

Single Coronary Artery with an Ectopic Origin and Interarterial Course: A Case Report and Review of the Literature

Timothy A. Mixon, MD, Linley E. Watson, MD

Division of Cardiology, Scott & White Memorial Hospital and Clinic, Scott, Sherwood and Brindley Foundation, Texas A&M University System Health Science Center College of Medicine, Temple, TX

A previously unreported variant of a single coronary artery arising between the aorta and pulmonary artery is presented. This variant had many high-risk features, so prophylactic coronary artery bypass grafting was recommended.

[Rev Cardiovasc Med. 2002;3(2):107–110]

© 2002 MedReviews, LLC

Key words: Coronary anomaly • Sudden death • Ischemia

Among nonatherosclerotic causes of myocardial ischemia, coronary anomalies are a rare cause. While coronary anomalies occur at a rate of approximately 0.2% to 1.3% in adult angiographic series,^{1,2} the subset that leads to clinically important sequelae such as ischemia, infarction, or sudden death is considerably lower. Single coronary arteries may most commonly be associated with these outcomes if one or more arterial segments travels between the ascending aorta and the pulmonary infundibulum.

We present a previously unreported variant of a single coronary artery arising superior to the coronary sinuses with the proximal portion of both the right coronary artery and the left main coronary artery traveling between the great vessels, leading to severe myocardial ischemia.

Case Report

A 45-year-old African American woman presented for evaluation of numerous, intermittent episodes of chest pain occurring over 4 to 6 weeks. The pain was located in the left side of her chest with radiation to the left arm, occurring both at times of rest and during light exertion. Past medical history was negative; there was no history of syncope, rheumatic disease, or prior cardiac evaluation. Her only cardiac risk factor was smoking a pack of cigarettes per day; she took no medications chronically. Physical exam and baseline electrocardiogram were normal.

During an exercise thallium scan, the patient developed chest pain after walking for 4 minutes on the treadmill (standard Bruce protocol), with concomitant 1 mm ST depression throughout the precordial leads. Exercise was terminated at 5 minutes due to persistent pain. Nuclear scintigraphy revealed a normal resting ejection fraction, but stress images showed a pattern of global ischemia with left ventricular dilatation.

Coronary angiography was subsequently accomplished utilizing a 7 French Voda (Boston Scientific, Maple Grove, MN) catheter to cannulate a single, ectopic ostium. The single coronary artery arose above the level of the coronary sinuses (Figure 1), bifurcating early into a

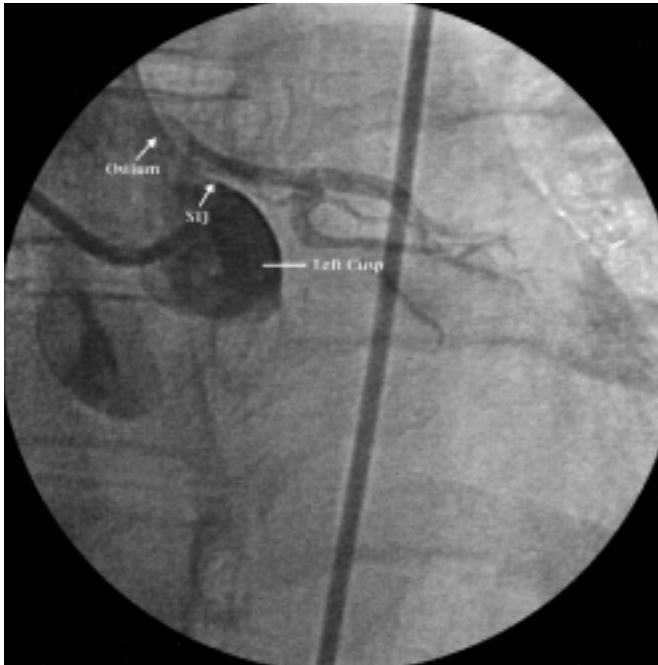


Figure 1. Root aortogram demonstrating the coronary ostium arising superior to the sinotubular junction. STJ, sinotubular junction.

