*Cardiology in the Young*

**cambridge.org/cty**

# Heart failure in an elderly man: where is that coronary?

Charlie J. Sang III[1], Stephen A. Clarkson[2], Elizabeth A. Jackson[2], Firas Al Solaiman[3] and Marc G. Cribbs[4]

[1]Internal Medicine – Pediatrics Residency Program, Department of Pediatrics, University of Alabama at Birmingham, Birmingham, AL, USA; [2]Division of Cardiovascular Disease, Department of Medicine, University of Alabama at Birmingham, Birmingham, AL, USA; [3]Section of Interventional Cardiology, Division of Cardiovascular Disease, Department of Medicine, University of Alabama at Birmingham, Birmingham, AL, USA and [4]Section of Adult Congenital Heart Disease, Division of Cardiovascular Disease and Pediatric Cardiology, University of Alabama at Birmingham, Birmingham, AL, USA

**Brief Report**

**Cite this article:** Sang CJ III, Clarkson SA, Jackson EA, Al Solaiman F, and Cribbs MG (2021) Heart failure in an elderly man: where is that coronary? *Cardiology in the Young* **31**: 1030–1033. doi: 10.1017/S104795112100007X

Received: 12 November 2020
Revised: 28 December 2020
Accepted: 30 December 2020
First published online: 28 January 2021

**Keywords:**
Coronary vessel anomaly; cardiomyopathy; CHD

**Address for correspondence:**
M. G. Cribbs, MD, Section of Adult Congenital Heart Disease, Division of Cardiovascular Disease and Pediatric Cardiology, University of Alabama at Birmingham, 1700 6th Avenue South, Suite 9100, Birmingham, AL 35233, USA. Tel: 205 – 934 – 2173; Fax: (205) 975-5991. E-mail: mgcribbs@uabmc.edu

**Abstract**

Anomalous coronary arteries from the pulmonary artery are uncommon causes of heart failure in the adult population. This case demonstrates the unusual presentation in a patient with anomalous right coronary artery from the pulmonary artery and discusses the complex pathophysiology of this lesion and the role of guideline-directed medical therapy in the management of these patients.

## Key points

1. Anomalous coronary arteries from the pulmonary artery represent a rare cause of congestive heart failure with complex pathophysiology.
2. Multimodal imaging is helpful to assess for other congenital abnormalities as well as for surgical planning.
3. Surgical correction of anomalous right coronary artery from the pulmonary artery is typically recommended; however, management of anomalous right coronary artery from the pulmonary artery in adulthood is challenging, especially when the aetiology of congestive heart failure is often multifactorial.
4. Guideline-directed medical therapy may provide a reasonable alternative to invasive interventions in patients at elevated risk for cardiac surgery.

## History of present illness

A 70-year-old male with hypertension, chronic obstructive pulmonary disease, and tobacco abuse presented to clinic with 2 months of fatigue and decreased activity. He endorsed exertional dyspnoea, paroxysmal nocturnal dyspnoea, two-pillow orthopnoea, and a 10-pound weight gain during this time frame. Physical examination revealed an overweight male (body mass index 28.3) in no acute distress, clear lung fields with prolonged expiration, a regular cardiac rhythm, with normal rate, and no murmur. Mild jugular venous distention was observed. Extremities exhibited trace oedema to the mid-lower extremities.

## Investigations

On presentation, electrocardiogram demonstrated sinus rhythm, voltage criteria for left ventricular hypertrophy, lateral T-wave inversions, and prolonged QTc (483 ms). Chest X-ray revealed normal cardiac silhouette and clear lung fields. A pro-Brain natriuretic peptide was elevated at 1193 pg/ml (ref. 0–100 pg/ml). Echocardiogram revealed a mildly dilated left ventricle with severe dysfunction and inferior and posterior wall akinesis, with an ejection fraction of 17% (Supplementary Video 1). There was moderate mitral regurgitation. The right ventricle was mild to moderately dilated with moderately reduced function. Right ventricular systolic pressure was estimated at 60 mmHg. He was placed on guideline-directed medical therapy and referred for cardiac catheterisation.

