

Bicuspid AV with Aortopathy

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Disclosures



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- Grant Funding
 - Boston Scientific
- Board of Directors
 - Anticoagulation Forum

Clinical Case

- 55yo man is referred for new murmur
- Echocardiogram finds bicuspid AV with moderate stenosis
- Ascending aorta 43mm in diameter
- PMH: Hypertension, obesity (BMI 32)
- FM: Mother with aortic valve replacement (age 62), maternal uncle with abdominal aortic aneurysm
- SH: Former smoker (quit 10 years ago)
- Meds: Lisinopril

Key questions to consider:

- 1) Is there an underlying cause for his BAV and aortic aneurysm?
- 2) What imaging regimen is recommended?
- 3) When might surgical intervention be advised?

Prevalence of Bicuspid AV

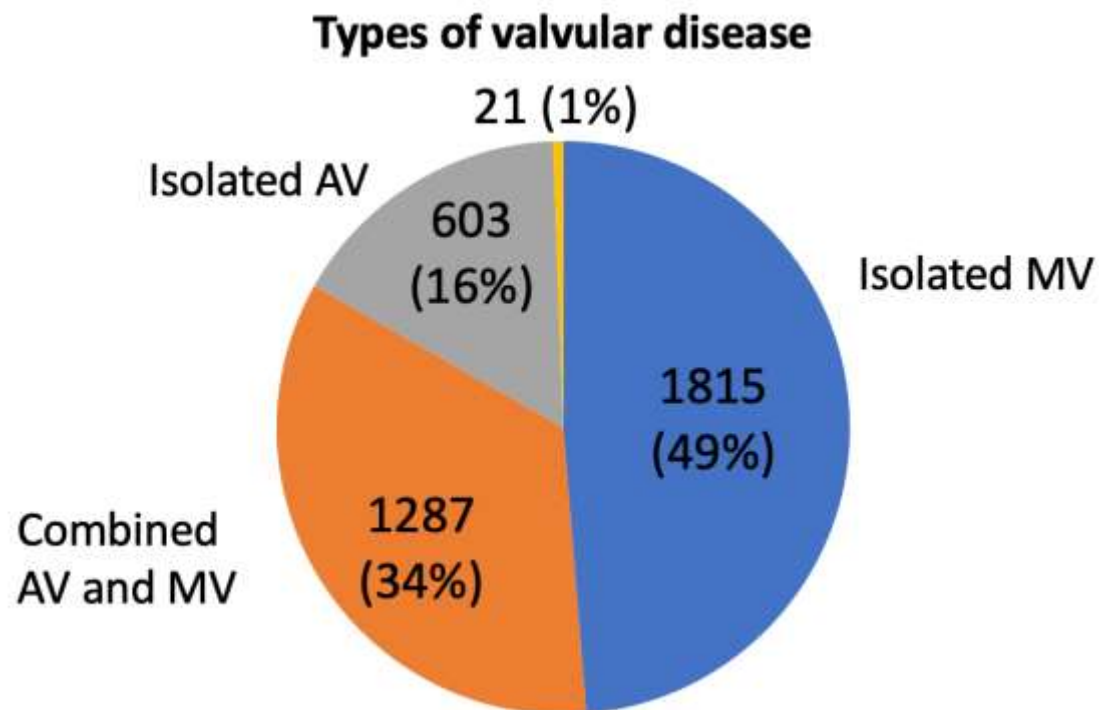
- 25,556 newborns in Copenhagen (2016-2018)
 - 0.8% with BAV
 - 2:1 male:female
- 1692 young men enrolling in Italy's military (1994-1996) undergoing echocardiogram
 - 0.8% with BAV

