Standard clinical sub-maximal stress-test protocols are inadequate for assessing the prognosis of most CAAs, while 48 h Holter monitoring repeated every 6–12

Table 3. Anatomical and pathophysiological details of the most important CAAs.

CAA	Anatomy	Pathophysiology
Separated origin of LCx and LAD	LAD and Cx arise from adjacent separate ostia in the LS	No hemodynamic impairment
Ectopic origin of LCx from RS/RCA	The Cx arises from the RS or proximal RCA with posterior to the AO course	No hemodynamic impairment. Accidental compression during valve replacement
Ectopic coronary origin from the AO	Origin from the proximal 2 cm of the ascending AO	Accidental crossclamped or transected during surgery
Intercoronary communication	Contiguity of AV branches of RCA and Cx resulting in a bidirectional flow	May serve as collateral source in case of coronary obstruction
Dual LAD Type I	The short LAD gives the septal branches, the long LAD runs in the AIVS	
II	The long LAD descends on the right ventricular side before reentering the AIVS	No hemodynamic impairment
III	The long LAD travels intramyocardially in the ventricular septum	Misinterpretation during bypass surgery
IV	The long LAD arises from the RCA	
Atresic/hypoplastic coronary artery	Congenitally absent or hypoplastic Cx or LM	Fixed myocardial ischemia
Myocardial bridge	Intramyocardial tunneling of epicardial coronary segements	Fixed and episodic myocardial ischemia;
Coronary artery fistula	Vessel arising from coronary artery branch and draining in a single chamber	Fixed myocardial ischemia or ventricle overload
Single coronary artery R	Ostium in the right sinus	Potential compression of the single coronary vessel with episodic ischemia, myocardial infarction and sudden death
L	Ostium in the left sinus	
I	Anatomical course of normal right or left coronary artery	
II	Single vessel arising from the proximal part of RCA or LAD	
III	LAD and Cx arise separately form proximal normal RCA	
Ectopic origin of LCA from PA	Blood flows from the RCA, passes via collaterals to the LCA and flows to PA	Fixed and episodic ischemia. Volume overload
Ectopic origin of RCA from the PA	Flow from the LCA via collaterals into the RCA and retrograde into the PA.	Congestive heart failure.Sudden death
Ectopic origin of LCA from the RS	LCA arises from the RS and passes anterior or posterior to the aorta or between the AO and PA or intramurally.	Potential squeezing of the intramural portion of the vessel with ischemia or
Ectopic origin of RCA form the LS	RCA arises from the LS and passes between the AO and PA or posterior	sudden death (unlikely for RCA)

AIVS: anterior intraventricular sulcus; AO: ascending aorta; CAA: coronary artery anomaly; CAD: coronary artery disease; LAD: left anterior descending coronary artery; LCx: left circumflex coronary artery; LCA: left coronary artery; LM: left main; LS: left sinus; PA: pulmonary artery; RCA: right coronary artery; RS: right sinus; RV: right ventricle.

months, or better, a subcutaneous electrocardiogram loop recording implant might be more effective.[12,13] Finally, intravascular pressure wire, intravascular Doppler flow wire and intravascular echocardiography coupled with pharmacological stress tests should be a valid adjunct to the standard functional assessment of CAAs before planning any endovascular or surgical interventions.

Special cases. (1): Myocardial bridges are likely to be prone to myocardial infarction and sudden death:[18] coronary spasm or intravascular clotting is likely to be due to additional pathophysiological changes which are capable of exacerbating neurogenic or autocrine changes in the vessels. Investigation tools now available in the catheterization lab can help to clarify the clinical relevance of such anomalies. Intravascular ultrasound demonstrates that the phasic systolic vessel compression is coupled with a persistent diameter reduction (“half moon” phenomenon), while intracoronary Doppler reveals an increased flow velocity (early diastolic “finger tip” phenomenon), a retrograde systolic flow and a reduced coronary reserve.[18]

