

# Spontaneous Coronary Artery Dissection

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PATIENT GUIDE 2022

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# Acknowledgements

The authors would like to pay special tribute to our patients and their families, who have taught us so much. We would like to thank the many reviewers of this guide who generously gave their time to review this guide and provided thoughtful comments and suggestions. Reviewers included Lesley Krempulec, Geraldine James, Colleen Nelson, anonymous patient reviewer, Kristen Hamilton, George Petropoulos, Katherine Farrand and Rebecca Stockford-Lehman. Thank you for helping to shape this final version.

# Introduction

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This guide is for people who have been affected with spontaneous coronary artery dissection (SCAD), their families, and anyone who wants a basic understanding of SCAD, its diagnosis, and management.

This guide does not replace treatment from your doctor. It can be used as an information resource and help with any questions you may have.

SCAD can be a cause of heart attacks. This guide has information on:

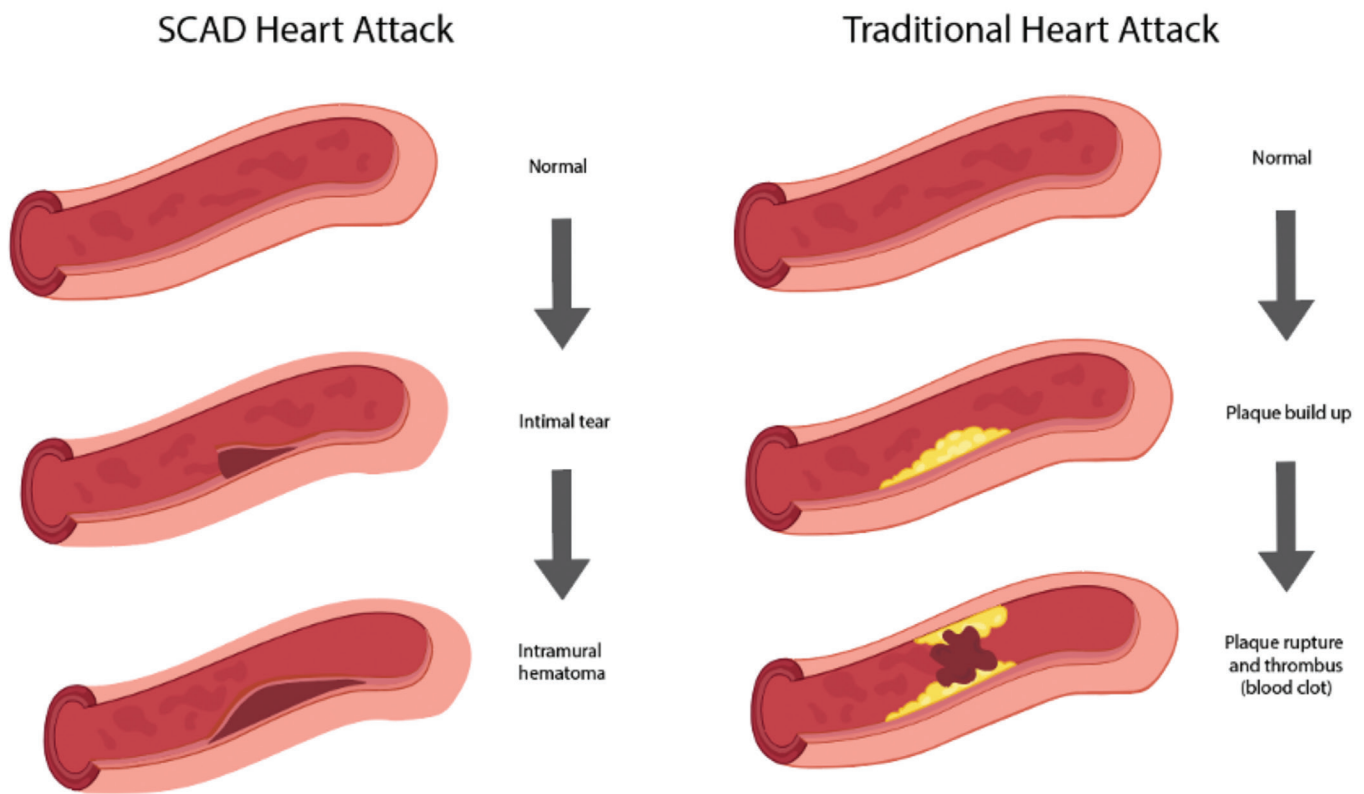
- SCAD risk factors
- Symptoms
- Diagnosis
- Treatment

We hope this guide helps you understand SCAD.

