BAV classification

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CENTRUM LEIDEN

FACULTY DISCLOSURE

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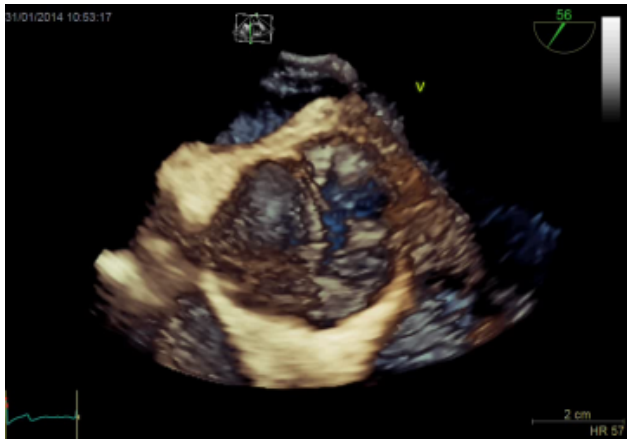
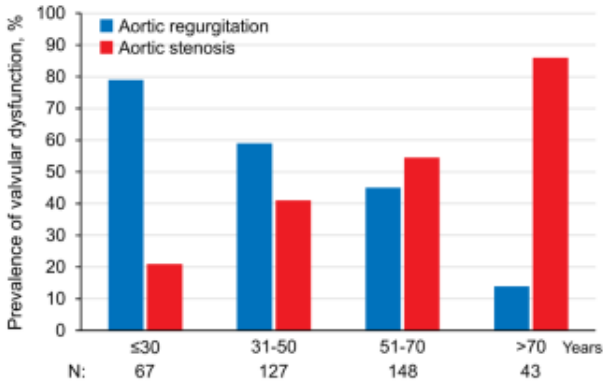
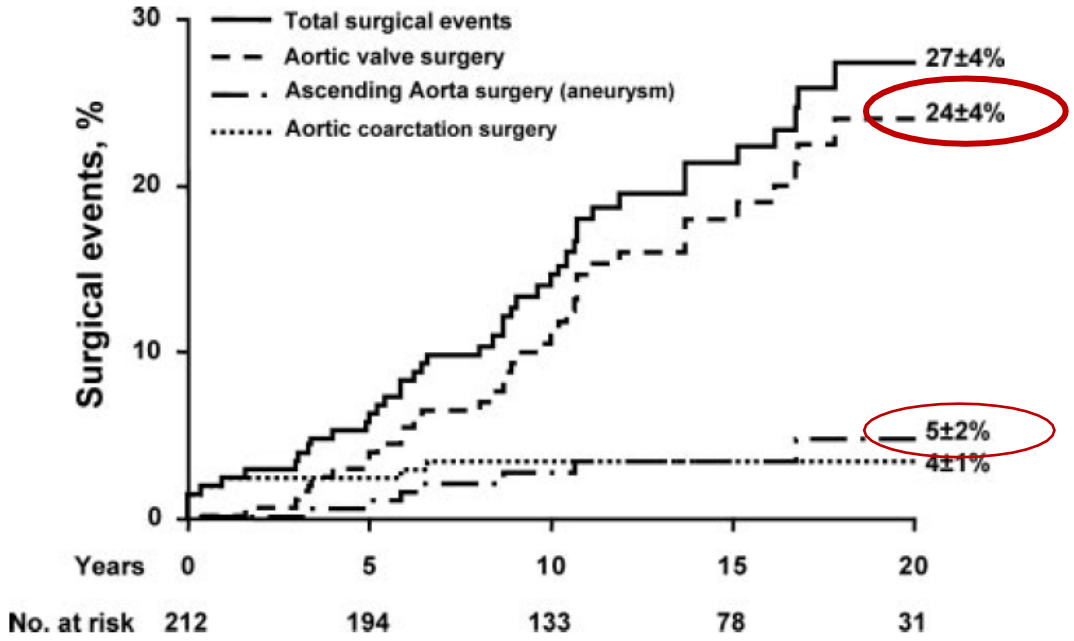
Bicuspid aortic valve



- The most prevalent congenital heart disease (2% of population)
- Associated with VALVULOPATHY
  - AR (younger age)
  - AS (older age)



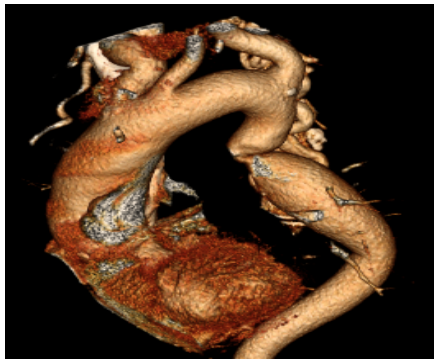
Evangelista et al.
Heart 2018



Michelena HI. Circulation. 2008;117:2776-2784.

- Associated with AORTOPATHY

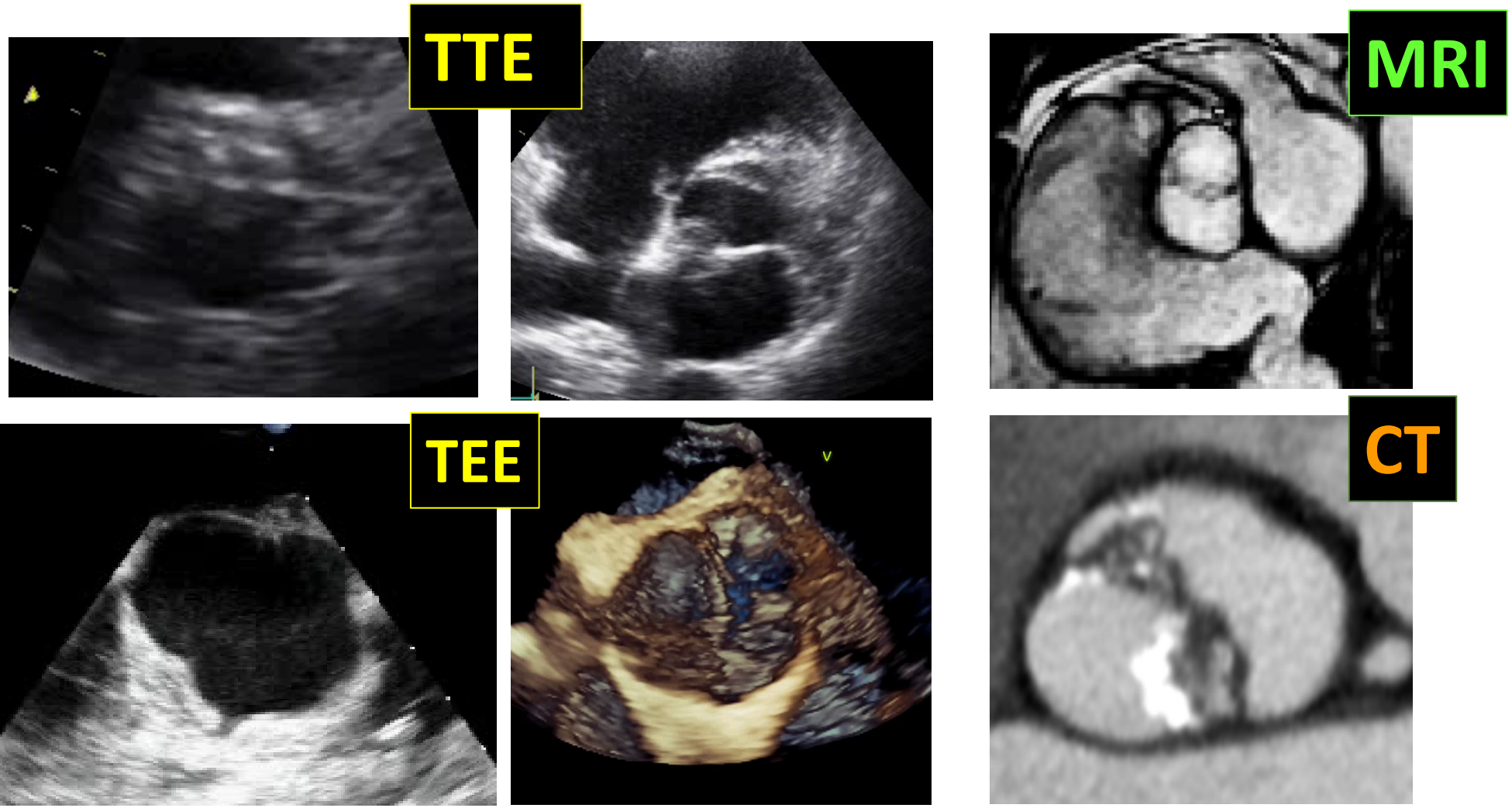
Bicuspid aortic valve: diagnostic approach



- **Diagnose BAV: morphology/phenotype:**
  - Raphe and cusp size and symmetry
  - Assessment of valve degeneration/calcification
- **Assessment of valve function: AR and AS**
- **Dimension thoracic aorta (aortopathy, exclude coarctation)**

Michelena et al. Progr CardioVasc Dis 2020

Imaging



Imaging

BAV	Echo	CT	CMR
Anatomy	+/++	++/+++	++/+++
Aortic stenosis	++	+	+
Aortic regurgitation	++	-	++
Aortopathy	+	++/+++	++/+++

BAV: new classification

- Based on imaging, pathology, surgery and clinical history evidence-based scientific data
- Covers all possible phenotypes and clinical presentations of the bicuspid valvulo-aortopathy
- Use English language and is intended for universal use encompassing clinical and basic research areas
- To optimize clinical practice and future research

TABLE 2 Critical Limitations of the Sievers Classification Compared to the New International Consensus

