

6th International Meeting on Aortic Diseases

New insights into an old problem CHU Liège, APF

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PHENOTYPES-GUIDED SURGICAL TREATMENT OF BICUSPID AORTOPATHY

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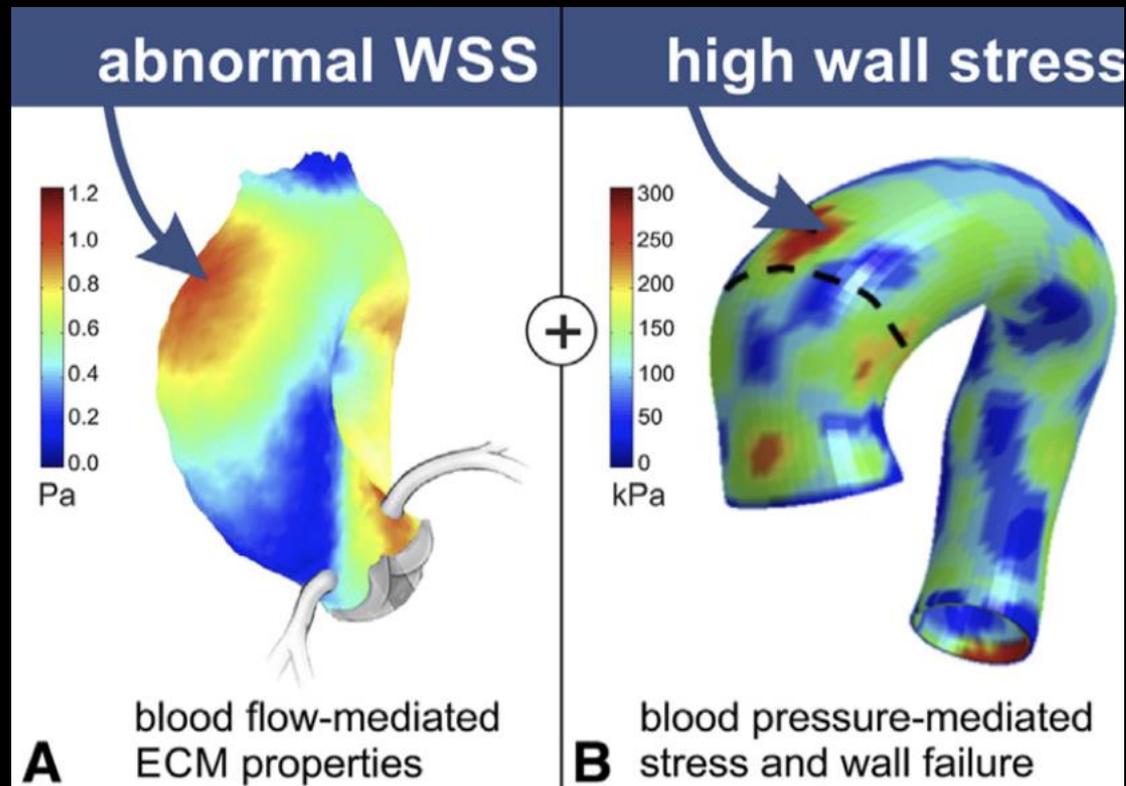


BICUSPID AORTIC VALVE

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?

Alex J. Barker, PhD,^a Michael Markl, PhD,^{a,b} and Paul W. M. Fedak, MD, PhD, FRCSC^{c,d}



“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

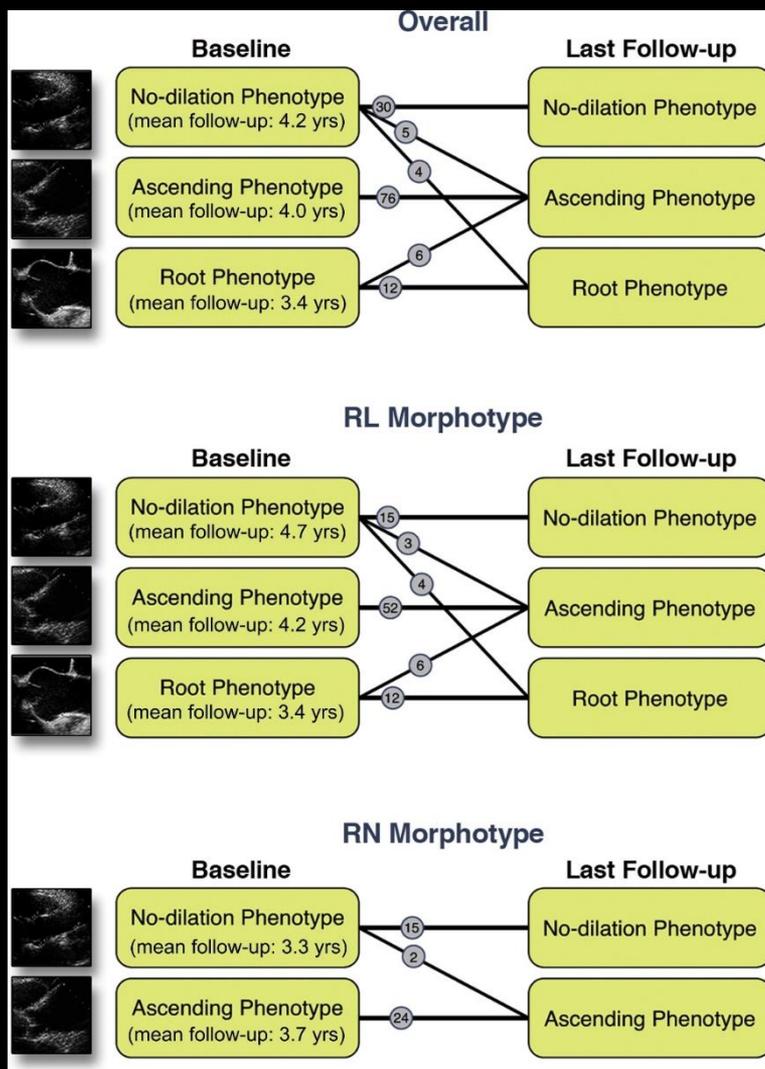
- Aortic enlargement already present at young age
- Incidence of dissection at smaller diameters
- Presence of familiar clusters
- Mutations in the NOTCH1 gene

