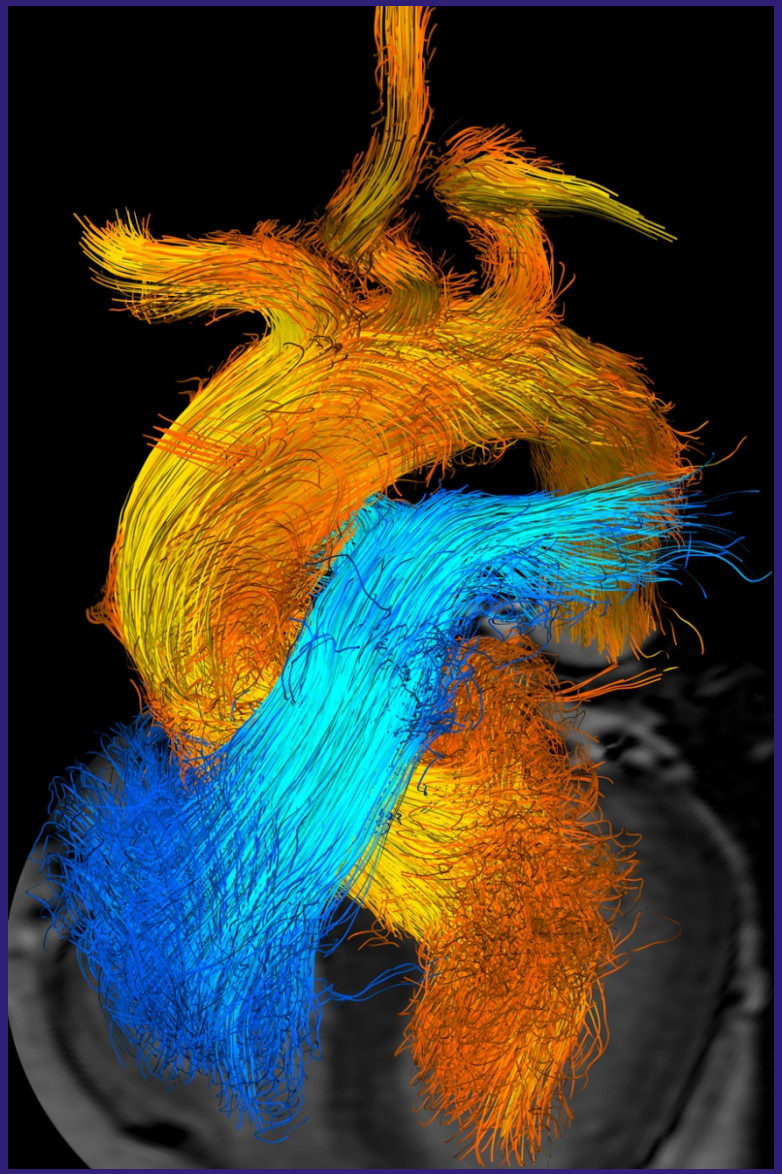


Aneurisma aortico in paziente con valvola tricuspide e bicuspide. Quando inviarlo al cardiochirurgo.

Dr. Francesco Baldascino

EZIOPATOGENESI

Table 1
Etiologies of ascending aortic dilatation.

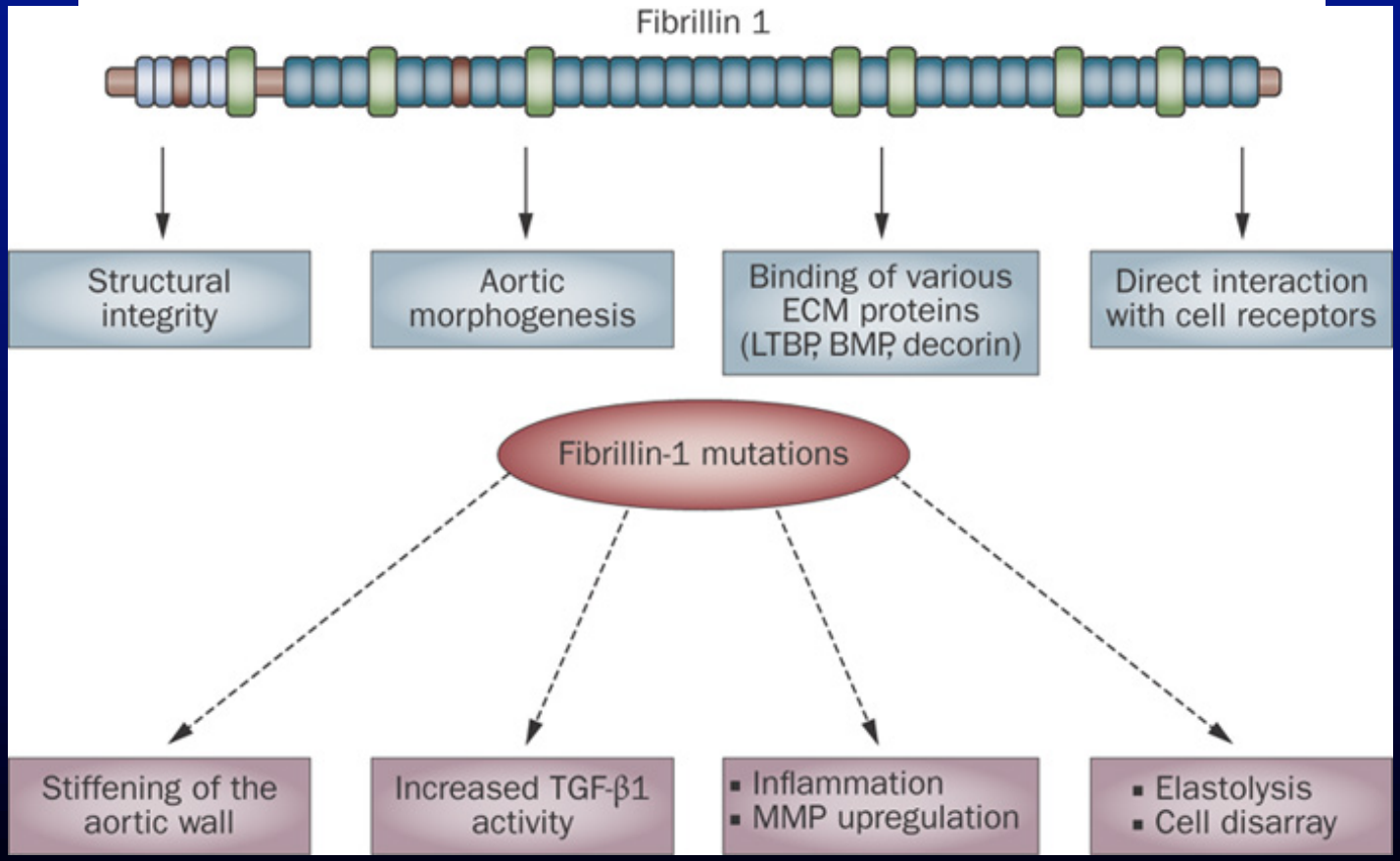
Congenital	Hereditary	Acquired
Bicuspid aortic valve Tetralogy of Fallot	Connective tissue disease	Hypertension Infectious
	- Marfan syndrome - Ehlers–Danlos syndrome - Loeys–Dietz syndrome - Arterial tortuosity syndrome - Aneurysms osteoarthritis syndrome	- Syphilitic - Bacterial infections - Viral - Fungal
	Familial aortic syndromes	Auto-immune
	- Autosomal dominant - Sporadic	- Takayasu - Behçet disease - Idiopathic aortitis
		Post-traumatic Chronic aortic dissection AV fistula Post-stenotic

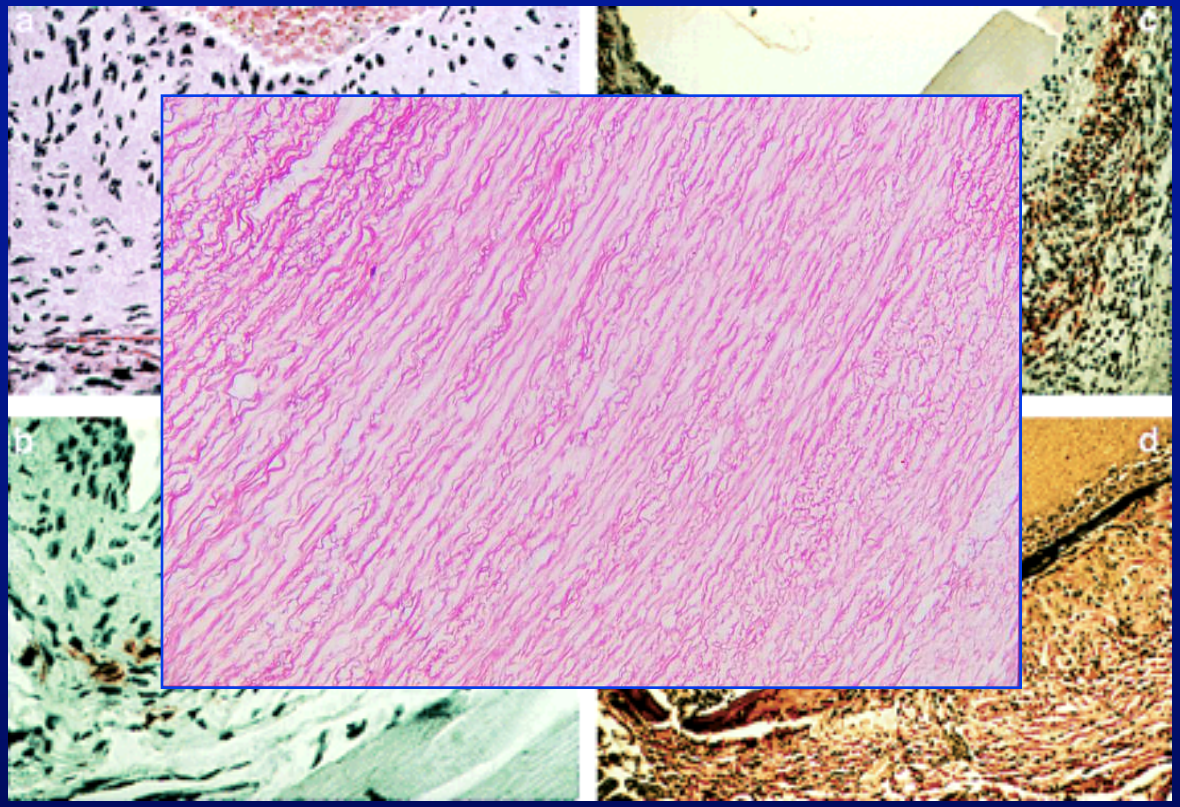
IJC Heart & Vasculature 6 (2015) 91–100

Thoracic aortic aneurysm is a genetic disease

Table 7. Genetic Syndromes Associated With Thoracic Aortic Aneurysm and Dissection

Genetic Syndrome	Common Clinical Features	Genetic Defect	Diagnostic Test
Marfan syndrome	Skeletal features (see text) Ectopia lentis Dural ectasia	*FBN1* mutations*	Ghent diagnostic criteria DNA for sequencing





Histopathology and immunohistochemistry of homozygous mgR mice examined between 3 and 6 mo of postnatal life. (*a*) Intimal hyperplasia and disorganized elastic lamellae at 3 mo; hemotoxylin and eosin. (*b* and *c*) Horseradish peroxidase immunostaining using mAb F4/80 specific for mature macrophages performed on 3-mo (*b*) and 5–1/2-mo (*c*) specimens. (*c*) Extensive macrophage infiltration at the adventitial border associated with aneurysmal dilatation. (*d*) Bony metaplasia with hematopoiesis within the vessel wall at the junction of the aortic annulus and the ascending aorta at 6 mo; Verhoeff–van Gieson. (×350.)

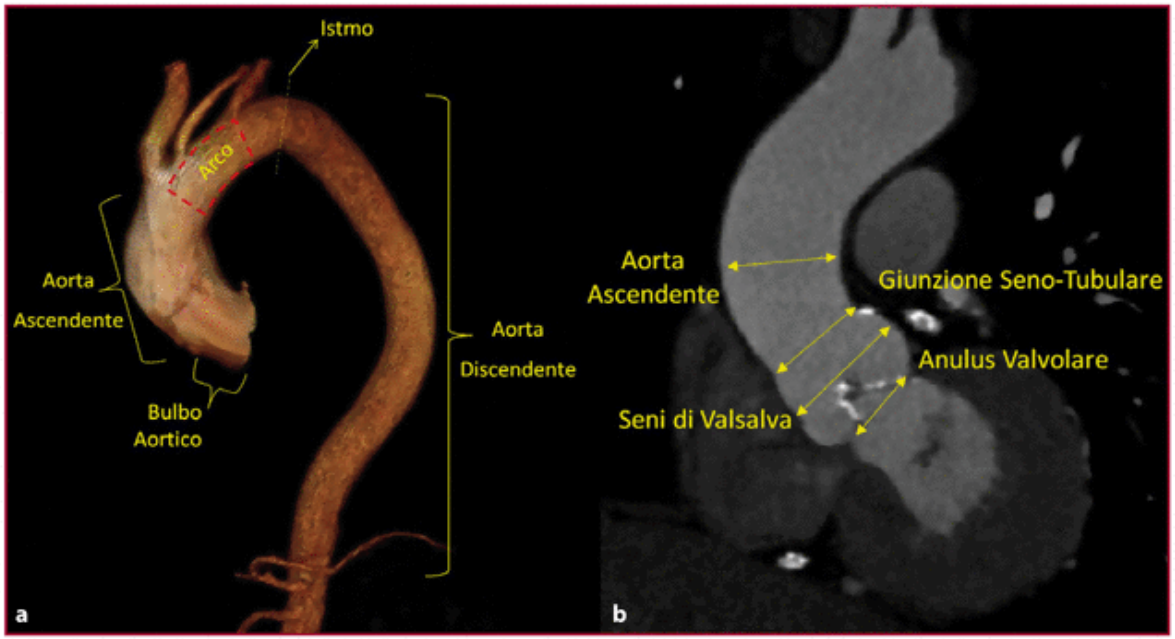


Fig. 7.1 Anatomia dell'aorta toracica (**a**) e del tratto ascendente (**b**)