Sievers and Schmidtke[34] Type of Limitation	Specific Sievers Limitation	International Consensus
Comprehension and retention	Not language-intuitive: Types: 0, 1 and 2	Language-intuitive: Types: fused, 2-sinus and partial fusion
Unable to define all BAV phenotypes	Type 0 does not differentiate between a fused BAV with no raphe and a 2-sinus BAV	Fused types may have raphe or not, 2-sinus types do not have raphe
Lack of prerepair symmetry assessment	Non-existent	Fused types require assessment of symmetry for surgical repair planning
Lack of recognition of BAV phenotypes	Does not recognize partial fusion (forme fruste), does not recognize fused BAV with no raphe	Recognizes partial fusion (forme fruste) Recognizes fused BAV with no raphe, which is different than 2-sinus BAV
Lack of recognition of aortopathy phenotypes	Non-existent	Aortic phenotypes: root, ascending and extended
Includes a non-BAV congenital aortic valve abnormality	Type 2 is not BAV, is unicuspid aortic valve, incompletely defined	Does not include unicuspid aortic valves
Evidence-based	Anatomical pathology only	Imaging, anatomical pathology, surgical-functional pathology, clinical-associations

Michelena et al J Thorac Cardiovasc Surg. 2021

Clinical Prognostic subgroups

Bicuspid Aortic Valve Nosology

Complex- presentation Valvulo-aortopathy

- Associated syndromes, and/or
- Associated congenital lesions, and/or
- Severe aortic coarctation, and/or
- Early/Accelerated valve dysfunction, and/or
- Early/accelerated aortopathy
- More common pediatric/young adult
- Requires surveillance
- Frequently requires early treatment

Increased risk:
- Endocarditis
- Aortic Dissection

Typical- presentation Valvulo-aortopathy

- Progressive valvulopathy and/or aortopathy
- More common young adult/adult
- Requires surveillance
- Usually requires eventual treatment

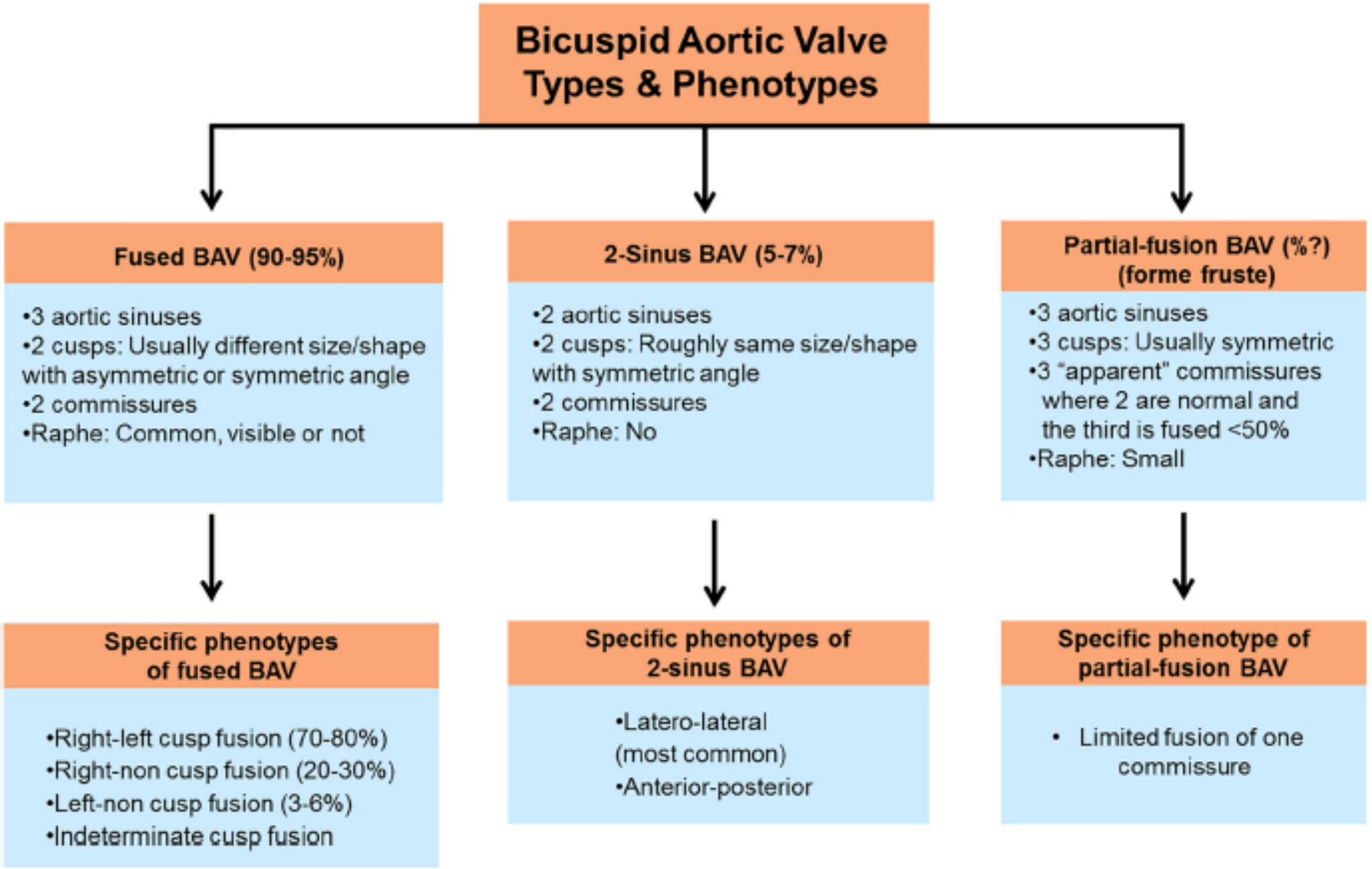
Increased risk:
- Endocarditis
- Aortic Dissection

Uncomplicated or Undiagnosed (retrospective definition)

- Mild or non-progressive valvulo-aortopathy
- Lifelong silent condition
- Never reaches clinical manifestations
- Sometimes diagnosed incidentally or by autopsy
- Requires surveillance if diagnosed

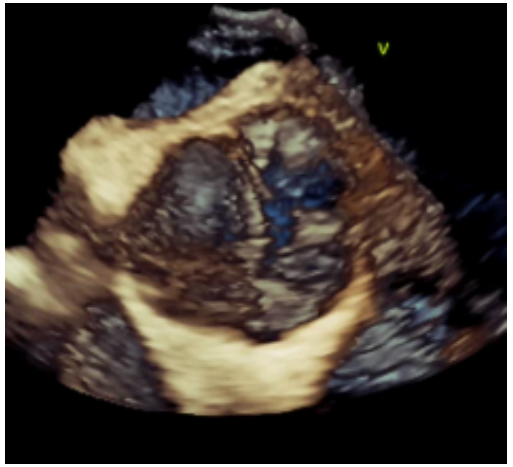
Michelena et al. Progr CardioVasc Dis 2020

BAV phenotypes: new classification

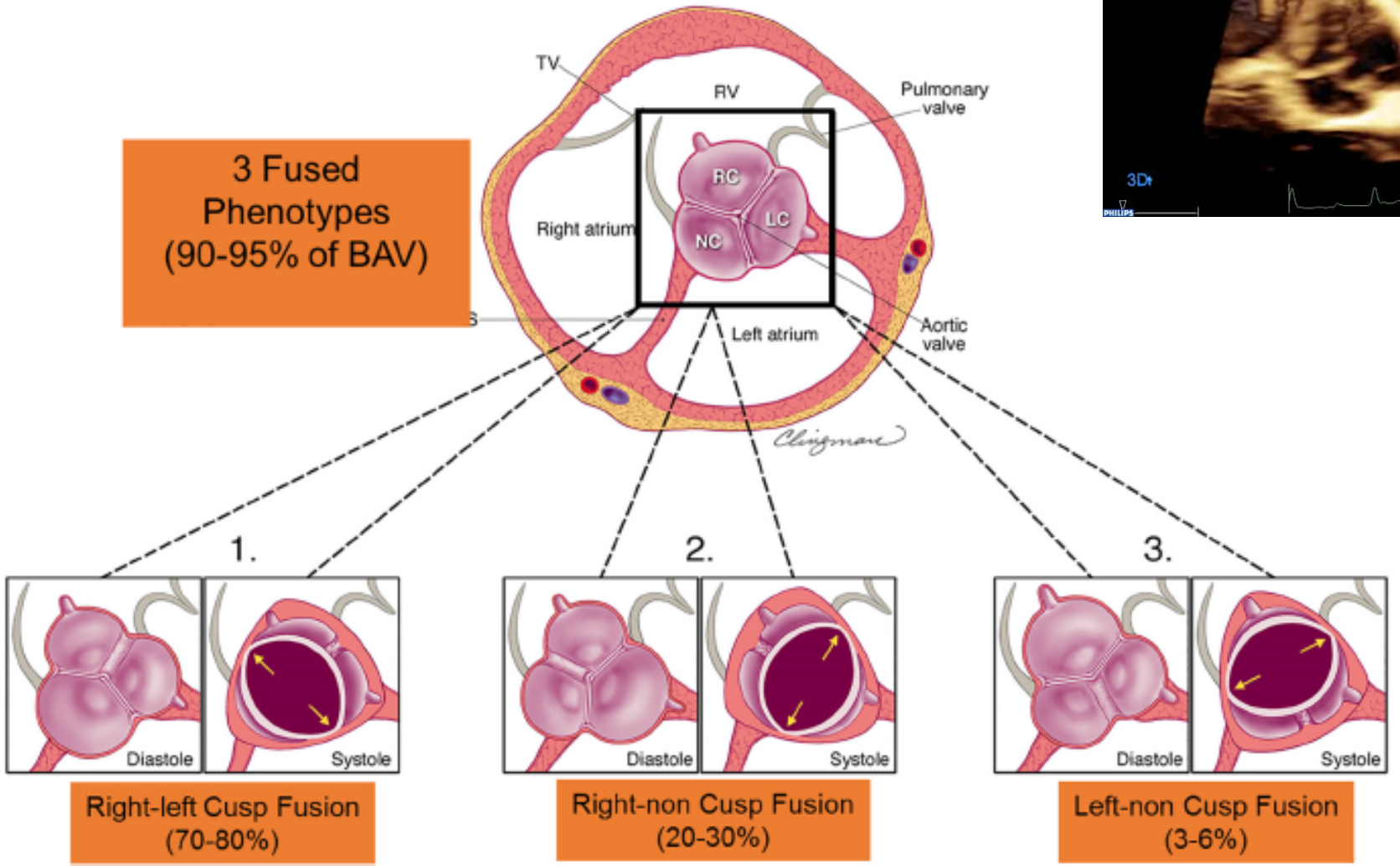


Michelena et al J Thorac Cardiovasc Surg. 2021

BAV phenotype: new classification



3 Fused Phenotypes (90-95% of BAV)



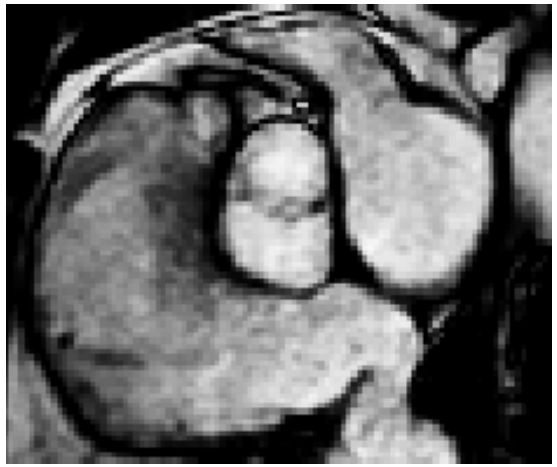
Right-left Cusp Fusion (70-80%)

