Management of

The Dilated Ascending Aorta in

Congenital Heart Disease

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AATS Congenital PGC 2013
No disclosures
Q65. What is the most aggressive aortopathy associated with rupture and dissection?

a. Bicuspid aortic valve syndrome
b. Marfan syndrome
c. Loeys-Dietz syndrome
d. Post-surgical repair
The Dilated Ascending Aorta in Congenital Heart Disease

- Bicuspid aortic valve syndrome
- Marfan syndrome
- Loeys-Dietz syndrome
- Turner syndrome
- Postsurgical
  - Conotruncal (TOF/Truncus/DORV)
  - Arterial switch
  - Norwood
  - Ross procedure
Aneurysm Morphology

Sinus

Ascending Aorta

Post-surgical
Complications of
Aortic Dilation (Aneurysm)

- Aortic dissection and rupture
- Aortic valve incompetence
- Compression of nearby structures
Aneurysm size and risk of complications

Elefteriades, Scientific American 2002
The 5 cm Rule

Elefteriades, JTCVS 2010
Bicuspid Aortic Valve

- 1-2% population
- 50-60% have ascending aorta dilatation
- Faster growth rates than trileaflet Ao valves
- Similar risk of rupture, except:
  - Aortic stenosis
  - Coarctation
- Intervention: > 5cm, growth > 0.5 cm/yr, valve pathology
Marfan syndrome aneurysm

- Most common inherited connective tissue disorder
- Ao dilation present at birth
- Principally sinus aneurysm; valve usually competent
- Rupture and dissection rare before age 12 but most common cause of death in adults
- Indications for surgery:
  - Root > 5cm
  - Growth > 1cm/yr
  - Progressive AI
  - dissection
Marfan Syndrome Shortens Life Expectancy by a Third
Surgical options for Marfan aneurysm

Bentall

VSRR: Remodeling

VSRR: Reimplantation
Caveats for Marfan Aneurysm

- Must replace sinuses
- Prophylactic arch replacement not necessary
- Reimplantation provides better anulus stabilization
Loeys-Dietz Syndrome

Hypertelorism

Arterial tortuosity and aneurysm

Bifid uvula
Natural history in 90 LDS patients

<table>
<thead>
<tr>
<th>Description</th>
<th>Details</th>
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<tbody>
<tr>
<td>Mean age at death (n = 22):</td>
<td>26.1 yrs (6m - 43 yrs)</td>
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<tr>
<td>Mean age at first surgery</td>
<td>18.7 yrs (14m - 38 yrs)</td>
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<tr>
<td>Mean age at first dissection</td>
<td>25.6 yrs (6m - 47 yrs)</td>
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<td>Surgery or death in childhood (&lt; 19yrs)</td>
<td>n = 26; 34%</td>
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<tr>
<td>Life-threatening events in pregnancy</td>
<td>7/11 pregnant women (64%)</td>
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<tr>
<td>Life-threatening events in pregnancy</td>
<td>(5 Aortic rupture; 2 uterine ruptures)</td>
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</tbody>
</table>

Cause of death:
- Thoracic aortic dissection: n = 13
- Abdominal aortic dissection: n = 6
- Subclavian artery dissection: n = 1
- Cerebral bleeding: n = 2

Arterial Aneurysms/dissections (n= 129):
- Ascending aorta: n = 68
- Abdominal aorta: n = 9
- Transverse aorta: n = 9
- Descending aorta: n = 9
- Thoracic circulation: n = 19
- Cerebral circulation: n = 9
- Abdominal circulation: n = 6
Loeys-Dietz Syndrome (LDS)

- Surgical approach similar to Marfan syndrome
- Earlier intervention
  - Children: Ao root > 3cm
  - Adults: Ao root > 4cm
- Require thorough imaging of entire vascular tree
Ascending Aorta Size in Turner Syndrome

Matura, Circ 2007

Intervention recommended at >3.5 cm or > 2-2.5 cm/m²
Aneurysms after repair of Congenital Heart Defects

- Conotruncal (TOF/Truncus/DORV)
- Arterial switch
- Norwood
- Ross procedure
Conotruncal Defects

- Virtually all TOF pts have dilated aortas
- Few surgically treated cases in literature (5 TOF, 5 P Atr) and most had >2+ AI
- Usually present > 20 years postop; rupture and dissection both reported
- Risk factors: P atresia, later repair, male
- Indications for surgery unclear
Histopathologic changes in ascending aorta and risk factors related to histopathologic conditions and aortic dilatation in patients with tetralogy of Fallot

Ujjwal K. Chowdhury, MCh, Diplomate NB, Anand K. Mishra, MS, Ruma Ray, MD, MRC (Path),
Mani Kalaivani, MSc (Biostatistics), Srikrishna M. Reddy, MS, and Panangipalli Venugopal, MCh

JTCVS 2008

- Significant fragmentation and loss of lamellar units
- Present from infancy
- Changes progress with age
Root dilation after arterial switch

- Common in late survivors
- Severe AI unusual (<5%)
- ? risk of rupture or dissection (no cases)
Root dilation is Common After the Arterial Switch

Schwartz, Circulation, 2004
Root dilation after Norwood

Common but no cases of rupture or dissection

Patch vs neoaorta?

Jonas, Ann Thor Surg, 2004
Aortic dilation after Ross procedure

- Late aortic dilation is common (both ascending aorta and neo-root)
- Risk factors include root replacement, young age at repair, BAV, Ao aneurysm
- Dissection reported but not rupture
VSRR after the Ross Procedure
(Luciani, JTCVS, 2010)
## Risk of Aortic Rupture or Dissection

<table>
<thead>
<tr>
<th></th>
<th>Infancy</th>
<th>Childhood</th>
<th>Adolescence</th>
<th>Adulthood</th>
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</thead>
<tbody>
<tr>
<td><strong>BAV</strong></td>
<td>−</td>
<td>+/−</td>
<td>+</td>
<td>+</td>
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<tr>
<td><strong>Marfan</strong></td>
<td>+/−</td>
<td>+/−</td>
<td>+</td>
<td>+</td>
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<tr>
<td><strong>Loeys-Dietz</strong></td>
<td>++</td>
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<td><strong>Post-Surgical</strong></td>
<td>−</td>
<td>−</td>
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</table>
Guidelines for Surgery

- Aneurysm > 5cm (except LDS: children 3cm, adults 4 cm)
- Dilation > 0.5cm/yr
- Aneurysm with progressive valve dysfunction
- Dissection (acute or chronic)
Z-scores: A word of caution

- Z-score describes distance from the mean, but not necessarily predictive of adverse events with precision
- At the tail of the curve, small change in measurement may lead to large increase in Z-score
- High Z-scores may lead to over-reaction
Summary

- Aortic aneurysm: a “growing” problem in congenital heart disease and late after repair of congenital heart defects
- Significant heterogeneity in morphology and risk for rupture and dissection among diagnostic groups
- Indications for surgery are not yet evidence based
- Risk of rupture is particularly high in LDS
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