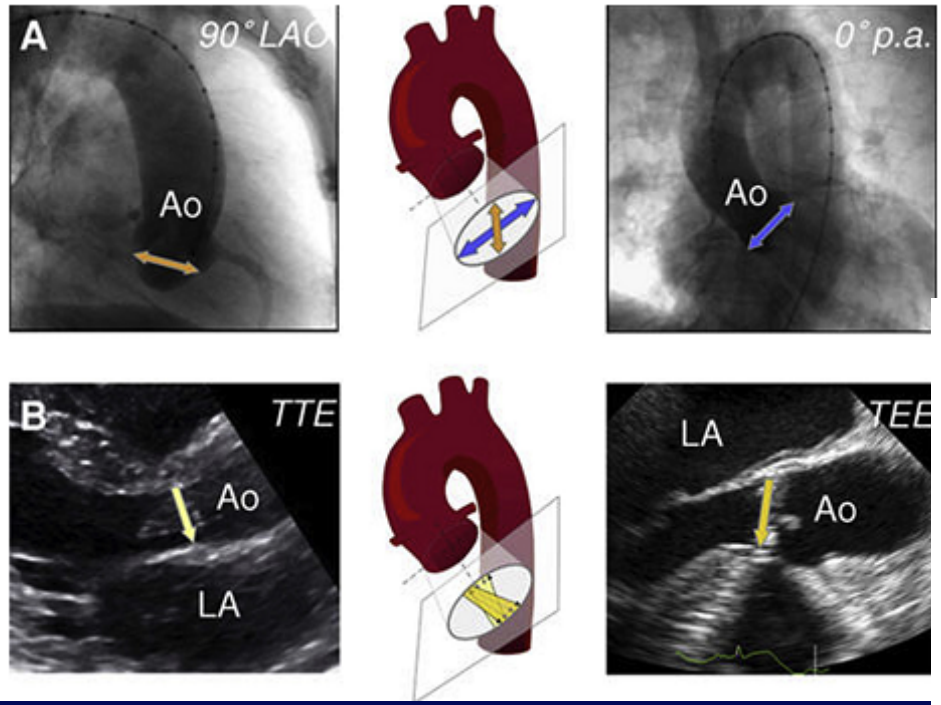
Tabella 7.1 Aorta toracica

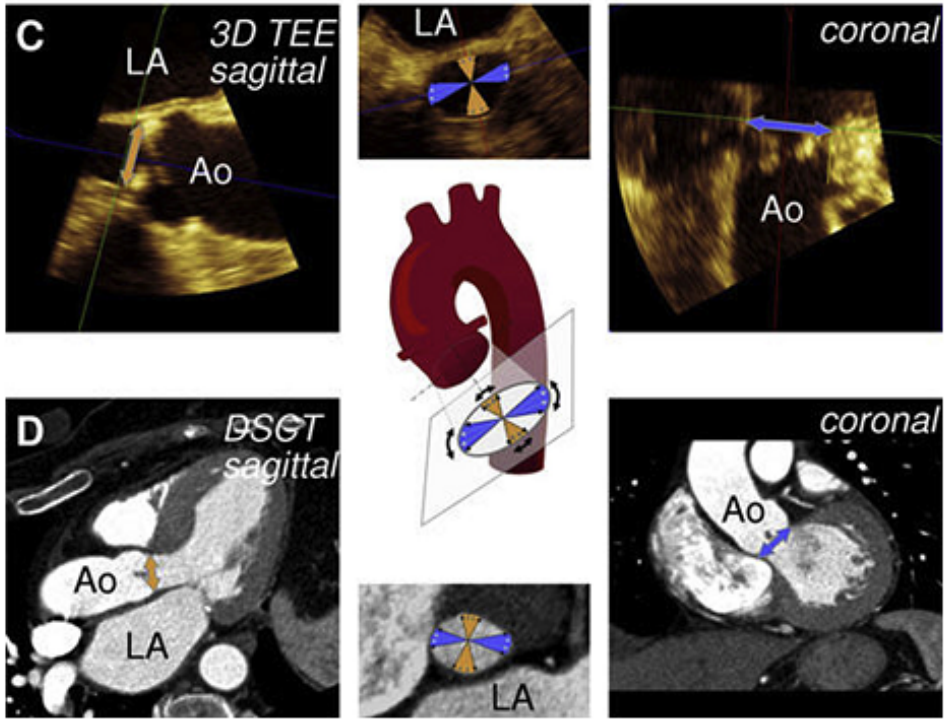
	Definizione	Diametri di riferimento	Maschi (diametro, mm)	Femmine (diametro, mm)	Rami collaterali
Tratto ascendente					
• Annulus	Tratto compreso tra la valvola aortica e l'origine del primo vaso sovraortico (tronco anonimo)	Seni di Valsalva	29-35	25-32	Coronaria destra
• Seni di Valsalva		Porzione tubulare	28-36	26-33	Coronaria sinistra
• Giunzione seno-tubulare					
• Porzione tubulare dell'aorta ascendente					
Arco aortico					
• Istmo	Tratto compreso tra il primo ramo sovraortico e il legamento arterioso (porzione prossimale e distale divise dall'origine della succlavia sinistra)	Arco prossimale	26-34	24-32	Tronco anonimo
		Istmo	21-30	20-27	Carotide comune sinistra
		Arco distale	23-31	21-27	Succlavia sinistra
Tratto discendente	Tratto compreso tra il legamento arterioso e lo iato diaframmatico	Iato Diaframmatico	22-28	22-28	Bronchiali Pericardio-mediastiniche Esofagee Intercostali Freniche superiori

Le fasi di una misura

- Quale grandezza misurare
  - Scopo/decisione/modello
- Quale unità di misura adottare
  - Convenienza/universalità/aspetti legali e scientifici/stabilità e ripetibilità
- Relazione fra la grandezza e l'udm
  - Risoluzione/precisione/accuratezza
- Il mondo esterno è isolato?
  - Influssi sullo strumento/ sul comparatore/sulla grandezza, generano incertezza

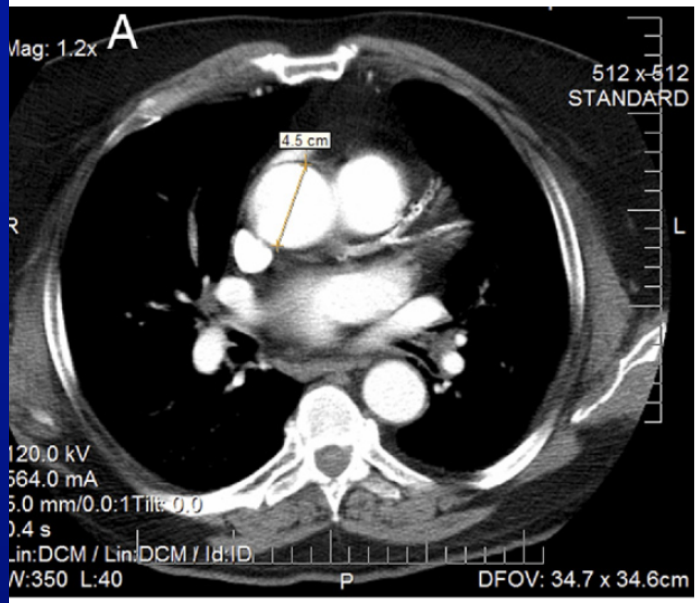
The question “How big the aorta?”

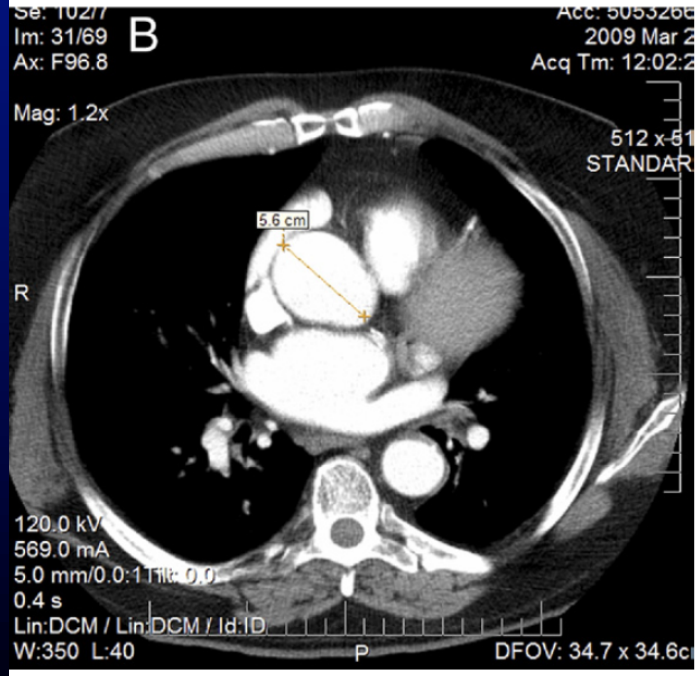




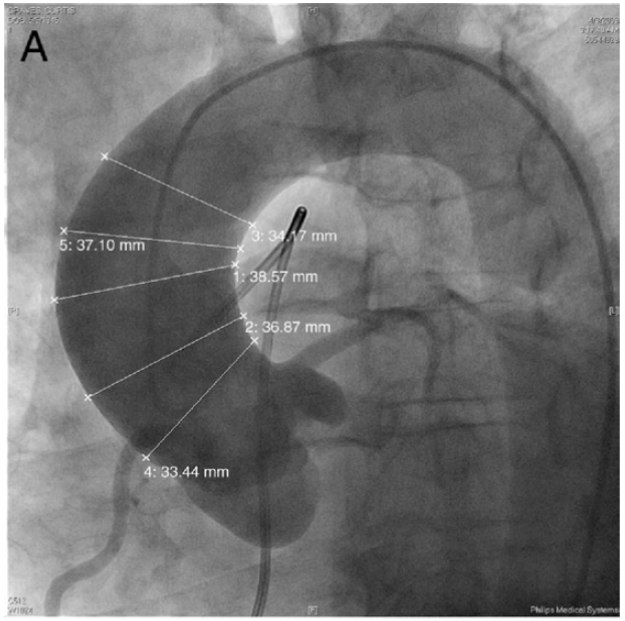
variability. In several studies, variability of measurement of proximal aortic diameters ranges from 1.6 to 5 mm.[8,23,24,27,28] Given this degree of variability, apparent small changes in proximal aortic diameters on serial computed tomographic examinations may be within the range of measurement error. Accordingly, for all imaging techniques, we recommend that changes of ≤3 mm by electrocardiographically gated CT and ≤5 mm without electrocardiographic gating be viewed with caution and skepticism.

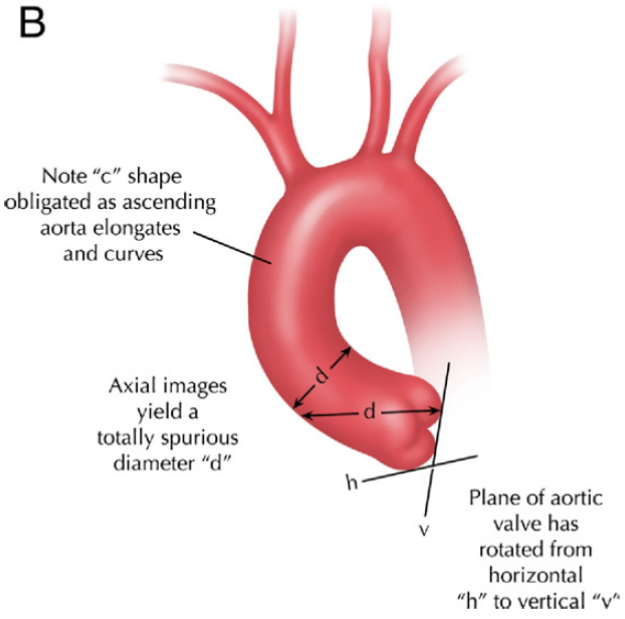
(J Am Soc Echocardiogr 2015;28:119-82.)





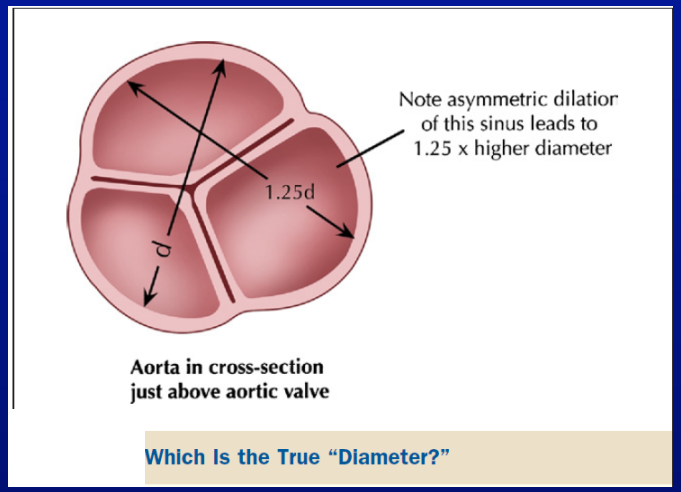
Limitations of Axial Imaging

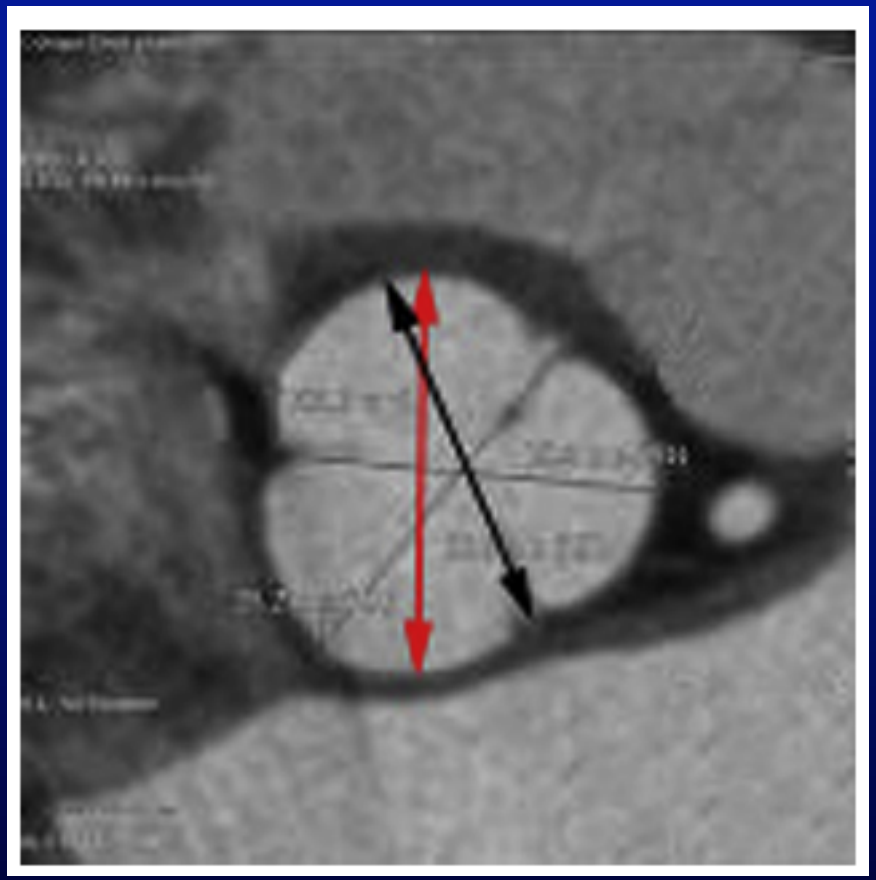




Confounding of Ascending Aortic Measurements Due to Elongation and "C" Shape of Ascending Aorta

The question “How big the aorta?”





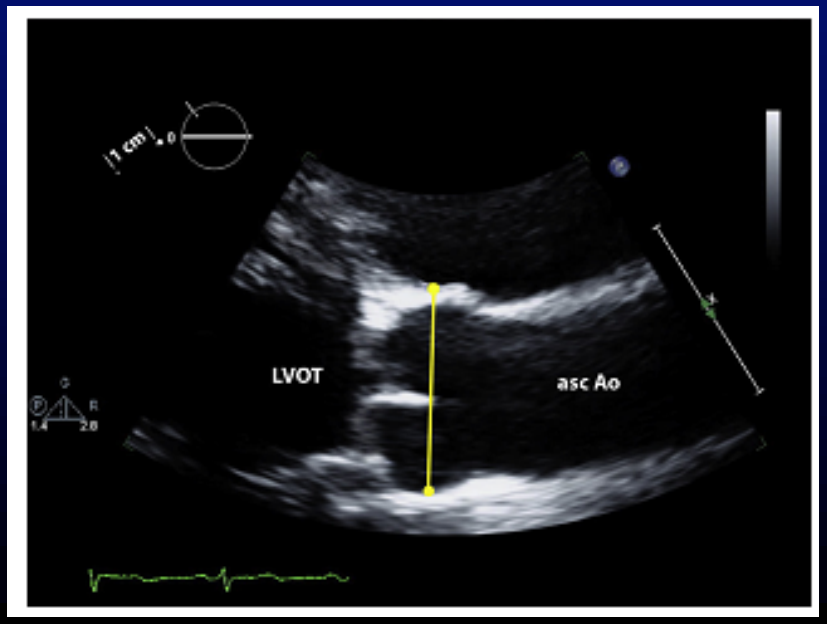


Figure 8 Computed tomographic scan image of aortic root illustrates that the mean difference of the aortic root diameter is about 2 mm larger measured by the anteroposterior diameter (*sinus-sinus*) shown by red arrow than by the sinus-commissure diameter (*black arrow*).