Right heart catheterisation revealed elevated right-sided filling pressures with right atrial pressure 17 mmHg, pulmonary artery pressure 74/35 (53) mmHg, pulmonary capillary wedge pressure 35 mmHg, and Fick cardiac index 1.6 L/minute/m2, with Qp:Qs of 1.7. Left heart catheterisation revealed a very large left main coronary artery. The right coronary artery could not be engaged. Injection of the left main revealed the left anterior descending artery, left circumflex artery, and filling of the right coronary artery in retrograde fashion. No significant atherosclerotic narrowing of the coronary vessels was observed. There also



CAMBRIDGE UNIVERSITY PRESS

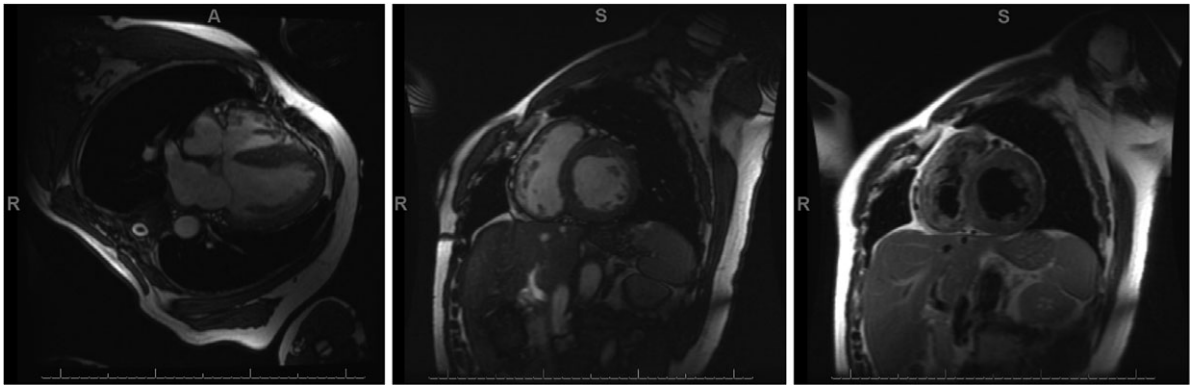


**Figure 1.** Cardiac MRI. Moderate asymmetric thickening of the midportion of the interventricular septum with patchy mid wall areas of late gadolinium enhancement. There is generalised moderate left ventricular myocardial hypertrophy. The right chambers are normal in size with mild right ventricular hypertrophy.

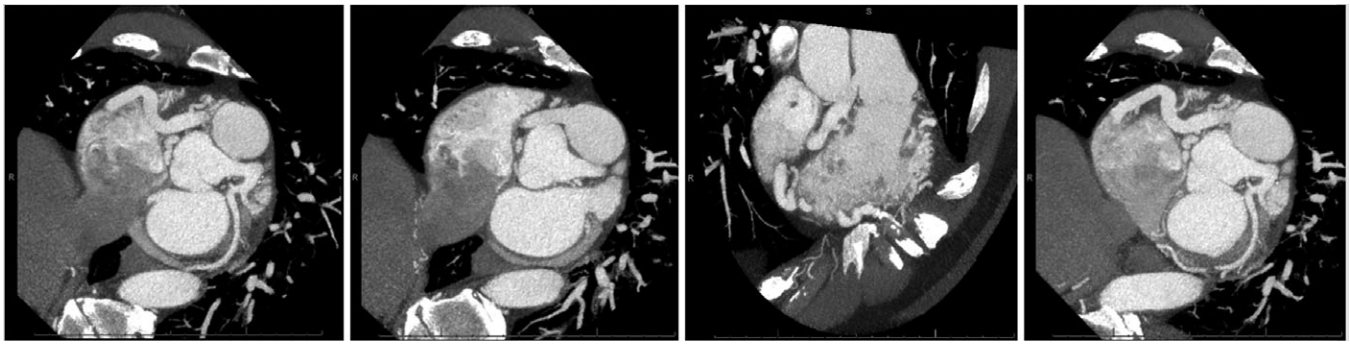

**Figure 2.** Cardiac CTA. There is a right coronary artery arising from the pulmonary trunk proximally with marked dilatation of all coronary arteries, with dilated small branches around the proximal pulmonary artery trunk as well as the proximal aorta. There is no significant coronary arterial atherosclerotic narrowing.

appeared to be a large fistula that arose from the distal right coronary artery/left circumflex artery system that emptied into the pulmonary artery (Supplementary Video 2). It was felt this represented either an anomalous right coronary artery arising from the main pulmonary artery that filled backwards into the pulmonary artery via direct collaterals from the left coronary system or a less likely right coronary artery arising from the left cusp and giving rise to a large fistula to the main pulmonary artery. Pulmonary artery angiogram was not performed to limit contrast use.