DIRECT SURGICAL CONSIDERATION

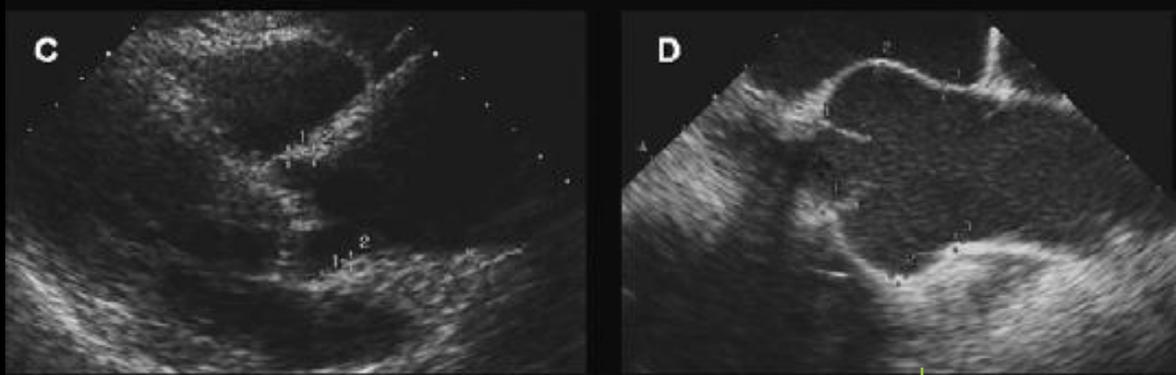
If the genetic theory is the main ethiology,
more aggressive aortic replacement is
warranted

Pattern of Ascending Aortic Dimensions Predicts the Growth Rate of the Aorta in Patients With Bicuspid Aortic Valve

A Della Corte et al. *J Am Coll Cardiol Img.* 2013;6(12):1301-1310



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

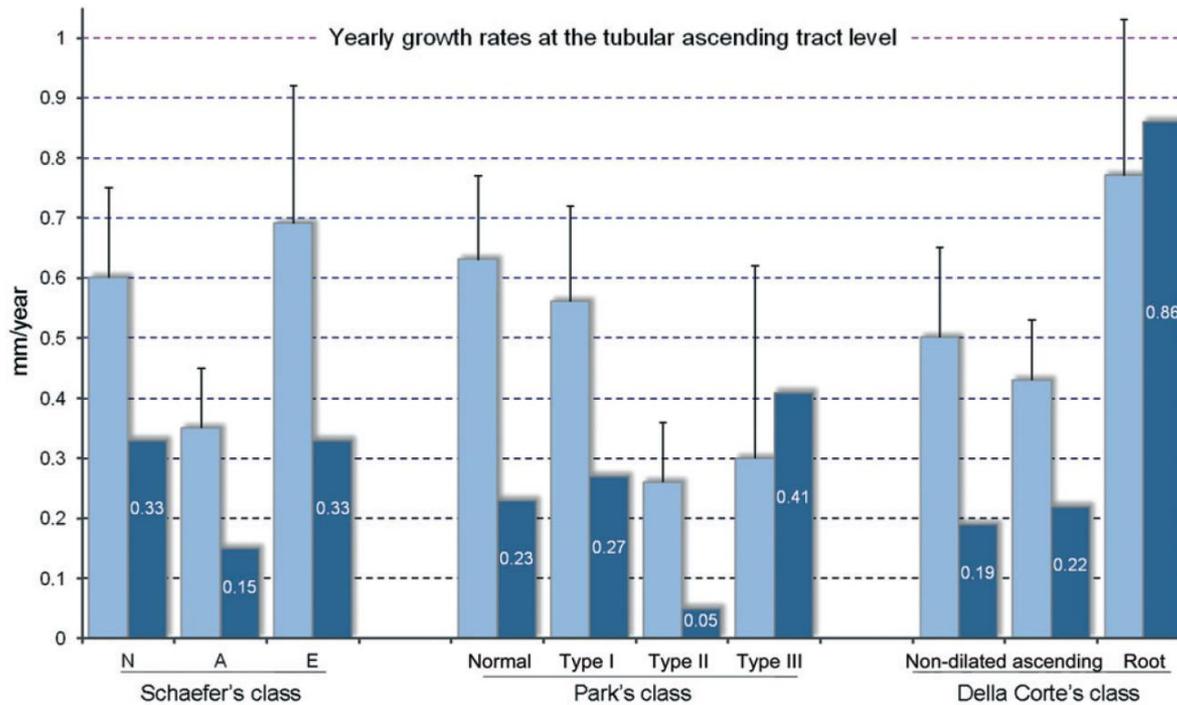


Figure 2: Differences in mean and median growth rates in different subgroups of BAV patients distinguished according to the three classification schemes. Light bars represent mean growth rate (\pm standard error of the mean) at the tubular ascending tract, dark bars represent the median values.

Root phenotype (type III, N) > Ascending phenotype
(Type II, A) > Non dilated phenotype

Message no. 3

Table 2

Univariate associations between BAV functional status and other clinical variables and prevalence of aortic dilation (AR > 1.1)

	Prevalence of ascending dilation (%)	<i>p</i> value	Prevalence of root dilation (%)	<i>p</i> value
Pts with \geq moderate stenosis	79	0.47	38	<0.001
Pts without \geq moderate stenosis	78		68	
Pts with \geq moderate regurgitation	84	0.03	68	0.003
Pts without \geq moderate regurgitation	75		51	
Pts with normal valve function	74	0.28	62	<0.001
Pts with predominant stenosis	79		39	
Pts with predominant regurgitation	84		74	
Pts with any degree of stenosis	81	0.24	47	<0.001
Pts with no degree of stenosis	75		70	
Pts with any degree of regurgitation	78	0.54	60	0.10
Pts with no degree of regurgitation	79		51	

