# The Vital Role of the Emergency Department

*in Improving Outcomes of* **Spontaneous Coronary Artery Dissection (SCAD)** 



A SCAD Alliance White Paper based on the 2014 SCAD Alliance Scientific Advisory Board Roundtable

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#### Dedication:

The Board of SCAD Alliance dedicates this white paper to the survivor network of patients and families around the globe, living their lives in testament to overcoming adversity and thriving. We also honor those who died from SCAD before their time and pledge to work diligently in their memory to save others.

"Much can be done to make the Emergency Department a safer place for patients and providers -- more specifically, to reduce diagnostic errors and missed diagnosis of acute cardiac ischemia. The cases discussed in this white paper effectively illustrate the challenges in considering the diagnosis of spontaneous coronary artery dissection (SCAD) and the specific issues around testing and treatment."

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#### Acknowledgements

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Special thanks to SCAD Alliance board members Kathleen Hoffman, PhD, and Katherine Leon for drafting and editing this document, and to James Feldman, MD, for his expertise in reviewing content.

Additional Information

For more on spontaneous coronary artery dissection, please see our online resources:

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# **Executive Summary**

"Time is muscle." Clinicians who work in cardiovascular patient care are faced with this reality every second, every minute, every day. During a heart attack, the vessels that feed the heart muscle are blocked and unable to provide oxygen. Every moment without oxygen means the death of heart muscle cells. Dead heart muscle does not regenerate; as you lose muscle, you lose the heart's ability to pump. That's why it is so important that heart attacks are recognized early. Medical interventions are essential to keep heart muscle alive. Patients experiencing Spontaneous Coronary Artery Dissection (SCAD) are no exception. When the artery dissects, patients suffer heart-related symptoms ranging from Acute Coronary Syndrome (ACS), heart attack and even sudden cardiac arrest.

Emergency Departments (ED) are on the frontline in the effort to save heart muscle.

## SCAD Alliance Scientific Advisory Rationale

The goal of the Roundtable panel was to identify the challenges involved with assessing and determining which tests are appropriate in determining next steps for spontaneous coronary artery dissection (SCAD) patients in the ED.

#### SCAD in the Emergency Department – Invisible?

In assessing SCAD patients, whose average age is 42 and have few if any risk factors for cardiovascular disease, ED staff face several initial challenges:

- What clues would signal the need for electrocardiogram (ECG) and troponin blood labs (the chemical marker of heart injury) if the patient is not perceived to be in cardiac distress because he or she appears to be too young and healthy for a heart attack?
- 2. If ECG is done, how do we identify "normal" for the patient if she or he is previously healthy in other words, no cardiac risk factors or medical documentation of cardiac history?
- 3. In a patient with possible SCAD, what is the appropriate diagnostic testing or imaging strategy?
- 4. How are these patients with symptoms, but with ECGs that do *not* demonstrate STEMI with elevated troponin tests, evaluated?

In reality, the SCAD patient in the midst of STEMI may not "look" like an emergency. She or he is "too young, too fit, to well" to signal heart disease. Compounding the problem for SCAD patients is the ever-increasing volume of non-emergency care delivered through our nation's EDs. The trend has broad implications.

#### Record High ED Use – A Universal Problem

With implementation of the Affordable Care Act (ACA), an increase in ED visits has been seen. Even beyond the impact of the ACA, research indicates that the number of people arriving at the ED with chest pain is rising every year.

Chest pain is a sentinel symptom of acute coronary syndromes (ACS) signaling heart attack. People experiencing chest pain and other symptoms consistent with myocardial ischemia (meaning lack of blood reaching the heart muscle) often go to their local hospital ED for care. They make up between 5-10% (or approximately 8-10 million) of the total number of ED visits in the U.S. every year.

Patients with ACS can also present with a broad range of symptoms (chest pain equivalents), especially women, older patients and those with diabetes. ACS can be heralded by shortness of breath, weakness, fatigue shoulder/arm/jaw pain and other complaints. Other serious diseases can also cause chest pain and these other symptoms including aortic dissection (a tear of the major blood vessel branching off the heart), pulmonary embolism (blood clot) and many other serious diseases as well as non-life threatening causes such as anxiety or muscle pain.

