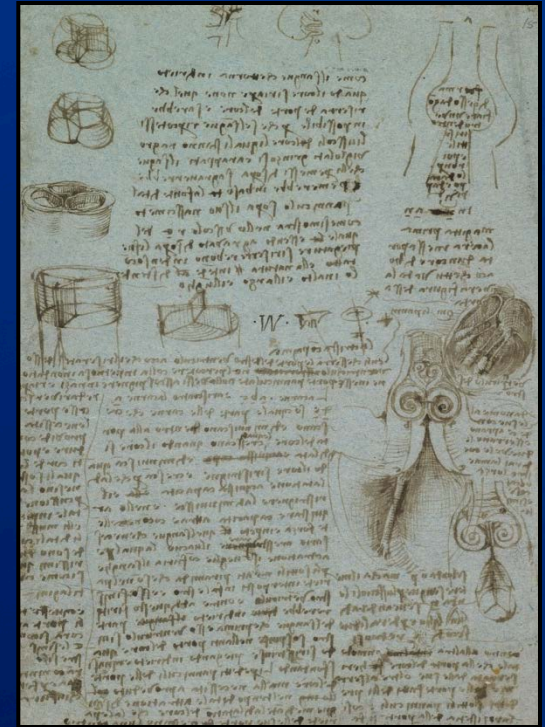


Aortic Valve Anatomy

Abnormalities and Natural History



Charles J. Bruce, MBChB, FCP(SA), FACC, FASE

Professor of Medicine

Director Structural Heart Disease Program

Mayo Clinic and Mayo Foundation, Rochester, MN

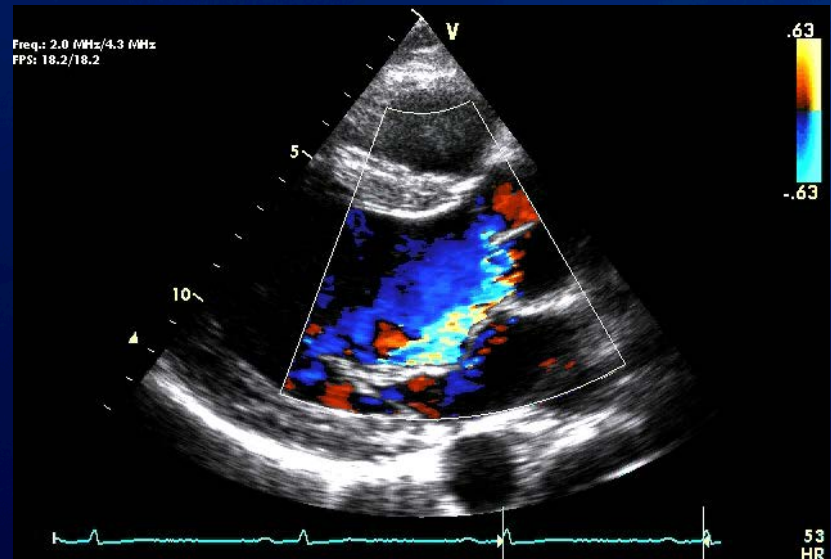
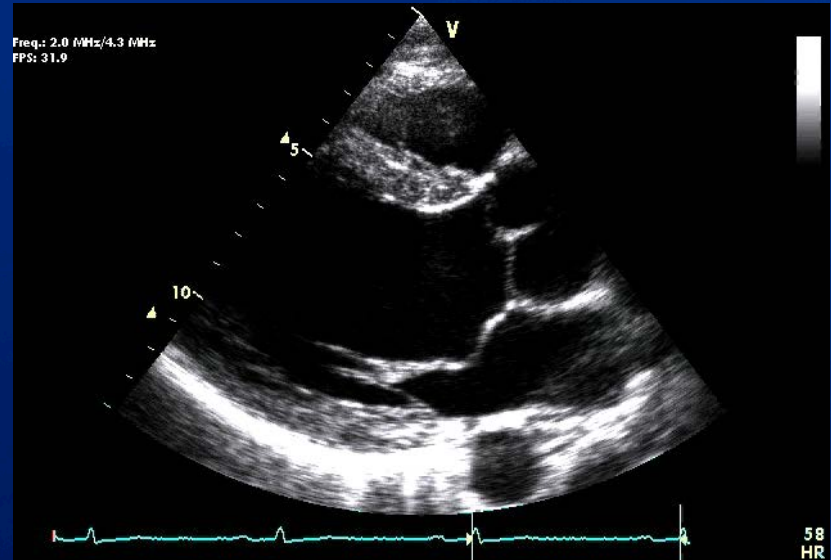
bruce.charles@mayo.edu

44 yo MD with Severe Aortic Regurgitation

- Incidental murmur few years ago
- Asymptomatic despite active lifestyle
- Paroxysmal AF
- Echo documents progressive LV enlargement
- Collapsing pulse
- $\frac{3}{4}$ diastolic decrescendo murmur LSB

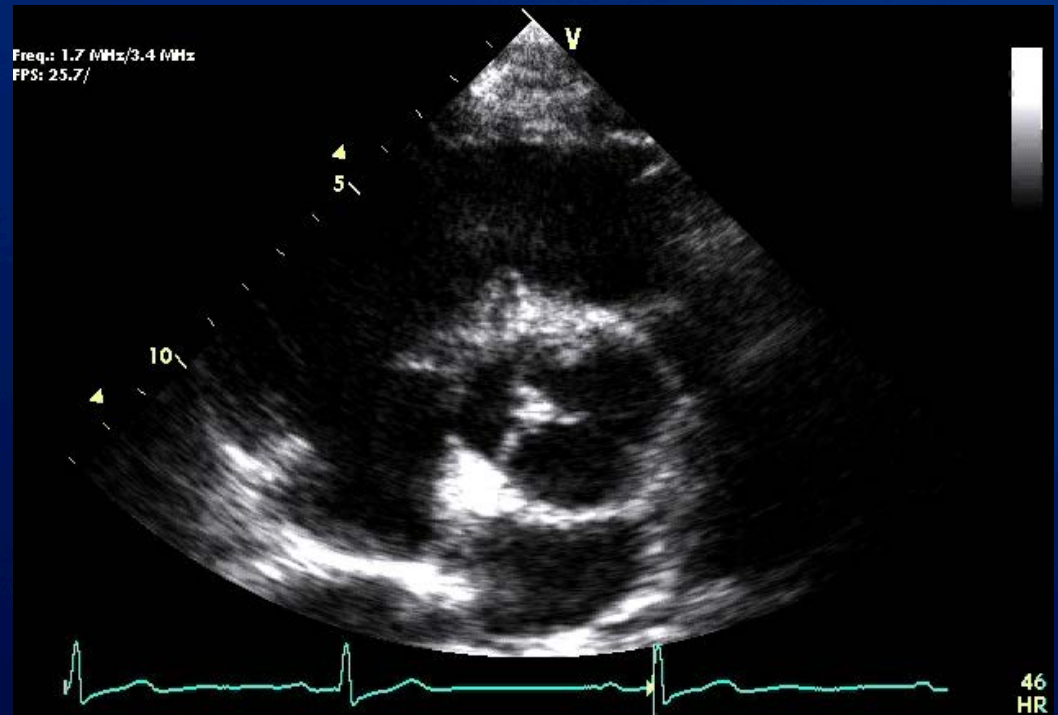
Echocardiogram

- Severe AR
- BAV without stenosis or calcification
- LVEDD 66mm
- EF 66%
- Mid ascending aorta 43mm



Echocardiogram

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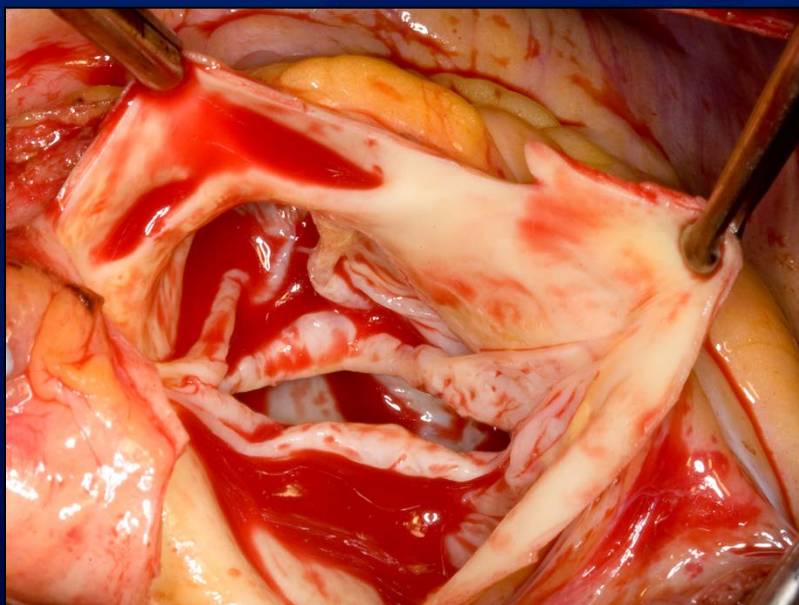
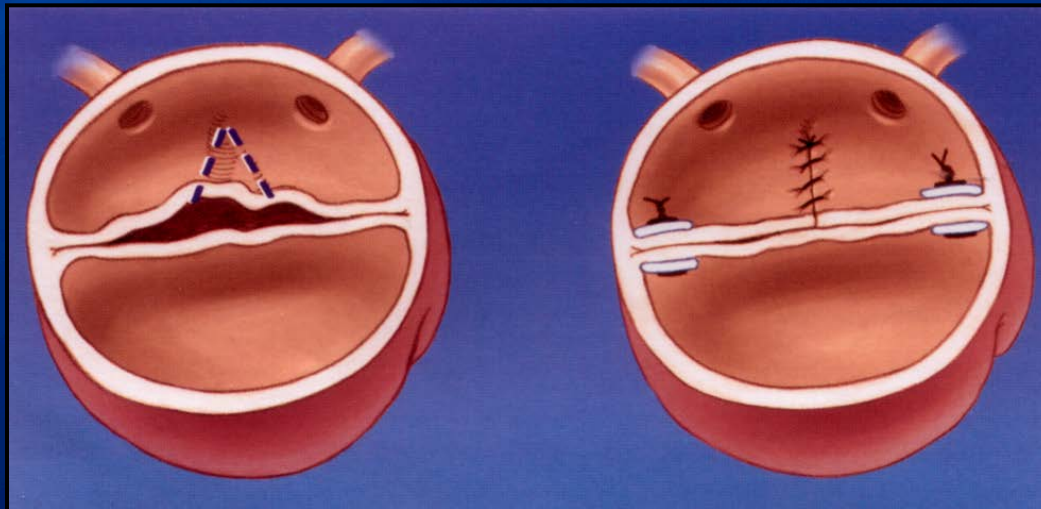