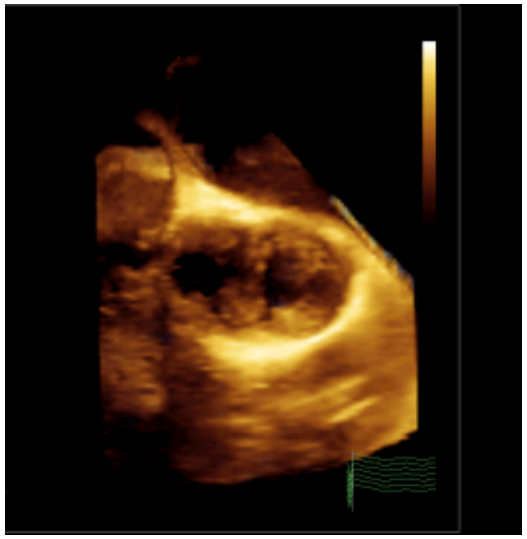
Right-non Cusp Fusion (20-30%)

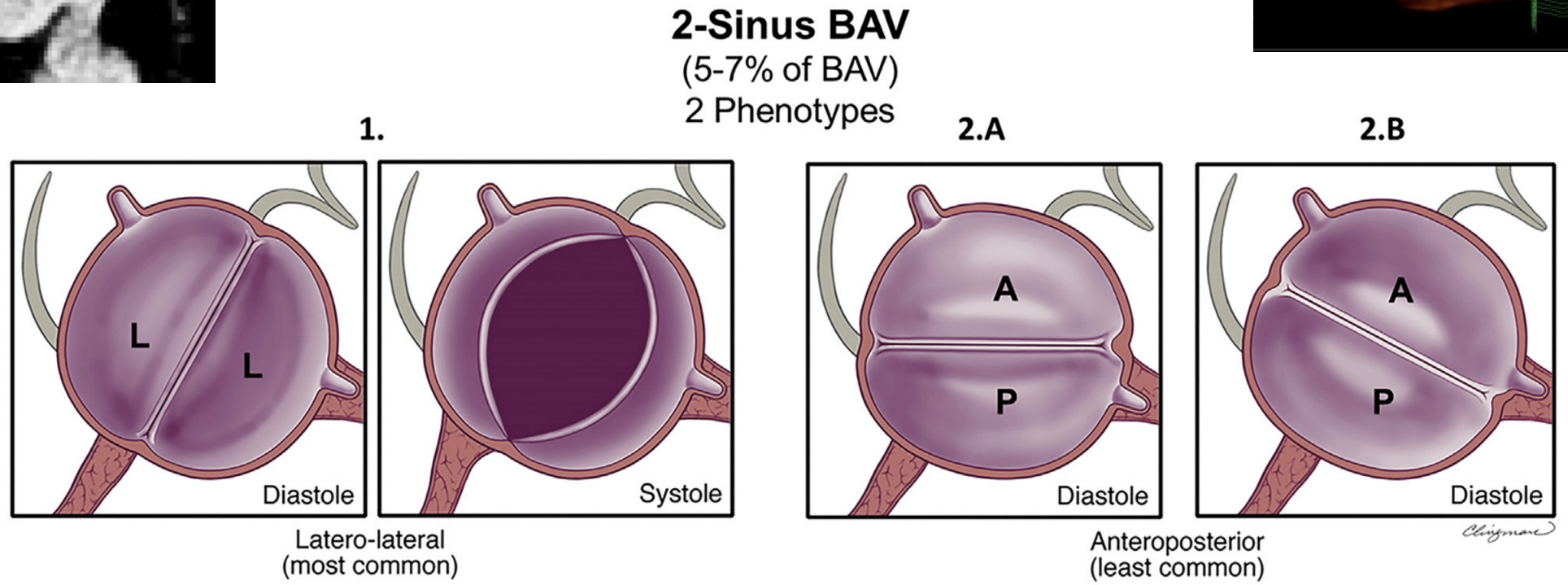
Left-non Cusp Fusion (3-6%)

