# Spontaneous Coronary Artery Dissection

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## Introduction

- **Pretty H**(1931)-1st **reported case**
- CAG : 0.2% incidence
- Mean age :40 yrs
- Men:30%,female:70%(peripartum)
- LAD(y) in ladies : most commonly

## Definition

- SCAD is defined as a non-traumatic and non-iatrogenic separation of the coronary arterial walls, creating a false lumen .
- This separation can occur between the intima and media or between the media and adventitia, with intramural hematoma (IMH) formation within the arterial wall that compresses the arterial lumen, decreasing antegrade blood flow and subsequent myocardial ischemia or infarction.

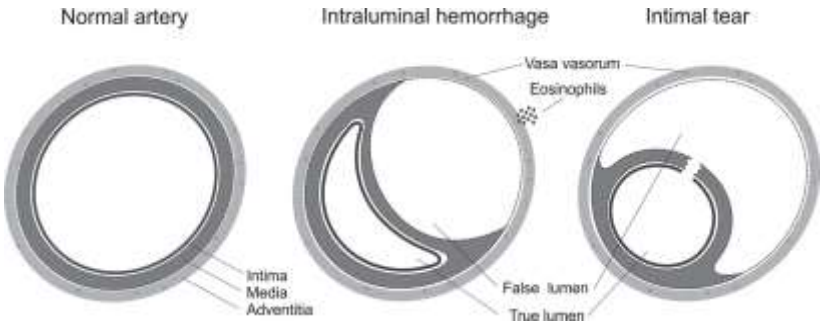
## Prevalence

- It is difficult to ascertain the true prevalence of SCAD as it is mosly under-diagnosed and have varying presentations from mild chest pains to sudden cardiac death.
- Older literature was mostly post-mortem reports but with recent awareness of the disease and improved imaging, SCAD cases are increasingly reported antemortem.
- In patients presenting with ACS, SCAD is noted to occur in 3-4%, diagnosed with optical coherence tomography (OCT)., *(old data)*

- Among stable patients presenting for routine coronary angiography, SCAD was diagnosed in 0.3% ,suspected these were delayed diagnoses following a previously undiagnosed ACS event.
- Among women presenting with ACS, the prevalence was reported to be higher at 8.7% among those <50 years old .
- In the subgroup of women presenting with ST elevation, prevalence of SCAD was even higher at 10.8% .

## Pathogenesis and pathophysiology

- There are two proposed mechanisms for the formation of IMH with SCAD.
- The first involves an intimal tear resulting in blood from the endoluminal space entering the intimal space, creating a false lumen filled with blood.
- The second mechanism of IMH formation is thought to be due to rupture of the vasa vasorum, which are small arterioles within the walls of arteries supplying blood to the walls. When such rupture occurs, blood can pool within the intramural space, creating a false lumen filled with hematoma.




Schematic diagram of the mechanisms of spontaneous coronary artery dissection.

- Eosinophilic infiltrates in arterial adventitia have also been observed in autopsies of peripartum women. It is believed that these eosinophilic granules cause breakdown of the medial-adventitial layer via lytic substances, predisposing the artery to dissection.
- However, it is unclear if the eosinophilic granules cause SCAD or is resultant of SCAD.

## Clinical implication

- Clinically, the mechanism of the tear is probably unimportant, and coronary angiography is suboptimal to visualize intimal tears.
- Intracoronary imaging with intravascular ultrasound (IVUS), and in particular OCT, has increased the detection of intimal rupture .
- The usual pathogenesis of ACS involves atherosclerotic plaque rupture that is different from non-atherosclerotic forms of SCAD.
- Dissections due to atherosclerosis tend to be less extensive, as medial atrophy and scarring from atherosclerosis limits propagation of the dissection.