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

ASCENDING PHENOTYPE

Schaefer's class

A shape

A shape

E shape

Park's class

Type I dilation

Type II dilation

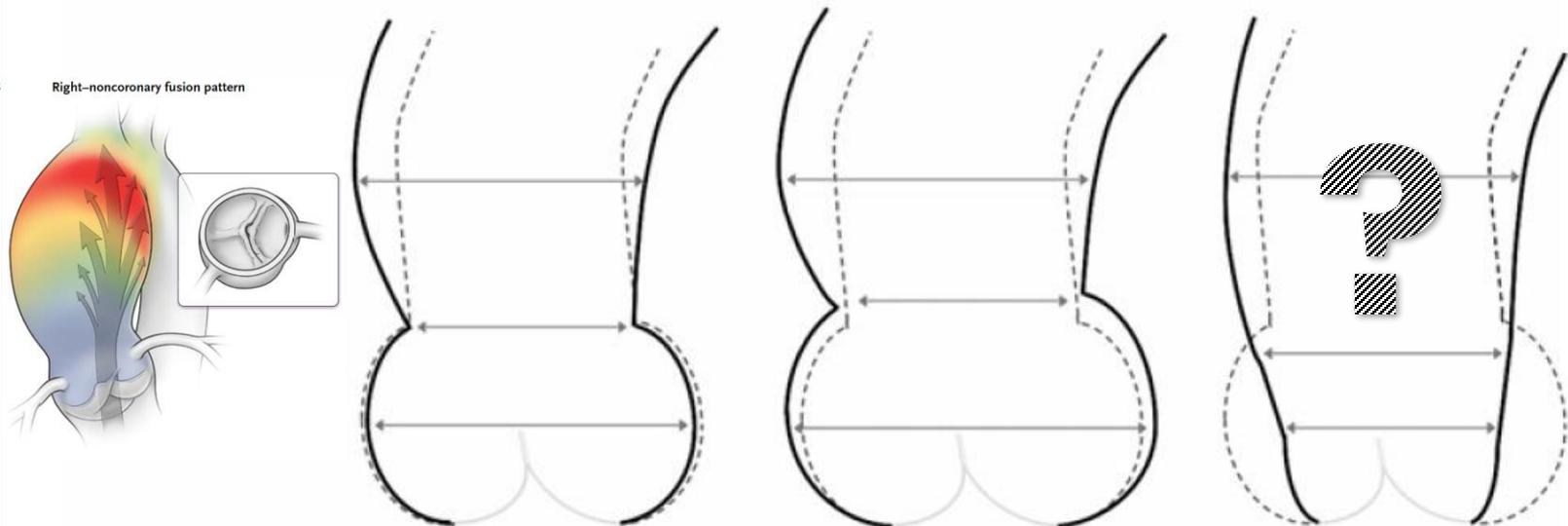
Type I dilation

Della Corte's class

Ascending phenotype

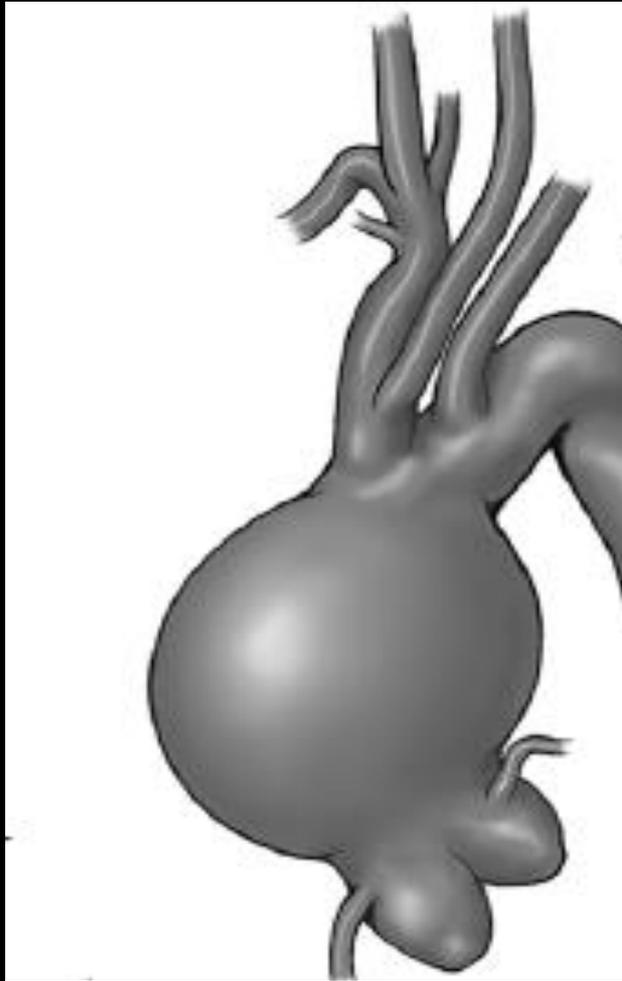
Ascending phenotype

Ascending phenotype

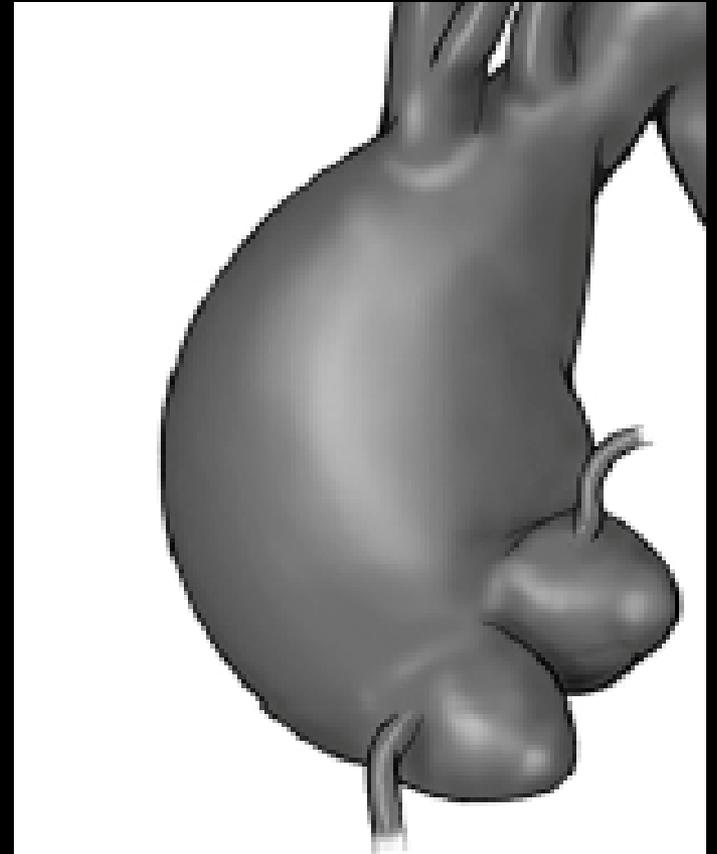


Usually older, R/N fusion, valve stenosis or normally functioning

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

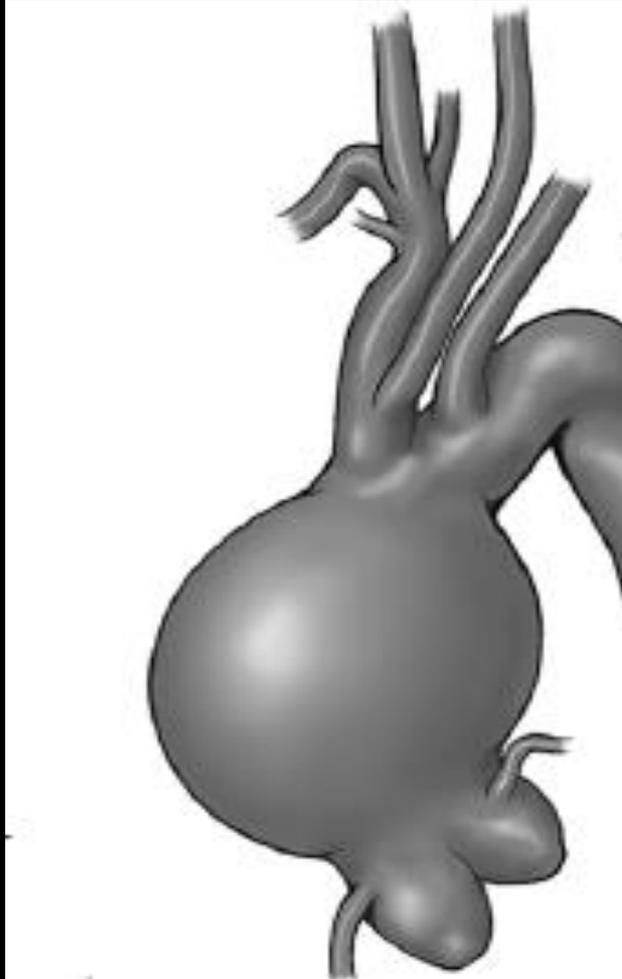


**AORTOPATHY TYPE II
(MOST FREQUENTLY IN L/R FUSION)**



AORTOPATHY TYPE I

(MOST FREQUENTLY IN R/N FUSION)