(J Am Soc Echocardiogr 2015;28:119-82.)

The question “How big the aorta?”

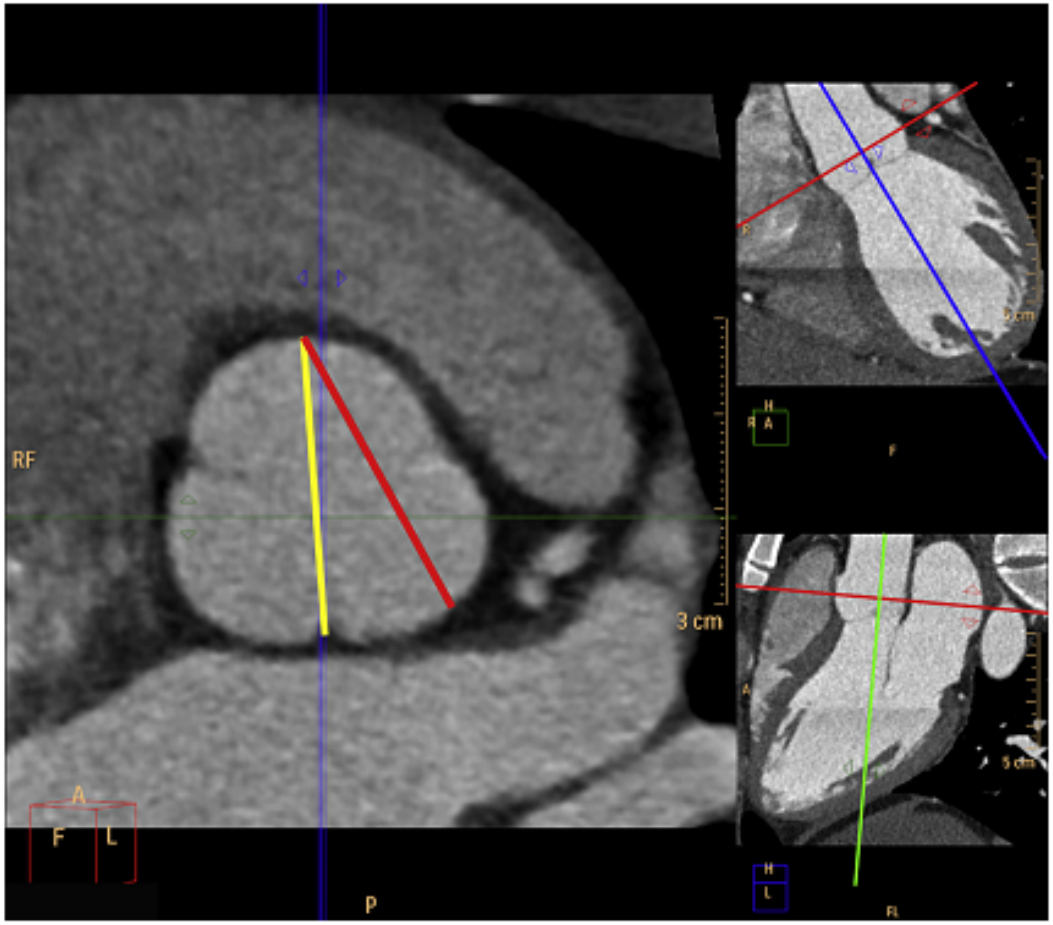


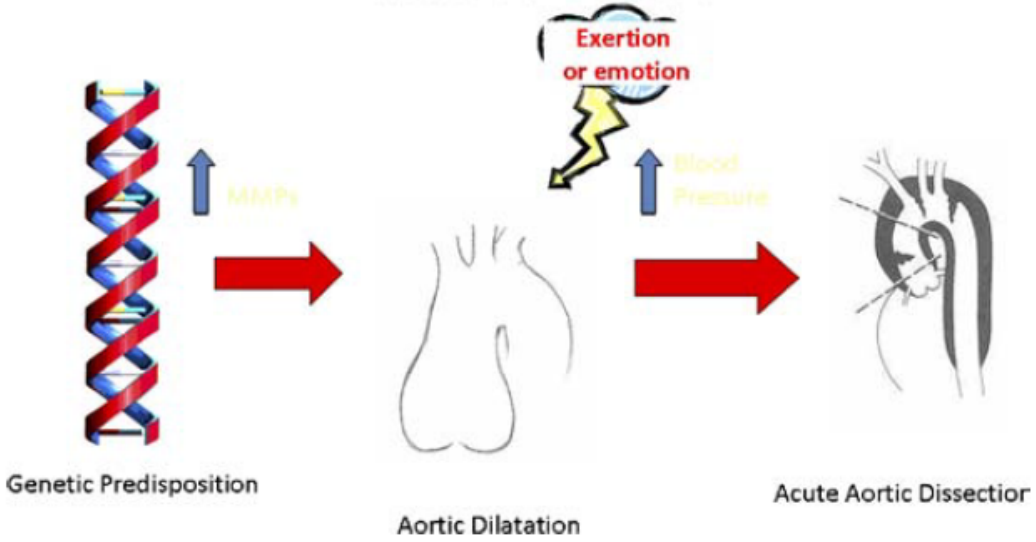
Figure 7 Aortic root measurements by CT. The aortic root diameter is commonly measured between the inner edges from one commissure to opposite sinus (*yellow line*) or from one sinus to another sinus (*red line*), as shown in the large image (*left*), which is a zoomed cross-sectional view of the aortic root at the sinus of Valsalva level using a double oblique image for orientation (shown in the *right panel*).

Table 17 Recommendations for choice of imaging modality for TAA

Modality	Recommendation	Advantages	Disadvantages
CT	First-line	• First-line technique for staging, surveillance • Contrast: enhanced CT and MRI very accurate for measuring size of all TAAs (superior to echocardiography for distal ascending aorta, arch, and descending aorta) • All segments of aorta and aortic branches well visualized	• Use of ionizing radiation and ICM • Cardiac motion can cause imaging artifacts
MRI	Second-line	• Ideal technique for comparative follow-up studies • Excellent modality in stable patients • Preferred for follow-up for younger patients • Avoids ionizing radiation • Can image entire aorta	• Examination times longer than CT • Benefits from patient cooperation (breath hold) • Limited in emergency situations in unstable patients and patients with implantable metallic devices • Benefits from gadolinium
TTE	Second-line	• Usually diagnostic for aneurysms effecting aortic root • Useful for family screening • Useful for following aortic root disease • Excellent reproducibility of measurements • Excellent for AR, LV function	• Distal ascending aorta, arch, and descending aorta not reliably imaged
TEE	Third-line	• Excellent for assessment of AR mechanisms • Excellent images of aortic root, ascending aorta, arch, and descending thoracic aorta	• Less valuable for routine screening or serial follow-up (semi-invasive) • Distal ascending aorta may be poorly imaged • Does not permit full visualization of arch vessels • Limited landmarks for serial examinations
Aortography	Third-line	• Reserved for therapeutic intervention • Useful to guide endovascular procedures	• Invasive; risk for contrast-induced nephropathy • Visualizes only aortic lumen • Does not permit accurate measurements

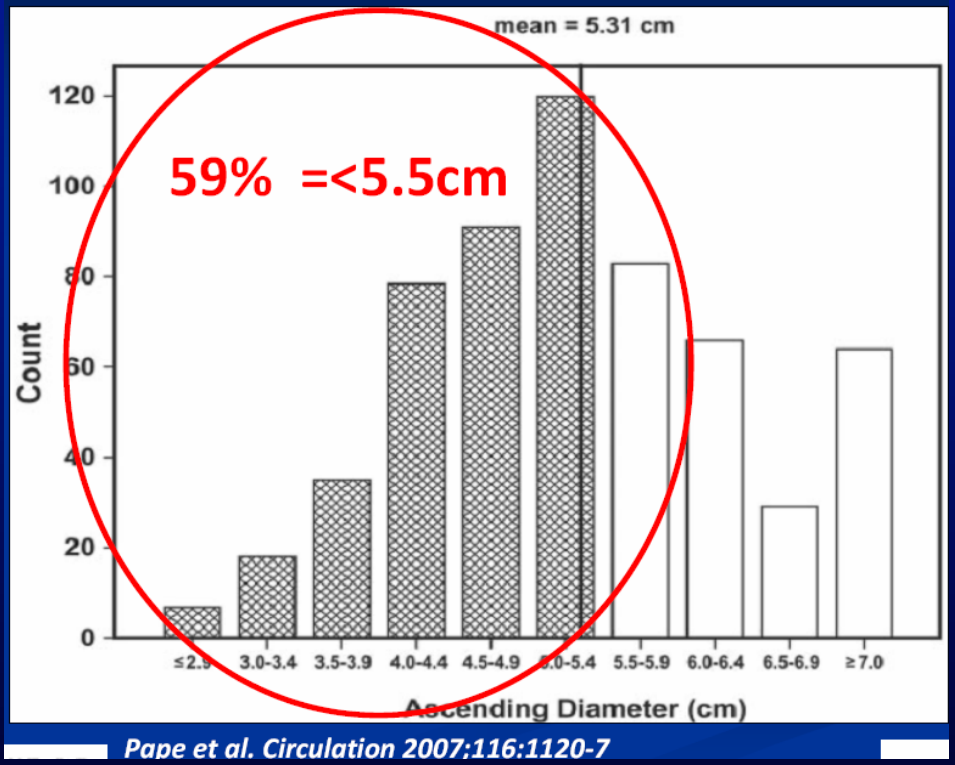
(J Am Soc Echocardiogr 2015;28:119-82.)

Why does dissection pick one point in time to occur?

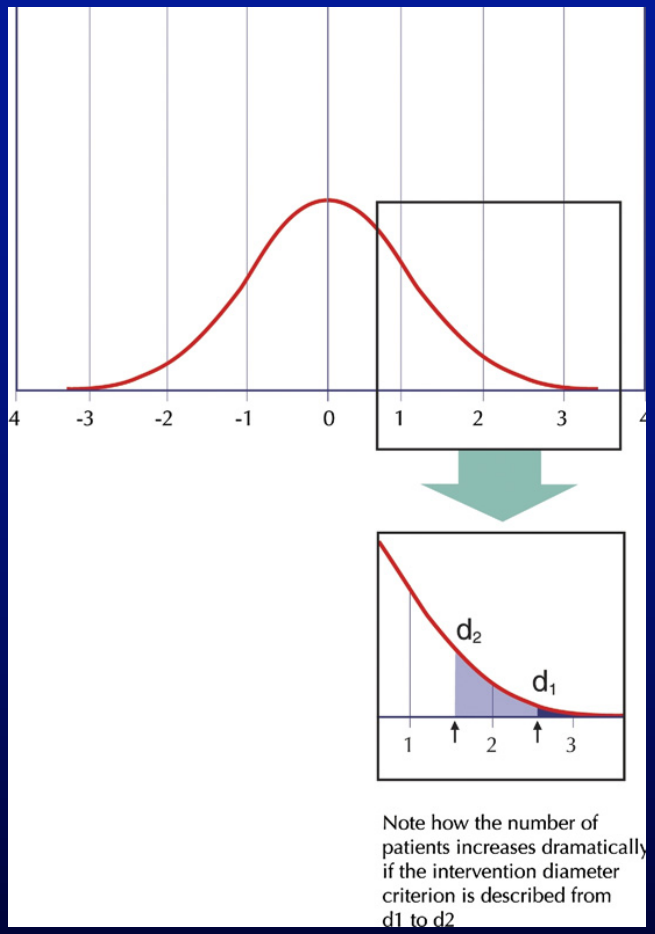


Conceptual Pathway
Leading to Acute Aortic Dissection

IRAD Registry



Pape et al. Circulation 2007;116:1120-7



Depiction of a normal distribution curve of aortic size (marked in SDs). Note how small the "tails" of such a curve are. Large aneurysms would reside far out in the tails. While dissections do occur at small dimensions, note how rapidly the at-risk group increases in number as the putative criterion diameter goes from d1 to d2. We anticipate that millions of Americans harbor small thoracic aortic aneurysms, making for a very large denominator of vulnerable patients, and a correspondingly low likelihood of dissection at small sizes. See the "Dissections Can and Do Occasionally Occur at Small Aortic Sizes" section for details. Figure illustration by Rob Flewell.

Age of complication for TAA.

	Mean age at presentation (years)
Hypertensive	64.2
Marfan syndrome	24.5
	34.4 (age at prophylactic surgery)
	39.4 (age at aortic dissection)
Bicuspid aortic valve	49
Familial (non syndromic)	56.8
Loeys–Dietz syndrome	19.8 (age at complications)
Ehlers–Danlos syndrome	No data

Diameter of ascending aorta at timing of complications.

	Mean size at complications (mm)[a]	Source
Hypertensive	60	(Davies, Kaple et al., 2007) [16]
Marfan syndrome	51	(Roman, Rosen et al., 1993) [19]
	56	(Kornbluth, Schnittger et al., 1999) [20]
	50–59	(Jondeau, Detaint et al., 2012) [17]
Bicuspid aortic valve	52	(Davies, Kaple et al., 2007) [16]
Familial (non-syndromic)	No data	No data
Loeys–Dietz syndrome	40–50	(Loeys, Schwarze et al., 2006) [8]
Ehlers–Danlos syndrome	No data	No data

[a] It is to be noted that the risk of complications is higher for bigger diameters in each of the preceding conditions.

The ascending aortic aneurysm: When to intervene?

Emile Saliba *, Ying Sia, In collaboration with Annie Dore, Ismael El Hamamsy

Montreal Heart Institute, 5000 Bélanger Street, Montreal, QC H1T 1C8, Canada
Hôtel Dieu de Montreal, CHUM — Centre Hospitalier de l'Université de Montréal, 3840 St Urbain St, Montreal, QC H2W 1T8, Canada

IJC Heart & Vasculature 6 (2015) 91–100

Determinants of Aortic Diameter:

- Role of age

Scandinavian Cardiovascular Journal. 2006; 40: 175–178

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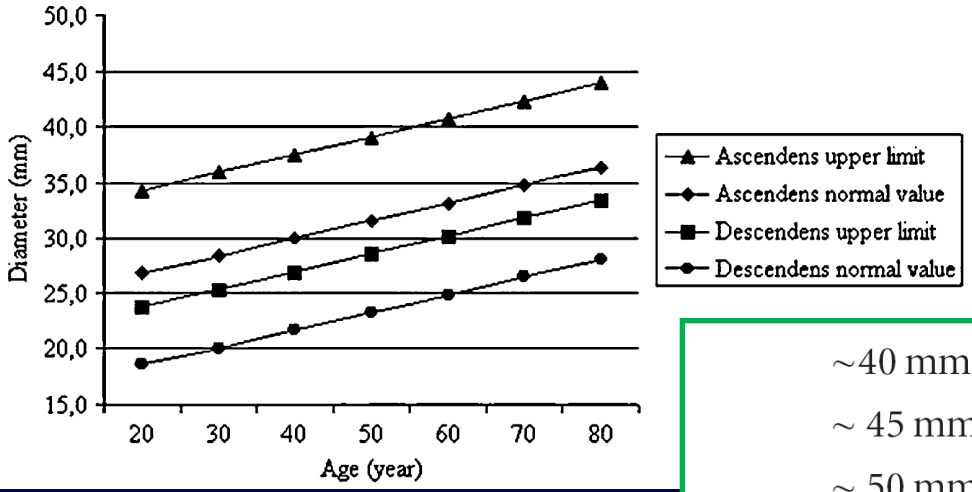
ORIGINAL ARTICLE

Thoracic aorta – dilated or not?

