# Aortic Root and Aortic Valve Repair: What is the Current State? What Does the Surgeon Need to Know from the Imager?

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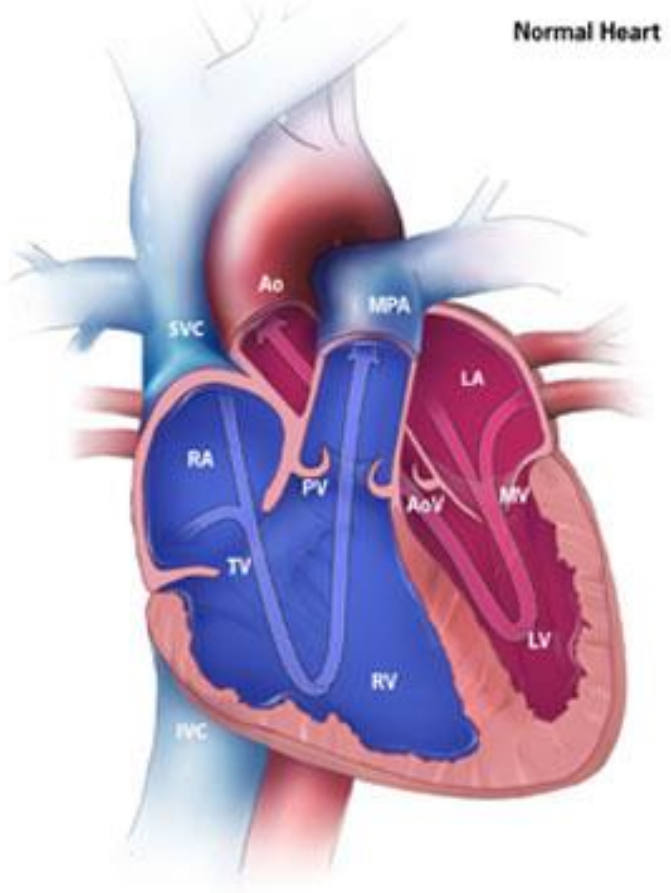
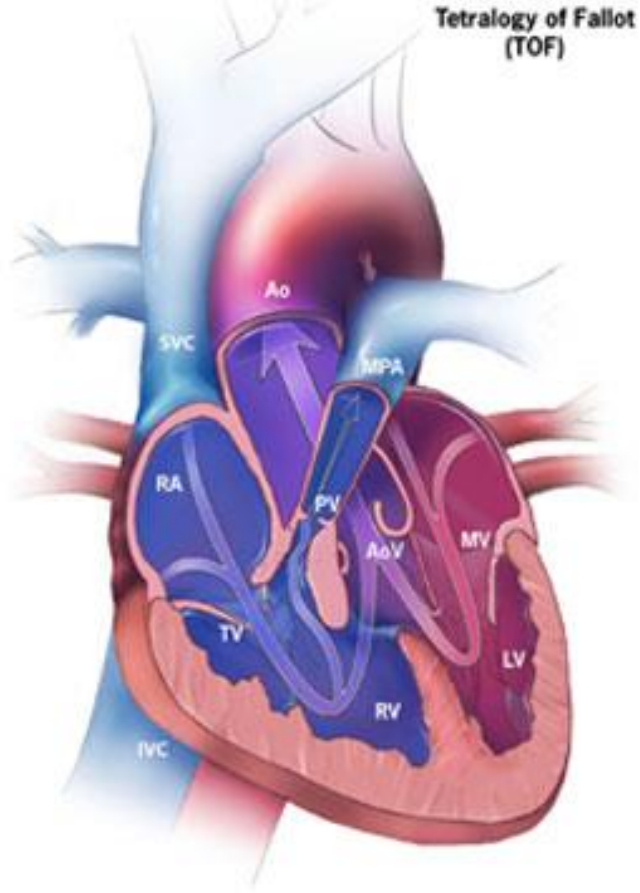
# Aortic root dilatation

- Aortic coarctation: BAV, syndromes
- Marfan S. and collagenopathies
- BAV
- Conotruncal anomalies: DORV, TGA, ToF, TA
- Single ventricle: Fontan, Norwood

# Hemodynamic causes:

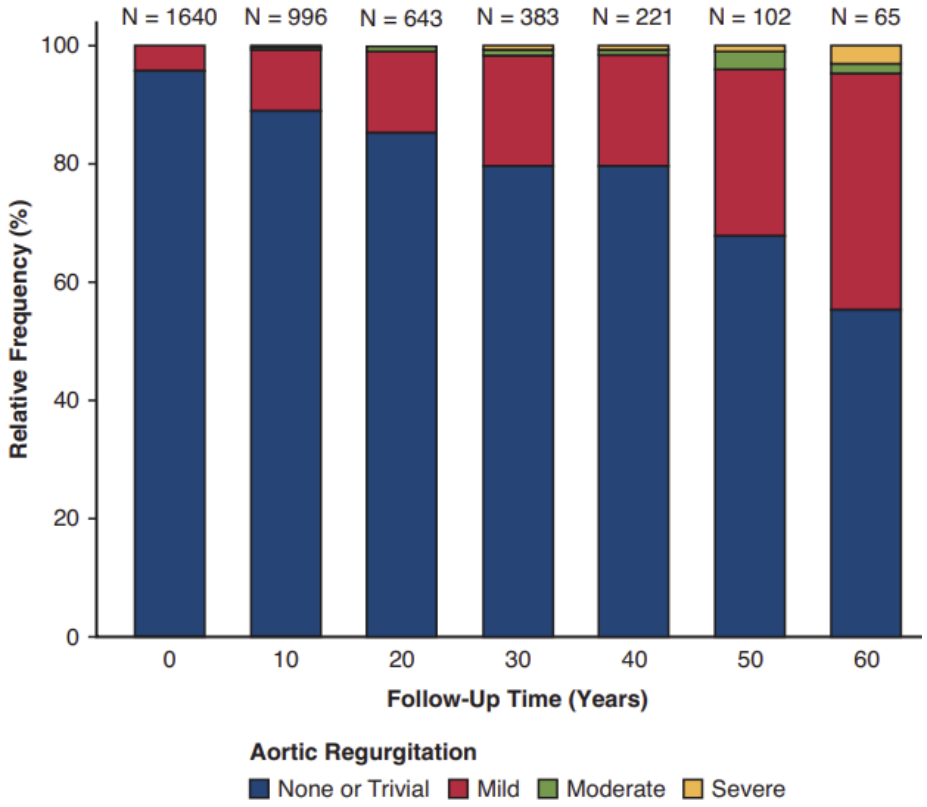


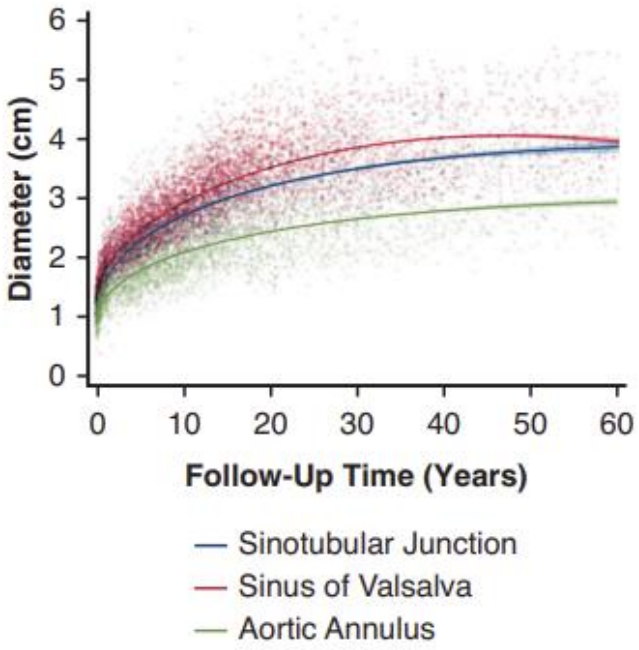



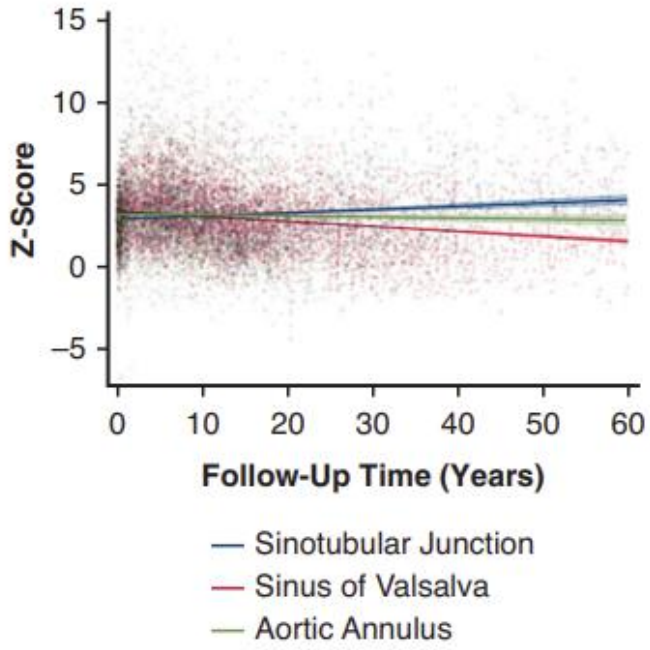

The earlier the correction

The lower the incidence of aortic dilatation

# Natural history of aortic root dilatation in ToF



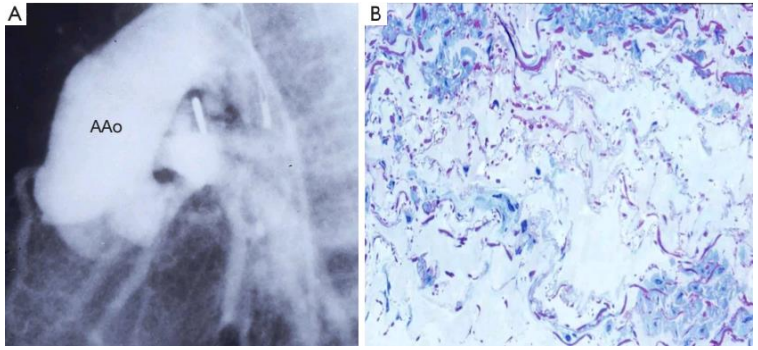




Incidence of dilatation > 30%
9 % of significant dilatation
Progression is rare

Sengupta A. Natural history of aortic root dilatation an pathologic aortic regurgitation in tetralogy of Fallot and its morphological variants. JTCVS 2023: in press

# Histologic/Genetic causes:

- Increased aortic stiffness
- Decreased distensibility
- Focal loss of smooth muscle cells in the tunica media
- Increased mucoid accumulation
- Fragmentation and disruption of elastic lamellae
- 22q11.2
- 14q23 (found both in BAV and HLHS)
- FBN1




Grade of disruption of elastic fibers

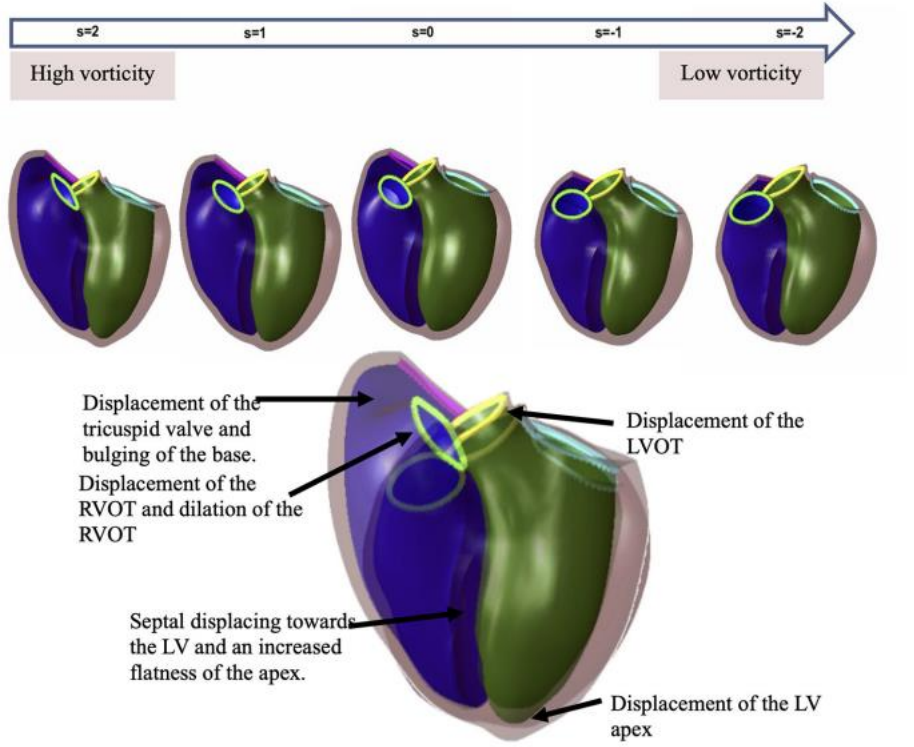
| | Normal | Grade 1 | Grade 2 | Grade 3 |
|---|---|---|---|---|
| Marfan | | | | 10 |
| AAE | | | | 5 |
| BAV AS | | 2 | 6 | 4 |
| BAV AR | | 4 | 3 | 3 |
| TOF | | | 9 | 6 |
| SV PS | | | 2 | 1 |
| TA PS | | | 2 | 1 |
| DORV | | | 1 | 1 |
| DOLV | | | 1 | |
| VSD | | | | 1 |
| Do.Ao A | | | 1 | |
| PTA | | | 3 | 2 |
| d-TGA | | | 6 | 2 |
| Controls | 21 | | | |

Timing could mitigate the histologic derangement as elastin deposition occurs in the first days after birth

Niwa K Aortic dilatation in complex congenital heart disease Cardiovasc Diagn Ther 2018; 8(6): 725-738
Francois K Aortopathy associated with congenital heart disease: A current literature review Ann Pediatr Cardiol 2015;8(1):25-36

# Geometric causes:

- Ventriculoarterial angular geometry
- Left ventricular systolic vorticity and supraphysiologic elical flow contribute to aortopathy




Schafer M Increased systolic vorticity in the left vetricular outflow tract is associated with abnormal aortic flow formations in tetralogy of Fallot. Int J Cardiovasc Imaging 2020; 36:691-700

# Indications:

- > 45 concomitant surgery on Ao valve
- > 45-50 mm Loeys-Dietz
- > 50 mm family history of aneurysm-dissection/ Marfan Syndrome
- > 50 mm and rapid aortic growth
- > 55 mm asymptomatic

- ACHD?