Echocardiogram

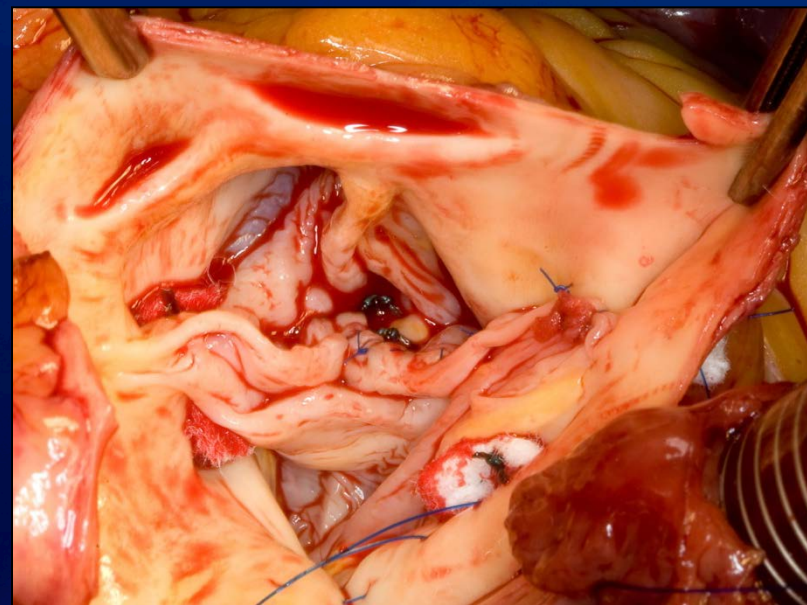
- Severe AR
- BAV without stenosis or calcification
- LVEDD 66mm
- EF 66%
- Mid ascending aorta 43mm

What would you advise?

1. No surgery
2. AV repair
3. AV replacement
4. AV repair and aortic repair
5. AV replacement and aortic repair

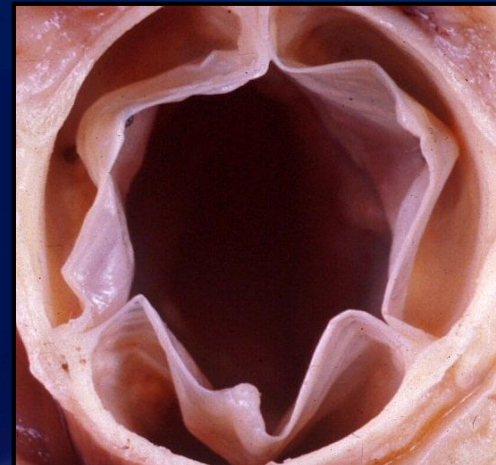
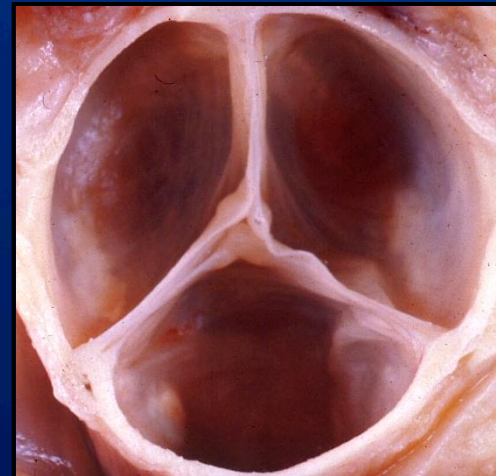
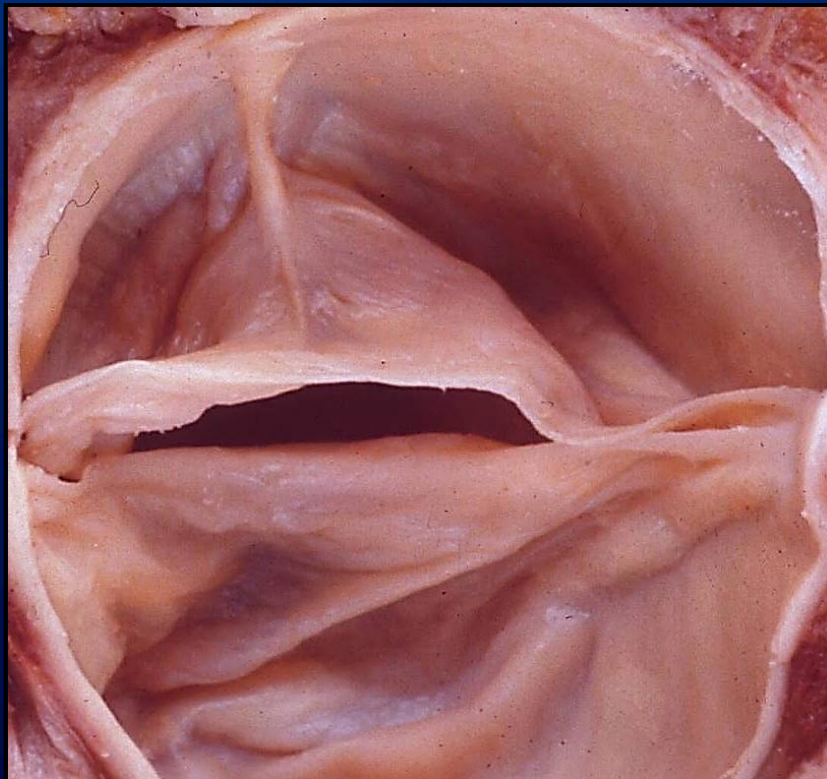


Before



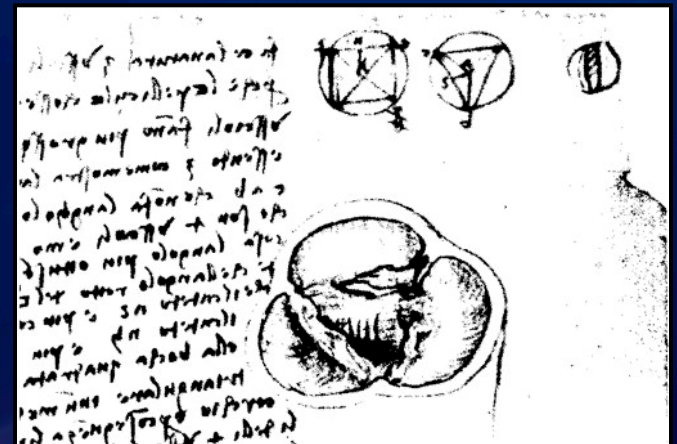
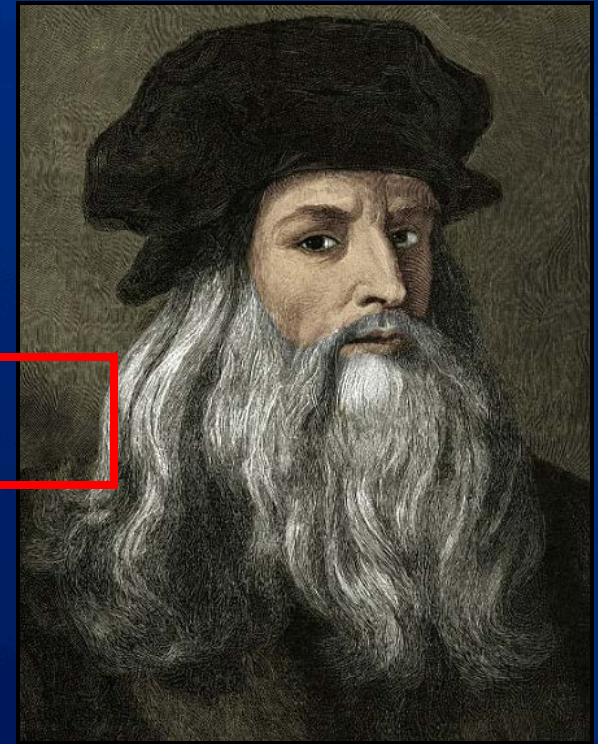
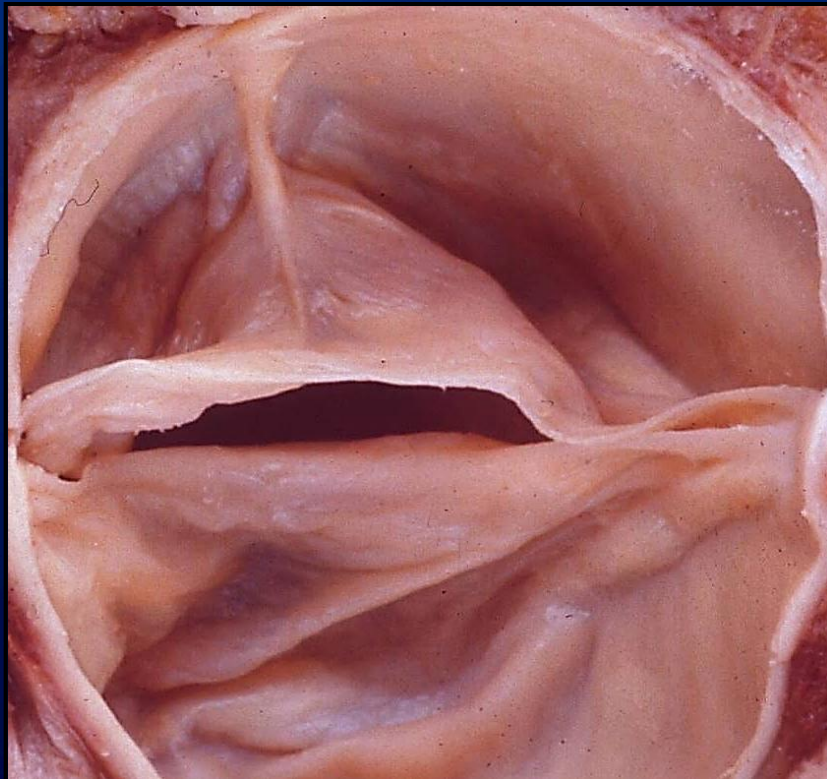
After

