

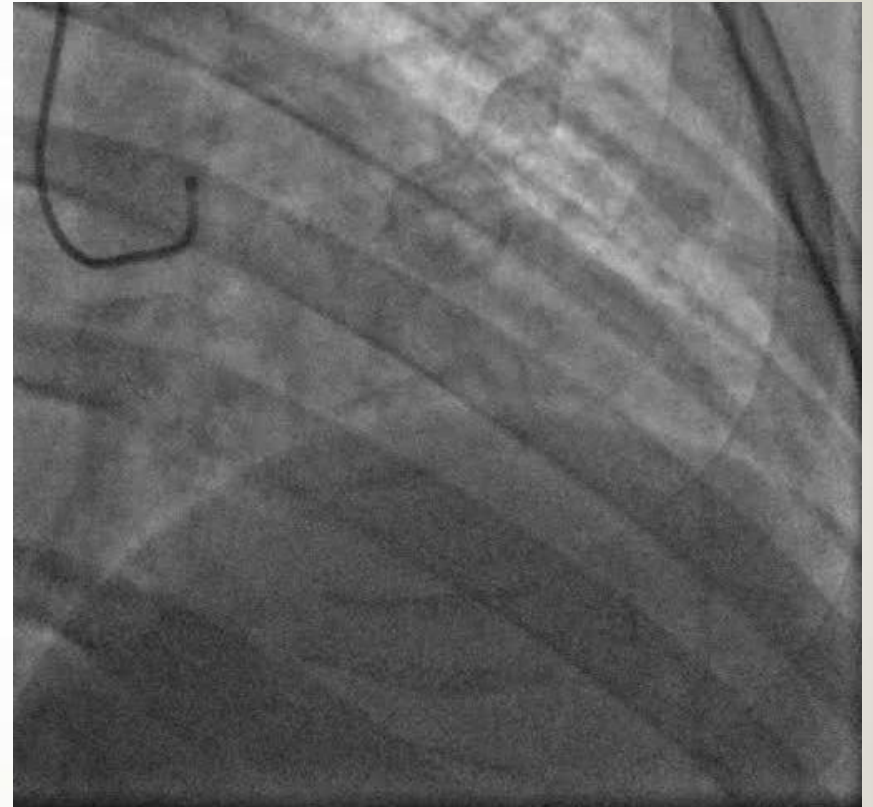
# Spontaneous Coronary Artery Dissection

Esther S.H. Kim, MD, MPH  
Director, Arteriopathy Clinic  
Associate Professor of Medicine  
Vanderbilt University Medical Center  
April 18, 2018

No Disclosures

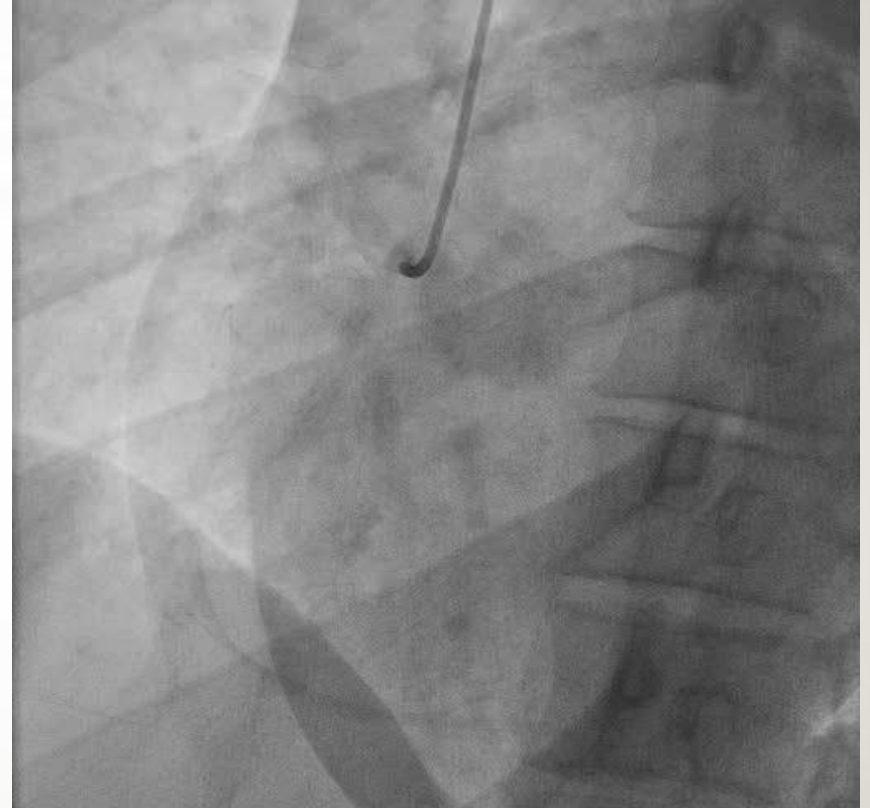
# Patient Presentation

- 33 year old woman
- 11 days postpartum
- Finishes nursing her baby, lays down and develops severe chest pressure
- Anterior STEMI → LHC



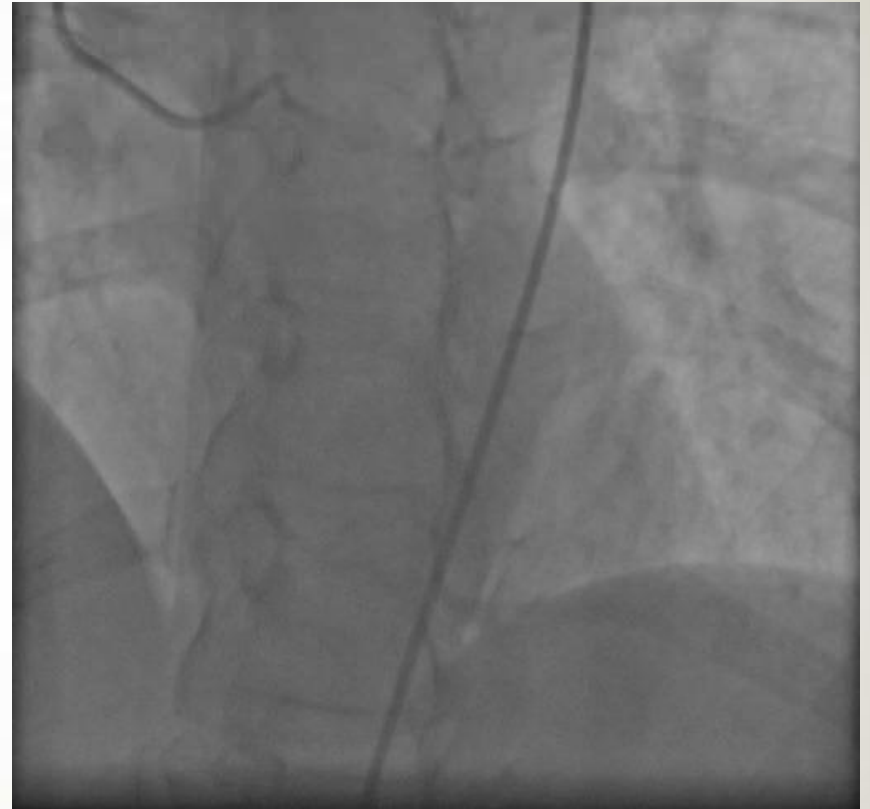
# Patient Presentation

- 42 year old woman just finished exercise class, felt faint and then felt chest pressure, nausea
- Inferior STEMI → LHC



# Patient Presentation

- 46 year old woman develops chest pain while driving
- Anterior STEMI → LHC



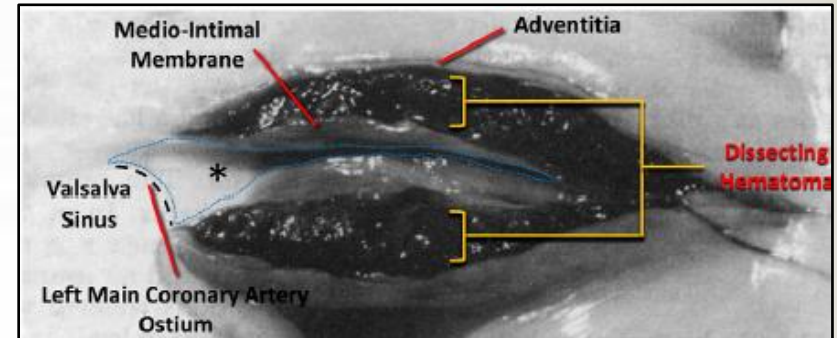
# Patient Presentation

- 61 year old nurse
- Chest pain at work
- NSTEMI → LHC



# Spontaneous Coronary Artery Dissection

- *Nontraumatic, noniatrogenic* separation of the coronary arterial wall by intramural hemorrhage creating a false lumen, with or without an intimal tear.
- Compression of arterial lumen by IMH or obstructed by dissection flap → myocardial ischemia or infarction



*Int J Cardiol 2014;175(8-20)*

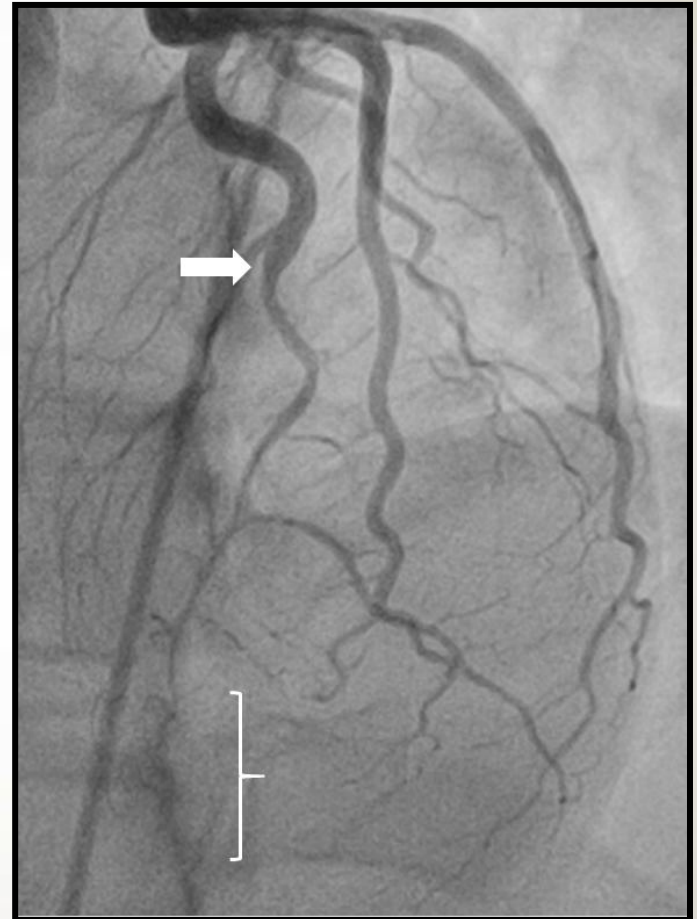


*Histopathology 2005, 47, 215–226*

# SCAD Epidemiology

- ♀ : ♂ = 9:1
- Prevalence uncertain
  - 1-4% of all ACS
  - 35% of ACS in ♀ ≤50 yrs
  - 43% of pregnancy-related MI
- ACS - most common presentation
- Mean age 45-53 years [20s – 80s]
- Rare (?) +/- underdiagnosed

1. Circ Cardiovasc Interv 2014;7:645-655
2. Eur J Cardiothor Surg 2009;35:250-4





# Who gets SCAD?

Middle-aged, white women with few traditional CV risk factors



<http://www.foxnews.com/health/2015/08/24/scientists-seeking-cause-mysterious-heart-attack-plaguing-young-women.html>

Smoking

Canada (n=27)	United States (n=189)
44.9 +/- 9.6	44.9 +/- 9.6
83%	92%



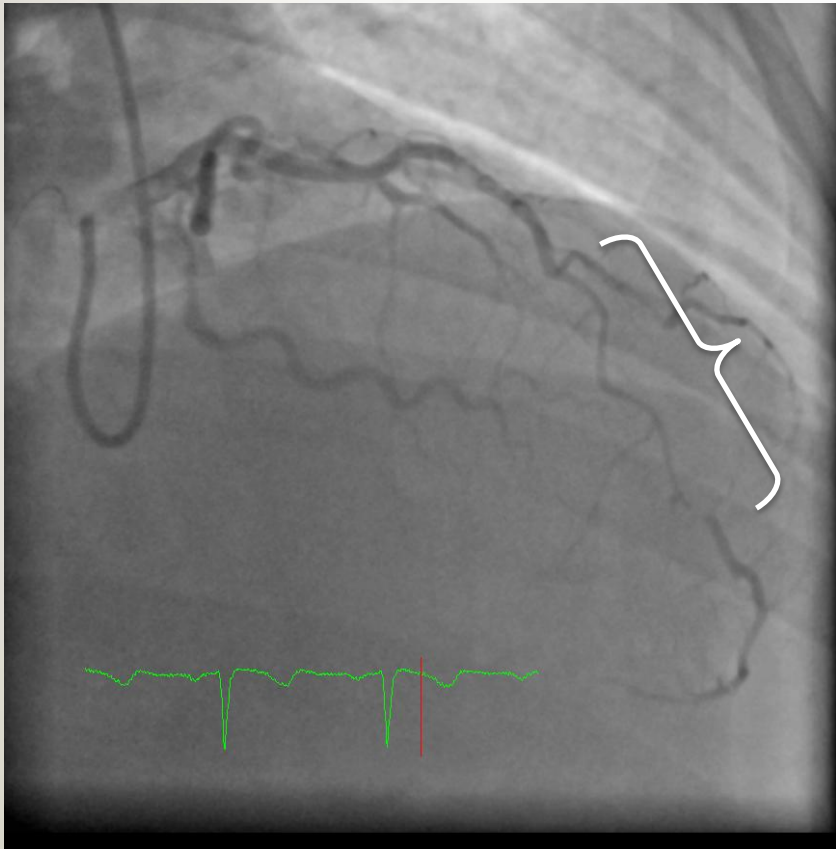
Katherine Leon  
Founder, SCAD Alliance



<https://news.vanderbilt.edu/2017/02/23/new-clinic-focuses-on-treating-complex-arterial-disorders/>