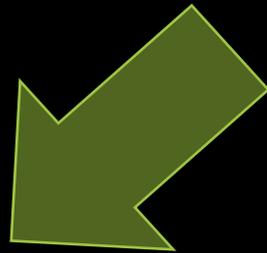
Characteristics:

Preserved ST junction

Tubular dilatation

Normal root dimension

ASCENDING PHENOTYPE (TYPE I AORTOPHATY)



- If surgical indication is for AS

What procedure if ascending is above 45 mm

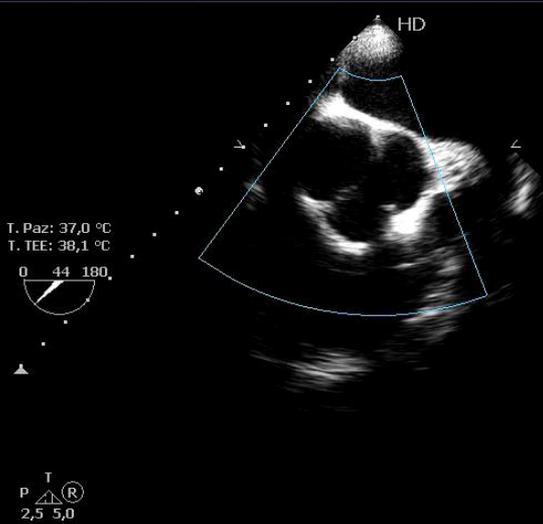
- If surgical indication is for the ascending (50 mm)

Replace at the STJ and spare/repair the valve



Aortic root 42.3 mm
ST-junction 33.1 mm
AA 53 mm

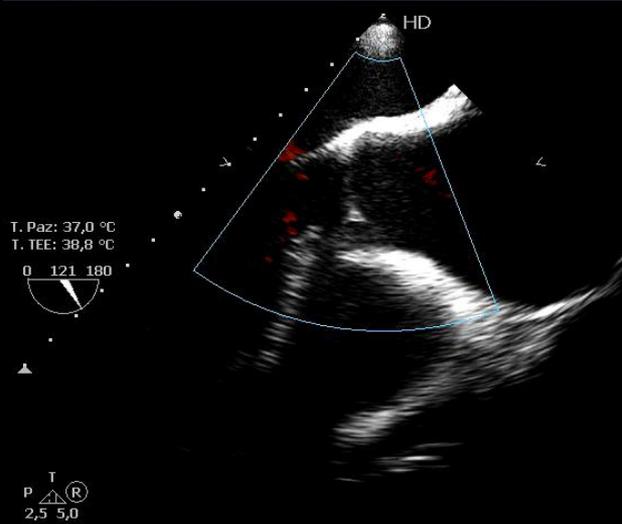
ceci, girolamo
11-03-15-151341



15/03/2011 PHILIPS
15.14.08

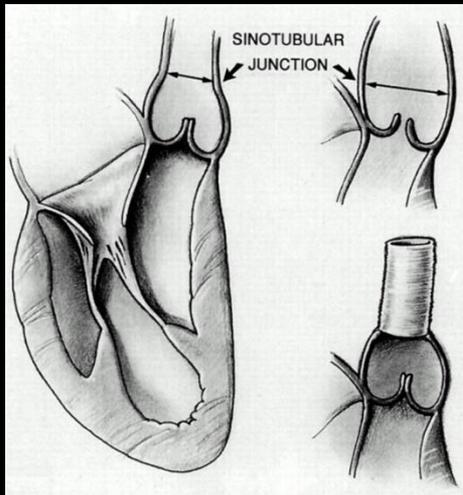


ceci, girolamo
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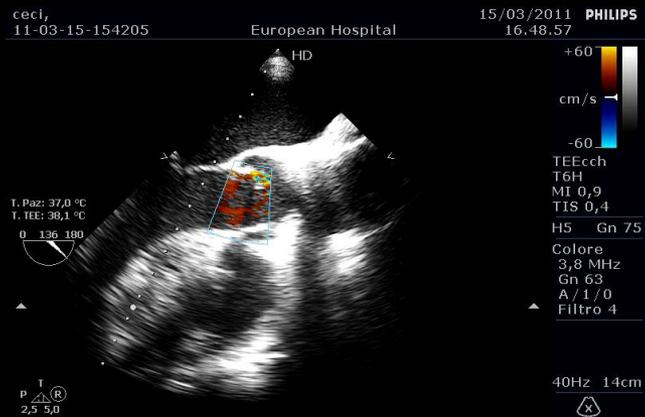