Michelena et al Progress in Cardiovascular Diseases 63 (2020) 419–424

BAV Phenotype: new classification

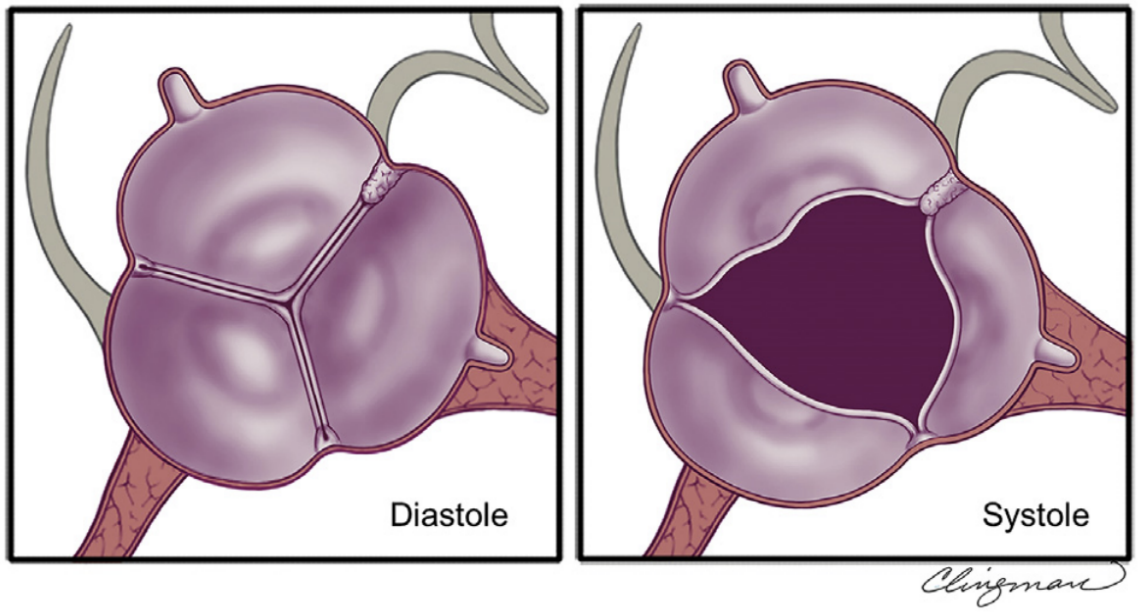




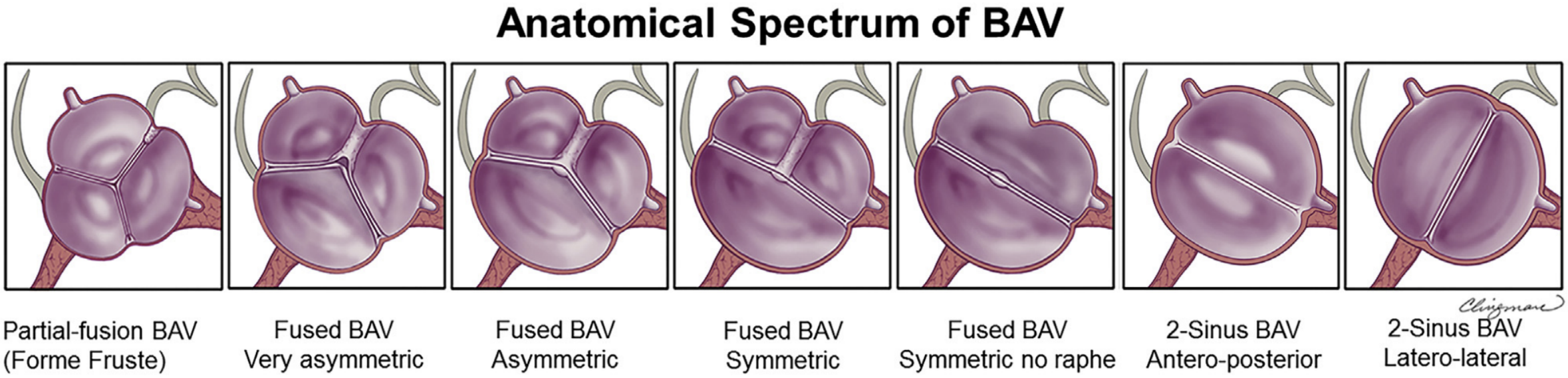


BAV Phenotype: new classification

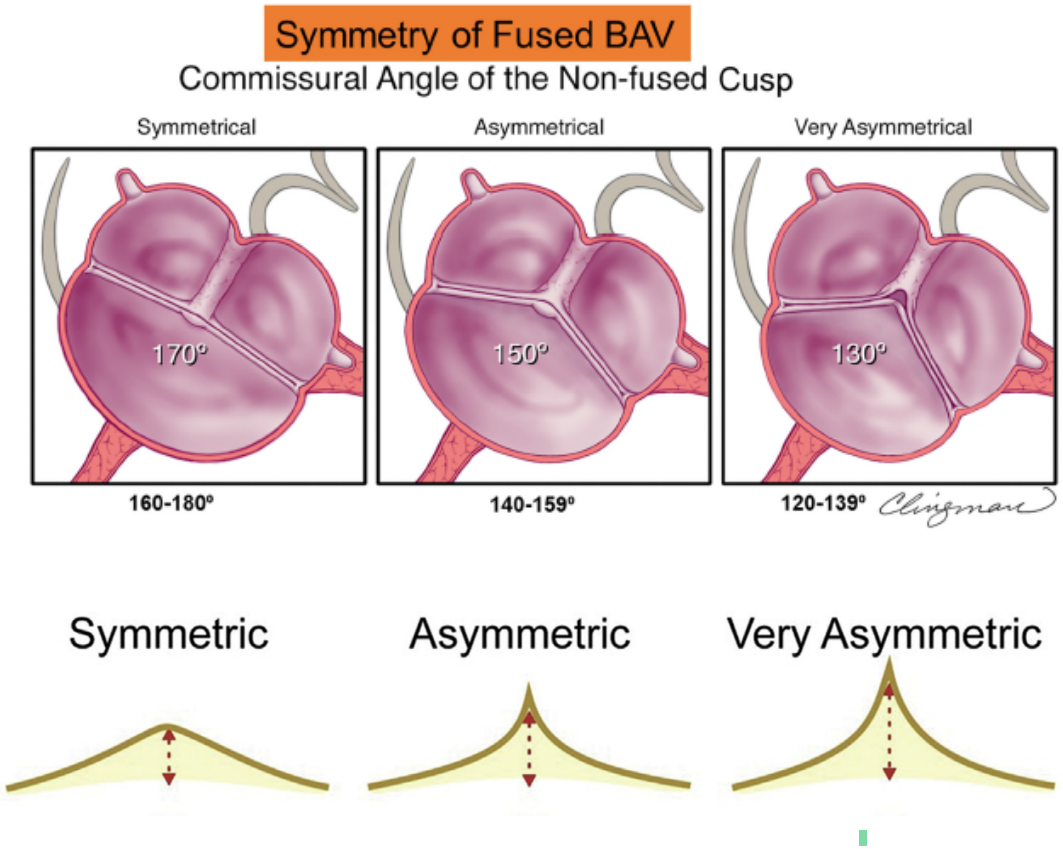
Partial-Fusion BAV
(Forme Fruste)
Short fusion of 1 commissure



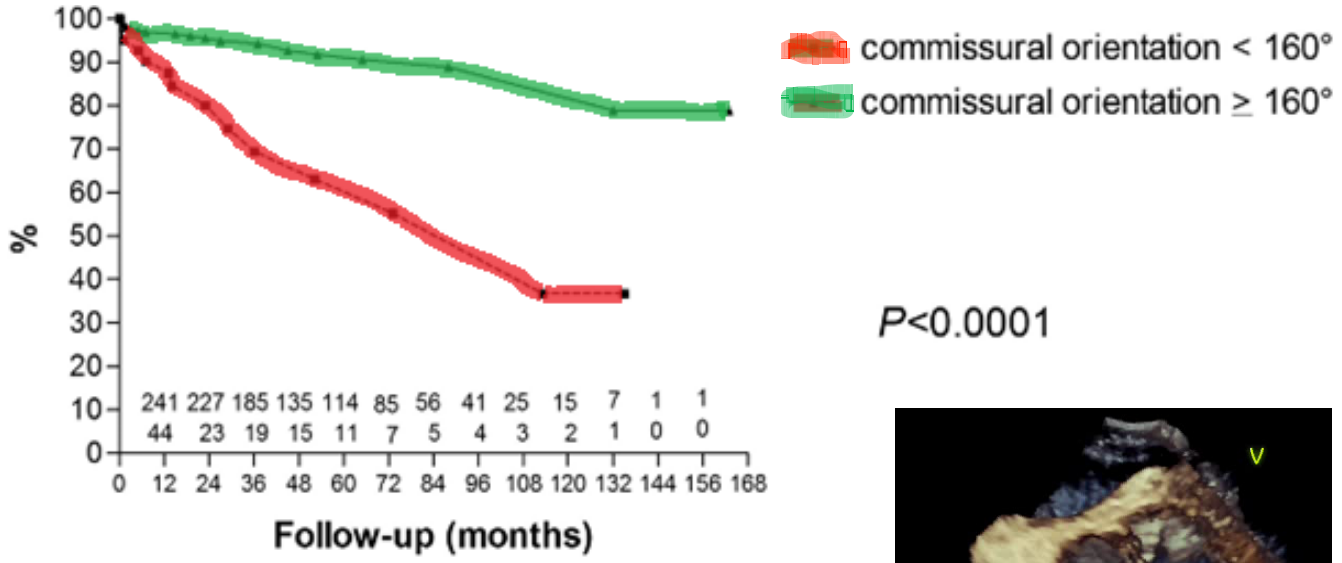
Gradient of embryology defects and “Bicuspidity”