# Risk of aortic rupture:

- 37 million admissions
- 12.000 dissections
- 6 with conotruncal disease
- OR: 1,2-1,7 (Marfan 92,9; BAV 10,4)

Thoracic aortic dissection and rupture (TAD) in congenital heart disease (CHD): diagnosis by age group.

| CHD diagnosis | All ages | | 0 to 17 years | | 18 to 44 years | | 45 to 64 years | | 65 + years | |
|---|---|---|---|---|---|---|---|---|---|---|
| | n TAD | n Died | n TAD | n Died | n TAD | n Died | n TAD | n Died | n TAD | n Died |
| BAV, n (%) | 94 | 10 | 5 (5%) | 1 | 39 (41%) | 4 | 37 (39%) | 4 | 13 (14%) | 1 |
| ASD, n (%) | 48 | 8 | 2 (4%) | 2 | 4 (8%) | 0 | 21 (44%) | 2 | 21 (44%) | 4 |
| Aortic coarctation, n (%) | 16 | 0 | 1 (6%) | 0 | 9 (56%) | 0 | 3 (19%) | 0 | 3 (19%) | 0 |
| VSD, n (%) | 12 | 4 | 1 (8%) | 1 | 3 (25%) | 0 | 3 (25%) | 0 | 5 (42%) | 3 |
| PDA, n (%) | 10 | 2 | 3 (30%) | 2 | 1 (10%) | 0 | 4 (40%) | 0 | 2 (20%) | 0 |
| Tetralogy of Fallot, n (%) | 3 | 1 | 1 (33%) | 0 | 1 (33%) | 0 | 1 (33%) | 1 | 0 (0%) | 0 |
| D-TGA, n (%) | 2 | 0 | 0 (0%) | 0 | 0 (0%) | 0 | 2 (2%) | 0 | 0 (0%) | 0 |
| Truncus arteriosus, n (%) | 1 | 0 | 0 (0%) | 0 | 1 (100%) | 0 | 0 (0%) | 0 | 0 (0%) | 0 |
| HLHS, n (%) | 1 | 1 | 1 (100%) | 1 | 0 (0%) | 0 | 0 (0%) | 0 | 0 (0%) | 0 |
| Fontan, n (%) | 1 | 0 | 0 (0%) | 0 | 0 (0%) | 0 | 1 (100%) | 0 | 0 (0%) | 0 |
| Other CHD, n (%) | 36 | 5 | 0 (0%) | 0 | 9 (13%) | 0 | 18 (20%) | 2 | 9 (17%) | 3 |

Note: CHD diagnoses are not mutually exclusive.
ASD: atrial septal defect. BAV: bicuspid aortic valve. D-TGA: D-transposition of the great arteries. HLHS: hypoplastic left heart syndrome. PDA: patent ductus arteriosus. VSD: ventricular septal defect.

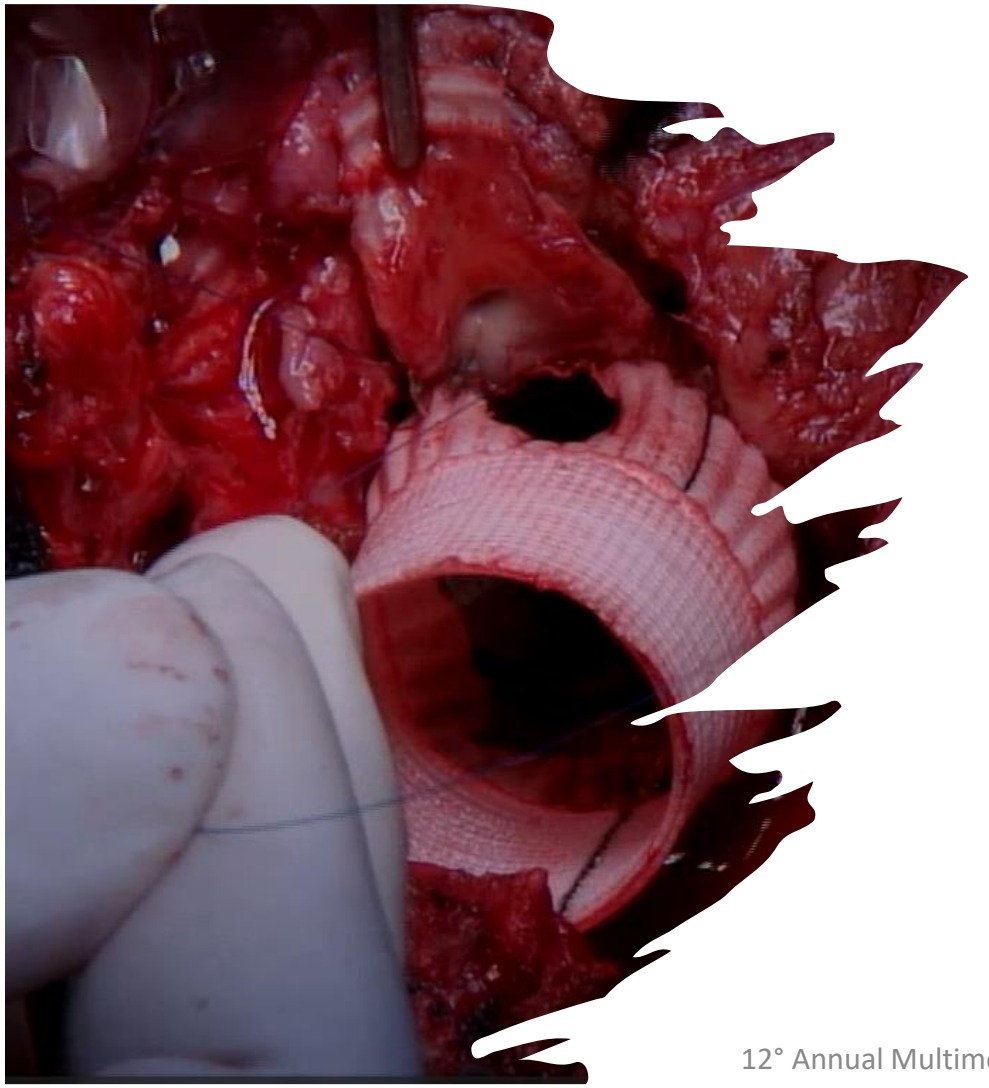
Frischertz BP Thoracic aortic dissection and rupture in conotruncal cardiac defects: A population-based study

# Indications in ACHD

- > 5,5 cm
- Concomitant surgery: residual VSD, Conduit replacement, AR
- AR: 4+
  - symptomatic
  - asymptomatic LV EF <50%
  - Concomitant surgery on the aorta or other valves

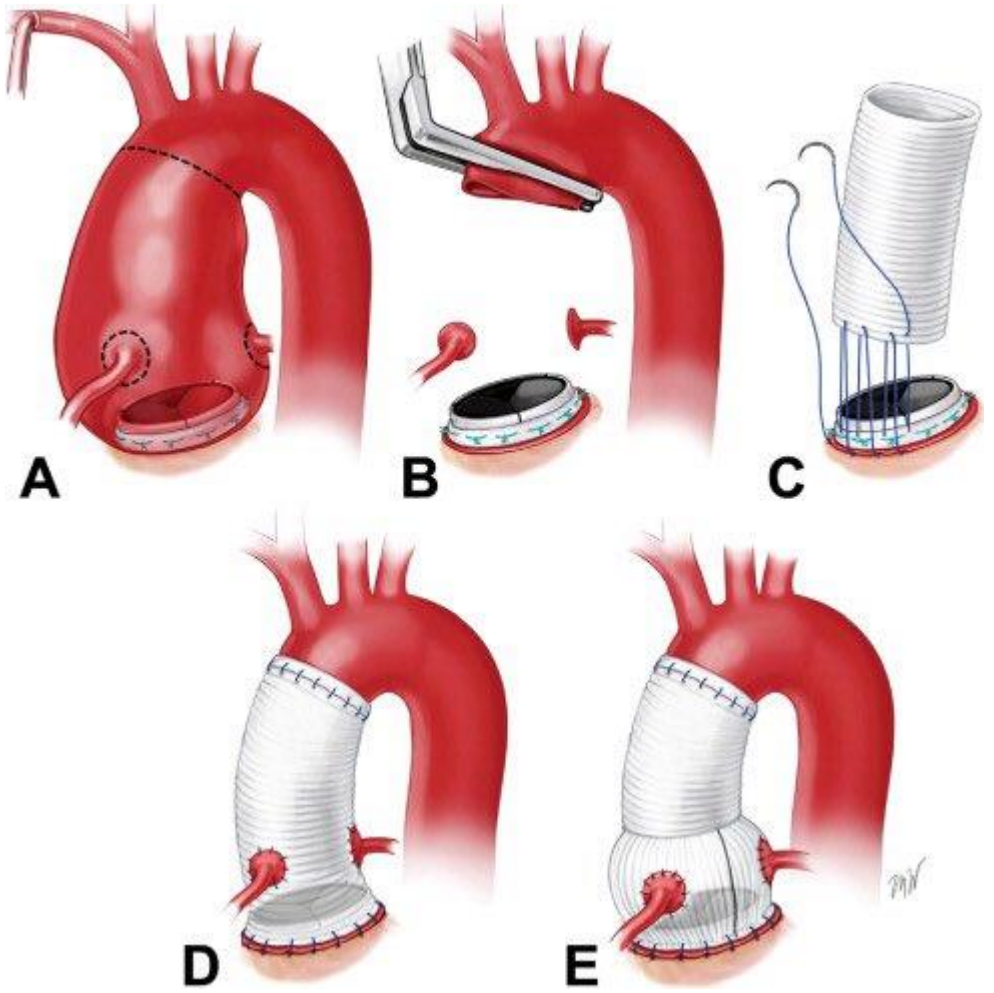
- > 27,5 mm/m2 (?)

# Type of operation



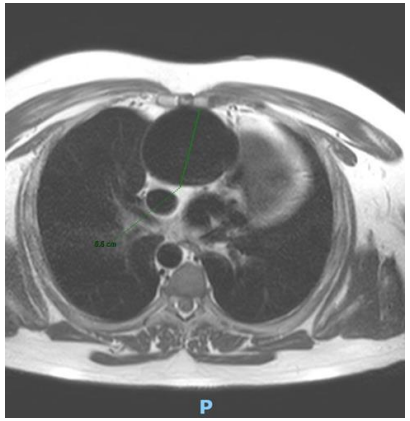
- Valve sparing:
  - Visualization of the aortic root and anatomy are different from normal heart
  - In presence of right sided conduit, the mobilization of the coronary arteries could be difficult
  - Presence of aortic valve regurgitation more than moderate
  - Lenght of cross clamp
- Bentall:
  - Mechanical: first choice in pts who already had multiple sternotomies
  - Biological: first choice in childbearing age, controindications to warfarin
  - Homograft: endocarditis

# Bentall operation



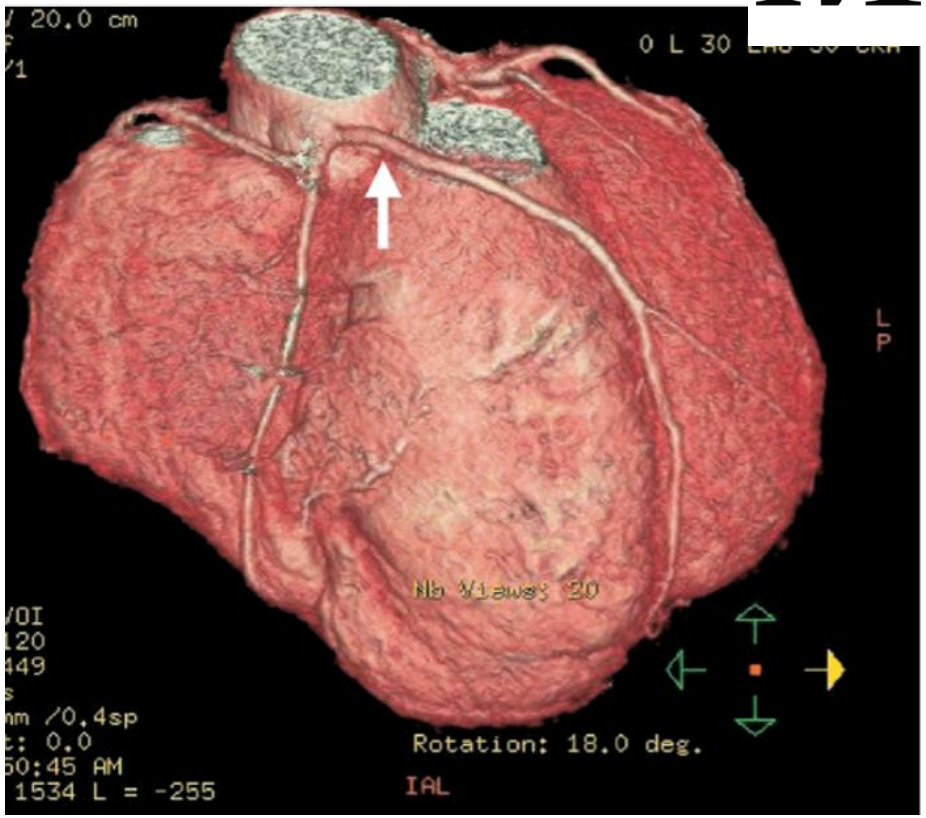
# Plan the re-entry:

- CT – MRI:
  - Distance of Ao and PA/conduit from the sternum
  - Coronary arteries: 15% of ToF pts and 30% of TGA pts have anomalous pattern of the coronary arteries
  - Single ventricle volume/function
  - MRI could suffer artifacts
- Periferal vessels doppler
- Echo: residual defects (VSD, PV, tunnel, LPA/MPA…)
- Check for AR: need for LV venting
- Echo contrast: check for residual L-R shunt




# Bentall in ToF:

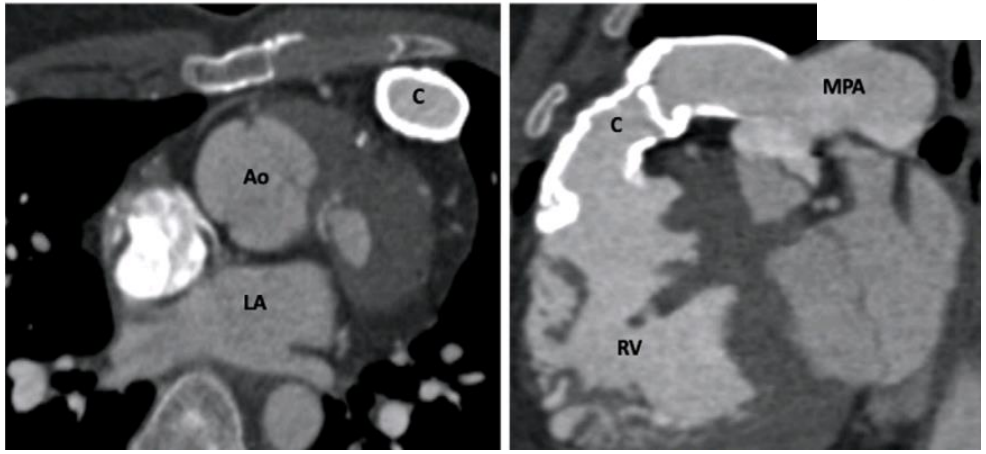
- Dilated more in the root. Aortic arch is normal
- Aorta is behind the sternum
- Anomalies of the coronary arteries are frequent (15%)