(2): Ectopic origin of LCA from the right sinus of Valsalva is differently significant depending on its origin and relationship with the aorta and pulmonary artery: “septal” subtype is the most common finding, while “between” one is rare but often dangerous.[19] Ectopic origin of the RCA from the left sinus of Valsalva, due to its route between aorta and pulmonary arteries and its occlusion during the expansion of the aorta, is a less dangerous anomaly.[20] The pathophysiology of these anomalies is complex: the proximal portion of the anomalous vessel often exits the aorta with an acute angle, creating functional or actual ostial stenosis. The proximal portion can also course between the aorta and pulmonary artery and can be impinged during exertion by pressure and volume expansion of the pulmonary artery against the aorta, causing perfusion mismatch; finally and probably most frequently, the proximal portion may course through the wall of the aorta resulting in narrowing of the lumen. Intravascular ultrasound interrogation during induced stress may be needed to assess risk of stress induced myocardial ischemia due to dynamic compression of the intramural segment of the anomalous vessel (Figure 4).[21]

5 Treatment notes

Surgery is preferred to correct the anomalous origin from the pulmonary artery,[22,23] and in cases of atresic coronary artery.[24] In all the other cases, the endovascular repair is considered when the CAA is involved in coronary atherosclerosis or when the anomaly per se causes ischemia or oveload as assessed by non-invasive tests.

5.1 Guiding catheters and wires

Percutaneous angioplasty of most CAAs subtypes remains a challenge for every interventional cardiologist. Unusual take-off angles, tortuosity, inability to engage the vessel with standard guiding catheter, and needs for more stiff guidewires often results in time consumption.

It is clear that major determinants for success are guiding catheter selection and placement of the balloon catheter within the very proximal portion of the anomalous vessel to facilitate guidewire advancement.[25] It may be useful keeping in mind some technical adjustments (Table 4).

There is a wide anatomical variety of anomalous vessel origin, which is difficult to organize in rigid scheme. Left Amplatz guiding or extra-back-up guiding catheters may be preferable in case of origin form the posterior sinus, whereas left or right vein bypass guiding catheters should be

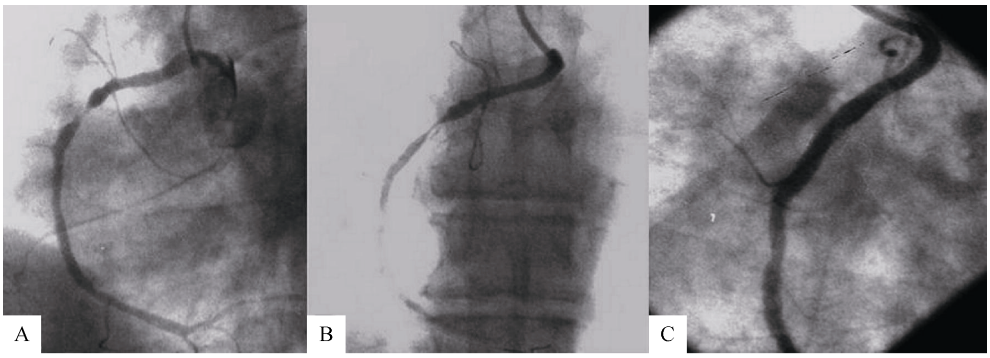

Figure 4. Challenging case of 81-year old patient with acute coronary syndrome and down-warded origin of the right coronary artery from the opposite sinus without intramural course (A); the ostium could not be cannulated correctly from the femoral approach with standard catheter, and it was cannulated through the radial artery approach with a Champ catheter (B); stenting with a drug eluting stent was accomplished from radial artery approach (C).

Table 4. Authors' experience technical tips and tricks to help endovascular treatment of some specific CAA subtypes.