# What is spontaneous coronary artery dissection (SCAD)?

Spontaneous coronary artery dissection (SCAD), is when there is a sudden tear in the wall of one or more blood vessels in the heart. This results in a buildup of blood between the layers of the artery wall. As blood collects between the layers of the wall, the vessel narrows. This impairs blood flow to the heart which causes a heart attack.

**Figure 1: SCAD heart attack versus traditional heart attack.**



# How common is SCAD?

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SCAD makes up 4% of heart attacks across the world. In women less than or 50 years of age, SCAD is the cause of 35% of heart attacks. SCAD is the number one cause of heart attacks in pregnancy.

# Who gets SCAD?

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SCAD is most common in middle-aged women between the ages of 45 to 55. Less than 10-15% of SCAD cases occur in males.



# What are potential causes and risk factors for SCAD?

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It is unclear what causes SCAD. In more than 20% of cases there is no known cause. We do know that:



- **Sex:** SCAD affects many more females than males.
- **Fibromuscular dysplasia (FMD):** This is a condition with abnormal twisted blood vessels throughout the body with areas of vessel narrowing and dilation. It most commonly affects blood vessels in the heart, the head and neck, and the kidneys. This condition has been found in more than 60% of SCAD patients.
- **Pregnancy:** SCAD can occur during and right after pregnancy. It has been shown to be more common in females who have had more than one pregnancy or births.
- **Emotional and physical stress:** Patients with SCAD have higher rates of physical and emotional stressors (for example: loss of a loved one, a job or a relationship, work stress.).
- **Connective tissue disorders:** This is a group of disorders involving tissue that connects structures of the body together. About 3 to 4% of SCAD is linked to underlying connective tissue disorders. These include Marfan's syndrome, Ehlers-Danlos and Loey-Dietz syndrome.
- **Hormonal therapy:** Although up to 10% of patients with SCAD were taking hormonal therapy, so far there is no conclusive evidence that hormonal therapy causes SCAD or increases the risk of SCAD recurrence.
- **Genetics:** SCAD is rarely associated with a family history. Very few genes have been identified that have an association with SCAD. Some of these gene mutations are familial including F11R and TLN1. Other gene mutations are spontaneous including TSR1, PHACTR1 and EDN1. Further research is needed to help understand whether there is a genetic basis for SCAD.



# How are SCAD heart attacks different from regular heart attacks?

Compared to regular coronary artery disease, SCAD patients have lower rates of traditional cardiovascular risk factors. SCAD patients have lower rates of high blood pressure, high cholesterol, diabetes, and smoking. Patients with SCAD report higher rates of emotional and physical stressors.

**Table 2: Key differences in demographics and associated conditions in SCAD compared with traditional coronary artery disease (CAD) (adapted from Adlam, García-Guimaraes, et al., 2019).**

	Traditional Heart Attack	SCAD Heart Attack
Sex		
Age < 65 yrs	+	+
High blood pressure	+	+
High cholesterol	+	+
Diabetes	+	-
Smoking	+	+
Pregnancy	+	+
Emotional stress	+	+
Strenuous exercise	+	+
Migraines	-	+
FMD	-	+

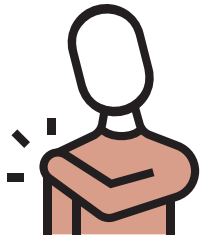
**Legend:**  Male  Female

# Signs and symptoms

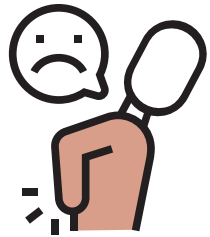
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The majority of patients with SCAD have new chest pain, much like a regular heart attack.