MATIAS HANNUKSELA[1], STEFAN LUNDQVIST[2] & BO CARLBERG[3]



The diameter increased by 0.12 – 0.20 mm (mean 0.17 mm) for year

~40 mm in a 20-year-old

~ 45 mm in a 40-year-old

~ 50 mm in a 60-year-old

~ 55 mm in an 80-year old

Table 1 Normal aortic root diameter by age for men with BSA of 2.0 m^2

	Age (y)					
	15–29	30–39	40–49	50–59	60–69	≥70
Mean normal (cm)	3.3	3.4	3.5	3.6	3.7	3.8
Upper limit of normal (cm) (95% CI)	3.7	3.8	3.9	4.0	4.1	4.2

Add 0.5 mm per 0.1 m^2 BSA above 2.0 m^2 or subtract 0.5 mm per 0.1 m^2 BSA below 2.0 m^2.[6]
CI, Confidence interval.

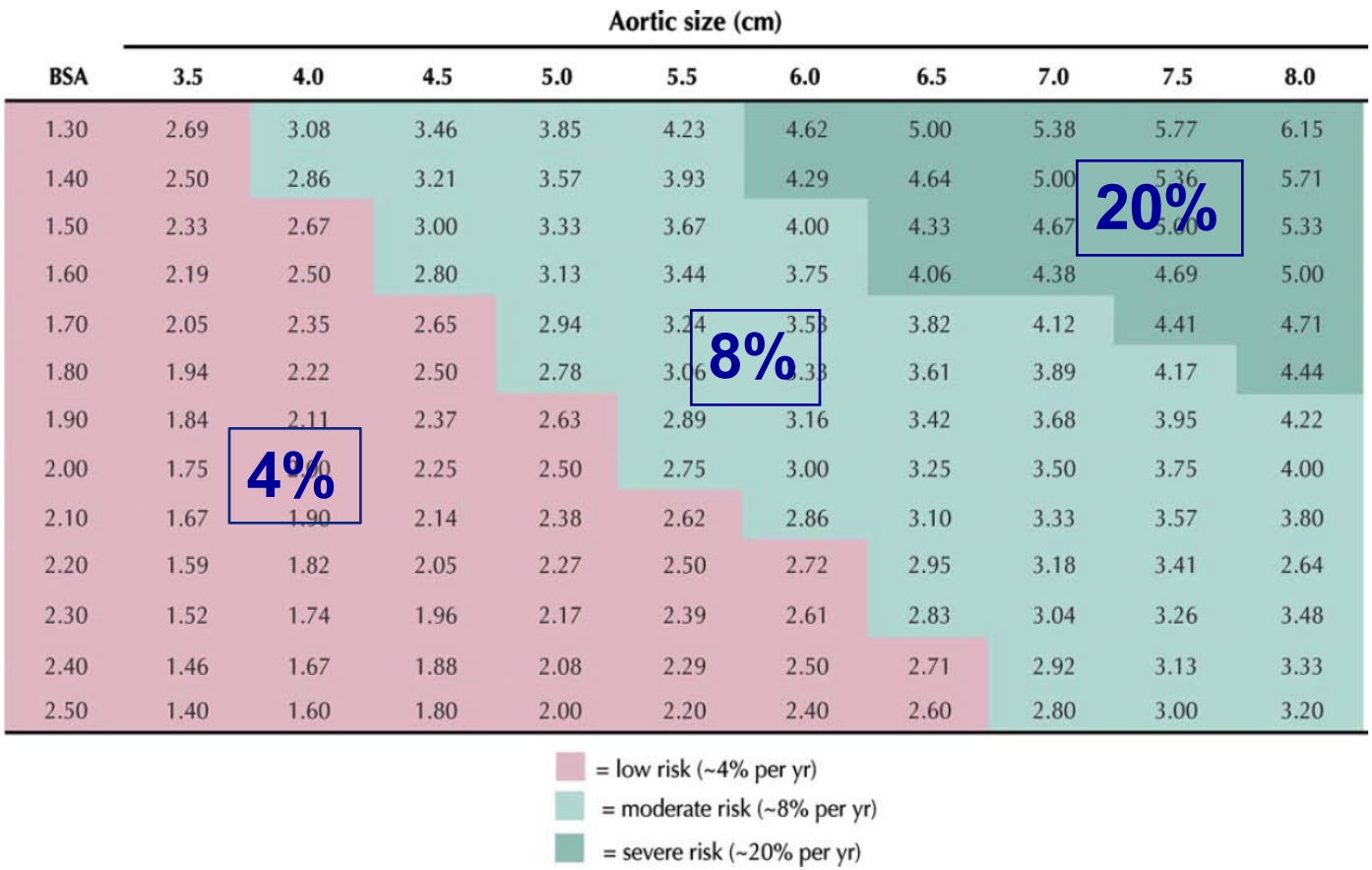
Table 2 Normal aortic root diameter by age for women with BSA of 1.7 m^2

	Age (y)					
	15–29	30–39	40–49	50–59	60–69	≥70
Mean normal (cm)	2.9	3.0	3.2	3.2	3.3	3.4
Upper limit of normal (cm)	3.3	3.4	3.6	3.6	3.7	3.9

Add 0.5 mm per 0.1 m^2 BSA above 1.7 m^2 or subtract 0.5 mm per 0.1 m^2 BSA below 1.7 m^2.[6]

(J Am Soc Echocardiogr 2015;28:119-82.)

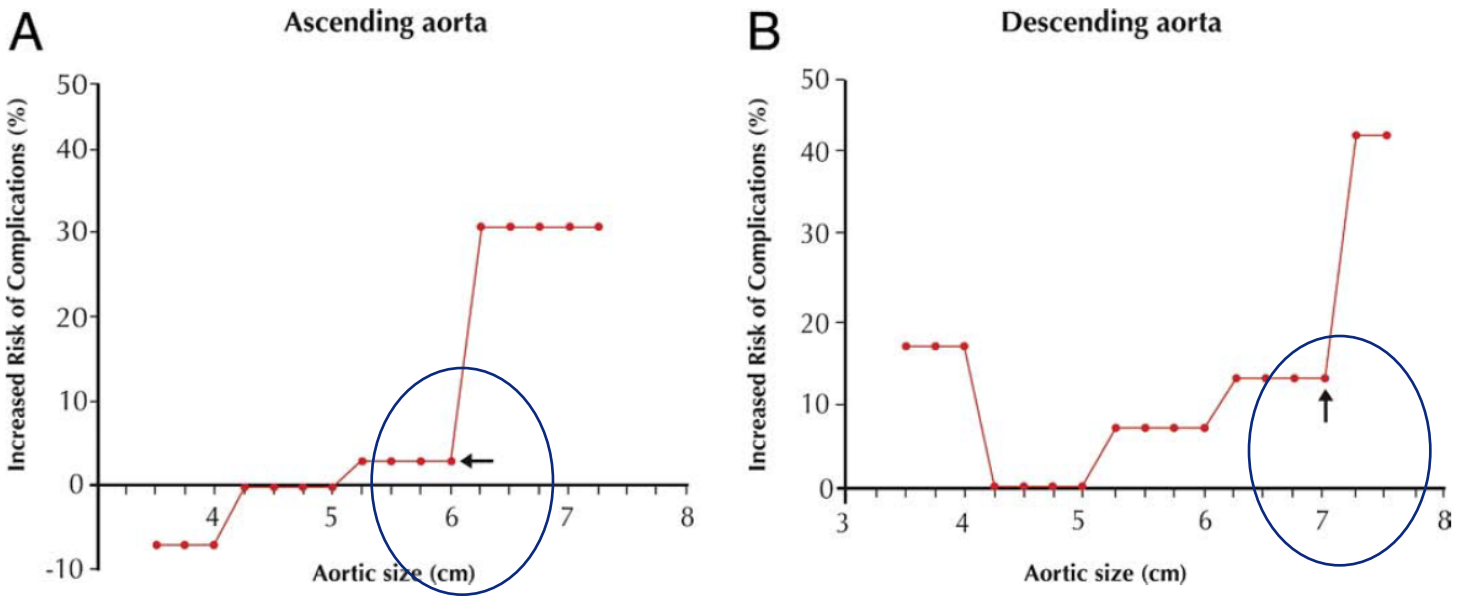
Aortic Size Index (Cross-Sectional Area/ BSA)



	Aortic size (cm)									
BSA	3.5	4.0	4.5	5.0	5.5	6.0	6.5	7.0	7.5	8.0
1.30	2.69	3.08	3.46	3.85	4.23	4.62	5.00	5.38	5.77	6.15
1.40	2.50	2.86	3.21	3.57	3.93	4.29	4.64	5.00	5.36	5.71
1.50	2.33	2.67	3.00	3.33	3.67	4.00	4.33	4.67	5.00	5.33
1.60	2.19	2.50	2.80	3.13	3.44	3.75	4.06	4.38	4.69	5.00
1.70	2.05	2.35	2.65	2.94	3.24	3.53	3.82	4.12	4.41	4.71
1.80	1.94	2.22	2.50	2.78	3.06	3.33	3.61	3.89	4.17	4.44
1.90	1.84	2.11	2.37	2.63	2.89	3.16	3.42	3.68	3.95	4.22
2.00	1.75	2.00	2.25	2.50	2.75	3.00	3.25	3.50	3.75	4.00
2.10	1.67	1.90	2.14	2.38	2.62	2.86	3.10	3.33	3.57	3.80
2.20	1.59	1.82	2.05	2.27	2.50	2.72	2.95	3.18	3.41	2.64
2.30	1.52	1.74	1.96	2.17	2.39	2.61	2.83	3.04	3.26	3.48
2.40	1.46	1.67	1.88	2.08	2.29	2.50	2.71	2.92	3.13	3.33
2.50	1.40	1.60	1.80	2.00	2.20	2.40	2.60	2.80	3.00	3.20

Aortic Size Index Nomogram

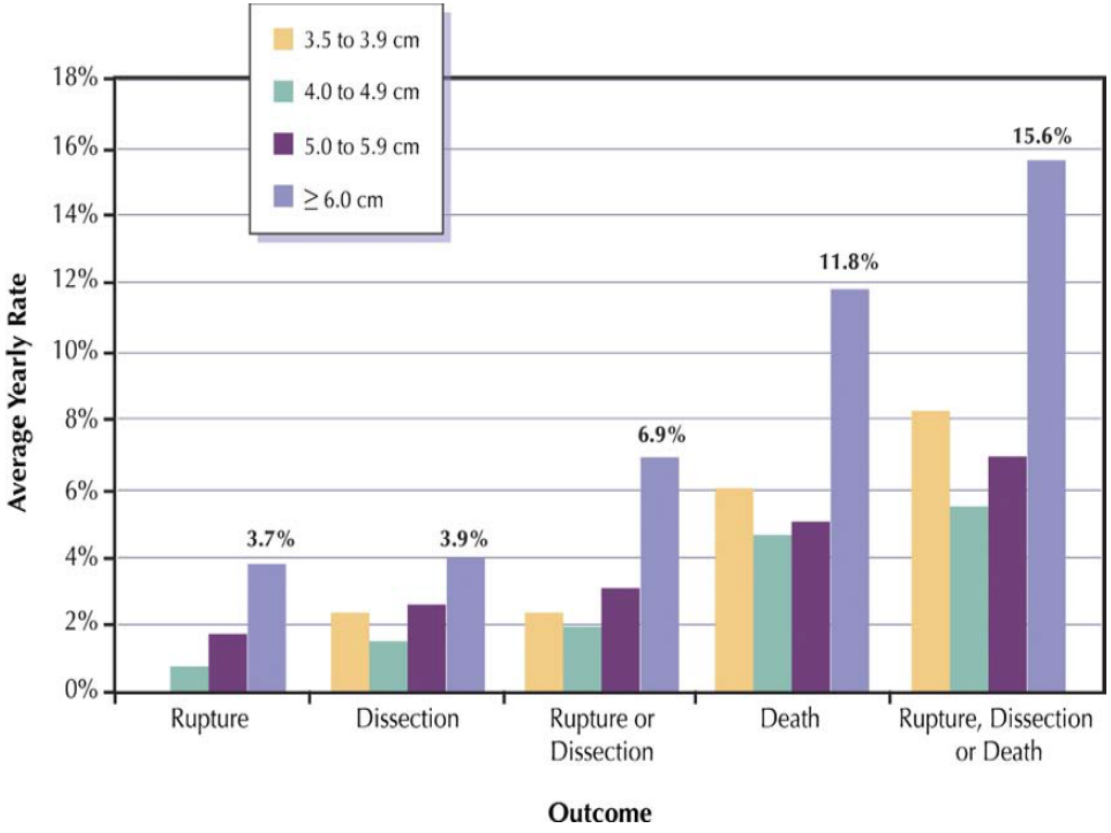
Ann Thorac Surg 2006;81:169-77



Depiction of "Hinge Points" for Lifetime Natural History Complications at Various Sizes of the Aorta

