Sudden Cardiac Arrest at the Finish Line:

In Coronary Ectopia, the Cause of Ischemia
Is from Intramural Course, Not Ostial Location

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A 26-year-old woman, a well-trained runner, had a sudden cardiac arrest just before crossing the finish line of a marathon. She was rapidly resuscitated and was later found to have an ectopic origin of the left coronary artery. This anomaly was surgically repaired by translocating the ostium from the right to the left sinus of Valsalva. Her difficult postoperative course prompted further coronary evaluation, which revealed severe stenosis of the neo-ostium. The patient underwent a second operation: this time, the stenosis was bypassed via a left internal mammary artery-to-left anterior descending coronary artery (LAD) graft. Hypoplasia of the LAD and spasm during manipulation caused the graft to fail, necessitating double-stent angioplasty of the left main ostium and the LAD 2 months later. At the patient's 6-month follow-up examination, she had no further evidence of functional ischemia, and she resumed jogging.

Because the mode and mechanism of the patient's condition and events were documented in unusual detail, this case furthers our understanding of sudden cardiac arrest in athletes who have rare coronary anomalies. We conclude that ectopia of a coronary artery does not itself cause potentially fatal ischemia. Rather, these events are due to the ectopic artery's intramural proximal course within the aortic media, which might result in critical stenosis by means of hypoplasia or lateral compression of the artery. (*Tex Heart Inst J* 2014;41(2):212-6)

Congenital coronary artery anomalies (CCAs) are important causes of sudden cardiac arrest (SCA).¹⁻³ We report the case of a patient in whom the mode and pathophysiology of SCA were documented to a thorough degree. The details of this case add to our knowledge of CCAs and the mechanisms of SCA in young athletes who have these rare conditions.

Key words: Cardiac surgical procedures; coronary stenosis/etiology; coronary vessel anomalies/classification/complications/diagnosis/surgery; coronary vessels/ultrasonography; death, sudden, cardiac/etiology/pathology/prevention & control; sinus of Valsalva/abnormalities; treatment outcome; ultrasonography, interventional

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Case Report

A 26-year-old woman, a well-trained and accomplished runner (body mass index, 16.7 kg/m²), collapsed 200 meters from the finish line of a 26.2-mile marathon after running for 3 hours 27 minutes. She was rushed to a nearby medical tent and was found to be pulseless. A rhythm strip revealed ventricular fibrillation. After undergoing brief chest compressions, she was given 1 mg of intravenous epinephrine followed by 2 direct-current cardioversions. Sinus rhythm was restored, and a pulse was palpable.

The patient was intubated and transported to a nearby hospital, where a 12-lead electrocardiogram showed sinus tachycardia with inferolateral ST-segment depression. She was rushed to our tertiary center. A peak troponin I level of 1.88 ng/mL and a creatine kinase level of 6,426 U/L were noted. Thirty-six hours after the SCA and after hypothermia was discontinued, she made a full neurologic recovery. She was soon weaned from mechanical ventilation.

On day 6 after the SCA, echocardiograms showed normal left ventricular (LV) wall thickness and motion. Cardiac magnetic resonance images revealed that the left main coronary artery (LMCA) originated ectopically from the right coronary sinus (Fig. 1) and that the proximal left main (LM) trunk probably had an intramural course, inside the aortic wall. The LMCA ran between the aorta and pulmonary artery (pursuing an interarterial or "preaortic" course) before branching into the left anterior descending coronary artery (LAD) and left circumflex coronary artery. A viability study showed only small focal areas of intramural hyperenhancement, suggesting a small scar, at the mid-anteroseptal LV wall. On day 9, coronary angiograms revealed an anoma-

lous origin of the left coronary artery (LCA) from the right sinus of Valsalva, with a straight-down course and severe ostial stenosis, the stenosis more apparent in some projections than in others and worse during systole (Fig. 2). Selective cannulation of the left coronary ostium was difficult to achieve.

The patient underwent surgical correction with the aid of extracorporeal circulation. The LMCA was detached with a button of aortic wall and was reimplanted into the left sinus. The aortic sinus wall was repaired with a pericardial patch to resuspend the anterior commissure of the aortic valve. During surgical inspection, the left coronary ostium was found to be “slit-like” and the LM trunk to be directed inferiorly and within the aortic sinus wall.