Other symptoms may include:



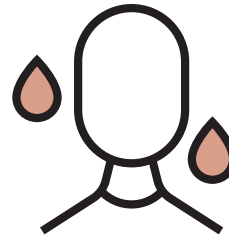
Pain in the arms, shoulders or jaw



Back pain



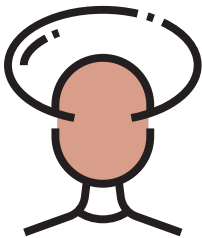
Nausea and vomiting



Sweating



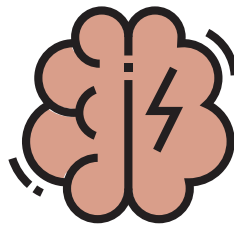
Shortness of breath



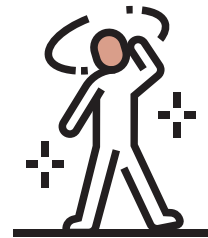
Dizziness or light-headedness



Extreme fatigue



Headache



Fainting



Palpitations/heart racing

Rarely, patients can have more severe symptoms including dangerous heart rhythms known as ventricular tachycardia and ventricular fibrillation (2-8%), cardiogenic shock (a life-threatening condition where your heart can't pump enough blood to your organs) or sudden death.

# Diagnosis

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The first tests used to diagnose SCAD are the same as those used to diagnose traditional types of heart attacks. Patients usually have abnormal electrocardiograms (ECGs) and elevated heart blood markers, known as troponins. The best test used to diagnose SCAD is a coronary angiogram.

A coronary angiogram is an imaging test during which contrast (x-ray dye) is injected into the arteries of the heart (coronary arteries) and x-ray images are used to look for artery narrowing. Classic findings on a coronary angiogram can help differentiate SCAD heart attacks from traditional heart attacks.

In some cases, more imaging from within the coronary arteries is needed to help make a diagnosis of a SCAD heart attack versus a traditional heart attack. These include optical coherence tomography and intravascular ultrasound. These are performed at the same time as the coronary angiogram, if needed.

Other non-invasive imaging studies such as computed tomography angiography or magnetic resonance angiography are less useful to make the initial diagnosis of SCAD, but help to screen other large blood vessels for abnormalities like FMD or aneurysms.

# Management

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Unlike traditional heart attacks which do not get better on their own, the majority of SCAD blood vessels heal with no treatment. Studies which looked at SCAD blood vessels with a repeat coronary angiogram 30 days after the first SCAD heart attack, showed that 95% of SCAD blood vessels had healed without treatment. This is why we don't treat most SCAD related heart attacks with invasive therapies.

Although a conservative strategy is used for the majority of patients, 2-8% of patients need invasive treatment because the tear and blood build up in the affected blood vessel does not resolve on its own or gets worse.

A few patients who do not improve without treatment or have severe SCAD heart attacks, need to have the blockage of the coronary artery fixed. This process is known as revascularization. Options for revascularization include percutaneous coronary intervention (PCI) and coronary artery bypass grafting (CABG).

## **Percutaneous Coronary Intervention (PCI)**

Percutaneous coronary intervention (PCI) may be considered in patients who have a SCAD heart attack with unstable features (ongoing chest pain, unstable condition, very large heart attack) or multiple coronary arteries affected by SCAD.

In these situations, the goal of PCI is to restore blood flow to the blocked SCAD blood vessel to minimize the damage to the heart. PCI uses a balloon to open the blockage in the coronary artery and possibly place a stent afterwards to keep the artery open. If needed, PCI can be performed immediately after the coronary angiogram is completed.