Cardiac magnetic resonance imaging revealed moderate concentric left ventricle hypertrophy and globally diminished contractility resulting in moderately depressed systolic function, with an ejection fraction of 37% (Fig 1). There was mild right ventricular hypertrophy with mildly decreased ejection fraction (39.7%) and contractility. There was no evidence of additional CHDs. There was noted to be abnormal late mid wall enhancement seen in the midportion of the interventricular septum as well as patchy areas of mid wall enhancement in the inferior lateral wall near the base, although these findings were noted to be subtle. Additionally, there was myocardial thinning of the inferior wall, making determiniation of viability difficult. Cardiac computed tomography angiogram revealed an anomalous origin of the right coronary artery from the pulmonary artery trunk with resultant cirsoid dilatation of all coronary vessels except for the left ventricular free wall branches (Figs 2 and 3). There was no significant coronary arterial atherosclerotic narrowing.

## Management and follow-up

With 2 months of strict adherence to guideline-directed medical therapy, including metoprolol, lisinopril, hydralazine (the patient did not tolerate nitrate therapy), and furosemide, the patient's symptoms resolved. He maintained close follow-up, and repeat echocardiogram 1 year later showed sustained improvement in the left ventricular ejection fraction to approximately 35–45% with continued mild dilation and global hypokinesis. Right ventricular dilation had resolved and function was mildly reduced. There was only trace tricuspid regurgitation, with estimated right atrial pressure of 0–5 mmHg. His case was discussed extensively in the paediatric and adult congenital cardiovascular multidisciplinary conference. The patient was estimated to have a 17% risk for significant perioperative morbidity or mortality based on the Society of Thoracic Surgeons risk assessment. Given his comorbidities, improvement in ejection fraction, and NYHA Class I functional capacity, surgical intervention on the anomalous right coronary artery was deferred and monitoring with regular follow-up was recommended. At the 2-year follow-up appointment, he continued to feel well and remained free from hospitalisations for heart failure.

## Discussion

Four congenital variants of anomalous coronary artery origins from the pulmonary artery have been described: anomalous left coronary artery from the pulmonary artery; anomalous right

https://doi.org/10.1017/S104795112100007X Published online by Cambridge University Press

