Journal of Clinical & Experimental Cardiology

Rabe et al., J Clin Exp Cardiolog 2017, 8:6
DOI: 10.4172/2155-9880.1000525

Case Report Open Access

# Challenges of Anomalous Coronary Artery Intervention: Case Series and a Review

**Timothy G Rabe**[1], **Amol A Bahekar**[2*] **and Sameer Arora**[1,2,3]

[1]*Campbell University School of Osteopathic Medicine, Lillington, NC, USA*

[2]*Division of Medicine-Cardiology, Cape Fear Valley Medical Center, Fayetteville, USA*

[3]*Division of Cardiology, University of North Carolina at Chapel Hill, USA*

[*]**Corresponding author:** Amol A. Bahekar, Division of Interventional Cardiology, Cape Fear Valley Medical Center, Fayetteville Heart Center, Fayetteville, NC 28304, USA, Tel: (910) 484 4100; Fax: (910) 615 5681; E-mail: abahekar@yahoo.com

**Received date:** April 24, 2017; **Accepted date:** June 01, 2017; **Published date:** June 03, 2017



**Abstract**

Catheter-directed angiography remains the gold standard both for diagnosis and treatment of a coronary artery anomaly (CAA). CAA intervention continues to remains a complicated task even in the hands of experienced operators. The greatest challenges faced in the management of CAAs are delays in identification and difficulty engaging the anomalous coronary artery. Operator experience in promptly identifying the anomaly and selection of the appropriate catheter is critical for successful CAA intervention. We discuss a series of cases where CAAs were encountered during angiography and review the diverse technical difficulties faced by operators during interventions of these congenital anomalies.

**Keywords:** Anomalous; Angiography; Coronary intervention; Angioplasty

## Introduction

Congenital malformation of the coronary arteries not associated with other congenital cardiac defects is referred to as coronary artery anomalies (CAAs) [1]. Typically, the right coronary artery (RCA) originates from the right cusp of the aorta and the left main coronary artery (LM) originates from the left cusp of the aorta further giving rise to the left anterior descending (LAD) and left circumflex arteries (LCx) [2]. An anomaly would be defined as any coronary vessel that falls outside of the two ostium three main artery systems. CAAs are frequently clinically silent and are only recognized in presence of concomitant cardiac disease or are found as an incidental finding via imaging or during autopsy [3,4]. Although the prevalence of CAAs is estimated to be anywhere between 0.2-2.4% [1,5,6], a majority of studies investigating the prevalence of CAAs are retrospective and therefore suffer from a selection bias due to the requirement for an indication to perform diagnostic testing.

The diagnosis and treatment of CAAs is challenging. The use of stress tests has been found to lack consistency [7-9]. Although multislice computed tomography and magnetic resonance angiography have shown some benefit as an imaging modality in patients suspected to have CAAs [10,11], catheter-directed angiography continues to remain the gold standard due to its diagnostic and therapeutic benefits. Low index of suspicion and lack of technical expertise in managing CAAs can potentially complicate angioplasty and lead to serious complications [12-14]. We will discuss a series of cases where we encountered CAAs during angiography, discuss the technical challenges they posed during angioplasty, and outline measures applied by operators to tackle these complicated clinical situations.

## Coronary Artery Anomalies Case Series

**LCx from the proximal RCA:** A 44-year-old man with a past medical history of coronary artery disease (CAD), hypertension, COPD, presented to the emergency department with angina. Initial electrocardiogram (EKG) revealed normal sinus rhythm with non-specific T wave abnormalities in the lateral leads and serial troponin I values were 7.050, 10.9 and 29.7. The patient was anticoagulated and coronary angiography was planned the following day. An FL4 (Boston Scientific) catheter was used to engage the left coronary artery which demonstrated a patent LM giving rise to the LAD and ramus arteries, both with minimal disease. An FR4 (Boston Scientific) catheter was used to engage the RCA which gave rise to the posterior descending artery (PDA) and posterolateral branches (PLB), all with minimal disease. Surprisingly, the LCx was found to originate from the proximal RCA and was totally occluded at its proximal segment (Figure 1a). After careful assessment, we decided to use a multipurpose catheter to engage the RCA. Subsequently, the LCx lesion was crossed with a Whisper wire (Abbott Vascular) and a Runthrough wire (Terumo Interventional System). A buddy-technique was utilized to provide additional support. A 2.0 × 12 mm Emerge balloon was used to dilate the proximal LCx and the mid-LCx lesion. This was followed by the deployment of a 2.5 × 32 mm Promus Premier Stent (Boston Scientific) to the proximal LCx lesion (Figure 1b and 1c).

