

## Anomalous right coronary artery arising from left coronary cusp with coexisting valvular heart disease

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### Abstract

We report an unusual case, a 50 year old female with an abnormal right coronary artery originating from the left coronary cusp. The patient, who had a history of hypertension presented with chest pain and shortness of breath to the emergency department. She was diagnosed with ischaemic heart disease (IHD) and had hypertension as one of the coronary risk factor. Echocardiography revealed poor progression of R waves. She was scheduled for echocardiography thereafter which revealed severe aortic stenosis with aortic root dilatation. The patient was discharged due to absence of any complications or other anomalies. This case is unique because of the simultaneously presenting valvular pathology, along with the anomalous origin of the right coronary artery which was detected, as an incidental finding, during coronary angiography.

**Keywords:** Right coronary artery, Left coronary cusp, Aortic stenosis.

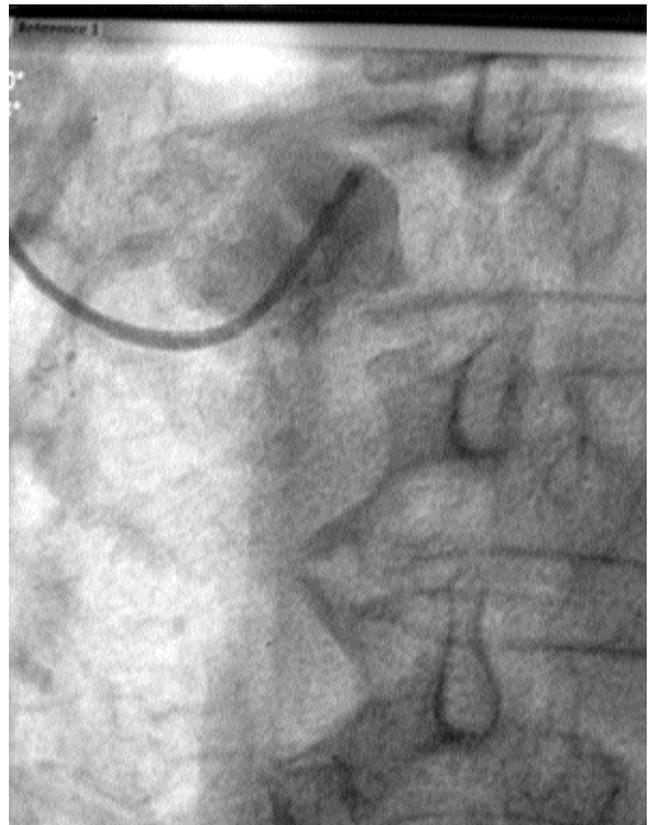
### Introduction

Coronary artery anomalies present rarely in the clinical setup. As few as about 1.3% of all the people undergoing coronary angiography are reported to have anomalous coronary arteries.<sup>1</sup> They are almost always asymptomatic and are found during a cardiac catheterization, cardiac surgery or autopsy.<sup>2</sup> While generally being clinically silent, they may sometimes present as arrhythmias, myocardial infarction, syncope or most commonly as sudden cardiac death (SCD) without any significant coronary artery disease.<sup>3</sup> Herein, we describe the case of a patient in whom coronary angiography was performed to evaluate the cause of chest pain and shortness of breath which then revealed anomalous right coronary artery arising from the left coronary cusp. We suggest that the young interventional cardiologists take a note of this aberration since it becomes quite difficult to engage coronary

arteries and poses a risk for rupture.

### Case Presentation

A 50-year-old hypertensive woman without previous angina history presented to the emergency department with chest pain and shortness of breath. She described the pain as continuous, mild to moderate in intensity (NYHA- III) with no radiation to adjacent sites. The patient denied any history of profuse sweating. Shortness of breath was associated with exertion, orthopnoea, productive cough and palpitations. She was suspected to have Ischaemic Heart Disease (IHD). She was neither obese, nor hypertensive and denied any forms of



**Figure-1:** Left Anterior Oblique (LAO) cranial view showing origin of RCA and LMCA from left coronary cusp.

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**Figure-2:** LAO caudal view showing origin of RCA and LMCA from left coronary cusp.

