Bicuspid Aortopathy - to replace or not to?
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**SCORE: 1234**
Agenda

I. Status quo (5‘)
II. Evidence (10’)
III. Guidelines (5‘)
IV. New risk markers (10‘)
I. Status quo

- Case Scenario

La pratica deve essere edificata sopra la buona teorica
(Practice must always be founded on sound theory)
Leonardo Da Vinci
I. Clinical Scenario

- 62 y/o male, NYHA II, FH: positive for BAV, one unclear sudden death, CVRF: arterial hypertension
- Bicuspid combined valvular dysfunction with leading stenotic component (4 m/sec)
- Valve Type: one Raphe, fused left-right coronary cusps
- Root Geometry: unbalanced (150 degrees)
- Sinotubular definition
- Aortic annulus: elliptical, max. 29mm – Aortic root: 42mm, Ascending Aorta: 45mm, rate last 2 years: 1mm/year

What to do with the ascending aorta?
Fictional patient – real attendings
I. Clinical Scenario

Attending Surgeon 1
- Thickness of ascending aortic tissue?
- Reduction Plasty

Attending Surgeon 2
- Modified Bentall / CVG

Attending Surgeon 3
- Leave aorta alone, no indication

Attending Surgeon 4
- Leave root alone
- Wheat procedure

➢ Surgeon preference
➢ Tissue „quality“ characteristics?
II. Evidence

- Bicuspid Aortopathy
- Aortopathy Clusters

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Leonardo Da Vinci"
Ascending Aorta – dilated or not?

Upper normal diameter based on patient age:
D (mm) = 31 + 0.16 \times \text{age (years)}

For extreme BMI:
D (mm) = 21 + 0.14 \times \text{age (years)} + (0.41 \times \text{BMI})

20 y/o average 27mm, 34 still “normal”
80 y/o average 37mm, 44 still “normal”

50% increase in Diameter
Ectasia-Aneurysm
20 y/o – 40mm
40 y/o – 45mm
60 y/o – 50mm
80 y/o – 55mm

Ascending Aorta – growth, but how fast?


Bicuspid Aortopathy

The aortopathy of bicuspid aortic valve disease has distinctive patterns and usually involves the transverse aortic arch

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N= 64 BAV patients
CTA or MRA of thoracic aorta
  • Aortoventricular junction
  • Sinuses of Valsalva
  • Sinotubular junction
  • Tubular ascending aorta
  • Proximal to innominate artery
  • Distal to innominate artery
  • Proximal to LSCA
  • Distal to LSCA
  • Proximal descending aorta

Bicuspid Aortopathy – related to valve type?

III. Guidelines

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III. Guidelines

Decision in aortic diameters **50-55mm** based on
- Patient age
- Body size
- Comorbidities
- Type of surgery
- Risk factors:
  - Family history
  - Hypertension
  - Coarctation
  - Rapid growth > 2mm/year

Aortic diameters $\geq 55\text{mm}$ surgery should be performed irrespective of valve function

In cases of BAV, surgery of the ascending aorta is indicated in case of:

- **aortic root** or ascending aortic diameter >55 mm.
- aortic root or ascending aortic diameter >50 mm in the presence of other risk factors.
- aortic root or ascending aortic diameter >45 mm when surgical aortic valve replacement is scheduled.

I. Back to Clinical Scenario

- 62 y/o male, NYHA II, FH: positive BAV, one unclear sudden death, CVRF: arterial hypertension
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- Valvular indication: ascending aorta indicated
- Without valvular indication: ascending indicated (risk factors)
- With or without valvular indication: root not indicated (?)
IV. New Markers
- Serum Enzyme Ratios
- Biomechanics
IV. New Markers: Serum Enzyme Ratios

- MMP serum levels not related to aneurysm size
- Different MMP / TIMP ratios in aneurysms of different sizes

IV. New Markers: Serum Enzyme Ratios

IV. Biomechanics: Flow Patterns

IV. Biomechanics - WSS

IV. Biomechanics: Flow Patterns

- Normalized displacement of flow from vessel center
- 25 patients, no significant valve dysfunction, F-U 4.5 years
- Faster aortic growth in pts with displaced flow

IV. Biomechanics - Geometry

- Angle LV/aorta significantly larger in the R/L group than in the R/N group
- LV/aorta angle indicator of indexed aortic diameter
- Angle LV/aorta and angle jet/aorta predictive of mid-ascending phenotype

Take Home Messages

- Diameters + Growth rate + Risk factors
- All guidelines: level of evidence C
- No differentiation between root / ascending aorta
- Serum Enzyme Ratios
- Biomechanics: Flow patterns, shear stress, root geometry, valve type
Bicuspid Aortopathy - to replace or not to?

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Stress – MMP2/TIMP1-4 - Stiffness

Hypotheses

- Stretch regulates MMP-2 activity and TIMP-1-4 protein levels in ascending aortic tissue
- Modulation of MMP-2 activity in human and murine *ex vivo* tissue is sufficient to alter tissue stiffness and regulate TIMP-1-4 protein expression
- TIMP-1-4 are released from aortic tissue and their serum levels are related to aortic mechanical properties

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IV. New Markers: Serum Enzyme Ratios

III. Guidelines

III. Guidelines