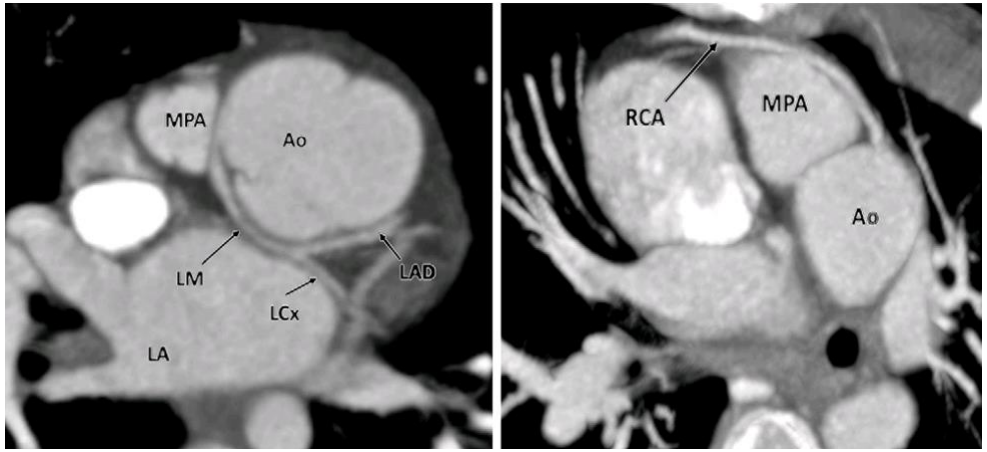



Dearani JA Management of the aortic root in adult patients with conotruncal anomalies Semin Thorac Cardiovasc Surg Pediatr Card Surg Ann 2009;12:122-129
Francois K Aortopathy associated with congenital heart disease: A current literature review Ann Pediatr Cardiol 2015;8(1):25-36

# Bentall in TGA:

- Dilatation is present in 50-60% of pts
- Dilated more at the sinus level. Aortic arch is normal
- Progression of AR over time is slow. Freedom from aortic root reoperation is 95% at 25 yrs
- Risk factors: previous PAB, VSD; age at op > 1 yr
- Pulmonary artery is behind the sternum
- CT is mandatory to assess distance of PA from the sternum and to check coronary arteries
- Coronary arteries are side by side

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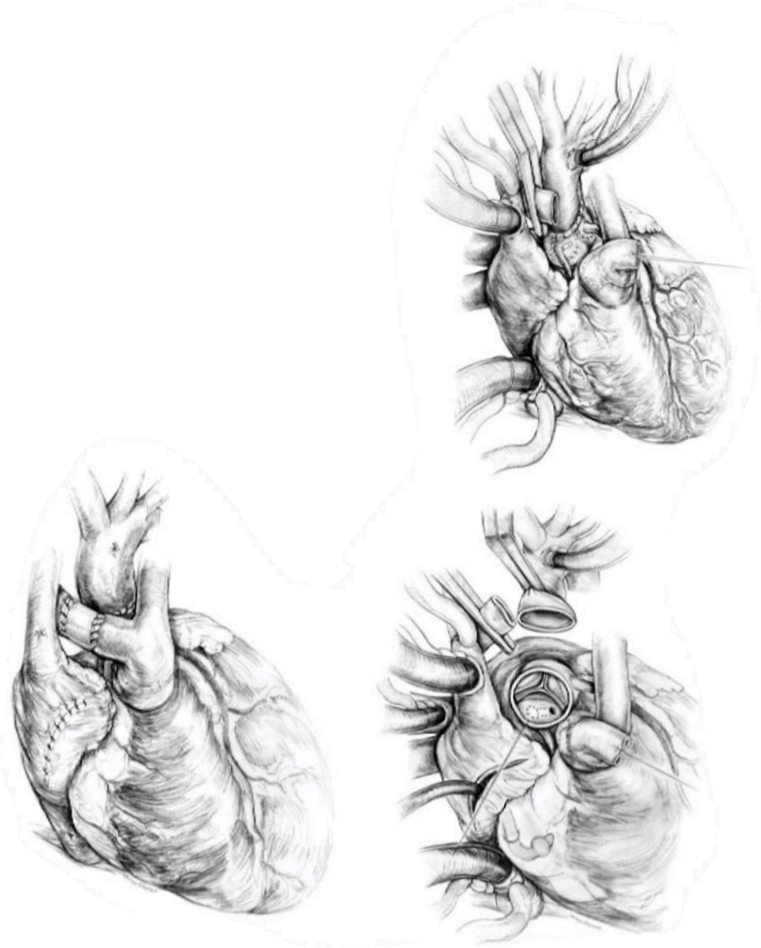
Angeli E Late reoperations after neonatal arterial switch operation for transposition of the great arteries Eur J Thorac Surg 2008;34:32-36

# Bentall in TGA:



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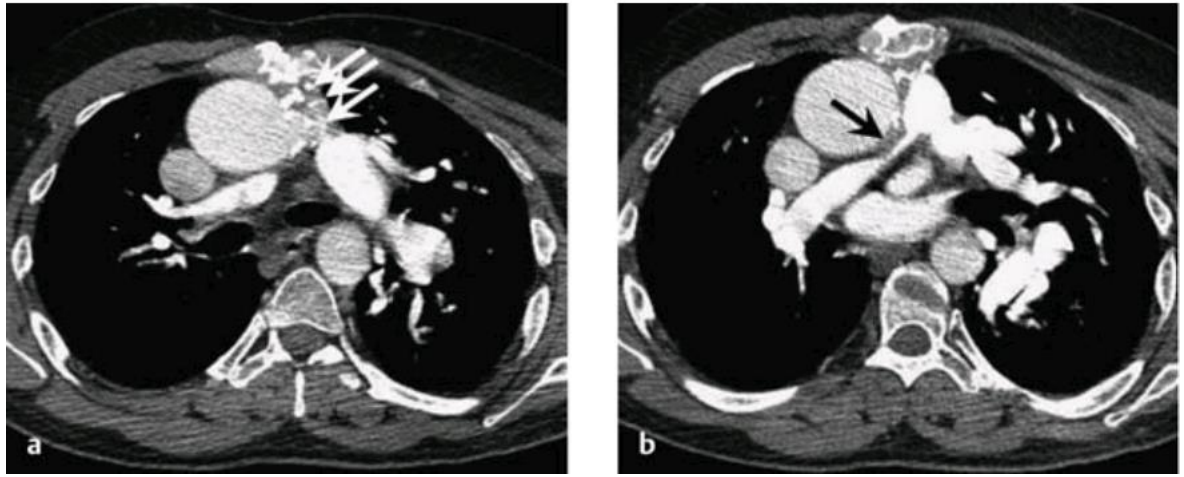
Angeli E Late reoperations after neonatal arterial switch operation for transposition of the great arteries Eur J Thorac Surg 2008;34:32-36

# Bentall in TA:

- Conduit is behind the sternum
- Harvesting of the coronary ostia could be difficult
- Calcification of the aorta
- Pathological pulmonary arteries
- Often it is a twin root

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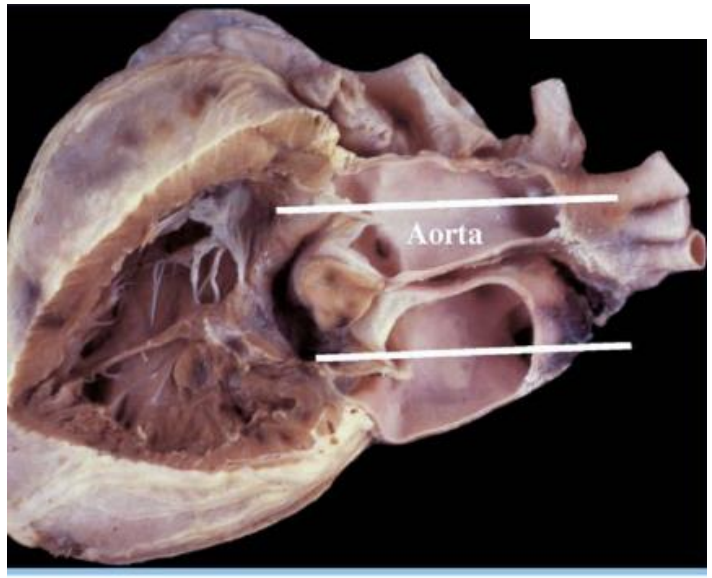
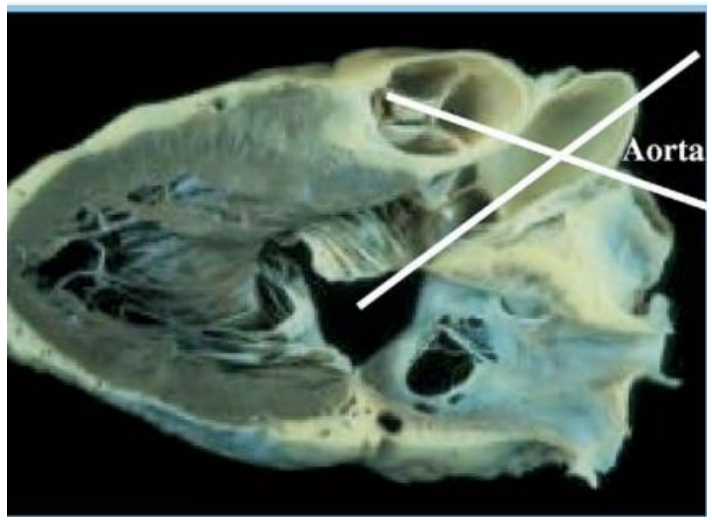
Francois K Aortopathy associated with congenital heart disease: A current literature review Ann Pediatr Cardiol 2015;8(1):25-36

# Bentall in DORV:

- Conduit could be behind the sternum
- Orientation of the aortic anular plane is different from normal
- Valve sparing is demanding due to different orientation of the aortic root

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# Bentall in UVH (Fontan/Norwood):

- After Fontan: dilatation of the root, normal descending aorta
  - >90% of Fontan pts with z-score > than 2 after 10 yrs
- After Norwood: dilatation of both ascending and descending aorta
  - ¼ pts with AR after 1 yr F-up, rare increase to more than mild
- Minimize cross clamp time

Dearani JA Management of the aortic root in adult patients with conotruncal anomalies Semin Thorac Cardiovasc Surg Pediatr Card Surg Ann 2009;12:122-129
Francois K Aortopathy associated with congenital heart disease: A current literature review Ann Pediatr Cardiol 2015;8(1):25-36

# Focus on Aortic Root:

- Enlarged aortic root is a common finding in CHD and is multifactorial
- Risk of dissection is rare but described for diameter > 60 mm
- Bentall should be considered if there is need for concomitant surgery
- Re-entry should be carefully planned
- Carefully inspection for residual defects

# Aortic Valve:

- The bicuspid aortic valve: most common congenital anomaly
- Aortic valve regurgitation in conotruncal anomalies
- Congenital aortic stenosis
- Quadricuspid aortic valve in truncus