Thus emergency providers face the challenge of screening a very large number of patients with these complaints and not missing life-threatening causes of symptoms. In fact, although the focus of ACS is often patients with chest pain (in approximately 6 million cases), the reality is that a much larger number of patients who are screened for ACS are actually experiencing atypical symptoms such as fatigue or jaw pain, as in the post partum example above.

The statistics reveal that many people visiting the ED are not admitted: in 2011 there were more than 5 times the number of visits to the ED as admissions to the hospital.<sup>1</sup>

#### Case #1

While breastfeeding her second child three days after he is born, a 28-year-old mother begins feeling cold and clammy. She is perspiring, having strange indigestion and feeling exhausted. She begins experiencing pain in her upper back and jaw. After trying to talk herself out of the pain, she confides in her husband. He too is hesitant to believe that the symptoms are serious, but as they worsen, he insists that he should take her to the ED. She describes herself as "a fit and healthy, clean living mother with...no significant health

worries." At the hospital, she is evaluated for pulmonary embolism and then discharged with no diagnosis.

#### Patients with No History of Coronary Artery Disease

According to a 2010 report,<sup>2</sup> 55-60% of patients who came into the ED with chest pain had no history of coronary artery disease and no electrocardiographic abnormalities. Clearly, this presents a challenge in diagnosing heart attack. Yet, the morbidity and mortality of patients with chest pain who are sent home is 2 to 3 times that of those that are admitted to the hospital.

EDs report a range of "missed" acute coronary events. Greater than 20% of malpractice awards against ED physicians are a result of these overlooked events.

#### Case #2

A 54–year-old lifelong athlete experiences "arm and chest pain intermittently for more than a year." Two cardiologists run standard treadmill tests and diagnose acid reflux and stress-related responses.

This patient story highlights a very crucial point: all tests have limitations and there can be false negative and false positive tests. His history of lifelong exercise and exercise tolerance suggest a lowered risk for cardiovascular disease – which, in this case, is interpreted as a false negative. Unless a patient-centered discussion takes place and reveals clues to a possible alternative diagnosis, his test result would prevent appropriate treatment.

#### Why Are These People Missed?

Partly it may be due to the "risk stratification" systems being used for diagnosis. These include the thoroughness of patient's history, the results of an electrocardiogram and perhaps a set of markers that indicate there has been injury to the heart muscle. If the patient has no prior history of cardiac problems or coronary artery disease, no irregular ECG readings or no abnormal troponin levels, the probability of a heart attack is around 6%. A confirmatory study like plain exercise treadmill testing (or other noninvasive test) is the most widely used test before discharge in order to exclude unstable angina (threatened heart attack) where the results of ECG and cardiac biomarker testing could be negative.

Although data has suggested that with newer ultrasensitive troponin, "unstable angina" may become less of a concern. However, the present generation of tests and testing done dependent

on the presence of pain or not, duration of pain, etc. can still be negative with unstable angina. In reality, serial ECG, troponin and noninvasive tests (stress ECG, ECHO or perfusion scans) can be negative and patients may return to the ED with Acute Myocardial Infarction.

#### **Cultural Issues May Play a Role**

Another reason may also be cultural. One study published in the New England Journal of Medicine in 2000 found that women were 7 times more likely than men to be sent home midheart attack from hospital EDs. More recently it's been reported that women are 20% more likely than men to visit the ED and yet more likely to not be admitted to the hospital after that visit than men (an over 20% higher discharge rate).

Perhaps there is a bias in expectations (i.e., men have heart attacks, women don't). If bias does play a role, it unfortunately creates more time wasted, more potential muscle damage and more debilitation for patients.

Another cultural issue that figures into the mix is the tendency of women to delay seeking care longer than men do. The therapies to treat myocardial infarction, if delivered within two hours of the onset of symptoms, can prevent MI and also reduce the risk of heart failure. One study revealed various reasons women delay seeking care, including the tendency to minimize their symptoms; knowing they were having a heart attack but not wanting to disturb others; and a general reluctance to call 9-1-1 for themselves.<sup>4</sup>

#### A Medical Mystery

The first published description of SCAD in the literature occurred in 1931. The patient's attending physician describes the last hours of a 42-year old woman whom he had known well. She presented with pain and severe vomiting but only after her death did an autopsy reveal a "dissecting aneurysm" of the coronary artery.