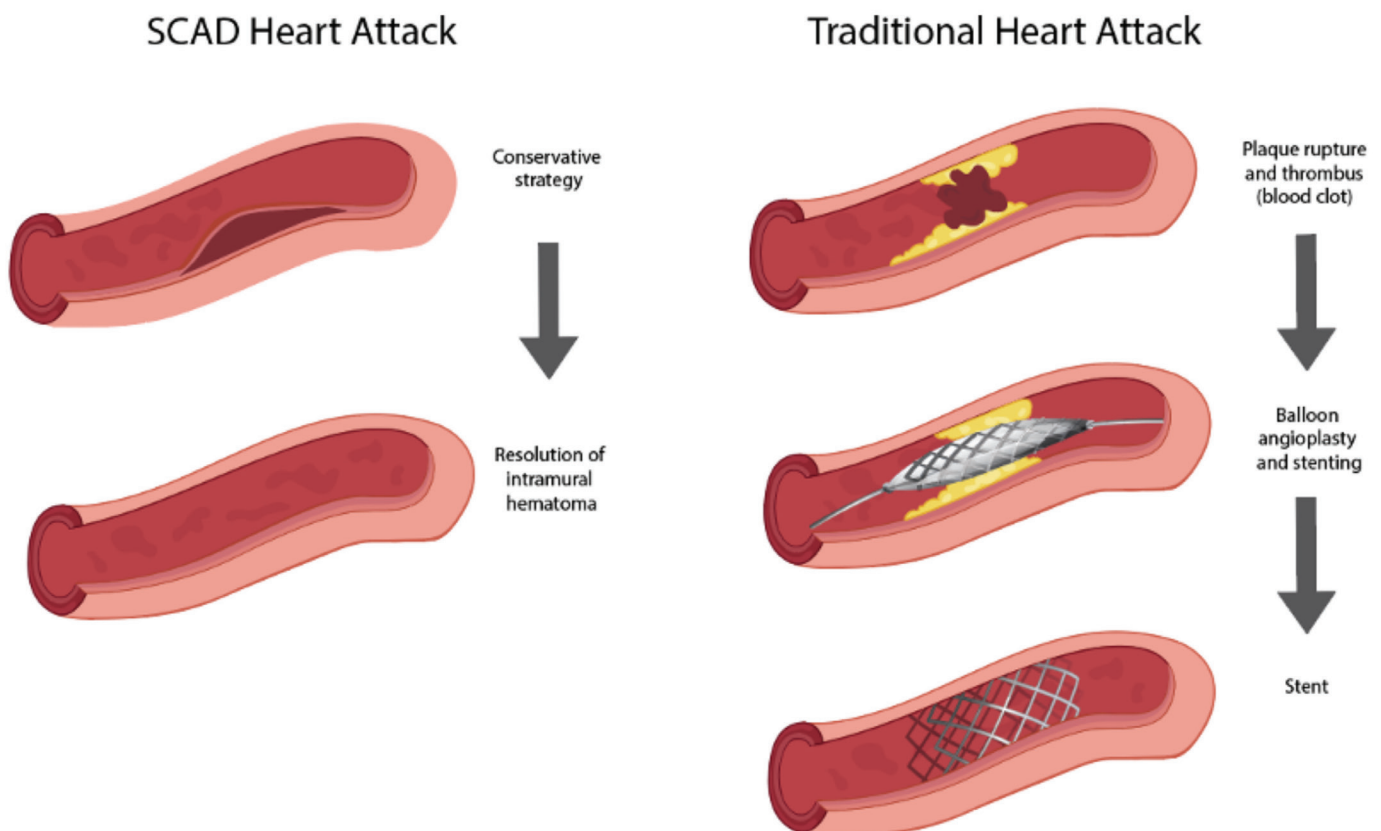
PCI is usually a lifesaving intervention in SCAD heart attacks. Some complications of PCI include recurrent coronary artery blockages in the stent, incorrect stent size for the blood vessel and residual blockages despite stent placement. Patients who require PCI for a SCAD heart attack have a higher risk of requiring a repeat procedure for recurrent coronary artery blockages compared to patients who can be managed with a conservative strategy.