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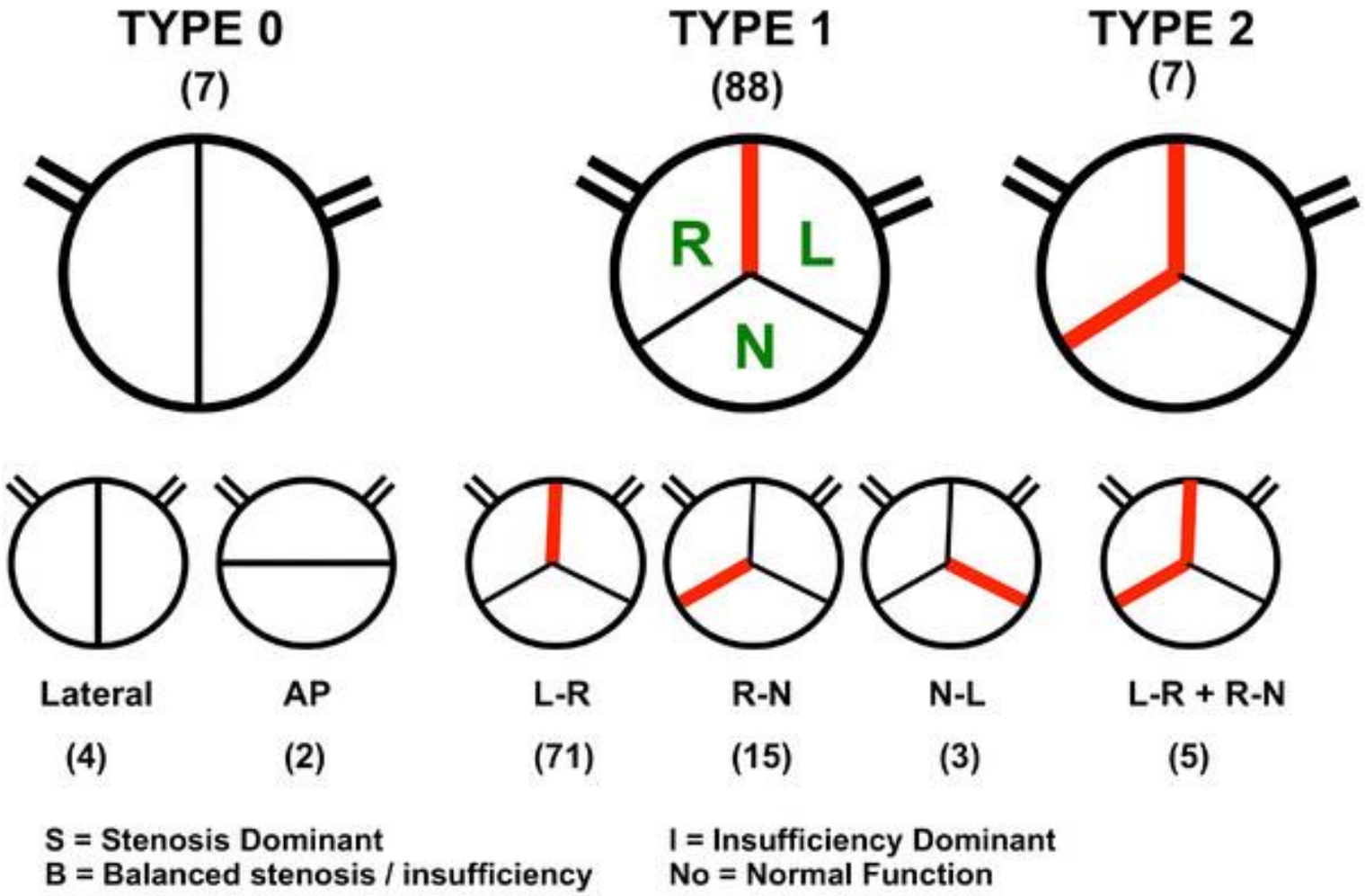
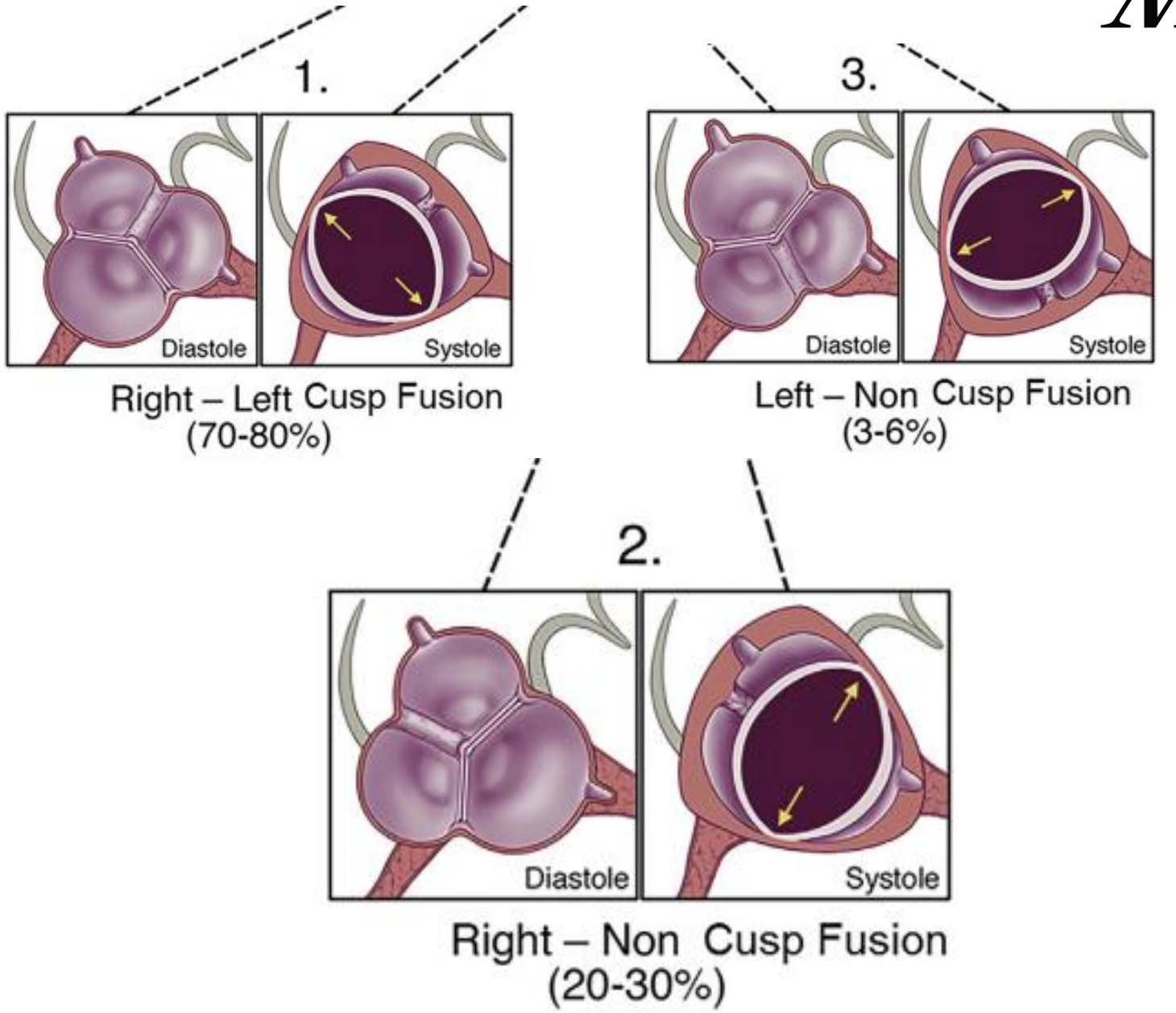
# Sievers Classification

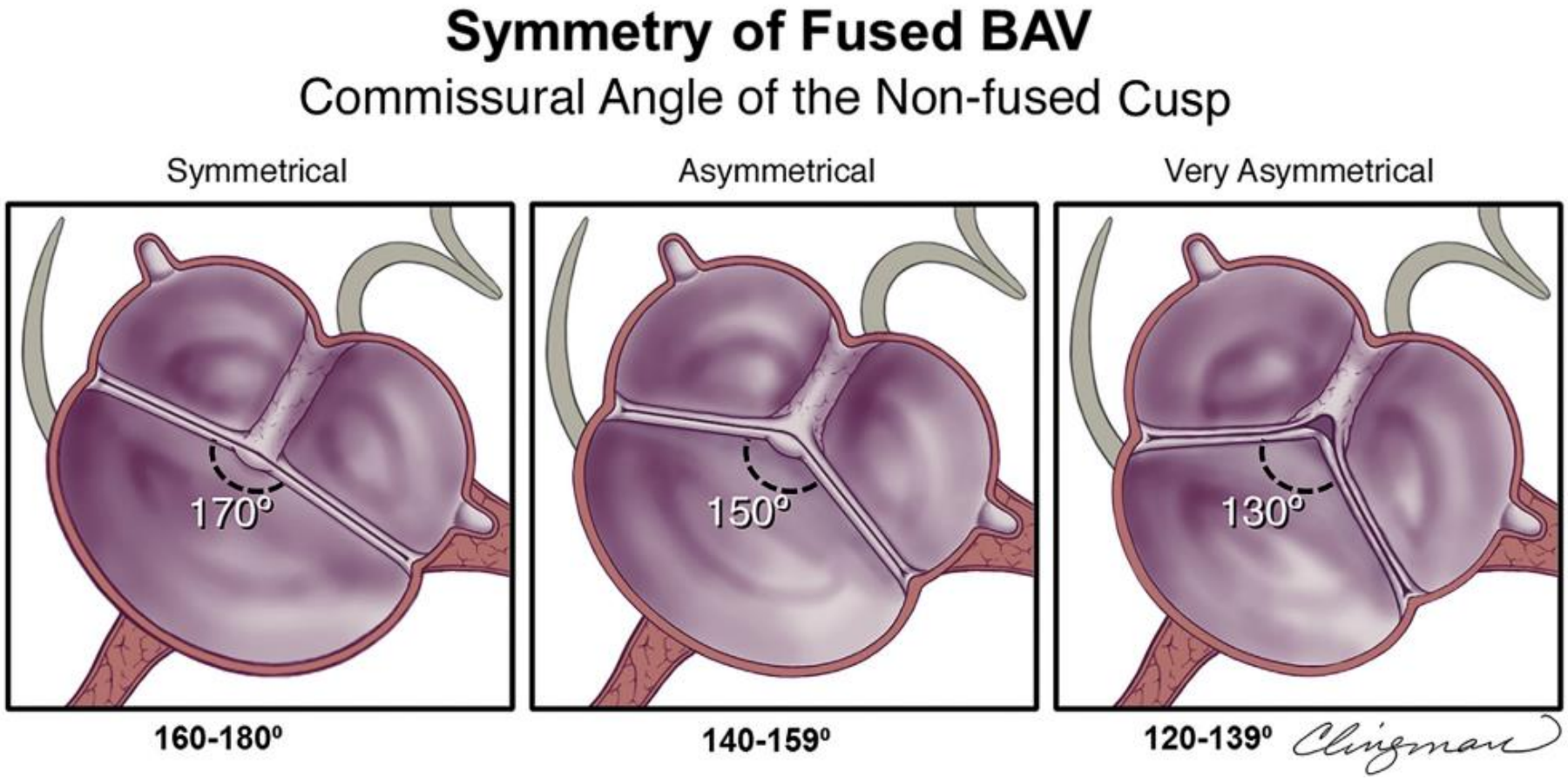


# Fused BAV

- Three sinus of Valsalva
- 2 cusps
- 2 commissures
- Raphe  common, visible or not