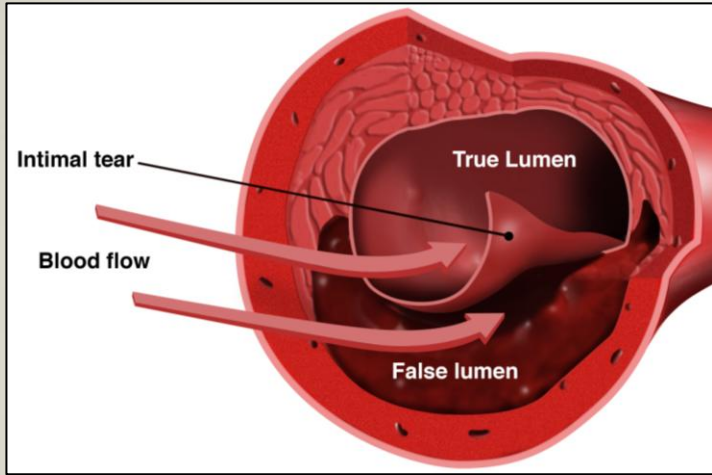
32.8%
51.4%
34.3%

# Clinical Presentation



- ACS with +troponin, the “rule”
- VT/VF, SCD 3-11%
- Cardiogenic shock 2-5%
- LV WMA common early on but overall LV function often preserved

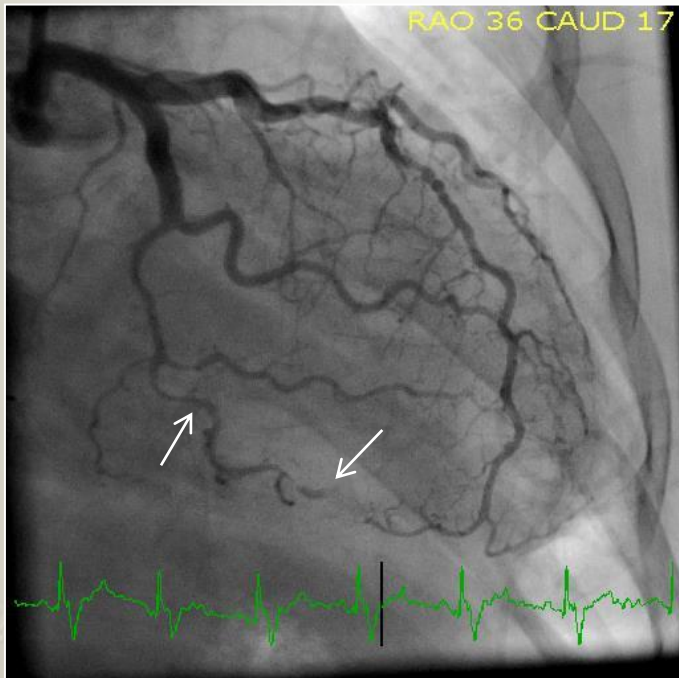
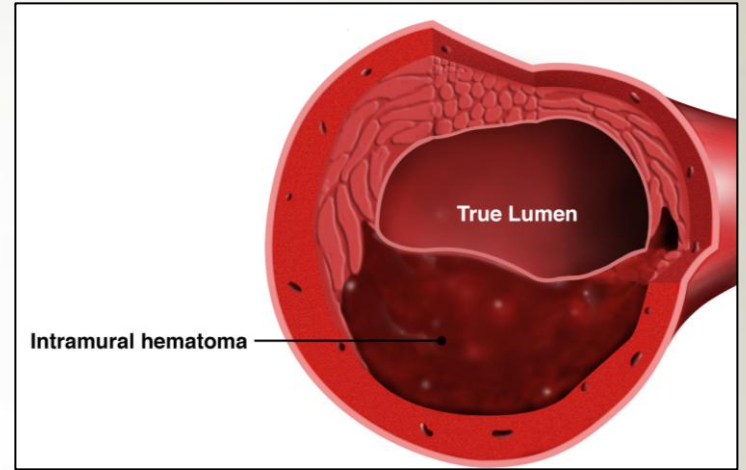
# SCAD Pathophysiology



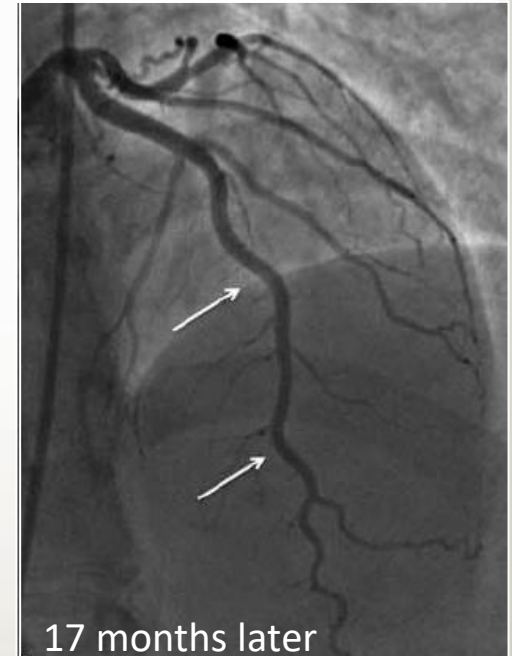
Intima intact?

Yes →

← No



Atherosclerosis mimic  
Type 3 SCAD

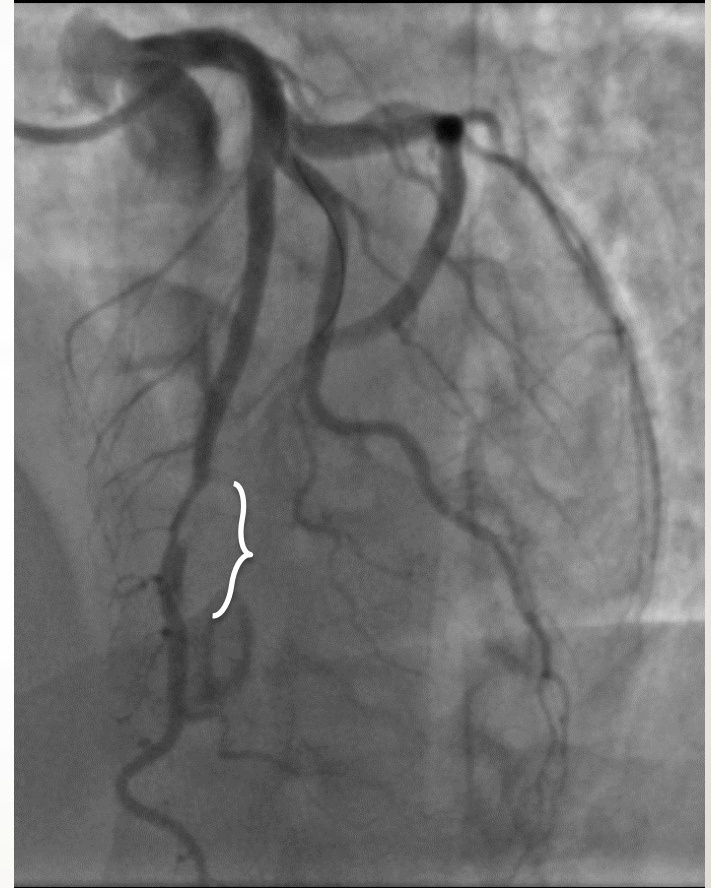
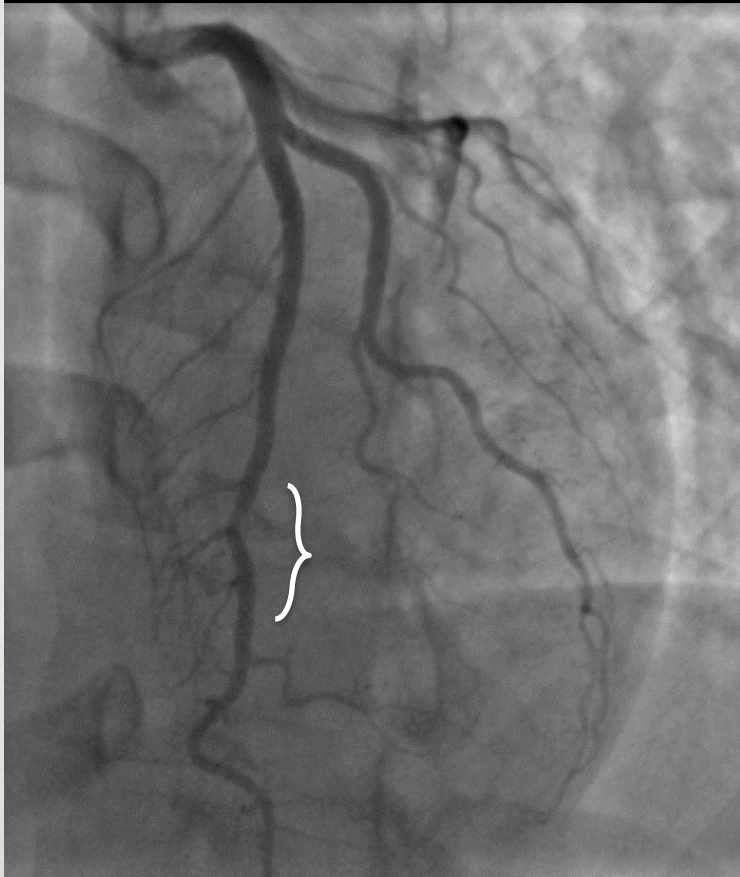


# Acute Management of SCAD

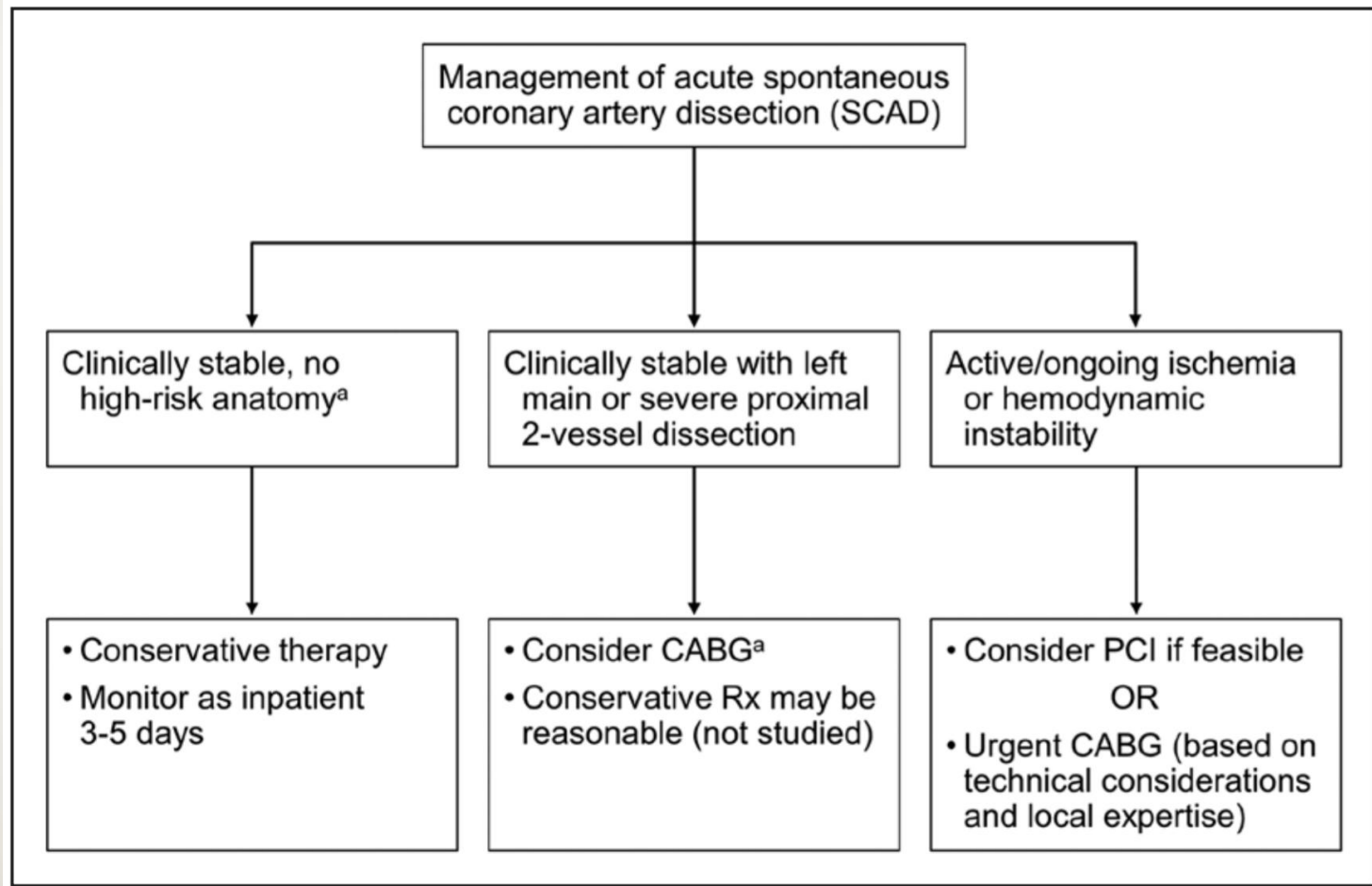


- SCAD  $\neq$  plaque rupture
- The tools we have to treat ACS were designed for plaque rupture
- Anticoagulation, lysis  
?benefit vs harm
- PCI ?benefit vs harm

