Published in final edited form as:

Catheter Cardiovasc Interv. 2017 June 01; 89(7): 1239–1241. doi:10.1002/ccd.26971.

A Heart Within a Heart

Edward T. Carreras, MD¹, Maya Barghash, MD¹, Michael M. Givertz, MD¹, and Deepak L. Bhatt, MD, MPH¹

¹Division of Cardiovascular Medicine, Brigham and Women's Hospital, Harvard Medical School, 75 Francis Street, Boston, MA 02115

Abstract

A 44-year-old man with a history of end-stage dilated cardiomyopathy status-post orthotopic cardiac transplant 14 years ago presented for coronary angiography in preparation for re-operative tricuspid valve replacement. Coronary angiography revealed an anomalous origin of the left coronary artery, with a common coronary trunk arising from the right coronary cusp and bifurcating into right and left main coronary arteries. Interestingly, the right and left coronary arteries coursed to form the shape of a heart, hence, a heart within a heart!

Introduction

In the following discussion, we report a case of an anomalous origin of the left main coronary artery arising from the right coronary cusp in a patient status-post heart transplant. The transplanted heart is supplied by a common coronary trunk arising from the right coronary cusp and bifurcates into right and left main coronary arteries, which course to form the shape of a heart, providing a unique and interesting display of this rare anomaly.

Corresponding Author: Deepak L. Bhatt, MD, MPH, FACC, FAHA, FSCAI, FESC, Executive Director of Interventional Cardiovascular Programs, Brigham and Women's Hospital Heart & Vascular Center, Professor of Medicine, Harvard Medical School, 75 Francis Street, Boston, MA 02115, dlbhattmd@post.harvard.edu.

Funding:

There was no funding for this manuscript.

Conflicts of interest:

Dr. Carreras has no relevant disclosures.

Dr. Barghash has no relevant disclosures.

Dr. Givertz has no relevant disclosures.

Dr. Deepak L. Bhatt discloses the following relationships - Advisory Board: Cardax, Elsevier Practice Update Cardiology, Medscape Cardiology, Regado Biosciences; Board of Directors: Boston VA Research Institute, Society of Cardiovascular Patient Care; Chair: American Heart Association Quality Oversight Committee; Data Monitoring Committees: Duke Clinical Research Institute, Harvard Clinical Research Institute, Mayo Clinic, Population Health Research Institute; Honoraria: American College of Cardiology (Senior Associate Editor, Clinical Trials and News, ACC.org), Belvoir Publications (Editor in Chief, Harvard Heart Letter), Duke Clinical Research Institute (clinical trial steering committees), Harvard Clinical Research Institute (clinical trial steering committee), HMP Communications (Editor in Chief, Journal of Invasive Cardiology), Journal of the American College of Cardiology (Guest Editor; Associate Editor), Population Health Research Institute (clinical trial steering committee), Slack Publications (Chief Medical Editor, Cardiology Today's Intervention), Society of Cardiovascular Patient Care (Secretary/Treasurer), WebMD (CME steering committees); Other: Clinical Cardiology (Deputy Editor), NCDR-ACTION Registry Steering Committee (Chair), VA CART Research and Publications Committee (Chair); Research Funding: Amarin, Amgen, AstraZeneca, Bristol-Myers Squibb, Eisai, Ethicon, Forest Laboratories, Ischemix, Lilly, Medtronic, Pfizer, Roche, Sanofi Aventis, The Medicines Company; Royalties: Elsevier (Editor, Cardiovascular Intervention: A Companion to Braunwald's Heart Disease); Site Co-Investigator: Biotronik, Boston Scientific, St. Jude Medical; Trustee: American College of Cardiology; Unfunded Research: FlowCo, PLx Pharma, Takeda.

All authors had a role in preparing this manuscript.

Case report

A 44-year-old man with a history of antiphospholipid antibody syndrome, venous thromboembolism, chronic hepatitis B, and end-stage dilated cardiomyopathy status-post orthotopic cardiac transplant 14 years ago presented with chest pain and acute on chronic heart failure. Initial noninvasive workup revealed severe tricuspid regurgitation due to a flail tricuspid leaflet. He was referred for coronary angiography in preparation for re-operative tricuspid valve replacement.