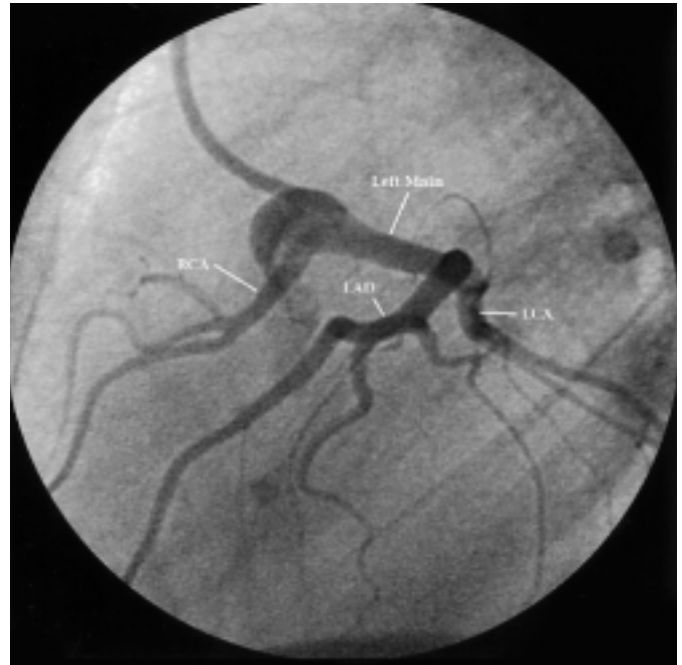


Figure 2. Coronary angiogram in the 30-degree left anterior oblique position, showing single coronary artery giving rise to all three epicardial vessels. RCA, right coronary artery; LAD, left anterior descending artery; LCX, left circumflex artery.

right coronary artery and a left main artery with normal terminal distribution (Figure 2). The initial course of the arteries in relation to the aorta and the pulmonary artery was

not known with certainty, however. An 80% stenosis of a medium-sized diagonal branch was the only evidence of atherosclerotic disease, but this could not fully explain the high

risk findings of the exercise thallium scan, including early widespread ischemic electrocardiographic changes and ventricular dilatation.

An electron beam computed tomography was obtained the following day, which revealed the origin of the single coronary artery to arise between the ascending aorta and the pulmonary infundibulum

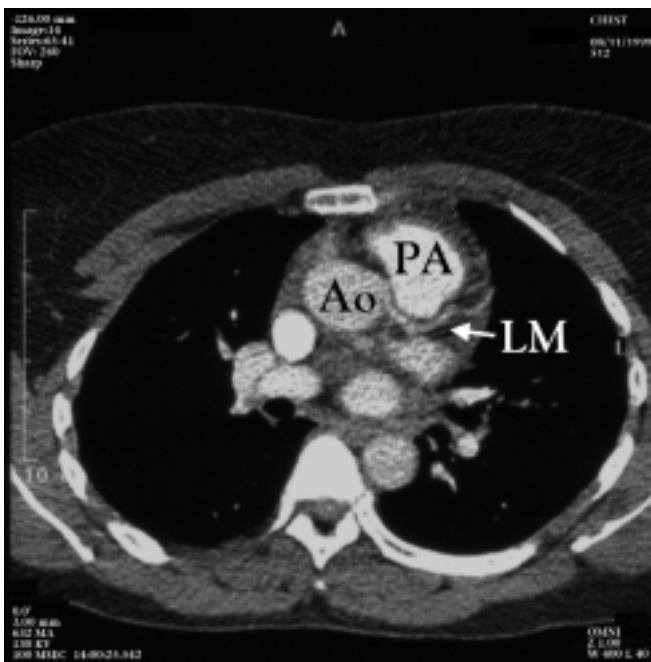
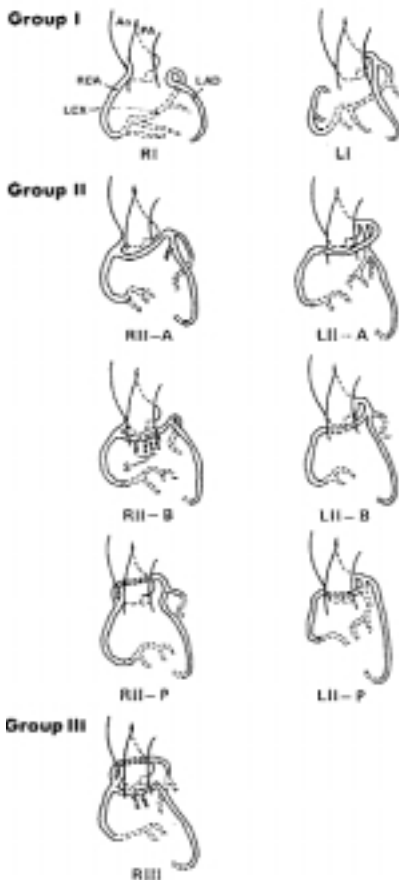