Bicuspid Aortic Valve Disease



Courtesy William D Edwards, MD

Bicuspid Aortic Valve Disease

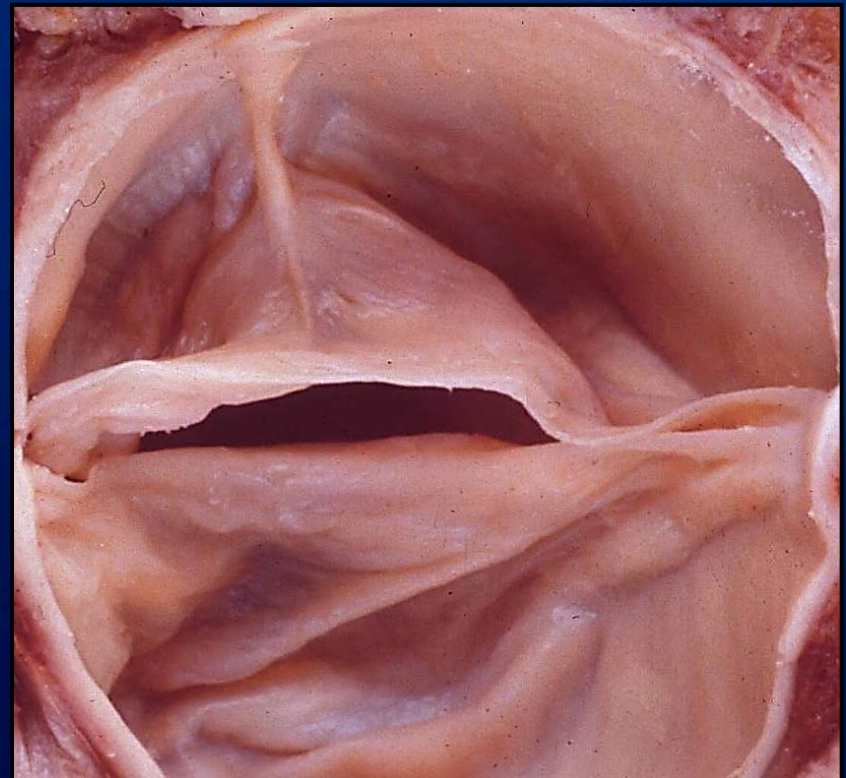


Bicuspid Aortic Valve Disease

- Most common congenital heart defect
 - Prevalence 0.5% to 2%
- Male predominance ~ 3:1
- AD with reduced penetrance; NOTCH1
- BAV in 9% of 1st degree relatives

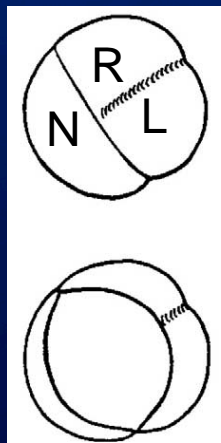
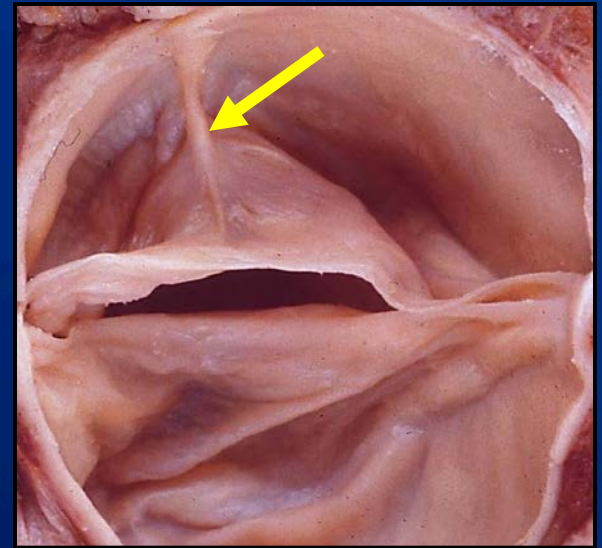
Non-valvular findings 50% adults

- Aortic dilatation most common
- Coarctation (50 - 75% BAV)
- Hypoplastic left heart syndrome



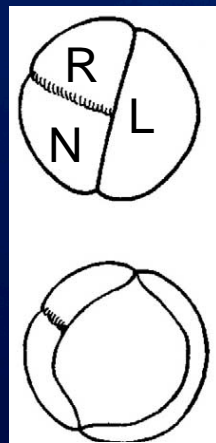
Pathology

- 2 unequal-sized leaflets
- Larger leaflet has central raphe
- Morphologic patterns