## Causes

- Non-atherosclerotic SCAD is typically a culmination of disease pathways that predispose arterial beds to injury.
- These include fibromuscular dysplasia (FMD), multiple pregnancy, systemic inflammation (systemic lupus erythematosus, Crohn's disease, polyarteritis nodosa and sarcoidosis), connective tissue disorder (Marfan's syndrome,Ehler Danlos, cystic medial necrosis), hormonal therapy, and coronary artery spasm .

| Table 1 Etiology of non-atherosclerotic SCAD |
|---|
| Predisposing arteriopathy |
| Fibromuscular dysplasia |
| Pregnancy: history of multiple pregnancy, peri-partum |
| Connective tissue disorder: Marfan's syndrome, Ehler Danlos syndrome, cystic medial necrosis, fibromuscular dysplasia |
| Systemic inflammation: systemic lupus erythematosus, Crohn's disease, polyarteritis nodosa, sarcoidosis |
| Hormonal therapy |
| Coronary artery spasm |
| Idiopathic |
| Precipitating stress events |
| Intense exercise (aerobic or isometric) |
| Intense emotional stress |
| Labor & delivery |
| Intense Valsalva-type activities (e.g., severe repetitive coughing, retching/vomiting, bowel movement) |
| Cocaine, amphetamines, met-amphetamines, beta-HCG |
| SCAD, spontaneous coronary artery dissection. |

# Women and SCAD

- SCAD has been observed in women who are peripartum or with multiple prior pregnancies.
- Hormonal changes during pregnancy are thought to alter normal elastic fibres, impair collagen synthesis and mucopolysaccharide content, causing weakened media.
- Progesterone is thought to be the culprit hormone, and estrogen, on the other hand, creates a hypercoagulable state.
- The weakened arterial walls (dissection ) &&& prothrombotic state ___increases the risk of false lumen creation and thrombosis.

- The hemodynamic changes during late pregnancy can also precipitate SCAD. The aorta undergoes microstructural changes secondary to increased shear stress from augmented cardiac output and increased circulatory volume during pregnancy. This could also extend to the coronary arteries .
- During labor, the increased intraabdominal pressures could also predispose more arterial stress leading to SCAD.
- Patients with peri-partum SCAD may also have underlying predisposing arteriopathies such as FMD

## Predisopsing factors

- In patients with underlying predisposing arteriopathies, there can often be precipitating stressors such as intense exercise or emotional stress, which may trigger the SCAD event.
- Intense exercises, particularly an isometric type, can increase cardio-circulatory stresses and shear forces against the coronary arterial wall. Other aerobic type exercises, and activities that cause intense Valsalva-type straining have also been shown to precipitate SCAD.
- Related to arterial integrity, a recent study showed that coronary artery tortuosity is present at a higher proportion in patients with SCAD as compared to patient with normal coronary arteries.
- Also , Repeat dissections were common within tortuous segments.

## SCAD and FMD

- A strong association between SCAD and FMD , *a condition that also predominantly* affects women.
- FMD is a non-inflammatory, nonatherosclerotic disorder of the arterial vasculature that leads to arterial stenosis, occlusion, aneurysm or dissection.
- It can involve any small to medium-sized arterial beds, especially the renal and internal carotid arteries. The etiology of FMD is unknown, however, hormonal influences had been proposed and a small proportion may be genetic.
- Since 90% of cases affect women , sex hormones were thought to influence development of FMD, with some similarities to SCAD.

- Fibroplasia, hyperplasia and aneurysmal changes can occur in all three segments of the arterial wall, intima, media and adventitia with FMD, and can result in weakened arterial walls.
- These processes predispose segments of the artery to dissections, which may explain the strong association.

## Presentation and patient characteristics

- Patients presenting with SCAD have a spectrum of clinical presentation, and fortunately, the majority appear to present with ACS with good in-hospital prognosis.
- Ventricular arrhythmia occurred in 8-14% of patients .
- mostly all cases of SCAD presented with troponin-positive ACS, with most cases presenting with ST-elevation MI, and NSTIMI.

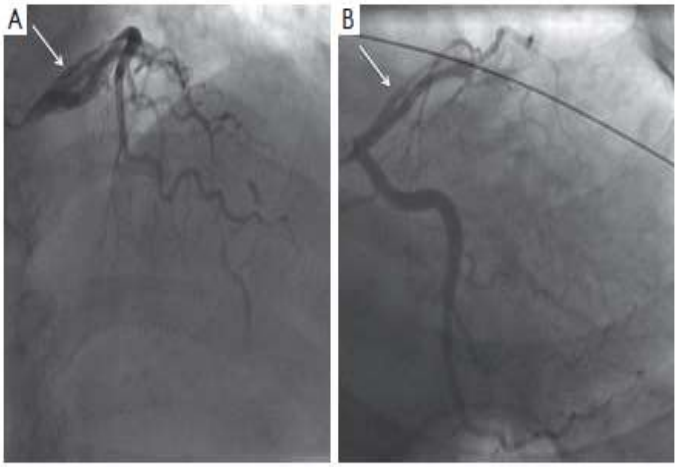
## Clinical features that raise suspicion of SCAD