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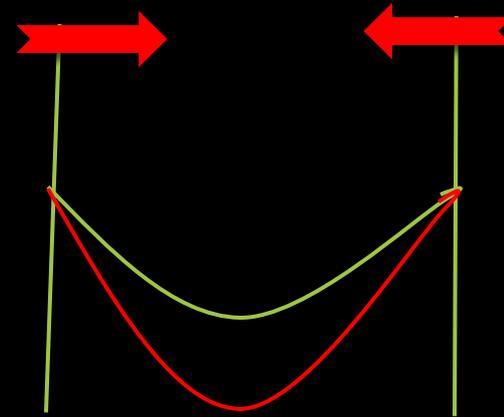
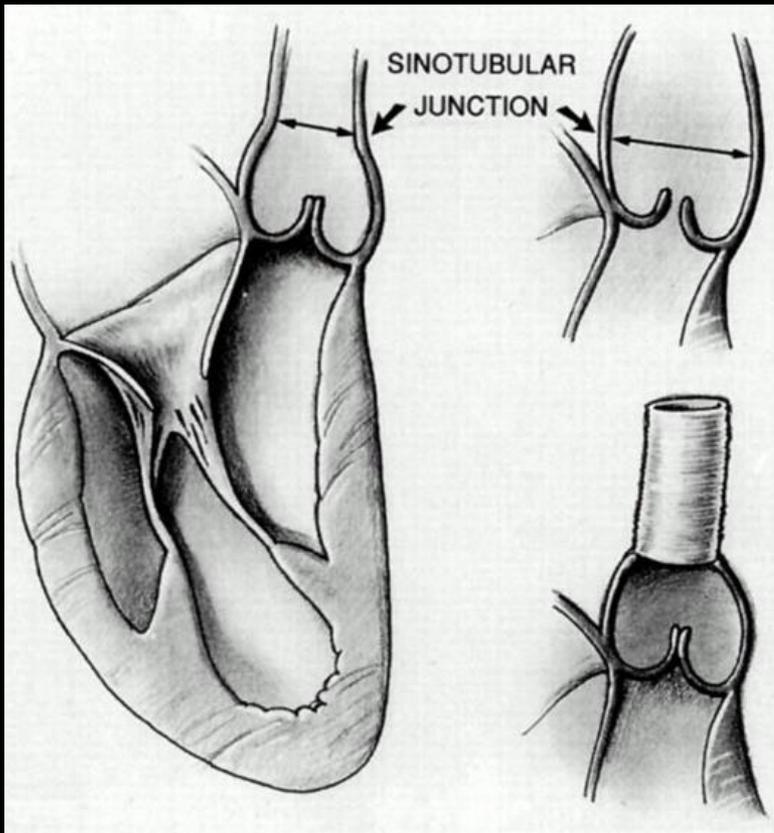




Aortic root 33 x 39.1 mm
ST-junction 28 mm (graft)

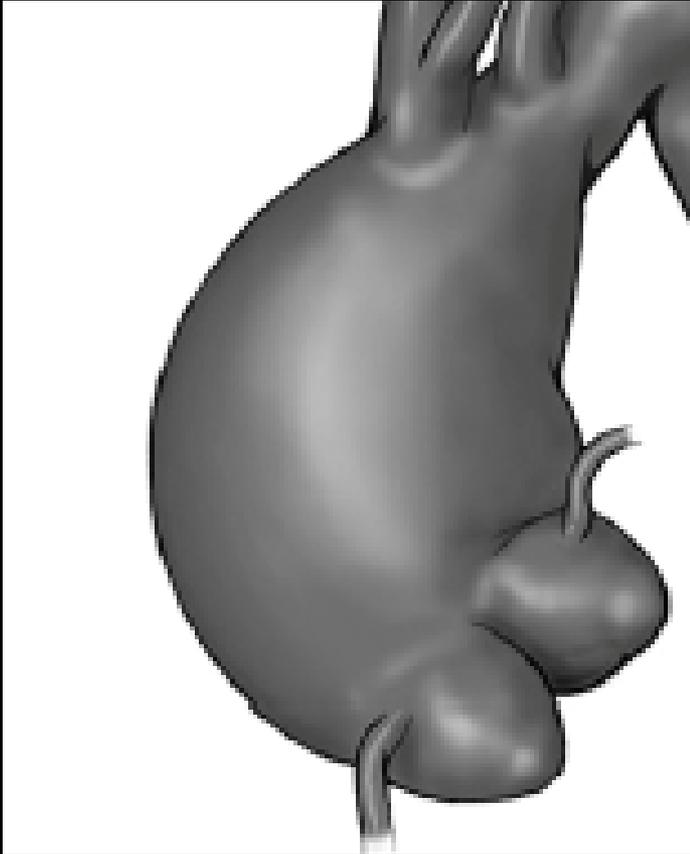


THE DANGER OF REMODELING THE ST JUNCTION



What works for tricuspid valve does not work for bicuspid!

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

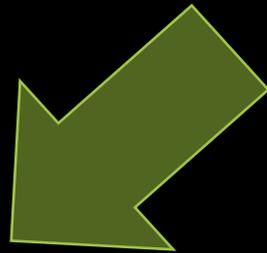


Characteristics:

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 repla
cement

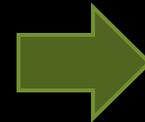
WHEN TO REPLACE THE ROOT

- Preserved STJ



> 45 mm

- Effaced STJ (type II, N or E)
- Asymmetric sinus dilatation



40-45 mm

ASYMMETRIC DILATATION IN BAV

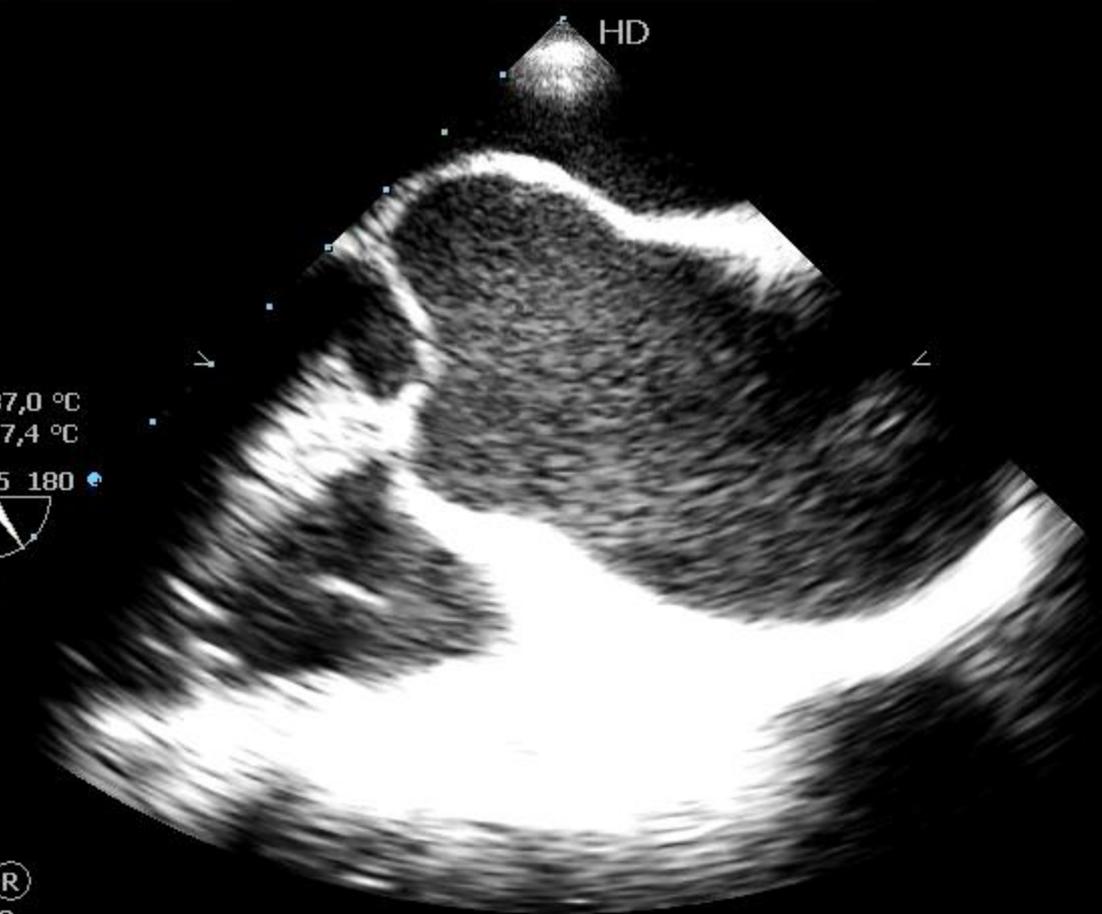
Type 0 or 1, fusion L/R with a large NC sinus