CAA subtypes	Technical remarks
Origin from the thoracic aorta	Cobra, hook and multipurpose guiding catheter
LCx from right sinus	Left Amplatz, Judkins right and right Amplatz guiding catheters
LCx from RCA	Standard Judkins right, modified Judkins right and Champ 1-2 (Medtronic Corp.) guide catheters
	Extrasupport guidewire
	Balloon support
Origin from the opposite sinus	Left Judkins or Champ (Medtronic Corp.) guiding catheter for RCA
	Right or left Amplatz guiding catheter for LCA
	Moderately stiff guidewire: BMW heavy weight, Whisper MS or ES (Abbot corp.)
	IVUS guidance
	Drug-eluting stents in intramural segments of the anomalous vessel.
Single coronary artery	Moderately stiff guide wires: BMW heavy weight -Whisper MS or ES (Abbot corp.)
	Ballon support

CAA: coronary artery anomalies; IVUS: Intravascular ultrasound; LCA: left coronary artery; LCx: left circumflex; RCA: right coronary artery.

used in case of origin above the omolateral or controlateral sinus. Peripheral interventions guiding catheters such as the cobra or hook or renal angioplasty guiding catheters may be useful during percutaneous interventions in patients with ectopic high origin from the ascending aorta.

The ectopic origin of LCx from the right sinus of valsalva is the most frequent CAA. Since its retro-aortic course makes it relatively inaccessible to surgical revascularization, coronary angioplasty may be preferable.[26] Left Amplatz, Judkins right and right Amplatz guiding catheters have been suggested as the catheter of choice in the case of origin from the right sinus, whereas Judkins right, modified Judkins right and multipurpose guide catheters have been used in cases of origin from the right coronary artery. Use of extra-support guidewire and balloon support in order to advance the guidewire should be always considered.[27]

Challenging cases are represented by patients with ectopic origin from the opposite sinus: superimposed CAD has been reported to be suitable for endovascular treatment:[28] large guide catheters (left Amplatz No. 2–3), unusual types of guide catheter (Champ 1–2) and extra support guide wire should be considered in order to provide more back-up, to overcome the tortuosity and facilitate the lesion crossing (Figure 4).[29] Intravascular ultrasound (IVUS) guidance has been suggested as the main tool for successful angioplasty in such anomalies (Figure 5).[30] Treatment of the functional squeezing of the proximal intramural segment of the anomalous vessels has been suggested but need further investigations.

Critical technical issues remains the difficult cannulation and adequate back-up support with standard guide catheter due to the ectopic ostium which is usually juxtacommissural; the risk of aortic root dissection as a consequence of aggressive coronary dilation due to the hypoplasia of the intramural