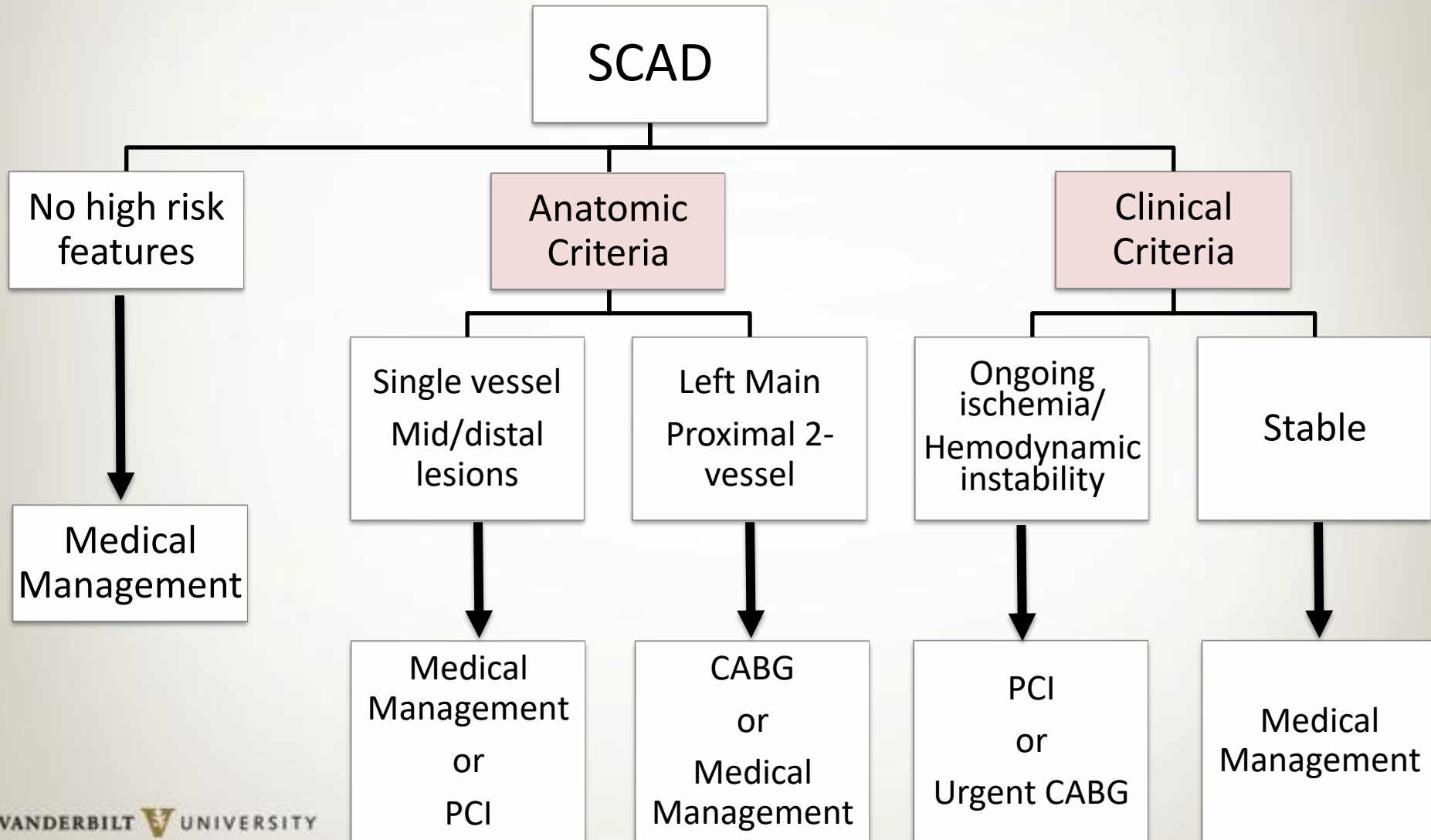
# Conservative Therapy for SCAD



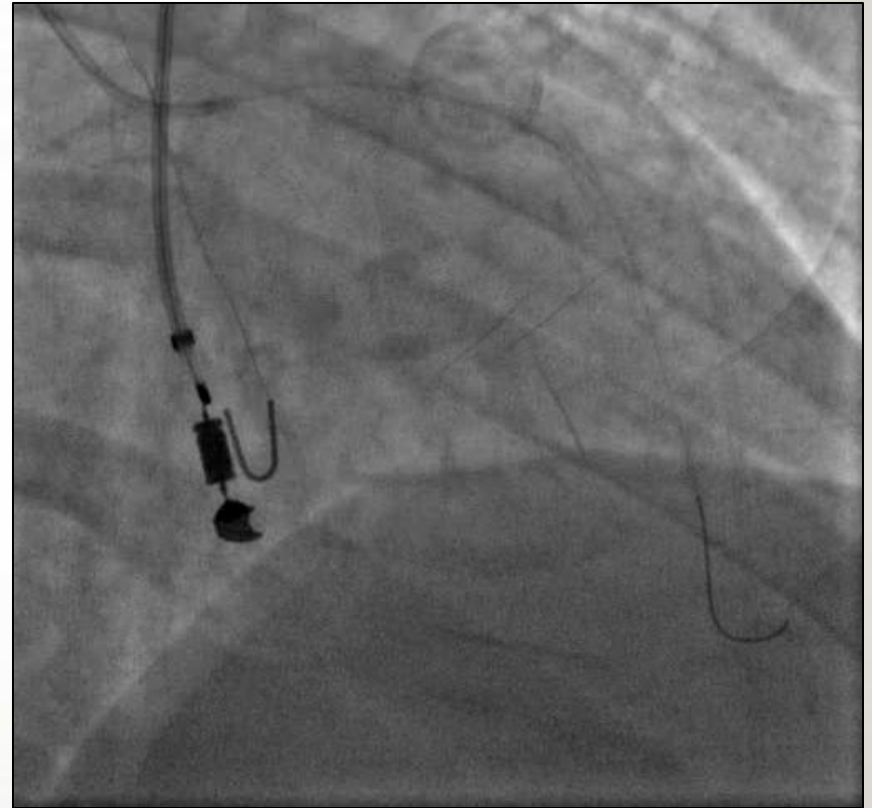
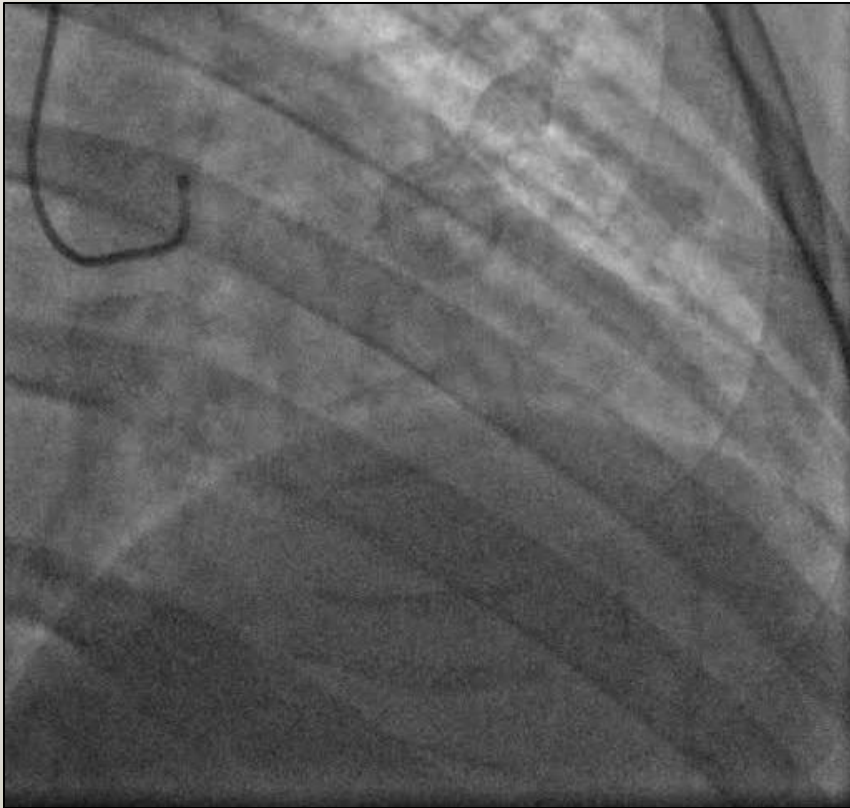
# Acute Management of SCAD



# Acute Management of SCAD



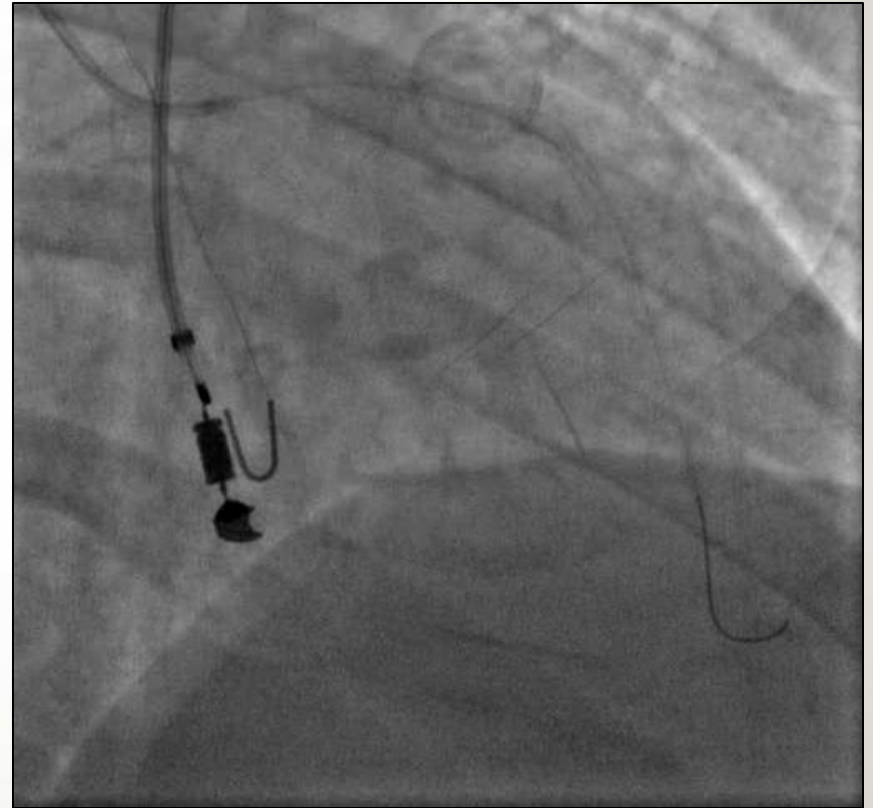
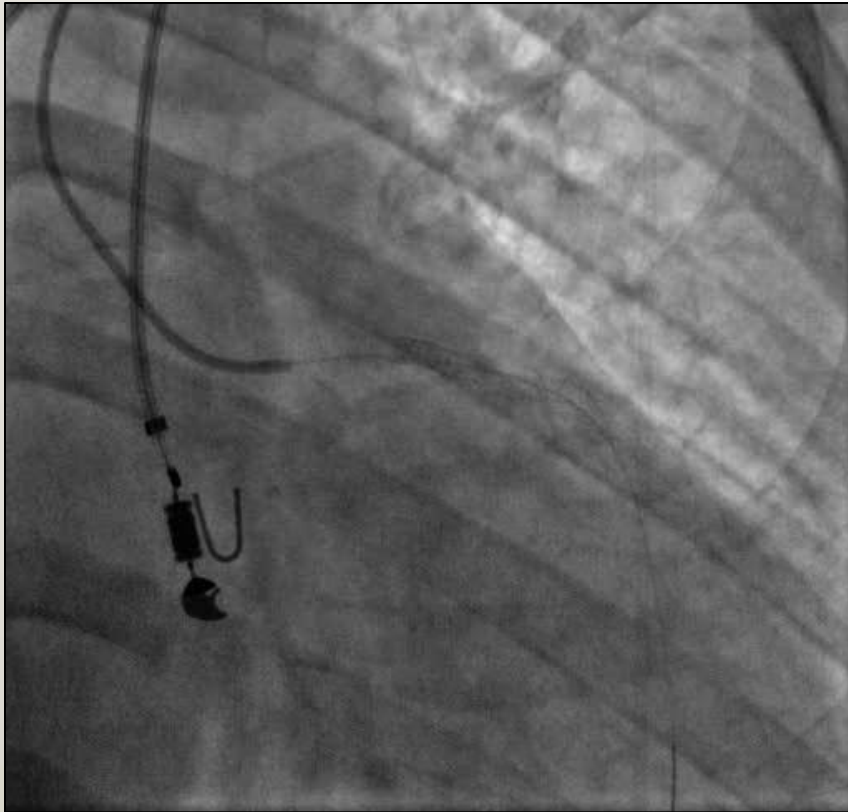
# 33 year old woman, post-partum