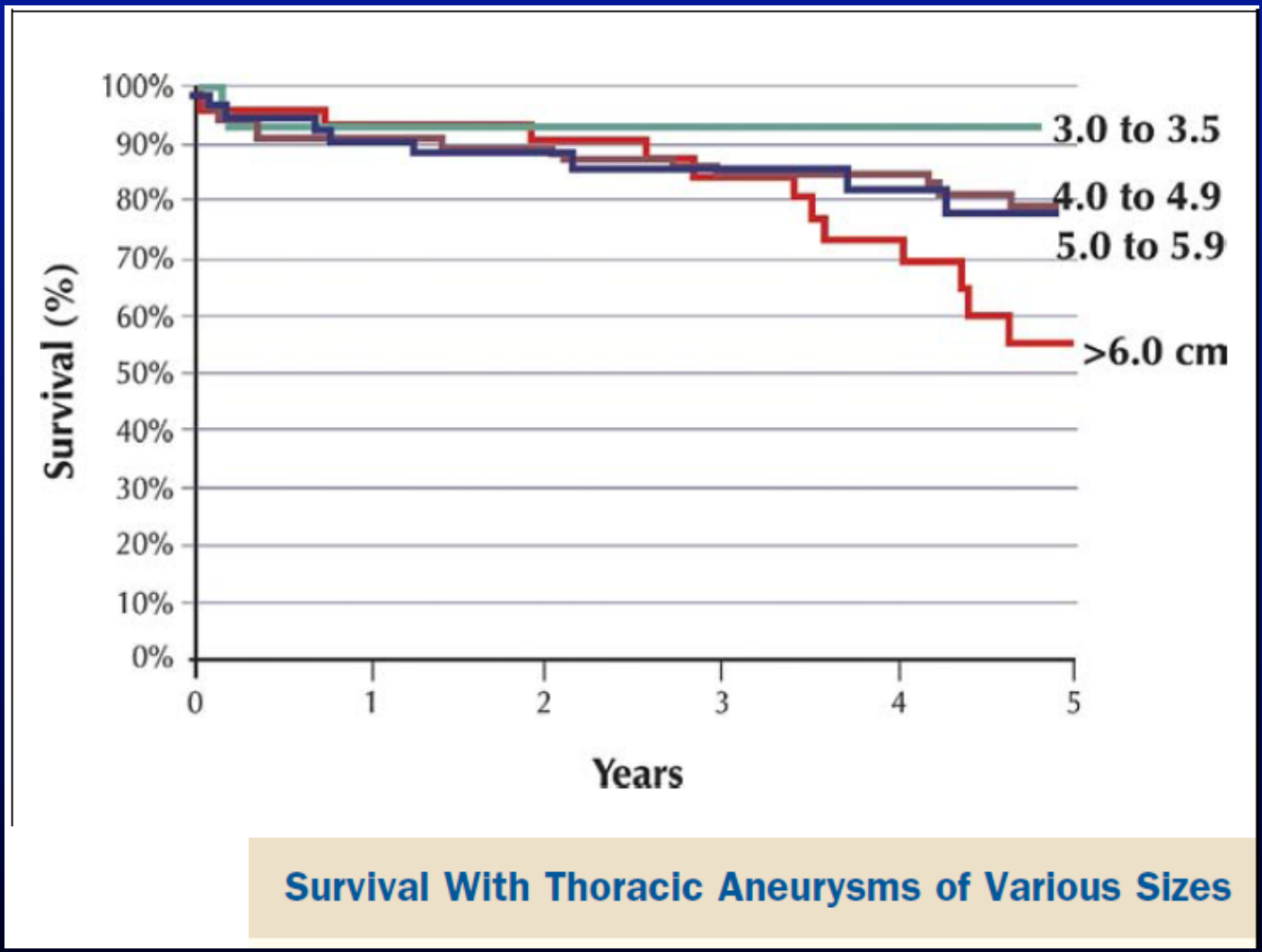
An individual with thoracic aortic aneurysm incurs a 34% lifetime risk of rupture or dissection by the time that his or her ascending aorta reaches a diameter of 6 cm

Elefteriades and Farkas – Thoracic aortic aneurysm 2010



Yearly Rates of Rupture, Dissection, or Death Related to Aortic Size

Elefteriades and Farkas – Thoracic aortic aneurysm 2010



Survival With Thoracic Aneurysms of Various Sizes

Elefteriades and Farkas – Thoracic aortic aneurysm 2010

L’aneurisma toracico è una patologia a prognosi potenzialmente infausta

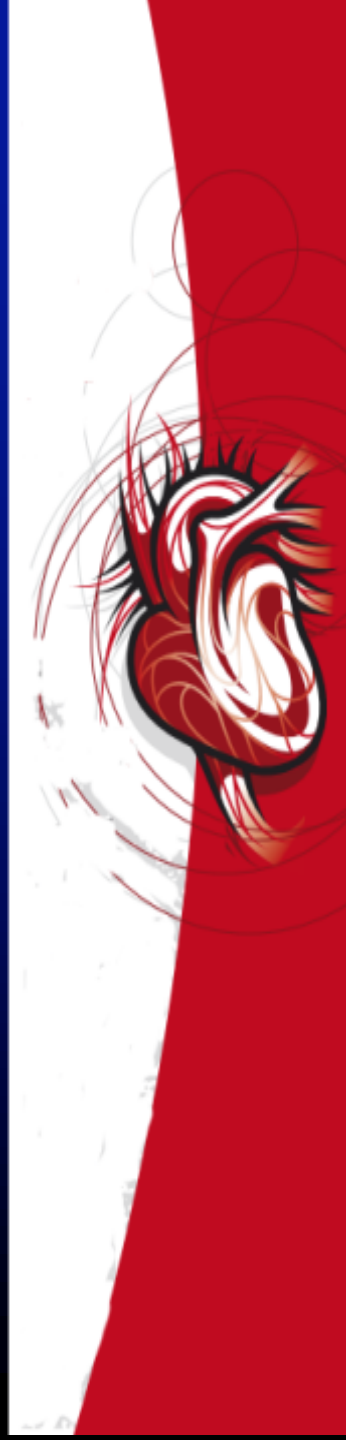
Gli aneurismi di dimensioni > 6 cm:

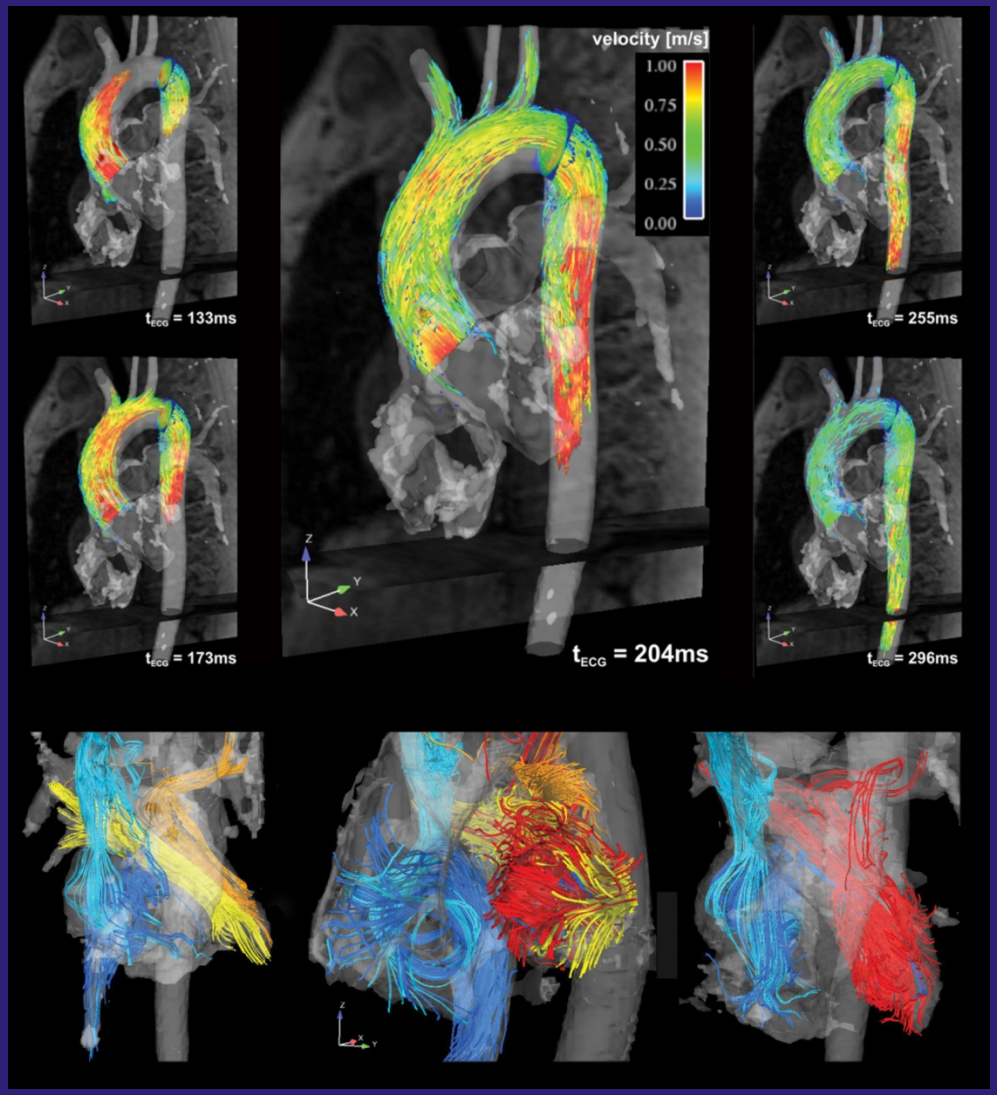
- rischio di rottura o dissezione 6.9% anno
- rischio cumulativo di morte, rottura o dissezione 15.6%/anno.

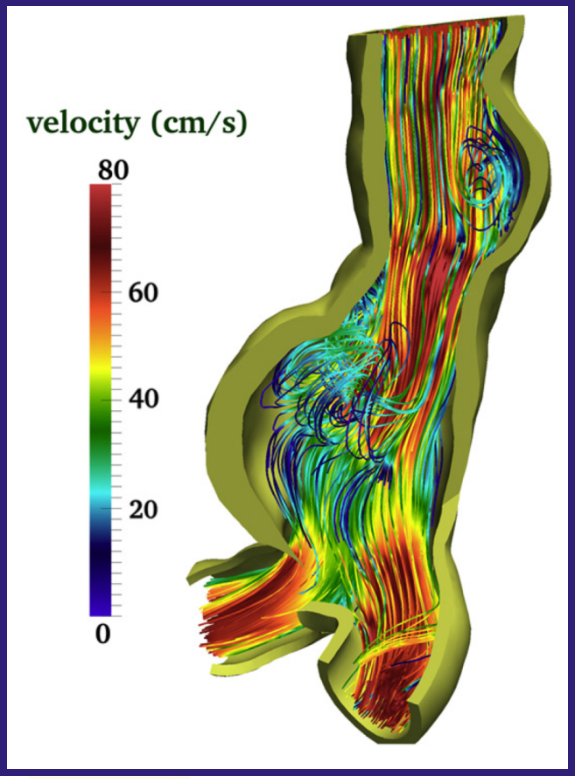
I sintomi si verificano quando insorgono le complicanze (dissezione/rottura) o quando tali complicanze sono imminenti

L’intervento in emergenza è gravato da un rischio operatorio molto più elevato

Elefteriades JA et Farkas 2010 by the American College of Cardiology Foundation

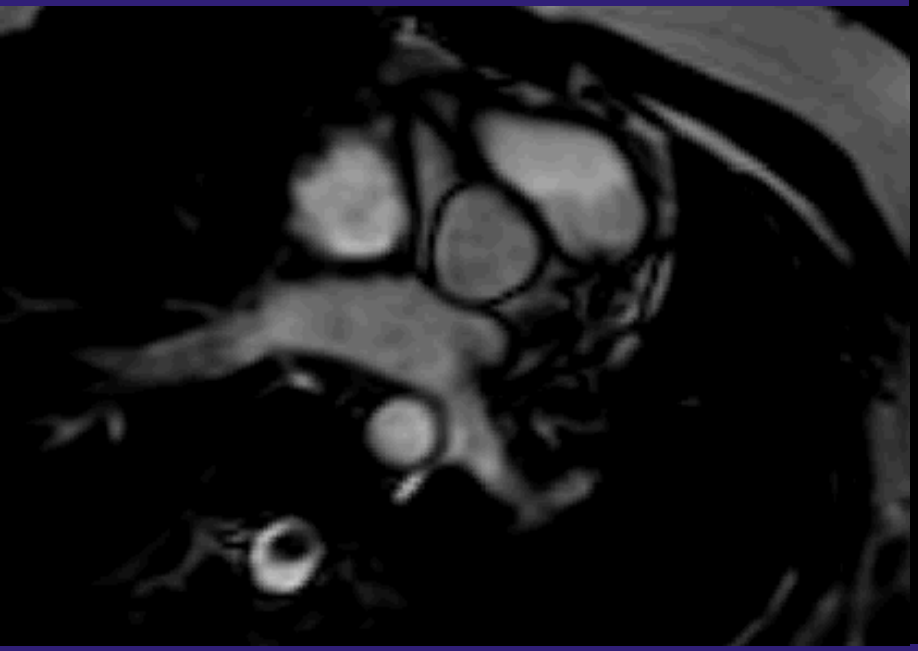


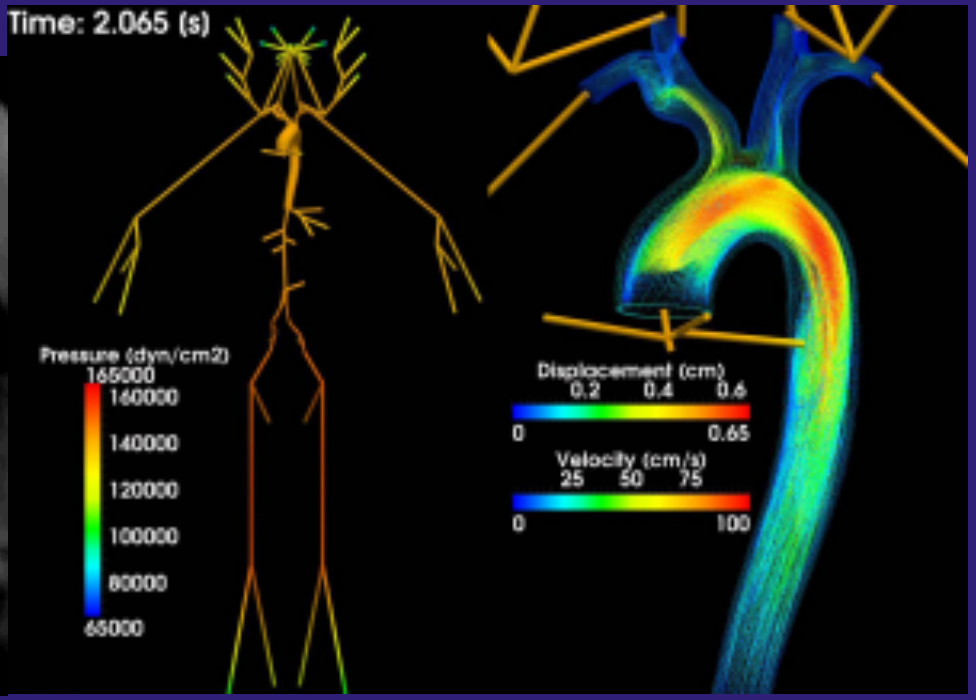




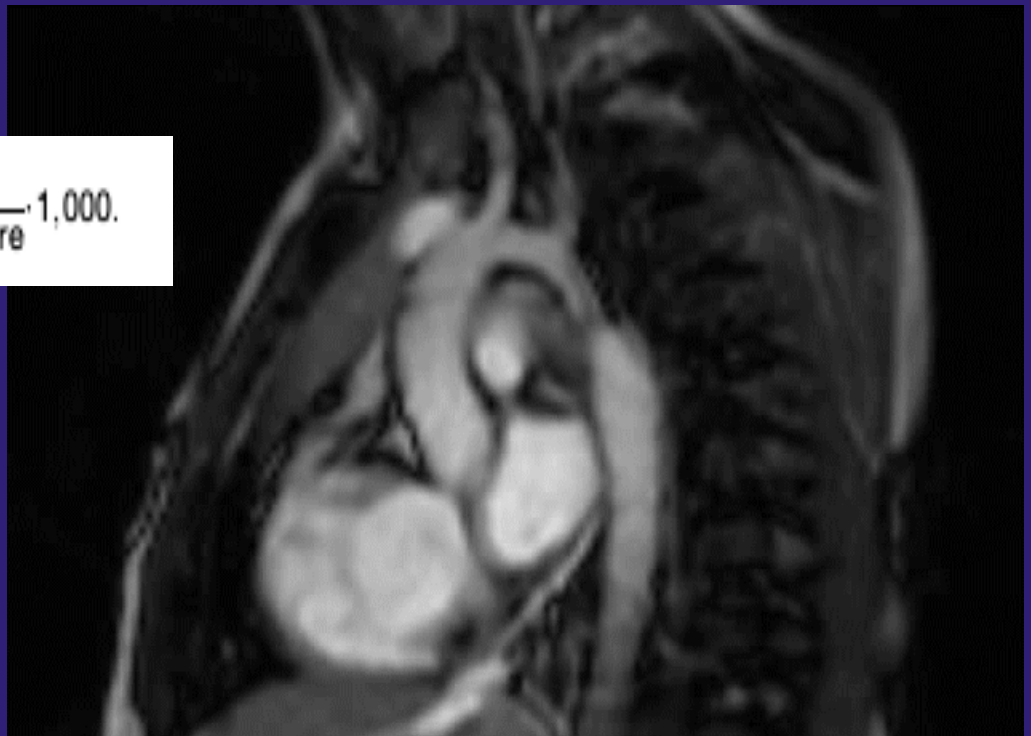
Reported normal values for invasively determined PWV measurements in middle-aged humans are 4.4 ± 0.4 m/sec in the aortic root, 5.3 ± 0.2 m/sec in the proximal descending thoracic aorta, 5.7 ± 0.4 m/sec in the distal thoracic descending aorta, 5.7 ± 0.4 m/sec in the suprarenal abdominal aorta, and 9.2 ± 0.5 m/sec in the infrarenal aorta.[32]