~1% of of people
have Bicuspid AV

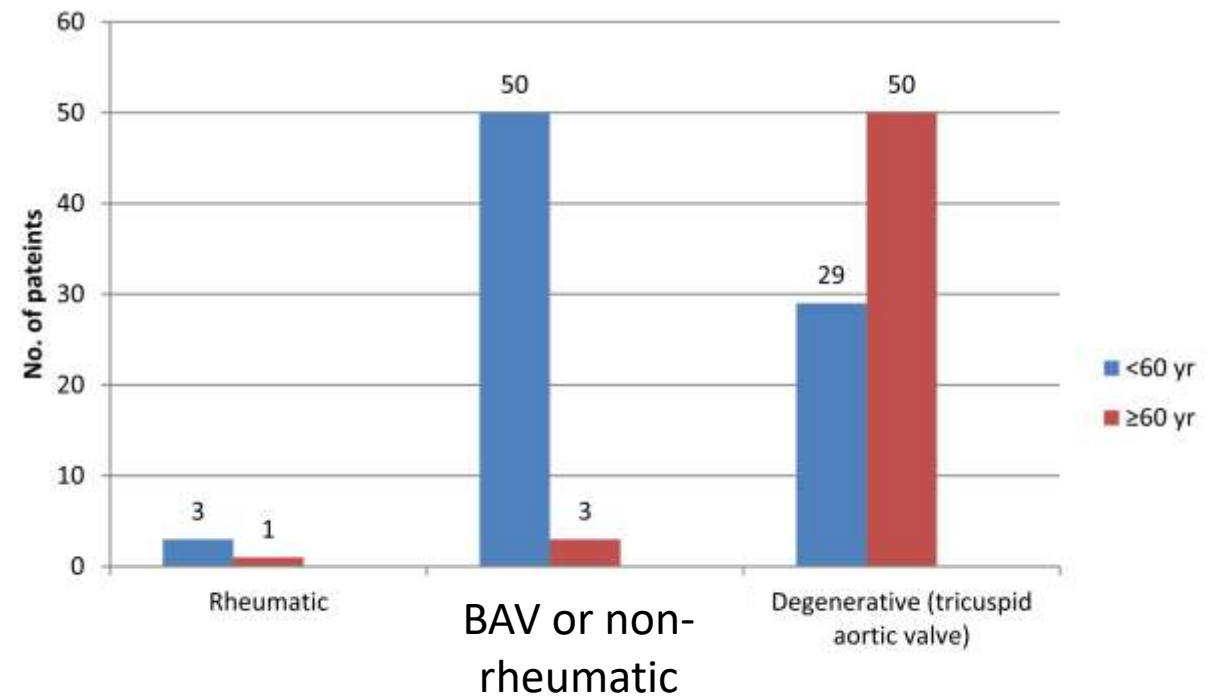
Men > Women

Bicuspid AV in India

- 3728 new cases of valve disease
 - 2014-2016
 - Sanjay Gandhi Postgraduate Institute of Medical Sciences



Etiology of Acquired Isolated Aortic Stenosis



Is Bicuspid AV Genetically-linked?

Univ of Ottawa

- 30 consecutive pts with BAV
- 17/186 (9.1%) relatives with BAV

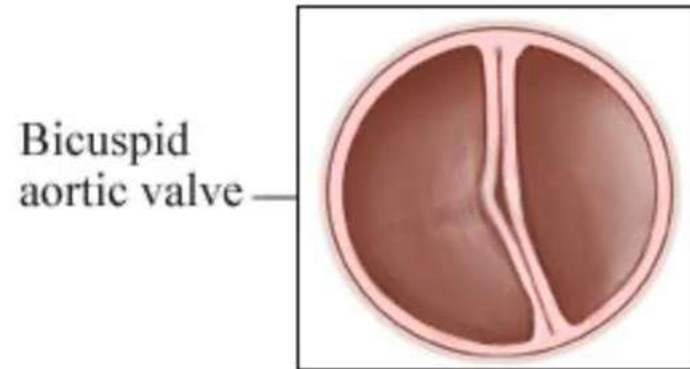
Cincinnati Children's Hospital

- 50 patients with BAV
- 74/309 (24%) relatives with BAV

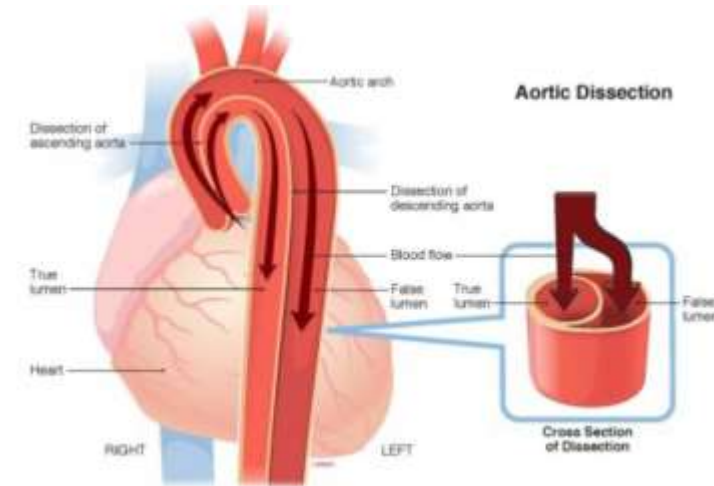
Genetic causes may include:

- NOTCH1
- TGFB2, MAT2A, GATA5, SMAD6, LOX, ROBO4, TBX20
- Turner Syndrome

Bicuspid AV and Aortopathy



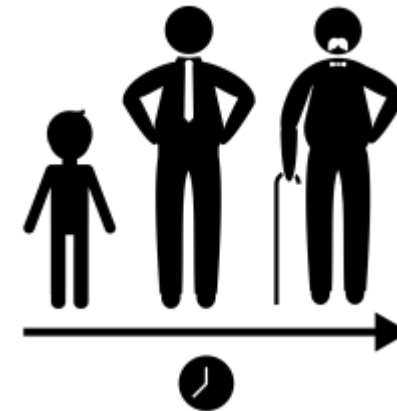
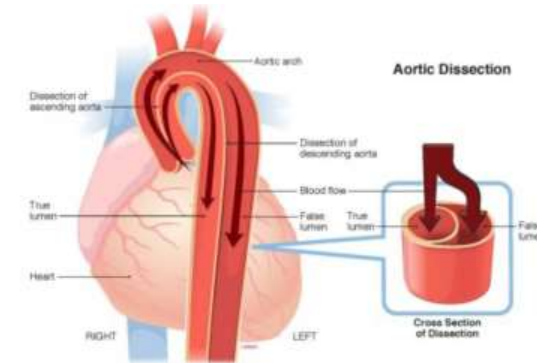
- Common (~1%)
- Men > women
- Many genetic etiologies
- Younger than degenerative valve disease



What is the link between BAV and aortic disease?