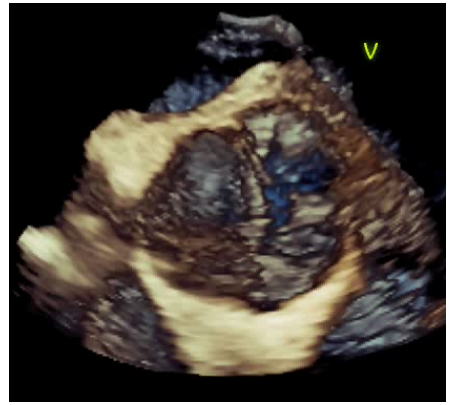


BAV Morphology: new classification



Freedom from reoperation after aortic valve repair

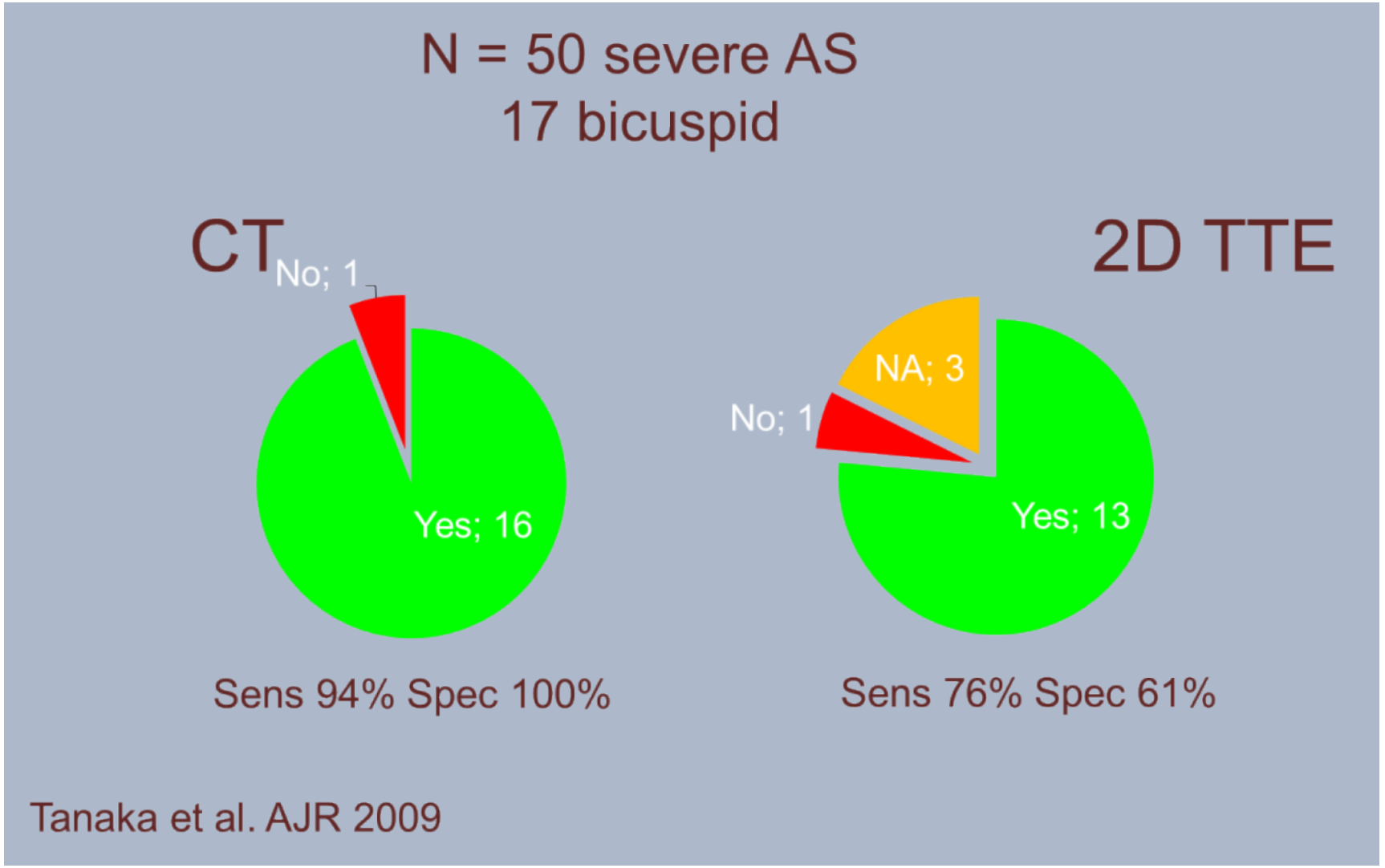




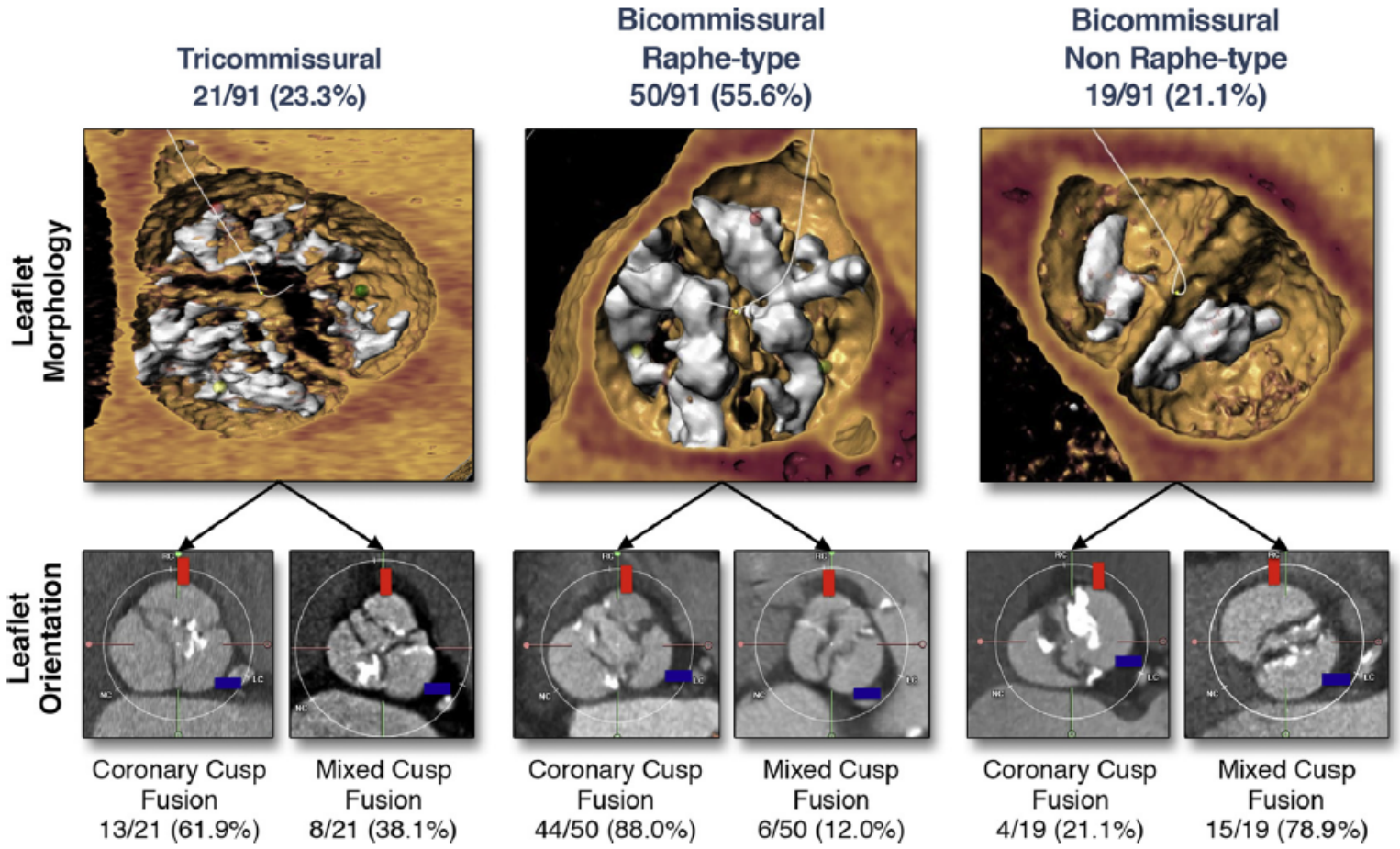
Michelena et al Progress in Cardiovascular Diseases 63 (2020) 419–424

Aicher et al Circulation 123 (2011) 178-185

BAV configuration: CT superior over TTE

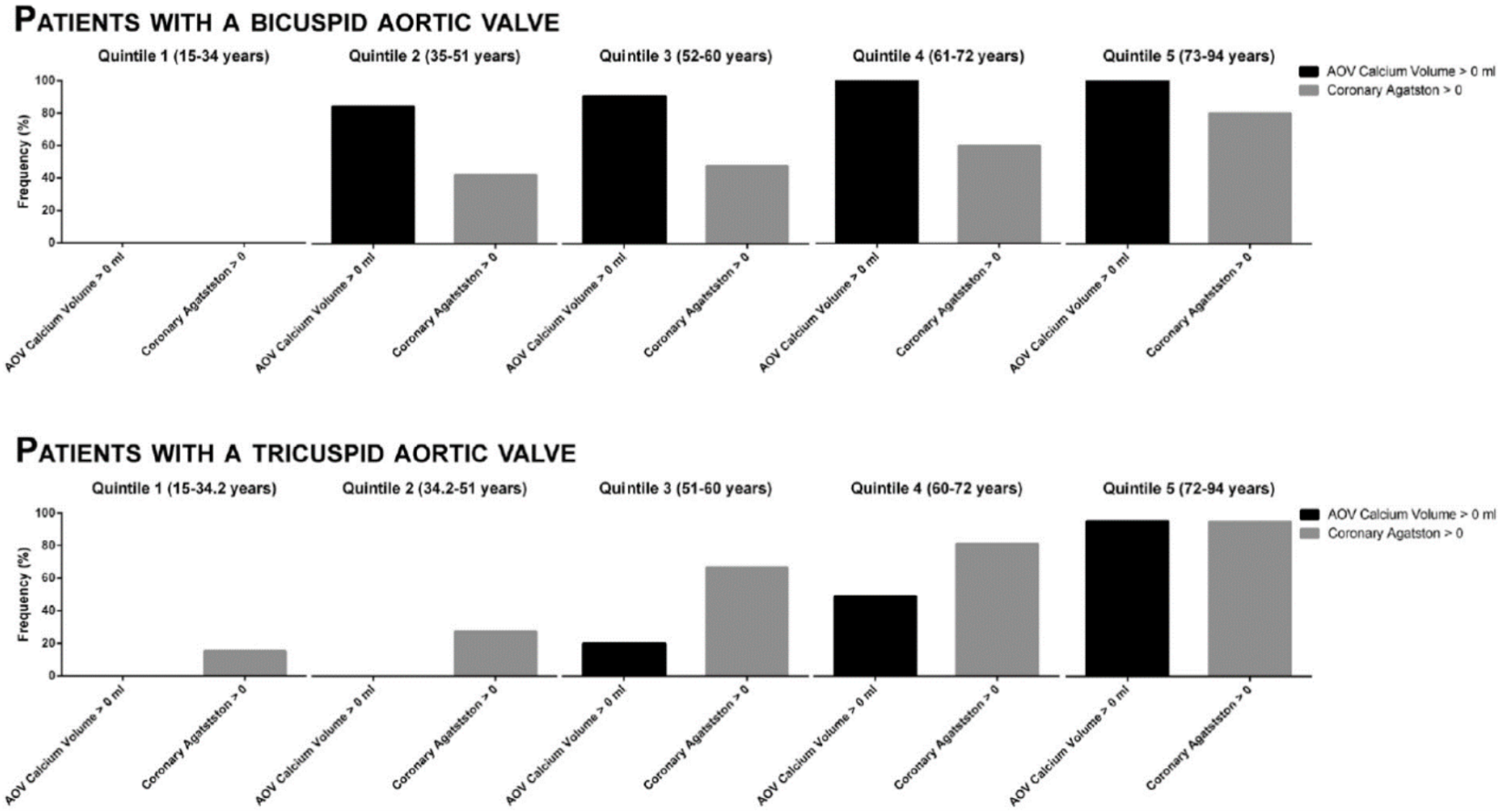


Precise BAV configuration



Jilaihawi et al. JACC Imag 2016

BAV calcification – assess by MDCT



Philippe van Rosendael et al AJC 2016

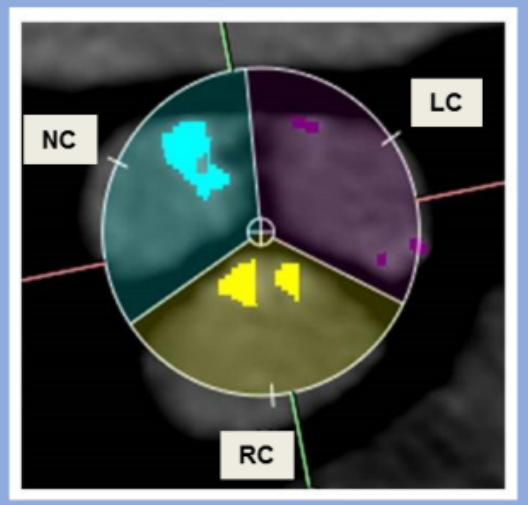
Ca deposits in BAV (vs TAV):

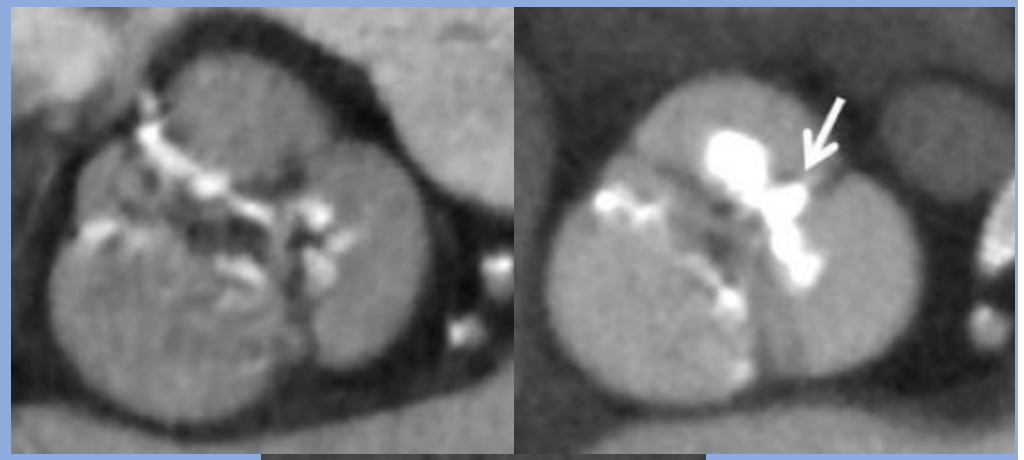
- starts younger age
- more Ca in valve than in coronary arteries

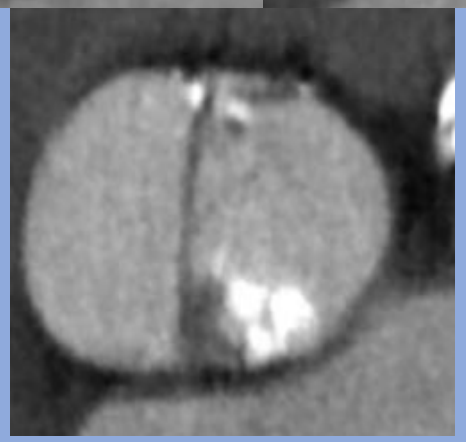
Location of Ca deposits in BAV:

- 60% raphe
- 59% in cusp with origin of the LM

BAV calcification (by MDCT): at the commissures and leaflet tips

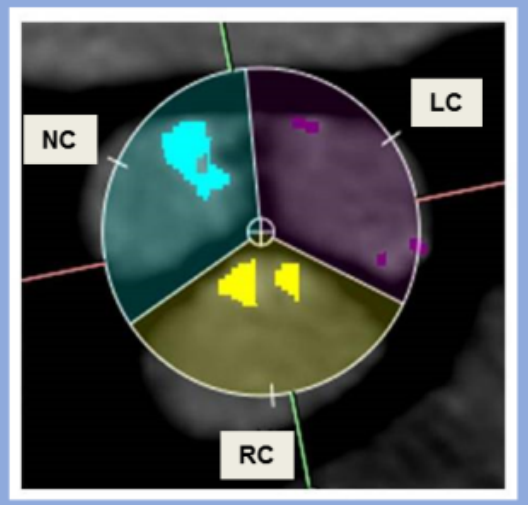


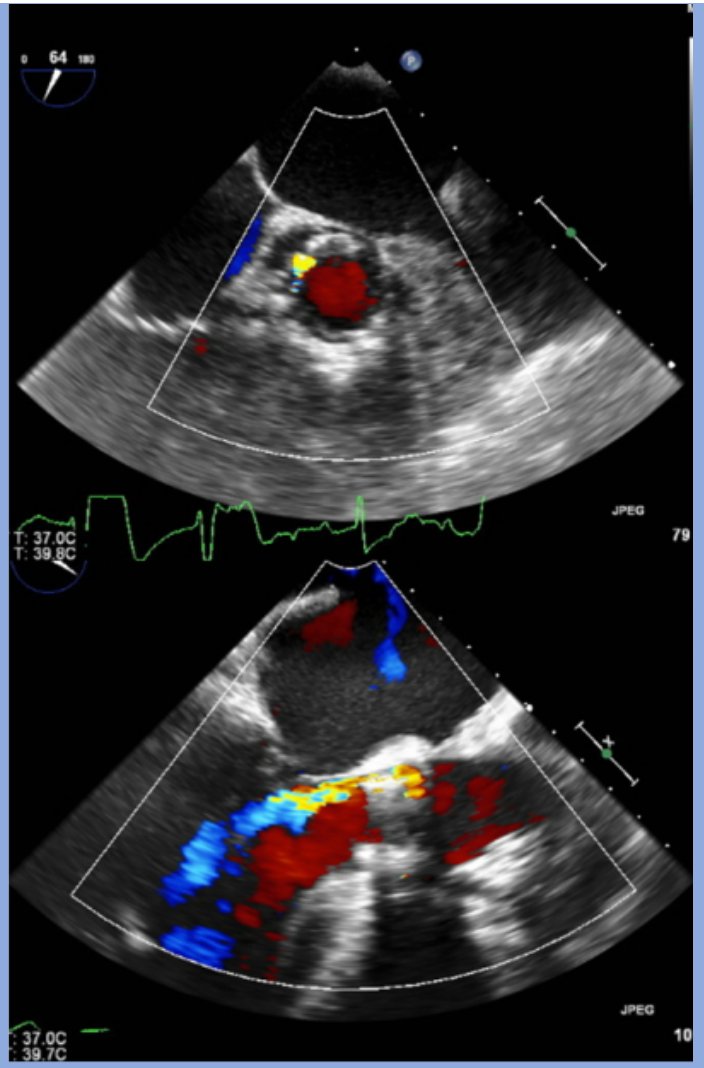


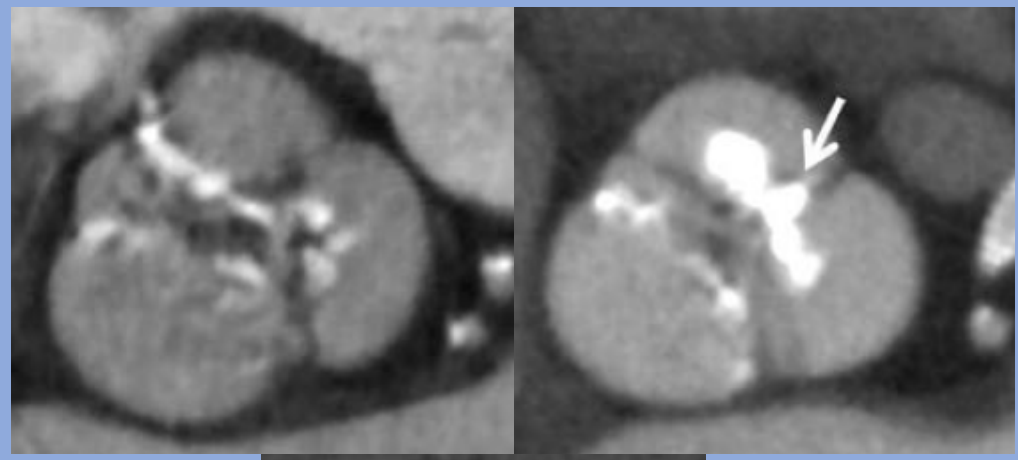


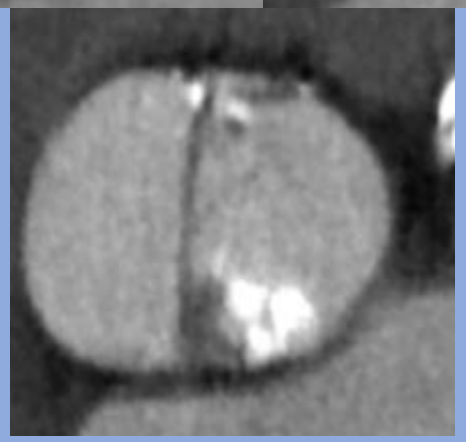
Ewe et al. Am J Cardiol 2011

BAV calcification (by MDCT): important for PVL (AR) after TAVR





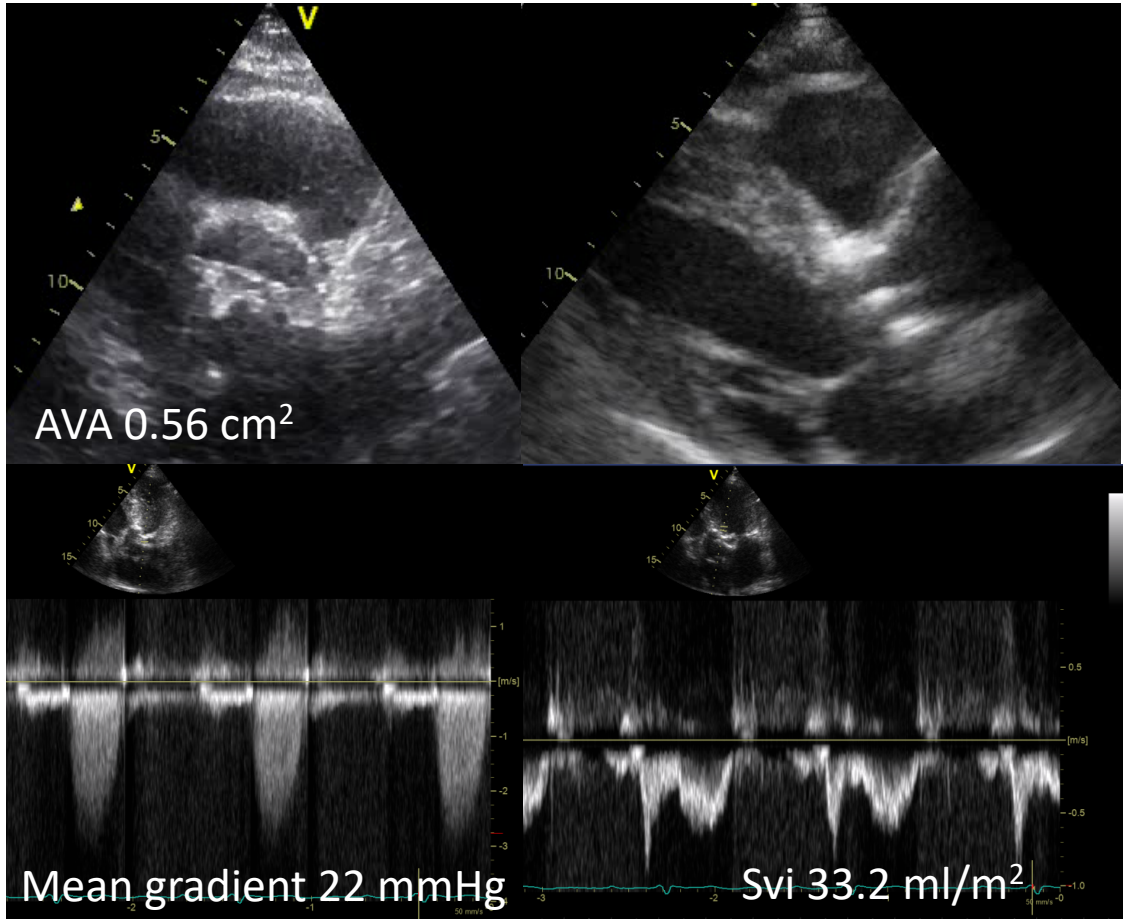


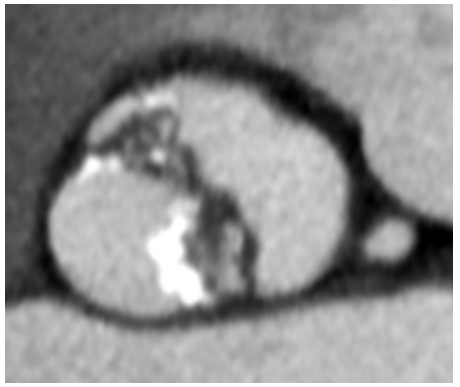


Ewe et al. Am J Cardiol 2011

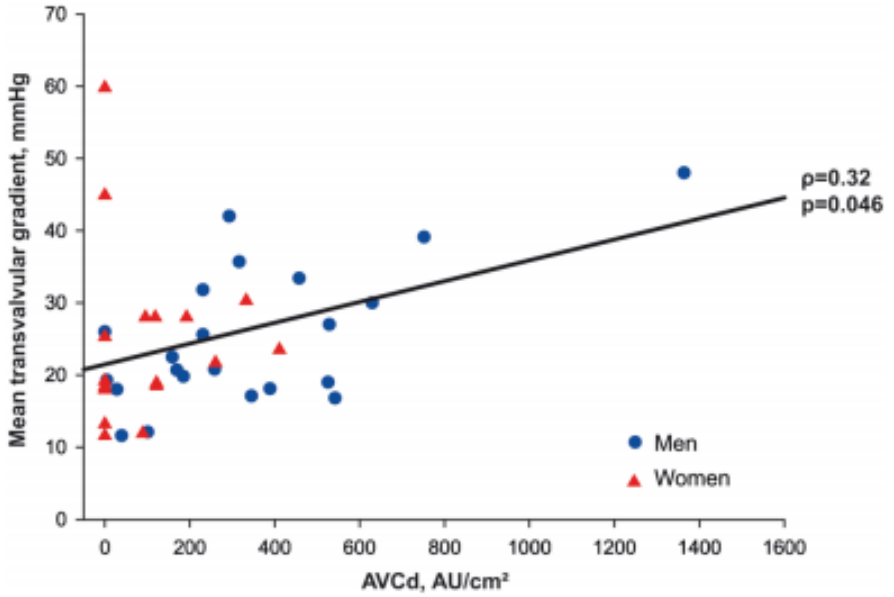
BAV – Valve Function

Aortic stenosis