### Anterior displaced RCA ostium within the right cusp of the aorta

A 70-year-old man with a past medical history of hypertension, diabetes mellitus type 2 and CAD presented to the emergency department with substernal chest pain at rest. Three sets of serial troponin I measurements were within normal reference limits and EKG did not demonstrate any new ischemic changes. However, due to the non-resolving nature of chest pain at rest along with presence of multiple risk factors, coronary angiography was planned. Coronary

angiography revealed LM branched to form the LAD and LCx. There was a proximal LAD stent without restenosis. The rest of the LAD and the LCx were free of significant disease. An FR4 catheter was used to non-selectively engage the RCA which was found to have an anteriorly displaced take off in the right cusp. The dominant RCA had mild stenosis in the proximal segment and 80-90% stenosis in the mid-segment (Figure 2a). The anomalous take off of the RCA anterior to its typical position was difficult to engage selectively using FR4 catheter. A 0.75 AL guide was used to selectively engage RCA and the lesion was successfully crossed with a Runthrough wire. A whisper wire was placed in the PDA for additional support. Balloon predilation was accomplished with a 2.5 × 12 mm Emerge balloon and a 2.5 × 16 mm Promus Premier Stent was deployed in the mid-RCA (Figure 2b). Final angiography after post dilation with a 2.75 × 12 mm noncompliant balloon showed excellent angiographic result (Figure 2c). In addition to anterior take off, there was an acute angulation of the proximal RCA which made it extremely challenging to deploy balloon and stent in the mid-segment. Using an "aggressive" guide along with buddy wire technique made this intervention possible.

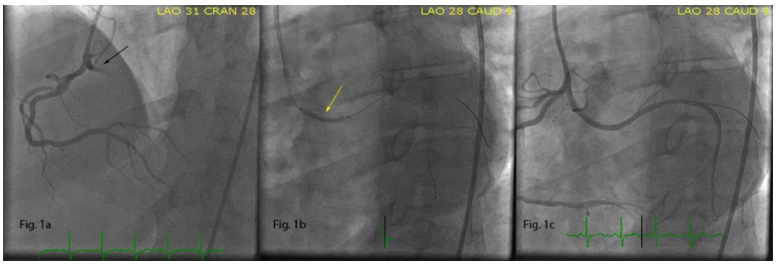


**Figure 1:** (a) Angiogram demonstrating a totally occluded anomalous LCx (black arrow) originating from the proximal RCA. (b) Angiography demonstrating balloon angioplasty and stent placement (yellow arrow) of the anomalous LCx with lead wires in a buddy wire technique. (c) Angiography showing a patent anomalous LCx post-intervention.

