MHIF FEATURED STUDY: HITSOVA

OBJECTIVE:
Prove that danaparoid use is not inferior to argatroban in terms of efficacy in HIT

ENDPOINTS:
1. Approximately Day 14 end study medication
2. Subject considered a treatment responder if none of the following occur @D44
   • New or extended venous and/or arterial thrombosis, including gangrene/skin necrosis
   • All-cause mortality
   • Unplanned amputation, including ischemic gut resection

CONDITION: Heparin Induced Thrombocytopenia
PI: Nedaa Skeik, MD

RESEARCH CONTACTS:
Carina Benson - carina.benson@allina.com 612-863-4393 | pager: 612-654-5542
Jane Fox - jane.fox@allina.com | 612-863-6289

SPONSOR: Aspen Global, Inc.

OPEN AND ENROLLING: Please Refer Patients!
MHIF FEATURED STUDY:
HITSOVA

CONDITION:
Heparin Induced Thrombocytopenia

PI:
Nedaa Skeik, MD

DESCRIPTION:
Open-Label, Randomized, Active Controlled, Multi-Centre Phase 3 Study to Evaluate the Safety and Efficacy of Danaparoid vs Argatroban in Treatment of Subjects with Acute HIT (HITSOVA study)

CONDITION:
Heparin Induced Thrombocytopenia

PI:
Nedaa Skeik, MD

RESEARCH CONTACTS:
Carina Benson: carina.benson@allina.com | 612-863-4393 | pager: 612-654-5542
Jane Fox: jane.fox@allina.com | 612-863-6289

SPONSOR:
Aspen Global, Inc.

OPEN AND ENROLLING:
Please Refer Patients!

EXCLUSION
• Cardiac surgery within 44 days
• Intra-aortic balloon pump or VAD
• Life expectancy less than study duration of 44 days
• Spinal/epidural access within past 48 hrs
• Severe hepatic impairment (Child-Pugh Class C)
• Active bleeding
• Hemorrhagic cerebrovascular accident within previous 3 mos.
• Severe, uncontrolled hypertension (>180/110 mmHg)
• Long-term (>3wks) HD or continuous renal replacement

CRITERIA LIST/ QUALIFICATIONS:

Inclusion
Males or females aged ≥2 weeks
Subjects with suspected HIT by 4Ts of >3 and with reduction of platelet count of ≥ 30% at either:
 a) Between Day 4 and 14 of the start of heparin exposure OR
 b) At Day 1 of heparin exposure with pre-treatment with heparin within the last 30 days

Have adequate renal function: Glomerular filtration rate ≥ 15 mL/min/1.73 m²
Minneapolis Heart Institute Foundation® Cardiovascular Grand Rounds

Title: Non-Atherosclerotic Vasculopathies
Speaker: Nedaa Skeik, MD, FACC, FACP, FSVM
Section Head, Vascular Medicine
Medical Director, Vein Center
Medical Director, Anticoagulation and Thrombophilia Clinic
Medical Director, Vascular Lab
Minneapolis Heart Institute® at Abbott Northwestern Hospital

Date: January 13, 2020
Time: 7:00 - 8:00 AM
Location: Minneapolis Heart Institute Building, Suite 100, Learning Center

OBJECTIVES
At the completion of this activity, the participants should be able to:
1. Identify abdominal non-atherosclerotic arterioapthies
2. Analyze different underlying etiologies
3. Determine differential diagnoses and management planning

DISCLOSURE POLICY & STATEMENTS
Allina Health, Learning & Development intends to provide balance, independence, objectivity and scientific rigor in all of its sponsored educational activities. All speakers and planning committee members participating in sponsored activities and their spouse/partner are required to disclose to the activity audience any real or apparent conflict(s) of interest related to the content of this conference.

The ACCME defines a commercial interest as “any entity” producing, marketing, re-selling, or distributing health care goods or services consumed by, or used on, patients. The ACCME does not consider providers of clinical service directly to patients to be commercial interests - unless the provider of clinical service is owned, or controlled by, an ACCME-defined commercial interest.

Moderator(s)/Speaker(s)
Dr. Nedaa Skeik has disclosed the following commercial interests: Consultant- Boston Scientific; Speaker’s Bureau- Janssen, Boehringer Ingelheim, Pfizer, Bristol-Myers Squibb

Planning Committee
Dr. Alex Campbell, Jake Cohen, Jane Fox, Dr. Kevin Harris, Dr. Kasia Hryniewicz, Rebecca Lindberg, Amy McMeans, Dr. Michael Miedema, Dr. JoEllyn Moore, Pamela Morley, Dr. Scott Sharkey, Maia Hendel and Jolene Bell Makowesky have disclosed that they DO NOT have any real or apparent conflicts with any commercial interest as it relates to the planning of this activity/course. Dr. Mario Gössl has disclosed the following relationships - Grant/Research Support: Edwards Life Sciences; Consultant: Abbott Vascular, Caisson; Speaker’s Bureau: Edwards Lifesciences. Dr. David Hurrell has disclosed the following relationship -Chair, Clinical Events Committee: Boston Scientific. Dr. João Cavalcante has disclosed the following relationships -Grant/Research Support: Boston Scientific, Medtronic, Abbott Vascular, Circle Cardiovascular Imaging, Siemens Healthineers; Consultant: Boston Scientific, Medtronic; Speaker's Bureau: Medtronic, Siemens Healthineers; Honoraria: Medtronic, Siemens Healthineers.
NON-ENDORSEMENT OF COMMERCIAL PRODUCTS AND/OR SERVICES

We would like to thank the following company for exhibiting at our activity.

Amgen

Boehringer Ingelheim

Accreditation of this educational activity by Allina Health does not imply endorsement by Allina Learning & Development of any commercial products displayed in conjunction with an activity.

A reminder for Allina employees and staff, the Allina Policy on Ethical Relationship with Industry prohibits taking back to your place of work, any items received at this activity with branded and or product information from our exhibitors.
Non-Atherosclerotic Abdominal Vasculopathies

Nedaa Skeik, MD, FACC, FACP, FSVM, RPVI
Associate Professor of Medicine
Section Head, Vascular Medicine
Medical Director, Thrombophilia/Anticoagulation Clinic
Medical Director, Vein Practice
Medical Director, Vascular Laboratory
Minneapolis Heart Institute® Abbott Northwestern Hospital

Disclosures

Consulting and speaking for Pfizer, BMS, J&J, B.I. and BSC:
Not relevant to this talk

No financial conflict related to this talk

Cases were modified for education purposes
Learning Objectives

- Discuss different non-atherosclerotic abdominal vasculopathies, case presentations
- Cover underlying etiologies (e.g. FMD, SAM, connective tissue disorders, vasculitis,..)
- Provide some tools to help with differential diagnoses
- Summarize management planning and outcome
- Share our center experience

Background

- **Problem:** Mesenteric and or renal artery dissection, aneurysm, stenosis, thrombosis or vasculitis
- **Epidemiology:** Very rare with great clinical significance. Very scarce literature!
- **Presentation:** Incidental → arterial rupture and life-threatening bleed!
- **Diagnosis/Management:**
  - No consensus on diagnostic process, management or follow up strategy
  - Hard to make a differential diagnosis (e.g. FMD, SAM, vasculitis,..)
  - Management can be very challenging
- **Outcome:** Varies based on underlying etiology
Etiologies

• Fibromuscular dysplasia (FMD)
• Segmental arterial mediolysis (SAM)
• Connective tissue disorders (vEDS, LDS, Marfan)
• Localized vasculitis of the GI tract (LGVT) or systemic vasculitis (e.g. PAN, ANCA-vasculitis)
• Median arcuate ligament syndrome (MALS)
• Trauma
• Isolated disease (dissection or aneurysm)

Case 1

• Patient: 74 y/o F
• PMH: CAD (remote history of MI) and uncontrolled HTN
• Presentation: abdominal and neck pain
• Family history: 3 sisters and one daughter with history of irregular vessels!
• VS: BP: 160/90 mmHg P: 80/min regular
• Exam: right side carotid bruit and mild epigastric tenderness
• Labs: ESR: 35 and CRP: 2.7
CTA of Abdomen/Pelvis

CTA of Neck
Diagnosis?