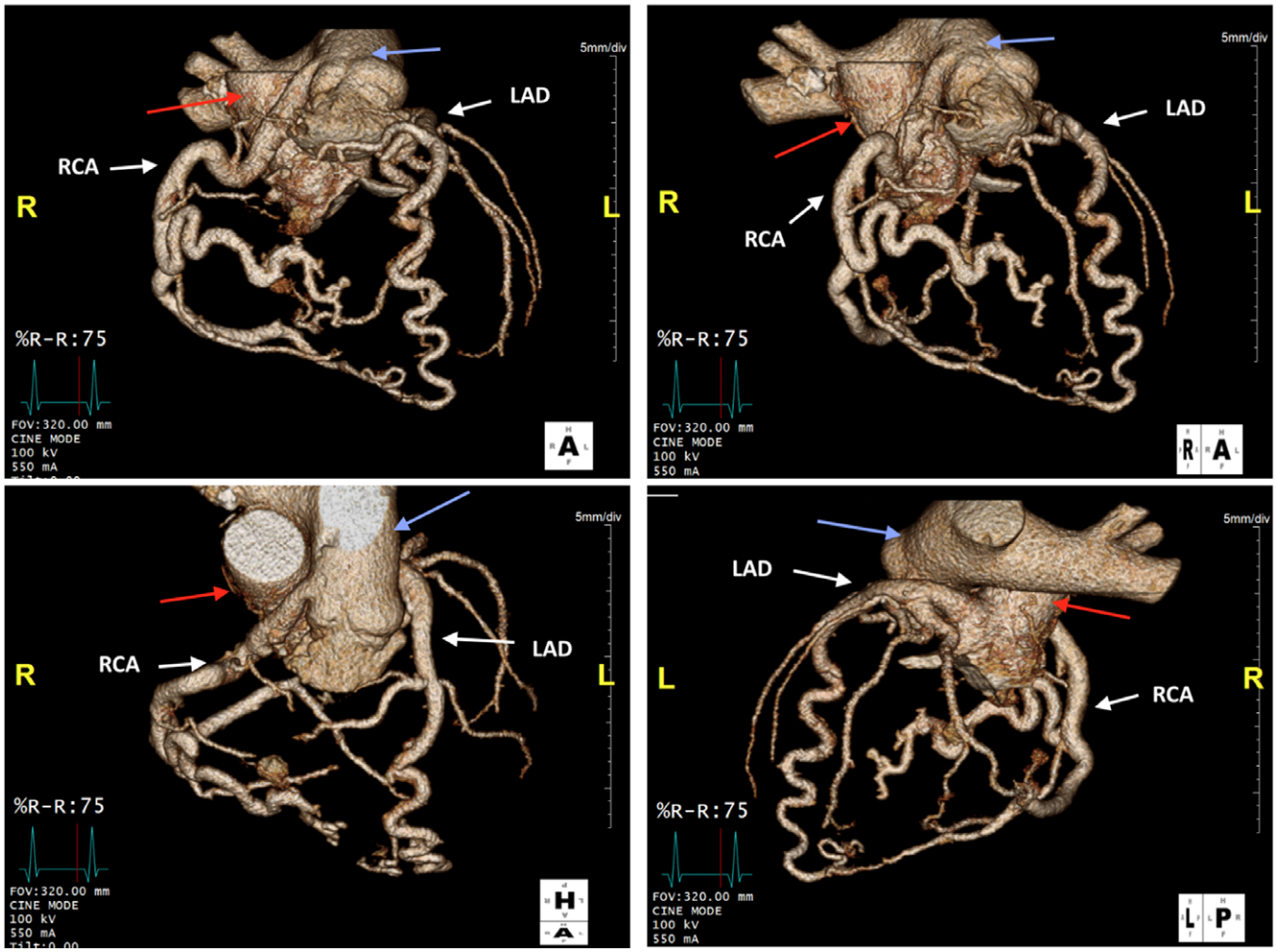


**Figure 3.** Three-dimensional coronary reconstruction. Three-dimensional reconstruction revealing the origin of the right coronary artery arising from the pulmonary artery trunk (blue arrow). The left main coronary arises from the aortic root (red arrow). There is marked dilation of all coronary arteries. RCA: Right Coronary Artery; LAD: Left Anterior Descending Artery.

coronary artery from the pulmonary artery; anomalous origin of both coronaries; or an accessory artery from the pulmonary artery, with the first two being the most often encountered and clinically relevant.[1] Of these, anomalous right coronary artery from the pulmonary artery lends itself to survival into adulthood.[1] Estimated incidence of anomalous right coronary artery from the pulmonary artery is 0.002% in adult patients undergoing routine coronary angiography.[2] The embryologic events leading to anomalous right coronary artery from the pulmonary artery remain largely unknown, although the incorrect fusion of coronary buds probably occurs during coronary artery development in the 4–6th week of gestation.[1]

The clinical presentation of anomalous right coronary artery from the pulmonary artery is variable, both in age of onset and type of symptoms reported.[1,3] In patients with symptoms, the age of onset depends on collateralisation between the left and right coronary system.[1,4] Soon after birth, pulmonary artery pressures decrease dramatically, and if adequate collateralisation has not occurred, patients can experience cardiac dysfunction in the areas supplied by the right coronary artery.[1] In contrast, if collateralisation is adequate, coronary flow from the left system may compensate, providing adequate perfusion to the entire heart.[1] However, as seen in this case, the collateralisation eventually developed into a significant inter-coronary left-to-right shunt and created a previously described coronary steal phenomenon, thereby reducing myocardial oxygen delivery with resultant myocardial ischaemia.[5] This ischaemia, combined with long-standing hypertension and tobacco abuse, contributed to left ventricular dysfunction in the present case. Pulmonary artery pressures were increased due to elevated left-sided filling pressures and chronic pulmonary disease, which subsequently led to the development of right-sided heart failure.