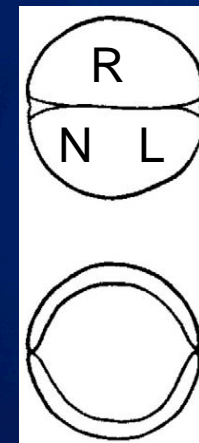
Fusion
left and
right

> 60%



Fusion
right and
non

15-25%



Fusion
left and
non

< 5%

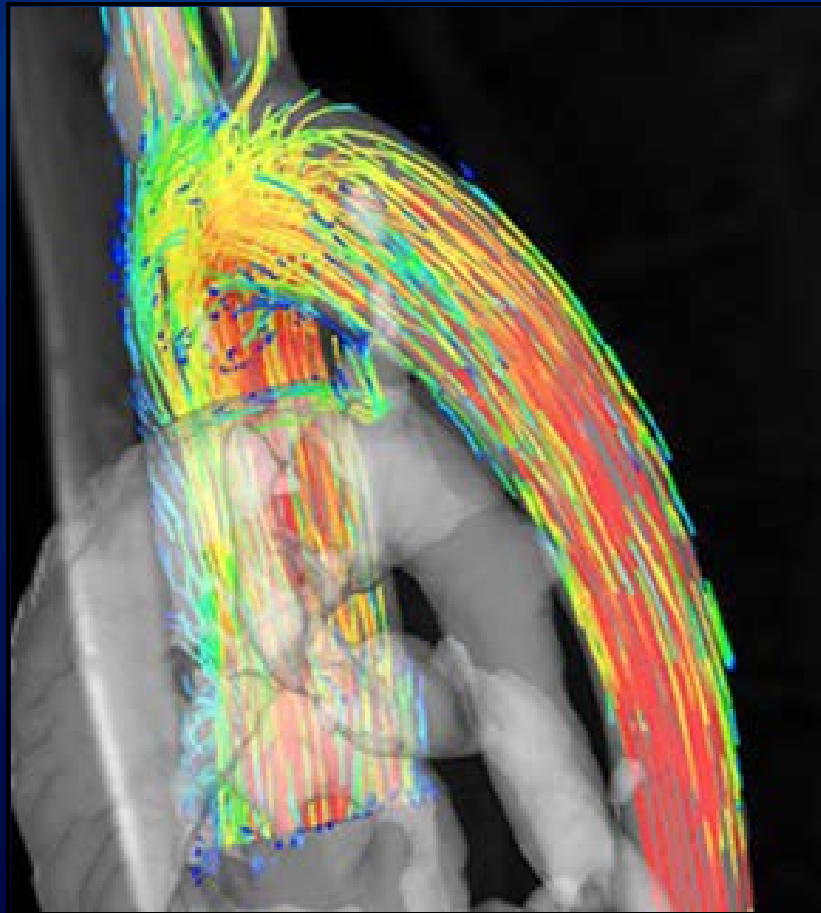
Coarctation
AS progress more rapid

AS in childhood
More AR

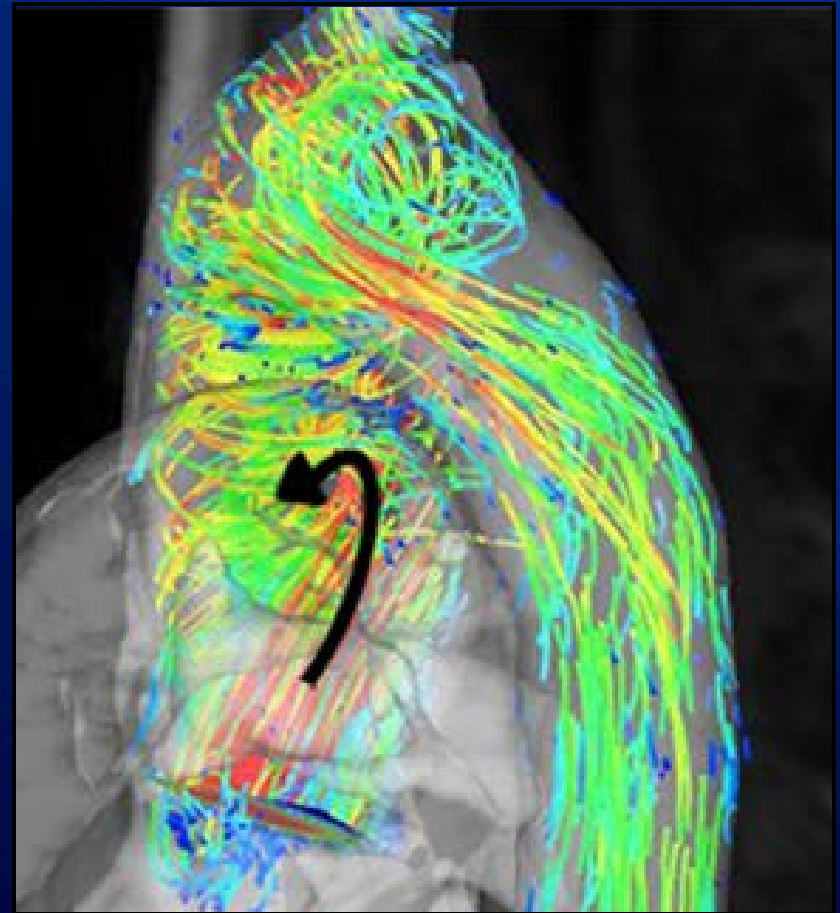
No assoc with AS

Cusp Fusion Pattern and Aortic Outcome

Right-Non Associated with Larger Aortic Dimensions

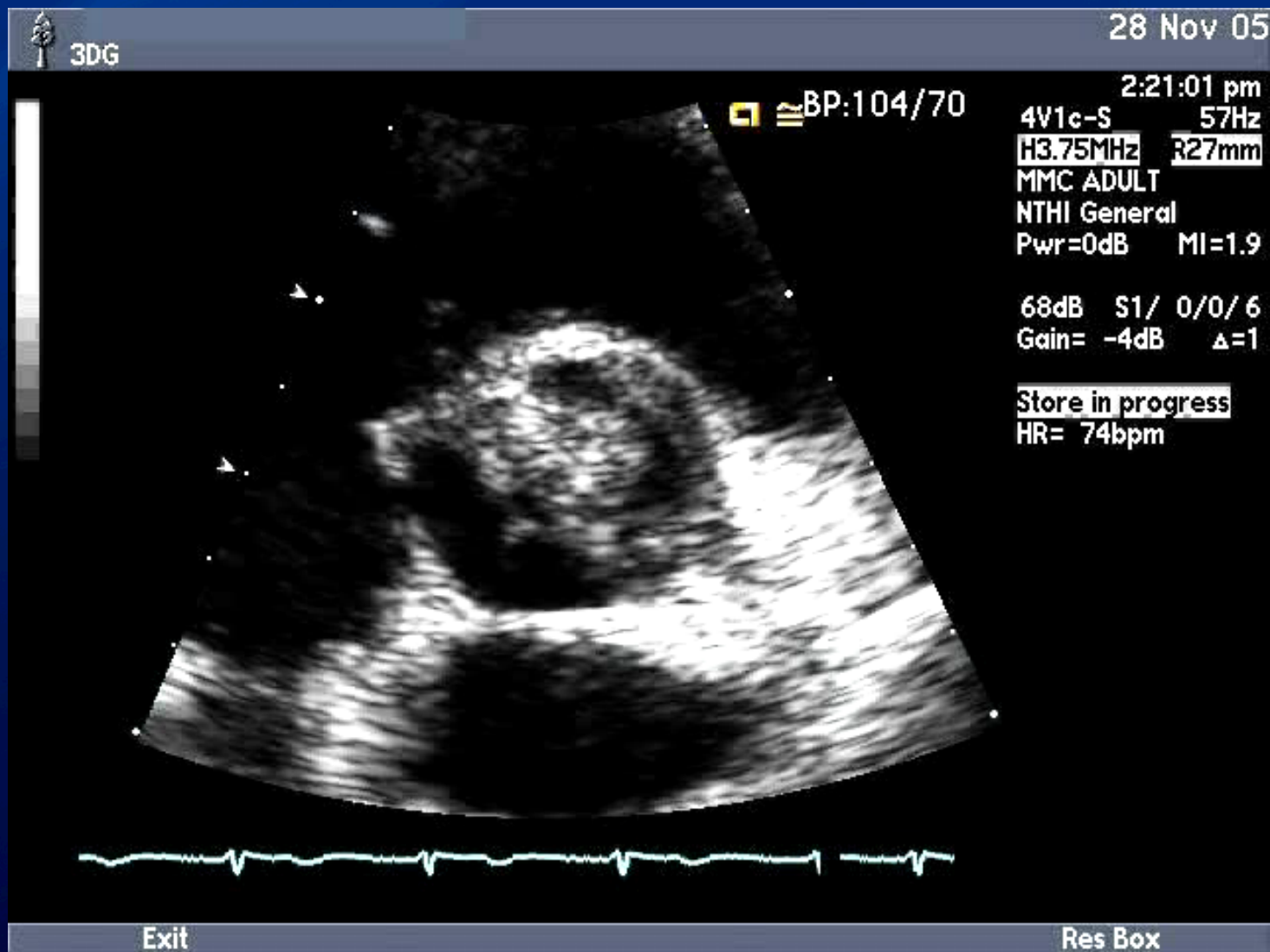


Normal flow

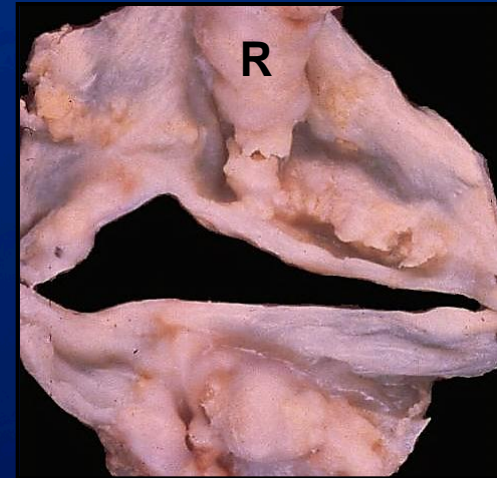
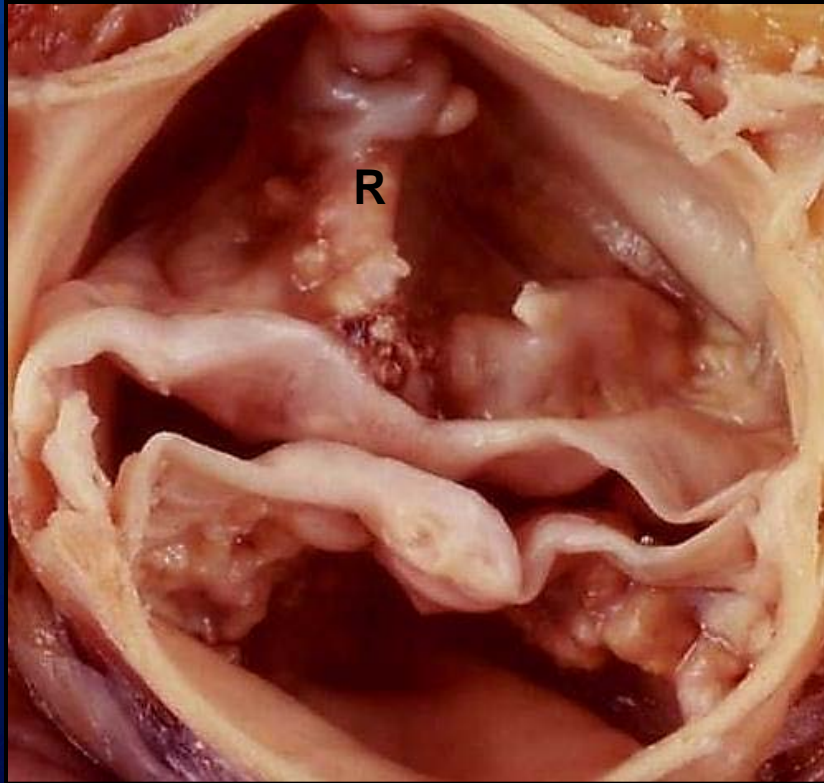


Right-handed helical flow

Bicuspid AV



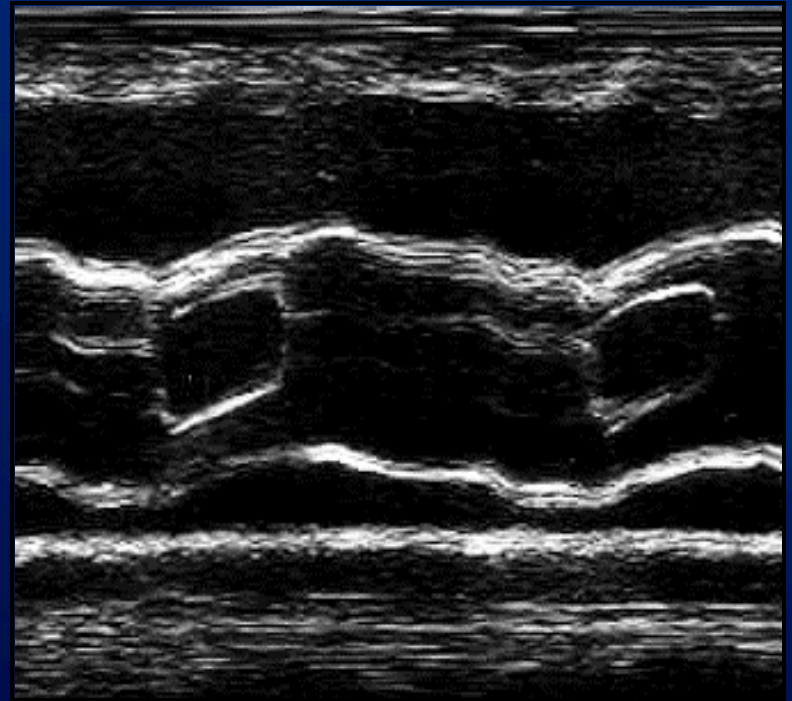
Bicuspid Aortic Valve Stenosis



Raphe: congenital line of fusion
Calcification: along raphe first

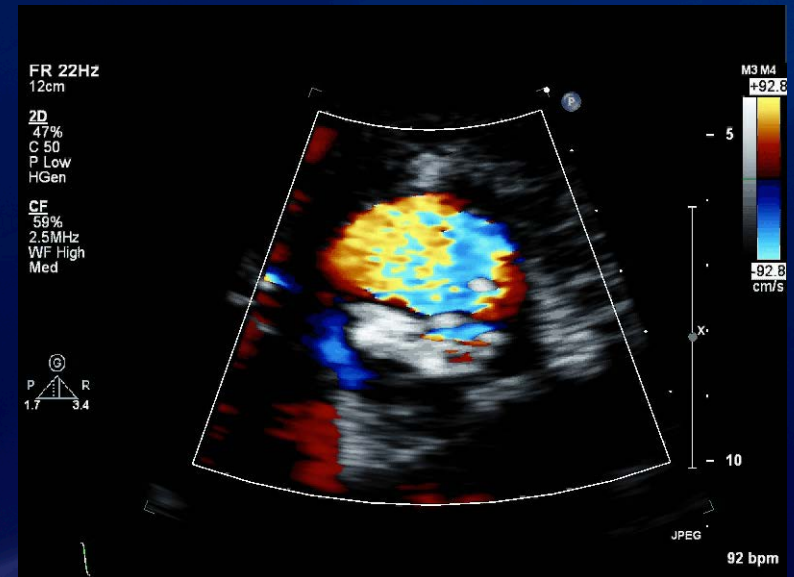
Pathophysiology: Aortic Stenosis

- Restricted cusp motion + eccentric coaptation
- Elliptical orifice area + flow turbulence
- Early valve degeneration and Ca^{++}
- AS up to decade earlier than tricuspid AV's