- A- FMD
- B- SAM
- C- vEDS
- D- LDS
- E- Vasculitis

Diagnosis: Fibromuscular Dysplasia (FMD)

- Meets criteria by *First international Consensus on the Diagnosis and Management of FMD*:
  - Focal or multifocal FMD lesion (alternating stenosis and dilation) in one vascular bed +/- aneurysm, dissection, or tortuosity in another vascular bed
  - Age and gender
  - Beading appearance and history of SCAD
  - No significant wall thickening or elevated ESR/CRP
  - Genetic testing was negative for connective tissue disorders

- Less likely SAM: biopsy not feasible!

- Management: ASA, good BP control, and imaging surveillance.
Case 2

- **Patient:** 47 y/o M
- **PMH:** None
- **Presentation:** neck pain, headache followed by abdominal pain
- **Family history:** No relevant history
- **VS:** BP: 170/95 mmHg, P: 105/min regular
- **Exam:** Neck and diffuse abdominal tenderness
- **Labs:** ESR: 27 and CRP: 4.54

---

**CTA of Abdomen/Pelvis**
CTA Abdomen/Pelvis

CTA Neck
Diagnosis?

- A- FMD
- B- SAM
- C- vEDS
- D- LDS
- E- Vasculitis

Diagnosis: Segmental Arterial Mediolysis (SAM)

- Dissection and or an aneurysm with or without organ infarction in multiple mesenteric and or renal arteries with exclusion of other vasculopathies (e.g. FMD), and no significant concurrent arterial wall thickening (< 3mm) or inflammatory markers' elevation

- Biopsy: lysis of the outer arterial media resulting in dissection and pseudoaneurysm

- Age and gender
- Dramatic presentation
- Genetic testing was negative for connective tissue disorders
- No significant wall thickening or significant ESR/CRP elevation

- Management: hematoma evacuation, blood pressure control, ASA, and imaging surveillance.

Case 3

- **Patient:** 38 y/o M
- **PMH:** Tobacco abuse
- **Family History:** Not relevant history
- **Presentation:** Abdominal pain, nausea and vomiting
- **VS:** BP: 145/85 mmHg, P: 86/min regular
- **Exam:** Generalized abdominal tenderness
- **Labs:** ESR: 14 and CRP 2.23

CTA of Abdomen and Pelvis
Diagnosis?

- A - FMD
- B - SAM
- C - vEDS
- D - LDS
- E - Vasculitis

Diagnosis: Vascular Ehlers Danlos Syndrome (vEDS)

- Genetic testing: *novel frameshift* variant in COL3A1
- One major criteria and positive genetic testing

- Younger patient
- Multiple arterial involvement and pathologies: aneurysms and dissection
- No significant inflammatory markers elevation
- *No suggestive family history!*
- *No connective tissue manifestations!*

- Management: ASA, BP control and imaging surveillance. Celiprolol !!
Case 4

- **Patient:** 50 y/o F
- **PMHX:** Obesity, HTN, and tobacco abuse
- **Family History:** No relevant history
- **Presentation:** Abdominal pain
- **VS:** BP: 156/87 mmHg, P: 86/min regular
- **Exam:** Severe epigastric tenderness
- **Labs:** ESR 48 and CRP: 22.67

CTA of Abdomen/Pelvis
Diagnosis: Isolated Vasculitis of the GI Tract (LVGT)

- Significant wall thickening (> 3mm) and elevated ESR/CRP
- Condition worsened within 3 weeks on ASA only
- Improved significantly on prednisone for 4 weeks

- No rheumatological symptoms
- Autoimmune work up was negative
- Negative genetic testing for connective tissue disorder
- No other arteriopathies
- Tissue biopsy was not feasible!

- Management: Prednisone taper, ASA and imaging surveillance.
Case 5

- **Patient:** 63 y/o M
- **PMH:** of AVR (St. Jude mechanical), DM, HTN, HL
- **Presentation:** Abdominal pain and nausea
- **Family history:** No relevant history
- **VS:** BP: 171/99 mmHg, P: 100/min regular
- **Exam:** mechanical S2, no murmur. Abdominal general tenderness
- **Labs:** WBC: 23,300 Hgb: 15.5, and PLT: 580,000. Over 2 years, INR levels (2.1 - 4.4)
Diagnosis?

- A - Antiphospholipid syndrome
- B - HIT
- C - Essential thrombocythemia
- D - Malignancy
- E - Aortic Vegetation

Diagnosis: Essential Thrombocythemia

- Elevated PLT
- Positive JAK 2 mutation
- Unremarkable TEE
- Other malignancy work up was negative
- No recent heparin exposure
- Negative antiphospholipid syndrome work up

Management: ASA, warfarin and hydroxyurea with resolution of the thrombi
Case 6

- **Patient:** 51 y/o F
- **PMHX:** none
- **Presentation:** Postprandial epigastric abdominal pain with nausea and weight loss (20 pounds in 1 year)
- **Family history:** No relevant history
- **VS:** Normal
- **Exam:** Epigastric tenderness with epigastric bruit with expiration
- **Labs:** CBC, CMP, LFT's, lipase were normal

CTA of Abdomen/Pelvis
Diagnosis?

- A- FMD
- B- SAM
- C- vEDS
- D- Median Arcuate Ligament Syndrome (MALS)
- E- Vasculitis

Diagnosis: Median Arcuate Ligament Syndrome

- Typical symptoms and images
- No other explanation
- Negative work up: EGD, colonoscopy, celiac, depression, ..
- Management:
  - Ganglion block: good response
  - Median arcuate ligament release
  - ASA

Differential Diagnoses!

Non-atherosclerotic Abdominal Vasculopathies

Single Center Retrospective Analysis of Patients: (Age: 18-60)
Presented with NAV Between 01/2000 and 12/2015

• Pathologies:
  - Arterial stenosis
  - Wall thickening
  - Dissection
  - Aneurysm
  - Pseudoaneurysm

• Arterial Involvement:
  - Renal
  - Celiac
  - Hepatic
  - Splenic
  - Mesenteric arteries

Data Collection

- **Demographics**: date of birth, gender
- **Comorbid Conditions**: DM, HTN, HL, current or history of tobacco use, PAD, and CAD
- **Presenting symptoms**
- **Physical Exam**
- **Medications**
- **Laboratory data and diagnostic imaging**: at initial presentation and at follow-up visits
Fibromuscular Dysplasia (FMD)

- Non-atherosclerotic arterial disease with abnormal cellular proliferation and wall architecture.
- Primarily manifests as beaded (multifocal) or focal lesions in medium or small-sized arteries.
- Phenotype now expanded to include arterial dissection, aneurysm, and tortuosity.
- Most commonly affects the renal and extracranial carotid and vertebral arteries, but nearly all arterial beds may be affected, and multi-vessel involvement is common.
- 90% female predominance!

**Diagnostic Criteria:**
- Stenosis, occlusion, dissection, or aneurysm in the renal and/or extracranial cerebrovascular arteries with concurrent string of beading appearance (multifocal FMD) or circumferential stenosis (focal FMD) in the setting of no aortic involvement and no significant inflammatory markers' elevation.
Segmental Arterial Mediolysis (SAM)

- Rare but serious non-atherosclerotic, noninflammatory vasculopathy of unknown etiology.
- Characterized by vacuolar degeneration and lysis of the medial layer, often resulting in dissection, aneurysm, occlusion, or stenosis.
- Commonly affects the abdominal aortic branches, such as the celiac, mesenteric, and/or renal arteries with occasional carotid, cerebral, and coronary artery involvement.
- Slightly more common in males!

**Diagnostic Criteria:**
- Dissection and or an aneurysm with or without organ infarction in *multiple* mesenteric and or renal arteries with exclusion of connective tissue disorders, FMD, or other vasculopathies, and no significant concurrent arterial wall thickening (< 3mm) or inflammatory markers' elevation.