# 33 year old woman, post-partum

Final Result



# Poor Short Term Outcomes with PCI

1. Antegrade and retrograde extension of dissection/IMH → worsening obstruction
2. “weak” arteries → susceptibility
  - catheter-induced dissection of unaffected proximal segments (3.4% in SCAD pts vs <0.2% non-SCAD)
3. Distal lesions too small for stents
4. Guide wires in the false lumen



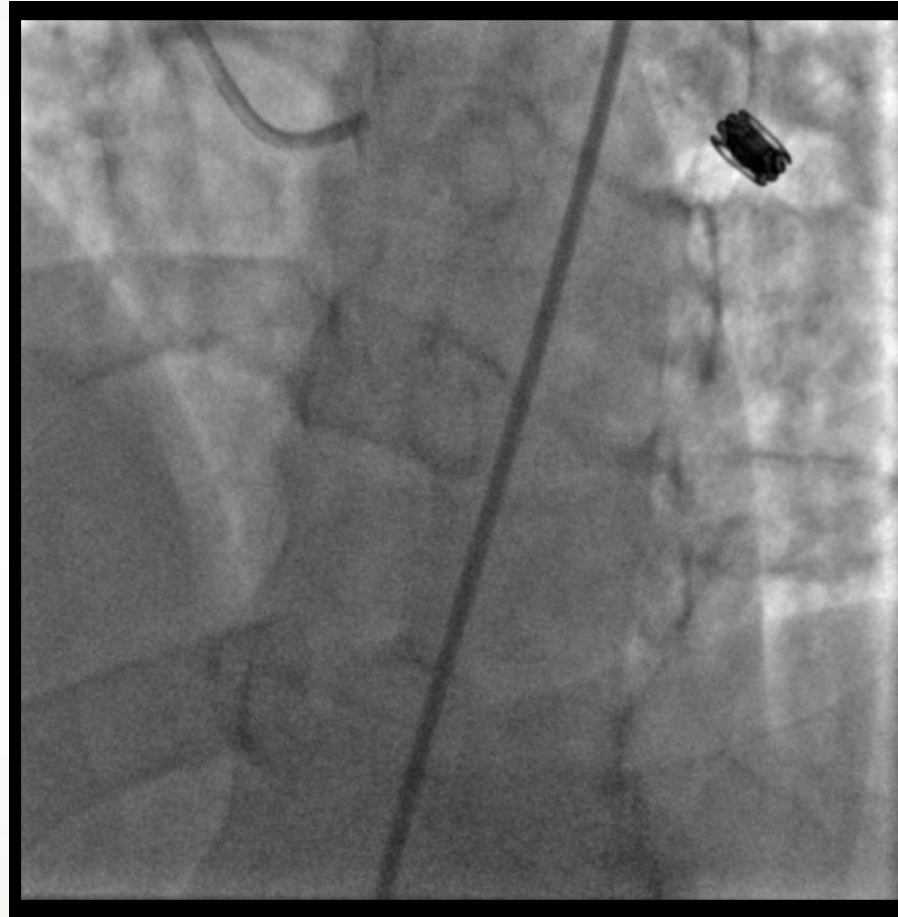
*Sealing intima may displace IMH proximally → loss of flow*

# Short Term Outcomes

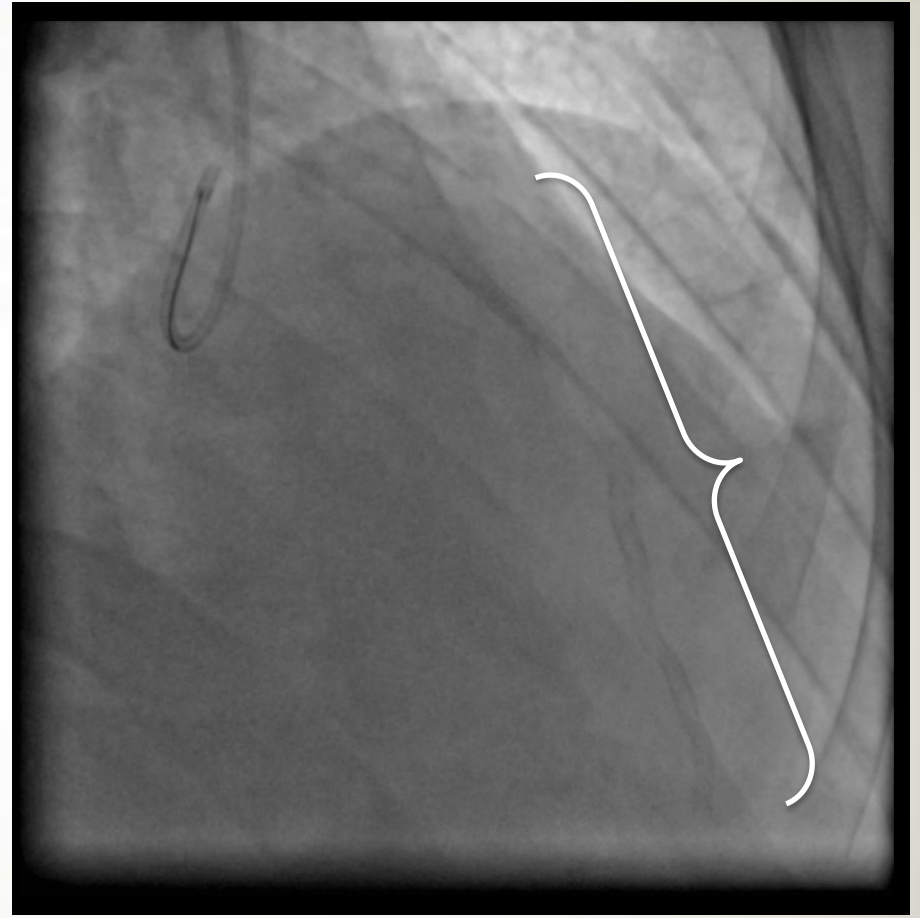
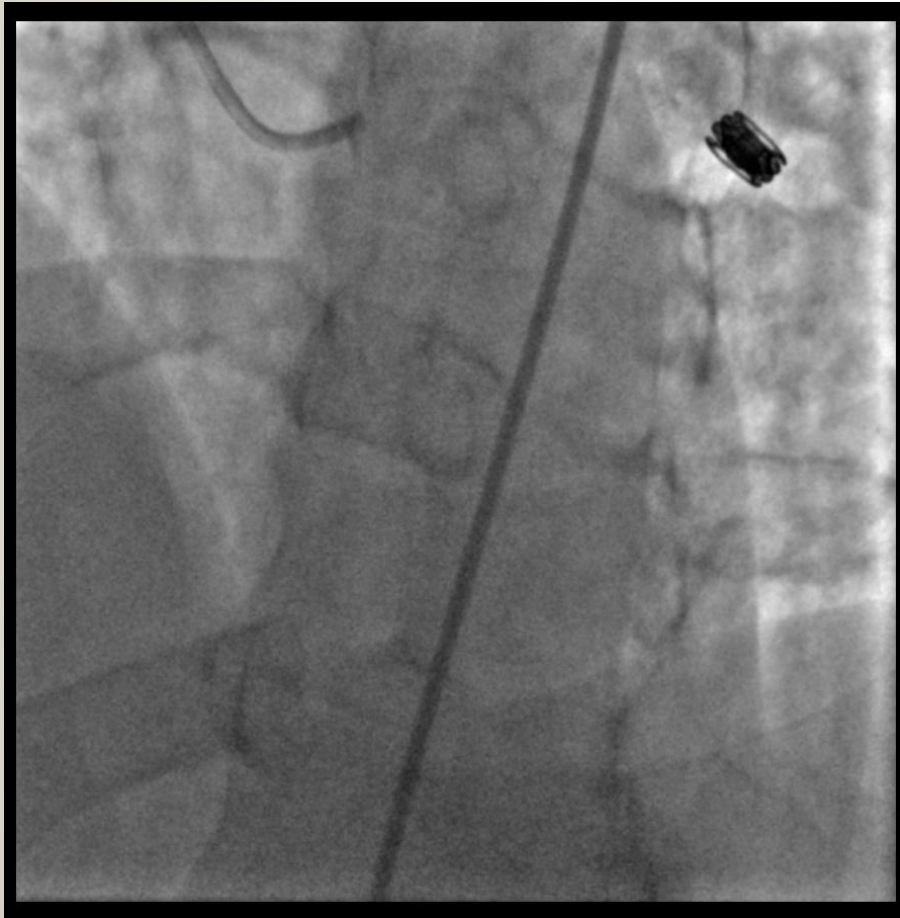
	Year	N	Medical Therapy n=patients (%)	Crossover to Revasc n=patients (%)	All in- hospital Revasc n=patients (%)	All PCI (% success)	All CABG (% success)	In-hospital MI n=patients (%)	In-hospital urgent revasc n=patients (%)	In hospital mortality
Saw	2017	327	272 (83.2%)	9 (3.3%)	61 (18.7%)	54 (43.1%)	7 (NR)	15 (4.6%)	14 (4.8%)	0
Tweet	2014	189	94 (49.7%)	8 (8.5%)	103 (54.4%)	97 (47.4%)	20 (94.1%)	NR	26 (14%)	1 (0.53%)
Lettieri	2015	134	78 (58.2%)	2 (2.6%)	58 (43.3%)	55 (72.5%)	8 (87.5%)	7 (5.2%)	7 (5.2%)	3 (2.2%)

- Treatment with medical therapy 49.7-83.2%
- PCI success rates 43.1% - 72.5% (vs >95% in ASO)
- In hospital MI 4.6-5.2%
- Cross-over to revascularization 2.6-8.5%
- In hospital mortality 0-2.2%

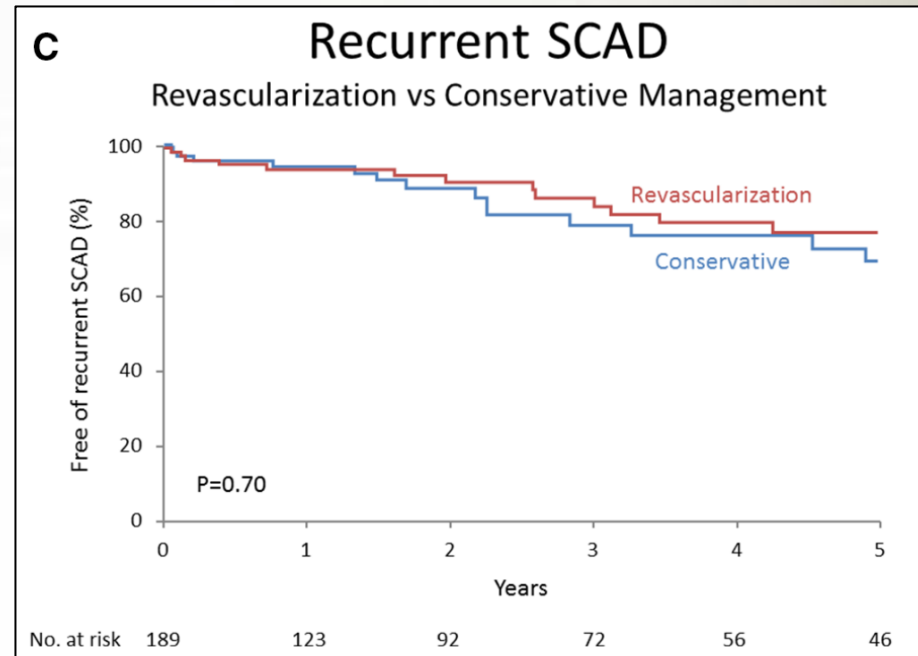
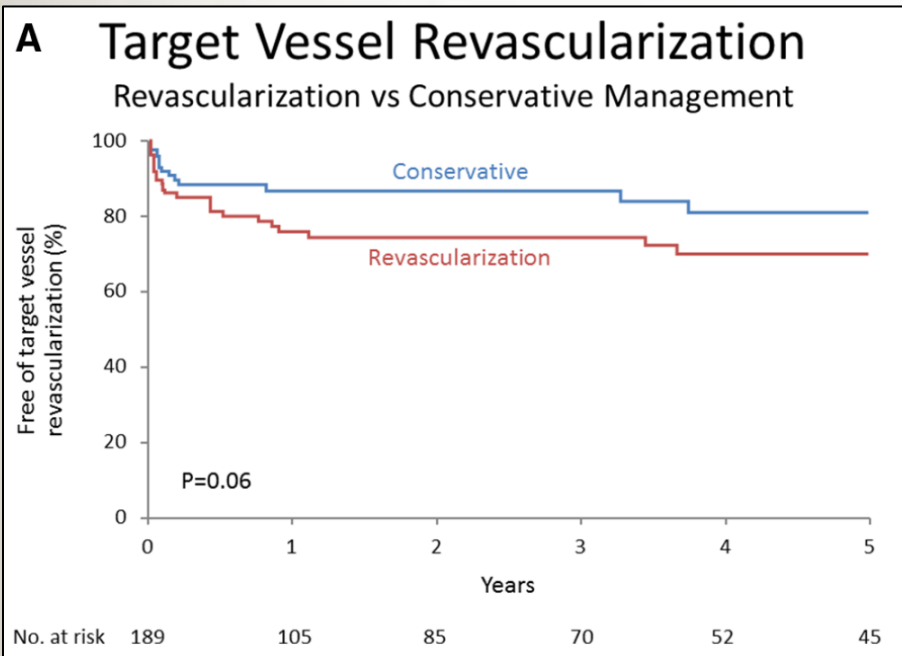
# 37 year old woman chest pain after sneezing



4 months and 6 LHC later...