*Myocardial infarction in young women (especially age ≤50)*
*Absence of traditional cardiovascular risk factors*
*Little or no evidence of typical atherosclerotic lesions in coronary arteries*
*Peripartum state*
*History of fibromuscular dysplasia*
*History of relevant connective tissue disorder: Marfan's syndrome, Ehler Danlos syndrome, cystic medial necrosis, fibromuscular dysplasia*
*History of relevant systemic inflammation: systemic lupus erythematosus, Crohn's disease, ulcerative colitis, polyarteritis nodosa, sarcoidosis*

# Investigations

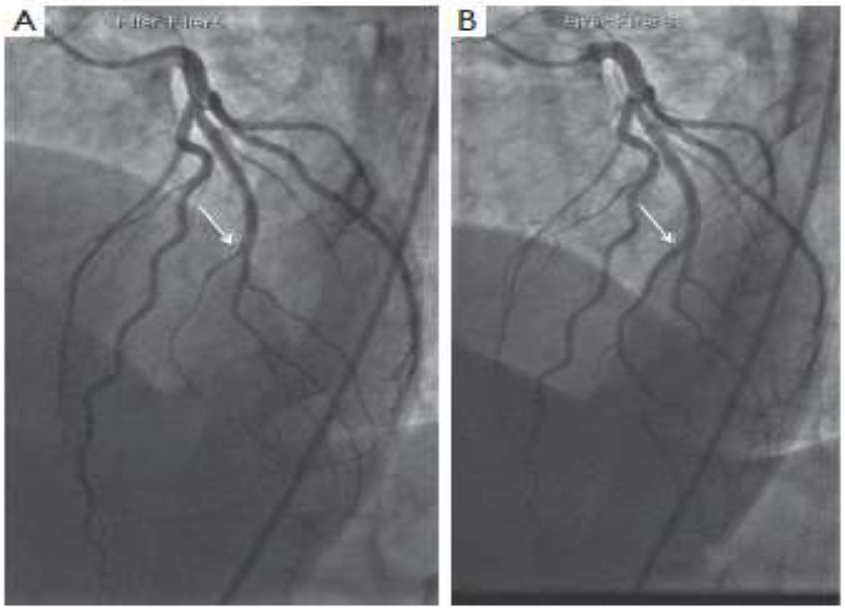
# Imaging

- ***Angiography***
- The early case reports and series on SCAD had relied on post-mortem diagnosis. Current widespread availability of coronary angiography enabled earlier diagnosis of SCAD.
- There are three distinct angiographic appearances and patterns of SCAD to aid diagnosis :
- (I) Type 1 (evident arterial wall stain): this is the pathognomonic angiographic appearance of SCAD with contrast dye staining of the arterial wall with multiple radiolucent lumens .

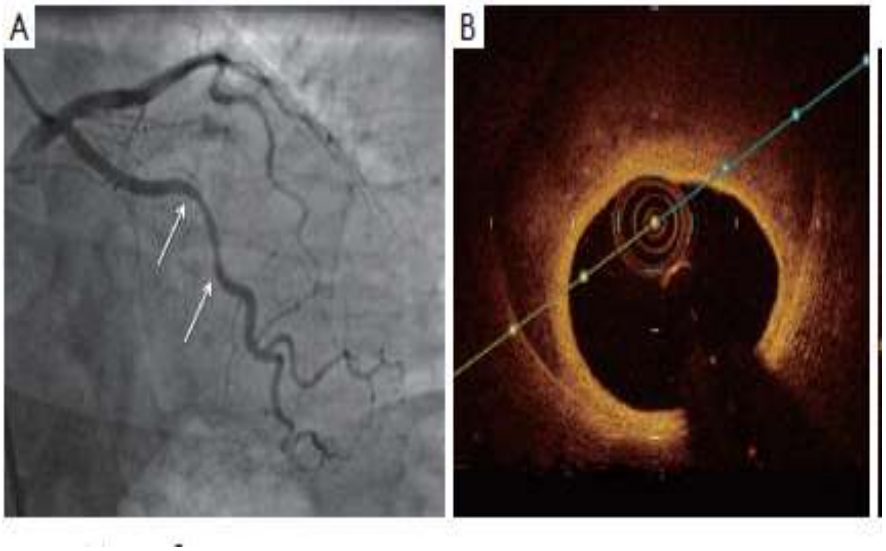



Type 1 SCAD. (A) Cranial projection showing double-lumen (arrow) with contrast dye hang-up in the proximal LAD, and occluded mid LAD from SCAD; (B) caudal projection showing double-lumen (arrow) of proximal LAD from SCAD.