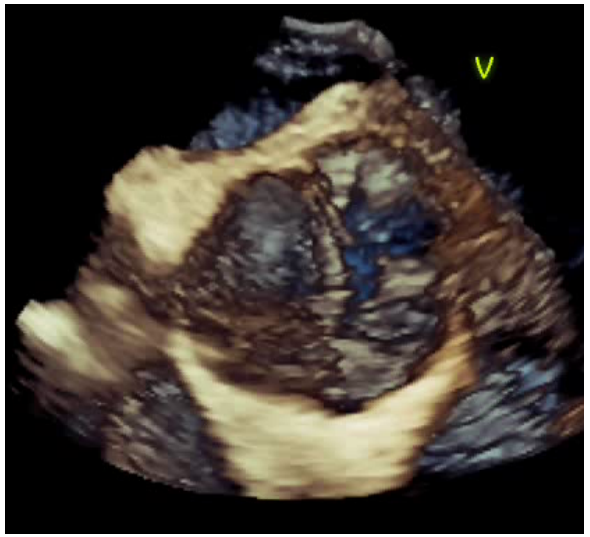
AVC?

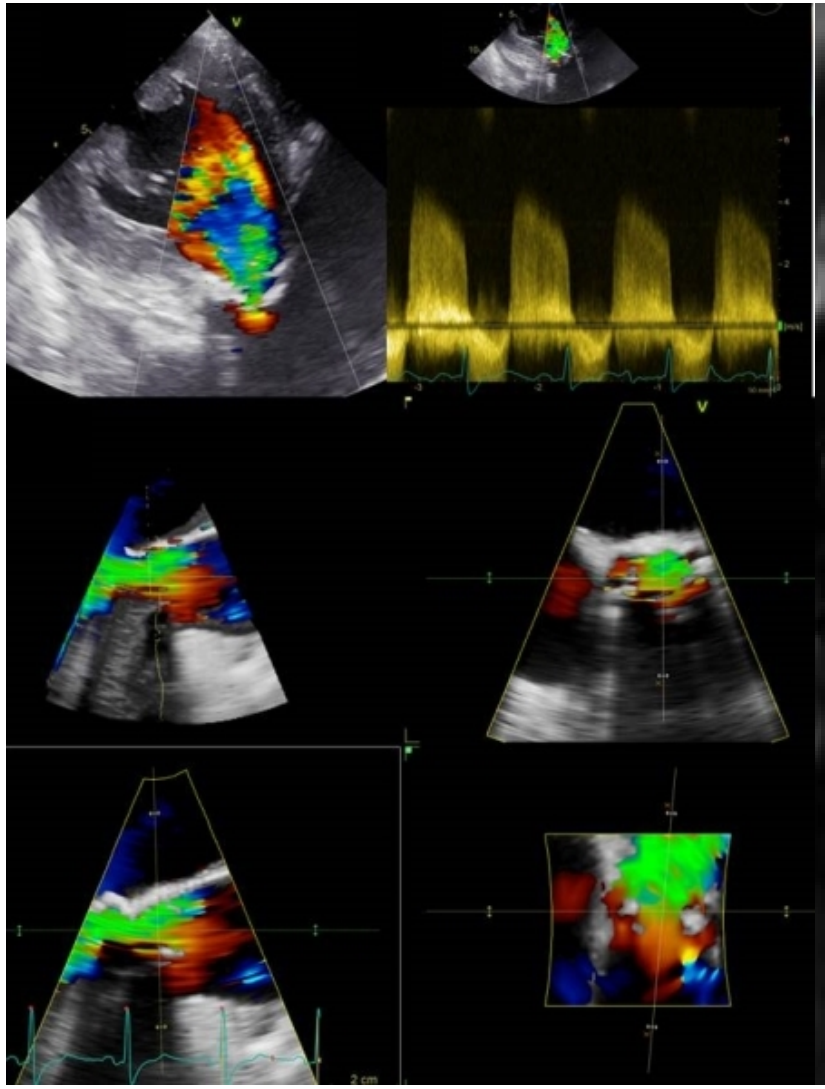


Shen M, et al. Heart 2017;103:32–39

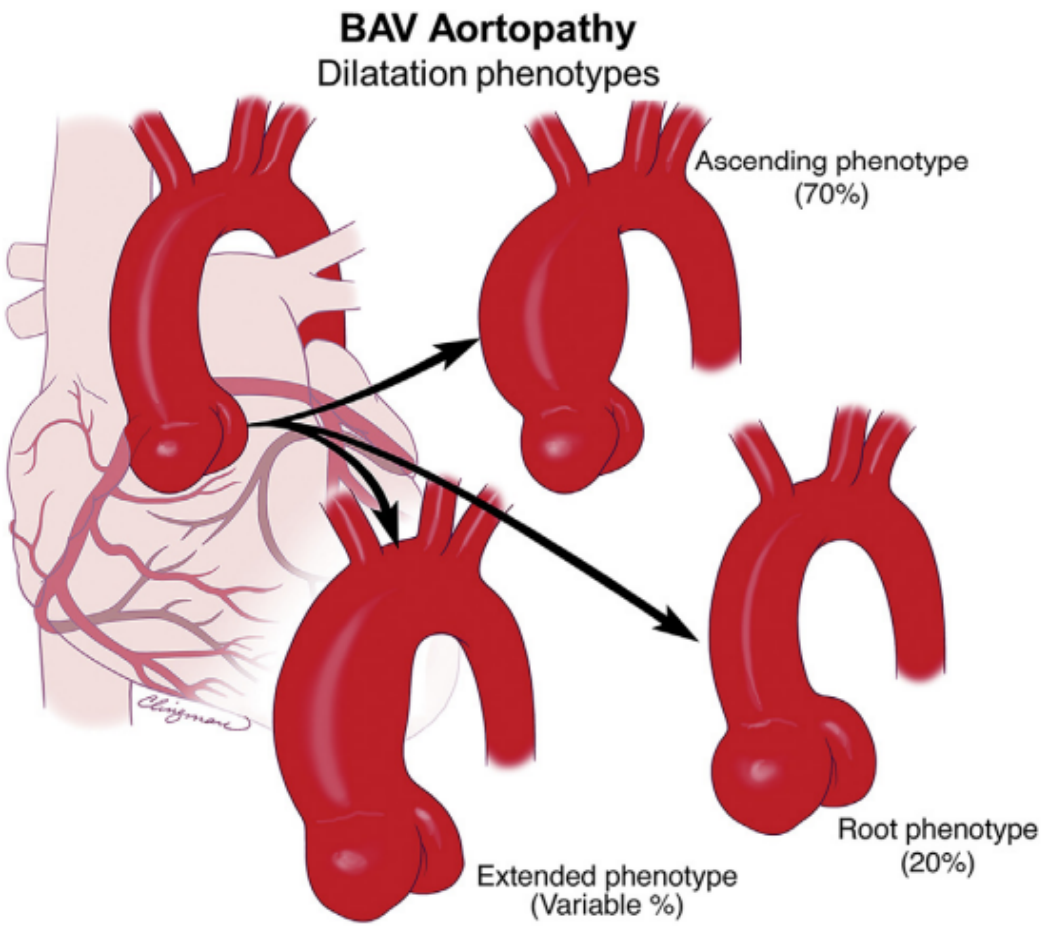
BAV – Valve Function

Aortic regurgitation





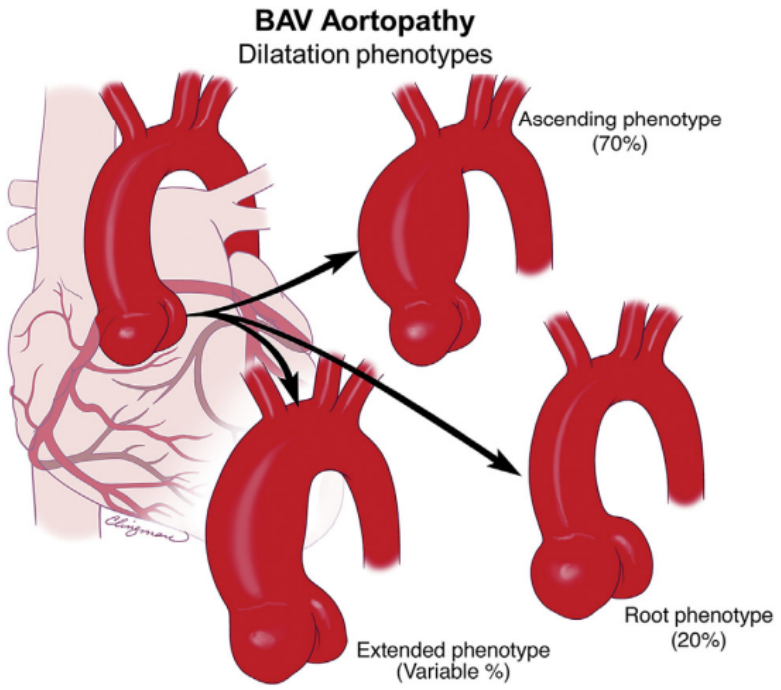
BAV aortopathy: new classification



- Older age
- More associated with AS
- Associated with RCC-NCC fusion BAV

- Younger age
- Associated with LCC-RCC fusion BAV and male sex
- Faster tubular-ascending aorta dilatation and AR
- Most likely to be associated with genetic cause
- Risk factor for dissection