Localized Vasculitis of the GI Tract (LVGIT)

- Single organ vasculitis of the GI tract.
- Imaging: wall thickening +/- stenosis, occlusion or aneurysm of GI arteries.
- Diagnosis: symptoms + images +/- path. Always presumptive and requires exclusion of systemic illness at the time of diagnosis and over a 6-month follow-up period.

**Diagnostic Criteria:**
- Acquired GI manifestations (including abdominal pain, nausea or vomiting, diarrhea, weight loss, melena) and a histopathological evidence of vasculitis in a GI specimen; or high-probability angiographic findings (smooth segmental narrowing, dilatation, occlusion or aneurysms affecting one or more GI arteries) with significant wall thickening (≥ 3mm) and the absence of vessel changes consistent with atherosclerosis, FMD or SAM).
- Inflammatory markers can be normal or elevated!

Vascular Ehlers-Danlos Syndrome (vEDS)

- Life-threatening arteriopathy caused by genetic alteration of COL3A1 gene.
- **Major criteria:** arterial rupture, intestinal rupture, uterine rupture or positive family history.
- **Minor criteria:** thin translucent skin, characteristic facial appearance (thin face, large appearing eyes, thin lips and nose), acrogeria, hypermobility of small joints, tendon and muscle rupture, talipes equinovarus, early onset varicosities, pneumothorax, gingival recession.
- **Two major criteria** are considered highly specific.
- **Two minor criteria** requires further testing to confirm the diagnosis of EDS.
- **Genetic testing:** sequence and deletion/duplication testing of the COL3A1 gene.

**Diagnostic Criteria:**
- Positive Villefranche criteria and genetic testing (COL3A1 gene).

Loeys-Dietz Syndrome (LDS)

- **Diagnosis consideration:** hypertelorism, a bifid uvula and/or cleft palate and generalized arterial tortuosity with widespread vascular aneurysm and dissection.
- **Diagnosis confirmation:** genetic alteration in 5 genes: transforming growth factor beta receptors 1 or 2, SMAD family member 3, or transforming growth factor beta 2 or 3, resulting in 5 LDS subtypes with less functional protein.
- **LDS type 3** (mutations of SMAD3): characterized more specifically by aortic and arterial aneurysms and dissections.

**Diagnostic Criteria:**
- Phenotype and genotype consistent with LDS
Median Arcuate Ligament Syndrome (MALS)

- Celiac artery compression by median arcuate ligament
- Arterial and neurogenic syndrome
- Diagnosis of exclusion

**Diagnostic Criteria:**
- Combination of symptoms (e.g. postprandial abdominal pain, weight loss), typical CTA findings of celiac artery compression by the median arcuate ligament with post-stenotic dilation or aneurysm, and increased velocity during exhalation compared with inhalation in an ultrasound.


---

**Demographics and Comorbidities**

<table>
<thead>
<tr>
<th></th>
<th>All Patients (n=118)</th>
<th>FMD (n=30)</th>
<th>Isolated aneurysm (n=29)</th>
<th>MALS (n=18)</th>
<th>LVGT (n=9)</th>
<th>Isolated Dissection (n=6)</th>
<th>MP/GP (n=6)</th>
<th>Trauma (n=5)</th>
<th>SAM (n=5)</th>
<th>EDS (n=3)</th>
<th>Takayasu's Arteritis (n=2)</th>
<th>PAN (n=1)</th>
<th>LDS (n=1)</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Age (Year)</strong></td>
<td>50.5 (39.0, 54.0)</td>
<td>60 (43, 54)</td>
<td>52 (39, 54)</td>
<td>50 (38, 51)</td>
<td>51 (41, 55)</td>
<td>45.5 (35, 54)</td>
<td>56 (54, 59)</td>
<td>50 (42, 59)</td>
<td>54 (52, 55)</td>
<td>41 (24, 53)</td>
<td>35 (26, 36)</td>
<td>51 (43, 59)</td>
<td>34</td>
</tr>
<tr>
<td><strong>Male, (%)</strong></td>
<td>42 (35.6)</td>
<td>3 (10.0)</td>
<td>7 (24.1)</td>
<td>4 (22.2)</td>
<td>4 (43.8)</td>
<td>6 (100)</td>
<td>4 (66.7)</td>
<td>5 (100)</td>
<td>5 (100)</td>
<td>3 (100)</td>
<td>1 (33.3)</td>
<td>0 (0)</td>
<td>0 (0)</td>
</tr>
<tr>
<td><strong>Hypertension, (%)</strong></td>
<td>61 (52.1)</td>
<td>18 (60.0)</td>
<td>12 (41.4)</td>
<td>6 (33.3)</td>
<td>4 (44.4)</td>
<td>3 (50.0)</td>
<td>2 (40.0)</td>
<td>4 (80.0)</td>
<td>5 (100)</td>
<td>2 (66.7)</td>
<td>2 (66.7)</td>
<td>2 (100)</td>
<td>1 (100)</td>
</tr>
<tr>
<td><strong>Tobacco Use, (%)</strong></td>
<td>51 (43.6)</td>
<td>16 (53.3)</td>
<td>10 (35.7)</td>
<td>6 (33.3)</td>
<td>5 (33.3)</td>
<td>3 (50.0)</td>
<td>2 (33.3)</td>
<td>3 (60.0)</td>
<td>3 (60.0)</td>
<td>1 (33.3)</td>
<td>1 (33.3)</td>
<td>0 (0)</td>
<td>1 (100)</td>
</tr>
<tr>
<td><strong>Hyperlipidemia, (%)</strong></td>
<td>20 (17.1)</td>
<td>6 (20.0)</td>
<td>5 (17.2)</td>
<td>0 (0)</td>
<td>2 (22.2)</td>
<td>2 (33.3)</td>
<td>1 (20.0)</td>
<td>0 (0)</td>
<td>2 (40.0)</td>
<td>0 (0)</td>
<td>1 (33.3)</td>
<td>1 (50.0)</td>
<td>0 (0)</td>
</tr>
<tr>
<td><strong>Diabetes, (%)</strong></td>
<td>10 (8.6)</td>
<td>4 (13.3)</td>
<td>4 (13.8)</td>
<td>0 (0)</td>
<td>0 (0)</td>
<td>1 (20.0)</td>
<td>0 (0)</td>
<td>0 (0)</td>
<td>0 (0)</td>
<td>0 (0)</td>
<td>0 (0)</td>
<td>0 (0)</td>
<td>1 (50.0)</td>
</tr>
<tr>
<td><strong>CAD, (%)</strong></td>
<td>10 (8.6)</td>
<td>2 (6.7)</td>
<td>3 (10.3)</td>
<td>0 (0)</td>
<td>1 (11.1)</td>
<td>1 (16.7)</td>
<td>0 (0)</td>
<td>0 (0)</td>
<td>1 (20.0)</td>
<td>0 (0)</td>
<td>1 (33.3)</td>
<td>1 (50.0)</td>
<td>0 (0)</td>
</tr>
<tr>
<td><strong>PAD, (%)</strong></td>
<td>6 (5.1)</td>
<td>3 (10.0)</td>
<td>0 (0)</td>
<td>1 (5.6)</td>
<td>0 (0)</td>
<td>0 (0)</td>
<td>0 (0)</td>
<td>1 (20.0)</td>
<td>0 (0)</td>
<td>1 (33.3)</td>
<td>0 (0)</td>
<td>0 (0)</td>
<td>0 (0)</td>
</tr>
</tbody>
</table>