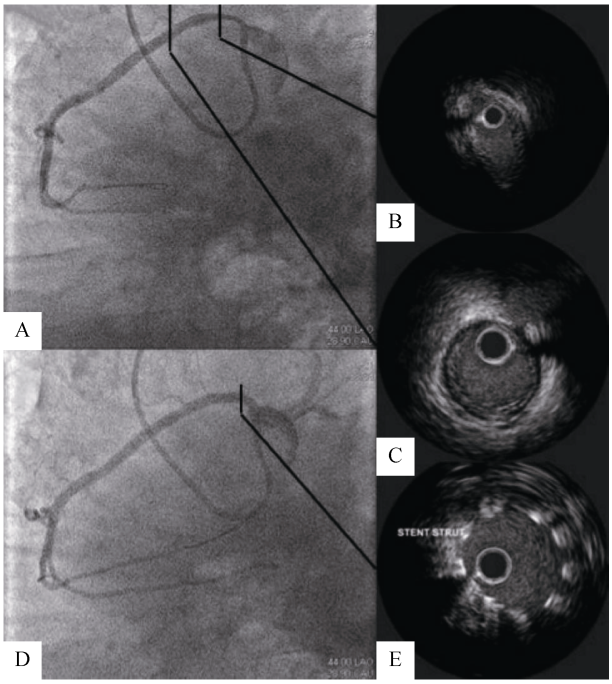

Figure 5. Acute coronary syndrome in a 67-year old patient with anomalous origin of the left main from the opposite sinus. (A): by injecting through a left Amplatz 2.0 6F guiding catheter an anomalous origin or the right coronary artery from the opposite sinus (ACAOS) with a tight stenosis of the proximal course of the vessel was apparent; (B): an IVUS examination revealed a 10-mm hypoplastic intramural course within the aortic wall of the proximal portion of the vessel, a phenomenon previously described as "intramural course"; (C): the around shaped structure of the coronary artery was absent in the proximal portion of the artery that presented an oval and laterally compressed appearance with a marked reduction of the vessel luminal area; (D) coronary angioplasty and stenting were accomplished by means of a 3.0-15 mm DES with angina and EKG resolution; (E): the IVUS control demonstrated good vessel apposition of the stent. DES: drug eluting stent; EKG: electrocardiogram; IVUS: intravascular ultrasound.

segment of the anomalous vessel; the absence of intimal thickening in the intramural segment in which there is no adventitia; and finally, the likelihood more frequent restenosis due to the phasic compressive forces which certainly persist in patients with ectopic origin from the opposite sinus, even after stent implantation. Probably surgical revascularization remains the recommended management, although the result of CABG, reimplantation of the ectopic vessel and translocation of the pulmonary artery in order to solve the problem of an aortopulmonary scissor remain less than ideal. Moderately, stiff guide-wire as the Whisper ME or ES or the BMW Heavy weight (Abbot Corp.) can allow for a more coaxial position of the guiding catheter, leading to a proper opacification of the vessel and helping the correct stent placement.[29]

Finally single coronary artery (Figure 6) with superimposed CAD still remains a challenge for endovascular specialists (Figure 7). Although successful endovascular management has been reported by many authors,[31,32] the amount of jeopardized myocardium involved in proximal lesions of the single coronary artery is in our opinion an absolute indication for surgical revascularization, whereas lesions of the distal segments or branches may be treated with angioplasty and stenting. Guiding catheter selection depends on specific cases. Moderately stiff guide wires are required to reach distal coronary segments and to allow stent-balloon system accessing to lesion.

5.2 Special considerations: myocardial bridges

Atherosclerosis was distinctly suppressed in the LAD

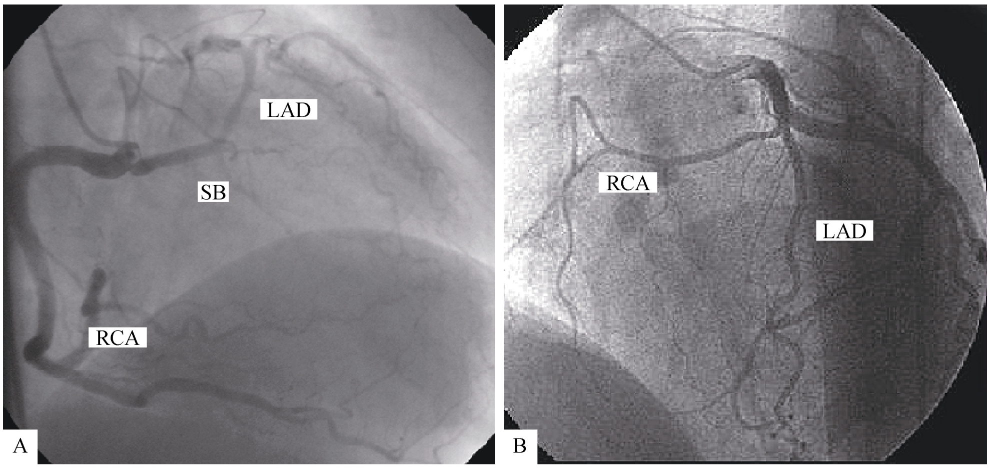

Figure 6. Single coronary artery. (A): origin of LCA from the RCA with septal course and absence of the LCx in a 70-year old female; (B): origin of RCA from the LAD with interarterial course confirmed by MRI in a 75-year old male. LAD: left anterior descending coronary artery; LCA: left coronary artery; LCx: left circumflex artery; RCA: right coronary artery; SB: side branch.