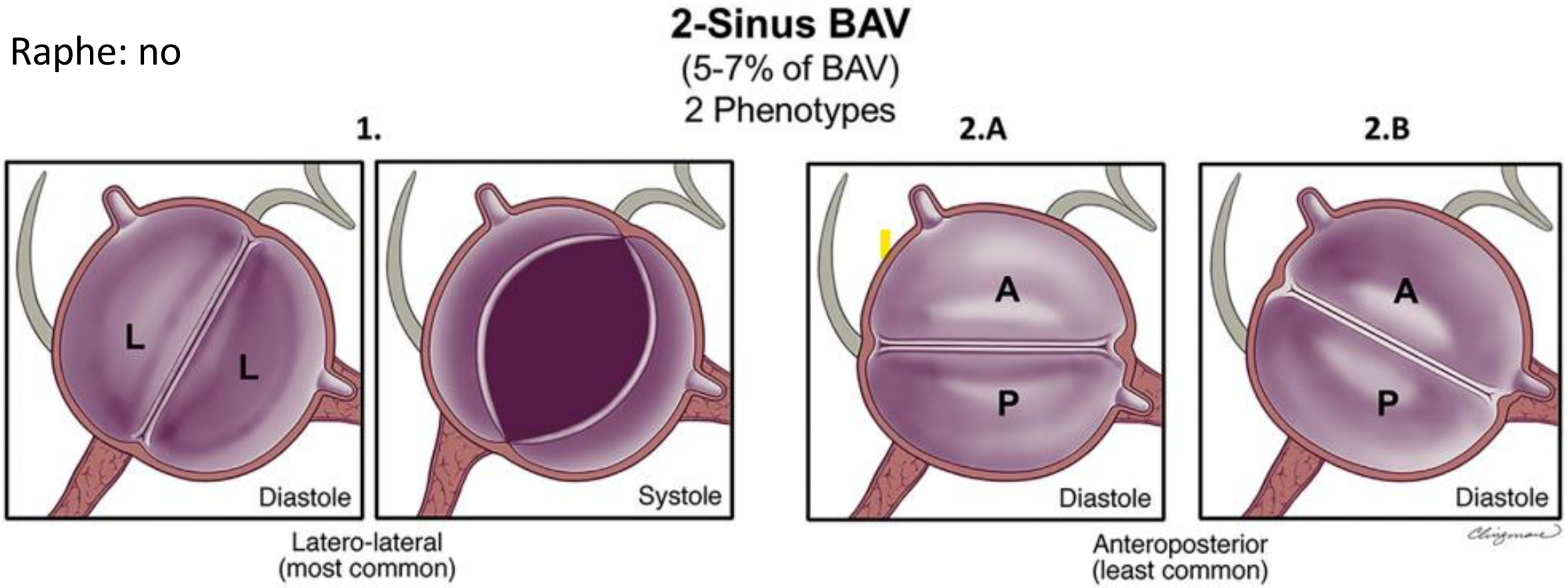


# Orientation of the commissures



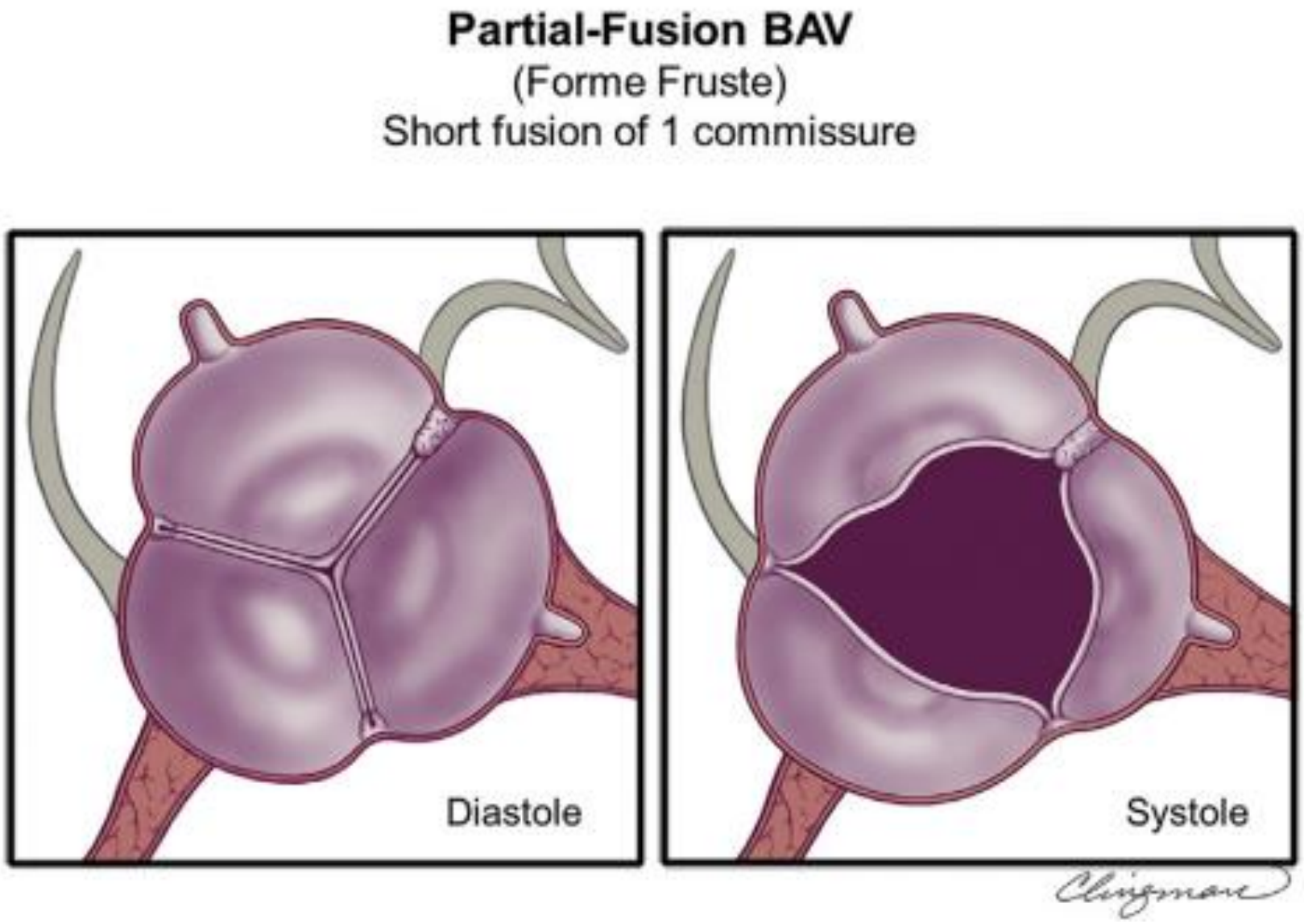
# 2 sinus BAV

- 2 sinuses
- 2 cusps
- 2 commissures
- Raphe: no

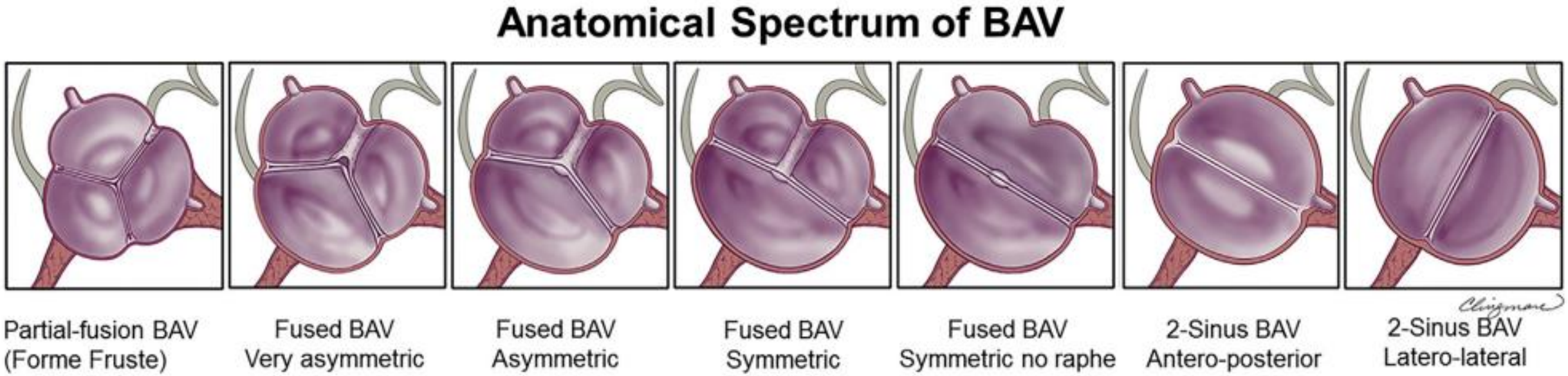


# Partial Fused BAV

- Three sinus of Valsalva
- 3 cusps
- 3 commissures (1 fused < 50%)
- Raphe: small

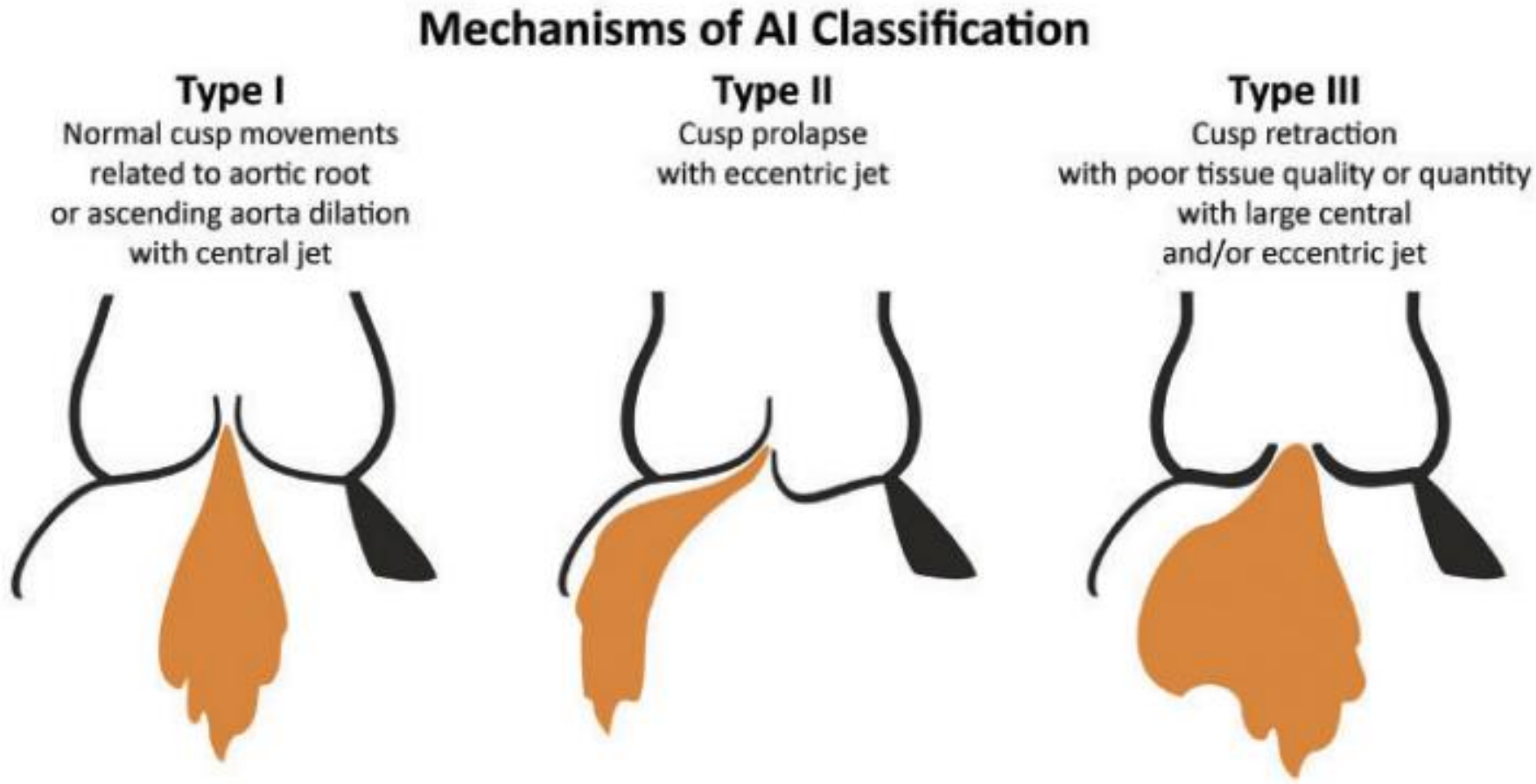


# Continuum….




Michelena H. JTCVS 2021; 162:e383-e414

# Functional classification



# Why repair?

Bouhout I. Semin Thorac Cardiovasc Surg Pediatr Card Surg Ann 2022:25:28-37

**Table 1** The Relative Merit of the Ross Procedure, Tissue Valves, Mechanical Prosthesis, Aortic Valve Repair, and Ozaki Procedure in Young and Middle-Aged Adults

| | Ross Procedure | Stented Tissue Valves | Mechanical Prosthesis | AVNeo (Ozaki) Procedure | Aortic Valve Repair |
|---|---|---|---|---|---|
| Survival | Equivalent to sex- and gender-matched general population up to 10 years after surgery | Under expected survival | Under expected survival | Excellent | Excellent |
| Valve-related complications | 0.5% per patient-year | 0.5-1% per patient-year | 1% per patient-year | No long-term data for this patient population, more early leaflet thrombosis/ stiffening? | Lower than prosthetic valves |
| Aortic valve reintervention | 1% per patient-year for AS<br>2% per patient-year for AR | 1-2% per patient-year | 0.5% per patient-year | No long-term data for this patient population, but expected in a majority at midterm | Expected in a majority at midterm after "complex repairs" |
| Quality of life | Restored quality of life | Uncertain | Lower quality of life when compared to the Ross | Restored QoL (midterm) | Restored QoL (midterm) |
| Pregnancy | Low risk of fetal and maternal complications | Low risk of fetal and maternal complications | Significant risk of fetal and maternal complications | Low risk of fetal and maternal complications | Low risk of fetal and maternal complications |
| Hemodynamic performance | Closest profile to native aortic valve | The lowest aortic orifice area<br>Up to 30% rate of PPM | Suboptimal hemodynamics<br>20-30% rate of PPM | Excellent initially, but likely continuous decline midterm | Residual and progressing AS and AR in "complex repairs" |
| Reproducibility | Expertise needed | High | High | High | Expertise needed |
| Anatomic milieu | Important | Less important | Less important | Less important | Very important |

AR, aortic regurgitation; AS, aortic stenosis; PPM, prosthesis-patient mismatch.

# Trends of aortic valve surgery

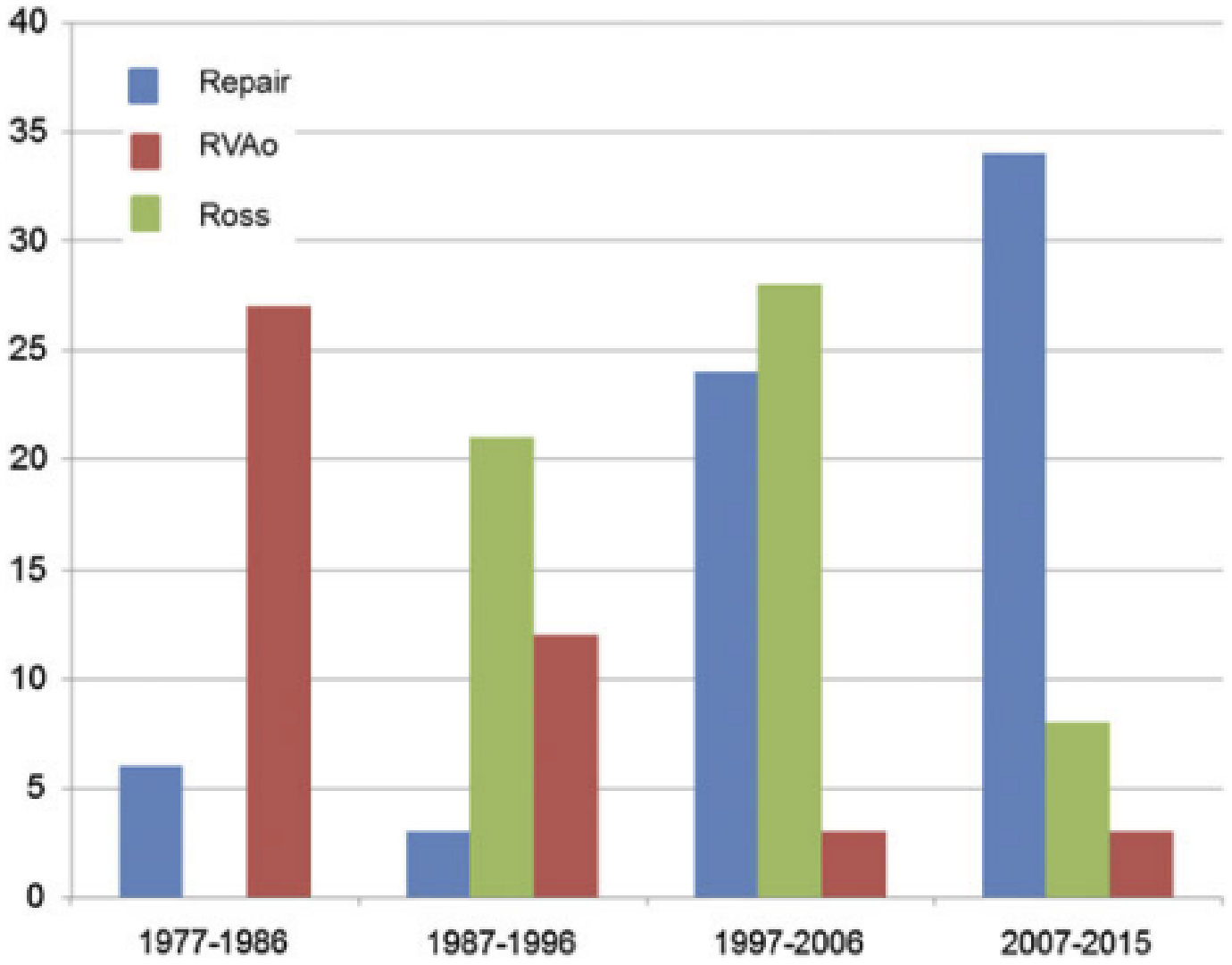
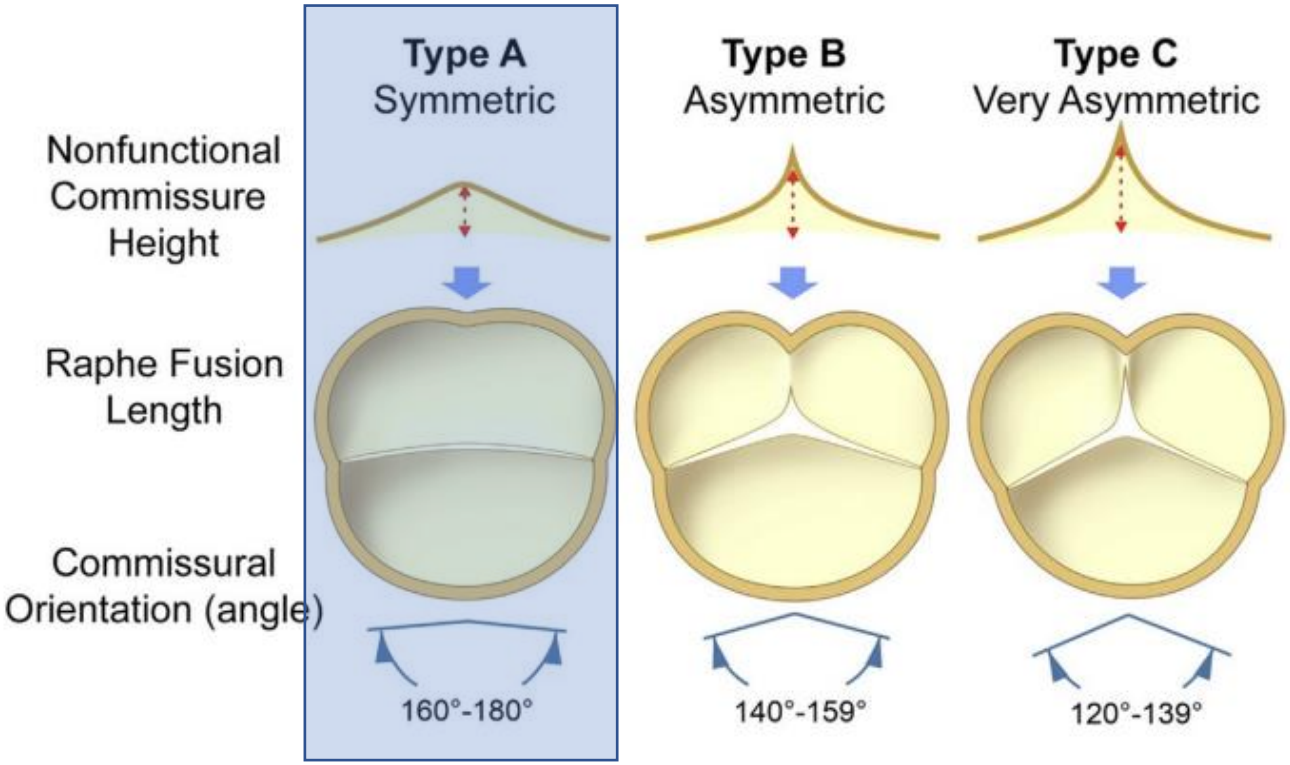
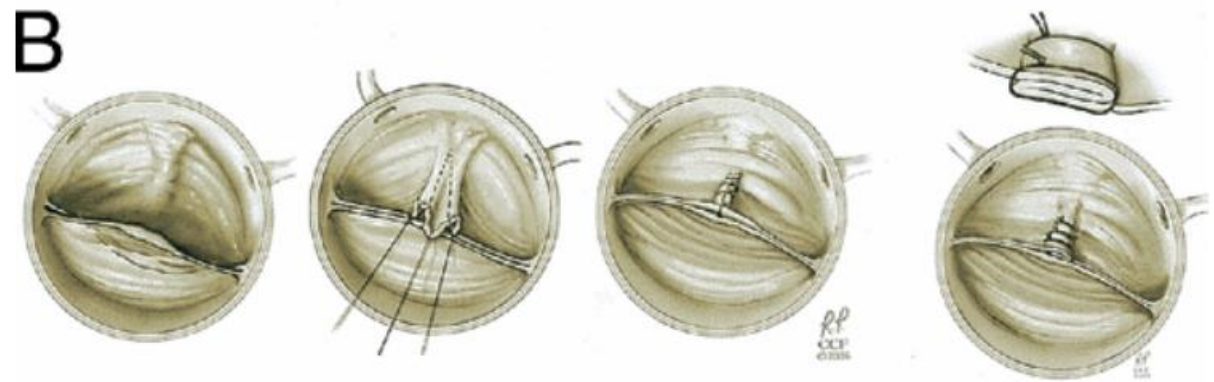