Aortic Dissection Risk in BAV

- High risk of aortic dissection
 - 9x higher BAV than TAV
- Dissection at younger age
 - 54 vs. 62 years



2022 ACC Guideline Recommendations



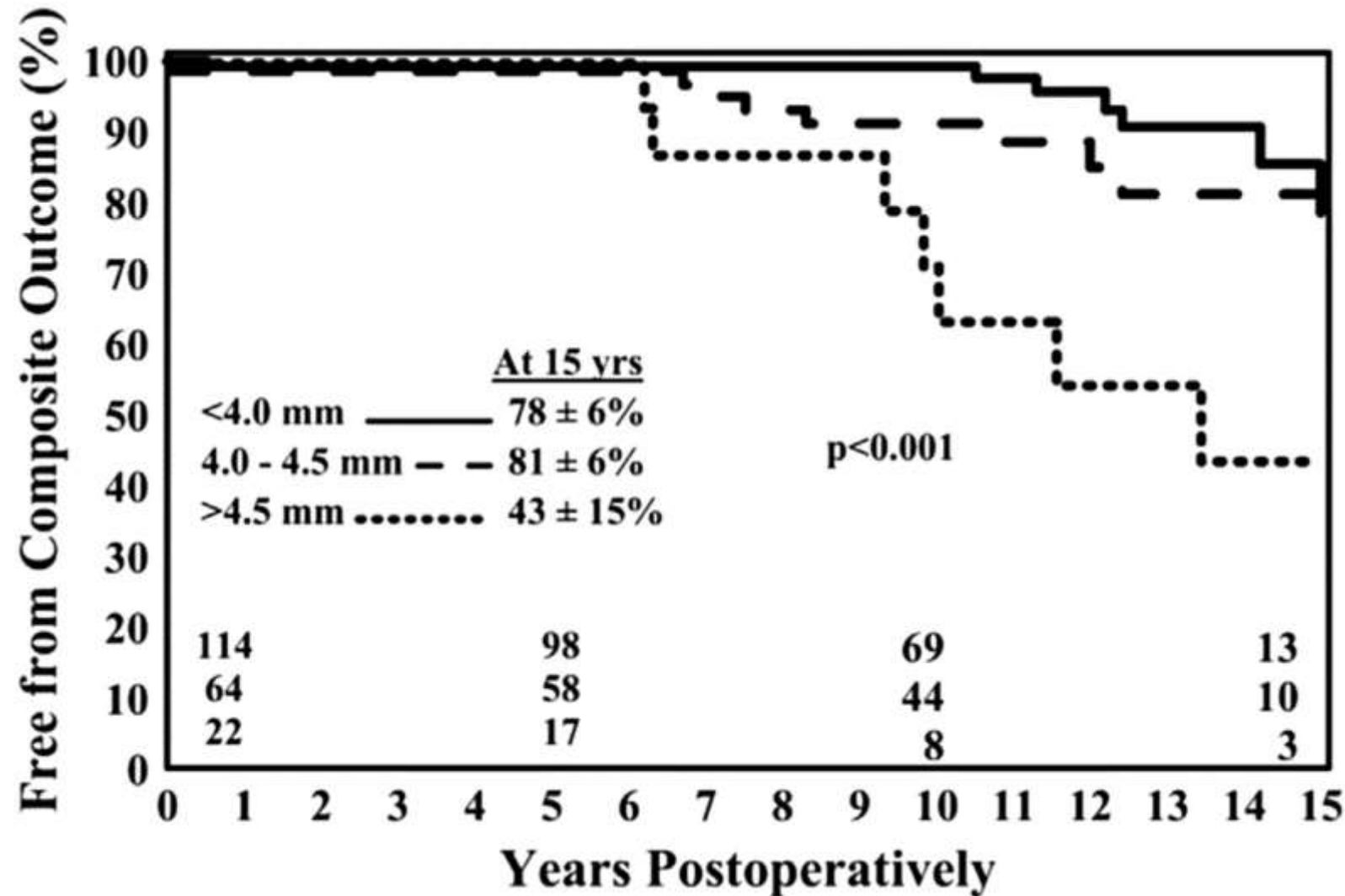
Class 1

- Always take a multi-generational family history
- Recommend genetic screening if BAV and thoracic aortic disease
 - Includes patients with Loeys-Dietz syndrome features
- Screen 1st degree relatives with echocardiogram if patients has BAV and dilated aorta

Class 2a

- Consider screening all 1st degree relatives with echocardiogram if BAV with normal aorta