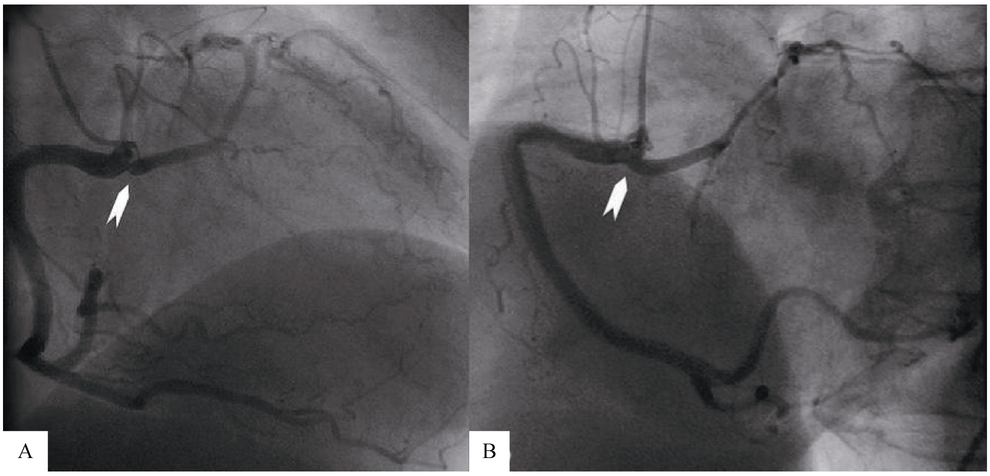

Figure 7. Challenging case of 65-year old patient with acute coronary syndrome and single coronary artery with an ostial stenosis of left coronary artery (A); good result of angioplasty and stenting (B).

intima beneath muscular bridges irrespective of their anatomic properties.[33] Nevertheless myocardial bridges that vigorously compress the LAD during cardiac systole due to physical exertion might cause life-threatening events even among superficially "healthy" individuals without coronary atherosclerosis.[34] In asymptomatic patients no intervention is warranted. Although medical therapy with beta-blockers has been shown to alleviate angina symptoms and ischemia, similar results have been achieved by nitrates or calcium channel blockers.[35] In the past, surgery has been proposed in cases of severe muscular bridge with or without CAD: minimally invasive coronary artery bypass grafting without cardiopulmonary bypass.[36] The implantation of endovascular stents in muscular bridges has been though to prevent external compression of the coronary artery by providing internal scaffolding and to normalize the hemodynamic alterations caused by the anomaly but had a very high restenosis rate of 36%,[37] also with first generation drug eluting stent (DES). Currently, patients with myocardial bridges deserve only medical therapy.

5.3 Special considerations: coronary artery fistula

Ischemia or overload-related symptom, are the main indications for treatment. In asymptomatic patients, all fistulas with moderate to severe shunting should be closed, since the potential long-term complications are serious.[38] Multiple origins such as a complex, plexiform and diffuse vascular anomaly resembling a hemangiomas, are preferably managed by surgery with surgical epicardial ligation or endocardial ligation,[39] which have a recurrence rate of 22.2% and 16.6%, respectively. Endovascular treatment is recommended in the case of fistulas originating from the proximal coronary artery segments as a conduit that later develops into a maze of capillary vessels and includes: stainless steel and platinum coils and vascular plugs. Interlocking detachable coils can be delivered through a 3F microcatheter coaxially introduced in a 5 or 6F guiding catheter by sizing the coil 10%–20% larger than the diameter of the vessel as measured when occluded (Figure 8). In high flow fistulas, temporary balloon occlusion may be necessary to allow proper coil delivery. Success is reported around 40%–92%,[40] and a comparison between endovascular coil embolization and surgical ligation showed similar early effectiveness, morbidity and mortality.[41] Use of the Amplatzer Duct Occluder and more recently the Amplatzer vascular plug has been reported to be effective in treating large coronary-cameral fistulas,[42,43] in which it is preferable to deliver umbrella device from the right ventricle in order to avoid coronary damage. Covered stent implantation to seal coronary fistulas has been reported in cases of concomitant