26 of 43
### Arterial Pathology

<table>
<thead>
<tr>
<th></th>
<th>All Patients (n=118)</th>
<th>FMD (n=30)</th>
<th>Isolated aneurysm (n=29)</th>
<th>MALS (n=18)</th>
<th>LVGT (n=9)</th>
<th>Isolated Dissection (n=6)</th>
<th>MP/GP (n=6)</th>
<th>Trauma (n=5)</th>
<th>SAM (n=3)</th>
<th>EDS (n=2)</th>
<th>Takayasu’s Arteritis (n=3)</th>
<th>PAN (n=1)</th>
<th>LDS (n=1)</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Aneurysm</strong></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td></td>
<td>70 (59.3)</td>
<td>24 (80.0)</td>
<td>29 (100)</td>
<td>3 (16.7)</td>
<td>1 (11.1)</td>
<td>0 (0)</td>
<td>4 (80.0)</td>
<td>5 (100)</td>
<td>2 (66.7)</td>
<td>0 (0)</td>
<td>1 (50.0)</td>
<td>1 (100)</td>
<td></td>
</tr>
<tr>
<td><strong>Stenosis</strong></td>
<td></td>
<td>48 (40.7)</td>
<td>11 (36.7)</td>
<td>1 (3.5)</td>
<td>18 (100)</td>
<td>9 (100)</td>
<td>1 (16.7)</td>
<td>0 (0)</td>
<td>0 (0)</td>
<td>4 (80.0)</td>
<td>0 (0)</td>
<td>3 (100)</td>
<td>0 (0)</td>
</tr>
<tr>
<td><strong>Dissection</strong></td>
<td></td>
<td>29 (24.6)</td>
<td>9 (30.0)</td>
<td>0 (0)</td>
<td>1 (5.6)</td>
<td>3 (33.3)</td>
<td>6 (100)</td>
<td>0 (0)</td>
<td>4 (80.0)</td>
<td>3 (60.0)</td>
<td>2 (66.7)</td>
<td>0 (0)</td>
<td></td>
</tr>
<tr>
<td><strong>Thrombosis</strong></td>
<td></td>
<td>29 (24.6)</td>
<td>7 (23.3)</td>
<td>6 (20.7)</td>
<td>1 (5.5)</td>
<td>2 (22.2)</td>
<td>5 (83.3)</td>
<td>0 (0)</td>
<td>1 (20.0)</td>
<td>3 (60.0)</td>
<td>2 (66.7)</td>
<td>1 (100)</td>
<td></td>
</tr>
<tr>
<td><strong>Renal Infarction</strong></td>
<td></td>
<td>13 (11.0)</td>
<td>6 (20.0)</td>
<td>0 (0)</td>
<td>0 (0)</td>
<td>1 (11.1)</td>
<td>4 (66.7)</td>
<td>0 (0)</td>
<td>0 (0)</td>
<td>1 (20.0)</td>
<td>1 (33.3)</td>
<td>0 (0)</td>
<td></td>
</tr>
</tbody>
</table>


### Arterial Involvement

<table>
<thead>
<tr>
<th></th>
<th>All Patients (n=118)</th>
<th>FMD (n=30)</th>
<th>Isolated aneurysm (n=29)</th>
<th>MALS (n=18)</th>
<th>LVGT (n=9)</th>
<th>Isolated Dissection (n=6)</th>
<th>MP/GP (n=6)</th>
<th>Trauma (n=5)</th>
<th>SAM (n=3)</th>
<th>EDS (n=2)</th>
<th>Takayasu’s Arteritis (n=3)</th>
<th>PAN (n=1)</th>
<th>LDS (n=1)</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Renal Artery</strong></td>
<td></td>
<td>52 (44.1)</td>
<td>25 (83.3)</td>
<td>7 (24.1)</td>
<td>1 (6.6)</td>
<td>1 (11.1)</td>
<td>5 (83.3)</td>
<td>6 (100)</td>
<td>1 (20)</td>
<td>2 (40)</td>
<td>2 (66.7)</td>
<td>2 (66.7)</td>
<td>1 (50.0)</td>
</tr>
<tr>
<td><strong>Right</strong></td>
<td></td>
<td>16 (30.9)</td>
<td>9 (36.0)</td>
<td>4 (57.1)</td>
<td>0 (0)</td>
<td>0 (0)</td>
<td>0 (0)</td>
<td>1 (100)</td>
<td>0 (0)</td>
<td>0 (0)</td>
<td>2 (66.7)</td>
<td>0 (0)</td>
<td></td>
</tr>
<tr>
<td><strong>Left</strong></td>
<td></td>
<td>15 (28.9)</td>
<td>5 (20.0)</td>
<td>3 (42.9)</td>
<td>1 (100)</td>
<td>1 (100)</td>
<td>4 (80.0)</td>
<td>0 (0)</td>
<td>0 (0)</td>
<td>0 (0)</td>
<td>1 (50.0)</td>
<td>0 (0)</td>
<td></td>
</tr>
<tr>
<td><strong>Bilateral</strong></td>
<td></td>
<td>22 (40.4)</td>
<td>11 (44.0)</td>
<td>0 (0)</td>
<td>0 (0)</td>
<td>0 (0)</td>
<td>1 (20.0)</td>
<td>6 (100)</td>
<td>0 (0)</td>
<td>2 (100)</td>
<td>1 (50.0)</td>
<td>0 (0)</td>
<td></td>
</tr>
<tr>
<td><strong>Celiac artery</strong></td>
<td></td>
<td>43 (36.4)</td>
<td>7 (23.3)</td>
<td>3 (10.3)</td>
<td>18 (100)</td>
<td>4 (44.4)</td>
<td>0 (0)</td>
<td>0 (0)</td>
<td>3 (60.0)</td>
<td>5 (100)</td>
<td>2 (66.7)</td>
<td>1 (33.3)</td>
<td>0 (0)</td>
</tr>
<tr>
<td><strong>Splanic artery</strong></td>
<td></td>
<td>31 (26.3)</td>
<td>9 (30.0)</td>
<td>17 (58.6)</td>
<td>0 (0)</td>
<td>2 (22.2)</td>
<td>0 (0)</td>
<td>0 (0)</td>
<td>1 (20.0)</td>
<td>1 (20.0)</td>
<td>0 (0)</td>
<td>0 (0)</td>
<td>1 (50.0)</td>
</tr>
<tr>
<td><strong>SMA</strong></td>
<td></td>
<td>23 (19.5)</td>
<td>3 (10.0)</td>
<td>2 (6.9)</td>
<td>2 (11.1)</td>
<td>5 (65.6)</td>
<td>1 (16.7)</td>
<td>0 (0)</td>
<td>3 (60.0)</td>
<td>3 (60.0)</td>
<td>0 (0)</td>
<td>2 (66.7)</td>
<td>1 (50.0)</td>
</tr>
<tr>
<td><strong>Hepatic artery</strong></td>
<td></td>
<td>8 (6.8)</td>
<td>2 (6.7)</td>
<td>0 (0)</td>
<td>0 (0)</td>
<td>1 (11.1)</td>
<td>0 (0)</td>
<td>0 (0)</td>
<td>0 (0)</td>
<td>2 (40.0)</td>
<td>1 (33.3)</td>
<td>1 (33.3)</td>
<td>0 (0)</td>
</tr>
<tr>
<td><strong>IMA</strong></td>
<td></td>
<td>4 (3.4)</td>
<td>0 (0)</td>
<td>0 (0)</td>
<td>0 (0)</td>
<td>0 (0)</td>
<td>0 (0)</td>
<td>0 (0)</td>
<td>1 (20.0)</td>
<td>0 (0)</td>
<td>1 (33.3)</td>
<td>1 (50.0)</td>
<td>0 (0)</td>
</tr>
</tbody>
</table>

## Vital Signs and Laboratory Findings

### SBP
- All Patients: 120 (115, 125) (n=95)
- FMD: 129 (120, 137) (n=24)
- Isolated aneurysm: 116.5 (110, 124.5) (n=24)
- MALS: 114 (105, 124) (n=16)
- LVGT: 134 (120, 145) (n=9)
- Isolated Dissection: 130 (115, 140) (n=1)
- MP/GP: 130 (124, 147.3) (n=1)
- Trauma: 124 (124, 124) (n=1)
- SAM: 140 (135, 132) (n=2)
- Takayasu's Arteritis: 110 (108, 146) (n=3)
- PAN: 110 (n=1)
- LDS: 102 (n=1)

