Current Trends in Aortic Syndromes

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NO DISCLOSURES

Aortic Syndromes

• Aortic Aneurysm
• Aortic Dissection
• Intramural Hematoma (IMH)
• Penetrating Arterial Ulcer
Objectives

- Spectrum of Aortic Syndromes
- Historical Milestones
- Risk Factors & Epidemiology
- Aortic Imaging Modalities
- Classification Schema
- Operative Techniques & Outcomes
- Endovascular Repair [TEVAR]

Historical Milestones

- 1760 Nicholls autopsy of King George II reveals intimal tear & aortic wall hematoma after he collapsed while straining on a commode
- 1761 Morgagni coins term 'aortic dissection'
- 1930 Erdheim describes histologic changes - cystic medionecrosis
- 1935 Gurin attempts repair by fenestrating iliac artery
- 1948 Contrast angiography introduced for diagnosis
- 1955 DeBakey surgically treats patients with primary repair
- 1956 Cooley & DeBakey: Cardiopulmonary bypass with selective anterior cerebral perfusion used (mainstay of contemporary aortic surgery)
- 1965 DeBakey classification schema;
- 1965 Wheat medical treatment to decrease bp & wall stress (dp/dt)
- 1975 Griep uses hypothermic circulatory arrest
- 1990 Endovascular stents
There is no disease more conducive to clinical humility than aneurysm of the aorta.

Sir William Osler

Risk Factors for Aortic Syndromes

• Connective Tissue Disorders
  – Marfan Syndrome
  – Ehlers-Danlos Syndrome
  – Loeys-Dietz Syndrome
• Degenerative
  – Atherosclerosis/Hypertensive
  – Cystic Medial Necrosis
• Congenital
  – Bicuspid Aortic Valve/Ascending Aortopathy
  – Coarctation of the Aorta
• Inflammatory
  – Arteritis/Vasculitis
  – Bacterial/Syphilitic
• Familial predisposition Aneurysm/Dissection
• Aortic Dissection
Epidemiology of Thoracic Aortic Aneurysms

- 30-60,000 deaths/year
- 18th most common cause of death (more than HIV)
- Frequency increasing
- Circadian-Diurnal variation
- Exact prevalence unknown
  - Anterior MI
  - Sudden cardiac death

Epidemiology of Aortic Dissections

- Estimated 2.6-3.5/100,000 patient years
- IRAD Database:
  - Mean age 63
  - 2:1 male predominance
  - Older patients: 72% have HTN, atherosclerosis, or previous heart surgery
  - Younger patients have Marfan’s or bicupsid AoV

Tsai et al, Circulation 2005
Criteria for Intervention on the Diseased Aorta

Diagnosis of Aortic Syndromes

- Poor prognosis after dissection/rupture mandates intervention before it occurs.
- Biomarker identifying risk/presence of aortic syndromes would help differentiate chest pain syndromes with different management.
  - Matrix metallo-proteinases
  - Circulating smooth muscle myosin chain
  - Inflammatory markers: CRP, fibrinogen, elastin fragments
  - Ribonucleic acid signatures
- Currently no available, reliable biomarker assay.

Elefteriades et al, J Am Coll Cardiol 2010
Annual Rates of Complications Related to Aortic Size

Hinge Points Defining Lifetime Risks

Size best criteria determining intervention

Imaging most reliable diagnostic tool
Aortic Imaging Modalities

Goals of Aortic Imaging
- Confirmation of Diagnosis
- Classification
- Tear localization & extent (dissection)
- Indicators of Emergency
  - Pericardial/Media/Intinal/Pleural hemorrhage
- Arch & Side-branch involvement

Aortic Imaging Modalities
- Each imaging modality is accurate for a specific portion of aorta
  - Need multiple modalities
- Compare images versus all previous images
- Changes < 3 mm imperceptible because aorta is dynamic structure
Echocardiography

- Available
- Crisp images
- Aortic root to STJ
- Assess for AI, tamponade, LV fxn
- TEE better for arch, prox desc Ao

Computed Tomography

- Widely available
- Excellent for distal ascending aorta, arch & head vessels, descending thoracic & abdominal aorta
- Axial cuts at root/valve level make measurement difficult
Computed Tomography/M2S

CT Axial, Coronal & Sagittal cuts facilitate 3-D reconstruction using specialized operative planning software

MRI

- Beautiful images
- Highly accurate
- Limited availability, especially in emergencies
Indications for Operative Intervention

Operative Indications For Ascending & Arch Aorta

- Size >5.5 cm atherosclerotic, degenerative or hypertensive aneurysms
- Size >5.0 cm with bicupsid aortic valve, connective tissue disorder, familial history of aneurysms/dissection
- Enlargement >0.5cm/6-12 months
- Symptomatic Aortic Valve Regurgitation
Operative Indications for Descending Aorta