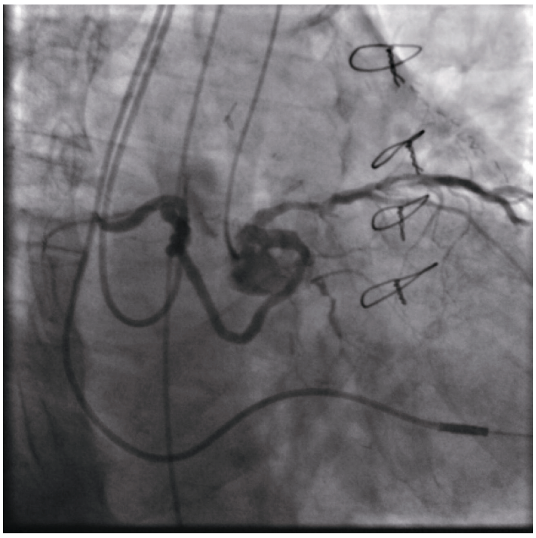

Figure 8. A coronary fistula with origin from the left stem, draining into the pulmonary artery in a patient with previous aorto-coronary bypass grafting in a 83-year old patient. The fistula was surgically excluded, because transcatheter embolization with an Amplatzer vascular plug failed because of the proximal tortuosity and calcification.

fistula and coronary stenosis in the same vessels segment but is not justified in the absence of atherosclerotic disease of the native vessel.[44]

In conclusion, CAAs represent a complex issue which rarely may involve cardiovascular professionals at different levels. A timely practical knowledge of the main issues regarding CAAs is important in the management of such entities.

Acknowledgement

None of the authors has any conflict of interest to declare.

References

1 Kardos A, Babai L, Rudas L, *et al*. Epidemiology of congenital coronary anomalies: a coronary arteriography study on a central European population. *Cathet Cardiovasc Diagn* 1997; 42: 270–275.
2 Angelini P Villanson S, Chan AV. Normal and anomalous coronary artery in humans. In *Coronary artery anomalies: a comprehensive approach*; Angelini P, Ed.; Lipincott Williams & Wilkins: Philadelphia, USA, 1999; 27–150.
3 Yamanaka O, Hobbs RE. Coronary artery anomalies in 126,595 patients undergoing coronary arteriography. *Cathet Cardiovasc Diagn* 1990; 21: 28–40.
4 Rigatelli G, Rigatelli A, Cominato S, *et al*. A clinical-angiographic risk scoring system for coronary artery anomalies. *Asian Cardiovasc Thorac Ann* 2012; 20: 299–303.

5 Nekkanti R, Mukhtar O, Nanda NC, *et al*. Transesophageal color Doppler three-dimensional echocardiographic assessment of left circumflex coronary artery fistula. *Echocardiography* 2002; 19: 573–575.

6 Ghadri JR, Kazakauskaite E, Braunschweig S, *et al*. Congenital coronary anomalies detected by coronary computed tomography compared to invasive coronary angiography. *BMC Cardiovasc Disord* 2014; 14: 81.

7 Clark RA, Marler AT, Lin CK, *et al*. A review of anomalous origination of a coronary artery from an opposite sinus of Valsalva (ACAOS) impact on major adverse cardiovascular events based on coronary computerized tomography angiography: a 6-year single center review. *Ther Adv Cardiovasc Dis* 2014 2014; 8: 237–241.

8 Pedon L, Rigatelli G, Zanchetta M, *et al*. Left main coronary artery atresia without induced ischemia. *Ital Heart J* 2000; 1: 769.