### DBP
- All Patients: 76 (66, 86) (n=95)
- FMD: 81 (70, 90.5) (n=24)
- Isolated aneurysm: 72 (65, 80) (n=24)
- MALS: 71 (65, 83) (n=16)
- LVGT: 68 (60, 77) (n=3)
- Isolated Dissection: 81 (75, 91.5) (n=1)
- MP/GP: 76 (74, 78) (n=1)
- Trauma: 74 (64, 100) (n=2)
- SAM: 82 (60, 86) (n=3)
- Takayasu's Arteritis: 60 (n=1)
- PAN: 58 (n=1)

### Hemoglobin
- All Patients: 13.3 (12.1, 14.6) (n=88)
- FMD: 13.1 (11.9, 14.5) (n=22)
- Isolated aneurysm: 13.1 (12.4, 14) (n=20)
- MALS: 14.05 (12.1, 14.7) (n=14)
- LVGT: 14 (13.8, 15) (n=8)
- Isolated Dissection: 11.1 (10.6, 12.2) (n=4)
- MP/GP: 14.7 (12.1, 15.6) (n=3)
- Trauma: 14.7 (12, 1.5) (n=3)
- SAM: 11.9 (9.2, 13.2) (n=1)
- Takayasu's Arteritis: 12.45 (12, 1.2) (n=2)

### ESR
- All Patients: 23 (16, 51) (n=39)
- FMD: 27 (15, 40) (n=7)
- Isolated aneurysm: 51 (14, 75) (n=7)
- MALS: 19 (16, 22) (n=3)
- LVGT: 23 (22, 42) (n=6)
- Isolated Dissection: 34 (30, 44) (n=7)
- MP/GP: 34 (32, 74) (n=6)
- Trauma: 18 (15, 70) (n=2)
- SAM: 10 (n=1)
- Takayasu's Arteritis: 33 (14, 54) (n=3)

### CRP
- All Patients: 1.2 (0.5, 3.5) (n=33)
- FMD: 0.65 (0.5, 6.8) (n=8)
- Isolated aneurysm: 3.35 (1.85, 5.85) (n=4)
- MALS: 0.9 (0.5, 1.3) (n=2)
- LVGT: 2 (0.6, 2.94) (n=9)
- Isolated Dissection: 13.25 (6.9, 19.6) (n=2)
- MP/GP: 1.2 (0.6, 1.8) (n=4)
- Trauma: 11.8 (n=1)
- SAM: 3 (1.2, 3.4) (n=2)
- Takayasu's Arteritis: 0.5 (n=1)

## Type of Imaging

### CTA/CT
- All Patients: 99 (83.9) (n=118)
- FMD: 24 (80.0) (n=30)
- Isolated aneurysm: 28 (86.6) (n=29)
- MALS: 14 (77.8) (n=18)
- LVGT: 9 (100) (n=8)
- Isolated Dissection: 6 (100) (n=6)
- MP/GP: 1 (100) (n=6)
- Trauma: 5 (100) (n=5)
- SAM: 3 (100) (n=5)
- Takayasu's Arteritis: 1 (100) (n=3)
- PAN: 1 (100) (n=2)
- LDS: 1 (100) (n=1)

### MRA
- All Patients: 21 (18.0) (n=118)
- FMD: 9 (30.0) (n=30)
- Isolated aneurysm: 1 (3.5) (n=9)
- MALS: 4 (22.2) (n=4)
- LVGT: 2 (22.2) (n=2)
- Isolated Dissection: 0 (0) (n=0)
- MP/GP: 0 (0) (n=0)
- Trauma: 1 (20.0) (n=1)
- SAM: 2 (66.7) (n=2)
- Takayasu's Arteritis: 0 (0) (n=0)

### DUS/US
- All Patients: 15 (12.7) (n=118)
- FMD: 3 (10.0) (n=30)
- Isolated aneurysm: 0 (0) (n=29)
- MALS: 5 (27.8) (n=18)
- LVGT: 0 (0) (n=8)
- Isolated Dissection: 2 (33.3) (n=6)
- MP/GP: 1 (16.7) (n=6)
- Trauma: 2 (20.0) (n=5)
- SAM: 1 (20.0) (n=5)
- Takayasu's Arteritis: 0 (0) (n=3)

## Presenting Findings and Symptoms

<table>
<thead>
<tr>
<th></th>
<th>All Patients</th>
<th>FMD (n=30)</th>
<th>Isolated aneurysm (n=29)</th>
<th>MALS (n=18)</th>
<th>LVGT (n=9)</th>
<th>Isolated Dissection (n=6)</th>
<th>MP/GP (n=5)</th>
<th>Trauma (n=5)</th>
<th>SAM (n=3)</th>
<th>EDS (n=3)</th>
<th>Takayasu’s Arteritis (n=2)</th>
<th>PAN (n=1)</th>
<th>LDS (n=1)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Abdominal/Flank Pain Abdominal/Flank Pain</td>
<td>65 (55.1)</td>
<td>14 (46.7)</td>
<td>8 (27.6)</td>
<td>13 (72.2)</td>
<td>6 (100)</td>
<td>1 (16.7)</td>
<td>5 (100)</td>
<td>4 (80.0)</td>
<td>2 (66.7)</td>
<td>1 (33.3)</td>
<td>1 (50.0)</td>
<td>0 (0)</td>
<td>0 (0)</td>
</tr>
<tr>
<td>Nausea/Vomiting Nausea/Vomiting</td>
<td>17 (14.4)</td>
<td>3 (10.0)</td>
<td>2 (6.9)</td>
<td>6 (33.3)</td>
<td>3 (33.3)</td>
<td>1 (16.7)</td>
<td>1 (16.7)</td>
<td>0 (0)</td>
<td>0 (0)</td>
<td>1 (33.3)</td>
<td>0 (0)</td>
<td>0 (0)</td>
<td>0 (0)</td>
</tr>
<tr>
<td>Diarrhea Diarrhea</td>
<td>8 (6.8)</td>
<td>2 (6.7)</td>
<td>0 (0)</td>
<td>4 (22.2)</td>
<td>0 (0)</td>
<td>1 (16.7)</td>
<td>0 (0)</td>
<td>1 (20.0)</td>
<td>0 (0)</td>
<td>0 (0)</td>
<td>0 (0)</td>
<td>0 (0)</td>
<td>0 (0)</td>
</tr>
<tr>
<td>Asymptomatic Asymptomatic</td>
<td>31 (26.3)</td>
<td>3 (10.6)</td>
<td>21 (72.4)</td>
<td>3 (16.7)</td>
<td>0 (0)</td>
<td>0 (0)</td>
<td>0 (0)</td>
<td>1 (20.0)</td>
<td>1 (33.3)</td>
<td>1 (33.3)</td>
<td>0 (0)</td>
<td>1 (100)</td>
<td>0 (0)</td>
</tr>
</tbody>
</table>