## Coronary Artery Bypass Grafting (CABG)

Coronary artery bypass grafting (CABG) is heart surgery to restore blood flow to a blocked coronary artery. It takes a blood vessel from another part of the body (usually chest wall or leg) and attaches it to the coronary artery above and below the blockage to bypass it. This new blood vessel is known as a graft. The number of grafts you will need is dependent on the number of blockages that need to be bypassed. This procedure is performed under general anesthesia, which means you will be unconscious during the operation.

A very small minority of patients with SCAD heart attacks undergo heart surgery. Patients would be considered for CABG if they have a life-threatening SCAD heart attack and PCI is not possible. Most patients who undergo CABG do well in the long term, with the highest risk occurring in the immediate period surrounding the surgery. In the long term, it is common for these grafts to become blocked in more than 60% of cases as the patient's SCAD blood vessel heals and the grafts are no longer required to redirect blood flow. Although graft closure is a long-term risk, the initial CABG operation is usually lifesaving.

**Figure 2: Management of SCAD heart attacks compared with traditional heart attacks.**



# What medications should I be on?

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There are no randomized controlled trials to guide medical therapy in SCAD. A randomized controlled trial involves researchers deciding randomly as to which participants in the trial receive a new treatment and which receive a placebo, or 'fake' treatment. This is how we study the benefits and side effects of medications to provide evidence for their use.

Some commonly prescribed medications include the following:

- Antiplatelet agents: These are mild blood thinners that stop cells in the blood (platelets) from sticking together and forming a clot. There is a lack of evidence to guide the duration of antiplatelet therapy and the number of antiplatelet medications to be used.
  - » Single antiplatelet therapy refers to treatment with one antiplatelet medication, usually low dose aspirin.
  - » Dual antiplatelet therapy refers to low dose aspirin plus a second antiplatelet agent which may be clopidogrel (Plavix), ticagrelor (Brilinta) or prasugrel (Effient).
  - » If a stent was placed, dual antiplatelet therapy is generally recommended for 1 year.
  - » If there is no stent used the use of single versus dual antiplatelet therapy is controversial. Currently, many centers prefer the use of a single antiplatelet medication (aspirin) while some recommend that patients are treated with dual antiplatelet therapy
- Beta-blockers: These are medications that slow your heart rate and may reduced the force that the blood vessels experience by the blood being pumped through. Only one study has shown benefit with beta-blocker use in SCAD patients. This study showed a lower rate of recurrent SCAD heart attacks in patients who took beta-blockers

- Anti-hypertensives: These are medications that aim to lower your blood pressure. It includes a wide variety of medications such as angiotensin-converting enzyme inhibitors (ACE inhibitors), angiotensin II receptor blockers (ARBs), calcium channel blockers (CCBs) and nitroglycerin among other classes. One study showed that high blood pressure was associated with a higher risk of recurrent SCAD heart attacks and therefore recommended the use of anti-hypertensive medications to treat high blood pressure. There is no evidence that these medications have direct benefit in SCAD, however they may be used as an antihypertensive to lower blood pressure.
  - » Angiotensin-Converting Enzyme (ACE) Inhibitors: These medications relax (dilate) the blood vessels and decrease the blood volume to lower blood pressure.
  - » Angiotensin II Receptor Blockers (ARBs): These medications are similar to ACE inhibitors. They relax (dilate) the blood vessels and lower blood pressure.
  - » Calcium-channel blockers (CCBs): These medications lower blood pressure by preventing calcium from entering the cells of your heart and arteries.
  - » Nitroglycerin: This medication lowers blood pressure by relaxing (dilating) blood vessels, and may also help with symptoms of chest pain after SCAD, which is not so uncommon.

There are some situations where medications should be used. These include if the patient underwent PCI or if the patient has a weak heart, also known as left ventricular dysfunction or heart failure. In these situations, medical therapy would often include the following:

- Percutaneous coronary intervention (PCI): Dual antiplatelet therapy for at least one year. This includes aspirin plus a second antiplatelet either clopidogrel (Plavix), ticagrelor (Brilinta) or prasugrel (Effient).
- Left ventricular dysfunction: Medical therapy is based on the heart failure guidelines. It often includes sacubitril/valsartan (Entresto), an ACE inhibitor or ARB as well as a beta-blocker, a mineralocorticoid receptor antagonist and an SGLT-2 inhibitor.