9 Serota H, Barth III CW, Seuc CA. Rapid identification of the course of anomalous coronary arteries in adults: The "dot and eye" method. *Am J Cardiol* 1990; 65: 891–898.

10 Rigatelli Gl, Rigatelli G. Coronary artery anomalies: what we know and what we have to learn. *Ital Heart J* 2003; 4: 305–310.

11 Nguyen T, Phuoc TT, Rigatelli GL. Angiographic views. In *Handbook of interventional cardiology*; Nguyen T, Ed.; Blackwell Publishing Inc.: New York, USA, 2003; 25–61.

12 Angelini P, Velasco JA, Flam S. Coronary anomalies. Incidence, pathophysiology, and clinical relevance. *Circulation* 2002; 105: 2449–2454.

13 Rigatelli GL, Docali G, Rossi P, *et al*. Angiographic Congenital coronary artery anomalies classification revisited. *Int J Cardiovasc Imag* 2003; 19: 361–366.

14 Dabizzi RP, Teodori G, Barletta GA, *et al*. Associated coronary and cardiac anomalies in the tetralogy of Fallot. An angiographic study. *Eur Heart J* 1990; 11: 692–704.

15 Dabizzi RP, Barletta GA, Caprioli G, *et al*. Coronary artery anatomy in corrected transposition of the great arteries. *J Am Coll Cardiol* 1988; 12: 486–491

16 Pasquali SK, Hasselblad V, Li JS, *et al*. Coronary artery pattern and outcome of arterial switch operation for transposition of the great arteries: a meta-analysis. *Circulation* 2002; 106: 2575–2580.

17 Angelini P, Lopez A, Lutechnowski R, *et al*. Coronary arteries in crisscross heart. *Tex Heart Inst J* 2003; 30: 208–213.

18 Bourassa MG, Butnaru A, Lesperance J, *et al*. Symptomatic myocardial bridges: overview of ischemic mechanisms and current diagnostic and treatment strategies. *J Am Coll Cardiol* 2003; 41: 351–359.

19 Ripley DP, Saha A, Teis A, *et al*. The distribution and prognosis of anomalous coronary arteries identified by cardiovascular magnetic resonance: 15 year experience from two tertiary centres. *J Cardiovasc Magn Reson* 2014; 16: 34.

20 Opolski MP1, Pregowski J, Kruk M, *et al*. Prevalence and characteristics of coronary anomalies originating from the opposite sinus of Valsalva in 8,522 patients referred for coronary computed tomography angiography. *Am J Cardiol* 2013; 111: 1361–1367.

21 Angelini P, Velasco JA, Ott D, *et al*. Anomalous coronary artery arising form the opposite sinus: descript features and pathophysiologic mechanism, as documented by intravascular ultrasonography. *J Invasive Cardiol* 2003; 15: 507–514.

22 Azakie A, Russell JL, McCrindle BWl. Anatomic repair of anomalous left coronary artery from the pulmonary artery by aortic reimplantation: early survival, patterns of ventricular recovery and late outcome. *Ann Thorac Surg* 2003; 75: 1535–1541.

23 Takeuchi S, Imamura H, Katsumoto K. New surgical method for repair of anomalous left coronary artery from pulmonary artery. *J Thorac Cardiovasc Surg* 1979; 78: 7–11.

24 Reul RM, Cooley DA, Hallman GL. Surgical treatment of coronary artery anomalies: report of a 37 1/2-year experience at the Texas Heart Institute. *Tex Heart Inst J* 2002; 29: 299–307.

25 Topaz O, DiSciascio G, Goudreau E, *et al*. Coronary angioplasty of anomalous coronary arteries: notes on technical aspects. *Cathet Cardiovasc Diagn* 1990; 21: 106–111.

26 Blanchard D, Ztot S, Boughalem K, *et al*. Percutaneous transluminal angioplasty of the anomalous circumflex artery. *J Interv Cardiol* 2001; 14: 11–16.