# Poor Long-Term Outcomes with PCI

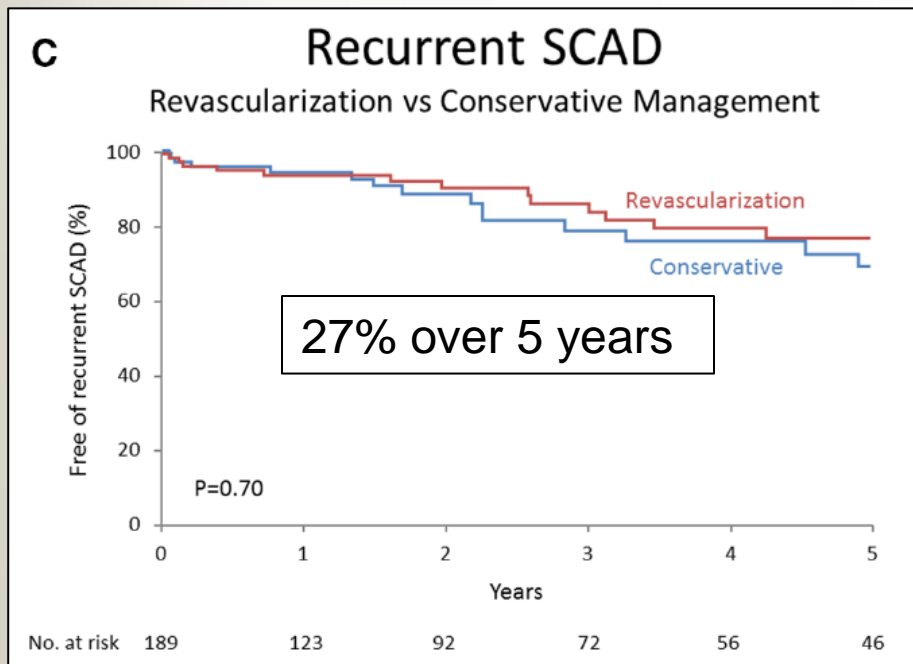


- Long lesion lengths requiring long or multiple stents increases risk of ISR, IST
- IMH resorption → malapposition of stents increasing risk of ISR and stent thrombosis

# Recurrence

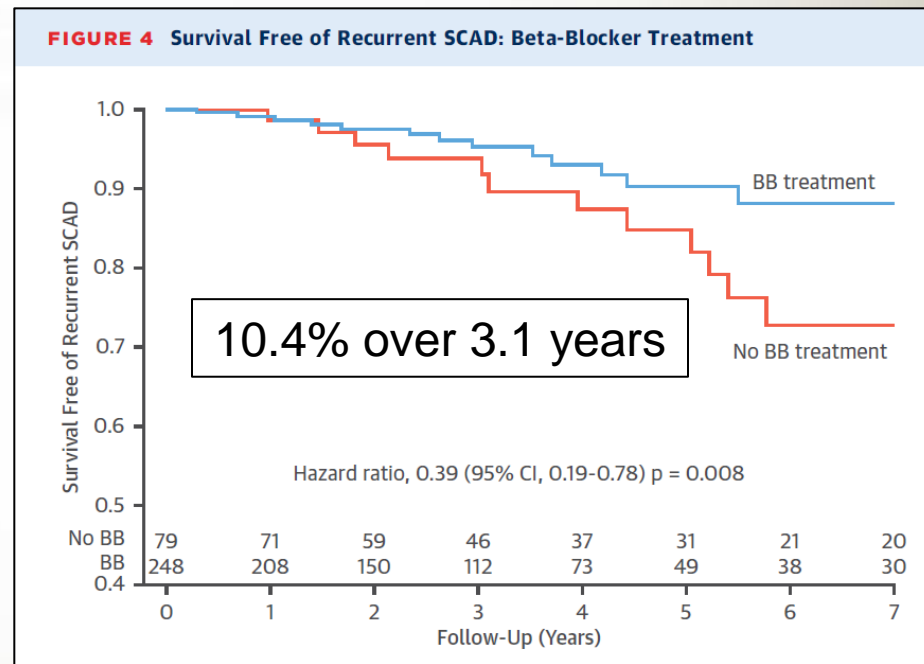
(not to be confused with extension)

## Mayo Series (n=189)



*Circ Cardiovasc Interv.* 2014;7:777-786

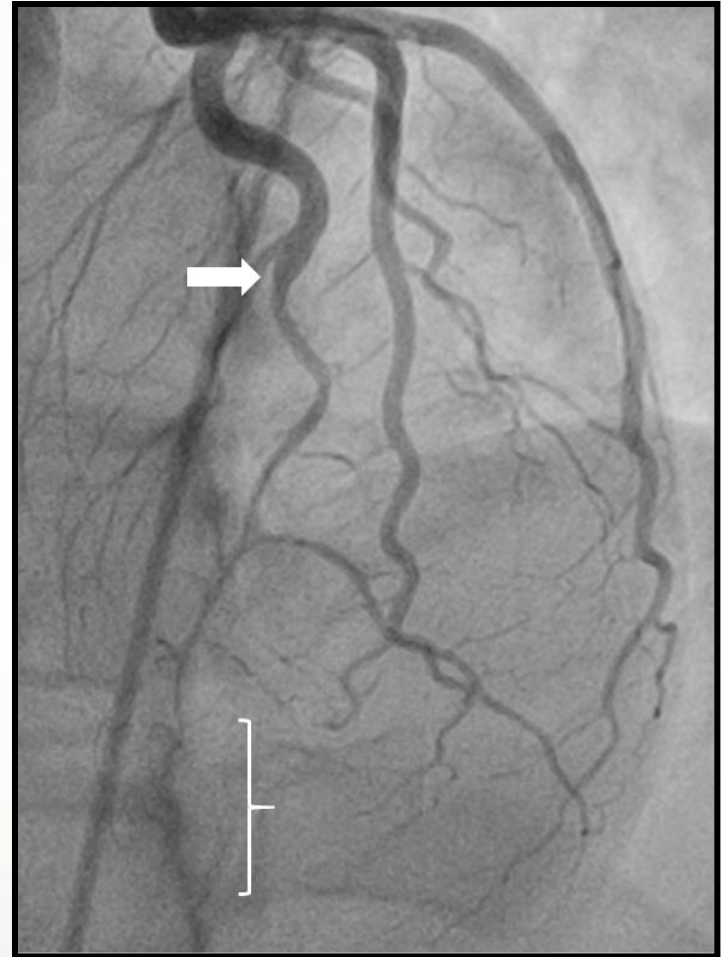
## Vancouver Series (n=327)



*JACC* 2017;70:1148-58

# Patient Presentation

- 45 year old woman with no significant cardiac history
- Acute onset chest pain at work
- NSTEMI → LHC
- PCI attempted – no change in appearance of LAD lesion
- Discharged on aspirin and clopidogrel
- She presents to clinic to establish cardiology care





# Spontaneous Coronary Artery Dissection

## Clinical Questions

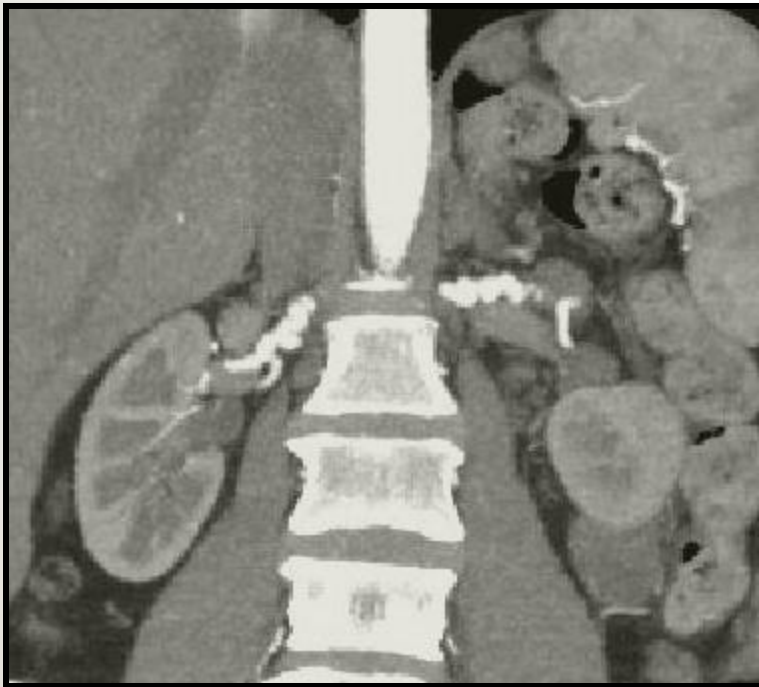
- Are further evaluations necessary
- What history should I obtain?
- What physical exam findings should I be looking for?
- What laboratory tests should I order?
- What imaging tests do I order?
- Do I need to consult genetics?

## Root Questions

- Is SCAD a disease in isolation?
- What is the etiology of SCAD?
- How do I diagnose extracoronary abnormalities?
- Is SCAD a genetic disorder?