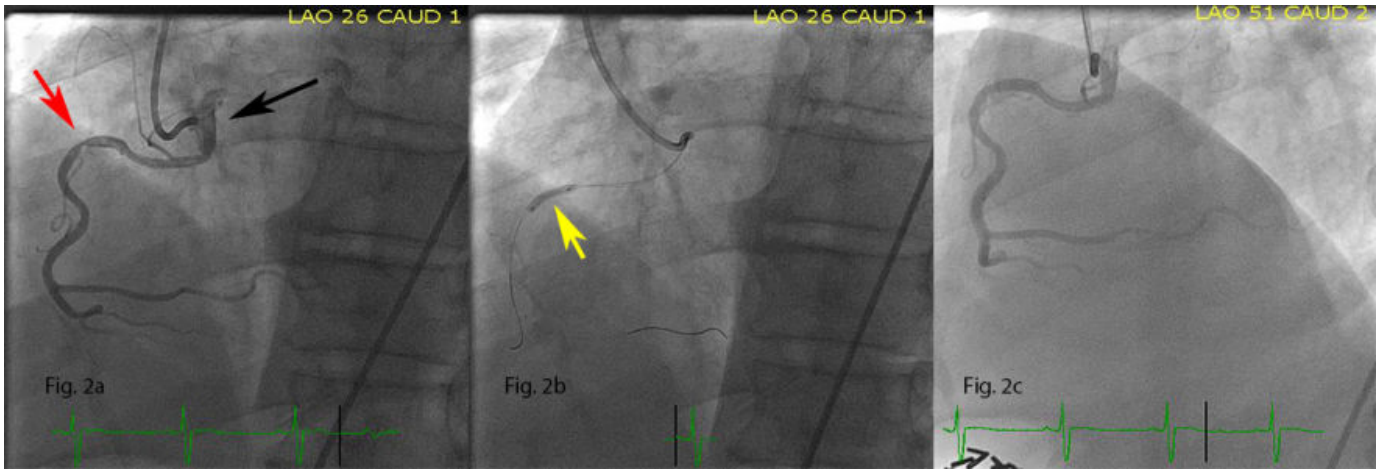


**Figure 2:** (a) Angiogram of RCA with tortuous anterior takeoff (black arrow) and a mid-segment RCA lesion with 80-90% stenosis (red arrow). (b) Balloon angioplasty and stent placement (yellow arrow) of the mid-segment RCA lesion. (c) Post-intervention angiogram demonstrates a patent mid-segment RCA.

## RCA originating in the left cusp of the aorta with a proximal RCA lesion

A 51-year-old male with a past medical history of CAD, HIV, and 25 pack year smoking history presented to the emergency department with exertional chest pain and shortness of breath. Initial troponin I was elevated at 0.051. The Radial artery was accessed with a 6 French Terumo Glidesheath and a TIG 4 (Terumo Interventional systems) catheter was used to engage the left and right coronary arteries. The LM was free of disease and branched into the LAD and the LCx. The mid-LAD had a focal area of stenosis of 90-95%. The LCx gave rise to a large obtuse marginal (OM)-1 artery with 80-90% stenosis and an OM-2 which was small and mildly diseased. The RCA was found to anomalously originate from the left cusp of the aorta. The proximal segment of the anomalous RCA demonstrated 70-80% stenosis and the mid-RCA was ectatic (Figure 3a). The distal RCA gave off the PDA and PLB branches which were mildly diseased. Since the RCA anomalously originated from the left cusp, we were unsuccessful in selectively engaging the RCA with AL 1, LBU 3, FL 3.5, or FL 4. Finally, a LBU 3.5 was used to engage the RCA and the RCA lesion was crossed with a Runthrough wire. A choice PT extra used in buddy wire fashion allowed for additional support during the proximal RCA intervention. The RCA was dilated using a 2.5 × 12 mm Emerge balloon and a 3.0 × 20 mm Promus Premier Stent was delivered to the proximal RCA. Post dilation of the stent was accomplished using a 3.0 × 15 non-compliant

balloon leading to good angiographic result (Figure 3b). This case demonstrates using left coronary guide to intervene anomalous RCA originating from left coronary cusp.

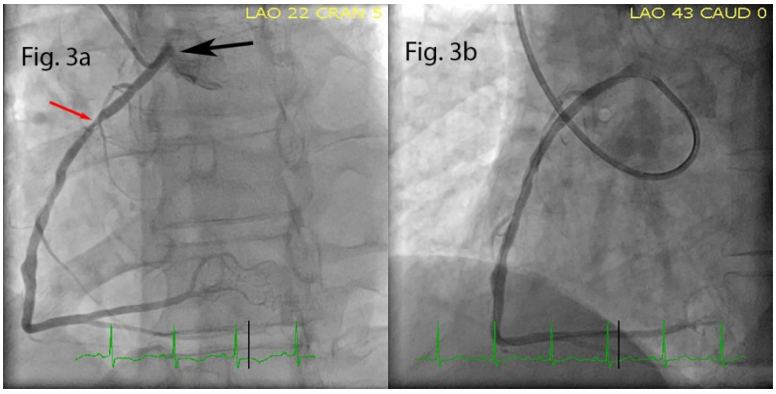


**Figure 3:** (a) Angiogram demonstrating an anomalous RCA originating from the left cusp of the aorta (black arrow) and 70-80% lesion in the proximal RCA (red arrow). (b) The anomalous RCA following balloon angioplasty and stent placement in the proximal segment.