Physicians and nurses often see people with non-specific symptoms. When someone arrives at the ED with vague symptoms—perhaps upper body discomfort, fatigue, chest tightness but who are otherwise physically fit, young, and with no history or risks of heart disease, it's little wonder they tell the patients, "Go home, take it easy" or "Take an antacid."

But what if the most overlooked symptom of this particular disorder is death? As one physician told Katherine K. Leon, "SCAD isn't rare. It's rare to meet a survivor."

Here's what happens in a spontaneous coronary artery dissection or SCAD event. Coronary arteries, the arteries that feed the heart, have three layered walls. With SCAD, the wall develops a

tear, causing blood to flow between the layers of the arterial wall. Pieces of the arterial wall can act as a flap, stopping the flow of blood and causing a heart attack or even sudden death. Early research estimates 80 percent of the people experiencing SCAD are women.

What's scary is that it's really not known how many people are dying from SCAD each year. This disorder is stealthy and abrupt. No symptoms; then suddenly, a piece of the coronary artery—the one that feeds the muscle of the heart—splits. Either a flap develops or a clot forms and stops the blood flow. This formerly healthy person mysteriously suffers a heart attack. In severe cases and depending on the artery involved, a "widow maker" heart attack may result. Most family members don't get autopsies to see what caused their loved one's sudden cardiac death.

When examining the statistics of people who experience spontaneous coronary artery dissection, we find that SCAD is more likely to occur in people ages 30 to 50, people who are otherwise healthy and who don't have risk factors for heart disease. SCAD is more likely to occur in women. These issues, coupled with the general information about ED usage and delaying behavior of women who need to seek care at the ED, are deeply disconcerting.

# Summary of the SCAD Alliance 2014 Scientific Advisory Board Roundtable

After a general review of SCAD, the roundtable participants discussed two recent cases, from the cardiologists' perspective as well as the ED specialist's view.

#### Incidence

The incidence of spontaneous coronary artery dissection has been difficult to determine. Some say it is between .1% and 1.1% of the attacks that occur. However this rarity is beginning to be questioned. In fact, with advanced imaging the prevalence of SCAD may be as high as 1% to 4% of acute coronary syndrome.<sup>5</sup>

Although the average age of SCAD incidence is 42 years, spontaneous coronary artery dissections have occurred in people who are teenagers and septuagenarians.

#### **Cardiologist's Perspective**

Consider the following cases, presented by cardiologist Malissa Wood, MD, co-director of the Corrigan Women's Heart Health Program at the Massachusetts General Hospital Heart Center.

#### Case # 3

A 35-year old woman in the 30<sup>th</sup> week of pregnancy came to the hospital with chest pain and premature labor. A C-Section was performed.

The physicians started blood draws to measure troponin levels. The patient's peak troponin level was 2.8. She also had non-specific ECG changes, which then became diffuse. Her diagnosis: a subtotal occlusion of the LAD.

Using Optical Coherence Tomography (OCT), which is one of the best available imaging technologies for SCAD, physicians diagnosed artery dissection. (OCT can discern the dissection and blood clot causing the blockage versus plaque build-up occluding the artery. OCT is a risky procedure for arteries prone to dissection because it requires a catheter.) For this patient, the OTC clarified that the dissection was in the intimal layer of the artery wall and involved a hematoma (localized swelling filled with blood).

#### A Disorder of Younger Women

Women (80%) are more likely to have SCAD than men and between 20 and 25% of SCAD occur around the last months of pregnancy or during the first few months after delivery.  $^{6}$ 

In women under 50, an estimated 40% of the heart attacks that occur appear to be a result of a spontaneous coronary artery dissection.

#### No "Normal" Precursors

As Dr. Wood's example illustrates, a key feature of the disorder is that SCAD occurs in people

without the "normal" precursors, for example the build-up of plaque, or atherosclerosis.

Potential risk factors for SCAD have been identified. These include:

- 1) Fibromuscular dysplasia (FMD), a condition in which the walls of arteries experience irregular growth and may result in narrowing, aneurysm, and dissection.
- 2) Out of the ordinary, extreme stress and extremely high, untreated blood pressure have been associated with SCAD events.
- 3) Lupus and Polyarteritis nodosa are diseases that cause inflammation of blood vessels and are linked to SCAD.