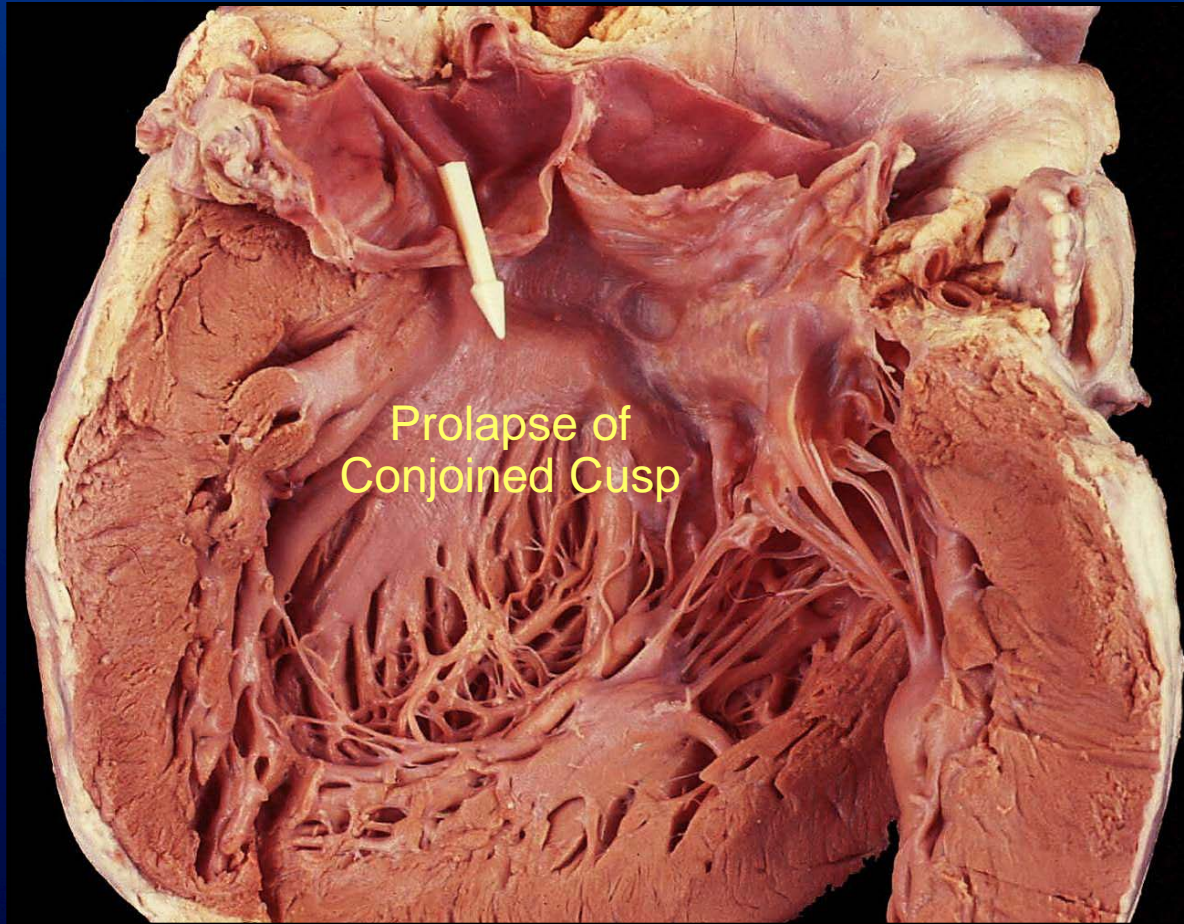
Pathophysiology: Aortic Regurgitation

- AR often mild to moderate concurrent with AS
- Mechanism of AR
 - 15 to 20% incomplete closure
 - Cusp prolapse
 - Aortic root dilatation
 - Endocarditis