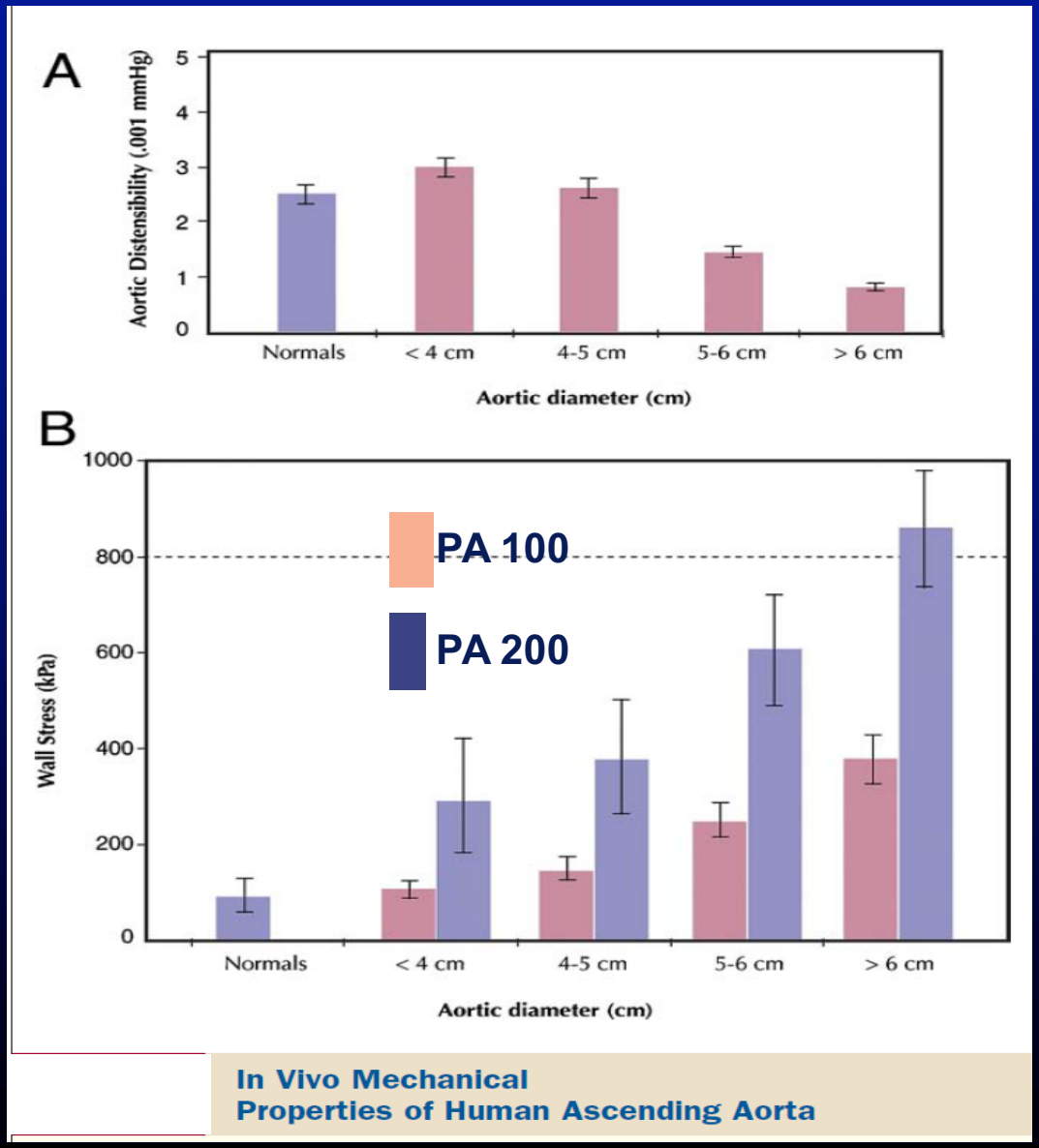
Immagini di risonanza magnetica cardiovascolare 4D che mappano la velocità e la direzione del flusso ematico durante il battito cardiaco. In alto, i colori delle linee visualizzano le velocità del flusso ematico in cinque diversi istanti della sistole in un'aorta toracica. Sotto, i colori indicano le diverse origini del flusso. Le immagini sono tratte da Comprehensive 4D velocity mapping of the heart and great vessels by cardiovascular magnetic resonance, pubblicato da Michael Markl e collaboratori su "Journal of Cardiovascular Magnetic Resonance" (2011;13:7).





$$\text{Distensibility}\left(10^{-3}\cdot\text{mm Hg}^{-1}\right) = \frac{\text{Area}_{\text{systole}} - \text{Area}_{\text{diastole}}}{\text{Area}_{\text{diastole}}\cdot\text{Pulse pressure}}\cdot 1{,}000.$$

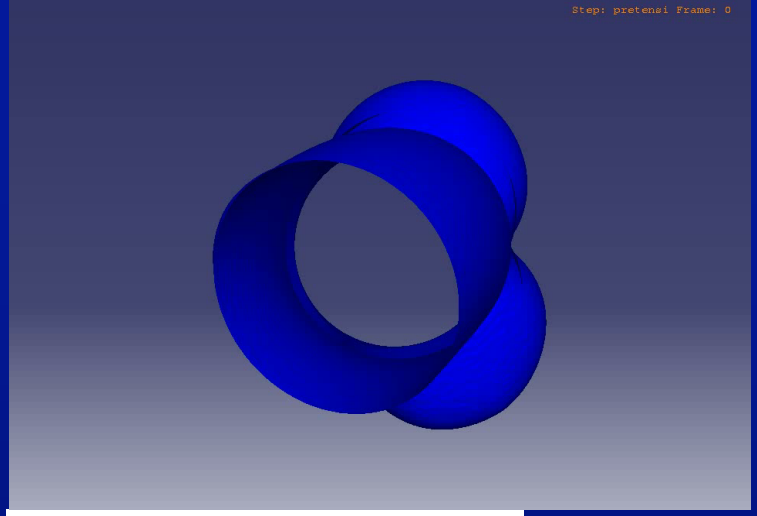


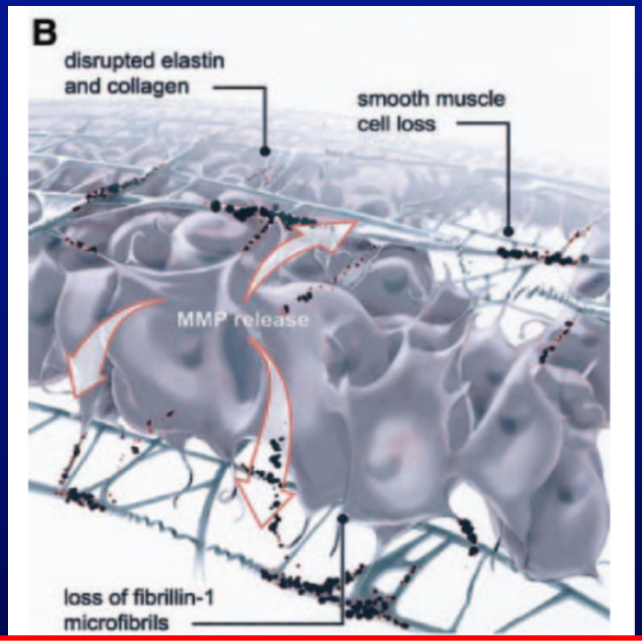


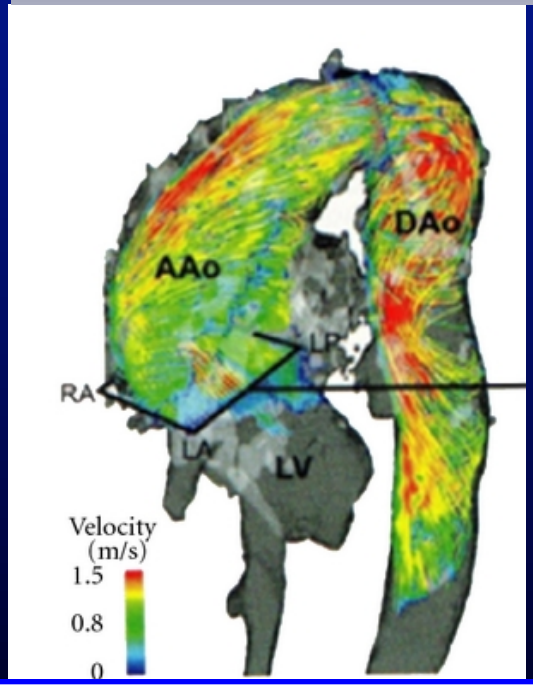
In Vivo Mechanical Properties of Human Ascending Aorta

Debated questions:

- haemodinamic factors on a genetic base?







Circulation

JOURNAL OF THE AMERICAN HEART ASSOCIATION

American Heart Association.

Ascending Aortic Dilatation Associated With Bicuspid Aortic Valve : Pathophysiology, Molecular Biology, and Clinical Implications

Thomas M. Tadros, Michael D. Klein and Oz M. Shapira

ELSEVIER

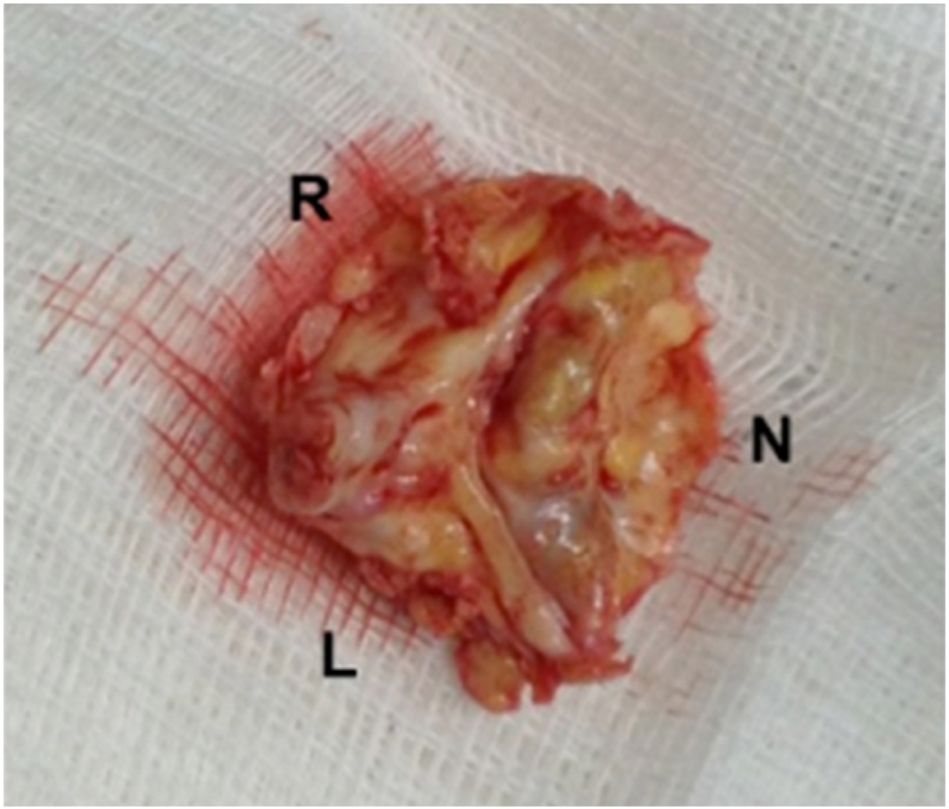
European Journal of Cardio-thoracic Surgery 39 (2011) 805–806

EUROPEAN JOURNAL OF CARDIO-THORACIC SURGERY

www.elsevier.com/locate/ejcts

Editorial

The role of hemodynamics in bicuspid aortic valve disease☆



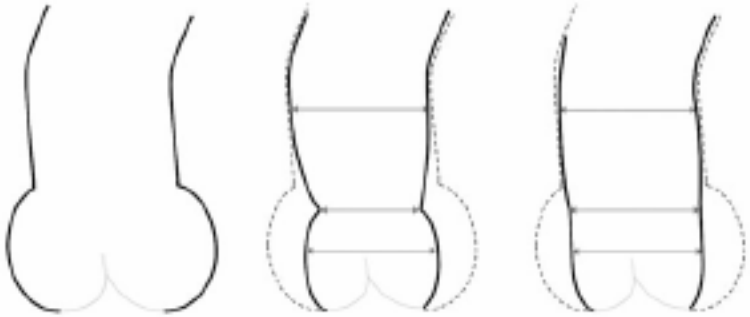
Photograph of surgically excised bicuspid aortic valve, demonstrating severe calcific stenosis. The left (L) and right (R) cusps are fused with a prominent calcified raphe, opposed to a calcified noncoronary cusp (N).

aortic stenosis is the most common presentation affecting 75% patients undergoing surgery for BAV disease, while insufficiency is the reason for intervention in only 13–16% of BAV patients

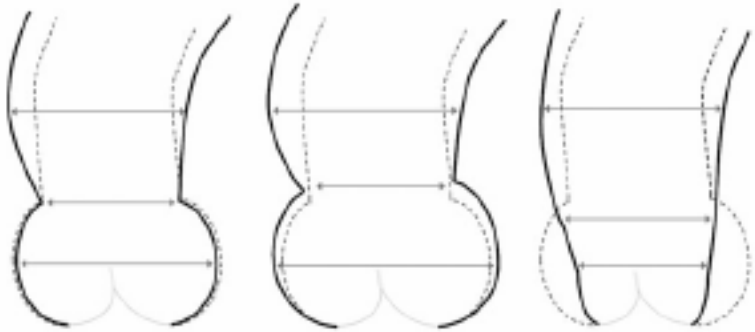
Bicuspid Aortic Valve Disease and Ascending Aortic Aneurysms: Gaps in Knowledge

