Current Therapy for Thoracic Aortic Disease

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OBJECTIVES

• Review established guidelines for treatment of thoracic aortic disease
• Update on evaluation and diagnosis of aortic aneurysm and dissection
• Discussion of new and evolving techniques in surgical treatment of thoracic aortic disease
Thoracic Aortic Disease

- Thoracic aortic disease is usually asymptomatic until something acute and catastrophic occurs
- Imaging is the cornerstone of therapy
  - Expense, radiation, contrast
  - Identification of higher risk individuals
  - Intervene before the catastrophe
Aortic Anatomy

- **Root** = aortic annulus and sinuses to the level of the sinotubular junction
- **Ascending** = sinotubular junction to innominate artery
- **Arch** = innominate artery to left subclavian
- **Descending** = left subclavian to diaphragm
Aortic Anatomy

- Ascending
- Arch
- Root
- Descending
The Aorta – Size Matters

• The magic number is 4

Table 3. Normal Adult Thoracic Aortic Diameters

<table>
<thead>
<tr>
<th>Thoracic Aorta</th>
<th>Range of Reported Mean (cm)</th>
<th>Reported SD (cm)</th>
<th>Assessment Method</th>
</tr>
</thead>
<tbody>
<tr>
<td>Root (female)</td>
<td>3.50 to 3.72</td>
<td>0.38</td>
<td>CT</td>
</tr>
<tr>
<td>Root (male)</td>
<td>3.63 to 3.91</td>
<td>0.38</td>
<td>CT</td>
</tr>
<tr>
<td>Ascending (female, male)</td>
<td>2.86</td>
<td>NA</td>
<td>CXR</td>
</tr>
<tr>
<td>Mid-descending (female)</td>
<td>2.45 to 2.64</td>
<td>0.31</td>
<td>CT</td>
</tr>
<tr>
<td>Mid-descending (male)</td>
<td>2.39 to 2.98</td>
<td>0.31</td>
<td>CT</td>
</tr>
<tr>
<td>Diaphragmatic (female)</td>
<td>2.40 to 2.44</td>
<td>0.32</td>
<td>CT</td>
</tr>
<tr>
<td>Diaphragmatic (male)</td>
<td>2.43 to 2.69</td>
<td>0.27 to 0.40</td>
<td>CT, arteriography</td>
</tr>
</tbody>
</table>

Circulation 2010;121:1551
Genetics of Aortic Disease

- Marfan syndrome = fibrillin-1
- Loeys-Dietz syndrome = TGFBR
- Ehlers-Danlos syndrome = COL3A1
- Familial aortic disease = vascular smooth muscle cell contractile dysfunction
  - Actin (ACTA2)
  - Myosin (MYH11)
Genetics of Aortic Disease

- Familial thoracic aortic disease
  - About 20% have affected relative
  - Most common gene alteration is ACTA2 (10-14% of families)
  - Autosomal dominant inheritance
  - Decreased penetrance in women
  - Presence of genetic mutation but lack of syndrome features
Genetics of Aortic Disease

• Bicuspid aortic valve
  • The most common congenital heart defect (0.9-1.4% of population)
  • Possible autosomal dominant pattern in certain families
  • Loss of elastic fibers, cystic medial necrosis, and altered smooth muscle alignment in aorta
• 9x increase in aortic dissection
Screening – Genetic Aortic Disease

- Obtain a careful family history if you suspect genetic aortic disease.
- Most patients do *not* need expensive serologic or tissue testing.
- CT and echocardiography are the mainstay of aortic imaging.
- Prompt referral to aortic center is indicated for any suspicious patient.
Screening – Familial Aortic Disease

- **Class I**: Aortic imaging is recommended for first-degree relatives of patients with aneurysm or dissection.

- **Class IIa**: If one or more first-degree relatives is found to have aneurysm or dissection, imaging of second-degree relatives is reasonable.
Screening – Bicuspid Aortic Valve

- Class I: First-degree relatives of patients with a bicuspid aortic valve, premature onset of thoracic aortic disease with minimal risk factors, and/or a familial form of thoracic aortic aneurysm and dissection should be evaluated for the presence of a bicuspid aortic valve and asymptomatic thoracic aortic disease.