addiction. On examination, her blood pressure was 110/80 mmHg and pulse was 97 per minute. She was afebrile with a respiratory rate of 32 per minute. Upon CVS examination, the apex beat was heard at the 5th Intercostal Space (ICS). This was accompanied by a 4/6 ejection systolic murmur at the upper sternal border, which radiated to the carotids. A sustained thrill at the 2nd intercostal space, right of the sternum was also present. Other systemic examinations were unremarkable except for vertigo and mild dizziness. Initial Laboratory results were within normal ranges. Electrocardiograms (ECGs) revealed sinus tachycardia with a heart rate of 150 beats per minute and poor progression of R-waves. No ST-segment changes were observed. Echocardiography showed mild to moderate Left Ventricular Dysfunction with an Ejection Fraction of 40%, along with severe aortic stenosis and a dilated aortic root. It also revealed an Aortic Valve Peak Pressure Gradient (PPG) of 103 mmHg and Mean Pressure Gradient (MPG) of 75 mmHg. Initial therapy comprised of acetylsalicylic acid, beta-blockers and angiotensin-converting-enzyme inhibitor (ACEIs) and

the patient was scheduled for coronary angiography, via the right femoral artery route. The angiogram revealed a dominant, normal but aberrant Right Coronary Artery which arose from the back of the Left Coronary Cusp (Figure-1 and 2). The Right Coronary Artery could not be engaged due to the abnormal origin. The Left Main Coronary Artery (LMCA), Left Anterior Descending (LAD) and Left Circumflex Artery (LCX) were found to be of normal origins with no signs of pre atherosclerotic lesions.

The patient was scheduled for AVR (Aortic Valve Replacement). Keeping in view the known association of this anomaly and adverse cardiac effects, she was kept on anti-atherosclerotic therapy following angiography up to 3rd day post angiography. The patient was counseled and was subsequently discharged in a very stable state, without any further therapy for the coronary artery anomaly.

### Discussion

Coronary artery anomalies are an incidental finding. Yamanaka and Hobbs reported that of all the people undergoing coronary angiography, anomalous coronary arteries are detected in only about 1.3%.<sup>1</sup> Anomalous right coronary artery (RCA) from the left coronary sinus is reported to have an incidence of 0.13%.<sup>2</sup> These anomalies are mostly benign and are often detected upon routine cardiac catheterizations. It is well documented that coronary artery anomalies, particularly erroneous right coronary artery poses greater risks for myocardial infarction, syncope, ischaemia and most commonly sudden cardiac death (SCD). While other conditions can be present in patients of any age group, SCD is most commonly present in younger patients (<35 years) and is a common cause of death in asymptomatic patients, among other causes.<sup>2</sup> Certain predisposing factors such as intra-arterial course, slit like orifice, acute angle takeoff and anomalous origin from the aorta increases the risk of the development of SCD.<sup>3</sup> Anomalous RCA also increases the risk for epicardial atherosclerotic lesions<sup>4</sup> via two mechanisms namely the steal phenomenon and the accelerated atherosclerosis<sup>5</sup> and these are more commonly found in patients with Aortic Valve diseases. Hence, coronary artery anomalies associated with occlusive coronary artery disease should be included in the differential diagnoses of cases where extensive ST segment changes are observed.

The anomalous RCA originating from the left sinus was categorized into 4 types by Kragel and Roberts, which are as follows: RCA arises (i) from within the left sinus, (ii) from above the left sinus, (iii) directly above the commissure between the left and right cusps and (iv) from a common

ostium with the left main coronary artery.<sup>6</sup> The ostial shape was discovered to be a determinant of clinical significance. Hence, selective cannulation and engaging the coronary artery becomes a hassle owing to the unusual location of the ostium of an anomalous RCA. Similarly, failure to adequately engage the anomalous vessel may lead to poor angiographic visualization, an erroneous diagnosis and ultimately percutaneous coronary intervention (PCI) failure due to lack of guide support. Keeping this in view, a wide variety of guide catheters have been used in cases where anomalous RCA was found. These include the 6F Judkins left guide catheter (JL 5.0, 4.0), AL (Amplatz Left) catheter, Voda, EBU (extra backup guide catheter) and Loya which is a modification of the Amplatz. Recently, a new catheter, the Loya catheter has been reported to be of value for this difficult engagement. Lorin et al, published two cases of stenting in an anomalous RCA by use of a 6F Judkins left 5.0 guiding catheter through right radial access after being unable to cannulate the anomalous RCA through the femoral artery approach.<sup>7</sup> Similarly, Topaz et al reported the successful use of Amplatz AL-1 guiding catheter in 2 cases.<sup>7</sup>