Cardiology Research and Practice Volume 2012, Article ID 145202

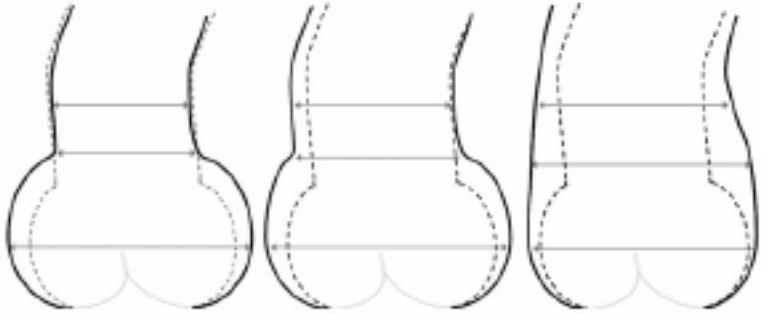
Schaefer's class	N shape	A shape	E shape
Park's class	normal	normal	normal
Della Corte's class	Non-dilated	Non-dilated	Non-dilated



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



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



N shape - ascending < sinuses > ST

A shape - ascending > sinuses > ST

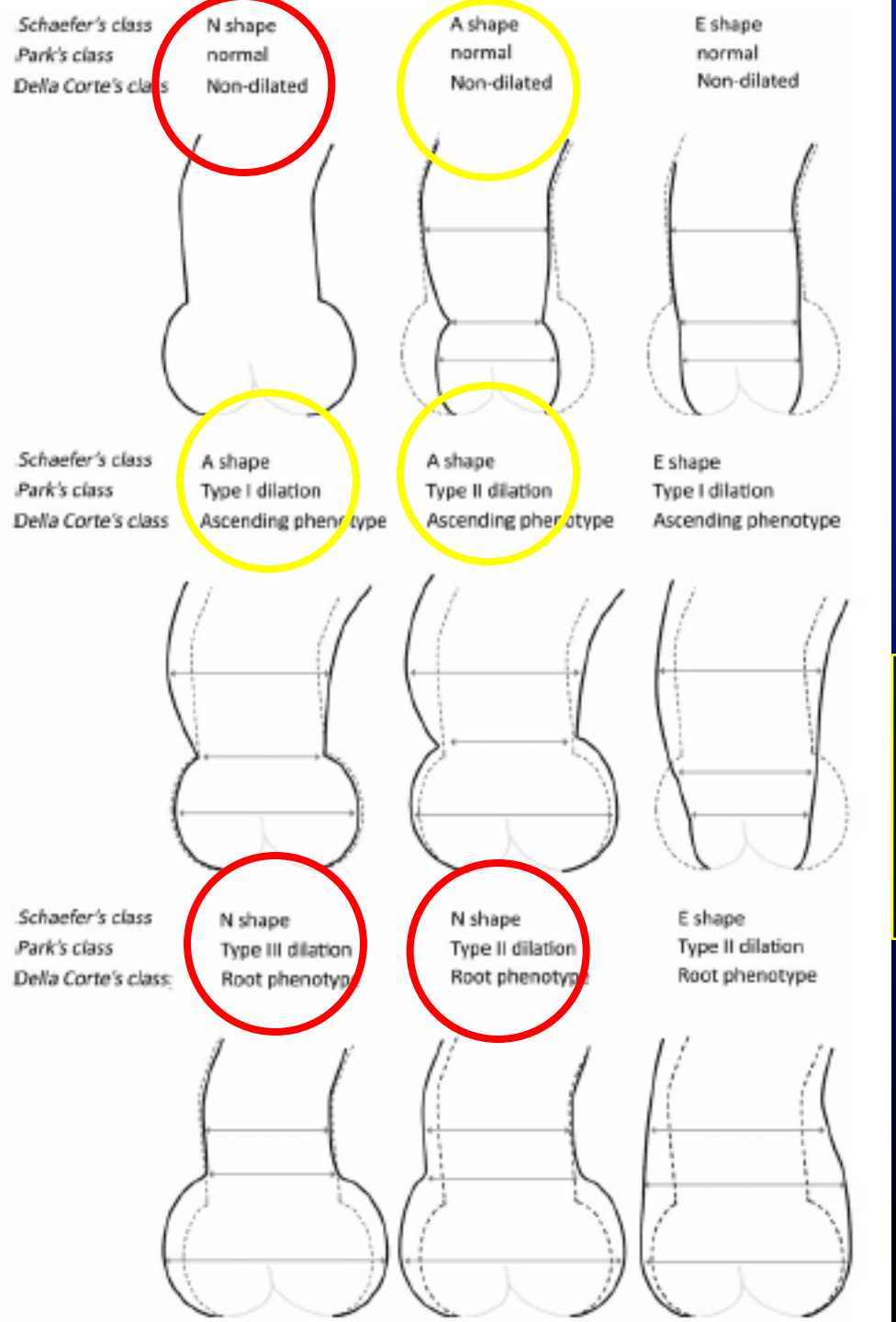
E shape - sinuse < ST

Type I - dilatation located at the ascending trant only

Type II – involving both tre ascending and the root

Type III – confined to the root

European Journal of Cardio-Thoracic Surgery 46 (2014) 240–247
doi:10.1093/ejcts/ezt621 Advance Access publication 14 January 2014



Fusion L-R coronary cusps, most commonly presented with Type N root anatomy

Fusion NCC - R coronary cusps, most commonly presented with Type A root anatomy

European Journal of Cardio-Thoracic Surgery 46 (2014) 240–247
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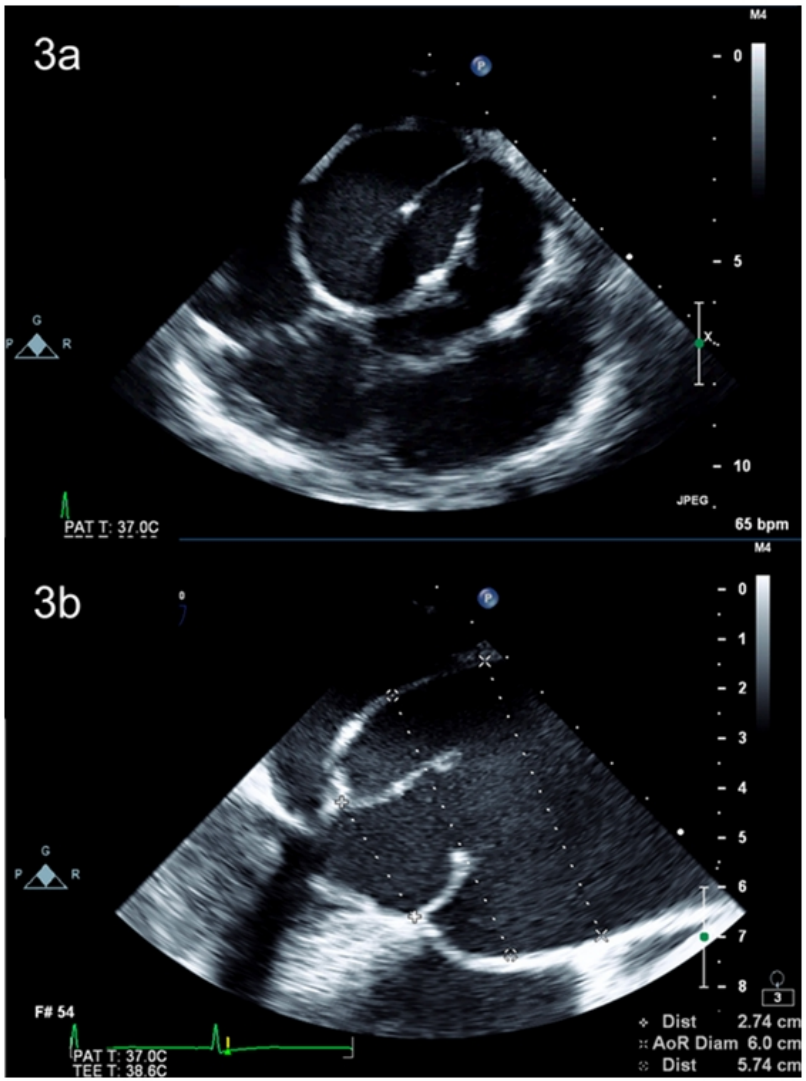


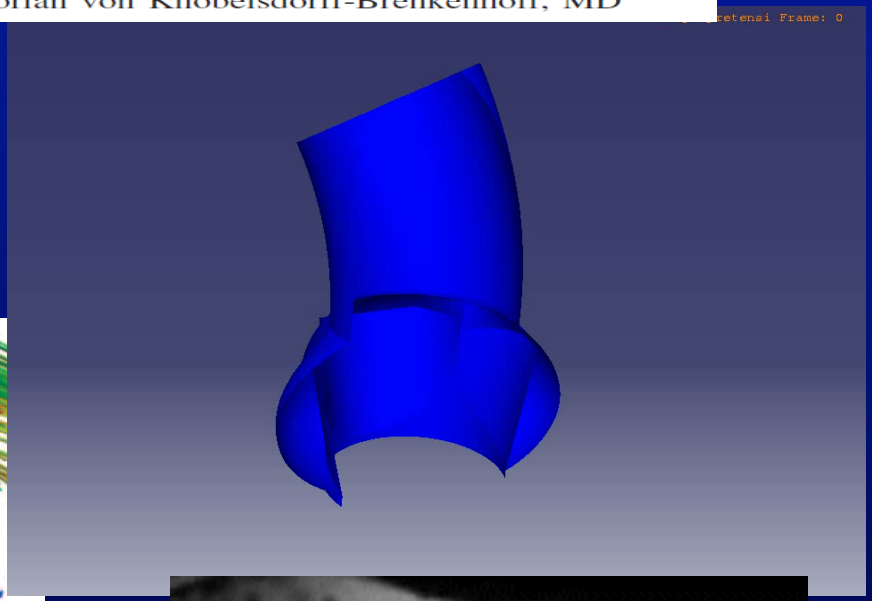
(a) Transesophageal echocardiogram demonstrating a bicuspid aortic valve in short-axis view, with left-right coronary cuspal fusion (Sievers' classification type 1, L-R, insufficient), and moderate-severe aortic insufficiency on colour flow Doppler (not shown).

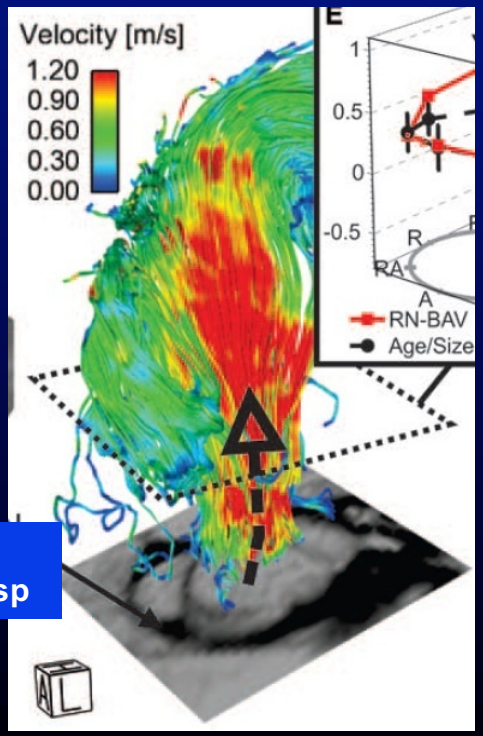
(b) Transesophageal echocardiogram demonstrating a dilated aortic root and ascending aorta in long-axis view along with a bicuspid aortic valve.

Original Article

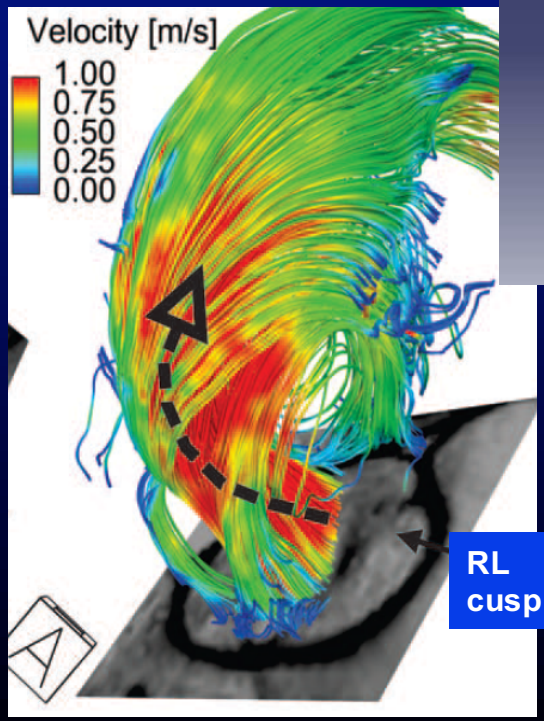
Bicuspid Aortic Valve Is Associated With Altered Wall Shear Stress in the Ascending Aorta

Alex J. Barker, PhD; Michael Markl, PhD; Jonas Bürk, MD; Ramona Lorenz, MS; Jelena Bock, MS; Simon Bauer, PhD; Jeanette Schulz-Menger, MD; Florian von Knobelsdorff-Brenkenhoff, MD

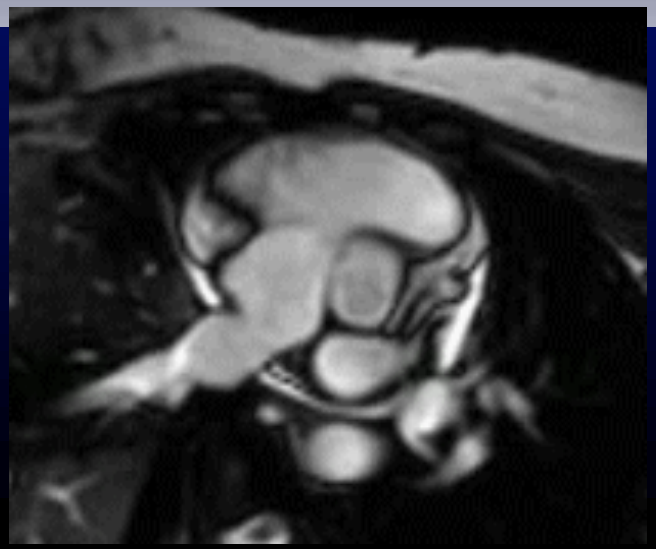




RN cusp

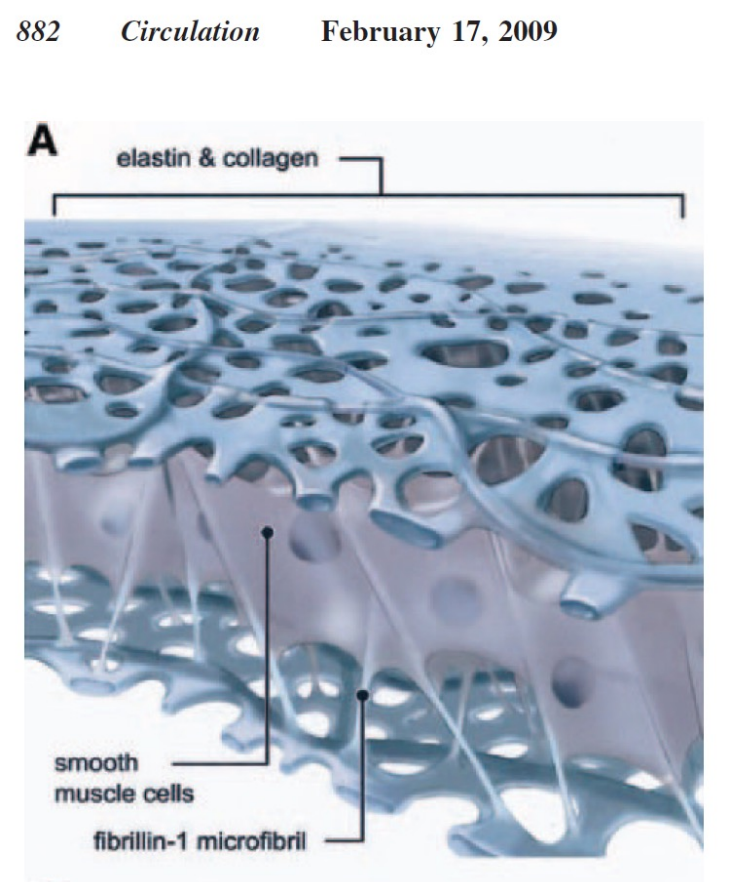


RL cusp



Indications for aortic surgery in BAV patients: a debated question

- Debated questions (?):
  - Genetic Aortic Wall Abnormality?