# Is SCAD a Disease in Isolation?

**CTA Aorta**

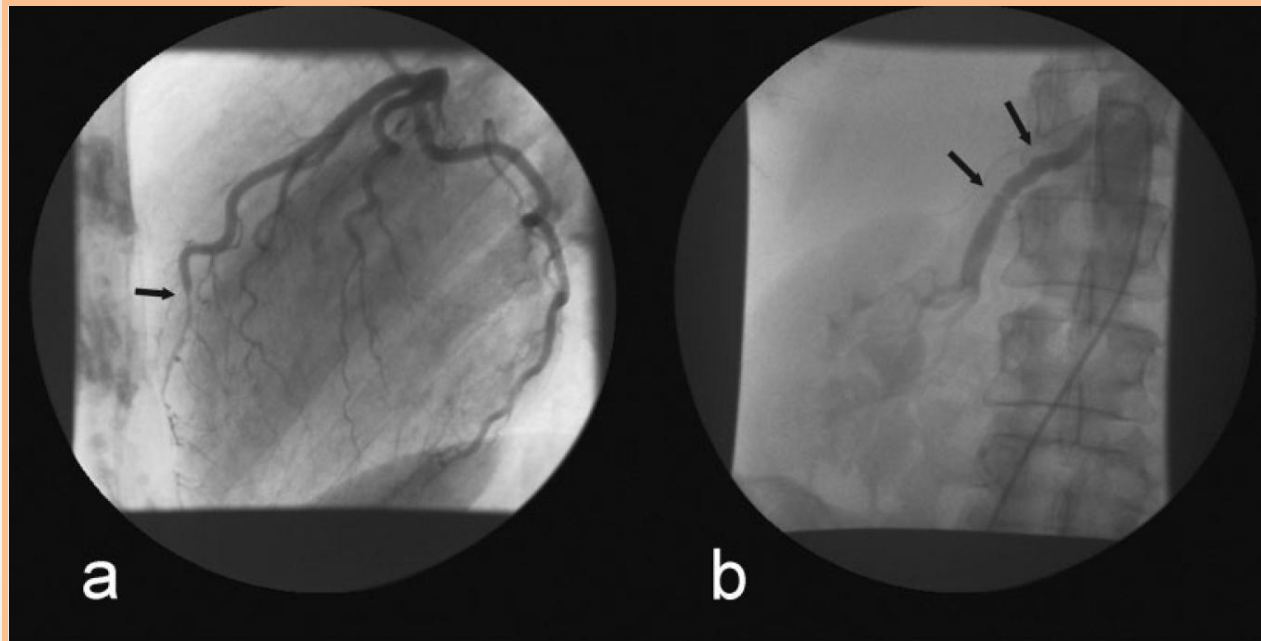


**CTA Neck**



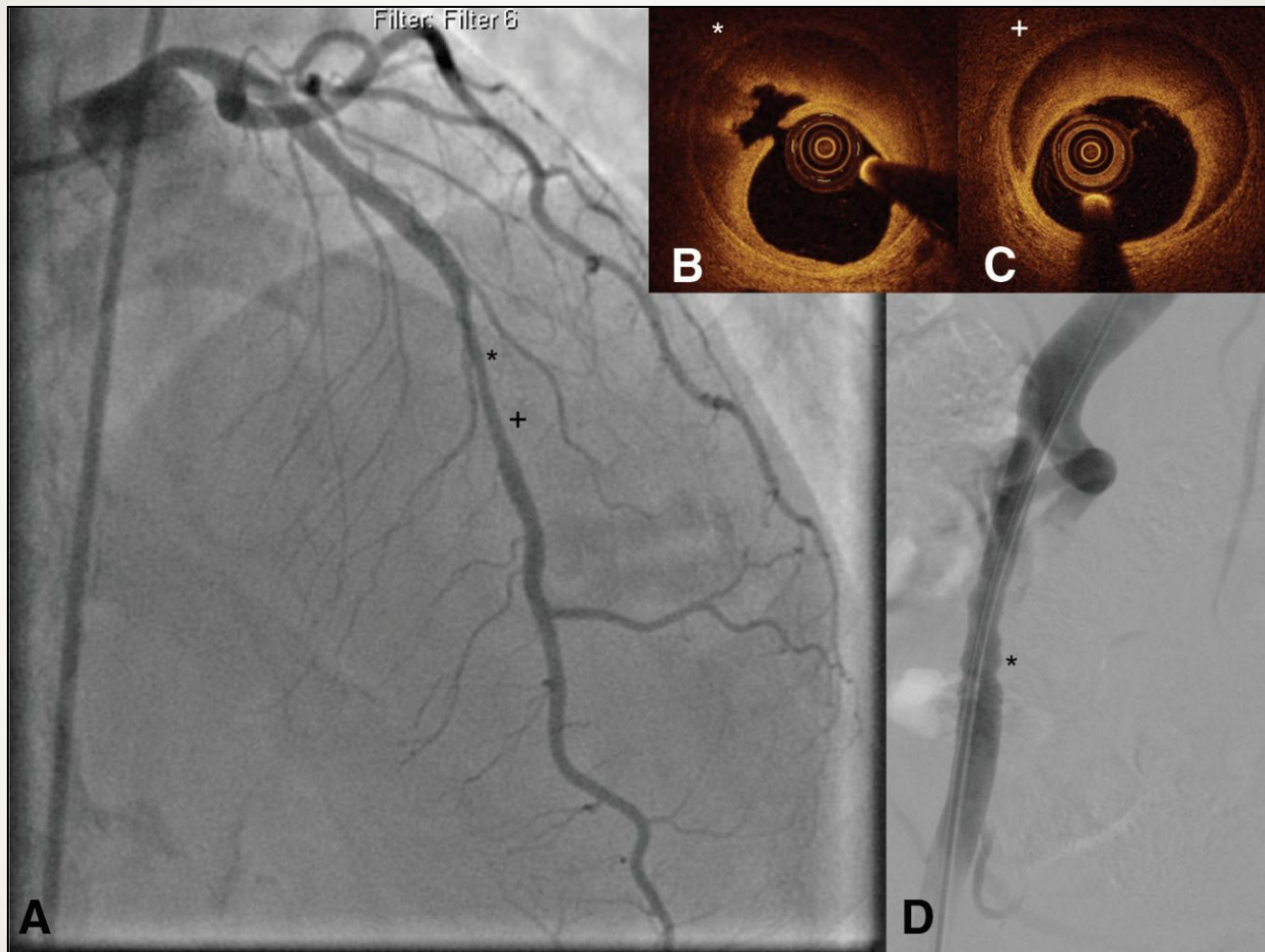
45 F multivessel SCAD

# Is SCAD a Disease in Isolation?



- First publication showing SCAD angiogram image next to extracoronary FMD image
- 7 women with ACS, unusual coronary angiography, renal artery FMD. Authors proposed this was coronary appearance of FMD
- “...coronary dissection is a condition that we considered....None of the patients demonstrated angiographic findings of a double lumen, spiral lucency or contrast staining.”

# SCAD Is Not A Disease In Isolation



Spontaneous Coronary Artery Dissection in Patients With Fibromuscular Dysplasia. Saw J et al. (Circ Cardiovasc Interv. 2012;5:134-137.)

# SCAD Is Not A Disease In Isolation

## Canadian Cohort

- N=168
- 86.3% complete screening
  - 72.0% FMD
  - 10.1% cerebral aneurysm

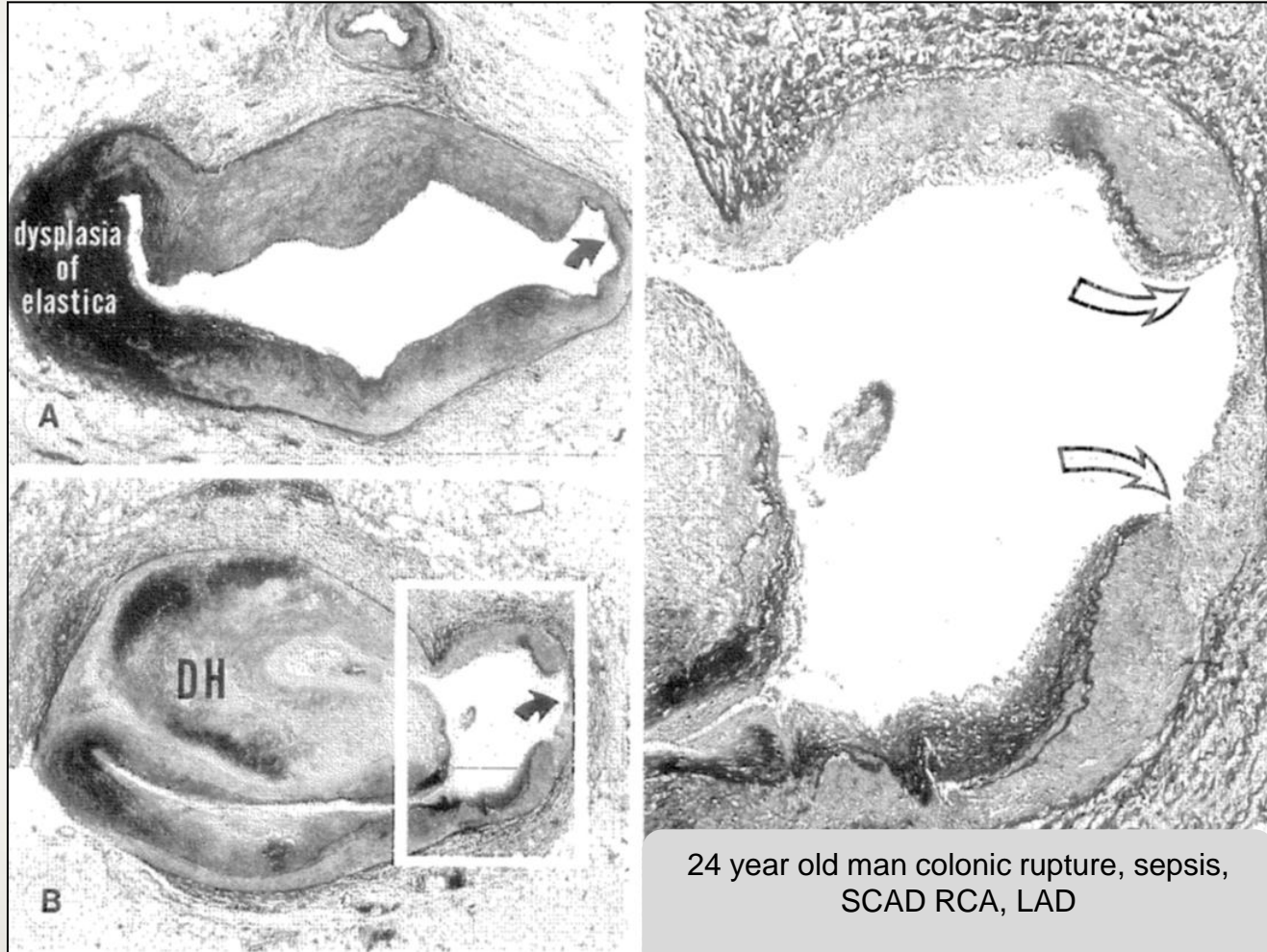
## US Cohort

- N=115
- 72% complete screening
  - 52% FMD
  - 34.8% cerebral imaging
    - Of those, 23% brain aneurysm

Circ Cardiovasc Interv. 2014;7:645-655

Am J Cardiol 2015: 115(12): 1672-7

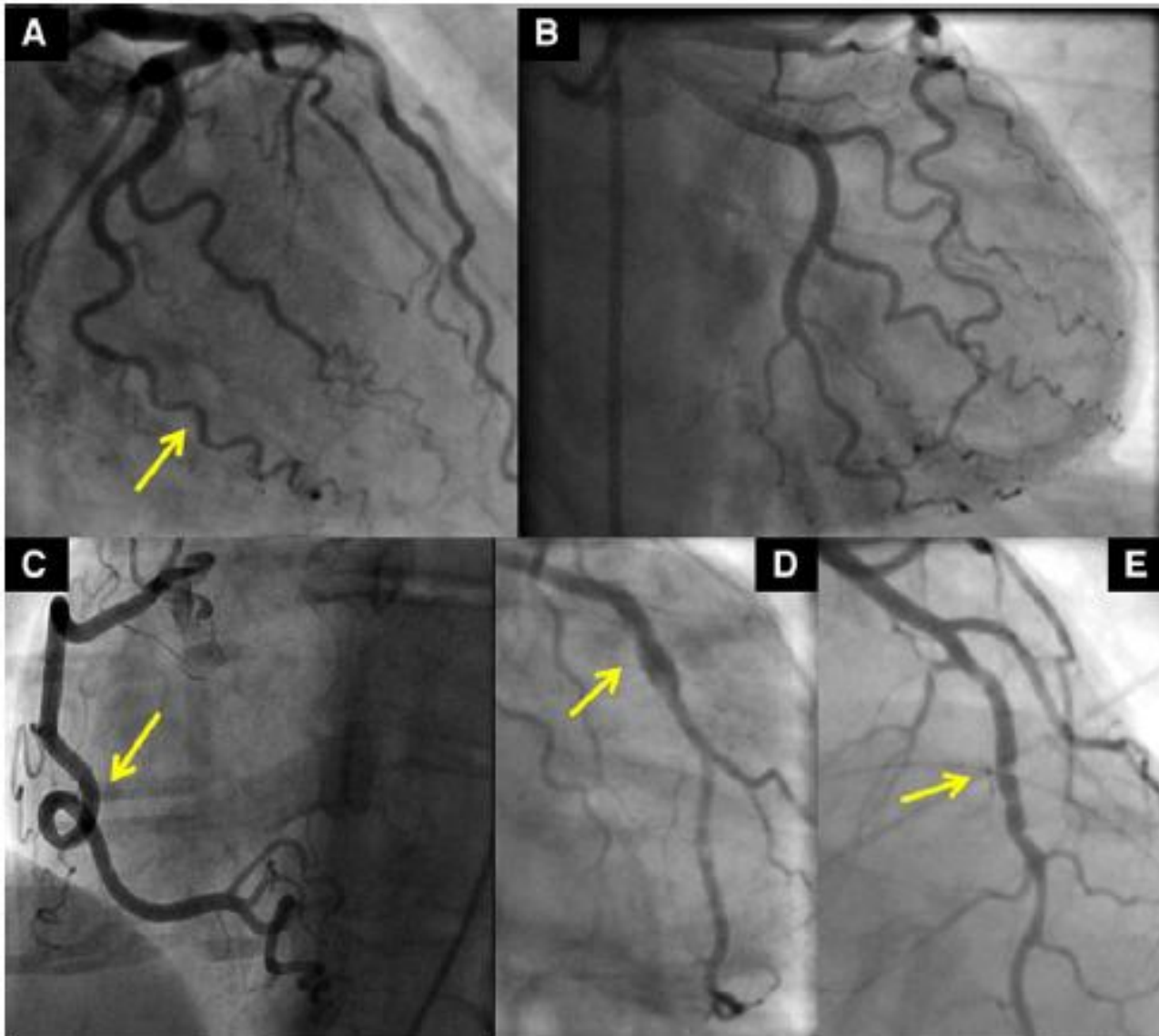
# SCAD Is A Manifestation of Coronary FMD



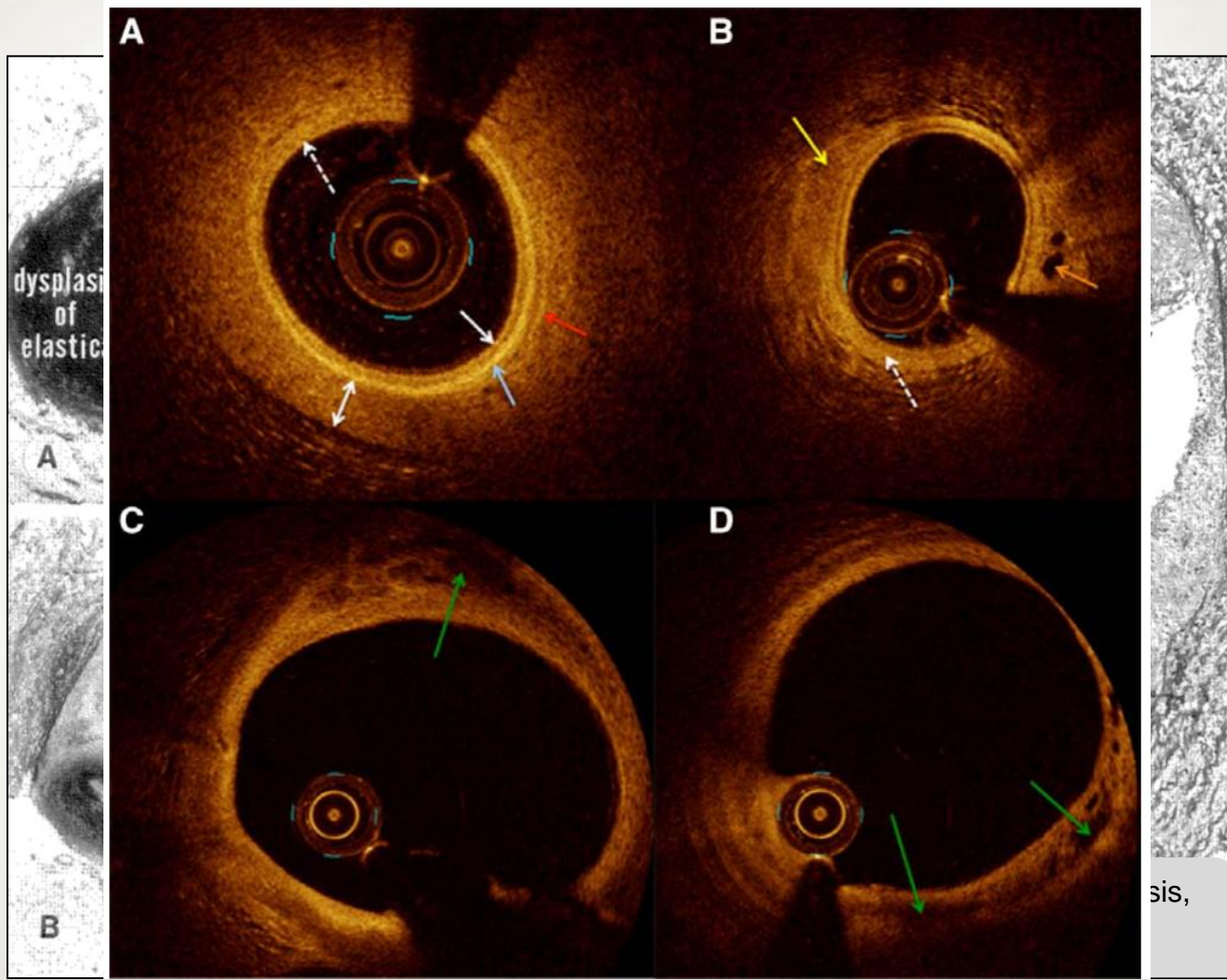
24 year old man colonic rupture, sepsis,  
SCAD RCA, LAD