27 Nguyen T, Douglas J, Hermiller J, *et al*. Guides and wires. *J Interv Cardiol* 2001; 14: 113–123.

28 Ng W, Chow WH. Successful angioplasty and stenting of anomalous right coronary artery using a 6 French Left Judkins #5 guide catheter. *J Invasive Cardiol* 2000; 12: 373–375.

29 Rigatelli G, Cardaioli P. Endovascular therapy for congenital coronary artery anomalies in adults. *J Cardiovasc Med* (*Hagerstown*) 2008; 9: 113–121.

30 Hariharan R, Kacere RD, Angelini P. Can stent-angioplasty be a valid alternative to surgery when revascularization is indicated for anomalous origination of a coronary artery from the opposite sinus? *Tex Heart Inst J* 2002; 29: 308–313.

31 Kim D1, Jeong MH, Lee KH, *et al*. Successful primary percutaneous coronary intervention in a patient with acute myocardial infarction and single coronary artery ostium. *Korean Circ J* 2012; 42: 284–287.

32 Moustafa SE, Lesperance J, Gosselin G. Percutaneous coronary intervention on a single coronary artery: case report. *Int J Cardiol* 2009; 131: e118–e119.

33 Ishii T, Ishikawa Y, Akasaka Y. Myocardial bridge as a structure of "double-edged sword" for the coronary artery. *Ann Vasc Dis* 2014; 7: 99–108.

34 Ishikawa Y, Akasaka Y, Akishima-Fukasawa Y, *et al*. Histopathologic profiles of coronary atherosclerosis by myocardial bridge underlying myocardial infarction. *Atherosclerosis* 2013; 226: 118–123.

35 Ural E, Bildirici U, Celikyurt U, *et al*. Long-term prognosis of non-interventionally followed patients with isolated myocardial bridge and severe systolic compression of the left anterior

descending coronary artery. *Clin Cardiol* 2009; 32: 454–457.

36 Pratt JW, Michler RE, Pala J, *et al*. Minimally invasive coronary artery bypass grafting for myocardial muscle bridging. *Heart Surg Forum* 1999; 2: 250–253.

37 Kunamneni PB1, Rajdev S, Krishnan P, *et al*. Outcome of intracoronary stenting after failed maximal medical therapy in patients with symptomatic myocardial bridge. *Catheter Cardiovasc Interv* 2008; 71: 185–190.

38 Yuksel S, Yasar E, Nar G, *et al*. Prevalence and characteristics of coronary-cameral communications in adult patients: coronary angiographic analysis of 16,573 patients. *Med Princ Pract* 2014; 23: 336–339

39 Cheung DL, Au WK, Cheung HH, *et al*. Coronary artery fistulas: long-term results of surgical correction. *Ann Thorac Surg* 2001; 71: 190–195.

40 Raufi MA, Baig AS. Coronary artery fistulae. *Rev Cardiovasc Med* 2014; 15: 152–157.

41 Qureshi S, Reidy J, Alwi M, *et al*. Use of interlocking detachable coils in embolization of coronary arteriovenous fistulas. *Am J Cardiol* 1996; 78: 110–113.

42 Wang C, Zhou K, Li Y, *et al*. Percutaneous transcatheter closure of congenital coronary artery fistulae with patent ductus arteriosus occluder in children: focus on patient selection and intermediate-term follow-up results. *J Invasive Cardiol* 2014; 26: 339–346.

43 Wiegand G, Sieverding L, Bocksch W, *et al*. Transcatheter closure of abnormal vessels and arteriovenous fistulas with the Amplatzer vascular plug 4 in patients with congenital heart disease. *Pediatr Cardiol* 2013; 34: 1668–1673.

44 Balanescu S, Sangiorgi G, Medda M, *et al*. Successful concomitant treatment of a coronary-to-pulmonary artery fistula and a left anterior descending artery stenosis using a single covered stent graft: a cse report ans a literature review. *J Interv Cardiol* 2002; 15: 209–213.