Surgical strategy of choice for this kind of anomaly has been a pressing issue in cardiac medicine. There are a few options which include coronary reimplantation, coronary artery bypass and unroofing of the intramural segment (ostiotomy). De Lello et al suggested that RCA reimplantation to the aorta should be the ideal treatment.<sup>8</sup> Whilst this is a proposal put forth recently, a coronary artery bypass without ligating the RCA has been a surgical strategy for more than a decade because it eliminates the need to open the aorta, however it puts the patient at risk of graft thrombosis. Similarly surgical unroofing has also been proposed by many surgeons, as pointed out by Mustafa et al.<sup>9</sup> However, this technique cannot be employed in the aforementioned scenario because it carries a risk of aortic insufficiency even late after surgery.<sup>10</sup> Surgical treatment is often proposed in young, asymptomatic patients while older patients are kept on conservative therapy like exercise restriction. Likewise, stents are indicated only in those patients with atherosclerotic changes in the anomalous vessels. Unfortunately, long-term impact of surgical intervention in patients with anomalous right coronary artery from left sinus of valsalva (LSOV) is uncertain and further studies need to be conducted to reach a definitive conclusion.

## Conclusion

The unusual course and location of the artery proves to

be a great challenge for young interventional cardiologists. Therefore perioperative angiograms are very helpful for surgeons to carefully operate on these anomalous vessels thereby eliminating the risk to rupture the arteries. In particular, the selection of an appropriate guiding catheter is critical to ensure selective angiography, proper assessment of lesion characteristics and facilitate successful delivery of appropriate devices. In this case, extra caution must be taken by the surgeon since aberrant origin of the right coronary artery may interfere with the valve replacement; the patient has to undergo to replace the stenosed aortic valve.

**Consent:** Informed consent was obtained from the patient to reproduce her case in this report.

**Conflict of Interests:** The authors declare that they have no conflict of interests.

## References

1. Yamanaka O, Hobbs R. Coronary artery anomalies in 126,595 patients undergoing coronary arteriography. *Cathet Cardiovasc Diagn.* 1990; 21: 28-40.
2. Yuan S, Tager S, Raanani E. Anomalous Origin of the Right Coronary Artery from the Left Coronary Sinus. *Chang Gung Med J.* 2009; 32: 455-8.
3. Lee B. Anomalous Right Coronary Artery From the Left Coronary Sinus With an Interarterial Course: Is It Really Dangerous?. *Korean Circulation J.* 2009; 39: 175.
4. Jim M, Siu C, Ho H, Lee S. Anomalous origin of the right coronary artery from the left coronary sinus is associated with early development of coronary artery disease. *J Invasive Cardiol.* 2004; 16: 466-8.
5. Chen I, Chao T, Tsai L. Simultaneous Anterior and Inferior Wall Myocardial Infarction in a Patient with Unusual Coronary Anatomy. *ActaCardiol Sin.* 2010; 26: 119-22.
6. Kragel A, Roberts W. Anomalous origin of either the right or left main coronary artery from the aorta with subsequent coursing between aorta and pulmonary trunk: Analysis of 32 necropsy cases. *Am J Cardiol.* 1988; 62: 771-7.
7. Uthayakumar K, Subban V, Lakshmanan A, Pakshirajan B, Solirajaram R, Krishnamoorthy J, et al. Coronary intervention in anomalous origin of the right coronary artery (ARCA) from the left sinus of valsalva (LSOV): A single center experience. *Indian Heart J.* 2014; 66: 430-4.
8. Di Lello F, Mruk J, Flemma R, Mullen D. Successful coronary reimplantation for anomalous origin of the right coronary artery from the left sinus of Valsalva. *J ThoracCardiovasc Surg.* 1991; 102: 455-6.
9. Hauser M. Congenital anomalies of the coronary arteries. *Heart.* 2005; 91: 1240-5.
10. Huynh N, Ross R, Burns P, C. George J. Anomalous Right Coronary Artery From Left Coronary Cusp With Inter-Arterial Course: Evaluation and Management. [online] [cited 2015 Oct 12]; Available from URL: <http://www.cathlabdigest.com/article/Anomalous-Right-Coronary-Artery-Left-Coronary-Cusp-Inter-Arterial-Course-Evaluation>.