

# Resuscitated sudden cardiac death due to diminutive coronary artery syndrome



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## Introduction

Diminutive coronary artery syndrome (DCAS) refers to myocardial ischemia occurring as a consequence of coronary artery hypoplasia. DCAS was first described in 1964 in a report of 2 previously healthy men, aged 18 and 25, who suffered acute myocardial infarctions (MI) and were found on rudimentary coronary angiography to have hypoplastic right coronary arteries.<sup>1</sup> Subsequent case reports have described an association between DCAS and sudden cardiac death (SCD), with postulated mechanisms of ischemia-induced or scar-mediated reentrant ventricular arrhythmia.<sup>2,3</sup> The incidence of DCAS is unclear and, to date, antemortem clinical criteria for this disorder have not been proposed, which may lead to substantial under-diagnosis.

Herein, we describe clinical and pathophysiologic features of a patient experiencing SCD owing to MI and found to have diminutive coronary arteries on angiography.

## Case report

A 53-year-old woman with no significant past medical history developed left-sided chest and arm pain while riding a motorcycle. She had no antecedent angina despite practicing mixed martial arts 3 times weekly. Prior to admission, she was stopped at a traffic light, lost consciousness, and fell to the ground. Cardiopulmonary resuscitation was initiated by witnesses and subsequently continued by emergency medical personnel. A rhythm strip demonstrated ventricular fibrillation (VF) and external defibrillation successfully restored sinus rhythm and spontaneous circulation.

An electrocardiogram obtained upon arrival to the emergency department demonstrated sinus rhythm with a normal QT interval and T-wave inversions localized to the anteroapical precordial leads, as illustrated in [Figure 1](#). T-wave inversions resolved after 24 hours. Coronary angiography revealed a hypoplastic left anterior descending artery that

measured <2.0 mm at its origin and that terminated in the mid myocardium. The left circumflex artery also measured 2.0 mm at the origin and gave rise to 2 small obtuse marginal branches with reduced vessel length, terminating in the second third of the lateral wall ([Figure 2](#)). The right coronary artery was a 2.5-mm vessel at its origin and supplied both posterior descending and posterolateral branches. There was no angiographic evidence of coronary atherosclerosis. Serum troponin level peaked at 28 ng/L (normal range <0.02 ng/dL). A transthoracic echocardiogram demonstrated distal septal, distal lateral, and apical wall hypokinesis. A cardiac magnetic resonance imaging (MRI) examination demonstrated no evidence of infiltrative disease or myocardial scar as assessed by late gadolinium contrast enhancement, but did reveal transmural edema of the septum and apex consistent with recent MI ([Figure 3](#)).

Her diagnostic evaluation revealed no evidence of long QT syndrome, arrhythmogenic right ventricular cardiomyopathy, hypertrophic obstructive cardiomyopathy, recent myocarditis, or Brugada syndrome. Erythrocyte sedimentation rate and C-reactive protein were measured to exclude vasculitis and were within normal limits. The patient underwent implantation of an automated implantable cardioverter-defibrillator (ICD) prior to discharge and has remained symptom free without recurrent syncope or tachyarrhythmia detected on device interrogation. An echocardiogram performed 6 months after the index hospitalization showed persistence of the aforementioned wall motion abnormalities but preserved overall left ventricular systolic function.

## Discussion

DCAS is a rare clinical entity comprising myocardial ischemia in the setting of coronary artery hypoplasia. The incidence of DCAS in the general population is unknown, as the condition is generally diagnosed at autopsy and reported in highly selected cohorts. In an autopsy study of 224 patients with coronary anomalies, 1 or more hypoplastic coronary arteries were identified in 5 patients (2.2%).<sup>3,4</sup> In another study of 158 competitive athletes who suffered SCD, hypoplastic coronary arteries were identified in <5% of cases at autopsy.<sup>5</sup>

**KEYWORDS** Diminutive coronary artery; Sudden cardiac death; Ventricular fibrillation; Myocardial infarction; Implanted cardiac defibrillator (*Heart Rhythm Case Reports* 2017;3:141–144)

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## KEY TEACHING POINTS

- Diminutive coronary artery syndrome (DCAS) refers to myocardial ischemia occurring as a consequence of coronary artery hypoplasia.
- DCAS is associated with an increased risk of sudden cardiac death, possibly owing to ischemia-induced or scar-mediated reentrant ventricular arrhythmia.
- Adenosine coronary flow reserve testing and acetylcholine provocative vasomotor testing may be helpful in distinguishing coronary microvascular disease from coronary artery hypoplasia.