**Figure 1:** Number of aortic valve (AV) repair (blue bar), AV replacement (red bar) and Ross procedures (green bar) by decade.

# Raphe plication

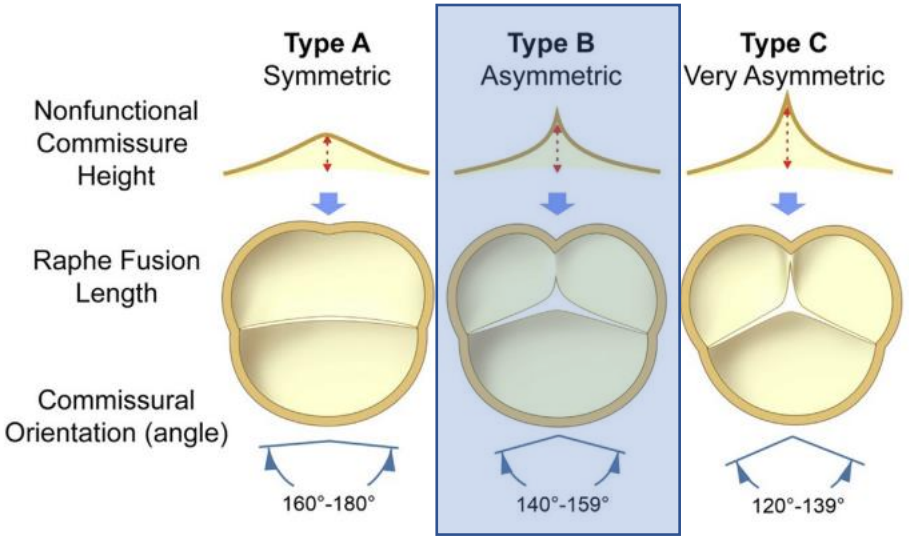




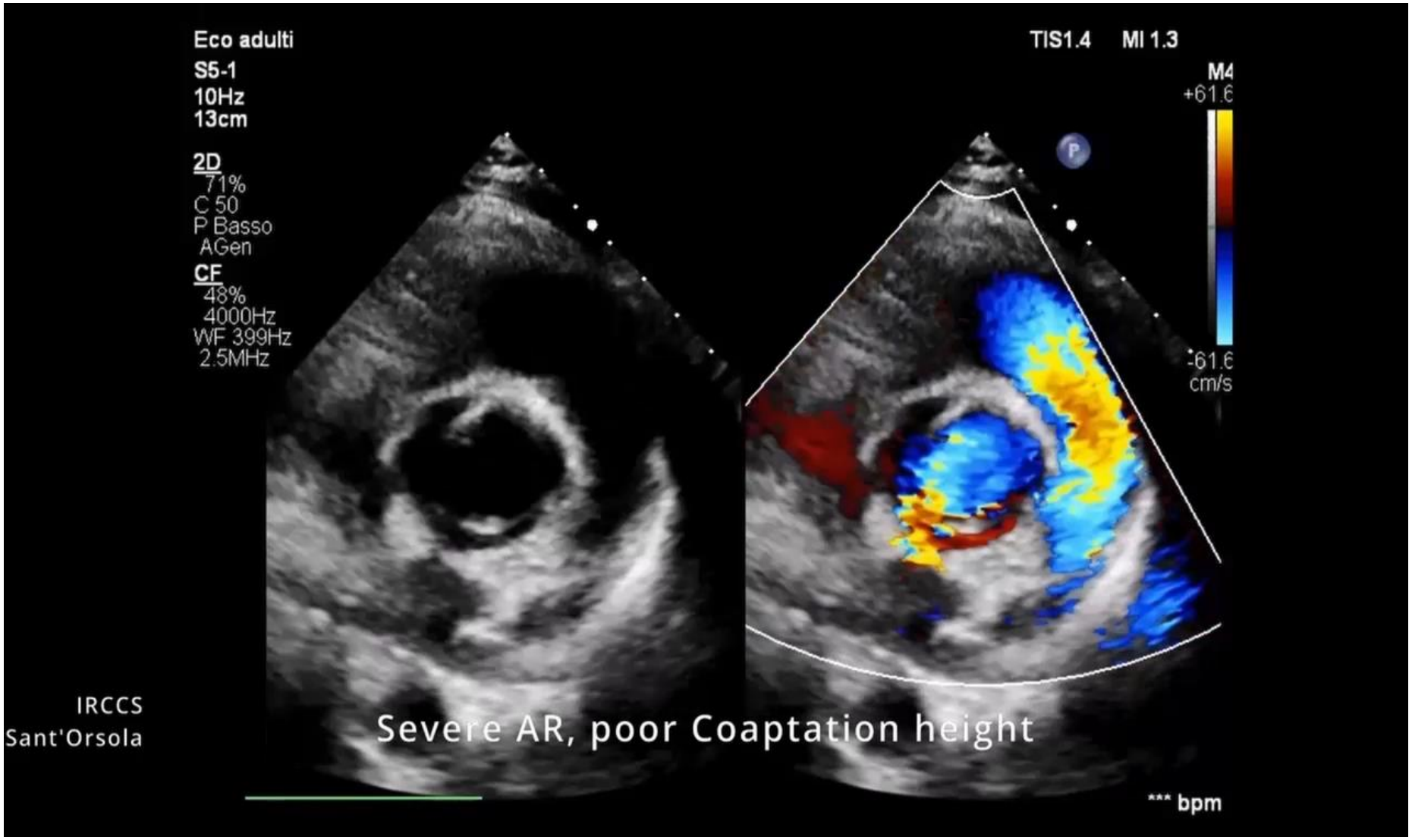

Petterson GB JACC 2008; 52:40-9

# Asymmetric

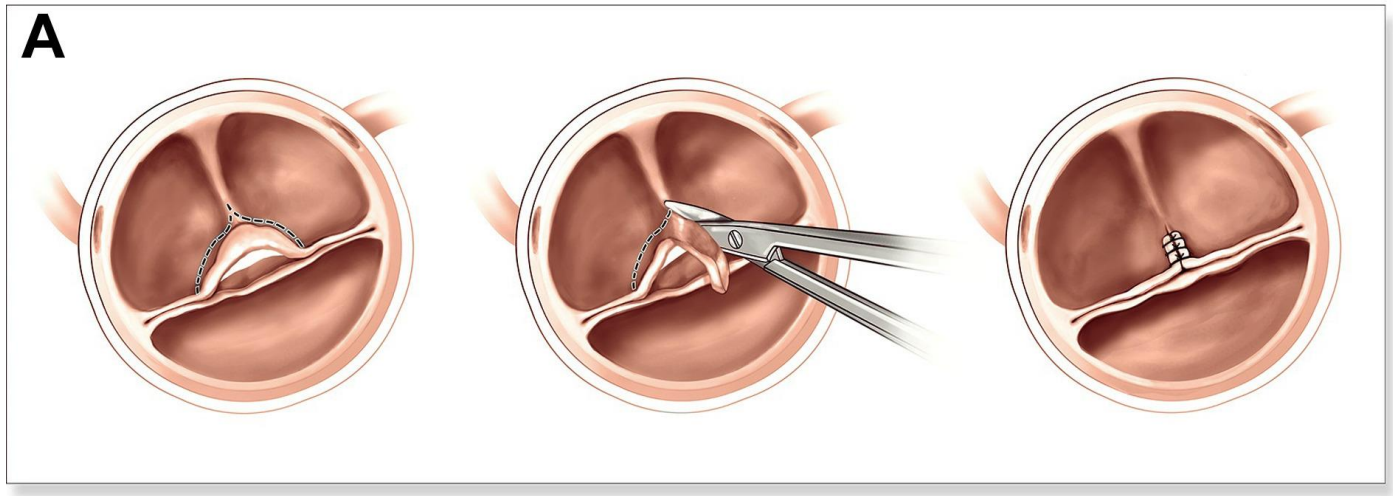

Type A
Symmetric
Type B
Asymmetric
Type C
Very Asymmetric
Nonfunctional Commissure Height
Raphe Fusion Length
Commissural Orientation (angle)
160°-180°
140°-159°
120°-139°

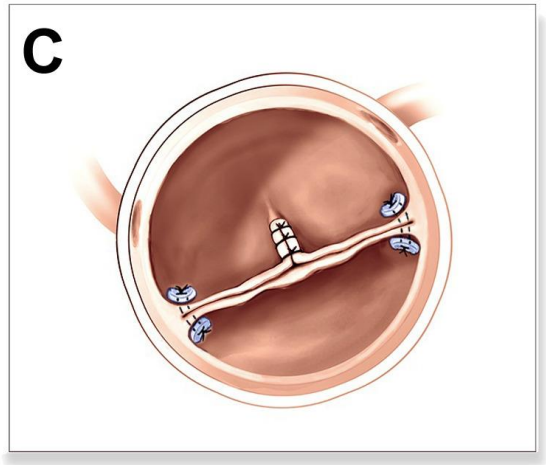

ZACEK

# Plication

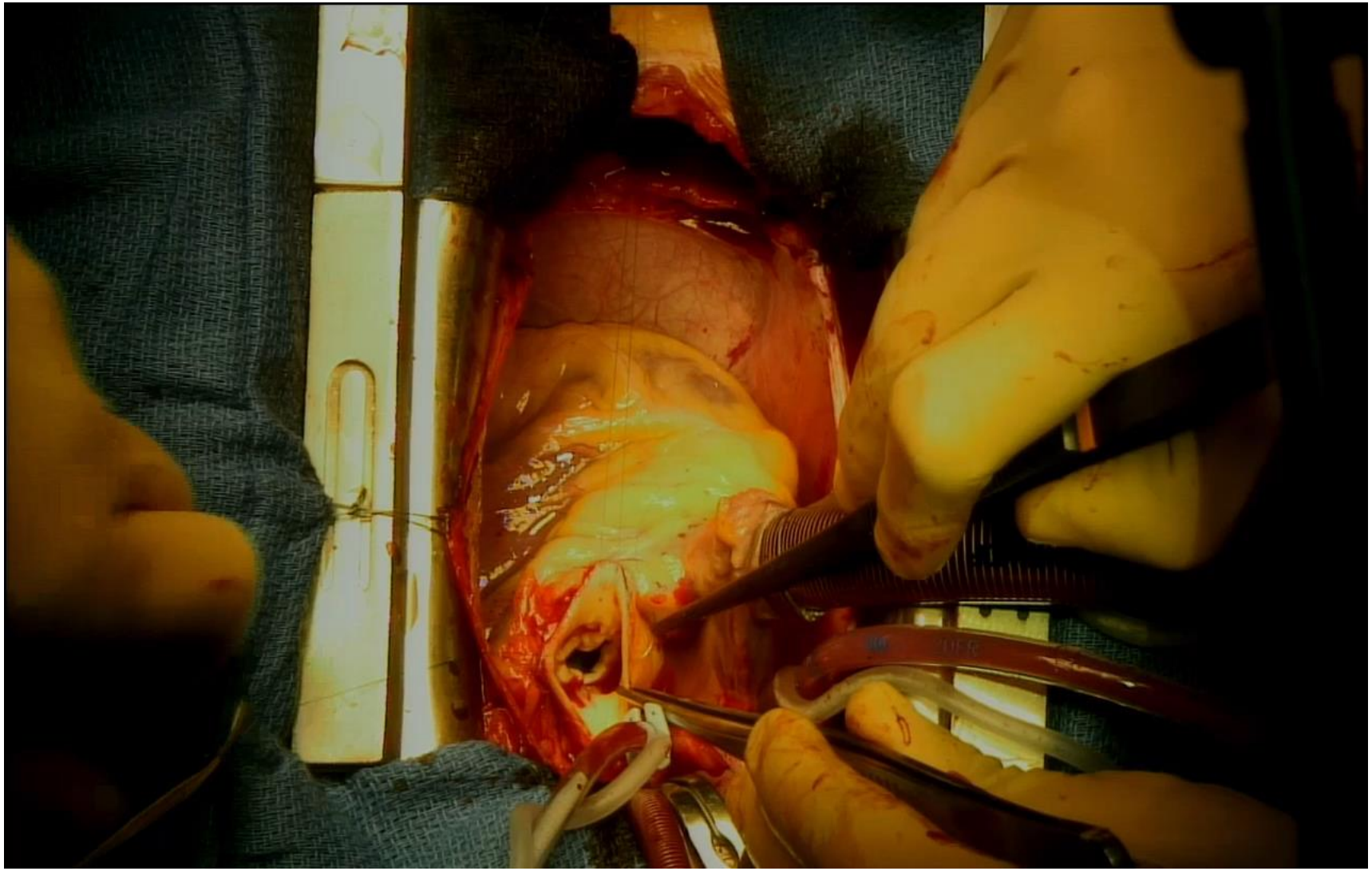

Eco adulti
S5-1
10Hz
13cm
2D
71%
C 50
P Basso
AGen
CF
48%
4000Hz
WF 399Hz
2.5MHz
TIS1.4 MI 1.3
M4
+61.6
-61.6
cm/s
IRCCS
Sant'Orsola
Severe AR, poor Coaptation height
*** bpm

