Spontaneous Coronary Artery Dissection

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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
SCAD Epidemiology

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

Who gets SCAD?
Middle-aged, white women with few traditional CV risk factors

<table>
<thead>
<tr>
<th>Canada (n=327)</th>
<th>United States (n=189)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Age 52.5 +/- 9.6</td>
<td>44.9 +/- 9</td>
</tr>
<tr>
<td>Sex 90.8%</td>
<td>92%</td>
</tr>
<tr>
<td>Race (White) 82.0%</td>
<td>95%</td>
</tr>
<tr>
<td>BMI 24.4 (21.5 - 29.3)</td>
<td>26 +/- 6</td>
</tr>
<tr>
<td>HPL 25.7%</td>
<td>31%</td>
</tr>
<tr>
<td>HTN 36.4%</td>
<td>22%</td>
</tr>
<tr>
<td>Smoking 9.8%</td>
<td>15%</td>
</tr>
</tbody>
</table>

Katherine Leon
Founder, SCAD Alliance

<table>
<thead>
<tr>
<th>Smoking</th>
</tr>
</thead>
<tbody>
<tr>
<td>32.8%</td>
</tr>
<tr>
<td>51.4%</td>
</tr>
<tr>
<td>34.3%</td>
</tr>
</tbody>
</table>


JACC 2017;70:1148-58
Am J Cardiol 2015;116:66-73
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

Type 1 SCAD

17 months later
Acute Management of SCAD

• SCAD ≠ 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

- 79-90% of patients with ACS from SCAD have angiographic "healing" weeks to months post-SCAD
- 7-10% day recurrent MI rate (extension of dissection), some requiring emergency revascularization
- Extended monitoring in the hospital (3-5 days)
Acute Management of SCAD

Management of acute spontaneous coronary artery dissection (SCAD)

Clinically stable, no high-risk anatomy
- Conservative therapy
- Monitor as inpatient 3-5 days

Clinically stable with left main or severe proximal 2-vessel dissection
- Consider CABG
- Conservative Rx may be reasonable (not studied)

Active/ongoing ischemia or hemodynamic instability
- Consider PCI if feasible OR
- Urgent CABG (based on technical considerations and local expertise)
Acute Management of SCAD

SCAD

No high risk features

Medical Management

Anatomic Criteria

Single vessel Mid/distal lesions

Medical Management or PCI

Left Main Proximal 2-vessel

CABG or Medical Management

Clinical Criteria

Ongoing ischemia/ Hemodynamic instability

PCI or Urgent CABG

Stable

Medical Management
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
## Short Term Outcomes

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

- 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 $\rightarrow$ malapposition of stents increasing risk of ISR and stent thrombosis

*Circ Cardiovasc Interv. 2014;7:777-786*
Recurrence
(not to be confused with extension)

Mayo Series (n=189)

27% over 5 years


Vancouver Series (n=327)

10.4% over 3.1 years

JACC 2017;70:1148-58
Patient Presentation

- 45 year old woman with no significant cardiac history
- Acute onset chest pain at work
- NSTEMI $\rightarrow$ 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

<table>
<thead>
<tr>
<th>Clinical Questions</th>
<th>Root Questions</th>
</tr>
</thead>
<tbody>
<tr>
<td>• Are further evaluations necessary</td>
<td>• Is SCAD a disease in isolation?</td>
</tr>
<tr>
<td>• What history should I obtain?</td>
<td>• What is the etiology of SCAD?</td>
</tr>
<tr>
<td>• What physical exam findings should I be looking for?</td>
<td>• How do I diagnose extracoronary</td>
</tr>
<tr>
<td>• What laboratory tests should I order?</td>
<td>abnormalities?</td>
</tr>
<tr>
<td>• What imaging tests do I order?</td>
<td>• Is SCAD a genetic disorder?</td>
</tr>
<tr>
<td>• Do I need to consult genetics?</td>
<td></td>
</tr>
</tbody>
</table>
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

### 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
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

FMD: Classification

Multifocal

Medial

Intimal

Adventitial/Periarterial

Focal


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

<table>
<thead>
<tr>
<th>Vascular Bed</th>
<th>Percentage</th>
</tr>
</thead>
<tbody>
<tr>
<td>Renal Artery</td>
<td>79.7%</td>
</tr>
<tr>
<td>Extracranial carotid</td>
<td>74.3%</td>
</tr>
<tr>
<td>Vertebral</td>
<td>36.6%</td>
</tr>
<tr>
<td>Mesenteric</td>
<td>26.3%</td>
</tr>
<tr>
<td>Lower extremity</td>
<td>60.0%</td>
</tr>
<tr>
<td>Intracranial carotid</td>
<td>17.0%</td>
</tr>
<tr>
<td>Upper extremity</td>
<td>15.9%</td>
</tr>
<tr>
<td>2 vascular beds</td>
<td>35.5%</td>
</tr>
<tr>
<td>3 vascular beds</td>
<td>21.9%</td>
</tr>
<tr>
<td>4 vascular beds</td>
<td>9.1%</td>
</tr>
</tbody>
</table>

FMD Presentation

- 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%)

Presenting symptoms/signs

- 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%

47 F HTN, headaches

Bilateral renal FMD, bilateral EIA FMD, R vertebral pseudoaneurysm
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%)