Aortic Size and Bicuspid AV

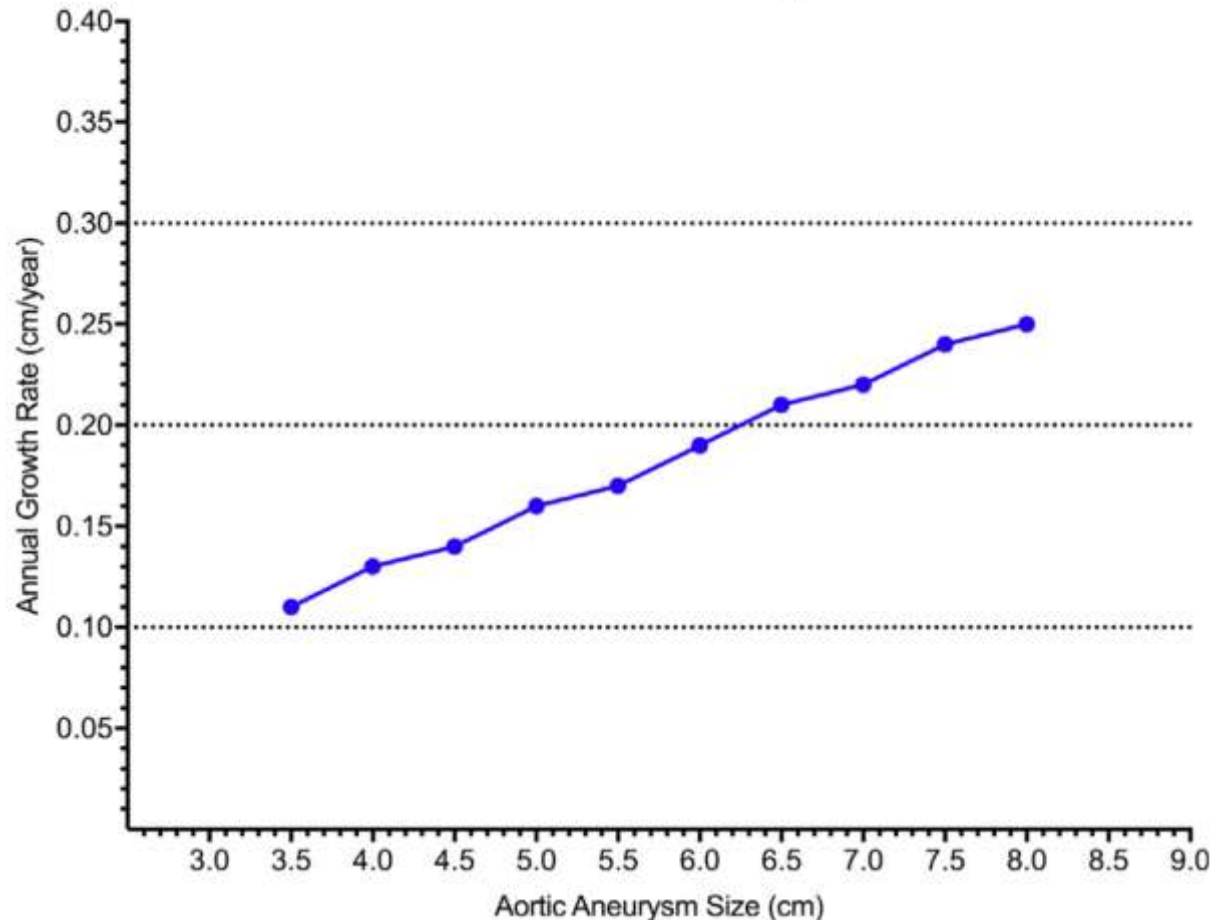


Patients with BAV s/p
valve replacement

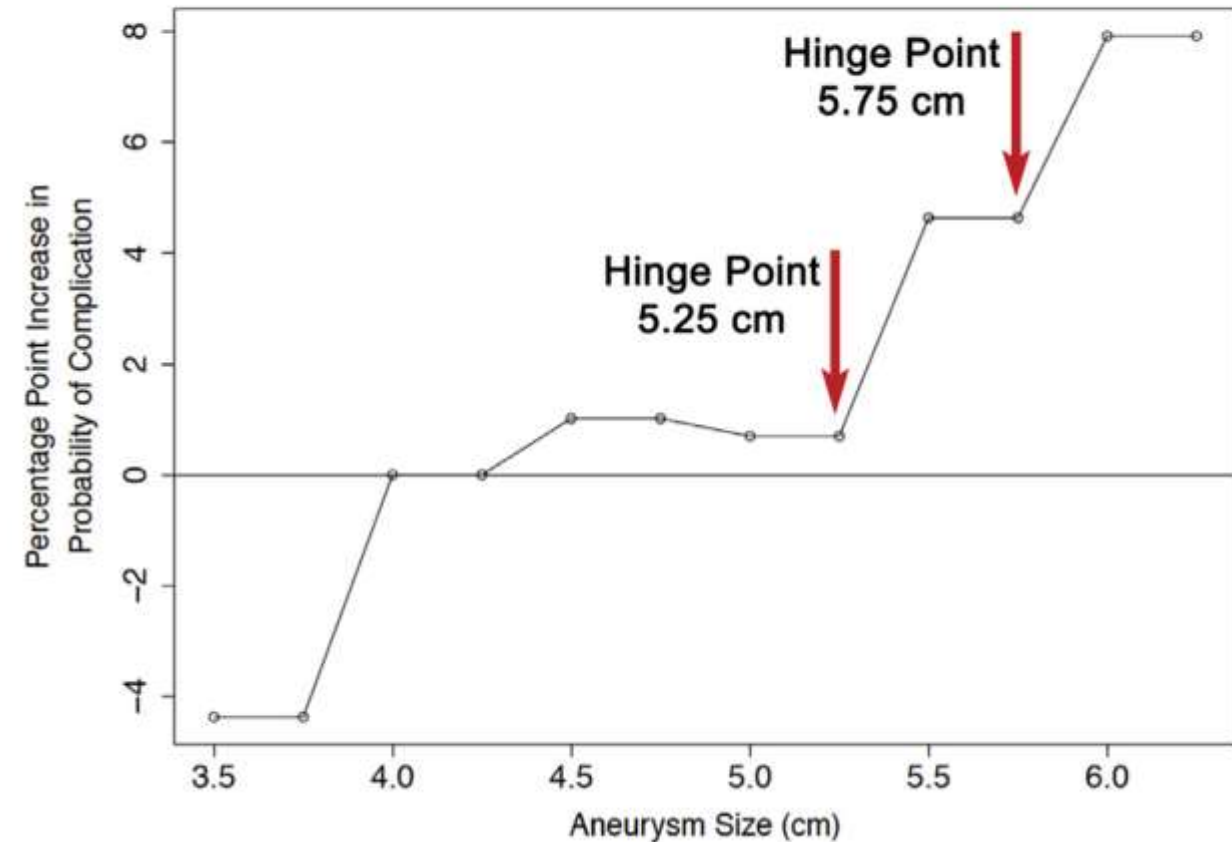
Risk of future aortic
pathology by baseline
size