## Management

<table>
<thead>
<tr>
<th></th>
<th>All Patients</th>
<th>FMD (n=30)</th>
<th>Isolated aneurysm (n=29)</th>
<th>MALS (n=18)</th>
<th>LVGT (n=9)</th>
<th>Isolated Dissection (n=6)</th>
<th>MP/GP (n=5)</th>
<th>Trauma (n=5)</th>
<th>SAM (n=3)</th>
<th>EDS (n=3)</th>
<th>Takayasu’s Arteritis (n=2)</th>
<th>PAN (n=1)</th>
<th>LDS (n=1)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Coil Embolization</td>
<td>17 (14.4)</td>
<td>6 (20.0)</td>
<td>7 (24.1)</td>
<td>1 (5.6)</td>
<td>1 (10.0)</td>
<td>0 (0)</td>
<td>0 (0)</td>
<td>1 (20.0)</td>
<td>0 (0)</td>
<td>0 (0)</td>
<td>1 (50.0)</td>
<td>0 (0)</td>
<td>0 (0)</td>
</tr>
<tr>
<td>Angioplasty/Stenting</td>
<td>14 (11.9)</td>
<td>5 (16.7)</td>
<td>1 (3.5)</td>
<td>3 (16.7)</td>
<td>0 (0)</td>
<td>2 (33.3)</td>
<td>0 (0)</td>
<td>1 (20.0)</td>
<td>0 (0)</td>
<td>1 (33.3)</td>
<td>1 (33.3)</td>
<td>0 (0)</td>
<td>0 (0)</td>
</tr>
<tr>
<td>Anticoagulation</td>
<td>14 (11.9)</td>
<td>5 (16.7)</td>
<td>0 (0)</td>
<td>1 (5.6)</td>
<td>2 (22.2)</td>
<td>1 (16.7)</td>
<td>0 (0)</td>
<td>1 (20.0)</td>
<td>3 (60.0)</td>
<td>1 (33.3)</td>
<td>0 (0)</td>
<td>0 (0)</td>
<td>0 (0)</td>
</tr>
<tr>
<td>Open Resection/Surgical Revascularization</td>
<td>12 (10.2)</td>
<td>4 (13.3)</td>
<td>4 (13.8)</td>
<td>3 (16.7)</td>
<td>0 (0)</td>
<td>0 (0)</td>
<td>0 (0)</td>
<td>1 (20.0)</td>
<td>0 (0)</td>
<td>0 (0)</td>
<td>0 (0)</td>
<td>0 (0)</td>
<td>0 (0)</td>
</tr>
<tr>
<td>Fibrinolysis</td>
<td>12 (10.2)</td>
<td>0 (0)</td>
<td>0 (0)</td>
<td>0 (0)</td>
<td>4 (44.4)</td>
<td>0 (0)</td>
<td>6 (100)</td>
<td>0 (0)</td>
<td>1 (20.0)</td>
<td>0 (0)</td>
<td>2 (90.0)</td>
<td>1 (100)</td>
<td>0 (0)</td>
</tr>
<tr>
<td>Splenectomy</td>
<td>6 (5.1)</td>
<td>2 (6.7)</td>
<td>4 (13.8)</td>
<td>0 (0)</td>
<td>0 (0)</td>
<td>0 (0)</td>
<td>0 (0)</td>
<td>0 (0)</td>
<td>0 (0)</td>
<td>0 (0)</td>
<td>0 (0)</td>
<td>0 (0)</td>
<td>0 (0)</td>
</tr>
<tr>
<td>Open MAL release</td>
<td>4 (3.4)</td>
<td>0 (0)</td>
<td>0 (0)</td>
<td>4 (22.2)</td>
<td>0 (0)</td>
<td>0 (0)</td>
<td>0 (0)</td>
<td>0 (0)</td>
<td>0 (0)</td>
<td>0 (0)</td>
<td>0 (0)</td>
<td>0 (0)</td>
<td>0 (0)</td>
</tr>
<tr>
<td>Laparoscopic MAL release</td>
<td>3 (2.6)</td>
<td>0 (0)</td>
<td>0 (0)</td>
<td>3 (16.7)</td>
<td>0 (0)</td>
<td>0 (0)</td>
<td>0 (0)</td>
<td>0 (0)</td>
<td>0 (0)</td>
<td>0 (0)</td>
<td>0 (0)</td>
<td>0 (0)</td>
<td>0 (0)</td>
</tr>
<tr>
<td>Nephrectomy</td>
<td>1 (0.9)</td>
<td>1 (3.33)</td>
<td>0 (0)</td>
<td>0 (0)</td>
<td>0 (0)</td>
<td>0 (0)</td>
<td>0 (0)</td>
<td>0 (0)</td>
<td>0 (0)</td>
<td>0 (0)</td>
<td>0 (0)</td>
<td>0 (0)</td>
<td>0 (0)</td>
</tr>
<tr>
<td>Conservative treatment</td>
<td>39 (33.1)</td>
<td>9 (30.0)</td>
<td>16 (55.2)</td>
<td>4 (22.2)</td>
<td>2 (22.2)</td>
<td>3 (100)</td>
<td>2 (40.0)</td>
<td>1 (20.0)</td>
<td>1 (33.3)</td>
<td>0 (0)</td>
<td>0 (0)</td>
<td>1 (100)</td>
<td>0 (0)</td>
</tr>
</tbody>
</table>

# Outcomes

<table>
<thead>
<tr>
<th></th>
<th>All Patients (n=118)</th>
<th>FMD (n=30)</th>
<th>Isolated aneurysm (n=29)</th>
<th>MALS (n=18)</th>
<th>LVGT (n=9)</th>
<th>Isolated Dissection (n=6)</th>
<th>MP/GP (n=6)</th>
<th>Trauma (n=5)</th>
<th>SAM (n=3)</th>
<th>EDS (n=2)</th>
<th>Takayasu’s Arteritis (n=1)</th>
<th>PAN (n=1)</th>
<th>LDS (n=1)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Symptoms relieved</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td></td>
<td>77/84 (91.7)</td>
<td>20/21 (95.2)</td>
<td>13/14 (92.9)</td>
<td>10/14 (71.4)</td>
<td>7/8 (87.5)</td>
<td>5 (100)</td>
<td>6 (100)</td>
<td>5 (100)</td>
<td>4 (100)</td>
<td>2 (100)</td>
<td>2 (100)</td>
<td>1 (100)</td>
<td>1 (100)</td>
</tr>
<tr>
<td>Symptoms recurred</td>
<td>12/83 (14.5)</td>
<td>3/21 (14.3)</td>
<td>2/13 (15.4)</td>
<td>3/14 (21.4)</td>
<td>1/2 (12.5)</td>
<td>0 (0)</td>
<td>2 (33.3)</td>
<td>1 (20)</td>
<td>0 (0)</td>
<td>0 (0)</td>
<td>0/2 (0)</td>
<td>0/1 (0)</td>
<td>0/0</td>
</tr>
<tr>
<td>Surgical Success</td>
<td>32/86 (27.1)</td>
<td>13 (43.3)</td>
<td>8 (27.6)</td>
<td>4 (22.2)</td>
<td>1 (11.1)</td>
<td>1 (16.7)</td>
<td>0 (0)</td>
<td>2 (40.0)</td>
<td>1 (20.0)</td>
<td>1 (33.3)</td>
<td>1 (33.3)</td>
<td>0 (0)</td>
<td>0 (0)</td>
</tr>
<tr>
<td>Imaging</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Improved, (%)</td>
<td>17 (30.4)</td>
<td>9 (40.9)</td>
<td>0 (0)</td>
<td>4 (44.4)</td>
<td>3 (60.0)</td>
<td>0 (0)</td>
<td>0 (0)</td>
<td>1 (100)</td>
<td>0 (0)</td>
<td>1 (33.3)</td>
<td>1 (33.3)</td>
<td>0 (0)</td>
<td>0 (0)</td>
</tr>
<tr>
<td>Unchanged, (%)</td>
<td>16 (28.6)</td>
<td>9 (40.9)</td>
<td>1 (33.3)</td>
<td>0 (0)</td>
<td>0 (0)</td>
<td>0 (0)</td>
<td>0 (0)</td>
<td>0 (0)</td>
<td>0 (0)</td>
<td>0 (0)</td>
<td>0 (0)</td>
<td>0 (0)</td>
<td>0 (0)</td>
</tr>
<tr>
<td>Worse, (%)</td>
<td>19 (33.9)</td>
<td>4 (17.1)</td>
<td>3 (13.6)</td>
<td>5 (55.6)</td>
<td>2 (40.0)</td>
<td>2 (40.0)</td>
<td>1 (20.0)</td>
<td>2 (40.0)</td>
<td>2 (66.7)</td>
<td>1 (33.3)</td>
<td>1 (33.3)</td>
<td>0 (0)</td>
<td>0 (0)</td>
</tr>
<tr>
<td>No F/U Done, (%)</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
</tbody>
</table>


# Summary

**Demographics:** 118 study participants, Age: 47.0 ± 9.9 years, Female: (64.4%)

**Comorbidities:** HTN (52.1%) and tobacco use (43.6%)

**Diagnoses:** FMD (25.4%), isolated aneurysms (24.6%), MALS (15.3%), LVGT (7.6%), isolated dissection (5.1%), MP/GP (5.1%), trauma (4.2%), SAM (4.2%), Takayasu’s arteritis (2.5%), EDS (2.5%), PAN (1.7%), and LDS (0.8%).

