PHENOTYPES-GUIDED SURGICAL TREATMENT OF BICUSPID AORTOPATHY

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BAV is not just a peculiar valve morphology; it is a disease of the ascending aorta
Assessing wall stresses in bicuspid aortic valve-associated aortopathy: Forecasting the perfect storm?

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Image: Diagram showcasing abnormal WSS and high wall stress, with possible sequence of events involving WSS, diameter, WSS, WS, and dissection.
“CLINICALLY NORMAL” BAV PHENOTYPES AND AORTOPHATY

Altered flow start a process of negative remodeling that might depend on the specific valve phenotype

Message no. 1
DIRECT SURGICAL CONSIDERATION

If the hemodynamic theory is the main etiology, AVR should stop the progression of aortic enlargement

But when is too late?
THE GENETIC FACTOR

• Aortic enlargement already present at young age

• Incidence of dissection at smaller diameters

• Presence of familiar clusters

• Mutations in the NOTCH1 gene
DIRECT SURGICAL CONSIDERATION

If the genetic theory is the main ethiology, more aggressive aortic replacement is warranted.
Root phenotype at presentation, not absolute baseline diameter, was an independent predictor of fast progression (>0.9 mm/year) for the ascending tract (OR: 14; p = 0.001). Fast growth was rarely seen in patients with the RL morphotype and ascending phenotype (6% at the root and 10% at the ascending level). In patients with BAV, the root phenotype may be a marker of more severe aortopathy, warranting closer surveillance and earlier treatment. The more common ascending phenotype proved to be a more stable disease entity, generally with slower progression.
It appears that the root phenotypes is more prone to progress into the ascending aorta rather than the ascending aorta progressing into the root.

Ascending phenotype

Root phenotype

Hemodynamic theory

Genetic theory

Message no. 2
Root phenotype (type III, N) > Ascending phenotype (Type II, A) > Non dilated phenotype

Message no. 3
BAV stenosis (often R/N) is more often associated with Ascending phenotype

Normal valve or some degree of regurgitation (often R/L) more often associated with Root phenotype

Message no. 4
GENERAL CONSIDERATIONS

- AVR (specially for AS) >> more conservative with the aortopathy

- Valve-sparing and/or leaflets repair >> more aggressive with the aortopathy

Message no. 5
TAILORED SURGICAL APPROACH
Usually older, R/N fusion, valve stenosis or normally functioning
AORTOPATHY TYPE I  
(MOST FREQUENTLY IN R/N FUSION)

AORTOPATHY TYPE II  
(MOST FREQUENTLY IN L/R FUSION)
AORTOPATHY TYPE I
(MOST FREQUENTLY IN R/N FUSION)

Characteristics:
- Preserved ST junction
- Tubular dilatation
- Normal root dimension
ASCENDING PHENOTYPE (TYPE I AORTOPHATATY)

- If surgical indication is for AS
  Wheat procedure if ascending is above 45 mm

- If surgical indication is for the ascending (50 mm)
  Replace at the STJ and spare/repair the valve
Aortic root 42.3 mm
ST-junction 33.1 mm
AA 53 mm
Aortic root 33 x 39.1 mm
ST-junction 28 mm (graft)
THE DANGER OF REMODELING THE ST JUNCTION

What works for tricuspid valve does not work for bicuspid!
AORTOPATHY TYPE II
(MOST FREQUENTLY IN L/R FUSION)

Characteristics:
- Normal or Effaced ST junction
- Sometimes NC sinus more dilated
ASCENDING PHENOTYPE (TYPE II AORTOPHATY)

If surgical indication is for AS

Bentall operation if root > 45 mm

• If surgical indication is for the ascending (50 mm) or AR

  Symmetrical root dilatation

  Valve sparing

  Asymmetrical root dilatation

  Single sinus replacement
WHEN TO REPLACE THE ROOT

• Preserved STJ
  > 45 mm

• Effaced STJ (type II, N or E)

• Asymmetric sinus dilatation
  40-45 mm
ASYMMETRIC DILATATION IN BAV

Type 0 or 1, fusion L/R with a large NC sinus
Non dilated annulus
Aortic root 45.4 x 36.9 mm

NC sinus 27 mm
Aortic root 34.9 x 30.6 mm
NC sinus 17 mm
ROOT PHENOTYPE

Usually male, young, L/R fusion with normal functioning or regurgitant valve
AORTOPATHY TYPE III
YOUNG MALE, DILATED ANNULUS)

AORTOPATHY TYPE II
(MOST FREQUENTLY IN L/R FUSION)
AORTOPATHY TYPE III
(ROOT PHENOTYPE, YOUNG MALE, DILATED ANNULUS)

Similar to Marfans usually with a dilated annulus (50 mm)

Bentall or Valve sparing procedure

Reimplantation

Always replace the root

In this peculiar type of BAV aoropathy the genetic might play a predominant role
Often these young patients reach the surgical indication because severe AR when the root diameter is well below 40 mm.

Valve repair or Always reimplantation ??
40yrs old
Although apparently is a non dilated phenotype
Max root diameter is 40

Valve sparing or Valve repair?
CONCLUSIONS

• Several interventional options are available for patients with BAV disease and aortic dilatation

• Root phenotype and associated AR need to be addressed more aggressively

• Planning and tailoring the operation to the single patients based on the actual knowledge is a good practice.