Spontaneous Coronary Artery Dissection (SCAD)

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The coronary arteries carry oxygen- and nutrient-rich blood to the heart, which allows it to pump and supply blood to the body. The coronary arteries can become blocked by various processes related to cholesterol (atherosclerotic) or other causes (nonatherosclerotic), which can cause heart attacks when the blockages are severe enough to impede blood flow to the heart. Although the atherosclerotic form is most common, nonatherosclerotic blockages (especially dissections/tears) are increasingly recognized as important causes of heart attacks, particularly in women. This article focuses on spontaneous coronary artery dissection (SCAD).

What Is SCAD?

SCAD is defined as a tear in the coronary arterial wall that is not related to trauma or medical instrumentation. This spontaneous tear can result from a disruption of the innermost lining of the arterial wall, causing blood to rush in from the lumen (center channel of the artery) into the arterial wall. Alternatively, separation of the arterial walls can occur as a result of bleeding caused by spontaneous rupture of small vessels that feed the arterial wall. The buildup of blood inside the arterial wall compresses the lumen, blocking blood flow to the heart.

How Frequent Is SCAD?

SCAD is considered to be rare in the general population, although it has been underdiagnosed, and its true prevalence is unknown. Overall, it is an infrequent cause of and accounts for only up to 4% of heart attacks.[1] However, SCAD proportionally causes a higher number of heart attacks in young women; ≈25% of women ≤50 years old had heart attacks caused by SCAD in 1 series.[2]

Who Is Affected by SCAD?

SCAD affects predominantly women in ≈90% of cases and can affect individuals of all ages and races. Historically, it was thought to affect mostly young women, but recent studies have shown that older and postmenopausal women could also suffer SCAD, even though the majority are <65 years old.[3] Affected patients tend not to have the typical risks for heart disease such as diabetes mellitus, smoking, or obesity. Instead, there are different risk factors for SCAD, including possible genetic predispositions to arterial weakening or fragility. The majority of cases have been sporadic without familial associations, but the genetic basis of SCAD has not been adequately explored.

What Causes SCAD?

Patients with SCAD typically have an underlying arterial disease and often have a precipitating stress event that triggers the SCAD (Table). Atherosclerosis-related SCAD is a very different condition and more often affects men; medical discussion of SCAD typically revolves around the nonatherosclerotic form. Historically, SCAD was thought to occur mostly during or after pregnancy (peripartum), but modern studies have shown that peripartum SCAD is relatively infrequent. Instead, the strongest link with SCAD is fibromuscular dysplasia (disease of abnormal cell growth in arterial wall), which has been observed in

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(*Circulation*. 2015;131:e3-e5. DOI: 10.1161/CIRCULATIONAHA.114.010773.)

***Circulation* is available at http://circ.ahajournals.org** **DOI: 10.1161/CIRCULATIONAHA.114.010773**

Table. Arterial Disease and Precipitating Events Associated With Nonatherosclerotic SCAD

Associated arterial disease
Fibromuscular dysplasia
Pregnancy: currently pregnant, recently gave birth, multiple previous pregnancies
Connective tissue disorders: Marfan syndrome, Loeys-Dietz syndrome, Ehler-Danlos syndrome type 4, cystic medial necrosis, α-1 antitrypsin deficiency, polycystic kidney disease, polycystic ovarian syndrome
Systemic inflammatory conditions: systemic lupus erythematosus, Crohn disease, ulcerative colitis, polyarteritis nodosa, sarcoidosis, Churg-Strauss syndrome, Wegener granulomatosis, rheumatoid arthritis, Kawasaki, giant-cell arteritis, celiac disease
Hormonal therapy: birth control pills, estrogen, progesterone, β-HCG, testosterone
Coronary artery vasospasm
Idiopathic (unknown cause)
Precipitating stress events
Intense exercises (isometric or aerobic forms)
Intense emotional stress
Labor and delivery
Intense bearing-down activities (eg, retching, vomiting, bowel movement, coughing)
Recreational drugs (eg, cocaine, amphetamines, methamphetamines)
Intense hormonal therapy (eg, β-HCG injections)

β-HCG indicates β-human chorionic gonadotropin; and SCAD, spontaneous coronary artery dissection.

up to 70% to 90% of SCAD patients.[3,4] Less frequently, SCAD has been associated with connective tissue disorders, systematic inflammatory diseases, and hormonal therapies (Table).[5] In a small proportion of SCAD patients, underlying genetic disorders affecting connective tissues that make up the arterial wall have been observed. Precipitating stress events such as intense emotional stresses (eg, death in the family, breakdown of marriage, job loss), physical stresses (eg, heavy weight lifting, intense aerobic workouts), drugs (eg, cocaine, methamphetamines, β-human chorionic gonadotropin), and bearing-down activities (eg, retching/vomiting, coughing, bowel movement) have commonly been reported as triggers for SCAD events.

What Are the Signs and Symptoms of SCAD?

Patients with SCAD can present with heart attacks, cardiac arrest, or sudden cardiac death. Symptoms of heart attacks in these cases can include chest, shoulder, arm, or epigastric pain, with or without shortness of breath and nausea/vomiting. The heart attack severity can range from mild to severe, which may compromise heart function, causing heart failure, low blood pressure, and other organ dysfunction. During hospitalization, heart enzymes and electrocardiograms often show changes consistent with heart attacks. Some may even have abnormal heart rhythms, causing cardiac arrest that can be lethal.

How Is SCAD Diagnosed?