**Pathologies:** Aneurysms (59.3%), stenosis (40.7%), dissection (24.6%), and thrombosis in 24.6%.

**Arterial Involvement:** renal (44.1%), celiac (36.4%), and splenic (26.3%).

**Images:** CT (83.9%), CBA (19.7%), MRA (18.0%), and duplex ultrasound (12.7%).
Labs: ESR and CRP diagnostic role is ??!!

Genetic Testing +/- Pathology: multiple arterial involvement+ beading: SAM vs. FMD vs. vEDS

Symptoms: abdominal pain (55.1%), nausea and vomiting (14.4%), diarrhea (6.8%) and (37.3%) were asymptomatic.

Management: Coil embolization (14.4%), angioplasty/stenting (11.9%), anticoagulation (11.9%), open resection/surgical revascularization (10.2%), prednisone (10.2%) and conservative treatment (33.1%).

Outcome: During a median follow-up time of 686 days the majority (91.7%) of patients had relief of their symptoms. Of the 37 pts who had flu imaging, 30.4% showed improvement, 28.6% remained unchanged, and 7.1% worsened.

Segmental Arterial Mediolysis
Systemic Review & Data Analysis of 143 Patients

- Literature review of all relevant SAM case studies from 2005 to 2018 yielded 126 individual SAM cases from 66 reports.

- Included papers: reported patient demographics (at least age and gender), arterial involvement, arteriopathy, and diagnostic strategy (imaging or labs or histology or symptoms).

- We identified 17 additional SAM cases from our center, bringing our analysis to 143 patients.
SAM Diagnosis

- **Our Diagnostic Criteria:**
  - Dissection and/or aneurysm with or without organ infarction in *multiple* mesenteric and/or renal arteries with exclusion of FMD, LVGT, connective tissue disorder, or other vasculopathies, and no significant concurrent arterial wall thickening (< 3 mm) or elevation of inflammatory markers (erythrocyte sedimentation rate (ESR) < 20 mm/h and C-reactive protein (CRP) < 5 mg/dL)

- Different authors used different diagnostic criteria!

- **Tissue diagnosis if possible:** Vacuolar degeneration and lysis of the medial layer of the arterial wall, often resulting in dissection, aneurysm, occlusion, or stenosis

- **Differential Diagnoses:** same process as the previous project

Demographics

<table>
<thead>
<tr>
<th>Variable</th>
<th>Value</th>
</tr>
</thead>
<tbody>
<tr>
<td>Age (year)</td>
<td>55 (48, 63)</td>
</tr>
<tr>
<td>Male</td>
<td>67.8 %</td>
</tr>
<tr>
<td>Hypertension</td>
<td>42.7 %</td>
</tr>
<tr>
<td>History of Tobacco Use</td>
<td>11.9 %</td>
</tr>
<tr>
<td>Hyperlipidemia</td>
<td>11.9 %</td>
</tr>
<tr>
<td>Diabetes</td>
<td>1.4 %</td>
</tr>
</tbody>
</table>
### Presenting Symptoms

<table>
<thead>
<tr>
<th>Presentation</th>
<th>%</th>
</tr>
</thead>
<tbody>
<tr>
<td>Abdominal/Flank Pain</td>
<td>79.7</td>
</tr>
<tr>
<td>Intra-abdominal bleeding</td>
<td>49.7</td>
</tr>
<tr>
<td>Nausea/Vomiting</td>
<td>16.1</td>
</tr>
<tr>
<td>Cerebrovascular symptoms</td>
<td>11.9</td>
</tr>
<tr>
<td>Melena/Hematochezia</td>
<td>5.6</td>
</tr>
<tr>
<td>Asymptomatic</td>
<td>4.9</td>
</tr>
<tr>
<td>Diarrhea</td>
<td>4.9</td>
</tr>
<tr>
<td>Shock</td>
<td>4.2</td>
</tr>
</tbody>
</table>


### Vital Signs and Labs

<table>
<thead>
<tr>
<th>Vital Signs and Labs</th>
<th>%</th>
</tr>
</thead>
<tbody>
<tr>
<td>Hypertension</td>
<td>42.4</td>
</tr>
<tr>
<td>Hypotension</td>
<td>25.4</td>
</tr>
<tr>
<td>Low Hemoglobin</td>
<td>50.8</td>
</tr>
<tr>
<td>Low Hematocrit</td>
<td>46.7</td>
</tr>
<tr>
<td>Elevated ESR</td>
<td>33.3</td>
</tr>
<tr>
<td>Elevated CRP</td>
<td>68.6</td>
</tr>
</tbody>
</table>

## Diagnostic Imaging

<table>
<thead>
<tr>
<th>Modality</th>
<th>%</th>
</tr>
</thead>
<tbody>
<tr>
<td>CT/CTA</td>
<td>77.6</td>
</tr>
<tr>
<td>CBA</td>
<td>35</td>
</tr>
<tr>
<td>Laparotomy</td>
<td>32.9</td>
</tr>
<tr>
<td>MRA</td>
<td>9.1</td>
</tr>
<tr>
<td>US/DUS</td>
<td>9.1</td>
</tr>
<tr>
<td>Other</td>
<td>9.8</td>
</tr>
</tbody>
</table>


## Arterial Involvement

<table>
<thead>
<tr>
<th>Arterial Involvement</th>
<th>%</th>
</tr>
</thead>
<tbody>
<tr>
<td>SMA</td>
<td>53.1</td>
</tr>
<tr>
<td>Hepatic</td>
<td>44.8</td>
</tr>
<tr>
<td>Celiac</td>
<td>35.7</td>
</tr>
<tr>
<td>Renal</td>
<td>25.9</td>
</tr>
<tr>
<td>Splenic</td>
<td>24.5</td>
</tr>
<tr>
<td>Cerebrovascular</td>
<td>13.3</td>
</tr>
<tr>
<td>IMA</td>
<td>10.5</td>
</tr>
<tr>
<td>Multiple</td>
<td>62.2</td>
</tr>
</tbody>
</table>

### Pathology Findings

<table>
<thead>
<tr>
<th>Pathologic Finding</th>
<th>%</th>
</tr>
</thead>
<tbody>
<tr>
<td>Aneurysm</td>
<td>76.2</td>
</tr>
<tr>
<td>Dissection</td>
<td>60.8</td>
</tr>
<tr>
<td>Rupture</td>
<td>45.5</td>
</tr>
<tr>
<td>Stenosis</td>
<td>18.9</td>
</tr>
<tr>
<td>Occlusion</td>
<td>16.9</td>
</tr>
<tr>
<td>&quot;String of Beads&quot;</td>
<td>14.7</td>
</tr>
<tr>
<td>Thrombosis</td>
<td>14.7</td>
</tr>
<tr>
<td>Pseudoaneurysm</td>
<td>11.9</td>
</tr>
<tr>
<td>Wall thickening</td>
<td>7.69</td>
</tr>
</tbody>
</table>


### Vascular Pathologies


![Vascular Pathologies Graph](image-url)
### Diagnostic Criteria

<table>
<thead>
<tr>
<th>Diagnostic Tool</th>
<th>%</th>
</tr>
</thead>
<tbody>
<tr>
<td>CT/CTA</td>
<td>59.4</td>
</tr>
<tr>
<td>Histology</td>
<td>44.1</td>
</tr>
<tr>
<td>CBA</td>
<td>29.4</td>
</tr>
<tr>
<td>Labs</td>
<td>10.5</td>
</tr>
<tr>
<td>Autopsy</td>
<td>5.6</td>
</tr>
</tbody>
</table>


### Diagnostic Tool

#### SAM Diagnostic Method Trendlines

![Graph showing trendlines for different diagnostic tools from 2005 to 2018](image)