Medications that should be avoided in SCAD, if possible, include triptans such as sumatriptan used for migraines, stimulants such as ADHD medications or cocaine, and hormonal therapy. The use of these medications after SCAD should be discussed with your doctors.

**Table 3: Key differences in the treatment of SCAD heart attacks compared with traditional heart attacks.**

	<b>SCAD Heart Attacks</b>	<b>Traditional Heart Attacks</b>
<b>Revascularization</b>		
PCI (balloon angioplasty ± stenting)	Conservative strategy. May consider PCI if large or unstable SCAD heart attack or multiple coronary arteries with SCAD	Recommended
CABG	When PCI is not possible	When PCI is not possible
<b>Medical Therapy</b>		
Antiplatelets	Lack of evidence to guide therapy. Dual antiplatelet therapy for 1 year if a stent is placed. In the absence of stent implantation, use of single versus dual antiplatelet therapy is controversial	Dual antiplatelet therapy for 1 year
Beta-Blockers	Recommended	Recommended
ACE Inhibitors	No clear benefit	Recommended
ARBs	No clear benefit	Recommended
Statins	No clear benefit	Recommended
<b>Lifestyle</b>		
Diet	No restrictions	Healthy heart diet
Physical activity	Moderate intensity exercise	Exercise intensity as tolerated
Stress Management	Recommended	Recommended
Smoking Cessation	Recommended	Recommended
<b>Additional Screening</b>		
FMD Screening	CTA from head to pelvis recommended	Not required



# What screening should I have after a diagnosis of SCAD?

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Abnormalities of the arteries throughout the body are common among SCAD patients, and a condition known as fibromuscular dysplasia (FMD) has been seen in more than 60% of patients.

Fibromuscular dysplasia is a condition that weakens the artery walls making them prone to tears or bulging. In FMD, patients can have abnormal blood vessels in many parts of the body, including the heart, head and neck, and kidneys. It is recommended that all patients with SCAD undergo blood vessel imaging from their head to pelvis to screen for findings consistent with FMD at the time of diagnosis. This is done using non-invasive imaging such as computed tomography angiography (CTA). If a patient has FMD they should have a follow up at least once a year. Follow-up may include clinical assessment, blood work and imaging. The timing and nature of imaging however, should be individualized according to the severity of the disease and any symptoms.

There is no commercially available genetic screening test for SCAD, however this is an active area of ongoing research.

Screening for connective tissue diseases including Marfan syndrome, Ehlers-Danlos syndrome and Loey-Dietz syndrome, may be considered if you have other clinical signs or symptoms consistent with these conditions.

Overall, we do not screen everyone for these conditions as they are rare. There is no recommended screening for autoimmune or inflammatory disorders as there is no clear association. Genetic counseling may be considered if there is a family history of inherited arteriopathy such as Marfan syndrome or a history of aneurysms.

# What is my risk for another SCAD related heart attack?

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Recurrent SCAD heart attacks have been reported to occur in approximately 10 to 30% of patients within 3 years of their initial SCAD heart attack. As we continue to follow more SCAD patients for longer periods of time, we will learn more about the chances of another SCAD heart attack. A higher risk of SCAD recurrence is associated with severe coronary artery twisting (tortuosity), FMD with migraines and high blood pressure.

To lower the risk of another SCAD event, you can maintain a healthy blood pressure, take medications to lower the shear stress on your blood vessels (beta-blockers) and maintain a healthy lifestyle that includes moderate intensity exercise.

Up to 50% of patients with SCAD have recurrent chest pains after their event, even though the artery is healed in most cases. In most patients these symptoms will get better over time, however sometimes hospital admission is necessary to assess for SCAD recurrence. If you develop recurrent symptoms, speak to your doctor, who can then decide what testing and treatment you may need for chest pain after SCAD.