- Class I: All patients with a bicuspid aortic valve should have both the aortic root and ascending thoracic aorta evaluated for evidence of aortic dilatation.
Indications for Ascending Aorta Replacement

• Class I indications
  • Symptomatic
  • Size:
    • Ascending 5.0-5.5 cm
      • Marfan, bicuspid 4.0-5.0 cm
    • Family history of dissection
    • Rapid growth (>0.5 cm/year)
    • Concomitant AVR and ascending > 4.5 cm
Indications for Aortic Aneurysm Replacement

- AHA Guidelines for aortic pathology beyond the ascending aorta
  - Arch = 5.5 cm or greater
  - Descending chronic dissection = open repair when 5.5 cm or greater
  - Descending degenerative or traumatic = endovascular repair
  - Thoracoabdominal = 6.0 cm
Surveillance of Aortic Disease - AHA guidelines

- CT or MRI is appropriate
- Repeat imaging for aortic dissection at 1 month, 3 months, 6 months, and annual
- Use same modality and same institution where possible
- MRI probably preferred in stable disease to reduce radiation over time
Medical Management

- Careful history: family history, bicuspid aortic valve, cerebral aneurysms, sudden/unexplained death
- Surveillance imaging
- β-blockers
- ACE/ARB
  - Losartan clinically decreases TGFβ signalling
Ascending Aneurysm
Ascending Aneurysm – Surgical Repair

Doty DB and Doty JR, Cardiac Surgery: Operative Technique, 2012
Ascending Aorta - Current Outcomes

• Hess et al (Florida)
• 44 patients underwent replacement ascending aorta/hemiarch
• Mean HCA = 12 minutes
• No operative mortality, stroke, or renal dysfunction
Aortic Root Aneurysm
Aortic Root Aneurysm – Surgical Repair

Doty DB and Doty JR, Cardiac Surgery: Operative Technique, 2012
Aortic Root and Ascending Aneurysm
Root + Ascending Aorta – Surgical Repair

Doty DB and Doty JR, Cardiac Surgery: Operative Technique, 2012
Root + Ascending Aorta – Current Outcomes

- Doty et al (Utah)
- 50 patients underwent replacement of root + ascending aorta/hemiarch
- Mean HCA = 13 minutes
- No operative mortality or stroke
- 98% freedom from reoperation
- No further aortic pathology
Marfan Aortic Root
Valve-Sparing Root Replacement

Doty DB and Doty JR, Cardiac Surgery: Operative Technique, 2012
Valve-Sparing Root Replacement
Valve-Sparing Root - Current Outcomes

- Cameron et al (Johns Hopkins)
- 19-year experience with valve-sparing root replacement for Marfan syndrome
  - 127 patients
  - 1 death
  - No reoperations with David I
  - Very low thromboembolism and endocarditis rates
Arch Aneurysm
Arch Aneurysm – Reimplantation Repair

Doty DB and Doty JR, Cardiac Surgery: Operative Technique, 2012
Aortic Arch - Current Outcomes

- Safi et al (Houston)
- 1316 arch repairs over 20-year period
- Operative mortality 10%
- Stroke 3%
- Higher risk in emergent, older, acute dissection, and ruptured aorta
Descending Aneurysm
Descending Aneurysm – Surgical Repair

Doty DB and Doty JR, Cardiac Surgery: Operative Technique, 2012
Descending Aneurysm – Surgical Repair

Doty DB and Doty JR, Cardiac Surgery: Operative Technique, 2012
Elefteriades et al (Yale)
130 patients underwent open surgical repair of descending thoracic aorta
Operative mortality 3%
Stroke 4%; paraplegia 7%
5-year survival 73%
Thoracoabdominal Aneurysm
Thoracoabdominal Aneurysm Open Repair
Thoracoabdominal Aneurysm Open Repair

Doty DB and Doty JR, Cardiac Surgery: Operative Technique, 2012
Thoracoabdominal Aorta - Current Outcomes