- 4) Some people who have had SCAD events have also had connective tissue disorders, like Marfan syndrome and Ehlers-Danlos syndrome.
- 5) Also, cocaine use has been associated with higher risk of SCAD.<sup>7</sup>

#### Importance of Correct Diagnosis for Appropriate Treatment

Dr. Wood shared case number 4 to show the importance of obtaining a correct diagnosis and the use of conservative treatment:

#### Case # 4

A 35-year old woman with obesity and high blood pressure presented with chest pain. The only family history was of a cousin who died of sudden cardiac death at age 35.

A coronary computed tomography angiogram (CTA) is a non-invasive way of finding blockages in the coronary arteries. Contrast material (dye) goes into the body via IV, and, using an advanced computed tomography (CT) technology, the doctor obtains 3-dimensional pictures of the heart. This patient received a CTA. The CTA was used to rule out pulmonary embolism and aortic dissection.

She also presented with a moderately abnormal ECG. She was admitted to the hospital and treated with heparin and a "cardiac cocktail." Eighteen hours after presenting, she showed a dissection in the Myocardial Bridge. Blood tests for troponin were taken at regular intervals. Troponin peaked and then the pain subsided.

An acute coronary syndrome (ACS), another term for coronary artery blockages, is different than one caused by atherosclerosis. The management both during the acute period and long term are unique. Specifically, there is a lower success rate of percutaneous coronary interventions (PCI) in people with SCAD. PCI is the procedure in which a hollow tube called a catheter is inserted into the blocked vessel and a stent is deployed to give structure to the artery so that the dissection can heal. Another technique is balloon angioplasty, in which the catheter contains a tiny balloon that is inflated at the blockage to compress the clot in the artery, making a larger opening for blood to flow.

PCI in SCAD has a 62% success rate while PCI in people with atherosclerotic involvement is 92%.<sup>6</sup> This makes sense, since the blockage is actually not plaque but rather a fragile part of the blood

vessel wall. Additionally, spontaneous healing of the artery walls has been found in SCAD patients. This indicates that using conservative interventions in stable patients is recommended.

In those who are treated conservatively, close monitoring in the hospital is required during the first four to five days. However this monitoring should be non-invasive because the blood vessels are fragile (often described as "friable" or tissue-thin) and can be damaged easily.

#### **Challenges for the Emergency Physician**

In the ED and throughout cardiac care in general, Dr. Wood identified specific challenges:

- How do we determine which patients to screen for ACS with ECG and troponin testing since many patients, especially those with SCAD, are not screened because of young age, low risk factor reporting, etc.
- 2) How do we interpret an ECG? What is "normal" for the patient? How do we identify abnormal for the patient if we do not have a pre-SCAD event ECG?
- 3) How can we obtain timely and accurate troponin blood draws, considering issues such as the duration of pain, interval in testing, re-sampling, knowledge of the specific assay, and diagnostic performance – particularly in the case of SCAD, when the patient is not perceived to be a cardiac patient.

#### **Emergency Department Perspective**

In response, Dr. Leana Wen described the overall challenges facing physicians in the ED.

Formerly attending physician and director of patient-centered care for the George Washington University Department of Emergency Medicine, Dr. Wen said the challenge for the ED physician is to figure out what is happening with all kinds of acute symptoms. "We stratify based on age and appearance," she reported. Factors involved in decision making include "making sure we are not over-testing."

Dr. Wen believes that some of the "difficulty arises when the patient has just started having chest pain. Until it is decided that there is a coronary issue, emergency physicians have trouble convincing an admitting physician (e.g., primary care physician, internist, family practice, cardiologist) to admit the patient." This is particularly the case for a hospital that does not have an Observation Unit or protocol that allows for the evaluation of patients who have symptoms suggestive of ACS but non-diagnostic ECGs. Considering Case #4, much was done to rule out a possible aortic dissection. Dr. Wen's question to Dr. Wood: How would you manage the patient differently at presentation in the ED?

#### **Cardiologist's Response**

Dr. Wood response was that historically you look at the risk factors first, then the symptoms and then ECG. It is very challenging with ECG changes in women. She believes that checking troponin levels is not a waste if the patient's story and ECG are suggestive. Her recommendations:

- 1) Assessment
- 2) Listen and interpret the patient's story
- 3) Have a low threshold to obtain an ECG and compare to prior ECG (if available). Consider having a system of ECG consultation or review. (Research shows ECG misclassification can be corrected – possibly prevented – using "real time" review to reduce errors.)
- 4) Check troponins
- 5) Use echocardiogram (ECHO) to provide information about tamponade, Left Ventricular Ejection Fraction (LVEF), dissection, or PE, and possibly screen for wall motion abnormalities (WMA).