giovinazzo, michele
11-02-18-145037

European Hospital

18/02/2011 PHILIPS
14.55.30



T. Paz: 37,0 °C
T. TEE: 37,4 °C
0 125 180

TEEcch
T6H
MI 1,4
TIS 0,9
H5 Gn 75
232dB/C1
C/1/1

T
P △ (R)
2,5 5,0

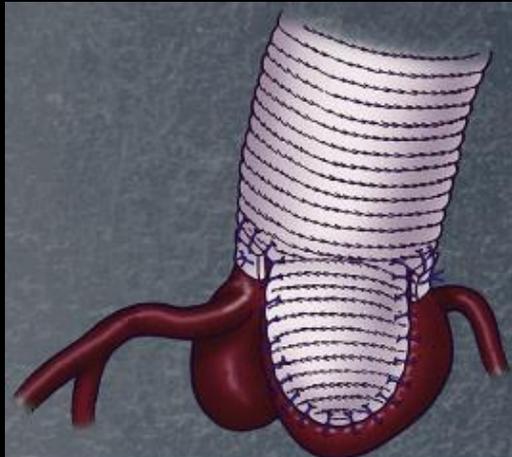
66Hz 11cm
⊗

Non dilated annulus

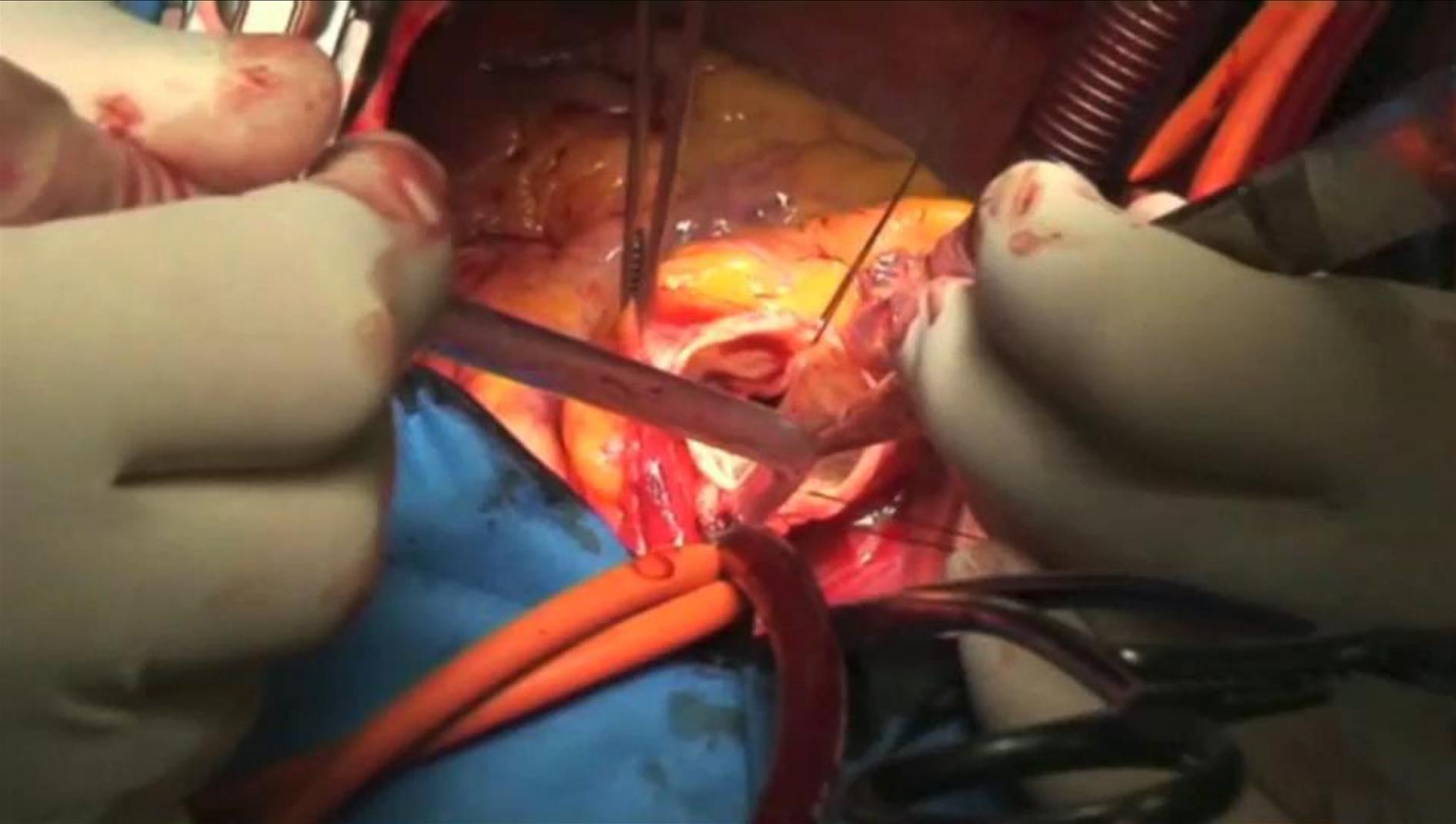


Aortic root 45.4 x 36.9 mm

NC sinus 27 mm



**Aortic root 34.9 x 30.6 mm
NC sinus 17 mm**



ROOT PHENOTYPE

Schaefer's class

N shape

N shape

E shape

Park's class

Type III dilation

Type II dilation

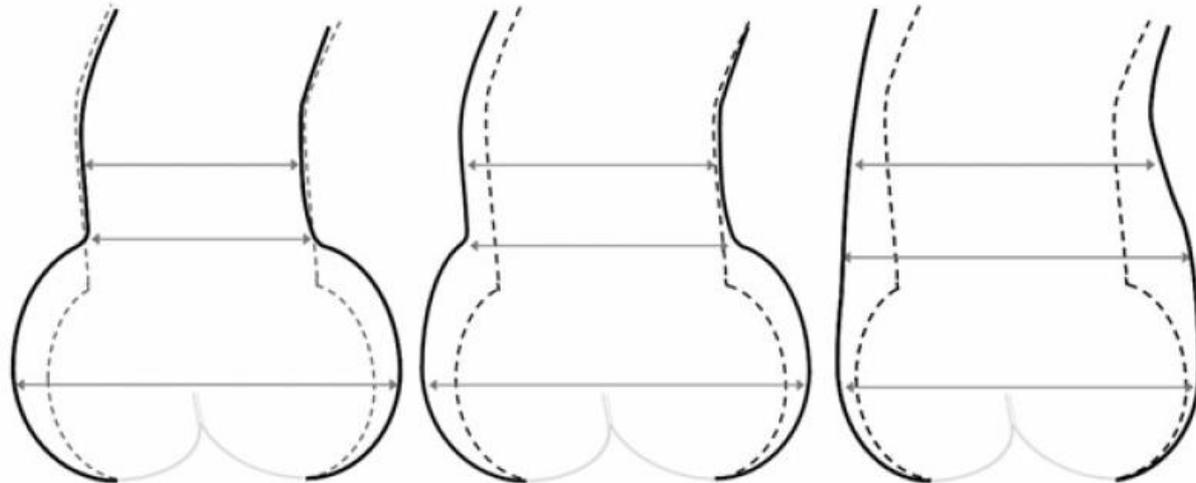
Type II dilation

Della Corte's class:

Root phenotype

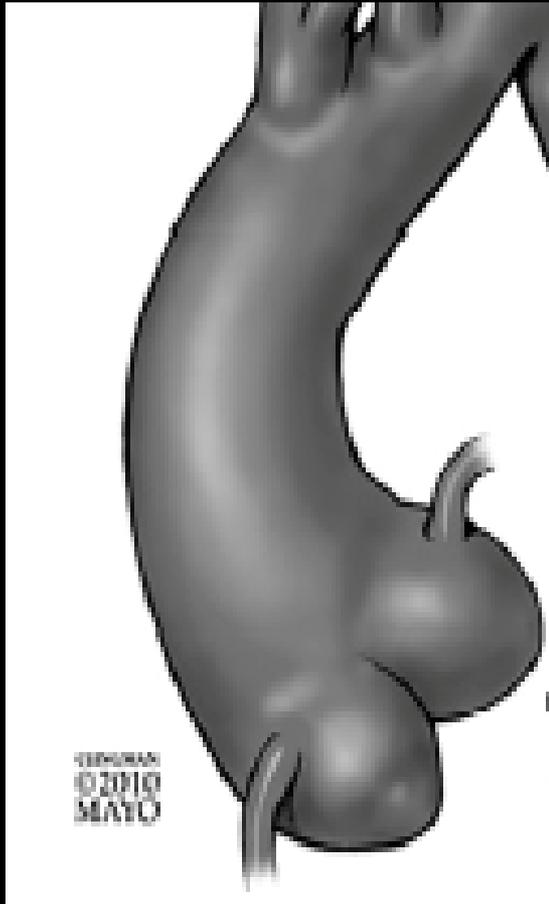
Root phenotype

Root phenotype

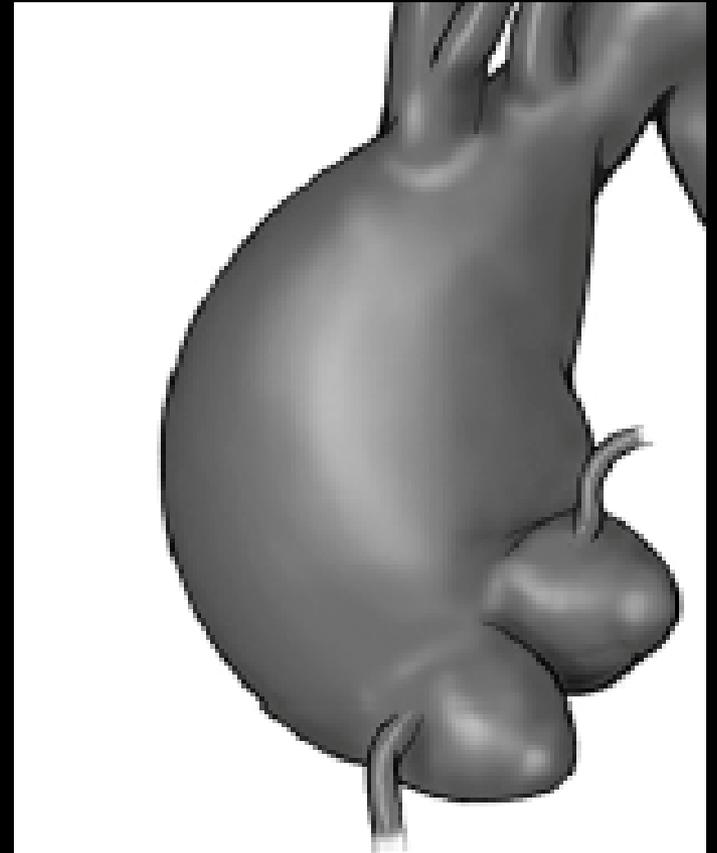


Usually male, young, L/R fusion with normal functioning or regurgitant valve

**AORTOPATHY TYPE III
(YOUNG MALE, DILATED ANNULUS)**

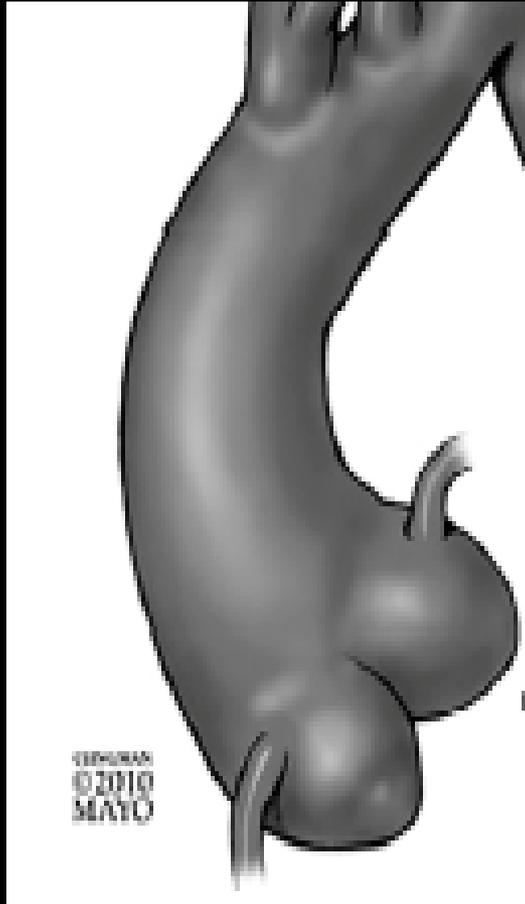


**AORTOPATHY TYPE II
(MOST FREQUENTLY IN L/R FUSION)**



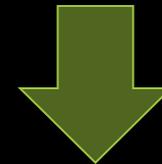
AORTOPATHY TYPE III

(ROOT PHENOTYPE, YOUNG MALE, DILATED ANNULUS)



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