- Kouchoukos et al (St. Louis)
- 218 patients
- Operative mortality 5%
- Spinal cord injury 3.5%
- Stroke 4%
- Renal failure 7%
Thoracic Endovascular Aortic Repair (TEVAR)

- Potential benefits:
  - Less invasive
  - Avoidance of CPB/HCA
  - Quicker recovery
  - Reduction in stroke and paralysis

- Disadvantages:
  - Failure to seal (endoleak)
  - Inability to treat entire pathology in areas of branching or extensive aortic disease
Thoracic Endovascular Aortic Repair (TEVAR)

- Proven and preferred therapy for descending thoracic aneurysms and chronic dissections
- Recent FDA approval for treatment of acute type B aortic dissections
- Growing interest in hybrid and extended applications to complex aortic pathology
Descending Aneurysm - Endovascular Repair

Doty DB and Doty JR, Cardiac Surgery: Operative Technique, 2012
TEVAR – CT Scan
Descending Aneurysm - Endovascular

- Bavaria et al (Pennsylvania)
- 140 patients underwent endovascular stent-grafting of descending aneurysm
- Operative mortality 2%
- Spinal cord injury 3%
- No difference in 2-year mortality compared to open surgical cohort
Expanding Endovascular therapy

- New techniques and approaches with goals of decreasing mortality/complications, including long-term development of other aortic problems
- Combining open operations with endovascular strategies
- Example: “hybrid” arch repair
  - Sternotomy with debranching of head vessels
  - Stent-graft into descending aorta
Type A Dissection – Arch Debranching
Hybrid arch debranching and endografting

- Roselli et al (Cleveland)
- 17 hybrid arch repairs for acute type A
  - Portion of stent-graft under arch cut out
  - Stent-graft manually sewn to head vessels and to proximal open graft
- No deaths
- 2 strokes (6%) and 2 paralysis (6%)
Hybrid arch debranching and endografting
Aortic Dissection

Type A

Type B
Aortic Dissection – CT

Type A

Type B
Aortic Dissection – Type A
Aortic Dissection – Type B
Type A Dissection - Current Outcomes

- Pagni et al (Louisville)
- 117 patients open surgical repair of type A dissection 2003-2011
- Operative mortality 19%
- 5-year survival 60%
Type B Dissection - Current Outcomes

- Estrera et al (Houston)
- Medical management = 10% hospital mortality
- Open surgical repair = 17% hospital mortality
- 5-year survival 75%
Acute Aortic Dissection – Type B Endovascular

- DISSECTION trial data - STS 2014
  - 20% presented with acute rupture
  - 100% technical success
  - 100% entry tear coverage
  - 8% all-cause mortality at 30 days
  - False lumen partial or complete thrombosis in 91%
Expanding Endovascular Repair of Aortic Disease

- Aortic dissection
- Chronic type B
- Acute, complicated type B
- Acute, uncomplicated type B
- Penetrating aortic ulcer
- Intramural hematoma
- Aortic transection
Aortic Dissection - Endovascular

- INSTEAD Trial for stable, uncomplicated type B dissection
- OMT vs. OMT + elective stent graft
- No difference in 2-year mortality
- Improved aortic remodeling after stent-grafting (91% vs. 19%)
Intramural Hematoma - AHA Guidelines

• An intramural hematoma should be treated similar to aortic dissection in that portion of the aorta

• Treatment of penetrating aortic ulcer remains controversial, but endovascular repair is a reasonable approach
Spectrum of Dissection

Aortic Dissection → Penetrating Ulcer → Intramural Hematoma
Penetrating Ulcer - Endovascular
Penetrating Ulcer - Endovascular
Intermountain Center For Aortic Disease

- Development of collaborative program for treatment of all forms of aortic and great vessel disease
- Screening and ongoing follow-up of patients with known aortic pathology
- Shared expertise of cardiovascular surgery, radiology, vascular surgery, and cardiology for optimal care
Conclusions

• Once an aortic patient, always an aortic patient
• High index of suspicion for screening
• Early referral for elective care is the safest and most effective treatment - 4.0 cm or larger, please refer early!

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