Diagnosis of anomalous right coronary artery from the pulmonary artery is most commonly made via coronary angiography[1] and, in the present case, was discovered incidentally during evaluation for ischaemic heart disease. Due to the invasive nature of angiography and inability to elucidate non-coronary anatomy, first-line imaging modalities have shifted to the use of cardiac computed tomography angiogram and cardiac magnetic resonance imaging.[6] While cardiac computed tomography angiogram, cardiac magnetic resonance imaging, and coronary angiography can establish the diagnosis, cardiac computed tomography angiogram offers superior spatial and temporal resolution.[6,7] Though not necessary for diagnosis, cardiac magnetic resonance imaging can be useful in pre-

https://doi.org/10.1017/S104795112100007X Published online by Cambridge University Press

operative risk stratification by providing additional information regarding both valvular and ventricular function.[6]

Management of anomalous right coronary artery from the pulmonary artery remains challenging, especially with regard to older adults who may have comorbid conditions that elevate the risk of cardiac surgery. In general, the treatment strategy has focused on surgical correction and attempts to create a two-coronary system either by reimplantation of the anomalous coronary or via ligation of the right coronary artery with subsequent bypass grafting.[1,7] The decision to operate in adults who are asymptomatic, however, is less clear and reports of long-term medical management are limited.[5,8] Alternative approaches include transcatheter closure using vascular plugs, a technique most often employed with haemodynamically significant coronary artery fistulae.[9] For the present case, guideline-directed medical therapy led to notable improvement in symptoms after 2 months. Though he remained at risk for worsening cardiac dysfunction related to myocardial steal, we felt the aetiology of his heart failure was multifactorial given his history of tobacco abuse, hypertension, and chronic lung disease. Given this, along with his age and comorbid conditions, conservative management was considered to be the safest course of action. Consideration of surgical correction would be readdressed should medical management fail. To date, the patient has remained free from hospitalisations and has maintained improvement in symptoms from an initial NYHA Class III presentation to Class I symptoms.

## Conclusion

This case demonstrates the pathophysiology of a rare congenital coronary defect. While this patient's presenting clinical picture was certainly multifactorial, this case also highlights the long-term implications of this congenital lesion in adulthood and the role of guideline-directed medical therapy in the management of adult patients with anomalous right coronary artery from the pulmonary artery.

**Acknowledgements.** None.

**Financial support.** There were no grants, contracts, or other forms of financial support used in the making of this report.

**Conflicts of interest.** Dr Jackson reports research funding from NIH and Amgen; editorial board membership: Circulation: Cardiovascular Quality and Outcomes; consulting: American College of Cardiology and McKesson, Inc.; Expert witness for DeBlase Brown Everly LLP.; and royalties for UpToDate. The remaining authors have nothing to disclose.

**Ethical standards.** This case does not include any protected health information and is exempt from Institutional Review Board protocol at this institution.

**Supplementary material.** To view supplementary material for this article, please visit https://doi.org/10.1017/S104795112100007X

## References

1. Guenther TM, Sherazee EA, Wisneski AD, et al. Anomalous origin of the right coronary artery from the pulmonary artery: a systematic review. Ann Thorac Surg 2020; 110: 1063–1071.
2. Yamanaka O, Hobbs RE. Coronary artery anomalies in 126,595 patients undergoing coronary arteriography. Cathet Cardiovasc Diagn 1990; 21: 28–40.
3. Radke PW, Messmer BJ, Haager PK, et al. Anomalous origin of the right coronary artery: preoperative and postoperative hemodynamics. Ann Thoracic Surg 1998; 66: 1444–1449.
4. Mantha Y, Feldman M, Assaf A, et al. Anomalous origin of a right coronary artery from the pulmonary artery in a middle-aged woman. JACC Case Reports 2020; 2: 464.
5. Halim SA, Wang A, Harrison JK. Anomalous right coronary artery origin from the pulmonary artery (ARCAPA) coexisting with obstructive atherosclerotic disease of the left coronary artery. J Invasive Cardiol 2011; 23: E260–261.
6. Gräni C, Buechel RR, Kaufmann PA, et al. Multimodality imaging in individuals with anomalous coronary arteries. JACC Cardiovasc Imaging 2017; 10: 471–481.
7. Stout KK, Daniels CJ, Aboulhosn JA, et al. 2018 AHA/ACC guideline for the management of adults with congenital heart disease. J Am College Cardiol 2019; 73: e81.
8. Farwati M, Shaker F, Nasser MM Anomalous origin of the right coronary artery from the pulmonary artery in a morbidly obese patient presenting with chest pain. Am J Case Rep 2018; 19: 992–997.
9. Behera SK, Danon S, Levi DS, et al. Transcatheter closure of coronary artery fistulae using the Amplatzer Duct Occluder. Catheter Cardiovasc Interv 2006; 68: 242–248.