Figure 3. Electron beam computed tomography scan revealing the proximal left main artery arising between the aorta and pulmonary artery. Ao, aorta; PA, pulmonary artery; LM, left main artery.

Coronary artery bypass grafting was recommended and successfully carried out.

(Figure 3). Based on the course of both the proximal right coronary artery (RCA) and the proximal left main coronary artery between the great vessels, coronary artery bypass grafting was recommended and successfully carried out with anastomosis of the right internal thoracic artery to the RCA, the left internal thoracic artery to the left anterior descending

Figure 4. Lipton's proposed classification of isolated single coronary artery. Key for abbreviations: R, right; L, left; RCA, right coronary artery; LCA, left coronary artery; LAD, left anterior descending coronary artery; LCx, left circumflex coronary artery; Ao, aorta; PA, main pulmonary artery; A, anterior; B, between and P, posterior to the great vessels. Reprinted with permission from Lipton MJ, Barry WH, Obrez I, et al. Isolated single coronary artery: diagnosis, angiographic classification, and clinical significance. *Radiology*. 1979;130:39-47.



(LAD), and a reverse saphenous vein graft from the proximal aorta to the first diagonal branch. Follow-up thallium scan revealed no further evidence of ischemia or chamber dilation, and the patient is symptom-free.

Discussion

Isolated single coronary artery is reported to occur in approximately 0.024% to 0.04% of the population.³ It may occur with (41%) or without (59%) associated congenital anomalies.⁴ Smith⁵ and subsequently

Lipton and colleagues⁶ have categorized numerous variations based on site of origin (right or left coronary cusp), number of branching vessels (1-3), and course in regards to the ascending aorta and pulmonary outflow tract (Figure 4).

Cheitlin and colleagues⁷ conducted an extensive review of autopsy data from the Armed Forces Institute of Pathology and first suggested that

that when the right coronary artery arises from the left cusp and travels between the great vessels (Lipton class LII-B)⁶ there was not a significant risk of MI or sudden death⁸; however, a growing number of reports brings this into question, especially when dealing with a right-dominant system.¹¹⁻¹⁴

An autopsy series by Kragel and Robert was the first to describe

The etiology of sudden death in these patients is unknown.

when the single ostium arises from the right cusp, and the left main artery traverses between the ascending aorta and the pulmonary outflow tract (Lipton class RII-B),⁶ the lesion may not be benign and the patient is at increased risk of myocardial infarction (MI) and sudden death, especially with exertion. To the contrary, if the left main artery travels anterior or posterior to both great vessels (Lipton class RII-A and RII-P respectively),⁶ or if it passes between the great vessels by way of an intramyocardial bridge (Lipton class RII-B intramyocardial),⁶ the risk of this anomaly leading to sudden death or MI is felt to be quite low, although the literature reports exceptions.^{7,10} It has been believed

anomalous coronary arteries arising outside of a coronary sinus of Valsalva.¹¹ Their series suggested that a high takeoff (that is, above the sinotubular junction), especially with an ostium occurring directly superior to a commissure, may place the patient at increased risk of an adverse outcome. This is evidenced by the fact that 5 of the 10 patients in this series with an RCA arising from above the commissure separating the right and left cusp and passing between the great vessels experienced sudden death attributed to the anomaly, a rate distinctly high for an RCA anomaly (Figure 5).