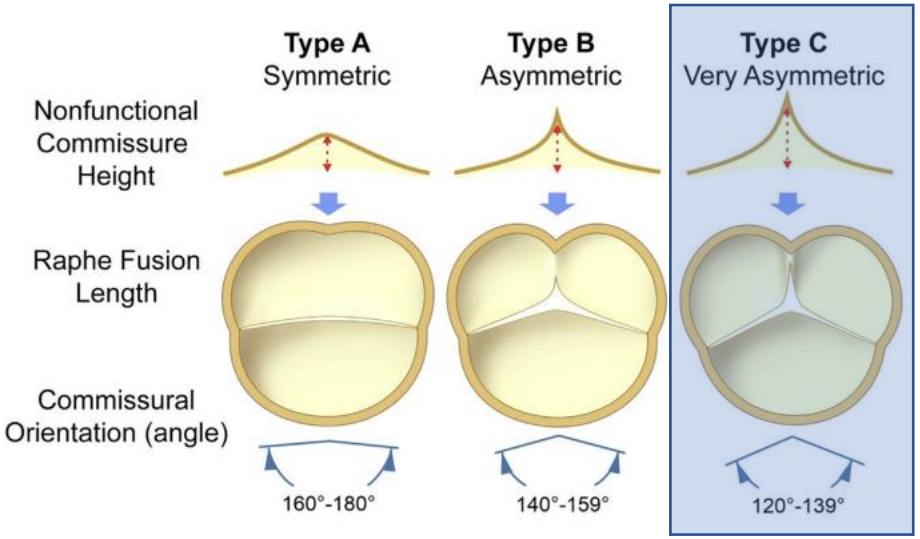
# Raphe resection



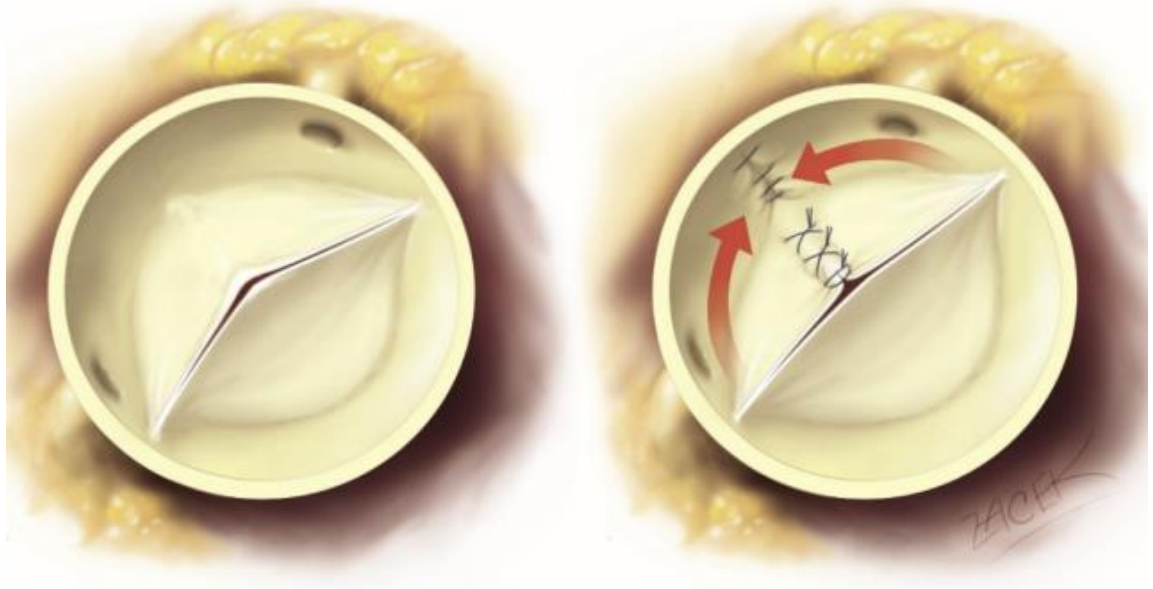



# Resection

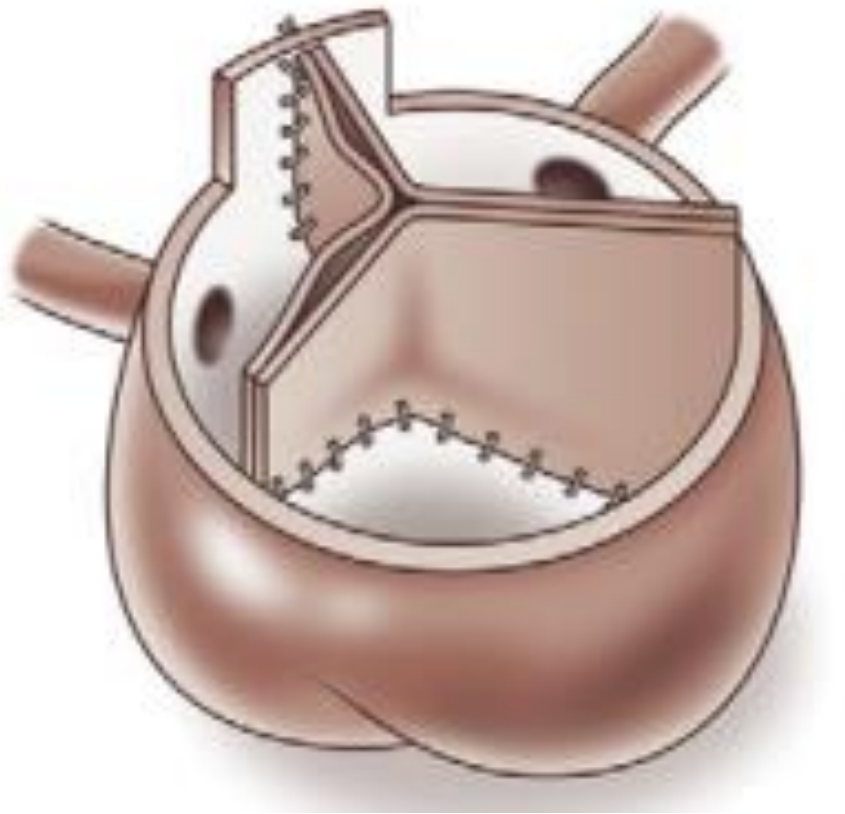




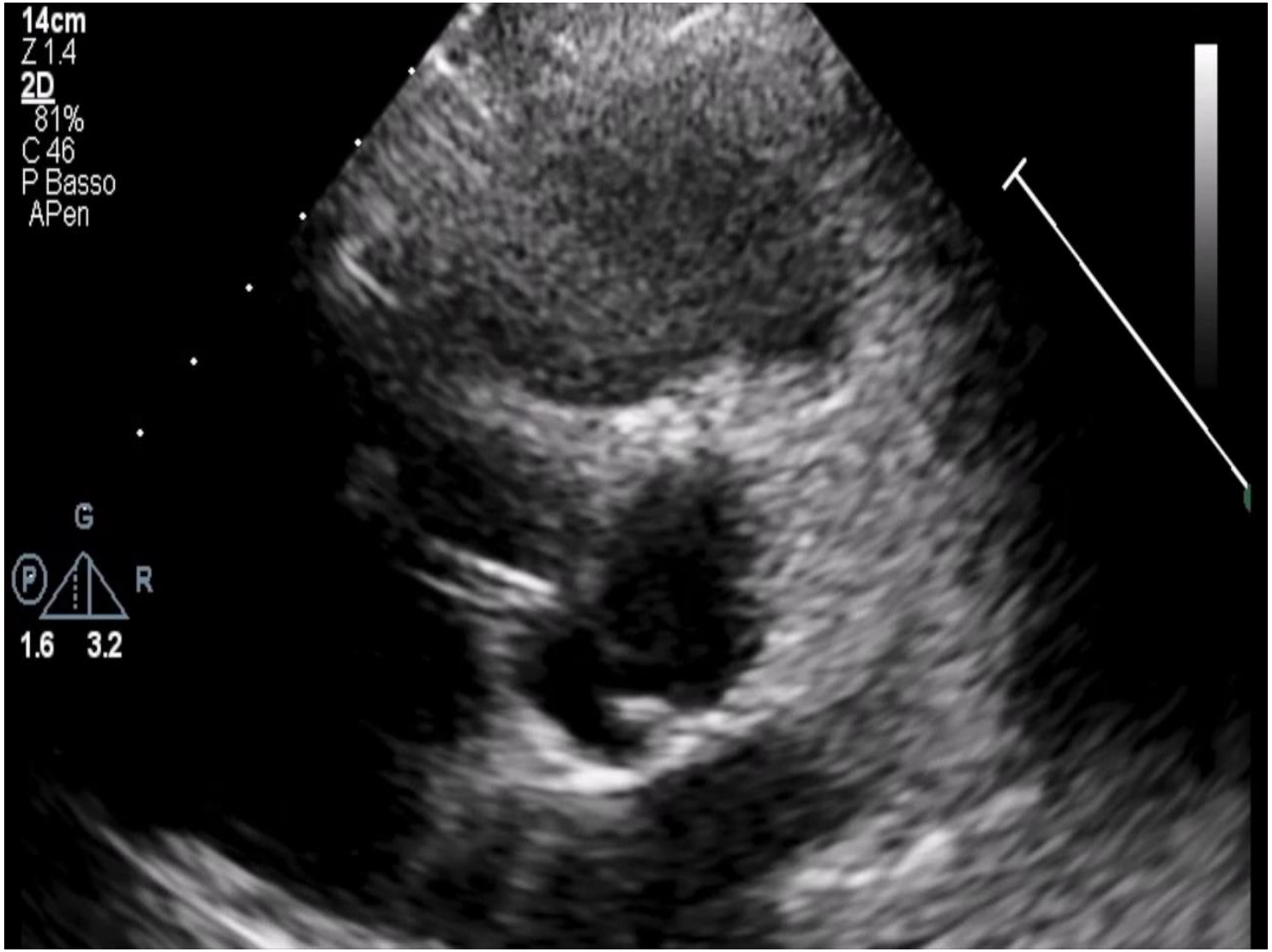

# Very asymmetric aortic valve



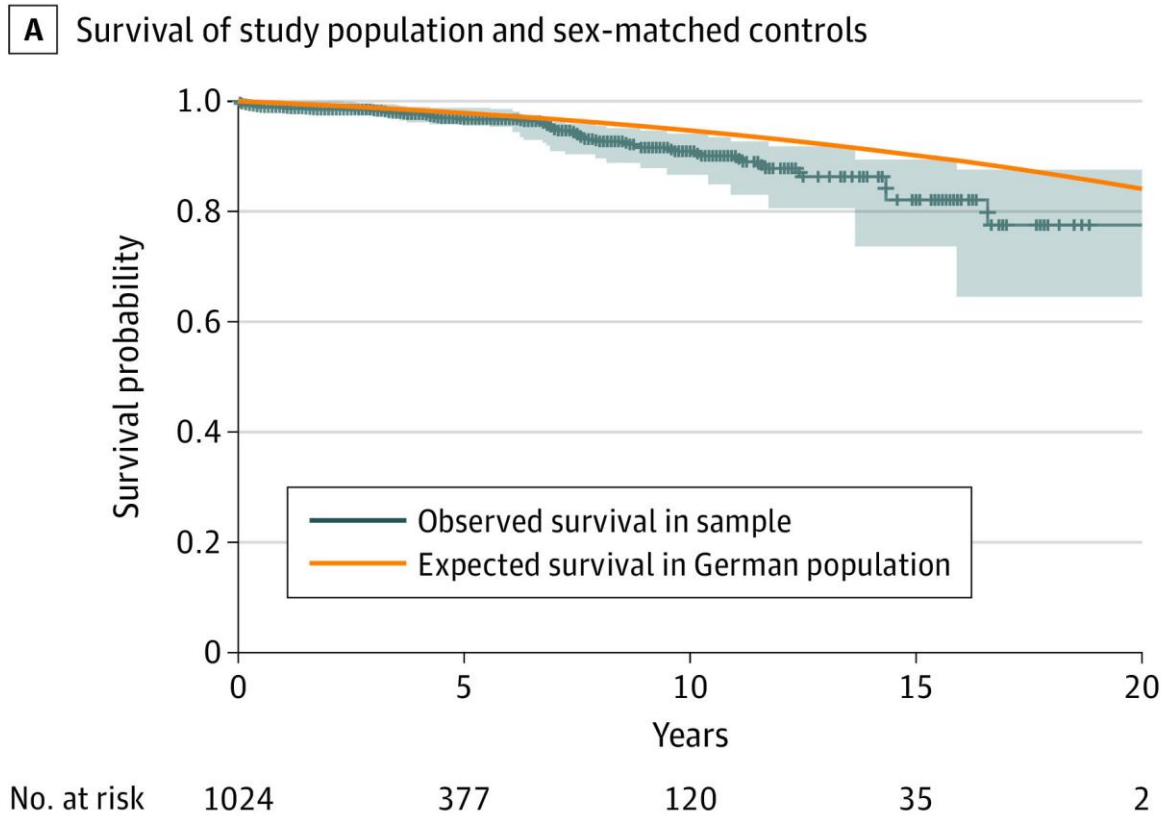
# Tricuspidalization

# Partial fused: Very asymmetric



# Results after long-term aortic valve repair

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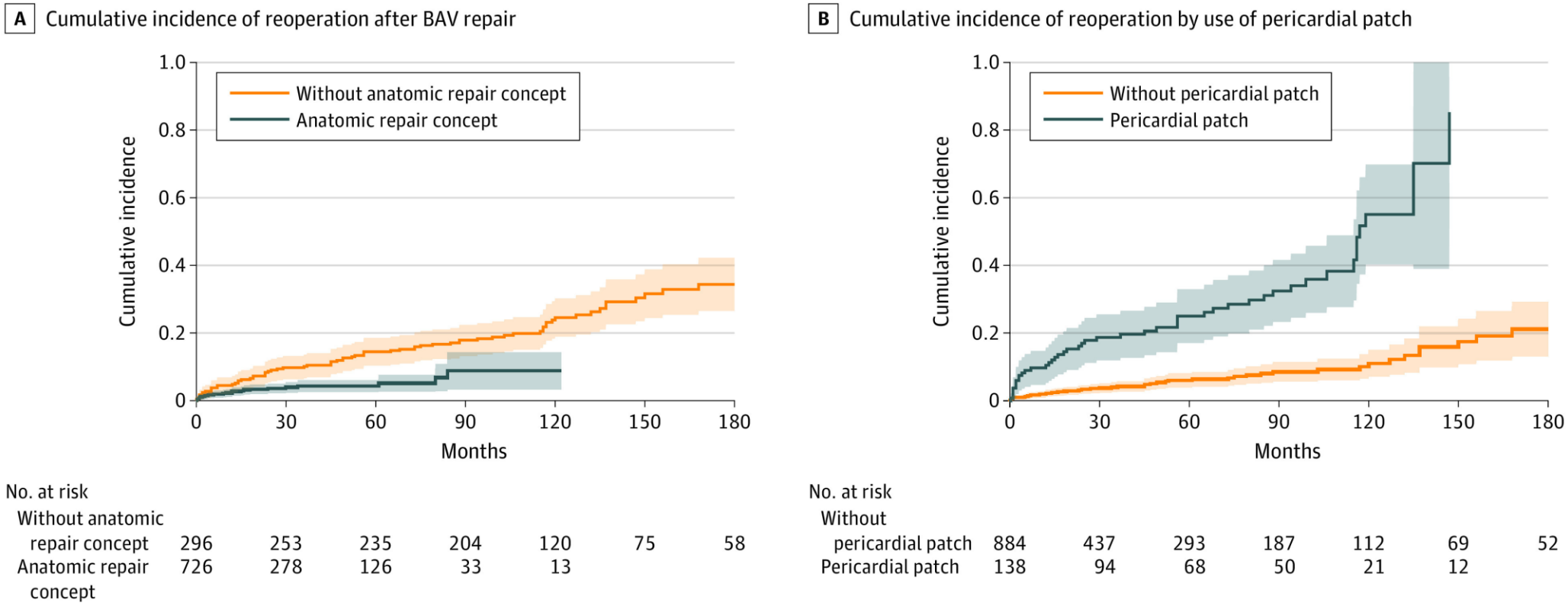
# Impact of an anatomic approach on results:

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# Risk factors

**Associations With Time to Reoperation From Fine-Gray Models**

| Characteristic | Crude model | | Adjusted model | |
|---|---|---|---|---|
| | SHR (95% CI) | *P* value | SHR (95% CI) | *P* value |
| Annuloplasty[a] | 0.52 (0.32-0.86) | .01 | 0.67 (0.37-1.19) | .17 |
| Commissural orientation[b] | | | | |
| Tricuspid-like vs symmetric[b] | 0.93 (0.43-2.03) | .86 | 0.74 (0.34-1.63) | .45 |
| Asymmetric without modification vs symmetric[b] | 3.87 (2.09-7.17) | <.001 | 1.95 (1.02-3.72) | .04 |
| Modified asymmetric vs symmetric | 0.79 (0.37-1.66) | .53 | 0.99 (0.46-2.12) | .97 |
| Cusp calcification[c] | 2.44 (1.63-3.64) | <.001 | 1.78 (1.14-2.77) | .01 |
| Pericardial patch | 5.25 (3.52-7.82) | <.001 | 5.25 (3.52-7.82) | <.001 |
| Root replacement[d] | 0.47 (0.31-0.72) | .001 | 0.71 (0.45-1.15) | .16 |

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# Results according to surgical performance

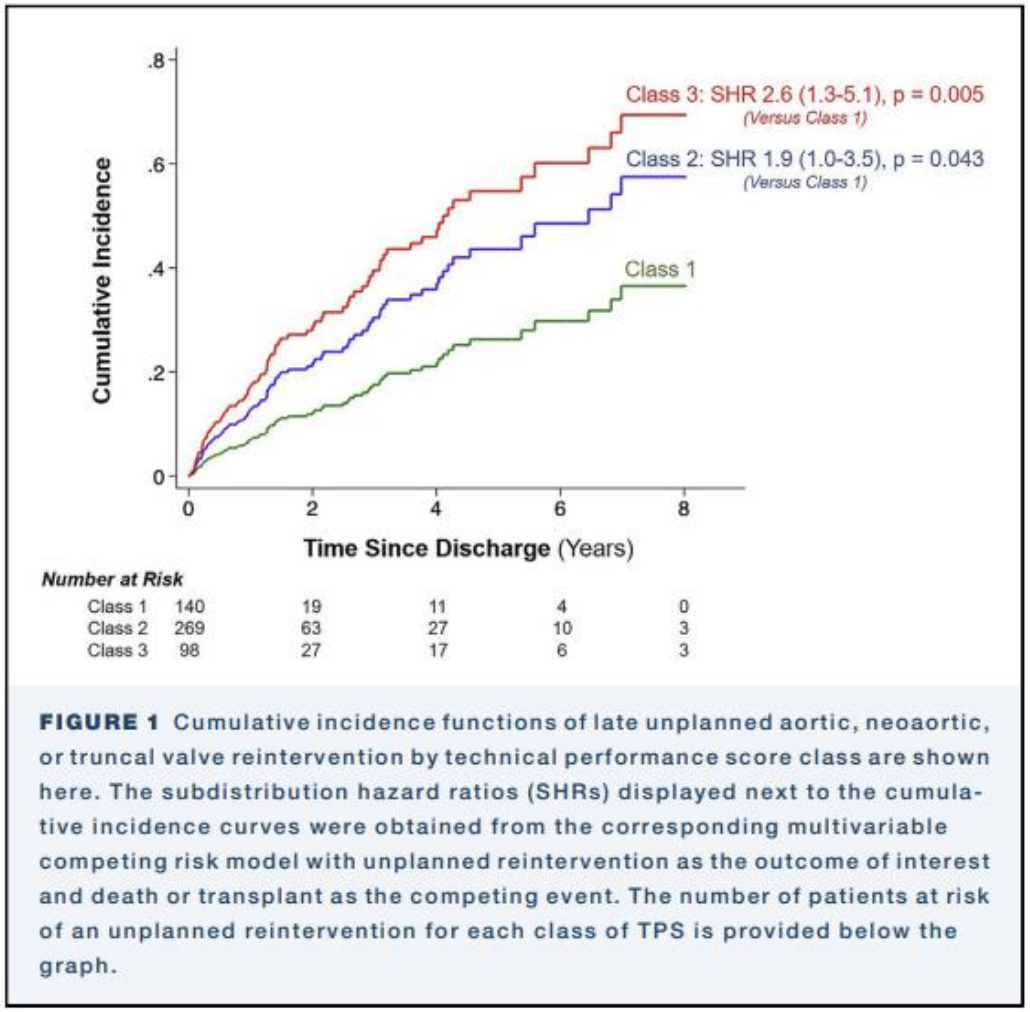
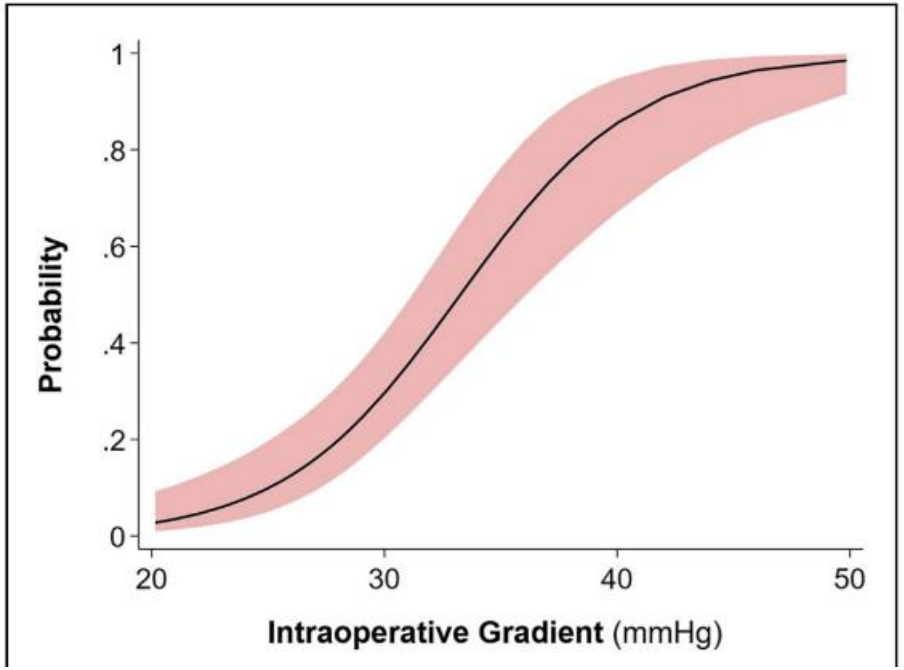



**FIGURE 1** Cumulative incidence functions of late unplanned aortic, neoaortic, or truncal valve reintervention by technical performance score class are shown here. The subdistribution hazard ratios (SHRs) displayed next to the cumulative incidence curves were obtained from the corresponding multivariable competing risk model with unplanned reintervention as the outcome of interest and death or transplant as the competing event. The number of patients at risk of an unplanned reintervention for each class of TPS is provided below the graph.




507 pts
Early deaths 4,3%

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# Results according to surgical performance

**TABLE 2 Cox Proportional Hazards Models of Postdischarge Mortality**

| | Univariable Analysis | | Multivariable Analysis | |
|---|---|---|---|---|
| Factor | HR (95% CI) | *P* Value | HR (95% CI) | *P* Value |
| Technical performance score[a] | | | | |
| Class 2 | 2.1 (0.5-9.8) | .33 | 2.3 (0.5-10.7) | .29 |
| Class 3 | 5.1 (1.1-23.3) | .037 | 5.3 (1.1-25.2) | .038 |
| Age[b] | | | | |
| Neonate (birth-1 month) | 1.2 (0.2-9.3) | .86 | 1.9 (0.2-15.5) | .57 |
| Infant (1 month-1 year) | 6.0 (2.5-14.5) | <.001 | 6.8 (2.6-17.9) | <.001 |
| Adult (≥18 years) | 0.5 (0.1-4.0) | .52 | 0.4 (0.04-3.3) | .38 |
| Prematurity | 0.7 (0.1-4.8) | .67 | 0.3 (0.03-3.6) | .38 |
| Noncardiac anomaly or syndrome | 0.9 (0.3-2.7) | .87 | 0.7 (0.2-2.5) | .61 |
| Preoperative risk factor[c] | 1.1 (0.4-3.1) | .81 | 1.3 (0.5-3.8) | .62 |
| Single-ventricle physiology | 2.0 (0.7-5.9) | .23 | 1.2 (0.3-5.5) | .81 |