Pathophysiology: AR

Cusp Prolapse



Courtesy William D Edwards, MD

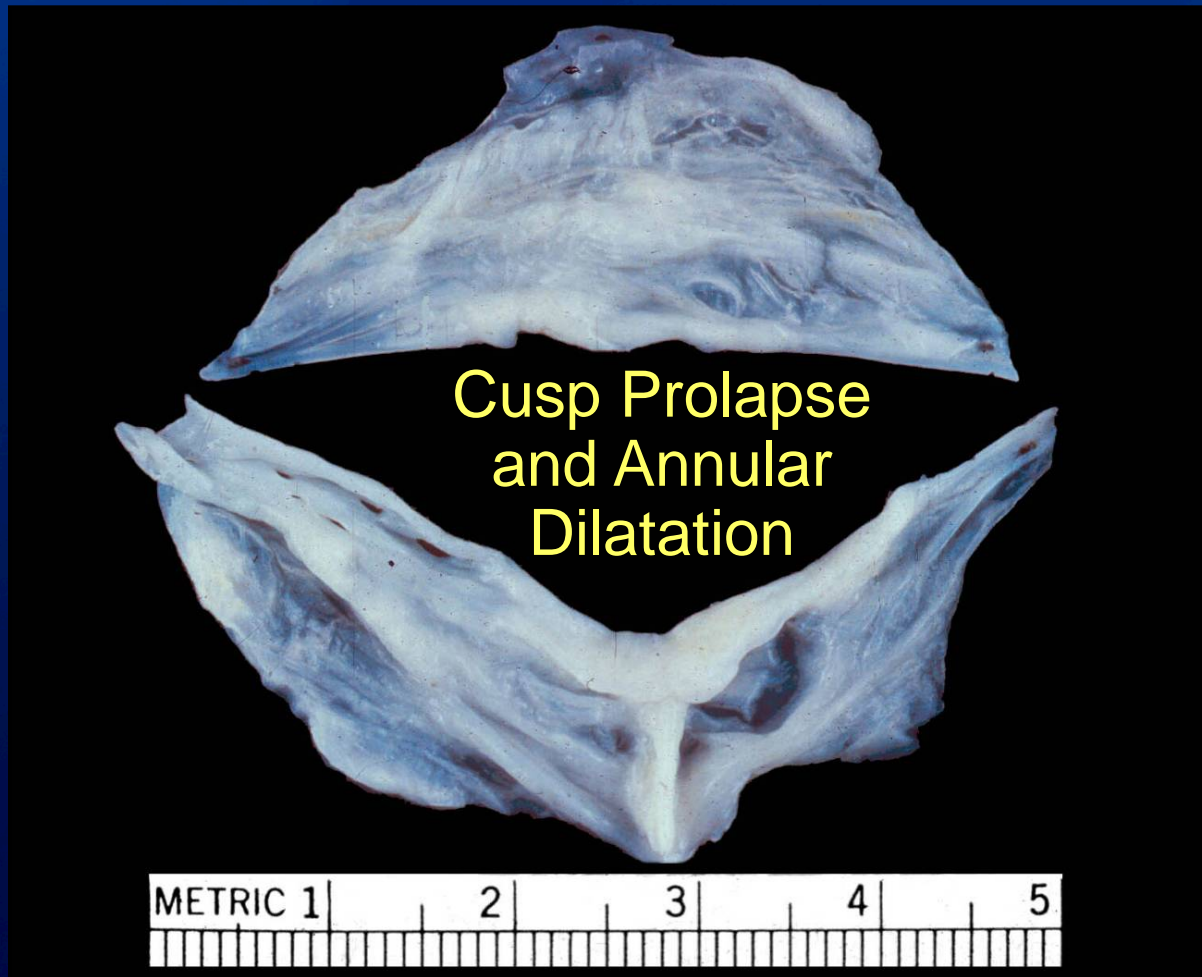
Pathophysiology: AR

Cusp Prolapse



Pathophysiology: AR

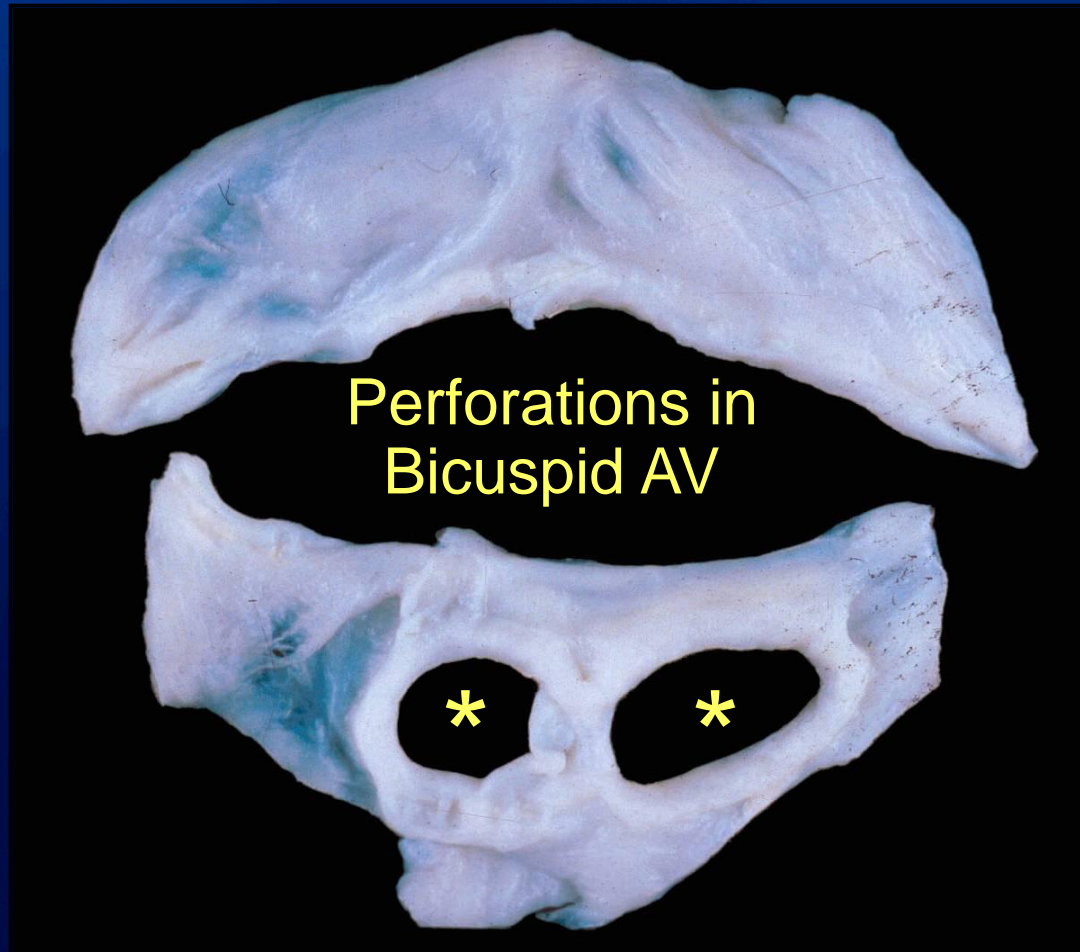
Annular dilatation and cusp prolapse



Courtesy William D Edwards, MD

Pathophysiology: AR

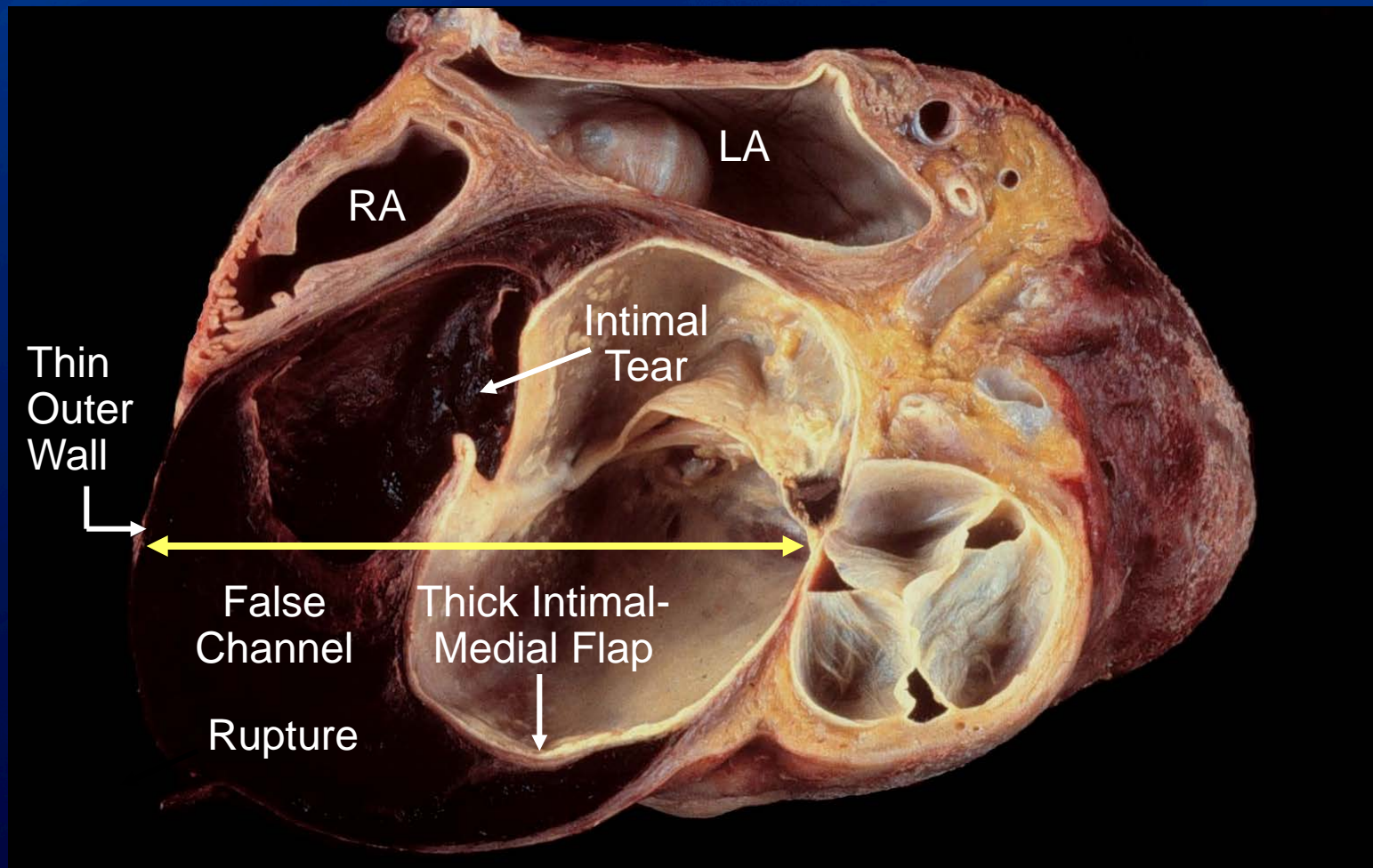
Endocarditis



Courtesy William D Edwards, MD

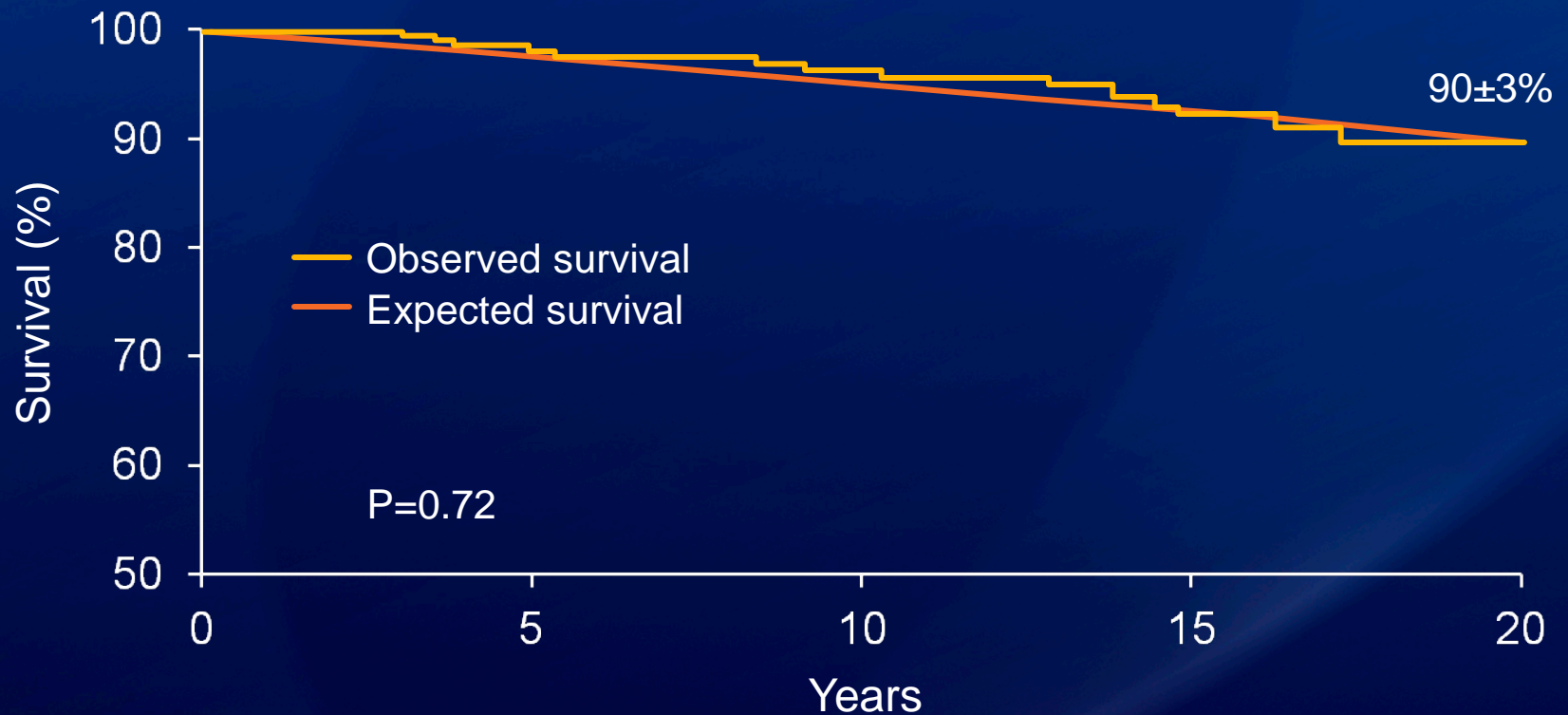