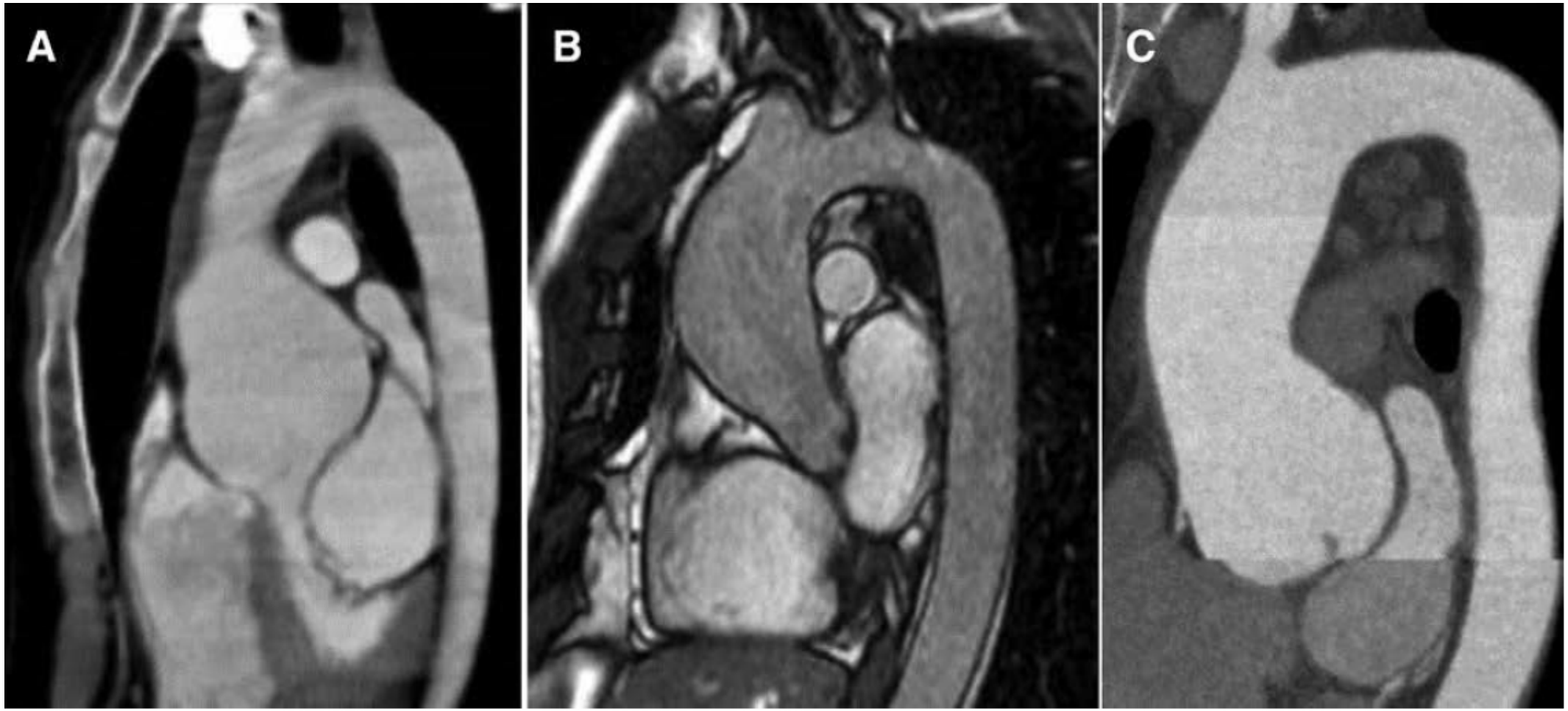
BAV aortopathy: new classification



Aortic disease in BAV			Valve disease in BAV	
Aortic root	20-30%	RL	23-70%	Aortic regurgitation (AR)
• **Earlier age** • **Male sex**				
Ascending aorta	60-80%	RN	14-50%	Aortic stenosis (AS)
• **Older age**				

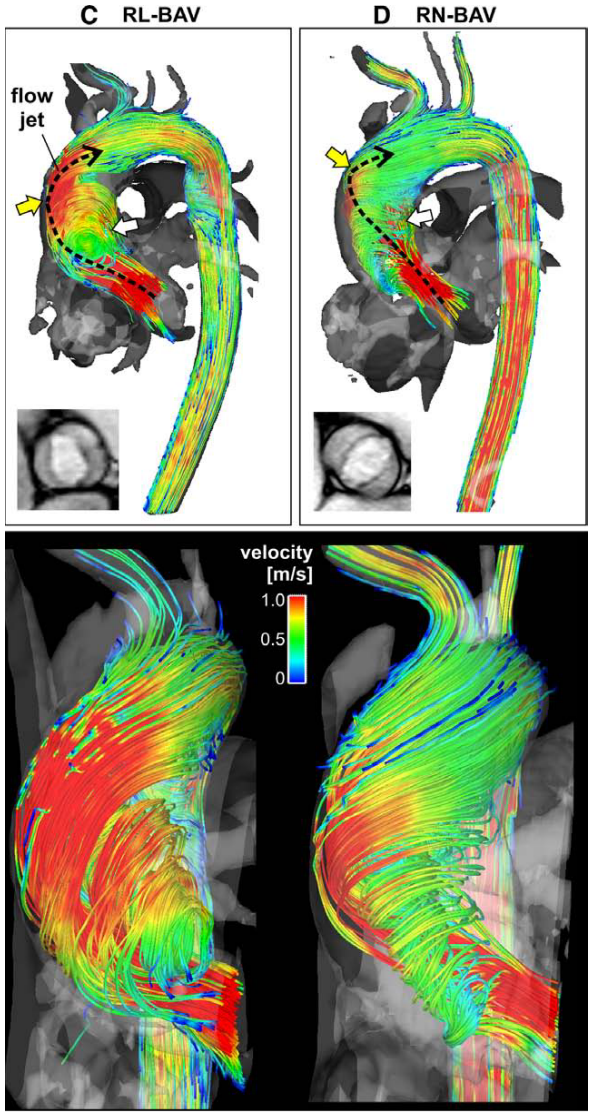
Michelena HI. Ann Thorac Surg. 2021 Sep;112(3):e203-e235.

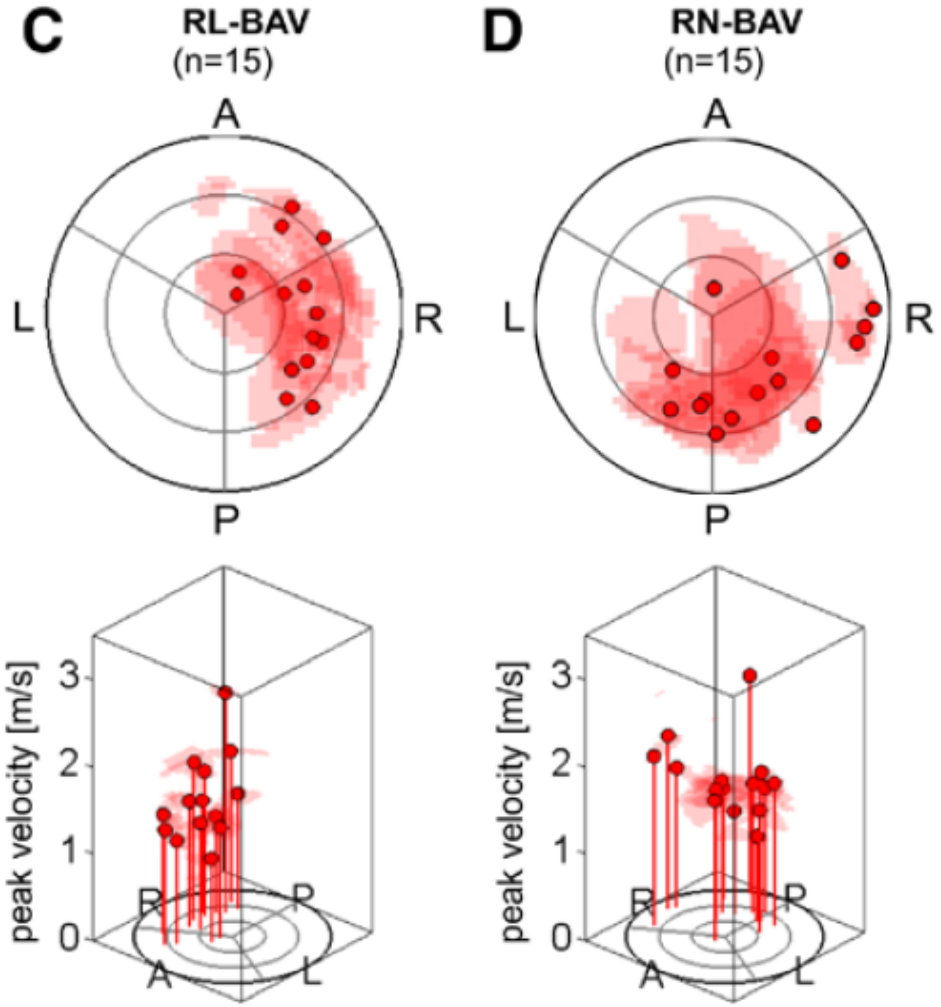
BAV - aortopathy



Kong et al. Circ CV Imaging 2017

BAV - aortopathy





Mahadevia et al. Circulation 2014

Conclusions