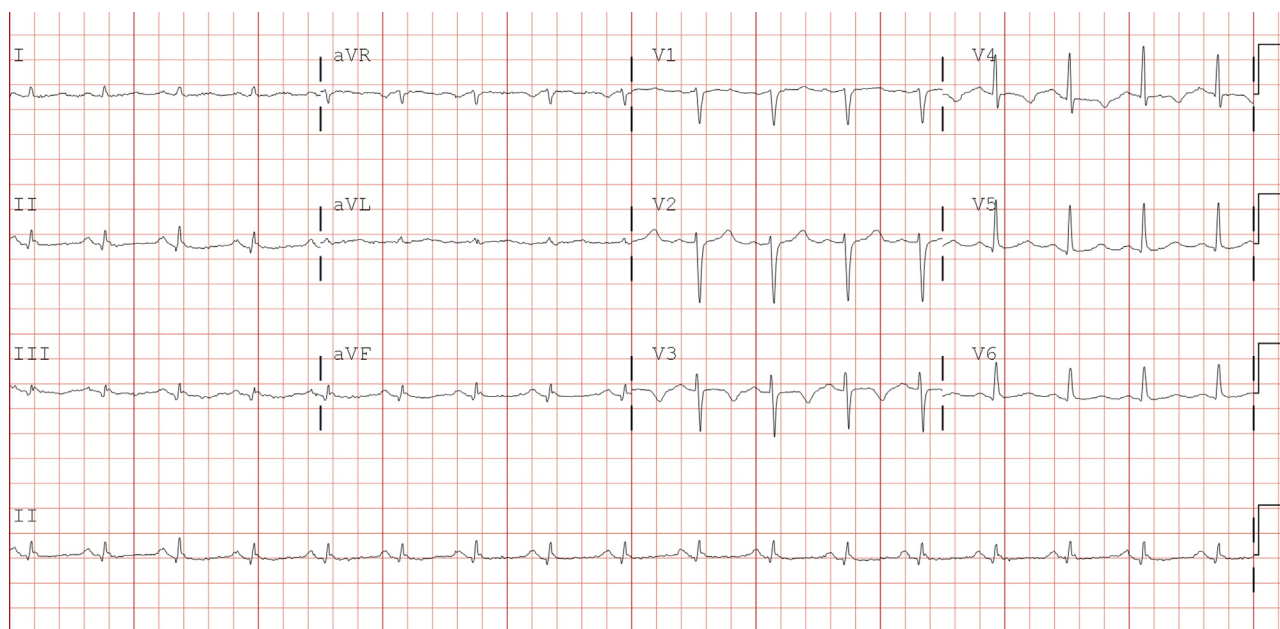
Ischemia-mediated ventricular arrhythmia is typically invoked as the mechanism of SCD in this population, but objective evidence to support this assertion is lacking in most reports.<sup>6,7</sup> Moreover, many of the described cases attributing SCD and MI to diminutive coronary arteries are confounded by the presence of other congenital abnormalities, including anomalous coronary arterial origin.<sup>8</sup> Importantly, previous definitions proposed for DCAS have required postmortem histopathologic evidence of reduced vessel length and diameter to make a definitive diagnosis.<sup>9</sup> In the current era, antemortem identification of patients with DCAS is essential to facilitate secondary prevention of SCD through ICD implantation.

In this case, SCD was clearly attributable to VF and myocardial ischemia/infarct, documented by electrocardiography, echocardiography, cardiac MRI, and elevated cardiac biomarkers. This occurred in the setting of diminutive coronary arteries without evidence of intracoronary