The patient’s recovery was complicated by low cardiac output and hemodynamic instability that necessitated prolonged vasopressor therapy. An electrocardiogram showed a new anteroseptal and lateral myocardial infarction, with ST-segment elevation. Echocardiograms revealed significant new impairment of LV function, with an LV ejection fraction (LVEF) of 0.30 and global hypokinesis. Twenty days after the SCA, the patient was discharged from the hospital with a prophylactic, wearable cardioverter-defibrillator.

The patient’s cardiac functional status only partially recovered. Two months postoperatively, she underwent

a treadmill test (standard Bruce protocol) because she wanted to resume physical training. She reached the 12th minute with no ventricular arrhythmias or ischemic changes. Follow-up coronary computed tomographic angiograms showed LM ostial stenosis. Results of selective coronary angiography confirmed severe obstruction of the LCA neo-ostium (Fig. 3). Consequently, the patient underwent an off-pump left internal mammary artery (LIMA)-to-LAD bypass. She had no perioperative complications, although the LAD was small in caliber and vulnerable to severe spasm during surgical manipulation. Subsequent cardiac magnetic resonance images showed that 18% of the total myocardium was scarred.

Fig. 1 Magnetic resonance image shows an ectopic origin of the left coronary artery from the right sinus of Valsalva. Note the positions of the aortic commissures (*) and of the right and left coronary artery proximal trunks. Although ostial stenosis of the left coronary artery could not be definitely diagnosed, an intramural course was suggested. The ostium of the left coronary artery (arrow) is just to the right of the anterior commissure.

L = left coronary sinus; LCA = left coronary artery; NC = noncoronary sinus; R = right coronary sinus; RCA = right coronary artery

Fig. 2 **A)** Systolic and **B)** diastolic frames from a subselective left coronary angiogram (mid-right anterior oblique projection) show ostial stenosis that is critically severe in systole (*) but less severe in diastole (obviously undergoing phasic changes), with a slit-like appearance. The right coronary artery has a co-dominant pattern.

Fig. 3 Selective left coronary angiogram shows critical left main ostial stenosis (#) and mid-left anterior descending coronary artery stenosis (*) at the distal anastomosis of the left internal mammary artery (LIMA) graft. Arrows show the ghost of the LIMA graft.

Two months after the bypass operation, the patient emergently presented with palpitations and dizziness precipitated by running on a treadmill. She was alert but pale and hypotensive, with ventricular tachycardia (200 beats/min). Upon emergency admission to the hospital, she underwent cardioversion and was started on antiarrhythmic therapy. Coronary angiograms showed severe, discrete narrowing of the left main ostium and the LAD at its distal anastomosis with the LIMA. The patient underwent percutaneous coronary intervention to treat the LMCA and mid-LAD lesion. During that procedure, intravascular ultrasonography (IVUS) revealed severe stenosis in the proximal LMCA; the diameter was 1.5×1.7 mm, compared with 4.5×5 mm in the distal portion (Fig. 4). The LM stenosis was related more to ostial hypoplasia than to lateral compression: the ostial cross-sectional area was 5.02 mm^2 and the distal LMCA area was 14.91 mm^2 , leading to a 66% area stenosis. The media was thickened (as expected, because it consisted of aortic media), but the intima was not. Two PROMUS Element™ Plus Everolimus-Eluting stents (Boston Scientific Corporation; Natick, Mass) were inserted: one (2.5×12 -mm) at the mid LAD and one (3×12 -mm) at the LM ostium (Fig. 5). If necessary in the future, the proximal stent can be dilated up to a 4.25-mm diameter.

After stent placement, the patient did well clinically. Because her LV function was persistently impaired (LVEF, 0.38), an automatic cardioverter-defibrillator was implanted. Eighteen months later, device interro-

gation revealed no shocks, and maximal stress testing evoked no functional ischemia. The patient returned to work and resumed jogging.