- Type 2 (diffuse stenosis of varying severity): this angiographic appearance is not well appreciated and is often missed or misdiagnosed.
- SCAD commonly involves the mid to distal segments of coronary arteries, and can be so extensive that it reaches the distal tip. There is an appreciable abrupt change in arterial caliber, with demarcation from normal diameter to diffuse narrowing. This diffuse (typically >20 mm) and usually smooth narrowing can vary in severity from mild stenosis to complete occlusion.




- (III) Type 3 (mimic atherosclerosis): this appearance is the most challenging to differentiate from atherosclerosis *and most likely to be misdiagnosed.*
- Angiographic features that favor SCAD are: (i) lack of atherosclerotic changes in other coronary arteries;
- (ii) long lesions (11-20 mm); (iii) hazy stenosis; and
- (iv) linear stenosis. Angiographer needs to have a high index of suspicion for SCAD *and* employ intracoronary imaging for such cases.




- There are no apparent differences in clinical presentations with the three angiographic classifications of SCAD.
- Type 1 SCAD should be easily diagnosed with angiography.
- Type 2 and 3 SCAD mostly intracoronary imaging for diagnosis.

## *Intracoronary imaging*

- The current gold-standard coronary angiography is an imperfect tool for the diagnosis of SCAD, because it is only a 2-dimensional luminogram.
- It is excellent to assess luminal narrowing; however, it is poor in assessing the arterial wall, where the key abnormalities occur with SCAD. The pathognomonic type1 angiographic SCAD appearance of multiple radiolucent lumen and contrast dye stains in the arterial wall can be demonstrated well on angiography; however, this is not the most frequent angiographic manifestation of SCAD. In fact, the most common angiographic SCAD appearance is the type 2 long diffuse stenosis, which is seen in 2/3 of SCAD cases.

- Type 3 angiographic appearance that mimics atherosclerosis is less
- So angiographic type 2 and 3 appearances of SCAD, contributing to under-diagnosis of SCAD.
- IVUS and OCT are both tools that allow angiographers to better visualize the arterial wall structure and composition.

## IVUS

- IVUS has a lower resolution (150-200 μm), but has deeper penetration allowing for the full vessel and extent of the IMH to be visualized.
- IVUS can delineate true and false lumens and detect IMH, which appears as a homogenous collection behind the intimal-medial membrane.
- A small study that utilized IVUS to detect SCAD showed IMH in all five patients who had angiographically normal coronary arteries

- OCT, on the other hand, is a much higher resolution (10-20 μm) modality and can visualize true lumen, false lumen, and even intimal tears .
- However, it has poorer penetration than IVUS, and may not visualize the full extent of the IMH .
- In comparative studies of IVUS and OCT , OCT was shown to be more sensitive and better at detecting more characteristics of SCAD than IVUS, especially for identifying intimal tears and flaps. Whereas, both OCT and IVUS were able to identify IMH equally.

## Other imaging modalities

- The majority of studies evaluating SCAD with computerized-tomography (CT) scans were done prior to the widespread use of cardiac CT angiography (CCTA) .
- Multi-detector CT scans were used in those cases to follow the course of SCAD and found the presence of IMH on CT 3 days post-event and near resolution on CT with only mild wall thickening at 10 days postevent.
- Despite improvements with CCTA technology, its resolution is still much lower than conventional angiography.
- Smaller diameter arteries (<2.5 mm) are not well visualized.

- Therefore, CCTA is not recommended as first-line imaging to rule out SCAD.
- However, CCTA might be useful as a non-invasive technique to assess arterial healing after SCAD of larger proximal-mid coronary arteries.

- Magnetic resonance imaging (MRI) has been used in detection of SCAD in one published case study .
- Hyperintensity was noted in the area seen to have an IMH on IVUS and CCTA. Full resolution of hyperintensity was noted at 23 days post-event.
- This finding is unlikely to be specific or sensitive to detecting SCAD. However, like CCTA, it can be a good non-invasive technique to assess for serial progression of arterial healing with minimal radiation exposure.