## RCA from the left cusp of the aorta with a lesion of the mid-RCA

A 66-year-old female with past medical history of hypertension presented to the emergency department with exertional chest pain. Troponin I measurements were normal. EKG was unremarkable. Nuclear stress test demonstrated moderate amount of ischemia in the RCA territory after which a coronary angiogram with a radial artery approach was planned. The right radial artery was accessed with a 6 French catheter. The LM was patent and free of disease. The proximal LAD was patent, mid and distal LAD segments were mild to moderately diseased. The LCx was a dominant vessel with mild diffuse disease. RCA was found to originate from the left cusp of the aorta with a 99% mid segment lesion (Figure 4a). Given previous experience, we decided to use LBU guide for this intervention. The RCA was engaged with a LBU 3 guide. The lesion was crossed with a run through wire. A choICE PT Extra support wire was placed in the PDA. The run through wire was subsequently advanced in the large acute marginal branch. Balloon angioplasty of the acute marginal branch was performed using a 2.5 × 12 mm Emerge balloon. Angioplasty of the RCA was performed using a 2.5 × 12 mm Emerge balloon, followed by deployment of a 2.25 × 20 mm Promus Premier Stent in the mid-RCA. A 2.25 × 15 mm Noncompliant balloon at 16 atm of pressure was used for postdilation (Figure 4b and 4c). This case again demonstrates successful use of left coronary guide along with "buddy wire" technique to intervene RCA with anomalous take off from left coronary cusp.

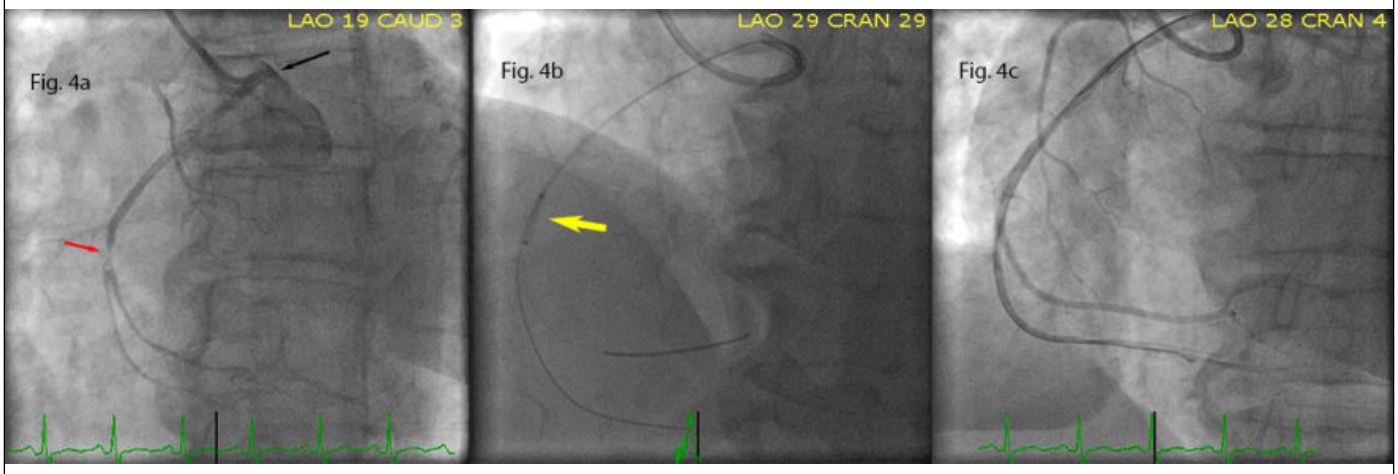


**Figure 4:** (a) Angiogram demonstrating RCA originating from the left cusp of the aorta (black arrow) with a 99% mid-segment lesion (red arrow). (b) Balloon angioplasty of the anomalous RCA mid-segment lesion (yellow arrow). (c) Post-intervention angiogram demonstrating a patent mid-segment anomalous RCA.

## Discussion

Catheter-directed angiography remains the diagnostic procedure of choice to identify CAAs. The factors which complicate angioplasty in the setting of CAAs include a delay in identification of the anomaly, location of the coronary ostium within the aorta, the proximal course of the vessel and angulation of the vessel [12,15]. Our case series describes four cases where we encountered CAAs during angiography and illustrate the technical difficulties faced by the operators. Furthermore, we reviewed similar existing cases in the literature and the potential complications associated with different types of CAAs.