# Pregnancy-Associated SCAD

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Pregnancy-associated SCAD heart attacks account for less than 5-17% of SCAD cases and 1.81 per 100,000 pregnancies. SCAD can occur at anytime during or after pregnancy, however more than 70% of cases occur post-partum with the majority within the first week. Patients who develop SCAD during pregnancy tend to be much sicker with larger sized heart attacks or more than one coronary artery affected with SCAD.

It is recommended that all patients who have had SCAD should avoid pregnancy due to the high risk of recurrent SCAD heart attacks. Approximately 20% of patients have recurrent SCAD heart attacks with pregnancy. Patients with SCAD should consider permanent solutions to avoid future pregnancies or consider taking contraception. It is recommended that SCAD patients avoid estrogen containing contraception options and instead used long acting progesterone only methods including the subdermal levonorgestrel implant and the levonorgestrel-releasing IUD.

# Living with SCAD

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- **Physical Activity:** SCAD symptoms have been associated with physical activity in 32% of patients. There are no strict recommendations to guide physical activity in SCAD patients. Physical activity recommendations should be individualized and take into consideration the patient's baseline physical activity and fitness level.
  - » Regular moderate-intensity exercise likely outweighs the theoretical risk of recurrent SCAD. Strenuous exercise and straining associated with heavy lifting can result in a sudden increase in your blood pressure and stress on your blood vessels causing recurrent SCAD. It is recommended that patients avoid lifting or carrying heavy objects that require prolonged straining, extreme endurance training and elite competitive sports.
- **Mental Health:** Many SCAD patients find it hard to adjust to a new diagnosis of 'heart attack'. SCAD often happens to younger people, in their early to mid-career, and for some women during their childbearing years, or while raising a young family. High rates of anxiety (41%), depression (32%), and posttraumatic stress disorder (28%) have been reported among SCAD patients. The prevalence of anxiety diminishes over time, but posttraumatic stress disorder (PTSD) and depression remained time independent. We recommend seeking support from a mental health professional, to help manage these symptoms, as needed.
- **Smoking:** Quitting smoking is recommended to protect your heart and cardiovascular system.
- **Diet:** There is no known association between diet and SCAD. It is recommended that patients avoid excess caffeine and caffeinated energy drinks.
- **Driving:** The Ontario Ministry of Transportation does not allow a person to drive after a heart attack for a minimum of one month for private driving and three months for commercial driving. Fitness to drive depends on many factors including the amount of heart damage and other medical conditions. You should discuss with your cardiologist when it is safe for you to drive again.
- **Sexual Activity:** Most people can return to sexual activity four to six weeks after a heart attack. Speak with your doctor about when it would be safe for you to resume sexual activity.

- **Cardiac Rehabilitation:** Cardiac rehabilitation is a program of exercise, education and counselling designed to help you recover after a heart attack in a safe environment. This personalized program will help you regain your strength and reduce your risk of having heart problems in the future. Your rehabilitation team may include a physician, nurse, occupational therapist, physiotherapist, physical educator, dietician, psychologist or psychiatrist, and social worker. It has been reported that women are only half as likely as men to attend and follow a rehabilitation program due to a variety of reasons. Take an active role in your recovery and speak to your doctor about cardiac rehabilitation options.

## Additional Resources

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- Your Cardiologist
- The SCAD Alliance: <https://www.scadalliance.org/>
- The Heart and Stroke Foundation: <https://www.heartandstroke.ca/heart/conditions/spontaneous-coronary-artery-dissection>
- Fibromuscular Dysplasia Society of America: <https://www.fmdsa.org/>
- FMD: <https://my.clevelandclinic.org/health/diseases/17001-fibromuscular-dysplasia-fmd>

## When to Seek Medical Attention

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Signs of a heart attack may include:

- Chest discomfort
- Upper body discomfort
- Sweating
- Nausea
- Shortness of breath
- Light-headedness



**If you have any of these signs, call 9-1-1 immediately.**







PR 60854 (2022/02/23)

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