Discussion

A trained athlete—an unknown carrier of a CCA—sustained a cardiac arrest during strenuous exertion. Her LV function was essentially preserved after immediate resuscitation (LVEF, 0.60). Surgical reimplantation was accompanied by severe LV impairment (LVEF, 0.30), which at first only partially resolved to 0.38 after revascularization with stent angioplasty. Recent investigations of CCAs point to several new, relevant conclusions. First, only a few types of CCAs (as anatomically described) can cause SCA or sudden cardiac death in the adult, and these conditions usually occur during exercise.^{4,7} Potentially fatal CCAs after infancy are essentially restricted to the group known as ACAOS (anomalous origin of a coronary artery from the opposite sinus, with an intramural course).^{8,9}

In these anomalies, the mechanism of ischemia (the main manifestation of coronary dysfunction) is consistently related to the presence of an intramural course, which results in variable degrees of both hypoplasia and lateral compression. Clinically, only IVUS can confirm and quantify the ischemic mechanism,^{8,9} a finding that has expanded the preliminary evidence of stenosis on cross-sectional histologic studies in sporadic autopsy cases.^{4,6} Under resting conditions, in patients with ACAOS, IVUS has documented variable severity of systolic compression of the ectopic artery, which usually seems to increase during simulated exertion upon catheterization.^{8,9} Maximal exercise increases oxygen demand and simultaneously worsens coronary narrowing, a potentially deadly combination. Nevertheless, individuals with ACAOS might experience ischemia only during extreme manifestations—SCA or sudden cardiac death is frequently the first reported event in these case—but not typically during submaximal clinical exercise protocols.⁶

In documenting the critical features of a dramatic event in a patient with ACAOS, the present case helps to further our understanding of similar episodes of SCA. The LMCA was ectopic, originating just to the right of the anterior commissure and coursing downward inside the aortic root wall. During the initial surgical repair, the ectopic origination itself was assumed to have caused the SCA episode, and reimplantation of the LCA ostium was accomplished. However, the patient's early postoperative course revealed the inadequacy of simple ostial translocation. Documentation of cardiac failure and selective myocardial damage in the left coronary territory provided substantial evidence of the critical severity of the residual LM intrinsic stenosis. As IVUS revealed, simply transferring the ostium to the

Fig. 4 Intravascular ultrasonographic images, obtained before and after percutaneous coronary intervention. Preoperative views of the **A**) left coronary artery (LCA) ostium and **B**) distal (extramural) left main trunk. In **A**, the circle of large squares represents the lumen of the internal elastic lamina (IEL); the circle of small squares represents the lumen (calibration marks at 1-mm intervals). In **B**, the circle of squares represents the IEL. **C**) Post-operative image of the ostial LCA stent (nominal diameter, 3 mm; postdilated diameter, 3.8 mm). The LCA ostium features a thin intima (0.2 mm) but a relatively thick media (0.6 mm). In this case, the main mechanism of stenosis was hypoplasia, as compared with the lumen at the distal LCA (**B**). The 3 cross-sectional images show the following features: in **A**, the internal elastic area (IEA) is 6.6 mm² (circle of large squares), and the luminal area is 3.14 mm² (circle of small squares). In **B**, the IEA is 15.3 mm² (circle of squares); in **C**, the area is 9.62 mm² (circle of squares). The area of stenosis is 80% before stent angioplasty and 38% afterwards. A similar stent can be redilated, if necessary, up to 4.25 mm in diameter.

proper sinus did not mitigate the intramural deformity of the proximal LM trunk.

In this regard, it is relevant to report the newly apparent spectrum of the “intramural LM” or L-ACAOS types (Fig. 6):

- Type 1: Origin from the right sinus, the most frequent form of L-ACAOS, with a longer intramural course inside the sinotubular junction (dextroposed L-ACAOS).⁹
- Type 2: Ectopic origin to the right of, and next to, the anterior commissure, with a downward course inside the aortic sinus, in front of the anterior aortic commissure (juxtacommissural right-sided L-ACAOS, as in our patient).
- Type 3: Origin from the noncoronary sinus, sometimes running intramurally at the sinotubular junction from the posterior-sinus origin (noncoronary

L-ACAOS, which also might not have any intramural course or stenosis).⁹

- Type 4: Normal origin at the left sinus of Valsalva, with a straight-down course inside the left aortic sinus, as reported by our group (orthoposed ostium L-ACAOS).¹⁰
- Type 5 (not shown in Fig. 6): Ostium above the left sinus of Valsalva, with or without an intramural course and stenosis (high-origin L-ACAOS).