Lie J.T. Hum Pathol 1987;18(6):654-6

# Can you see FMD in the Coronary Arteries?



# SCAD Is A Manifestation of Coronary FMD



Am J Cardiol 2015; 115(12): 1672-7  
Lie J.T. Hum Pathol 1987; 18(6):654-6



# Fibromuscular Dysplasia

- Non-inflammatory, non-atherosclerotic arteriopathy of unknown cause
- Manifests as arterial stenosis, aneurysm, dissection, arterial tortuosity
- Historically histopathologic diagnosis, currently primarily angiographic diagnosis



Stroke. 1982 Jan-Feb;13(1):46-52.

Int J Stroke. 2010 Aug;5(4):296-305.

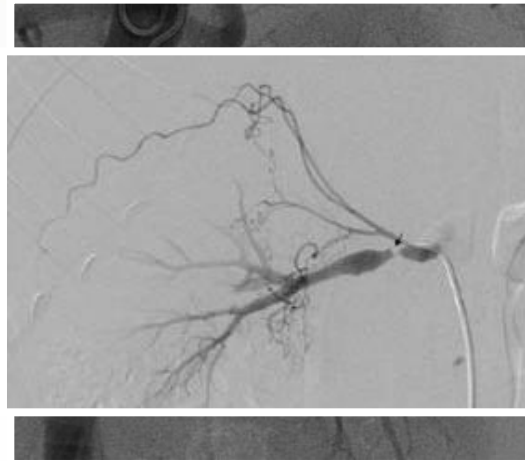
# FMD: Classification

**Multifocal**

Medial

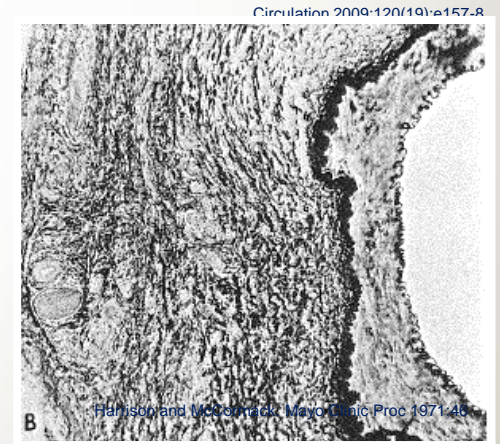
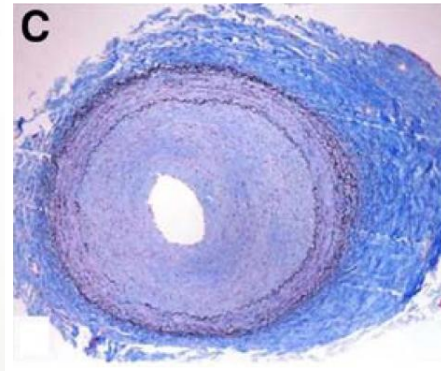
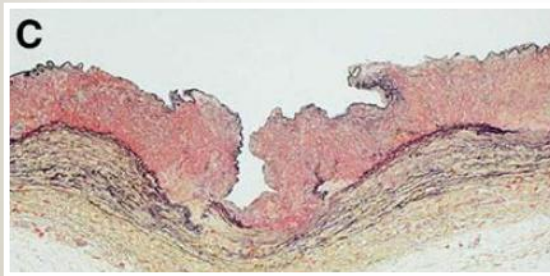
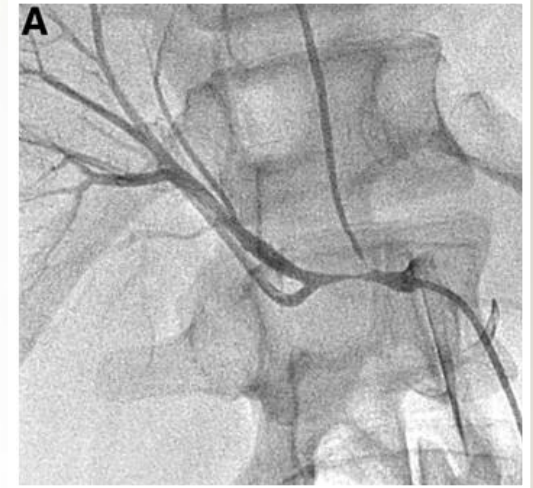


Intimal



**Focal**

Adventitial/Periarterial



Virmani R, Carter-Monroe N, Taylor AJ. Congenital anomalies and malformations of the vasculature. In: Creager MA, Beckman JA, Loscalzo J, eds. *Vascular Medicine: A Companion to Barunwald's Heart Disease*. 2nd ed. Philadelphia, PA: Elsevier Saunders; 2013.

Virmani R, Carter-Monroe N, Taylor AJ. Congenital anomalies and malformations of the vasculature. In: Creager MA, Beckman JA, Loscalzo J, eds. *Vascular Medicine: A Companion to Barunwald's Heart Disease*. 2nd ed. Philadelphia, PA: Elsevier Saunders; 2013.

# FMD is not an isolated arteriopathy

- Renal and carotid beds most commonly involved
  - 2/3 rule cervical/renal FMD
- Bruits are specific but not sensitive for detecting FMD (carotid bruit for FMD sens 45.4%, spec 93.7%)

## Vascular Distribution

Renal Artery	79.7%
Extracranial carotid	74.3%
Vertebral	36.6%
Mesenteric	26.3%
Lower extremity	60.0%
Intracranial carotid	17.0%
Upper extremity	15.9%
2 vascular beds	35.5%
3 vascular beds	21.9%
4 vascular beds	9.1%

Olin et al. Circulation 2012;125:3182.

# FMD Presentation

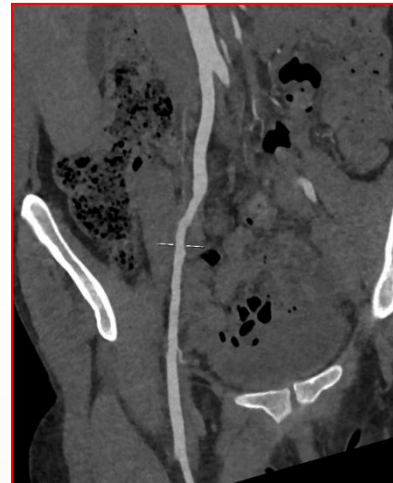
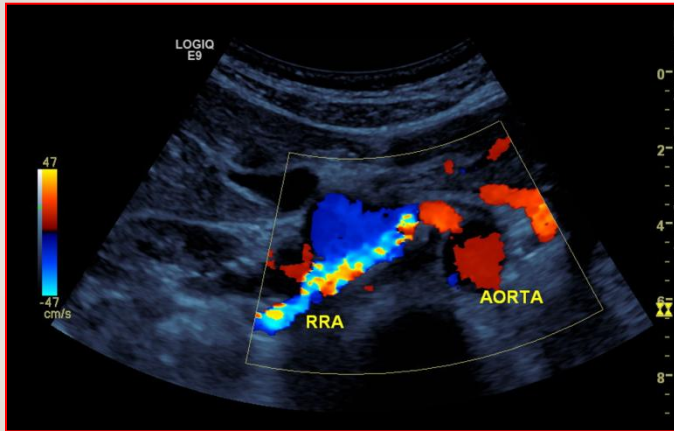
## Presenting symptoms/signs

- 9:1 F:M
- Mean age 55.7 ± 13.1 yrs
- Age range 18-86
- Mean 4.1 years delay in diagnosis
- Only 5.6% asymptomatic diagnosis
- Initial clinical presentation:
  - Hemispheric TIA (8.7%)
  - Stroke (6.9%)
  - Amaurosis fugax (5.2%)

Hypertension	63.8%
Headache	52.4%
Pulsatile tinnitus	27.5%
Dizziness	26.0%
Cervical bruit	22.2%
Neck pain	22.2%
Non-pulsatile tinnitus	18.8%
Chest pain/dyspnea	16.1%
Flank/abd pain	15.7%
Aneurysm	14.1%
Cervical dissection	12.1%

Olin et al. Circulation 2012;125:3182.

# 47 F HTN, headaches



Bilateral renal FMD, bilateral EIA FMD, R vertebral pseudoaneurysm

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Olin et al. Circulation 2012;125:3182.

# Aneurysm and dissection are not rare in patients with FMD

## Aneurysm

Renal	34%
Extracranial carotid	31%
Mesenteric/Celiac	13%
Intracranial	12.9%
Aorta	10%

## Dissection

Extracranial carotid	63.7%
Vertebral	20.7%
Renal Artery	11%
Mesenteric/Celiac	5.9%
Coronary	10.5%

21.7% of patients had at least 1

**41.7% of FMD patients in the Registry have had a dissection or an aneurysm**

patients had dissection

# FMD is a Morbid Disease...

- ~38% (348/913) have had  $\geq 1$  major vascular event(s) by enrollment
  - >50% renal procedures, endovascular
- 6.4% of procedures reported as technical failure
- 9.8% reported procedural complication
  - Arterial dissection most commonly reported procedural complication among Registrants

## Major Event at Enrollment

TIA	12.8%
Stroke	10.4%
Renal infarction	4.5%
SAH	2.4%
Mesenteric isch	1.6%
$\geq 1$ Therapeutic Vascular Procedure	50%



# What is the Etiology of SCAD?

Potential Risk Factors	Reported Prevalence among SCAD Patients
Fibromuscular Dysplasia	45-86%
Idiopathic/Unknown	20.8-44.8%
Hormonal Therapy	10-12.6% *
Multiparity ( $\geq 4$ births)	8.9-10% *
Pregnancy-related	12-40%, <5%
Connective Tissue Disorder	<<5% *
Systemic Inflammatory Disease	Case reports, 8.9% Vancouver cohort *inflammatory markers not routinely obtained
Precipitating Stressor Intense exercise, emotional stress, Valsalva, drugs	>50% report How much is too much, ?recall bias?

\* Prevalence in the general population?

Circulation

**AHA SCIENTIFIC STATEMENT**

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# Spontaneous Coronary Artery Dissection: Current State of the Science

A Scientific Statement From the American Heart Association

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**ABSTRACT:** Spontaneous coronary artery dissection (SCAD) has emerged as an important cause of acute coronary syndrome, myocardial infarction, and sudden death, particularly among young women and individuals with few conventional atherosclerotic risk factors. Patient-initiated research has spurred increased awareness of SCAD, and improved diagnostic capabilities and

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Sharonne N. Hayes, MD,  
FAHA, Chair  
Esther S.H. Kim, MD, MPH,  
FAHA, Co-Chair  
Jacqueline Saw, MD, FAHA,  
Co-Chair

Circulation 2018 Feb 22

# Life After SCAD

[Circulation](#)

**AHA SCIENTIFIC STATEMENT**

**Spontaneous Coronary Artery Dissection:  
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FAHA, Co-Chair  
Jacqueline Saw, MD, FAHA,  
Co-Chair

- **Chest pain** is common, with >60% of patient experiencing chest pain post MI
- **Anxiety and depression** among SCAD survivors is common
  - ~40% report hx depression/anxiety
  - ~33% are on meds/therapy for depression/anxiety after SCAD
- All patients post MI from SCAD should be referred for **cardiac rehabilitation**
  - *“The program should be tailored and individualized, taking into account not only cardiopulmonary factors such as ejection fraction but also patient age, pre-SCAD physical activity level, and patient-centered recovery goals”*

# Cardiac Rehab After SCAD

**Table 5. Research Priorities and Key Questions in SCAD**

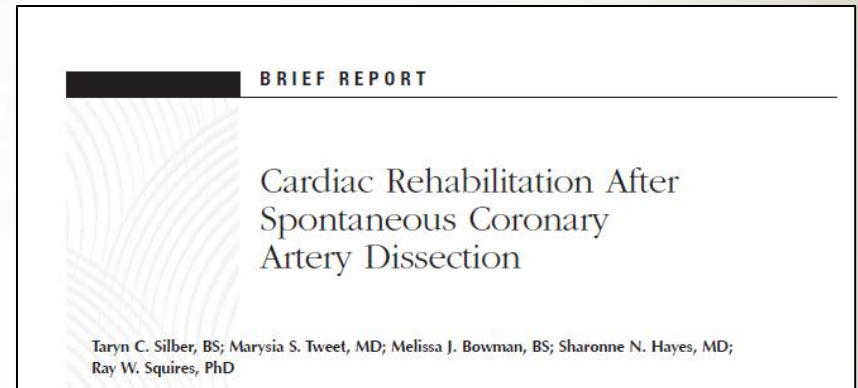
Epidemiology
What is the prevalence of SCAD in the general population?
Are there sex differences in the cause, presentation, diagnosis, and treatment of SCAD?
What is the recurrence rate of SCAD, and what are the determinants of recurrence?
Pathogenesis
Are there underlying genetic, hormonal, and environmental causes of SCAD, and what are the proportionate contributions of each?
What are the roles of physical and emotional stress and other potential triggers of SCAD?
What is the proportionate contribution of FMD and other systemic arteriopathies to the development and recurrence of SCAD?
Diagnosis
Under what circumstances and time frame and with what imaging method should follow-up coronary imaging be performed?
Treatment
What are the optimal approaches for initial revascularization?
What is the role of anticoagulation and antiplatelet therapy in SCAD immediately and after the event?
What are appropriate physical activity guidelines after SCAD?
What are the risks of exogenous hormone therapy and pregnancy after SCAD?