Ascending Aortic Aneurysm: Growth and Complications

Annual Growth of the Ascending Thoracic Aorta



Estimated Effect of Ascending Aortic Aneurysm Size on Risk of Complication



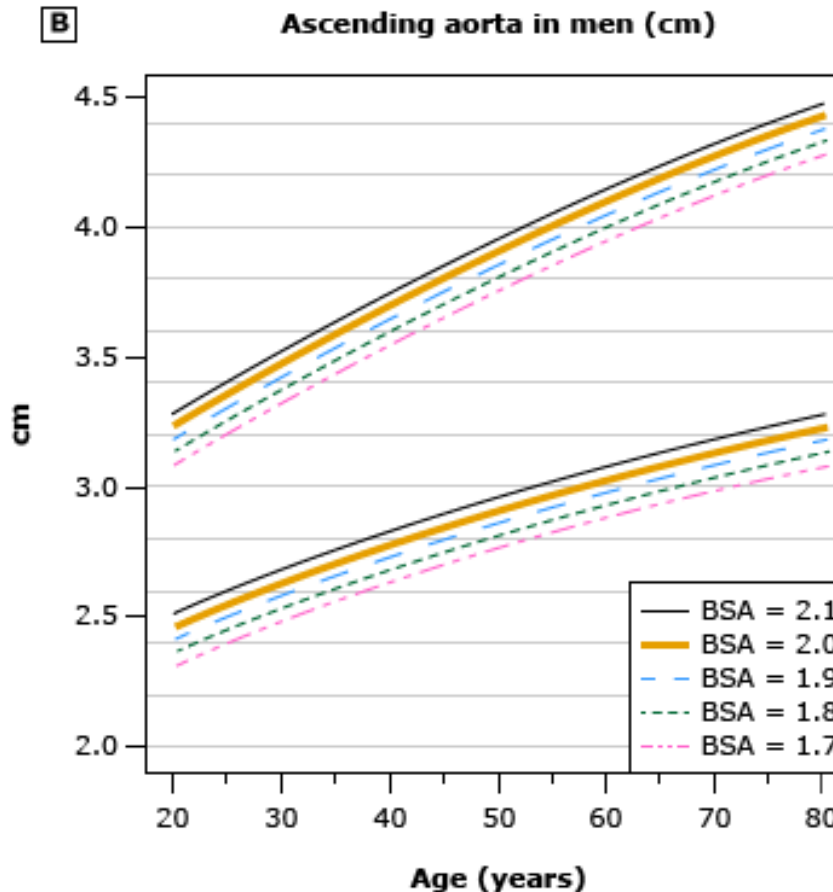
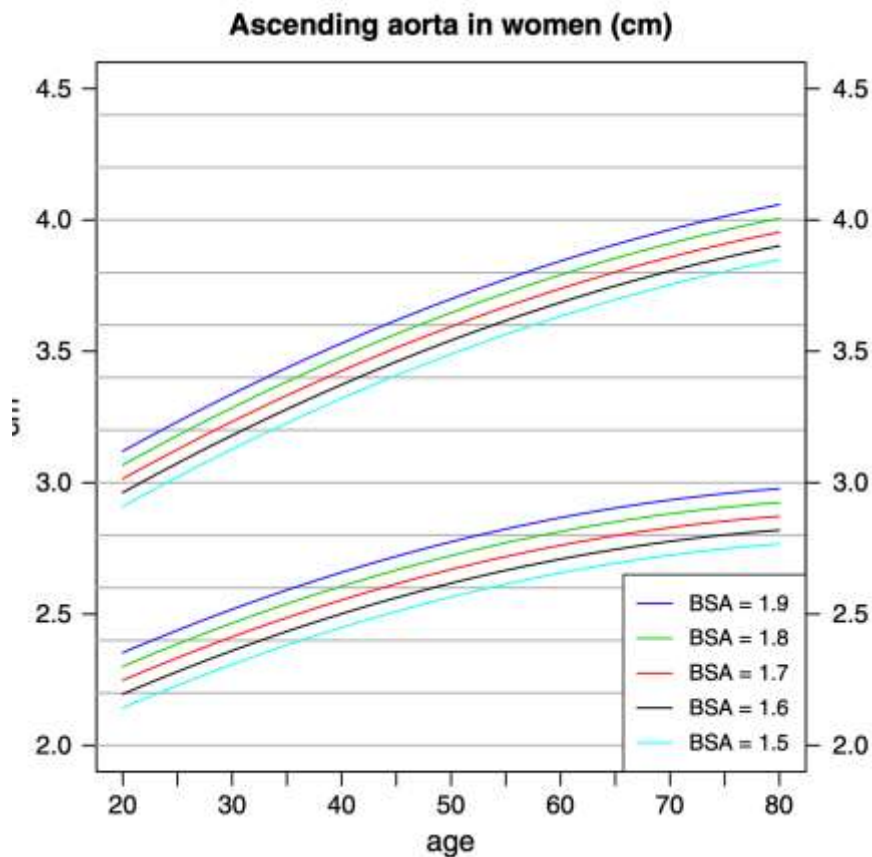
How & how often to Image?

- Start with baseline echocardiogram (Class 1)
- Obtain CT or MRI if unable to assess thoracic aortic anatomy/diameter (Class 2a)
- Follow up imaging in 6-12 months to assess rate of growth (Class 2a)
- Stable aortic aneurysm → surveillance imaging q6-24 months (Class 2a)

My Imaging Approach

Ascending Aortic Size	Imaging Interval
Normal (<4.0 cm)	Every 3-5 years
Mild dilation (4.0-4.4cm)	Every 2 years
Moderate dilation (4.5-4.9cm)	Every 1 year
Significant dilation (≥5.0cm)	Every 6 months & refer to surgeon