Bicuspid Aortic Valve

Association with Aortic Dissection

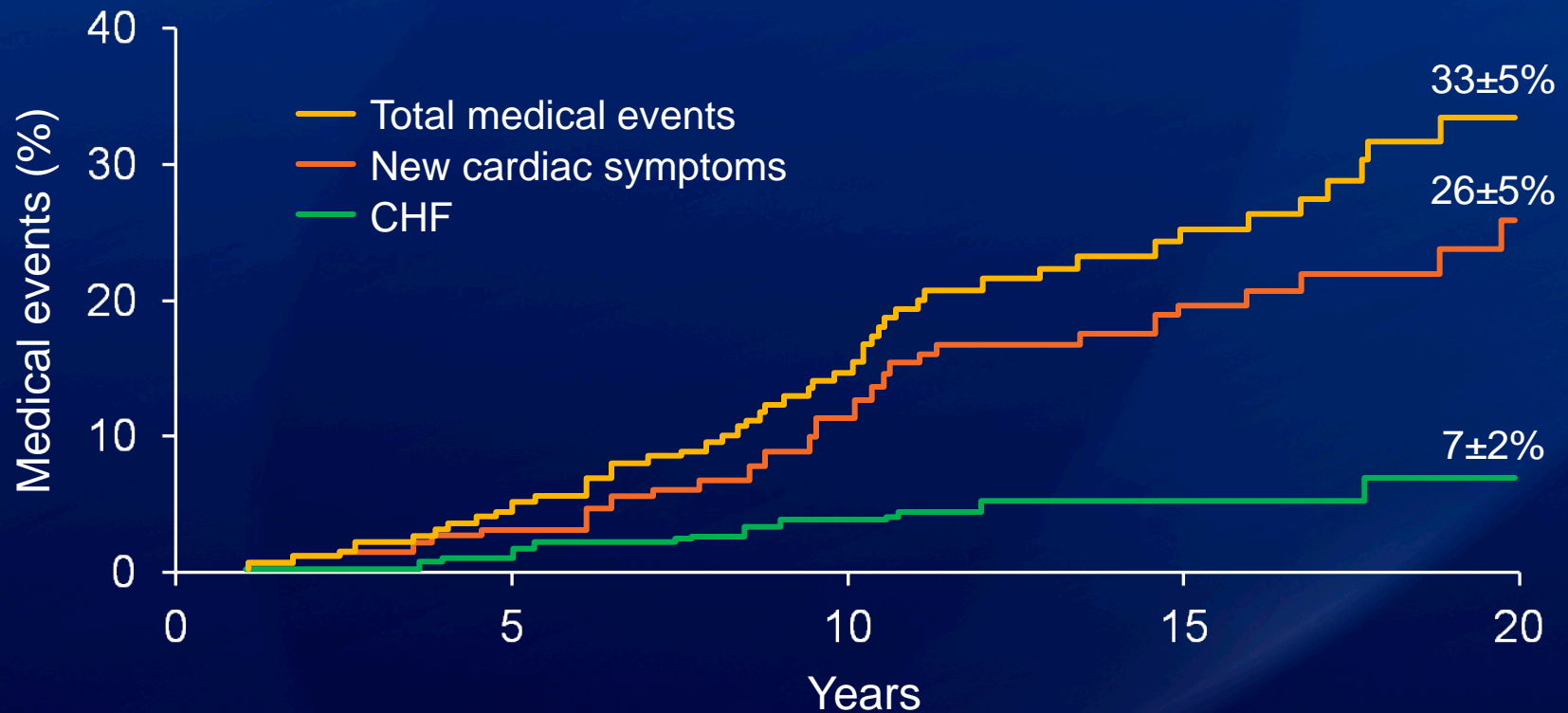


Courtesy William D Edwards, MD

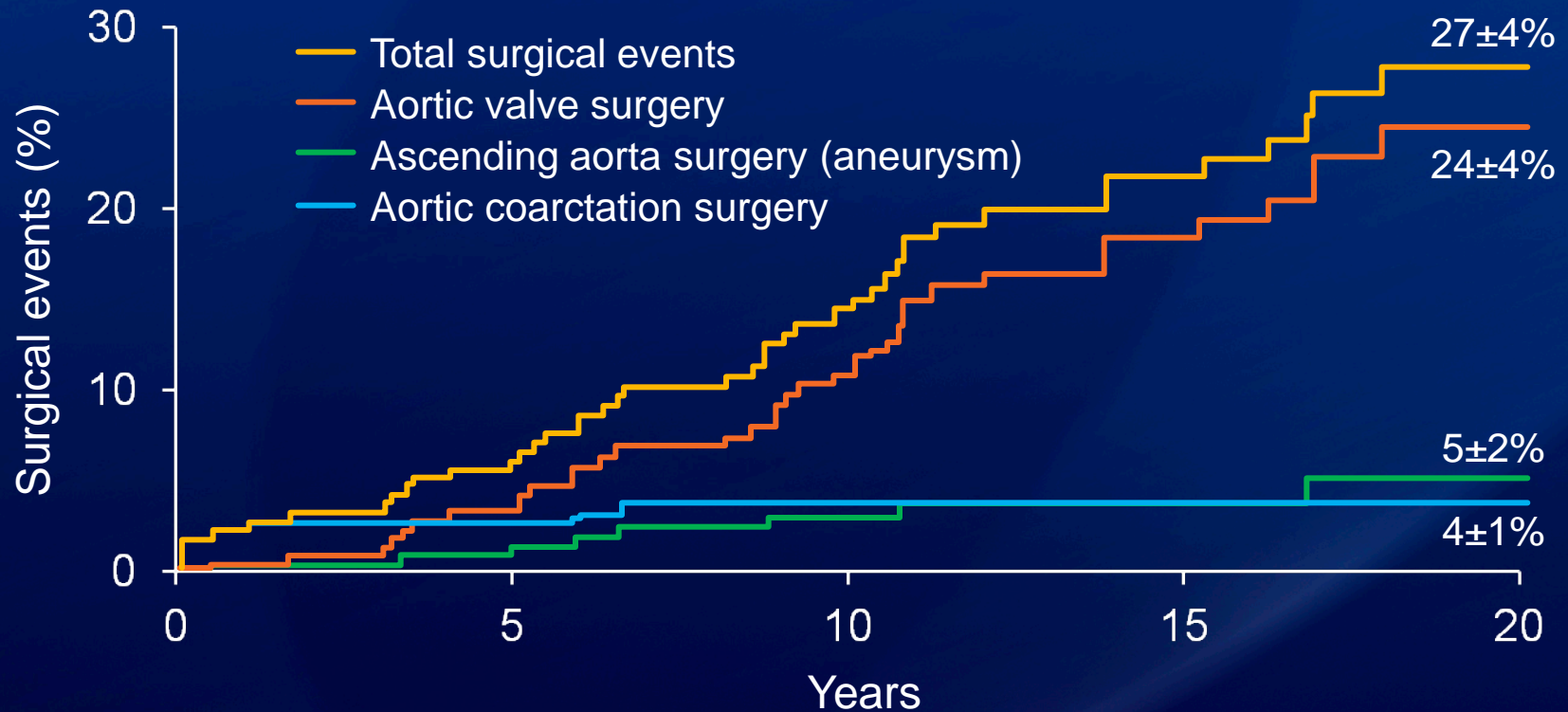
Natural History Asymptomatic Patients Normal Functioning or Minimally Dysfunctional BAV in Community



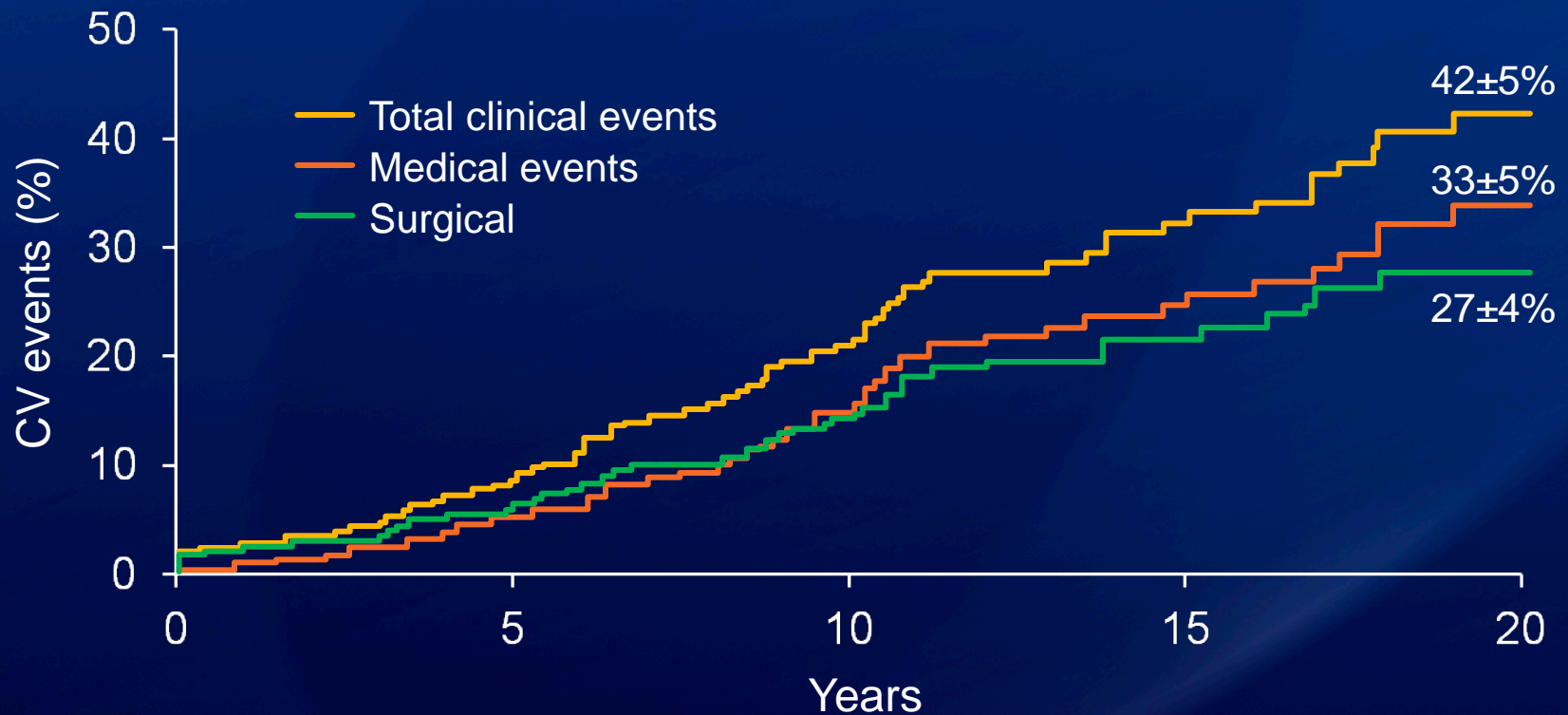
Natural History Asymptomatic Patients Normal Functioning or Minimally Dysfunctional BAV in Community