- Physical activity guidelines after SCAD do not exist
- Cardiac rehab represents the first step back to normal activity
  - Reassures patient
  - Reassures patient
- Few studies on cardiac rehab post SCAD exist

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# Cardiac Rehab After SCAD

- Retrospective study of 9 patients
- Standard CR, 1-2 weeks after d/c
- Participation mean 12.3 days, 28 sessions
- No cardiac sxs, no adverse events during exercise testing or training
- Results
  - Peak O2 uptake up 18%
  - 6 min walk distance up 22%
  - Mean body mass down 1.1 kg
  - Fat mass down 1.6 kg
  - Depression (PHQ-9) up 2.3 pts
  - Stress score down 1.3 pts



*Take home message →  
standard CR 1-2 weeks  
after SCAD is feasible and  
safe*

# Cardiac Rehab After SCAD

- VGH cohort, n=70
- SCAD-specific CR
  - 1 hr weekly
  - 15 min warm up, cool down
  - 30 min aerobic machines
  - Target HR 50-70% HRR
  - SBP <130 mmHg
  - Upper HR RPE “mod” to “somewhat difficult”
  - Resistance training 2-12 pound free weights, no >20 pounds
  - Peer-support group SCAD, counselling sessions

## Clinical Research

### The First Dedicated Cardiac Rehabilitation Program for Patients With Spontaneous Coronary Artery Dissection: Description and Initial Results

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**Table 1.** Description of the VGH SCAD-CR protocol

#### VGH SCAD-CR protocol

1. Weekly classes of SCAD survivors with targeted participation of 6 months
2. One-hour weekly exercise class consisting of 15-minute warm-up, 30-minute cardiovascular exercise on aerobic machines, and 15-minute cool-down
3. To reduce arterial shear stress, target exercise heart rate is 50%-70% of heart rate reserve on the basis of the entrance exercise treadmill test, and systolic blood pressure during exercise is limited to < 130 mm Hg
4. Exercise is adjusted to upper heart rate target to achieve rating of perceived exertion of “moderate” to “somewhat difficult”
5. Resistance training with 2- to 12-pound free weights to increase muscle strength, starting with lighter weights and progressing with strength gain
6. Patients are advised to avoid lifting weights > 20 pounds
7. A 20-minute educational session per week on heart-healthy nutrition, risk factors, and treatment of heart disease, and stress management, emphasizing women’s heart disease
8. Counselling, mindful living sessions, and peer-support from other SCAD survivors
9. Outside of formal sessions, participants can exercise in the supervised open gymnasium
10. Regular review of cardiac medications (eg, aspirin,  $\beta$ -blocker, angiotensin-converting enzyme inhibitor, statin) and coordination with the VGH SCAD clinic

# Cardiac Rehab After SCAD

**Table 5.** Comparison of chest pain and exercise METS at program start and exit

Variable	All patients (N = 70)	Completed 6 months (n = 28)	Completed $\geq$ 1 month (n = 48)
CP			
CP at program start	44 (62.9)	20 (71.4)	31 (64.6)
CP on program exit	26 (37.1)	8 (28.6)	15 (31.3)
<i>P</i> (start vs exit)	< 0.001	0.034	< 0.001
ETT			
ETT at program start: METS	10.1 $\pm$ 3.3	9.8 $\pm$ 3.4	9.9 $\pm$ 3.3
ETT on program exit: METS	11.5 $\pm$ 3.5	11.1 $\pm$ 3.7	11.5 $\pm$ 3.5
<i>P</i> (start vs exit)	< 0.001	< 0.001	< 0.001

Data are presented as mean  $\pm$  SD, or n (%), except where otherwise stated.

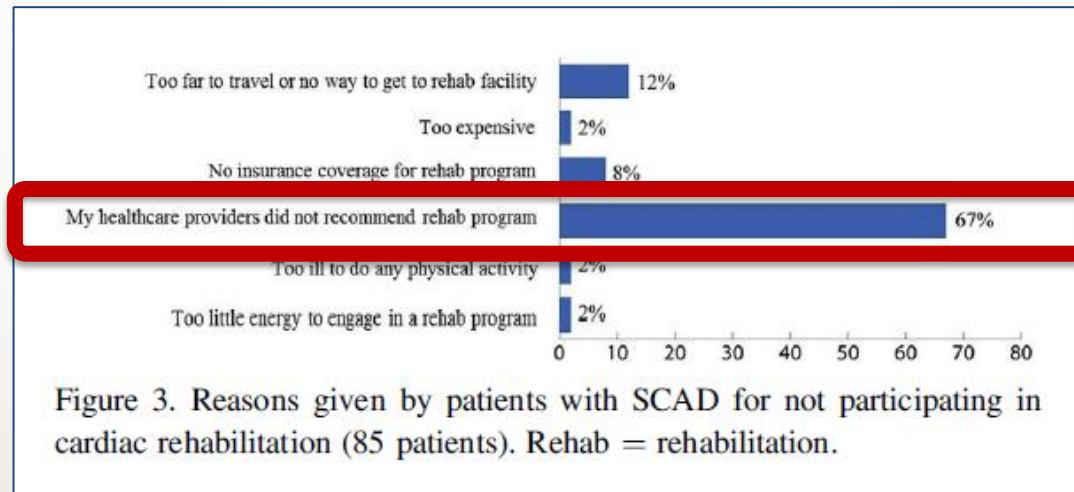
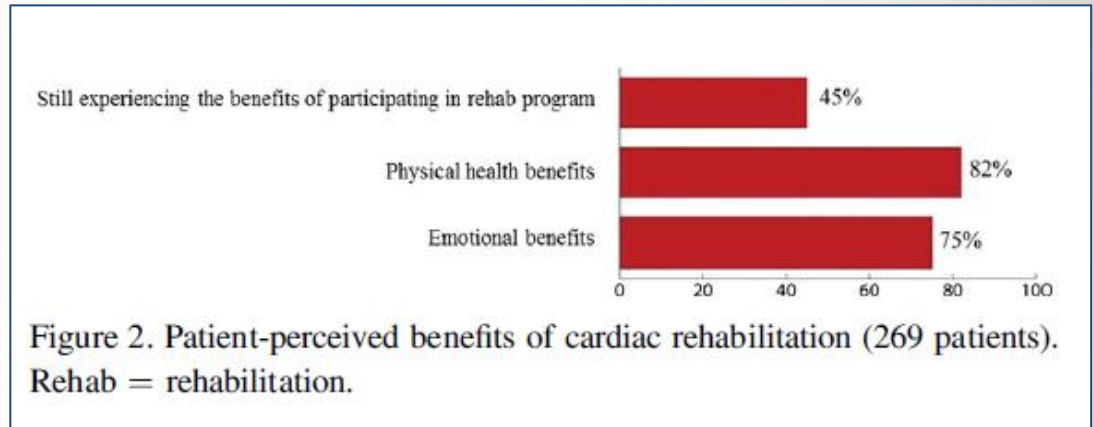
CP, chest pain; ETT, exercise treadmill test, METS, metabolic equivalents.

- Participation mean 12.4 +/- 10.4 wks
- Chest pain improved
- METs improved
- Improvement trend in anxiety/depression

*Take home message → modified CR can improve chest pain and EC. There is a “basement” of safety*

# Cardiac Rehab After SCAD

- Mayo virtual registry
- n=354
- 76%  $\geq 1$  CR session
- 66%  $>10$  CR session
- Average 18 +/- 12 sessions





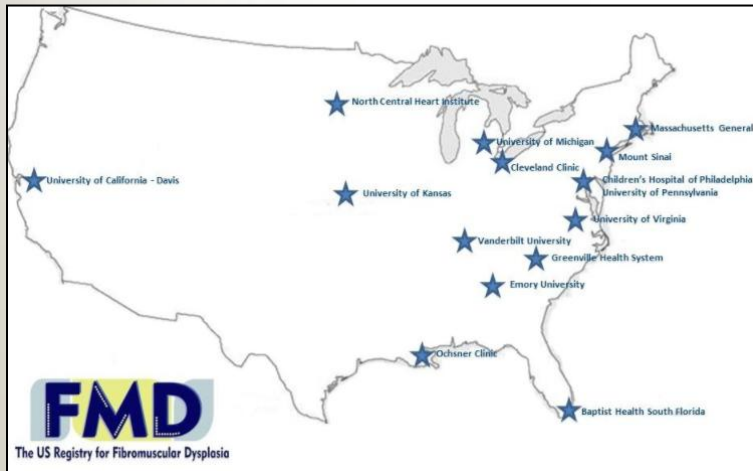
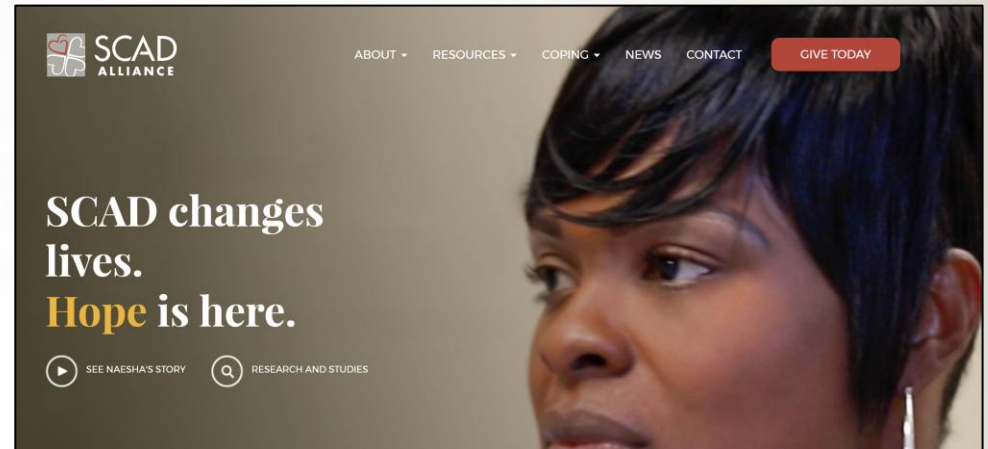
# Patient Perspectives to Consider

- “I found it too slow for me and left the program”
- “It was a waste of my time”
- “My own workout routine was more than the rehab program”
- “I tried going and did not think it was worthwhile”
- “Don’t have clear guidelines/feel at risk.”

# Life After SCAD

- Activity/Dissection Precautions
  - Avoid: heavy lifting, roller coasters, etc
  - Others depending on other arterial involvement
  - Rethink competitive sports

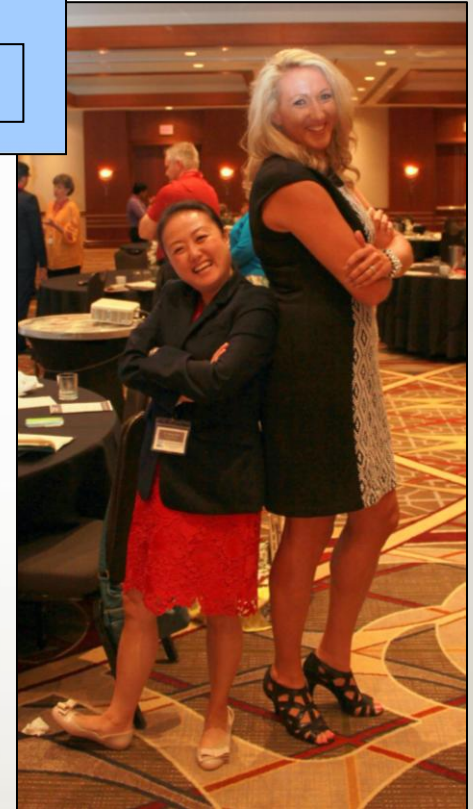
# SCAD/FMD Resources and Research



# SCAD PATIENT SYMPOSIUM

VANDERBILT HEART AND VASCULAR INSTITUTE

# 2017



# Summary

- SCAD is rare, but perhaps underdiagnosed
- Accurate diagnosis is critical for optimal treatment and further clinical investigations
- More than half the time, SCAD is not an isolated disease
- Cardiac rehab post-SCAD is safe and beneficial, but SCAD-specific protocols need to be developed and validated
- Be prepared to provide the patient with basic answers and resources to help them cope

Thank you