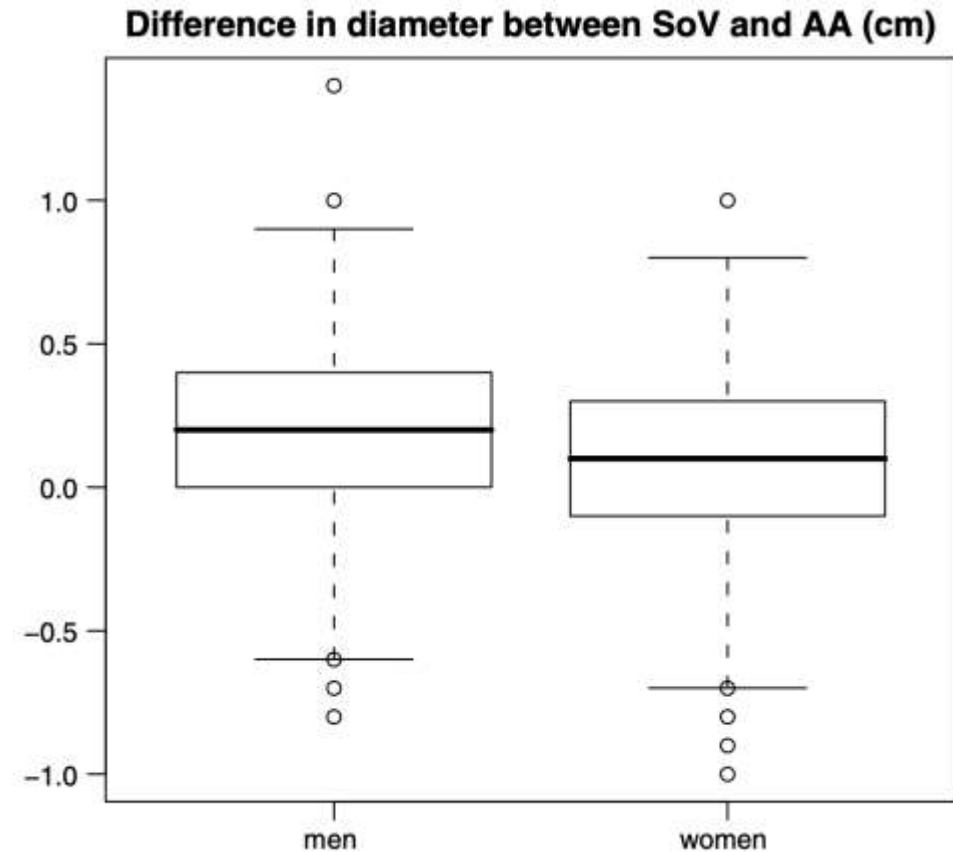
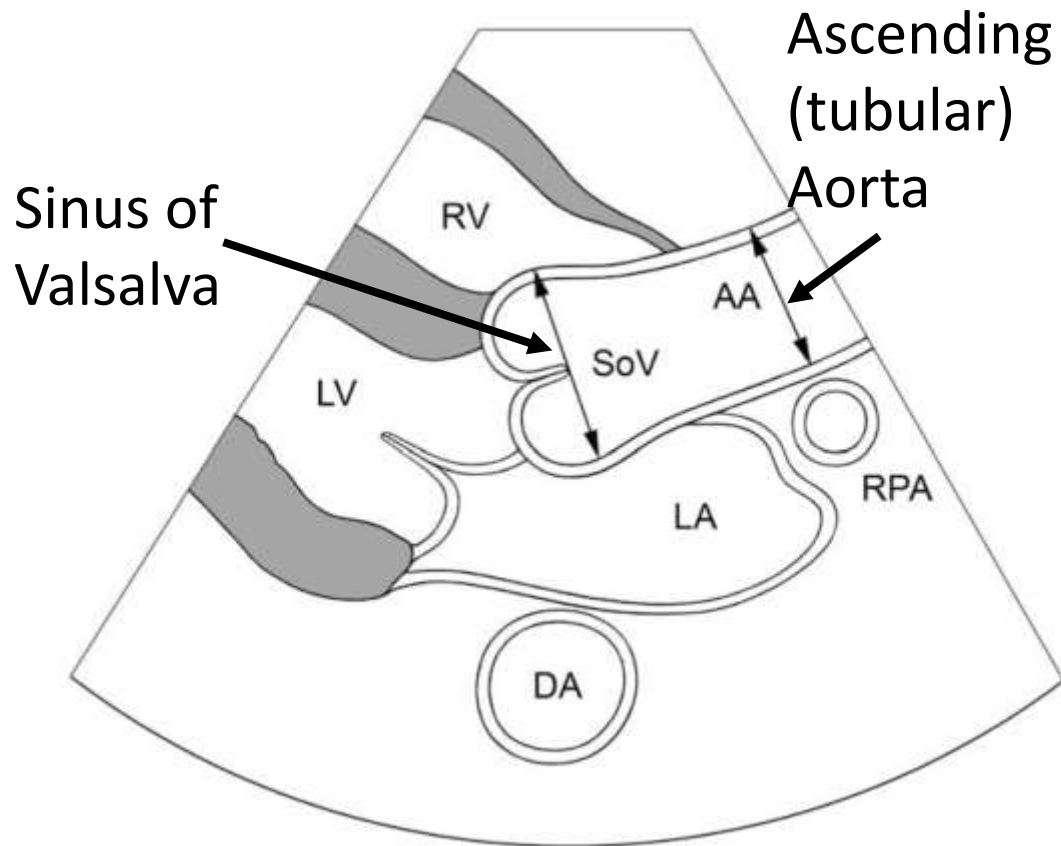
What Qualifies as Aortic Aneurysm?



Key Points:

- 1) Size increases with age
- 2) Size increases with body size
- 3) Size is larger in men vs. women

What Qualifies as Aortic Aneurysm?