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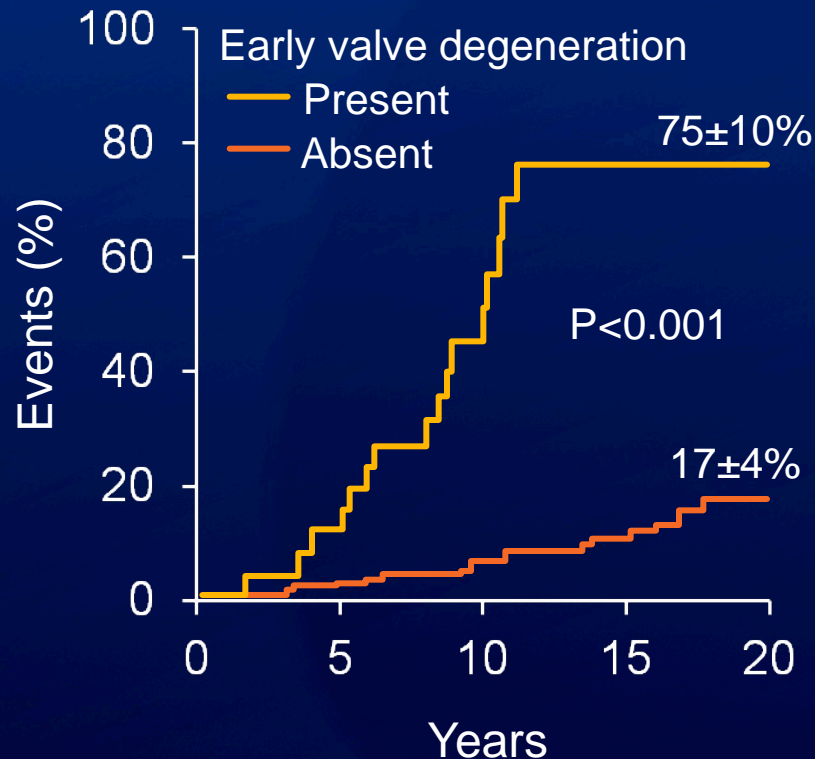


Natural History Asymptomatic Patients Normal Functioning or Minimally Dysfunctional BAV in Community

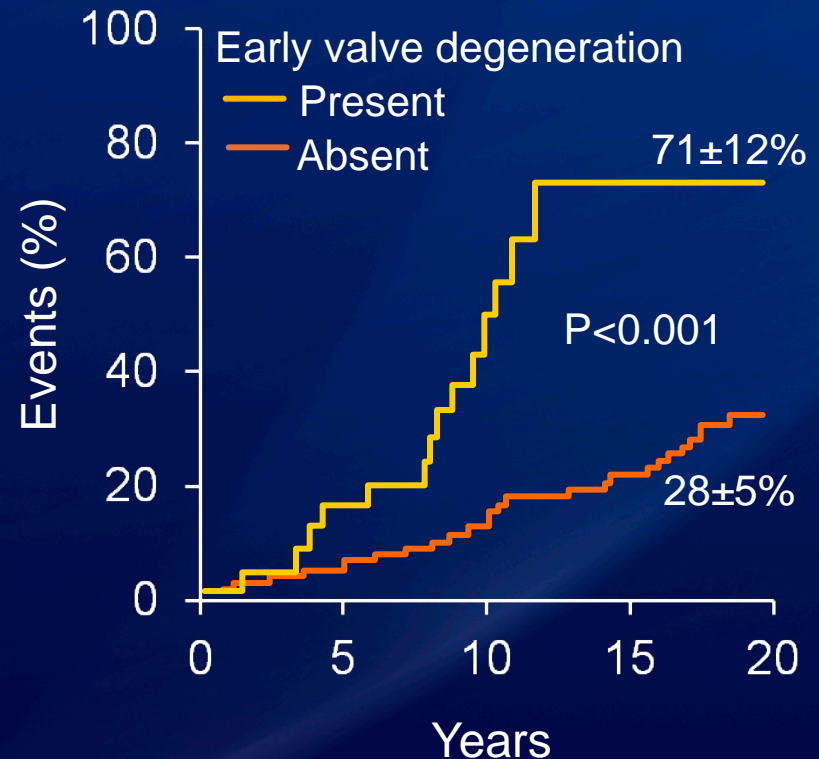


Natural History Asymptomatic Patients Normal Functioning or Minimally Dysfunctional BAV in Community

Aortic Valve Surgery

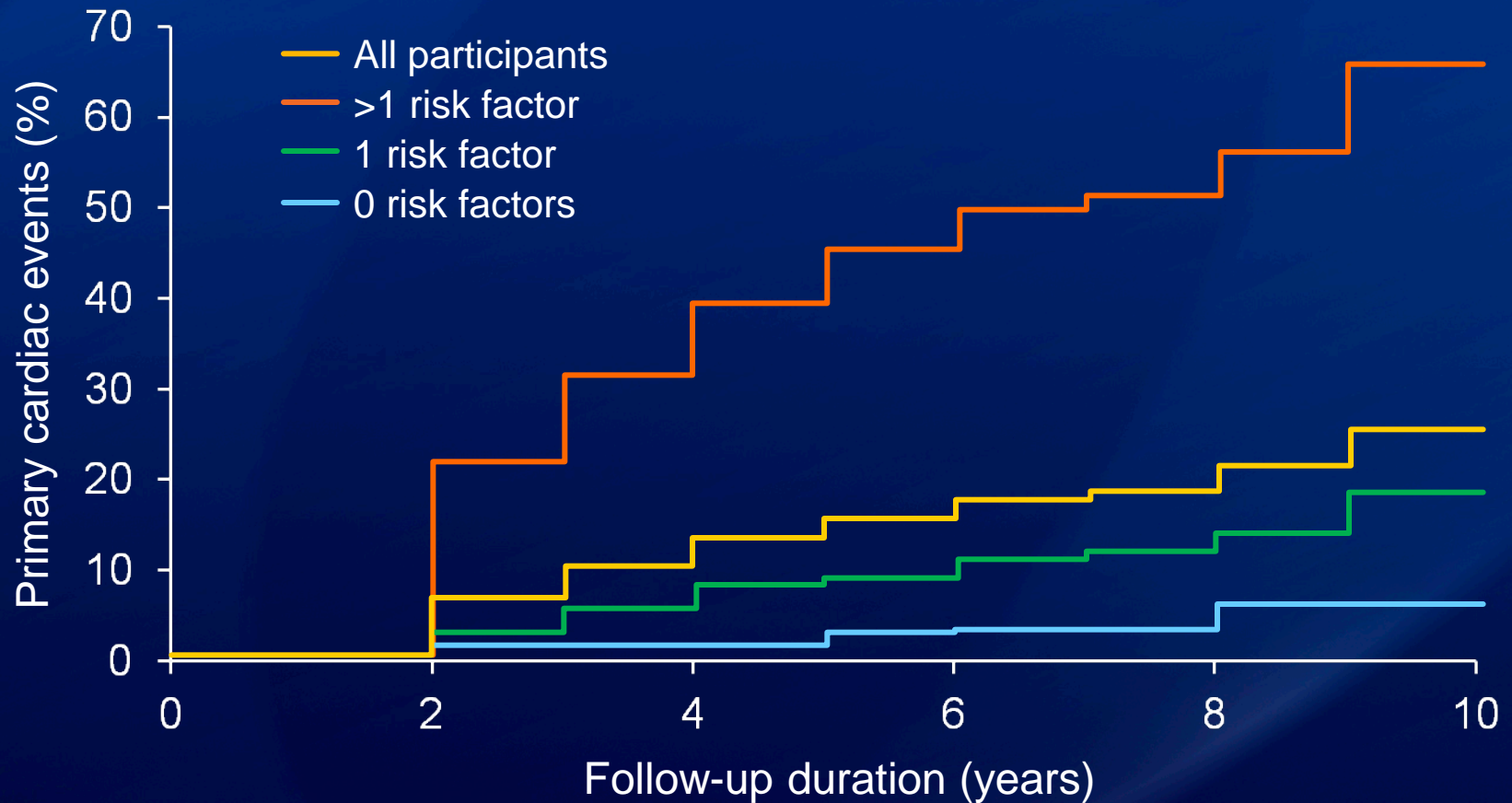


Medical Events



Michelen et al: Circulation, 2008

Cardiac Events in Adults With Bicuspid Aortic Valve



Tzemos et al: JAMA, 2008

Medical Therapy

- Aggressively treat HT
- Beta blockers to slow aortic dilatation (AHA/ACC IIa)
- ARB's: no evidence
- Vasodilators: only for HT
- Statins to slow progression of degenerative disease not endorsed by ACC/AHA

Indications for Intervention

ESC and ACC/AHA Guidelines

- Indications similar for AS or AR in tricuspid AV's
- Decision making involves assessment of BOTH valve function and aortic dimension
- ~30% adults will need aortic root surgery

Indications for Intervention

ESC and ACC/AHA Guidelines

Elective aortic repair

- Concomitant indication for elective AV repair or replacement AND Ao diameter > 45mm
- Asymptomatic with well functioning BAV AND
 - Ao diameter \geq 50mm
 - Diameter > 5mm/year
 - Family history AD, rupture or SD
 - Planned pregnancy when Ao > 45mm

Bicuspid Aortic Valve Disease

Take Home Points

- Its inherited
- Cusp morphology may be important
- Asymptomatic minimally dysfunctional valves confer no increased risk of mortality
- Morbidity is common
- Valve degeneration and age are important predictors of outcome
- Valve repair in selected cases

Bicuspid Aortic Valve Disease

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- Valve repair in selected cases
- **It's not just the valve!**