The etiology of sudden death in these patients is unknown, although there have been numerous postula-

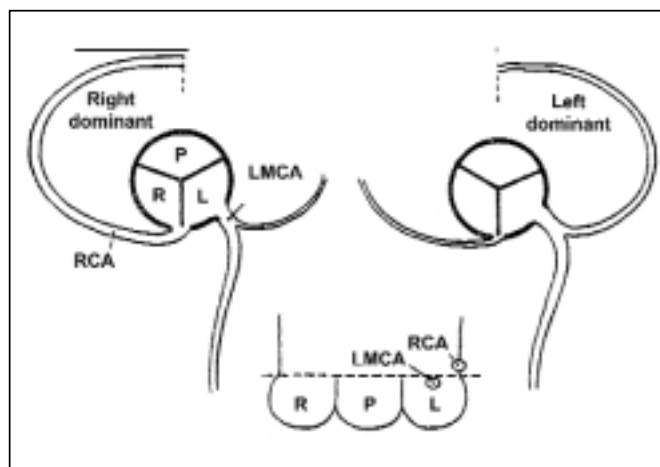


Figure 5. Coronary anatomy in cases of anomalous origin of the right coronary artery arising superior to the commissure between the right and left sinus of Valsalva. R, right; L, left; P, posterior/noncoronary cusp. Reprinted with permission from Kragel AH, Roberts WC. Anomalous origin of either the right or left main coronary artery from the aorta with subsequent coursing between aorta and pulmonary trunk: analysis of 32 necropsy cases. *American Journal of Cardiology* 1988;62:771-777.

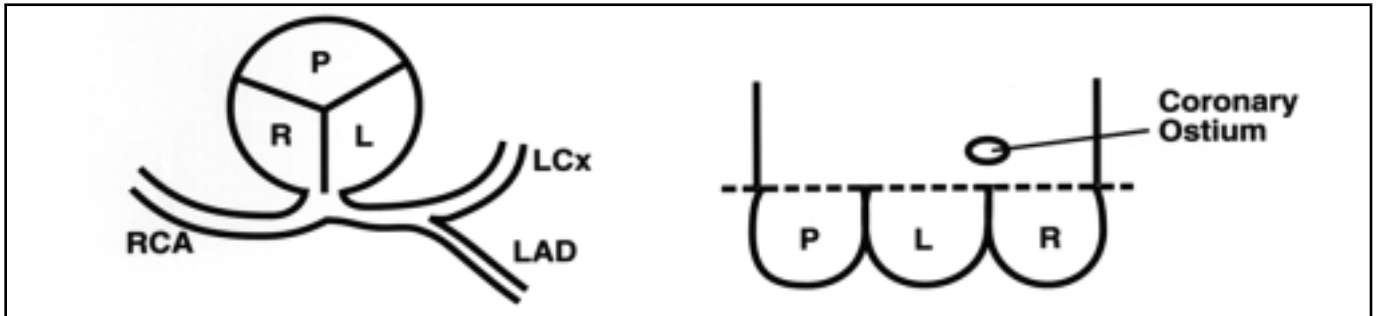


Figure 6. Coronary anatomy for case presented. The single coronary ostium arises between the left and right coronary cusp, superior to the sinotubular junction. Abbreviations as in Figure 5.

tions. Some have proposed that compression of the left main coronary artery between the aorta and pulmonary outflow may occur with vigorous exertion and increased cardiac output.^{15,16} Others suggest that exertion causes distention of the aorta that exaggerates the angulation at the origin of the vessel, creating a dynamic compression or “kinking.”¹⁷ Finally, Cheitlin and colleagues⁷ point out that many of the anomalous arteries have abnormal, slit-like orifices that may become compromised during exertion due to enlargement of the aorta.