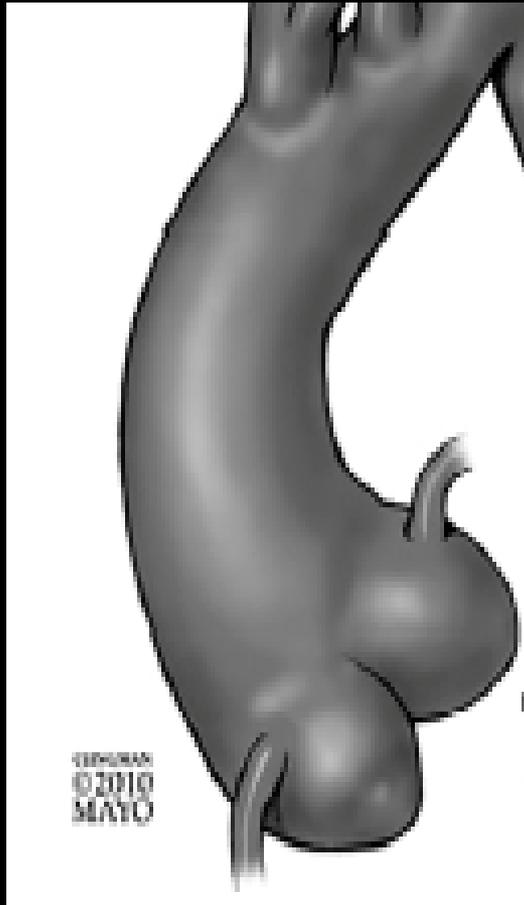
Bentall or Valve sparing procedure



Reimplantation

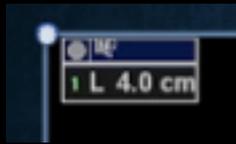
Always replace the root

In this peculiar type of BAV aortopathy 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 ??

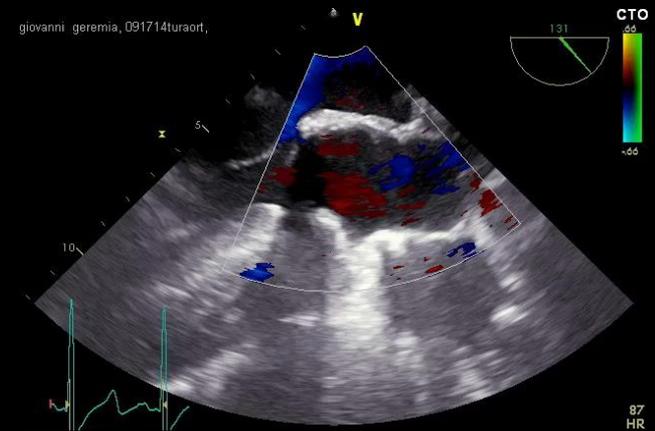
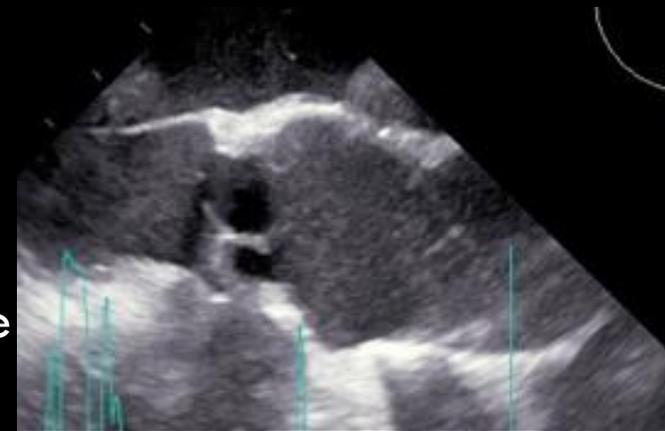
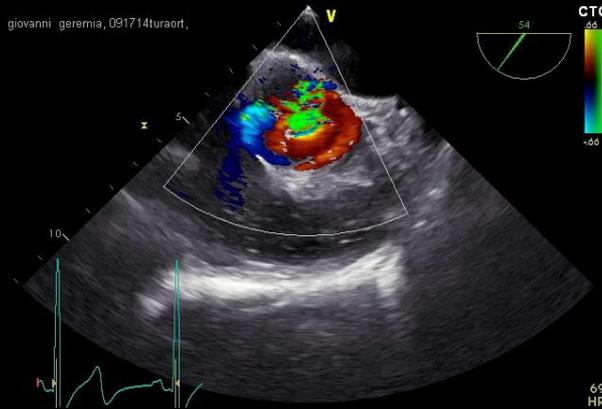
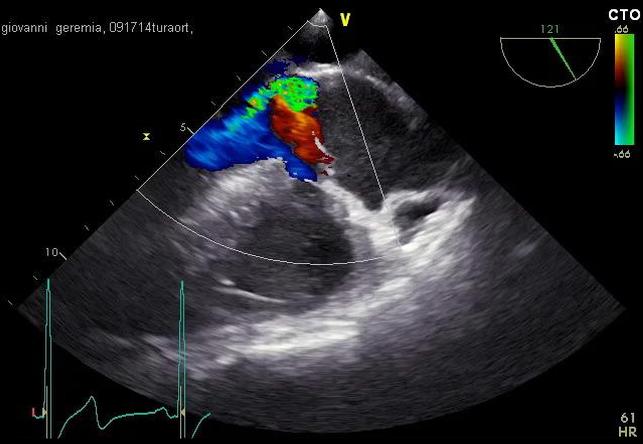


Preoperative

Postoperative

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.