## Management

- Early diagnosis is crusial for managing SCAD patients, because the use of unnecessary and potentially harmful pharmacologic therapies may be avoided.
- Unlike in the case of atherosclerotic coronary artery disease, there are noprospective randomized data that specifically address the management of SCAD.

## Medical therapy

- It is unclear if the standard ACS pharmaceutical agents are beneficial for SCAD.
- The role of antiplatelet therapy for SCAD patients not treated with stents is unclear.
- Considering the totality of evidence for aspirin in ACS and secondary prevention of CAD , along with low side-effect profile and bleeding risks, aspirin appears reasonable for acute and long-term SCAD treatment.
- The addition of clopidogrel for SCAD patients not treated with stents is also of uncertain benefit. Considering that a proportion of SCAD involves intimal tear, which is prothrombotic, this would empirically benefit from dual antiplatelet therapy.

- Reducing false lumen thrombus burden with antiplatelet agents could theoretically reduce true lumen compression .
- Routine administration of aspirin and clopidogrel for SCAD patients acutely; aspirin is then continued life-long and clopidogrel for up to 1 year.
- The role of new P2Y12 antagonists (prasugrel and ticagrelor) for SCAD is unclear. The role of GPIIb/IIIa inhibitors for acute SCAD management has also not been evaluated, but because of their greater potency, higher bleeding risk, and potential risk of extending dissection, they are not routinely used for SCAD.

- The role of anticoagulation for SCAD is controversial with the risk of dissection extension balanced by the potential benefit of resolving overlying thrombus and improving true lumen patency.
- Heparin agents are typically administered for ACS patients on hospital presentation; and discontinue heparin once SCAD is proven on angiography to avoid extension of IMH.

- Thrombolytic therapy should be avoided in SCAD, as there have been reports of harm and clinical deterioration due to extension of dissection and IMH .
- In the retrospective review by Shamloo *et al. , of 87 SCAD* patients who received thrombolysis, 60% had clinical deterioration requiring rescue percutaneous coronary intervention (PCI) or coronary artery bypass grafting (CABG). Thus, early coronary angiography is important if SCAD is suspected because medical management deviates from standard ACS therapy.
- But , in remote centers without onsite or established transfer algorithms for primary PCI, thrombolysis should not be delayed for ST-elevation MI patients since the overall frequency of thrombotic occlusion is much higher than SCAD.

- Beta-blockers reduce arterial shear stress and are beneficial in reducing coronary arterial wallstress, similar to the benefits in aortic dissection.
- The use of nitroglycerin may be useful in alleviating ischemic symptoms from overlying vasospasm during acute SCAD presentation, but are not routinely used
- long-term. Angiotensin-converting enzyme inhibitors are only routinely indicated when there is significant LV dysfunction after the MI (ejection fraction ≤40%; class 1 indication).
- The use of statins for non-atherosclerotic SCAD has not been studied, and its indicated in patients with preexisting dyslipidemia.

## Revascularization

- The decision to revascularize the dissected artery depends on the patient's clinical status and affected coronary anatomy. In most cases, conservative treatment is preferred for stable patients without ongoing pain.
- Patients with ongoing chest pain, ischemia, ST elevation, or hemodynamic instability should undergo PCI, especially when the dissection affects major arteries with sizable myocardial jeopardy .
- *Patients with dissected* left main or proximal segments of left anterior descending (LAD), circumflex or right coronary artery should be stented if feasible.

- When the dissected artery segment is distal, of small calibre, or when the dissection is extensive, stenting may not be practical. So conservative treatment is indicated.
- The success rate with PCI for SCAD is poor compared with atherosclerotic lesions.
- PCI of dissected coronary arteries can be challenging and often terminate with suboptimal results.