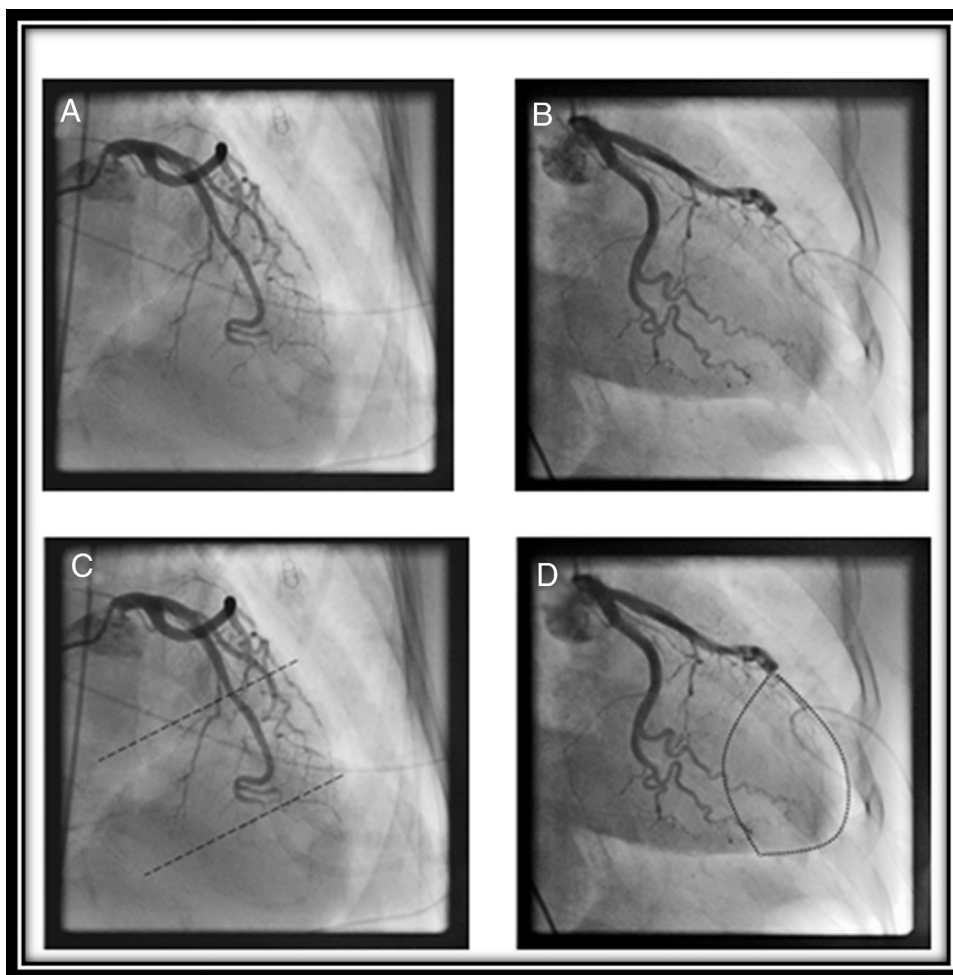
thrombosis or epicardial arterial stenosis. Importantly, the presence of focal segmental wall motion abnormalities in distal myocardial territories beyond a single coronary arterial distribution on echocardiography suggests that a “watershed” perfusion deficit may be pathophysiologically operational. This mechanism is analogous to that observed in both cortical and internal watershed strokes, whereby an infarct occurs owing to hypoperfusion of a terminal area subtended by >1 intracerebral artery.<sup>10</sup> Variations in vessel size are frequently encountered in the coronary vasculature, but the small distribution of 1 artery is typically compensated for by the greater distribution of another vessel. In the current case, the left coronary arteries were hypoplastic while the right coronary artery was relatively normal in size, leading to a watershed area, as evidenced by echocardiography and cardiac MRI. These findings suggest a causal connection between diminutive left coronary arteries and resultant ischemia and infarction.

Given these findings and review of previous literature, we propose the following antemortem clinical criteria to identify SCD or MI as a direct consequence of DCAS:

- (1) Angiographic evidence of diminutive coronary artery or arteries, defined as termination of the arterial tree less than or equal to two thirds the distance from coronary origin to the heart border and a vessel origin  $\leq 2$  mm diameter.
- (2) No angiographic evidence of atherosclerosis.
- (3) Electrocardiographic evidence of myocardial ischemia/infarction or elevation of plasma cardiac biomarkers.
- (4) Absence of an alternative causal explanation for SCD or MI.



**Figure 1** Electrocardiogram at presentation, demonstrating normal sinus rhythm with a QT interval of 390 milliseconds and inverted T waves in V3 and V4.