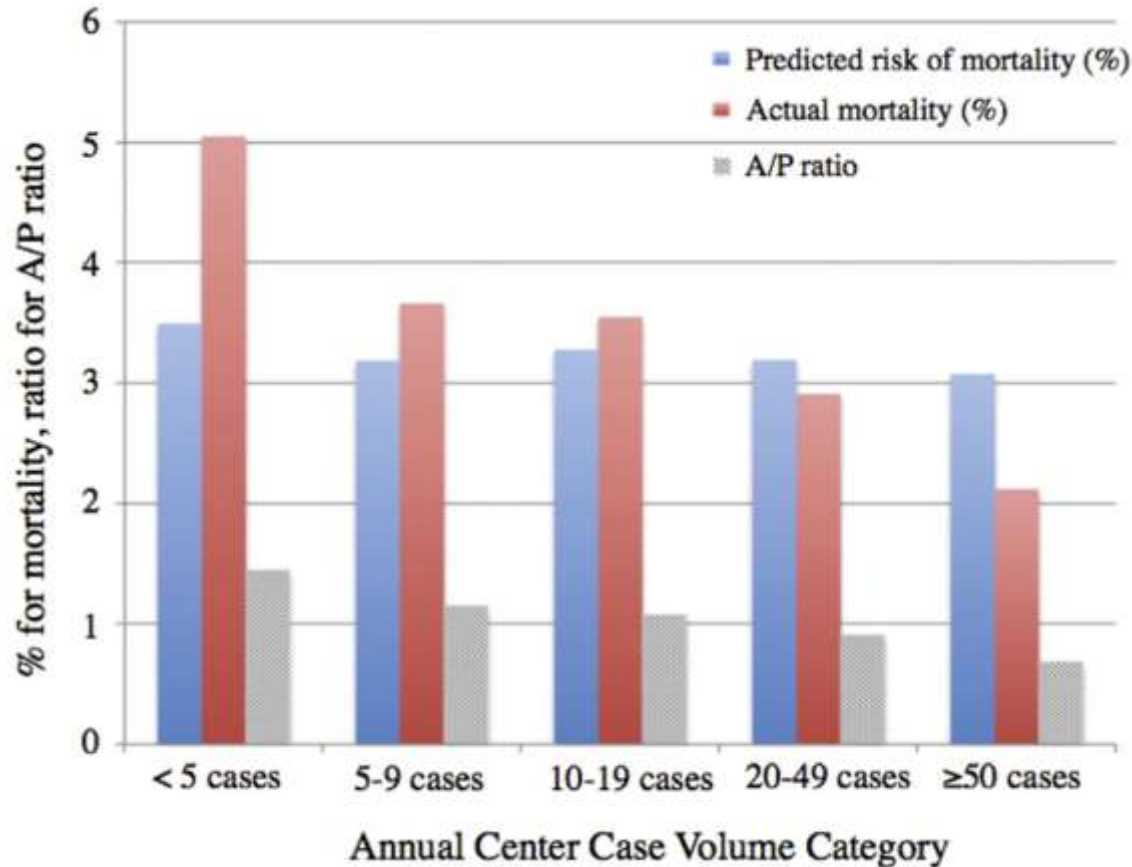


Thresholds for Surgery? Look at Height

		Aortic Size (cm)									
		3.5	4.0	4.5	5.0	5.5	6.0	6.5	7.0	7.5	8.0
Height (inches) (m)											
55	1.40	2.50	2.86	3.21	3.57	3.93	4.29	4.64	5.00	5.36	5.71
57	1.45	2.41	2.76	3.10	3.45	3.79	4.14	4.48	4.83	5.17	5.52
59	1.50	2.33	2.67	3.00	3.33	3.67	4.00	4.33	4.67	5.00	5.33
61	1.55	2.26	2.58	2.90	3.23	3.55	3.87	4.19	4.52	4.84	5.16
63	1.60	2.19	2.50	2.81	3.13	3.44	3.75	4.06	4.38	4.69	5.00
65	1.65	2.12	2.42	2.73	3.03	3.33	3.64	3.94	4.24	4.55	4.85
67	1.70	2.06	2.35	2.65	2.94	3.24	3.53	3.82	4.12	4.41	4.71
69	1.75	2.00	2.29	2.57	2.86	3.14	3.43	3.71	4.00	4.29	4.57
71	1.80	1.94	2.22	2.50	2.78	3.06	3.33	3.61	3.89	4.17	4.44
73	1.85	1.89	2.16	2.43	2.70	2.97	3.24	3.51	3.78	4.05	4.32
75	1.90	1.84	2.11	2.37	2.63	2.89	3.16	3.42	3.68	3.95	4.21
77	1.95	1.79	2.05	2.31	2.56	2.82	3.08	3.33	3.59	3.85	4.10
79	2.00	1.75	2.00	2.25	2.50	2.75	3.00	3.25	3.50	3.75	4.00
81	2.05	1.71	1.95	2.20	2.44	2.68	2.93	3.17	3.41	3.66	3.90

