

THE PRESENT AND FUTURE

STATE-OF-THE-ART REVIEW

Contemporary Review on Spontaneous Coronary Artery Dissection



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ABSTRACT

Spontaneous coronary artery dissection (SCAD) is gaining recognition as an important cause of myocardial infarction, especially in young women. There has been a surge in the diagnosis of SCAD in recent years, presumably due to an increased use of coronary angiography, and the clinical availability and application of high-resolution intracoronary imaging. The improved recognition and diagnosis, together with increased publications and attention through social media, have considerably raised awareness of this condition, which was once believed to be very rare. Recent publications of moderate to large contemporary case series have helped elucidate the early natural history, presenting characteristics (clinical and angiographic), underlying etiology, management, and cardiovascular outcomes with this condition, thus providing observations and important clinical insights of value to clinicians managing this challenging and perplexing patient cohort. The aim of our review is to provide a comprehensive contemporary update of SCAD to aid health care professionals in managing these patients in both the acute and chronic settings. (J Am Coll Cardiol 2016;68:297-312)
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Spontaneous coronary artery dissection (SCAD) is gaining recognition as an important cause of myocardial infarction (MI), especially in young women. The first report of SCAD, by Pretty (1) in 1931, was of a 42-year-old woman who died unexpectedly following repetitive retching and vomiting, which elicited coronary artery rupture from dissection of an atheromatous aneurysm. Over the next 8 decades, subsequent SCAD publications consisted mostly of isolated case reports and small case series. However, there has been a surge in the diagnosis of SCAD in recent years, presumably due to the increased use of coronary angiography in patients presenting with acute coronary syndromes (ACS), as well as the clinical availability and application of high-resolution intracoronary imaging (especially optical coherence tomography [OCT]) that enhances diagnosis (2-4). As such, of the ~1,500 reported

SCAD cases to date, about one-half were published in the past 5 years (5-15).

The improved recognition and diagnosis, together with the increased number of publications and attention through social media, have considerably raised awareness of this condition (16), once believed to be very rare. The first online community of SCAD patients on Inspire (17), developed by WomenHeart (The National Coalition for Women With Heart Disease) as a social networking site, has improved patient awareness. Simultaneously, publications of larger contemporary case series of SCAD have helped elucidate the early natural history, presenting characteristics (clinical and angiographic), the underlying etiology, the management, and the outcomes of this condition. These studies have provided observations and important clinical insights, reflected in expert opinions on management strategies, which are of



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**ABBREVIATIONS
AND ACRONYMS****ACS** = acute coronary syndrome**CABG** = coronary artery bypass graft**CTA** = computed tomography angiography**DAPT** = dual antiplatelet therapy**FMD** = fibromuscular dysplasia**IMH** = intramural hematoma**IVUS** = intravascular ultrasound**MACE** = major adverse cardiac events**MI** = myocardial infarction**MRA** = magnetic resonance angiography**OCT** = optical coherence tomography**PCI** = percutaneous coronary intervention**SCAD** = spontaneous coronary artery dissection**STEMI** = ST-segment elevation myocardial infarction**TIMI** = Thrombolysis In Myocardial Infarction

value to clinicians managing this challenging and perplexing patient cohort. The aim of our review is to provide a comprehensive contemporary update of SCAD to aid health care professionals managing these patients in both the acute and chronic settings.

DEFINITION OF SCAD

SCAD is defined as a spontaneous separation of the coronary artery wall that is not iatrogenic or related to trauma. As such, dissections due to blunt trauma, surgical instruments, or those that are catheter-induced are not deemed to be SCAD. Furthermore, contemporary usage of the term SCAD is typically reserved for the nonatherosclerotic variant, and most modern series exclude SCAD due to atherosclerotic coronary artery disease. For the purpose of our review, and because this disease is distinct from atherosclerotic disease, the term SCAD refers to “nonatherosclerotic SCAD” and is the focus of this paper.

EPIDEMIOLOGY

SCAD was previously, incorrectly believed to be very rare and to be frequently associated with pregnancy. Unfortunately, the true incidence and prevalence of SCAD in the general population is unknown due to significant underdiagnosis of this condition. For decades, SCAD diagnosis was plagued by a low clinical index of suspicion of seemingly healthy young women presenting with ACS who had not undergone coronary angiography. At the other extreme, SCAD patients who presented with sudden cardiac death may not have been captured in various databases. In addition, the current “gold standard” for a SCAD diagnosis (i.e., coronary angiography) has major limitations because it does not image the arterial wall, and many clinicians are unfamiliar with nonpathognomonic angiographic variants of SCAD. Furthermore, intracoronary imaging was infrequently used to aid diagnosis. Therefore, the previous reports of SCAD prevalence on coronary angiography of 0.2% to 1.1% were underestimates of the true prevalence of SCAD (18-20). In a recent Japanese series of 326 ACS patients who underwent routine OCT imaging, SCAD was diagnosed in 4% of cases (21). However, caution should be exercised in using this incidence estimate because 77% of these patients were men, and thus a proportion of these dissections were likely related to atherosclerosis. We suspect that a more accurate estimate of the SCAD prevalence in patients presenting

with ACS is 1.7% to 4% on the basis of modern series (13,21). In the extreme case of sudden cardiac death, SCAD was reported in 0.5% (8 of 1,647) of cases in an autopsy series (22), although this prevalence could be an underestimate.

The incidence of SCAD in young women has been further explored. In the older series by Vanzetto et al. (18) of women younger than 50 years of age who presented with ACS, the prevalence of SCAD was 8.7% (10.8% in patients with ST-segment elevation myocardial infarction [STEMI]). Contemporary series reported a much higher prevalence of SCAD in young women with ACS (22% to 43%). In a single-center Canadian series of women younger than 50 years of age who underwent coronary angiography (n = 177; mean age 45.4 years), SCAD was observed in 9.0%, and was the cause of MI in 24.2% (23). In a recent Japanese series, SCAD was the cause of MI in 35% (45 of 130) of women younger than 50 years of age (15). In a recent Australian series of women younger than 60 years of age (n = 23) who underwent angiography, SCAD accounted for 22.5% of ACSs (13). In the series by Elkayam et al. (24) of much younger women (mean age 34 years) with pregnancy-related MI, SCAD was the most frequent cause of MI, accounting for 43% of MIs (56 of 129), which occurred predominantly in the third trimester or post-partum phase. Therefore, the incidence of SCAD is not rare in young women presenting with ACS, especially those with pregnancy-related MI.

SCAD affects women in >90% of cases. In contemporary series that excluded patients with atherosclerotic causes, women accounted for 92% to 95% of the population with SCAD (6,9,10,12-14). The reported mean age ranged from 44 to 55 years in contemporary series, reflecting a relatively young to middle-age population (6,9,10,12-15). In the Vancouver cohort of 168 patients, 58% were ≥50 years of age, 62% of affected women were post-menopausal, and the oldest patient was 84 years of age (9). In another series, the oldest reported patient was 78 years of age (7). In terms of racial distribution, all races can be affected, although most of those in North America were Caucasians (81% to 83%) (7,9), which was similar to the racial distribution among patients who underwent percutaneous coronary intervention (PCI) in British Columbia.

MECHANISM AND PATHOPHYSIOLOGY

The arterial dissection with SCAD can occur within or between any of the 3 layers (intima, media, or adventitia) of the coronary artery wall. Two potential mechanisms for the initiation of arterial wall

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