In all such intramural variants, the mechanisms of LM trunk stenosis involve variable degrees of both hypoplasia and lateral compression inside the aortic media. To eliminate the stenosis, such an intramural segment needs to be trimmed or patched,¹¹ and either approach involves delicate techniques. Stent implantation, tested successfully in R-ACAOS, is theoretically also a primary alternative to surgery; however, it is generally considered

to be a 2nd choice, recommended chiefly for patients in whom surgery has failed or would be too risky.¹²

We have presented a case of SCA in a well-trained young marathon runner who was found to have a CCA

Fig. 5 Postoperative angiogram (right anterior oblique projection) shows the success of angioplasty in the stenotic segments of the ostial (#) and mid-left anterior descending coronary artery (*) in the proximal anomalous origin of the left coronary artery from the opposite sinus.

Fig. 6 Diagram shows 4 of the 5 known alternative types of anomalous left coronary artery with an intramural course (L-ACAOS). Type 5 (not shown) is highly similar to type 3 but has an abnormally high origin from the ascending aorta.

Cx = left circumflex coronary artery; L = left sinus of Valsalva; LAD = left anterior descending coronary artery; NC = noncoronary sinus; R = right sinus of Valsalva; RCA = right coronary artery

with unusual anatomic and functional features. In such cases, it is extremely important that the coronary anatomy be properly evaluated by means of intravascular imaging in order to clarify the mechanism of coronary dysfunction, properly plan for surgical repair, and follow up with interventions.

References

1. Eckart RE, Scoville SL, Campbell CL, Shry EA, Stajduhar KC, Potter RN, et al. Sudden death in young adults: a 25-year review of autopsies in military recruits. *Ann Intern Med* 2004;141(11):829-34.
2. Eckart RE, Shry EA, Burke AP, McNear JA, Appel DA, Castillo-Rojas LM, et al. Sudden death in young adults: an autopsy-based series of a population undergoing active surveillance. *J Am Coll Cardiol* 2011;58(12):1254-61.
3. Maron BJ, Haas TS, Doerer JJ, Thompson PD, Hodges JS. Comparison of U.S. and Italian experiences with sudden cardiac deaths in young competitive athletes and implications for preparticipation screening strategies. *Am J Cardiol* 2009;104(2):276-80.
4. Angelini P. Coronary artery anomalies--current clinical issues: definitions, classification, incidence, clinical relevance, and treatment guidelines. *Tex Heart Inst J* 2002;29(4):271-8.
5. Angelini P, Monge J. Coronary artery anomalies. In: Moscucci M, editor. *Baim & Grossman's cardiac catheterization, angiography, and intervention*. Philadelphia: Lippincott Williams & Wilkins; 2013. p. 335-53.
6. Frescura C, Basso C, Thiene G, Corrado D, Pennelli T, Angelini A, Daliento L. 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(7):689-95.
7. Angelini P. Coronary artery anomalies and sports activities. In: Lawless CE, editor. *Sports cardiology essentials: evaluation, management and case studies*. Chicago: Springer; 2010. p. 277-98.
8. Angelini P, Flamm SD. Newer concepts for imaging anomalous aortic origin of the coronary arteries in adults. *Catheter Cardiovasc Interv* 2007;69(7):942-54.
9. Angelini P, Walmsley RP, Liberos A, Ott DA. Symptomatic anomalous origination of the left coronary artery from the opposite sinus of Valsalva. Clinical presentations, diagnosis, and surgical repair. *Tex Heart Inst J* 2006;33(2):171-9.
10. Angelini P, Walmsley R, Cheong BY, Ott DA. Left main coronary artery originating from the proper sinus but with acute angulation and an intramural course, leading to critical stenosis. *Tex Heart Inst J* 2010;37(2):221-5.
11. Ito H, Takabayashi S, Kajimoto M, Shimpo H. Patch angioplasty and neo-ostium creation for intramural left coronary artery. *Interact Cardiovasc Thorac Surg* 2011;13(6):661-3.
12. Nef HM, Mollmann H, Mollmann S, Dill T, Hamm CW, Elsasser A. Primary stenting of the left main coronary artery with anomalous origin from the right sinus of Valsalva. *Int J Cardiol* 2007;114(1):137-8.