### Management

<table>
<thead>
<tr>
<th>Modality</th>
<th>%</th>
</tr>
</thead>
<tbody>
<tr>
<td>Coil Embolization</td>
<td>27.9</td>
</tr>
<tr>
<td>Abdominal organ surgery</td>
<td>23.5</td>
</tr>
<tr>
<td>Open artery repair</td>
<td>20.6</td>
</tr>
<tr>
<td>Antihypertensives</td>
<td>19.9</td>
</tr>
<tr>
<td>Anticoagulation</td>
<td>11.8</td>
</tr>
<tr>
<td>Antiplatelets</td>
<td>10.3</td>
</tr>
<tr>
<td>Angioplasty/Stenting</td>
<td>8.1</td>
</tr>
<tr>
<td>Conservative treatment</td>
<td>8.1</td>
</tr>
</tbody>
</table>


### Outcomes

<table>
<thead>
<tr>
<th>Outcomes</th>
<th>%</th>
</tr>
</thead>
<tbody>
<tr>
<td>Survived</td>
<td>93</td>
</tr>
<tr>
<td>Symptoms reported</td>
<td>70.6</td>
</tr>
<tr>
<td>Improved</td>
<td>90.6</td>
</tr>
<tr>
<td>Unchanged</td>
<td>5.2</td>
</tr>
<tr>
<td>Worsened</td>
<td>4.2</td>
</tr>
<tr>
<td>Imaging</td>
<td>58.8</td>
</tr>
<tr>
<td>Improved</td>
<td>66.3</td>
</tr>
<tr>
<td>Unchanged</td>
<td>17.5</td>
</tr>
<tr>
<td>Worsened</td>
<td>16.3</td>
</tr>
<tr>
<td>No F/U Reported</td>
<td>28.7</td>
</tr>
</tbody>
</table>

### Segmental Arterial Mediolysis (SAM) vs. Fibromuscular Dysplasia (FMD)

#### Demographics

<table>
<thead>
<tr>
<th></th>
<th>SAM</th>
<th>FMD</th>
</tr>
</thead>
<tbody>
<tr>
<td>Age at onset</td>
<td>50-60s</td>
<td>40-50s</td>
</tr>
<tr>
<td>Gender</td>
<td>67% Male</td>
<td>80-90% Female</td>
</tr>
<tr>
<td>History</td>
<td>Usually spontaneous</td>
<td>Drug-resistant hypertension, abdominal bruit, some cases with family history</td>
</tr>
<tr>
<td>Presentation</td>
<td>More dramatic and severe: Abdominal pain, intra-abdominal bleeding</td>
<td>Less severe: Asymptomatic, hypertension, abdominal pain, headaches</td>
</tr>
</tbody>
</table>


#### Arteries involved

<table>
<thead>
<tr>
<th>Arteries involved</th>
<th>SAM</th>
<th>FMD</th>
</tr>
</thead>
<tbody>
<tr>
<td>SMA</td>
<td>53%</td>
<td>Mesenteric 22%</td>
</tr>
<tr>
<td>Hepatic</td>
<td>45%</td>
<td></td>
</tr>
<tr>
<td>Celiac</td>
<td>36%</td>
<td></td>
</tr>
<tr>
<td>Splenic</td>
<td>25%</td>
<td></td>
</tr>
<tr>
<td>Renal</td>
<td>26%</td>
<td>66-75%</td>
</tr>
<tr>
<td>Cerebrovascular</td>
<td>13%</td>
<td>73-80%</td>
</tr>
</tbody>
</table>

## Segmental Arterial Mediolysis (SAM) vs. Fibromuscular Dysplasia (FMD)

<table>
<thead>
<tr>
<th>Imaging findings</th>
<th>SAM</th>
<th>FMD</th>
</tr>
</thead>
<tbody>
<tr>
<td>Aneurysm</td>
<td>76%</td>
<td>22-28%</td>
</tr>
<tr>
<td>Dissection</td>
<td>60%</td>
<td>17-22%</td>
</tr>
<tr>
<td>Rupture</td>
<td>45%</td>
<td>Not reported</td>
</tr>
<tr>
<td>String of Beads</td>
<td>15%</td>
<td>60-70%</td>
</tr>
<tr>
<td>Occlusion</td>
<td>17%</td>
<td>Not reported</td>
</tr>
</tbody>
</table>


**Imaging:**

- A: Right internal carotid artery (ICA) dissection (upper arrow) and irregularity (lower arrow)
- B: Right ICA beading appearance

**Histologic findings**

<table>
<thead>
<tr>
<th></th>
<th>Vascular degeneration of the arterial media</th>
<th>Fibrous or fibromuscular thickening of the arterial wall</th>
</tr>
</thead>
</table>

### Summary

**Demographics:** 143 study pts, median age: 55 years and 67.8% were males

**Comorbidities:** HTN (42.7%), tobacco abuse (11.9%), HL (11.9%), and DM (1.4%)

**Symptoms:** abdominal pain (79.7%), intra-abdominal bleeding (49.7%), nausea/vomiting (16.1%), cerebrovascular symptoms (11.9%), melena/hematochezia (5.6%), diarrhea (4.3%), shock (4.2%), and asymptomatic (4.9%)

**Involvement:** SMA (53.1%), hepatic (44.8%), celiac (35.7%), renal (25.9%), splenic (24.5%), cerebrovascular involvement (13.3%) and IMA (10.5%)

**Pathology:** aneurysm (76.2%), dissection (60.8%), rupture (45.5%), stenosis (16.9%), occlusion (16.9%), ‘string of beads’ (14.7%), thrombosis (14.7%), pseudo-aneurysm (11.9%), and wall thickening (7.7%)


### Summary II

**Diagnostic Tools:** CT/CTA (59.4%), histology (44.1%) CBA (29.4%), and autopsy (5.6%)

**Genetic Testing +/- Pathology:** DD: FMD, SAM or vEDS when multiple arteries are involved + beading

**Interventions:** Coil embolization (27.9%), abdominal organ surgery (23.5%), open arterial repair (20.6%) and angioplasty/stenting (8.1%)

**Medical Management:** antihypertensive (19.9%), anticoagulation (11.8%), antiplatelet agents (10.3%) and conservative treatment only (8.1%)

**Outcome:** Improvement (90.6%) of patients with a reported final outcome.
Of the 80 pts who had f/u imaging, 66.3% showed improvement, 17.5% remained unchanged, and 16.3% worsened.

### Our Approach

- **Multidisciplinary approach**: Vascular medicine, vascular surgery, IR, general surgery.
- Careful History and Physical.
- **Initial Labs**: CBC, CMP, LFT’s, lipase, amylase.
- **Images**: CT/CTA or MR/MRA.
- **Exclude Common Diagnoses**: gastroenteritis, pancreatitis, PUD, liver disease.
- **Further Work Up**: ESR/CRP, ANCA, tissue biopsy and genetic testing.
- **Use diagnostic definitions and tools**: Confirmed or presumed diagnoses!
- **Follow up**: based on diagnosis: CTA thin cuts in 1, 3, 6, and 12 months.

---

### Abdominal Vasculopathy Work-up Algorithm

**Suspicious Symptoms for NIVIs:**
- Abdominal Pain, Nausea, Vomiting, Diarrhea

**Label**:
- CBC, Creatinine, LFT, Lipase, Amylase

**Exclude Common Diagnoses**
- (e.g. Gastroenteritis, Peptic Ulcer Disease, Appendicitis, Pancreatitis, Liver Disease)

**Imaging**: CT or MRA of Abdomen and Pelvis
- Labs: ESR, CRP, ANCA, sgot and tissue biopsy
Final Summary

• Non-atherosclerotic abdominal vasculopathies are rare but can be life threatening.
• No consensus on some diagnostic criteria or work up strategy.
• CT angiogram has become the imaging of choice.
• ESR/CRP can be elevated in most pathologies and their role is questionable.
• Role of genetic testing and pathology (multiple pathologies in multiple vessels).

Final Summary II

• Most patients received conservative therapy.
• Endovascular therapy has been more frequently used.
• Most patients have had symptomatic improvement. Improvement on images varied.
• Multidisciplinary approach.
• More research is needed to address gaps in diagnoses and management.
Sunset, Gaza City

Thank You!
Sydney Olson
Jonathan Hyde
Dawn Witt
Ross Garberich

Questions?