- Size greater than 6.0 cm
- >5.5 cm connective tissue disorders, familial history aneurysm/dissection
  - Median size 5.4 cm in Type B dissection as indication is usually aneurysmal enlargement of false lumen
- Enlargement > 0.5 cm/6-12 months
- Symptomatic aneurysm
  - Persistent pain, rupture, visceral malperfusion,

Operative Indications for Aortic Dissections

<table>
<thead>
<tr>
<th>Dissection type</th>
<th>Operative Indication</th>
</tr>
</thead>
<tbody>
<tr>
<td>Acute</td>
<td></td>
</tr>
<tr>
<td>Type A</td>
<td>Presence</td>
</tr>
<tr>
<td>Type B</td>
<td>Rupture</td>
</tr>
<tr>
<td></td>
<td>Malperfusion</td>
</tr>
<tr>
<td></td>
<td>Progressive dissection</td>
</tr>
<tr>
<td></td>
<td>Failure of medical management</td>
</tr>
<tr>
<td>Chronic</td>
<td></td>
</tr>
<tr>
<td>Type A</td>
<td>Symptoms related to dissection (congestive failure, angina, aortic regurgitation, stroke, pain)</td>
</tr>
<tr>
<td></td>
<td>Malperfusion</td>
</tr>
<tr>
<td></td>
<td>Aneurysm</td>
</tr>
<tr>
<td>Type B</td>
<td>Symptoms related to dissection</td>
</tr>
<tr>
<td></td>
<td>Malperfusion</td>
</tr>
<tr>
<td></td>
<td>Aneurysm</td>
</tr>
</tbody>
</table>

Tsai et al, Circulation 2005
Aortic Syndrome Classification Schema

Aortic Dissection Classification

- **Classification**
  - Multiple systems
  - All based on location of intimal tear
  - DeBakey & Stanford classifications used most frequently
  - Stanford A:
    - Any dissection involving ascending aorta no matter primary tear
  - Stanford B:
    - Dissection involves only the descending aorta
### Crawford Classification

<table>
<thead>
<tr>
<th>Extent</th>
<th>Origin and Location</th>
</tr>
</thead>
<tbody>
<tr>
<td>I</td>
<td>Distal to L SCA to above renal arteries</td>
</tr>
<tr>
<td>II</td>
<td>Distal to L SCA to below renal arteries</td>
</tr>
<tr>
<td>III</td>
<td>6th IC space to below renal arteries</td>
</tr>
<tr>
<td>IV</td>
<td>12th IC space to iliac bifurcation</td>
</tr>
<tr>
<td>V</td>
<td>Below 6th IC space to above the renal arteries</td>
</tr>
</tbody>
</table>

### Penetrating Arterial Ulcers

#### Intramural Hematoma

Advanced imaging has defined precursors to aortic dissection/rupture

Likely all part of a continuum

*Tsai et al, Circulation 2005*
Intramural Hematoma

• Collection of blood within the wall of the aorta without an identified intimal tear

• Proposed pathology:
  – Vasovasorum rupture/aortic media abnormality
  – Continuum of aortic dissection: noncommunicating aortic dissection with thrombosed false lumen

Penetrating Arterial Ulcer

• Deep ulceration of atherosclerotic plaques can lead to:
  – IMH
  – Aortic Dissection
  – Perforation
  – Pseudoaneurysm

• Treatment of IMH/PAU based on aortic dissection classification
Operative Techniques and Outcomes

Pioneers of Aneurysm & Dissection Surgery

Operative Mortality 60%

Cooley DA, DeBakey ME JAMA 1956; 162:1158
Era of Modern Aneurysm Surgery

Bentall-Bono Procedure

Composite AVR
- Aortic Valve
- Coronary button (Bono modification)
- Asc Ao Graft
Ascending Aorta and Hemiarch Replacement

Total Arch and Elephant Trunk Replacement
Valve-sparing Aortic Root Replacement

Remodeling (Yacoub)  Reimplantation (David)

Repair of Type B Dissection Thoracoabdominal Aneurysm
Outcomes of Aortic Dissection

- Operative mortality
  - Type A: 7 -12%
  - Type B: 35-75%

- Surgical Long-term Survival
  - 1, 5, 10, 15 year
  - Type A: 67%, 55%, 37%, & 24%
  - Type B: 56%, 48%, 29%, & 11%

- Medical Long-term Survival
  - Type B: 73%, 58%
  - no significant survival difference versus surgical therapy