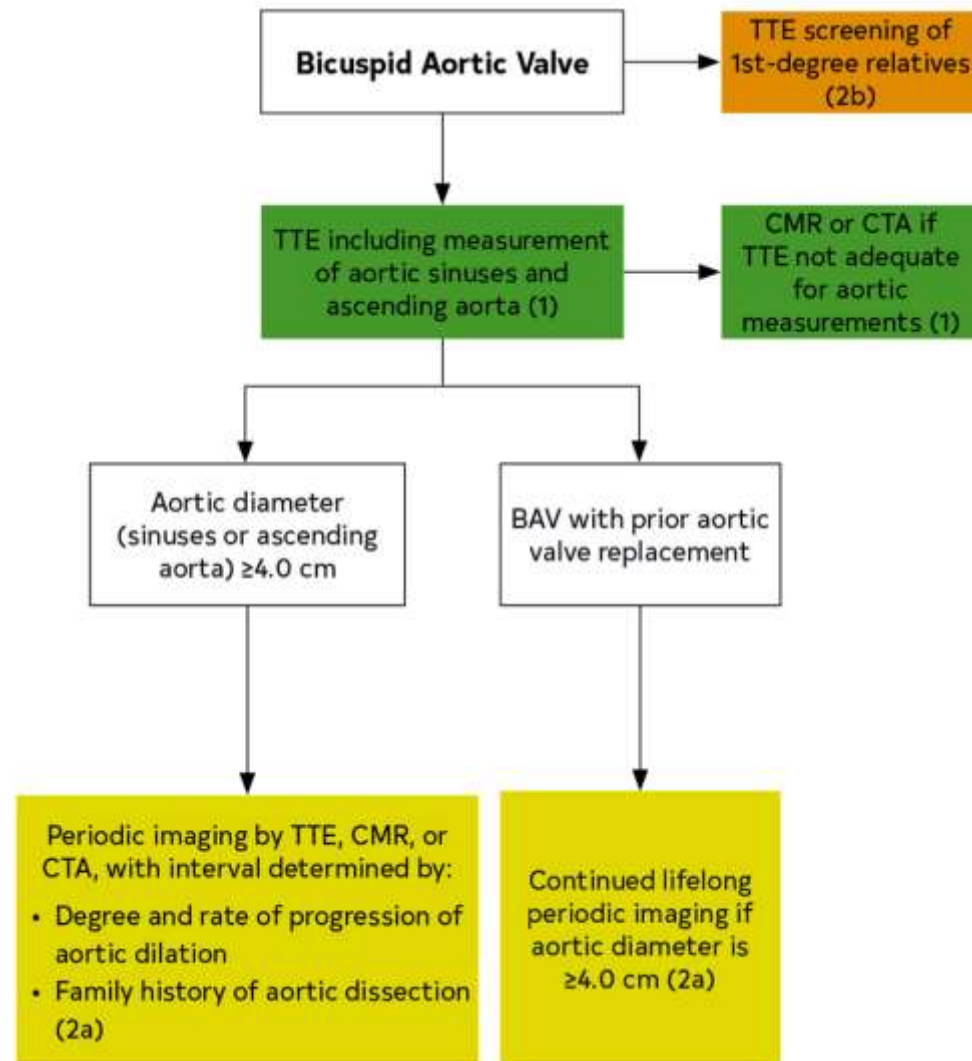
 = low risk (~ 4% per year)
 = moderate risk (~ 7% per year)
 = High risk (~ 12% per year)
 = severe risk (~ 18% per year)

Thresholds for Surgery? Consider Center Volume



Higher volume centers →
Lower actual mortality

Guideline Pathway



Medications to Prevent Aortopathy?

COR	LOE	RECOMMENDATIONS
1	B-NR	1. In patients with TAA and an average systolic BP (SBP) of ≥ 130 mm Hg or an average diastolic BP (DBP) of ≥ 80 mm Hg, the use of antihypertensive medications is recommended to reduce risk of cardiovascular events. ¹⁻³
2a	C-LD	2. In patients with TAA, regardless of cause and in the absence of contraindications, use of beta blockers to achieve target BP goals is reasonable. ^{1,4,5}
2a	C-EO	3. In patients with TAA, regardless of etiology and in the absence of contraindications, ARB therapy is a reasonable adjunct to beta-blocker therapy to achieve target BP goals. ⁶

Aortic Size Criteria	Standard Aortic Aneurysm	Non-syndromic hereditary Aortic Aneurysm	BAV Aortopathy
≥5.5cm	Surgery (Class 1)	Surgery (Class 1)	Surgery (Class 1)

CEL
R

Aortic Size Criteria	Standard Aortic Aneurysm	Non-syndromic hereditary Aortic Aneurysm	BAV Aortopathy
≥5.5cm	Surgery (Class 1)	Surgery (Class 1)	Surgery (Class 1)
5.0-5.4cm	Surgery at high-volume center (Class 2a) If other valve or cardiac surgery (Class 2a)	Surgery (Class 1)	Surgery if low surgical risk and high-volume center (Class 2b) If high risk features (Class 2a) <ul style="list-style-type: none"> - FHx of dissection - Rapid growth (>0.3cm/yr) - Coarctation - “Root phenotype”

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4.5-4.9cm	If other valve or cardiac surgery in high-volume center (Class 2a)	If high-risk features (Class 2a) <ul style="list-style-type: none"> - FHx dissection <5.0cm - FHx sudden death <50yo If other valve or cardiac surgery in high-volume center (Class 2a)	If other valve or cardiac surgery in high-volume center (Class 2a)

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Height-adjusted	Surgery if aortic area/height ratio ≥10 cm ² /m (Class 2a)		
Fast growth	≥0.6cm in 2yr OR ≥0.5cm in 1yr (Class 1)	≥0.6cm in 2yr OR ≥0.5cm in 1yr (Class 1)	

Return to the Case

- 55yo man with BAV aortopathy (43mm) and moderate aortic stenosis
- Is there an underlying cause for his BAV and aortic aneurysm?
 - Maybe – has family history, young age
 - Recommend genetic screening
- What imaging regimen is recommended?
 - Baseline echocardiogram
 - CT/MRI if not able to completely visualize thoracic aorta
 - Repeat imaging in 6-12 months (assess rate of growth), then every 2 years
- When might surgery be indicated?
 - $\geq 5.0\text{cm}$ if at high volume center
 - $\geq 4.5\text{cm}$ if valve or other cardiac surgery
 - If rapid growth ($\geq 0.6\text{cm}$ in 2yr, $\geq 0.5\text{cm}$ in 1 year)

Thank you!