**Figure 2** Coronary angiography. **A:** Cineangiography in the right anterior oblique (RAO) cranial projection demonstrated a diminutive left anterior descending artery (LAD) terminating in the mid anterior wall. **B:** RAO caudal views demonstrated small obtuse marginal branches and an absence of left-to-left collateral vessels. **C:** The LAD extended less than two-thirds of its expected course. **D:** A watershed zone between the LAD and left circumflex artery territories.

Current professional society guidelines include a class I indication for the use of ICDs in patients who have suffered SCD owing to ventricular tachycardia or VF in the absence of a reversible cause.<sup>11</sup> Our patient underwent ICD implantation but has not had recurrent arrhythmia. To date, arrhythmia surveillance data in DCAS patients have not been described, so the utility of device therapy in this population requires further study.

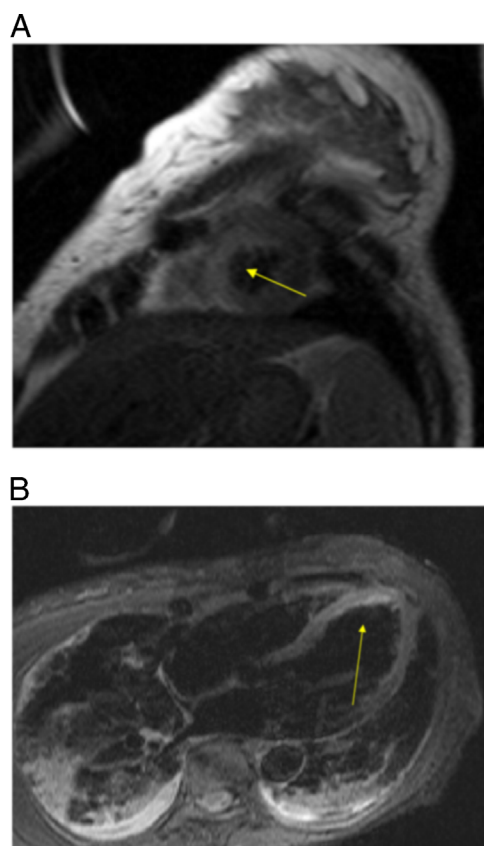
We acknowledge a number of important limitations to the current case description and proposed framework for evaluating this condition. Myocardial perfusion imaging demonstrating ischemia in the aforementioned watershed territories was not performed. However, echocardiography on presentation demonstrated wall motion abnormalities in the distal lateral wall, distal septum, and apex consistent with ischemia/infarction in the left anterior descending and circumflex coronary artery territories, which was corroborated by MRI. Based on the available data, we are unable to exclude the possibility that coronary microvascular disease provoked ischemia in this patient. Unfortunately, adenosine coronary flow reserve testing and acetylcholine provocative

vasomotor testing were not performed in this case. However, the presence of normal TIMI flow at angiography and an absence of atherosclerosis risk factors in this patient make this diagnosis unlikely.

The true prevalence of DCAS remains unknown in the general population. Even among patients undergoing angiography it is likely underestimated, since detailed anatomic information about size and extent of coronary artery distributions is not a feature of current registries, nor is this condition well known to cardiologists who are more familiar with anomalous coronary artery origin.<sup>12</sup> We speculate that this condition may be more frequent than appreciated, analogous to the original description of stress-induced cardiomyopathy as a cause for MI in the absence of epicardial atherosclerosis, where the prevalence may increase with greater clinician awareness.<sup>13</sup>

## Conclusions

To our knowledge, this is the first report to propose a pathophysiologic mechanism and diagnostic criteria for



**Figure 3** Cardiac magnetic resonance imaging. Gadolinium-enhanced magnetic resonance images demonstrating edema in the septum (A) and apex (B).

DCAS that do not involve an autopsy examination. DCAS is characterized by the triad of myocardial ischemia/infarction, angiographic evidence of coronary artery hypoplasia, and the absence of obstructive coronary artery disease. In the current case, angiography demonstrated diminutive coronary arteries

with cardiac imaging findings suggestive of watershed hypoperfusion. Patients with DCAS are likely at increased risk for recurrent ventricular arrhythmias and SCD and may benefit from electrophysiology evaluation and, ultimately, ICD therapy. Further studies characterizing the natural history and prognosis of DCAS patients are warranted.

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