An underlying issue for all U.S. emergency departments: the number one cause of malpractice is patients who are sent home despite presenting at the ED with coronary symptoms in the midst of a heart attack.

## **Recommendations for Emergency Department Staff**

To change and improve SCAD diagnosis the SCAD Alliance Scientific Advisory Board cardiologists provided these recommendations:

- 1) Conduct a thorough history, including a discussion of activity in week prior and month prior, connective tissue disorders, and any genetic information.
- 2) Recognize that some patients including those who are pregnant, peri-partum, have FMD, autoimmune disease or a connective tissue disorder, or use cocaine are at increased risk for SCAD even if young and without other classic risk factors for coronary artery disease.

- 3) Have a low threshold for obtaining an ECG to screen for ACS including in those who may complain of chest pain or other symptoms such as jaw pain, abdominal pain, weakness, fatigue, etc.
- 4) Increase the use of high sensitivity troponin testing.
- 5) Conduct bedside Echocardiogram. Point of Care (POC) ECHO can be a useful part of the ED evaluation as discussed previously, and if one identifies WMA, can assist in the evaluation.
- 6) Depending on results of ECHO, if WMA are seen (e.g., suggesting a problem with the Left Anterior Descending (LAD) artery), the results should be validated by Electrophysiology (EP).
- 7) Consult cardiology regarding imaging, especially for those patients at higher risk for SCAD (e.g., young patient, peri-partum with abnormal ECG, but not STEMI with positive troponin). While CT is a better test for SCAD than stress ECG, there are issues and concerns about doing CT, especially in young women who are considered low risk.
- 8) Based on history and status, a CT may offer better diagnostic guidance than a treadmill stress test (exercise tolerance test).
- 9) If suspicious of SCAD, do *not* order a treadmill test
- 10) Bottom line: recognize higher risk patients require cardiology consult for diagnosis and treatment. The pregnant woman illustrated in Case 3 may have had WMA, but she also was pregnant, had a "nonspecific ECG," and a positive troponin.

Dr. David Adlam, Senior Lecturer, Department of Cardiovascular Sciences, University of Leicester, reinforced these concepts:

- Imaging is key. If blood labs indicate a positive troponin and ECG change, refer patients to a center that has the capability of doing more advanced imaging and the ability to pursue percutaneous coronary intervention (PCI) or coronary artery bypass grafting (CABG), as indicated.
- 2) If the patient's condition is severe (i.e., experiencing refractory pain, is hemodynamically unstable or in shock, use a balloon pump and transfer him/her to a specialty facility.

#### Follow-up

Because SCAD has an estimated 10-year recurrence rate of up to 20% in women, the Scientific Advisory Roundtable participants agree it is important to continue to follow-up. All SCAD patients should do supervised cardiac rehabilitation and avoid weight lifting, body-building and competitive racing. Resistance training is recommended.

Patients are also advised to see a vascular specialist and geneticist to rule out associated conditions such as connective tissue disorders and fibromuscular dysplasia (FMD).

#### Conclusion

Because of the challenge of identifying patients with SCAD, future research is necessary to identify patients who are at risk for this disease and potentially develop preventive strategies.

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# **About SCAD Alliance**

SCAD Alliance is a unique 501c3 organization with an integrated approach to the challenges facing spontaneous coronary artery dissection (SCAD) patients and those who care about them. Our four-leaf "lucky" heart logo represents the torn artery we must overcome with the support of family, health partners, and researchers.

Our Mission – The SCAD Alliance mission is to be the leader in advancing the science of Spontaneous Coronary Artery Dissection (SCAD) through improved knowledge and cooperation among health care professionals, patients and their families. We pursue this mission by educating key audiences and fostering unique interdisciplinary research collaborations.

Our Vision – To empower each SCAD survivor with an accurate diagnosis, superior outcome, and answers.

With a focus on collaboration, SCAD Alliance has incorporated the many disciplines that play a role in researching spontaneous coronary artery dissection and caring for patients. We have crafted a Scientific Advisory Board whose members are expert in the fields of cardiology, vascular disease, clinical psychology, connective tissue disorders, obstetrics/gynecology, endocrinology, and genetics.

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