The first case demonstrated an anomalous origin of a totally occluded LCx from the proximal RCA. The low index of suspicion and difficulty in engagement of the LCx were the biggest hurdles in this case. Although the FR4 catheter was unsuccessful in engagement of anomalous LCx due to shape and location of the ostium, a timely change to a multipurpose catheter led to a successful cannulation. The use of buddywire technique permitted reduction of the curve and the tortuosity. Rivitz et al. [16] utilized a JR4 guiding catheter and Kimbris et al. [17] used a modified Judkins catheter with a heat gun to point the tip of the catheter to the right to successfully engage the anomalous LCx. In a series of cases reporting anomalous origin of LCx from RCA, Blanchard et al. used a multipurpose catheter in four out of five cases, along with additional guidewire support in three of the cases [18]. The use of multipurpose catheter comes with a high risk of dissection both of the ostium of the LCx and the RCA ostia and decannulation before contrast injection control has been recommended [18].

The second case showed an anteriorly displaced right ostium with angulation of the proximal RCA. Multiple stabilization techniques including a Run through buddy wire and whisper wire anchored in the PDA had to be employed to cross the mid-RCA lesion due to the acute angulation. The ostia of the coronary arteries may be situated anterior, posterior, or superior to their typical location within the aortic cusps; which may lead to interference by aortic valve leaflets or difficulty locating the ostium in the wall of the proximal aorta. A displaced RCA ostium is difficult to visualize, engage and revascularize; leading to prolonged procedure times, increased fluoroscopic exposure, and high failure rates during revascularization [19]. The various catheter stabilization techniques used in order to cannulate CAAs require operator proficiency and may increase complications during PCI [19-21]. In cases where catheter seating and backup support of the catheter is necessary to cannulate the vessel, calculation of the angles and length of the vessel during angioplasty has been suggested to be beneficial [12]. Calculating vessel parameters can help the operator choose the appropriate catheter, thereby avoiding the use of multiple catheters and limiting the amount of instrumentation required during the procedure [12].

The third and fourth cases illustrated the anomalous origin of RCA from the left cusp of the aorta. Both cases demonstrated successful use of left coronary catheter to intervene right coronary artery. The course and angulation of the anomalous RCA and suboptimum catheter guide support made it difficult to cannulate the lesion and restore blood flow. Various strategies including use of Amplatz left guiding catheter [22,23], Voda guiding catheter [24], oversized and undersized Judkins left guided catheter [25,26] has been utilized in the past to successfully intervene the RCA arising from the left cusp. Similarly, we used left coronary catheters to successfully intervene the RCA with no complications. Although this anomaly is usually benign, fatalities are not unknown and have largely been attributed to the mechanical compression of anomalous RCA between the aorta and the pulmonary artery [15]. However, the incidence of sudden death in asymptomatic patients with this anomaly is extremely low [27]. Therefore, anomalous origin of RCA from the left cusp can be followed without intervention, with recommendations for exercise restriction [27]. This is in contrast to the anomalous origin of LCA from the right cusp which carries a higher risk of adverse complications, including sudden cardiac death and therefore, is largely managed surgically [15,27].

Non-coronary interventions have also been shown to produce complications in patients with occult CAAs. Speziale et al. reported a case of a subtotal occlusion of an anomalous LCx leading to lateral myocardial wall infarction after a mitral valve repair [28]. Morimoto et al. reported a unique case of a left coronary artery originating from the posterior aortic sinus which was compromised from a new prosthetic aortic valve due to the increased pressure in the aortic root from the newly implanted valve [29]. An interesting report by Daher et al. investigating the occurrence of sudden cardiac arrest during anesthesia induction revealed an anomalous origin of the LCA from the right coronary cusp coursing behind the aorta near the left atrium during angiography [30].

CT and cardiac magnetic resonance (CMR) imaging have emerged as important diagnostic modalities in detection and classification of CAA. Gated multislice CT (MSCT) provides high spatial resolution and the capability of multiplanar reconstructions and therefore, facilitates the delineation of the precise course of all three coronary arteries and allows for correct classification in the anatomical classification system of coronary artery anomalies [31]. Conversely, CMR assists in the evaluation of cardiac morphology, myocardial tissue properties and myocardial function. In an interesting case, CMR demonstrated an inferoseptal defect in a patient with persistent exertional angina and normal exercise nuclear perfusion study [32]. Coronary CT demonstrated an anomalous origin of RCA. Stenting of the proximal RCA led to resolution of symptoms and normalization of CMR finding. Ripley et al. demonstrated the utility of cardiovascular magnetic resonance in their study to detect 172 CAAs over a 15 year period [33]. All of the detected cases were able to be anatomically classified based on the CMR images. These additional imaging modalities could be used in order to preemptively identify a CAA before performing angiography and may reduce procedural complication because the operator would be aware of the CAA.