The standard test to diagnose SCAD is coronary angiography, which uses an iodinated contrast agent to fill the lumen of coronary arteries and x-rays to image the lumen. This test allows visualization of narrowed arterial lumen and contrast staining inside the artery wall (Figure 1), but it often is inadequate to differentiate the cause of the narrowing. In some cases, further detailed imaging of the arterial wall with optical coherence tomography (Figure 2) or intravascular ultrasound is required to delineate the cause of narrowing, showing a tear or blood accumulation in the arterial wall with SCAD. Noninvasive tests like computed tomography or magnetic resonance angiography may miss SCAD involving smaller and more distal arteries and are therefore not definitive to rule out SCAD.

What Are the Treatment Options Available?

Most stable patients with SCAD without ongoing pain or electrocardiography changes can be treated medically. Conservative therapy has been associated with spontaneous arterial healing on subsequent angiograms.[3] Although there is no well-studied medication for SCAD treatment, long-term therapy with aspirin and β-blocker is commonly pursued.[6] Angiotensin-converting enzyme inhibitors may be administered in patients with large heart attacks and weak heart function. Statins may be administered in patients with underlying abnormal cholesterol levels. Patients with ongoing symptoms or who are unstable (low blood pressure or cardiac arrest) may be treated with coronary stenting or bypass surgery if necessary and feasible and sometimes may even require mechanical blood pressure support.

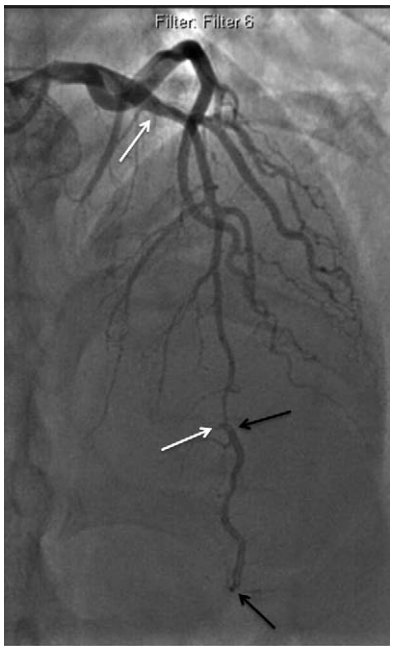

Figure 1. Long dissection of the left anterior descending artery showing long diffuse blockage (between the white arrows) and double-lumen (between the black arrows) in the artery representing different angiographic appearances of spontaneous coronary artery dissection.

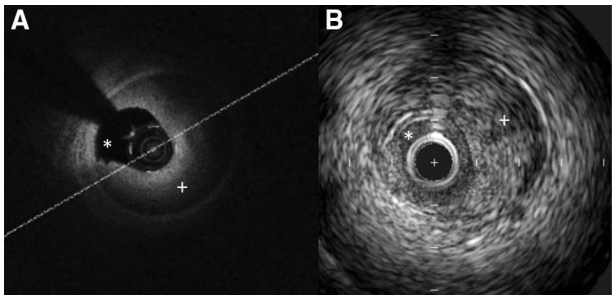

Figure 2. A, Optical coherence tomography showing blood in the artery wall (+) compressing the true lumen (*). **B**, Intravascular ultrasound showing blood in the artery wall (+) compressing the true lumen (*).

Are There Activity Restrictions for Patients With SCAD?

After a heart attack resulting from SCAD, patients should be monitored by cardiac specialists and referred to cardiac rehabilitation programs. It is common for cardiac specialists to advise avoiding intense isometric exercises, generally avoiding weights >20 to 30 pounds, and avoiding resistance training requiring marked bearing-down maneuvers. Routine aerobic activities appear relatively safe after SCAD and are encouraged to maintain physical well-being after heart attacks.

What Is the Outcome After SCAD, and What Follow-Up Care Is Recommended?

SCAD patients are at risk for recurrent cardiac events, including heart attack and recurrent SCAD, after their first event, necessitating long-term follow-up with cardiologists. Recurrent SCAD can occur in 13% to 18% of cases,[3,5] and patients should seek physician input for recurrent symptoms. Noninvasive stress testing is frequently done after a SCAD event to assess residual coronary arterial blockage. In addition, because of the association with fibromuscular dysplasia, SCAD patients are typically screened for fibromuscular dysplasia involvement in other organs. Sometimes artery outpouching (aneurysm) may be found, requiring further surveillance and management. Hormonal therapy that was thought to be the underlying cause should be discontinued. Evaluation for a genetic disorder affecting the arteries may be considered. Furthermore, SCAD survivors are advised to avoid future pregnancies.

Conclusions

SCAD is underdiagnosed and is an important cause of heart attack, especially in younger women, but it can also affect older postmenopausal women and men. Patients are usually treated conservatively but require long-term cardiac follow-up by cardiologists because recurrent SCAD and heart attacks can occur. Further research is needed to understand why SCAD occurs and to identify effective prevention and treatment strategies.

Disclosures

Dr Saw has received research grants from the Canadian Institutes of Health Research, AstraZeneca, Abbott Vascular, Servier, St. Jude Medical, and the University of British Columbia Division of Cardiology for research on SCAD. Dr Saw has also received speaker honoraria from Sunovion and speaker organization support from St. Jude Medical and AstraZeneca for SCAD research. The other authors report no conflicts.

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Circulation. 2015;131:e3-e5
doi: 10.1161/CIRCULATIONAHA.114.010773
Circulation is published by the American Heart Association, 7272 Greenville Avenue, Dallas, TX 75231

Print ISSN: 0009-7322. Online ISSN: 1524-4539