Aneurysm and dissection are not rare in patients with FMD

<table>
<thead>
<tr>
<th>Aneurysm</th>
<th>Dissection</th>
</tr>
</thead>
<tbody>
<tr>
<td>Renal</td>
<td>Extracranial carotid</td>
</tr>
<tr>
<td>Extracranial carotid</td>
<td>Vertebral</td>
</tr>
<tr>
<td>Mesenteric/Celiac</td>
<td>Renal Artery</td>
</tr>
<tr>
<td>Intracranial</td>
<td>Mesenteric/Celiac</td>
</tr>
<tr>
<td>Aorta</td>
<td>Coronary</td>
</tr>
</tbody>
</table>

21.7% of patients had at least 1 aneurysm

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

JACC 2016;68:176–85
JAMA Neurology 2017; Jul 17
FMD is a Morbid Disease...

- ~38% (348/913) have had ≥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

<table>
<thead>
<tr>
<th>Major Event at Enrollment</th>
<th>%</th>
</tr>
</thead>
<tbody>
<tr>
<td>TIA</td>
<td>12.8%</td>
</tr>
<tr>
<td>Stroke</td>
<td>10.4%</td>
</tr>
<tr>
<td>Renal infarction</td>
<td>4.5%</td>
</tr>
<tr>
<td>SAH</td>
<td>2.4%</td>
</tr>
<tr>
<td>Mesenteric isch</td>
<td>1.6%</td>
</tr>
</tbody>
</table>

≥1 Therapeutic Vascular Procedure 50%

O’Connor et al.  Oral Abstract SVM 2014
Gornik HG et al.  Oral Abstract ACC 2011
## What is the Etiology of SCAD?

<table>
<thead>
<tr>
<th>Potential Risk Factors</th>
<th>Reported Prevalence among SCAD Patients</th>
</tr>
</thead>
<tbody>
<tr>
<td>Fibromuscular Dysplasia</td>
<td>45-86%</td>
</tr>
<tr>
<td>Idiopathic/Unknown</td>
<td>20.8-44.8%</td>
</tr>
<tr>
<td>Hormonal Therapy</td>
<td>10-12.6% *</td>
</tr>
<tr>
<td>Multiparity (≥4 births)</td>
<td>8.9-10% *</td>
</tr>
<tr>
<td>Pregnancy-related</td>
<td>12-40%, &lt;5%</td>
</tr>
<tr>
<td>Connective Tissue Disorder</td>
<td>&lt;=5% *</td>
</tr>
<tr>
<td>Systemic Inflammatory Disease</td>
<td>Case reports, 8.9% Vancouver cohort</td>
</tr>
<tr>
<td></td>
<td>* inflammatory markers not routinely obtained</td>
</tr>
<tr>
<td>Precipitating Stressor</td>
<td>&gt;50% report</td>
</tr>
<tr>
<td></td>
<td>How much is too much, ?recall bias?</td>
</tr>
</tbody>
</table>

*Prevalence in the general population?*
AHA SCIENTIFIC STATEMENT

Spontaneous Coronary Artery Dissection: Current State of the Science
A Scientific Statement From the American Heart Association

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 findings of underlying syndromes led to advances in understanding and treatment.

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

• **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

- 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

Table 5. Research Priorities and Key Questions in SCAD

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

Circulation 2018 Feb 22
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
Cardiac Rehab After SCAD

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

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

<table>
<thead>
<tr>
<th>Variable</th>
<th>All patients (N = 70)</th>
<th>Completed 6 months (n = 28)</th>
<th>Completed ≥ 1 month (n = 48)</th>
</tr>
</thead>
<tbody>
<tr>
<td>CP at program start</td>
<td>44 (62.9)</td>
<td>20 (71.4)</td>
<td>31 (64.6)</td>
</tr>
<tr>
<td>CP on program exit</td>
<td>26 (37.1)</td>
<td>8 (28.6)</td>
<td>15 (31.3)</td>
</tr>
<tr>
<td>( P ) (start vs exit)</td>
<td>&lt; 0.001</td>
<td>0.034</td>
<td>&lt; 0.001</td>
</tr>
<tr>
<td>ETT at program start: METS</td>
<td>10.1 ± 3.3</td>
<td>9.8 ± 3.4</td>
<td>9.9 ± 3.3</td>
</tr>
<tr>
<td>ETT on program exit: METS</td>
<td>11.5 ± 3.5</td>
<td>11.1 ± 3.7</td>
<td>11.5 ± 3.5</td>
</tr>
<tr>
<td>( P ) (start vs exit)</td>
<td>&lt; 0.001</td>
<td>&lt; 0.001</td>
<td>&lt; 0.001</td>
</tr>
</tbody>
</table>

Data are presented as mean ± SD, or n (%), except where otherwise stated.
CP, chest pain; ETT, exercise treadmill test; METS, metabolic equivalents.

*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% ≥1 CR session
- 66% >10 CR session
- Average 18 +/- 12 sessions

![Figure 2. Patient-perceived benefits of cardiac rehabilitation (269 patients). Rehab = rehabilitation.](image)

![Figure 3. Reasons given by patients with SCAD for not participating in cardiac rehabilitation (85 patients). Rehab = rehabilitation.](image)

Am J Cardiol 2016;117:1604-1609
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
80 participants from 14 states
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