To conclude, prompt identification and engagement of the anomalous artery are the biggest hurdles to the operator in the management of CAAs. When combined with operator experience, angioplasty offers a safe therapeutic option for treatment of anomalous coronary arteries.

## Author Contributions

TG Rabe: Mr. Rabe had full access to all of the data in the study and takes responsibility for the integrity of the data and the accuracy of the data. Mr. Rabe contributed to the conception and design of the study and the manuscript drafting.

AA Bahekar: Dr. Bahekar was the operator of all the cases. Dr. Bahekar contributed to the manuscript drafting and critical revision of the manuscript.

S Arora: Dr. Arora had full access to all of the data in the study and takes responsibility for the integrity of the data and the accuracy of the data. Dr. Arora contributed to the conception and design of the study, the manuscript drafting, and the critical revision of the manuscript.

## Conflict of Interest Disclosures

T Rabe: Mr Rabe reports no conflict.

AA Bahekar: Dr Bahekar reports no conflict.

S Arora: Dr. Arora reports no conflict.

## References

1. Lluri G, Aboulhosn J (2014) Coronary arterial development: A review of normal and congenitally anomalous patterns. Clin Cardiol 37: 126-130.
2. Lipton MJ, Barry WH, Obrez I, Silverman JF, Wexler L (1979) Isolated single coronary artery: Diagnosis, angiographic classification, and clinical significance. Radiology 30: 39-47.
3. Eckart RE, Scoville SL, Campbell CL, Shry EA, Stajduhar KC, et al. (2004) Sudden death in young adults: A 25-year review of autopsies in military recruits. Ann Intern Med 141: 829-834.
4. Temperikidis P, Xanthos T, Stroumpoulis K, Koulouris S, Gavaliatsis I, et al. (2013) Primary congenital coronary artery anomalies: An angiographic study in greece. Hospital Chronicles 8: 171-178.
5. Mohsen GA, Mohsin KG, Forsberg M, Miller E, Taniuchi M, et al. (2013) Anomalous left circumflex artery from the right coronary cusp: A benign variant? J Invasive Cardiol 25: 284-287.
6. Yurtdas M, Gulen O (2012) Anomalous origin of the right coronary artery from the left anterior descending artery: Review of the literature. Cardiol J 19: 122-129.
7. Angelini P (2007) Coronary artery anomalies: an entity in search of an identity. Circulation 115: 1296-1305.
8. Basso C, Maron BJ, Corrado D, Thiene G (2000) Clinical profile of congenital coronary artery anomalies with origin from the wrong aortic sinus leading to sudden death in young competitive athletes. J Am Coll Cardiol 35: 1493-1501.
9. Waller BF (1985) Exercise-related sudden death in young (age less than or equal to 30 years) and old (age greater than 30 years) conditioned subjects. Cardiovasc Clin 15: 9-73.
10. Gharib AM, Ho VB, Rosing DR, Herzka DA, Stuber M, et al. (2008) Coronary artery anomalies and variants: technical feasibility of assessment with coronary MR angiography at 3 T. Radiology 247: 220-227.
11. Ten Kate GJ, Weustink AC, de Feyter PJ (2008) Coronary artery anomalies detected by MSCT-coronary angiography in the adult. Neth Heart J 16: 369-375.
12. Prashant PU (2012) Successful angioplasty of anomalous coronary arteries with total occlusions. J Invasive Cardiol 24: 228-232.
13. Saito T, Noguchi KF, Oikawa T (2011) Iatrogenic dissection of the anomalous-origin right coronary artery and left sinus of valsalva. J Invasive Cardiol 23: E51-E53.
14. Spokoyny I, LaBarbera M, Trilesskaya M (2012) Anomalous origin of the right coronary artery with intramural aortic course causing exercise-induced cardiac arrest. J Invasive Cardiol 24: 622.
15. Greet B, Quinones A, Srichai M, Bangalore S, Roswell RO (2012) Anomalous right coronary artery and sudden cardiac death. Circ Arrhythm Electrophysiol 5: E111-E112.