| Concomitant procedure | 2.4 (0.6-10.5) | .23 | 2.0 (0.4-9.0) | .38 |
| Valve repair category[d] | | | | |
| Neoaortic valve repair | 1.3 (0.4-4.5) | .67 | 0.8 (0.2-3.0) | .68 |
| Truncal valve repair | 0.7 (0.1-5.2) | .72 | 0.8 (0.1-7.2) | .81 |

[a]Class 1 as reference; [b]Age 1-17 years as reference; [c]Presence of at least 1 major preoperative risk factor, including cardiopulmonary resuscitation, shock, extracorporeal membrane oxygenation, malignant ventricular arrhythmia or high-grade atrioventricular block, mechanical ventilation, renal failure, liver failure, sepsis, necrotizing enterocolitis, stroke, seizure, or intracerebral hemorrhage, and prior noncardiac surgery; [d]Native aortic valve repair as reference. Model C-index: TPS only, 0.643; covariates only, 0.736; TPS and covariates, 0.787. HR, hazard ratio; TPS, technical performance score.

**TABLE 1 Competing Risk Models of Postdischarge Unplanned Aortic, Neoaortic, or Truncal Valve Reintervention**

| | Univariable Analysis | | Multivariable Analysis | |
|---|---|---|---|---|
| Factor | SHR (95% CI) | *P* Value | SHR (95% CI) | *P* Value |
| Technical performance score[a] | | | | |
| Class 2 | 2.1 (1.1-3.8) | .018 | 1.9 (1.0-3.5) | .043 |
| Class 3 | 2.9 (1.5-5.6) | .002 | 2.6 (1.3-5.1) | .005 |
| Age[b] | | | | |
| Neonate (birth-1 month) | 5.6 (3.1-9.8) | <.001 | 5.0 (2.7-9.3) | <.001 |
| Infant (1 month-1 year) | 2.0 (1.2-3.5) | .014 | 1.3 (0.7-2.4) | .44 |
| Adult (≥18 years) | 0.5 (0.3-1.2) | .12 | 0.7 (0.3-1.7) | .44 |
| Prematurity | 1.6 (0.8-3.1) | .21 | 1.3 (0.6-2.5) | .52 |
| Noncardiac anomaly or syndrome | 0.7 (0.4-1.2) | .17 | 1.0 (0.6-1.8) | .95 |
| Preoperative risk factor[c] | 1.3 (0.9-1.9) | .18 | 1.0 (0.7-1.6) | .91 |
| Single-ventricle physiology | 4.0 (2.5-6.2) | <.001 | 4.3 (2.3-8.2) | <.001 |
| Concomitant procedure | 1.3 (0.8-2.1) | .29 | 0.8 (0.5-1.4) | .42 |
| Valve repair category[d] | | | | |
| Neoaortic valve repair | 2.0 (1.2-3.5) | .010 | 0.8 (0.4-1.6) | .53 |
| Truncal valve repair | 3.1 (1.9-5.0) | <.001 | 2.4 (1.4-4.1) | .002 |

[a]Class 1 as reference; [b]Age 1-17 years as reference; [c]Presence of at least 1 major preoperative risk factor, including cardiopulmonary resuscitation, shock, extracorporeal membrane oxygenation, malignant ventricular arrhythmia or high-grade atrioventricular block, mechanical ventilation, renal failure, liver failure, sepsis, necrotizing enterocolitis, stroke, seizure, or intracerebral hemorrhage, and prior noncardiac surgery; [d]Native aortic valve repair as reference. Model C-index: Technical performance score only, 0.600; covariates only, 0.705; TPS and covariates, 0.744. SHR, subdistribution hazard ratio; TPS, technical performance score.

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# Conclusions

- AV repair is a viable option
- AV repair performs better when foreign material is avoided, although foreign material may provide better immediate results
- Foreign material is unavoidable under certain circumstances
- Poor quality of leaflet tissue is a risk factor for poor outcome
- A suboptimal result should be fixed immediately