Coronary angiography was performed via access from the right radial artery. A JR4 catheter was used to engage a common coronary trunk in the right aortic cusp. Imaging in the left anterior oblique caudal view revealed a common coronary trunk bifurcating into right and left main coronary arteries (Figure 1). The right coronary artery (RCA) was dominant and supplied a large posterior descending artery (Supplementary Video 1). The left main coronary artery (LMCA) bifurcated into left anterior descending (LAD) and left circumflex arteries (LCX) (Supplementary Video 2). There was a severe stenosis in the distal left circumflex artery after the take-off of the third obtuse marginal branch, as well as evidence of distal vessel pruning, consistent with cardiac allograft vasculopathy.

He later underwent a successful tricuspid valve replacement with a 29 mm St. Jude Epic bioprosthetic heart valve. His initial chest pain had resolved after diuresis and was ultimately attributed to heart failure. Coronary bypass grafting of the left circumflex artery was deferred at the time of surgery due to the distal location of the stenosis.

Discussion

An anomalous origination of a coronary artery from the opposite sinus is a rare congenital abnormality. Prior studies have estimated the global incidence at approximately 0.92% for anomalous RCA origin and 0.15% for anomalous LMCA origin.¹ The clinical impact of anomalous coronary origins is variable and dependent on the specific anomaly. For anomalous origination of the RCA from the left coronary cusp, the majority of patients have no symptoms and observation is recommended in the absence of clinical events.² In the rarer cases of anomalous origination of the LMCA from the right coronary cusp, the risk of coronary compression and sudden death are greater, and surgical correction is often recommended.²

The incidence and clinical impact of coronary artery anomalies in transplanted hearts is unknown, with only a few published case reports.^{3, 4} This is the first reported case to our knowledge of an anomalous origination of the LMCA from the right coronary cusp in a transplanted heart. There is one prior case report of an anomalous origination of the LAD from the pulmonary artery which was known at the time of transplant; in that case, surgical modification of the aortopulmonary groove was performed at the time of transplant to prevent future LMCA compression.⁵ In the case presented above, the anomaly was not known prior to transplant and was originally discovered at the time of the patient's first screening coronary angiogram after transplant. Further imaging at the time with cardiac magnetic resonance imaging revealed that the artery coursed anteriorly to the pulmonary

artery, implying a lower risk of coronary compression and ischemia. Surgical intervention was deferred and the patient was followed and observed, with no clinical symptoms related to his coronary anomaly during his 14 years post-transplant. Given his lack of symptoms and the anterior course of the coronary artery, the risk of compression was considered low enough that intervention was deferred again at the time of his tricuspid valve replacement.

Conclusion

To our knowledge, this is the first reported case of an anomalous origination of the LMCA from the right coronary cusp in a transplanted heart. Furthermore, the orientation of the coronary arteries in the shape of a heart provides a unique and interesting display of this anomaly.

Supplementary Material

Refer to Web version on PubMed Central for supplementary material.

References

1. Cheng TO. Anomalous coronary arteries. *Cardiology*. 1999; 91:74. [PubMed: 10393404]
2. Angelini P. Coronary artery anomalies: an entity in search of an identity. *Circulation*. 2007; 115:1296–305. [PubMed: 17353457]
3. Bresseleers D, Flameng W, Vanhaecke J. Anomalous origin of a coronary artery in a transplanted heart. *Acta Cardiol*. 1999; 54:93–6. [PubMed: 10378021]
4. Abudiab MM, Hakim FA, Fortuin DF, Scott RL. Anomalous coronary artery in a transplanted heart: a rare incidental diagnosis. *J Saudi Heart Assoc*. 2016; 28:46–8. [PubMed: 26778904]
5. Vasseur BG, Anderson MB, Freudenberger RS. Anomalous coronary artery in a transplanted heart: a technical modification. *J Heart Lung Transplant*. 2004; 23:1008–9. [PubMed: 15312832]

Figure 1.

Coronary angiography in the left anterior oblique caudal view revealing an anomalous coronary artery, with a common coronary trunk bifurcating into right coronary and left main coronary arteries in a patient with a transplanted heart. Note the course of the coronary arteries on the angiogram forms a heart – thus, a heart within a heart!