Tsai et al, Circulation 2005

Thoracic Endovascular Aneurysm Repair

T horacic
E ndo-
V ascualr
A neurysm
R epair
Endovascular Repair

- Appealing alternative option given open repair mortality 35-75%
- Many of those dying in medical therapy arm did die from complications of the dissection
- **Goals of Endovascular Repair**
  - Reconstruction of segment containing entry tear
  - Induction of thrombosis of false lumen
  - Reestablishment of true lumen and side-branch flow

Endovascular Devices

- MEDTRONIC TALENT
- Stanford Device
- GORE TAG
- COOK TX2
Five-Year Results of Endovascular Treatment with the Gore TAG Device Compared with Open Repair of Thoracic Aortic Aneurysms

<table>
<thead>
<tr>
<th></th>
<th>TEVAR</th>
<th>Open</th>
</tr>
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<tbody>
<tr>
<td>Operative Mortality</td>
<td>2.1%</td>
<td>11.7%</td>
</tr>
<tr>
<td>5-year Survival</td>
<td>68%</td>
<td>67%</td>
</tr>
<tr>
<td>Paraplegia/Paraparesis</td>
<td>2.8%</td>
<td>13.8%</td>
</tr>
<tr>
<td>Stroke</td>
<td>3.5%</td>
<td>4.3%</td>
</tr>
<tr>
<td>Major Adverse Events (MAE)</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Vascular complications</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Endoleaks</td>
<td>28%</td>
<td>70%</td>
</tr>
<tr>
<td>LOS</td>
<td>5-7 days</td>
<td>14-26 days</td>
</tr>
<tr>
<td>Return to Activity</td>
<td>30 days</td>
<td>8 months</td>
</tr>
</tbody>
</table>


Endoleaks

<table>
<thead>
<tr>
<th></th>
<th>Description</th>
</tr>
</thead>
<tbody>
<tr>
<td>I</td>
<td>Leak at attachment site</td>
</tr>
<tr>
<td></td>
<td>A Proximal end</td>
</tr>
<tr>
<td></td>
<td>B Distal end</td>
</tr>
<tr>
<td></td>
<td>C Iliac occlusion site</td>
</tr>
<tr>
<td>II</td>
<td>Flow from patent branch vessels</td>
</tr>
<tr>
<td></td>
<td>A Simple (1 branch)</td>
</tr>
<tr>
<td></td>
<td>B Complex (&gt;2 branches)</td>
</tr>
<tr>
<td>III</td>
<td>Graft Defect</td>
</tr>
<tr>
<td></td>
<td>A Leak at junction or disconnect of graft</td>
</tr>
<tr>
<td></td>
<td>B Graft disruption</td>
</tr>
<tr>
<td>IV</td>
<td>Graft Wall porosity</td>
</tr>
<tr>
<td>V</td>
<td>Endotension (aneurysm expansion-no endoleaks)</td>
</tr>
</tbody>
</table>
Randomized Comparison of Strategies for Type B: Aortic Dissection: Investigation of STEnt Grafts in Aortic Dissection Trial [INSTEAD]

- Role of TEVAR in improving outcomes of uncomplicated Type B dissection unknown
- Persistent false-lumen perfusion is a risk factor for adverse outcomes
- Uncomplicated chronic Type B aortic dissections

Nienaber et al., Circulation 2009

Randomized Comparison of Strategies for Type B: Aortic Dissection: Investigation of STEnt Grafts in Aortic Dissection Trial

- End-points
  - All-cause mortality at 2 years
  - Aorta-related death
  - Progressive aortic pathology
    - Additional surgery
Randomized Comparison of Strategies for Type B: Aortic Dissection: Investigation of STEnt Grafts in Aortic Dissection Trial

- No significant difference in mortality at 2 years
- No difference in aorta-related mortality
- Significant decreases in false-lumen diameter and rates of false-lumen thrombosis

Randomized Comparison of Strategies for Type B: Aortic Dissection: Investigation of STEnt Grafts in Aortic Dissection Trial

- Endovascular therapy failed to improve 2-year survival rate
  - Spinal injury complication: 2.8% vs 15%
- TEVAR definitely influences aortic remodeling
  - Study underpowered to evaluate mortality end-point
  - Mortality benefits not likely to be seen at 2 years
  - Remodeling may modulate late death related to Type B dissection
    - Aneurysm development/Late rupture 20-50% by 5 years
Conclusion

• Aortic Syndromes are increasing in incidence
• Diagnosis remains elusive
• Aortic Dissection is associated with high rates of morbidity & mortality with little change over the decades since repair became feasible
• Mid-term results of TEVAR with aneurysms are encouraging
• Mid-term results of TEVAR with aortic dissection are unclear
• Early referral for mildly dilated aortas or medically managed dissections/old dissections

Common questions

• Medication regimens
• Exercise limitations