16. Rivitz SM, Garratt KN (1989) Stenotic anomalous circumflex artery causing myocardial infarction following angioplasty of a right coronary artery stenosis. Cathet Cardiovasc Diagn 17: 105-108.
17. Kimbiris D, Lo E, Iskandrian A (1987) Percutaneous transluminal coronary angioplasty of anomalous left circumflex coronary artery. Cathet Cardiovasc Diagn 13: 407-410.
18. Blanchard D, Ztot S, Boughalem K, Ledru F, Henry P, et al. (2001) Percutaneous transluminal angioplasty of the anomalous circumflex artery. J Interv Cardiol 14: 11-6.
19. Graidis C, Dimitriadis D, Karasavvidis V (2012) Guideliner catheter facilitated percutaneous coronary artery intervention of an anteriorly displaced and heavily calcified right coronary artery. J Invasive Cardiol 24: E222-E224.
20. Devidutta S, Sharma G, Jagia P (2013) Coronary stenting in a patient with single coronary artery and double-vessel disease! J Invasive Cardiol 25: E147-50.
21. Thomas JA, Patel J, Latif F (2011) Successful coronary intervention of circumflex artery originating from an anomalous left main coronary artery using a novel support catheter: A case report and review of literature. J Invasive Cardiol 23: 536-539.
22. Charney R, Spindola-Franco H, Grose R (1993) Coronary angioplasty of anomalous right coronary arteries. Cathet CardiovascDiagn 29: 233-235.
23. Yip H, Chen MC, Wu CJ (2001) Primary angioplasty in acute inferior myocardial infarction with anomalous-origin right coronaryarteries as infarct-related arteries: Focus on anatomic and clinical features, outcomes, selection of guiding catheters and management. J Invasive Cardiol 13: 290-297.
24. Praharaj TK, Ray G (2001) Percutaneous transluminal coronary angioplasty with stenting of anomalous right coronary artery originating from left sinus of Valsalva using the Voda guiding catheter: a report of two cases. Indian Heart J 53: 79-82.
25. Cohen MG, Tolleson TR, Peter RH, Harrison JK, Sketch MH (2002) Successful percutaneous coronary intervention with stent implantation in anomalous right coronary arteries arising from the left sinus of valsalva: A report of two cases. Catheter Cardiovasc Interv 55: 105-108.
26. Ng W, Chow WH (2000) Successful angioplasty and stenting of anomalous right coronary artery using a 6 French Left Judkins #5 guide catheter. J Invasive Cardiol 12: 373-375.
27. Gersony WM (2007) J Am Coll Cardiol 50: 2083-2084.
28. Speziale G, Fattouch KF, Ruvolo GF, Fiorenza GF, Papalia UF, et al. (1998) Myocardial infarction caused by compression of anomalous circumflex coronary artery after mitral valve replacement. Minerva cardioangiol 46: 455-456.
29. Morimoto N, Okita Y, Okada K, Yamashita T, Matsumori M (2005) Aortic valve replacement for a case of anomalous origin of the left coronary artery from posterior sinus of valsalva with intramural aortic course. J Thoracic and Cardiovasc Surg 130: 1713-1714.
30. Daher M, Zanatta AR, Henz BD, da Silva MC, dos Santos SN, et al. (2012) Sudden cardiac arrest in general anesthesia as the first manifestation of anomalous origin of the left coronary artery. Rev Bras Anestesiol 62: 878-884.
31. Heermann P, Heindel W, Schülke C (2017) Coronary Artery Anomalies: Diagnosis and Classification based on Cardiac CT and MRI (CMR) - from ALCAPA to Anomalies of Termination. Rofo 189: 29-38.
32. Mikolich JR (2009) Cardiac magnetic resonance imaging and coronary computed tomography angiography in the diagnosis of anomalous coronary artery. J Am Coll Cardiol 53: 456.
33. Ripley DP, Saha A, Teis A, Uddin A, Bijsterveld P, et al. (2014) The distribution and prognosis of anomalous coronary arteries identified by cardiovascular magnetic resonance: 15 year experience from two tertiary centres. J Cardiovasc Magn Reson 16: 34.