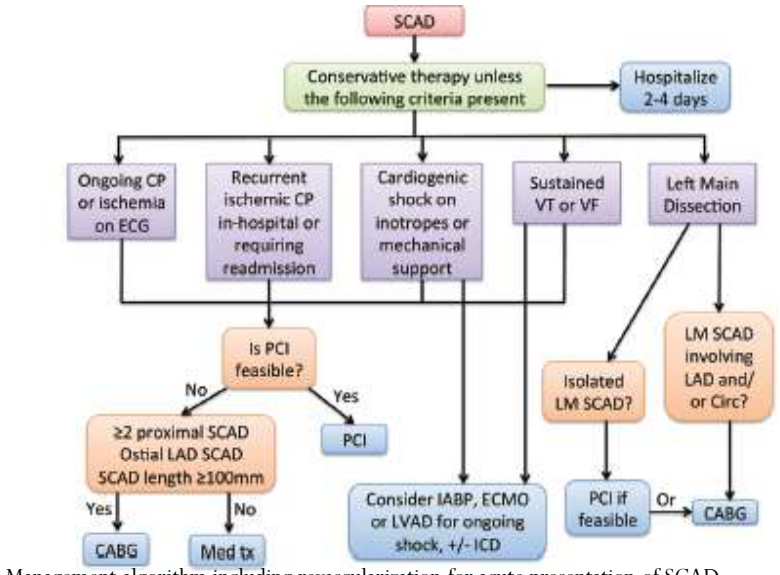
## Why ??

- To begin, it may be challenging to advance the coronary guidewire into the distal true lumen.
- The IMH of the dissected segment can also propagate antegradely or retrogradely with angioplasty, further compromising arterial blood flow and extending the dissection.
- The dissections often involve distal coronary arteries, which are too small to implant stents. Even if the dissected arteries are large, the dissections are often extensive requiring long stents and thus increasing the risks of restenosis.
- Furthermore, IMH resorb and heal over time, and may result in late strut malapposition, increasing the risk of very late stent thrombosis especially after cessation of dual anti-platelet therapy.
- ***Stent length ?? --- focal stenosis , long lesion.***
- ***BVS ??***

## CABG

- Emergency CABG should be considered if the dissection involves the left main.

- Lastly, the natural history of the dissected segments is such that the vast majority heals spontaneously, and patients appear to have good long-term outcome if they survive their initial event.
- Its recommend to reserve PCI for patients with ongoing chest pain and ischemia when the lesion is amenable to stenting, and to consider CABG for extensive dissections involving the left main.




Management algorithm including revascularization for acute presentation of SCAD.

# Repeated imaging

- There is no consensus as to repeat imaging after SCAD, irrespective of revascularization strategies.
- But, because a significant proportion of patients have recurrent chest pains after their initial event, its useful to repeat coronary angiography several weeks later to investigate potential ischemic causes of pain, and to assess arterial healing.

## Prognosis

- Initial mortality rates appeared to be over-estimated due mainly to post-mortem reporting and little ante-mortem data. A collection of case reports from 1980-2000 showed mortality rates ranging from 0% to 7% .
- More recent studies reported lower in-hospital mortality rates ranging from 1% to 5%.
- However, when analyzed with matched ACS controls, long-term mortality was lower for SCAD patients than ACS controls in this retrospective study, but cardiac events were similar in the two groups. SCAD patients can also present with recurrence of repeat SCAD, MIs, repeat or subsequent revascularizations, congestive heart failure, and other cardiac events.

## Rehabilitation

- Cardiac rehabilitation should be recommended to all SCAD patients. Many SCAD patients are not initially referred for cardiac rehabilitation, perhaps because of such factors as female gender, youth, overall good health, or fear of recurrent SCAD. This lack of rehabilitation has the effect of delaying patients' return to normal activity and mental health.
- Cardiac rehabilitation is particularly important in SCAD patients, who often have substantial anxiety, depression, and ongoing physical symptoms.
- The association of recent extreme physical activity with SCAD has led some healthcare providers to prohibit even minimal physical exertion, such as lifting more than 5 pounds. This approach has no evidence to support it and, if applied over a lifetime, has the potential to substantially increase the risk of such health conditions as osteoporosis, injuries from falls, obesity, and atherosclerosis.

- Until there is evidence that moderate exercise is harmful, it seems prudent to provide activity guidelines that include 30 to 40 minutes of moderate aerobic activity daily.
- Avoid of weightlifting and bodybuilding but generally encourage resistance training, with light weights and high repetitions, avoid competitive racing.
- Individuals with recurrent SCAD or extensive fibromuscular dysplasia (FMD) might need more conservative activity recommendations.

## Home message

- Knowledge is key of perception , you can`t see what you don`t know .
- Think (uncommon ) for uncommon presentation.
- Young female with ACS , and no classic RF for CAD , put in mind SCAD.
- Tailor your management strategy according to clinical scenario, no specific guidelines for SCAD.

I am always searching something better for YOU in this chaotic world.