Conclusion

We have presented a case of a previously unreported coronary anomaly in which a single coronary artery arises both superior to the sinotubular junction above the commissure separating the left and right coronary cusp (Figure 6), and between the ascending aorta and pulmonary infundibulum, so that the proximal portions of both the left main artery and the right coronary artery travel between the great vessels (see Figure 3). Either

anomaly alone has been associated with MI and sudden death, so the lesion was judged to be “high risk” on anatomical grounds. Both early widespread electrocardiographic abnormalities and scintigraphic features of global ischemia with ventricular dilatation provided objective evidence of a myocardium at significant risk. ■

References

1. Click RL, Holmes DR, Vlietstra RE, Kosinski AS, Kronmal RA. Anomalous coronary arteries: location, degree of atherosclerosis and effect on survival—a report from the Coronary Artery Surgery Study. *J Am Coll Cardiol.* 1989;13:531–537.
2. Yamanaka O, Hobbs RE. Coronary artery anomalies in 126,595 patients undergoing coronary arteriography. *Cathet Cardiovasc Diagn.* 1990;21:28–40.
3. Desmet W, Vanhaecke J, Vrolix M, et al. Isolated single coronary artery: a review of 50,000 consecutive coronary angiographies. *Eur Heart J.* 1992;13:1637–1640.
4. Sharbaugh AH, White RS. Single coronary artery. Analysis of the anatomic variation, clinical importance, and report of five cases. *JAMA.* 1974;230:243–246.
5. Smith JC. Review of single coronary artery with report of 2 cases. *Circulation.* 1950;1:1168–1175.
6. Lipton MJ, Barry WH, Obrez I, Silverman JF, Wexler L. Isolated single coronary artery: diagnosis, angiographic classification, and clinical significance. *Radiology.* 1979;130:39–47.
7. Cheitlin MD, De Castro CM, McAllister HA. Sudden death as a complication of anomalous left coronary origin from the anterior sinus of Valsalva, a not-so-minor congenital anomaly. *Circulation.* 1974;50:780–787.
8. Liberthson RR, Dinsmore RE, Fallon JT. Aberrant coronary artery origin from the aorta. Report of 18 patients, review of literature and delineation of natural history and management. *Circulation.* 1979;59:748–754.
9. Roberts WC. Major anomalies of coronary arterial origin seen in adulthood. *Am Heart J.* 1986;111:941–963.
10. Chaitman BR, Lesperance J, Saltiel J, Bourassa MG. Clinical, angiographic, and hemodynamic findings in patients with anomalous origin of the coronary arteries. *Circulation.* 1976;53:122–131.
11. Kragel AH, Roberts WC. 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(10 Pt 1):771–777.
12. Thompson SI, Vieweg WV, Alpert JS, Hagan AD. Anomalous origin of the right coronary artery from the left sinus of Valsalva with associated chest pain: report of two cases. *Cathet Cardiovasc Diagn.* 1976;2:397–402.
13. Frescura C, Basso C, Thiene G, et al. Anomalous origin of coronary arteries and risk of sudden death: a study based on an autopsy population of congenital heart disease. *Hum Pathol.* 1998;29:689–695.
14. Taylor AJ, Rogan KM, Virmani R. Sudden cardiac death associated with isolated congenital coronary artery anomalies. *J Am Coll Cardiol.* 1992;20:640–647.
15. Benson PA, Lack AR. Anomalous aortic origin of left coronary artery. Report of two cases. *Arch Pathol.* 1968;86(2):214–216.
16. Cohen LS, Shaw LD. Fatal myocardial infarction in an 11-year-old boy associated with a unique coronary artery anomaly. *Am J Cardiol.* 1967;19:420–423.
17. Jokl E, McClellan JT, Ross GD. Congenital anomaly of left coronary artery in young athlete. *JAMA.* 1962;182:572–573.

Main Points

- Isolated single coronary artery is reported to occur in approximately 0.024%–0.04% of the population.
- It may occur with or without associated congenital anomalies.
- When the single ostium arises from the right cusp, and the left main artery traverses between the ascending aorta and the pulmonary outflow tract, the patient is at increased risk of myocardial infarction (MI) and sudden death.
- If the left main artery travels anterior or posterior to both great vessels or passes between the great vessels by way of an intramyocardial bridge the risk of sudden death or MI is felt to be quite low.