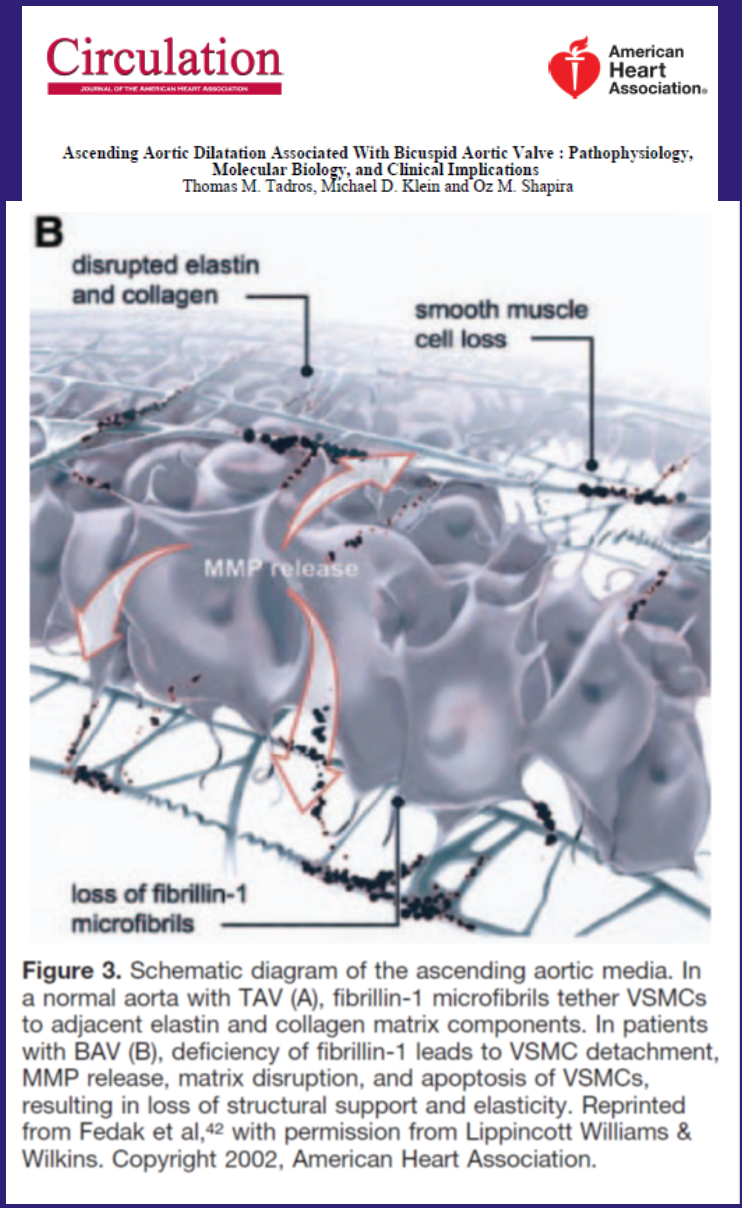
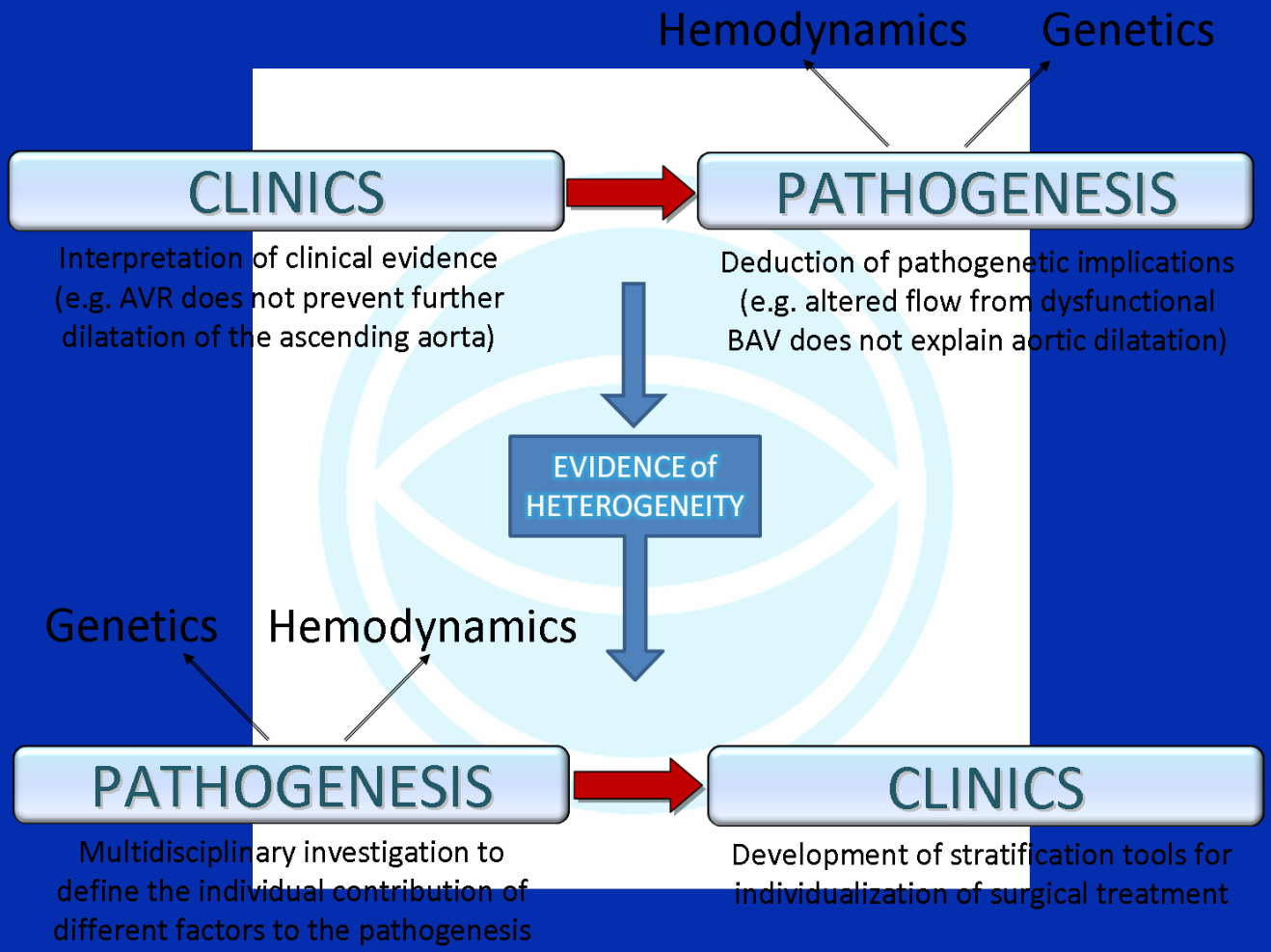


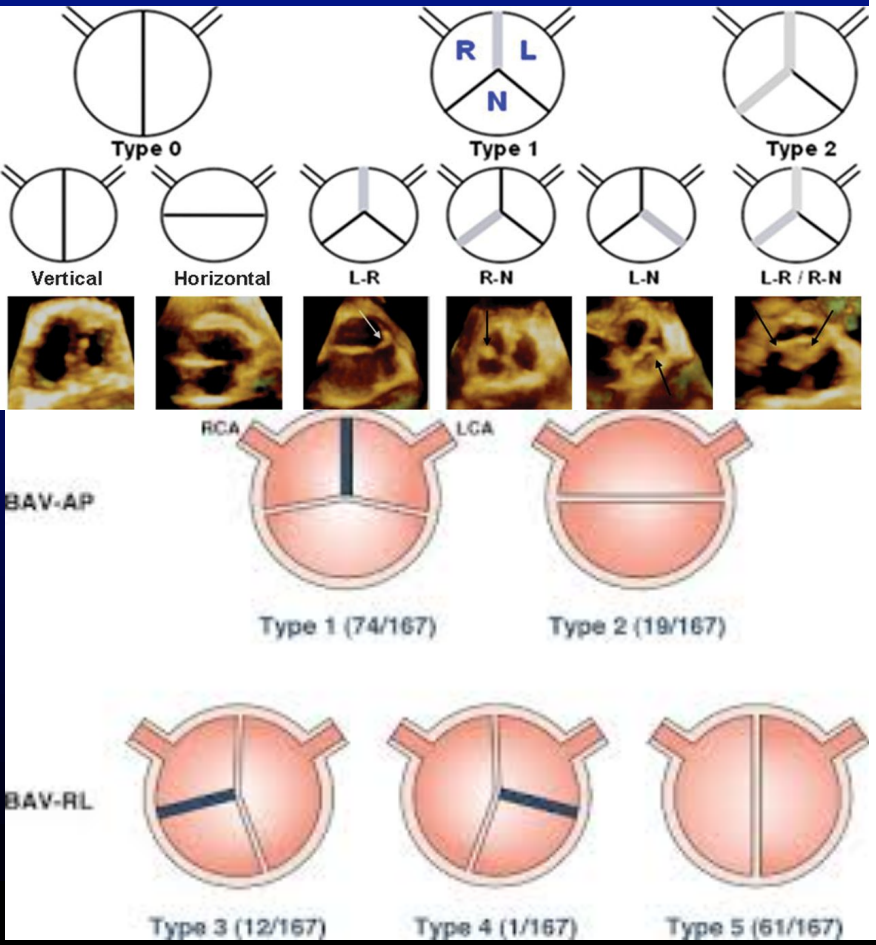
Figure 3. Schematic diagram of the ascending aortic media. In a normal aorta with TAV (A), fibrillin-1 microfibrils tether VSMCs to adjacent elastin and collagen matrix components. In patients with BAV (B), deficiency of fibrillin-1 leads to VSMC detachment, MMP release, matrix disruption, and apoptosis of VSMCs, resulting in loss of structural support and elasticity. Reprinted from Fedak et al,[42] with permission from Lippincott Williams & Wilkins. Copyright 2002, American Heart Association.

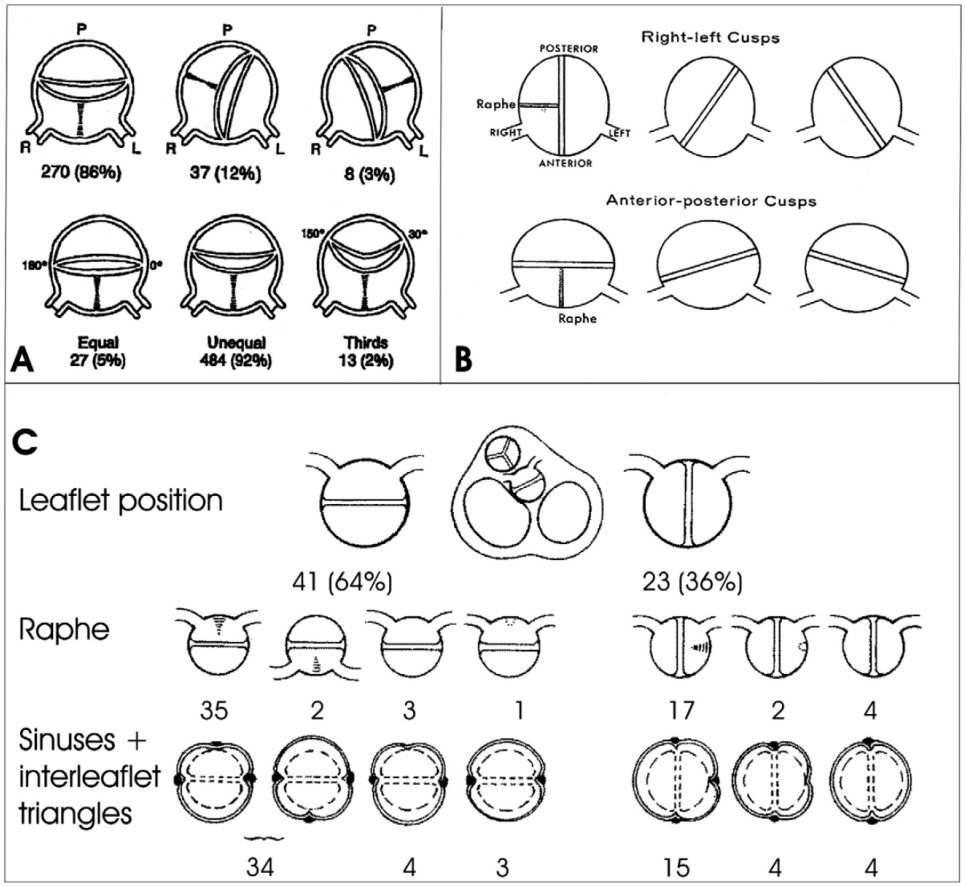

Hemodynamics
Genetics
CLINICS
PATHOGENESIS
Interpretation of clinical evidence (e.g. AVR does not prevent further dilatation of the ascending aorta)
Deduction of pathogenetic implications (e.g. altered flow from dysfunctional BAV does not explain aortic dilatation)
EVIDENCE of HETEROGENEITY
Genetics
Hemodynamics
PATHOGENESIS
CLINICS
Multidisciplinary investigation to define the individual contribution of different factors to the pathogenesis
Development of stratification tools for individualization of surgical treatment

Indications for aortic surgery in BAV patients: a debated question

Debated questions (?):

One Pathology/1000 Pathologies?





Debated questions (?):

Sievers Classification and prognosis

One Pathology/1000 Pathologies?
Phenotypes and Outcomes

Sievers and Schmidtke

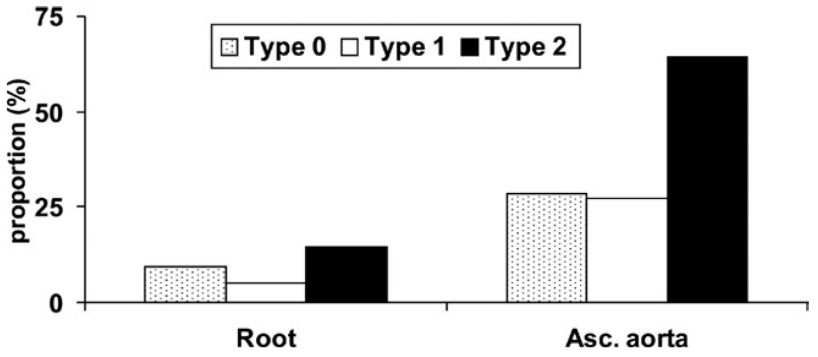
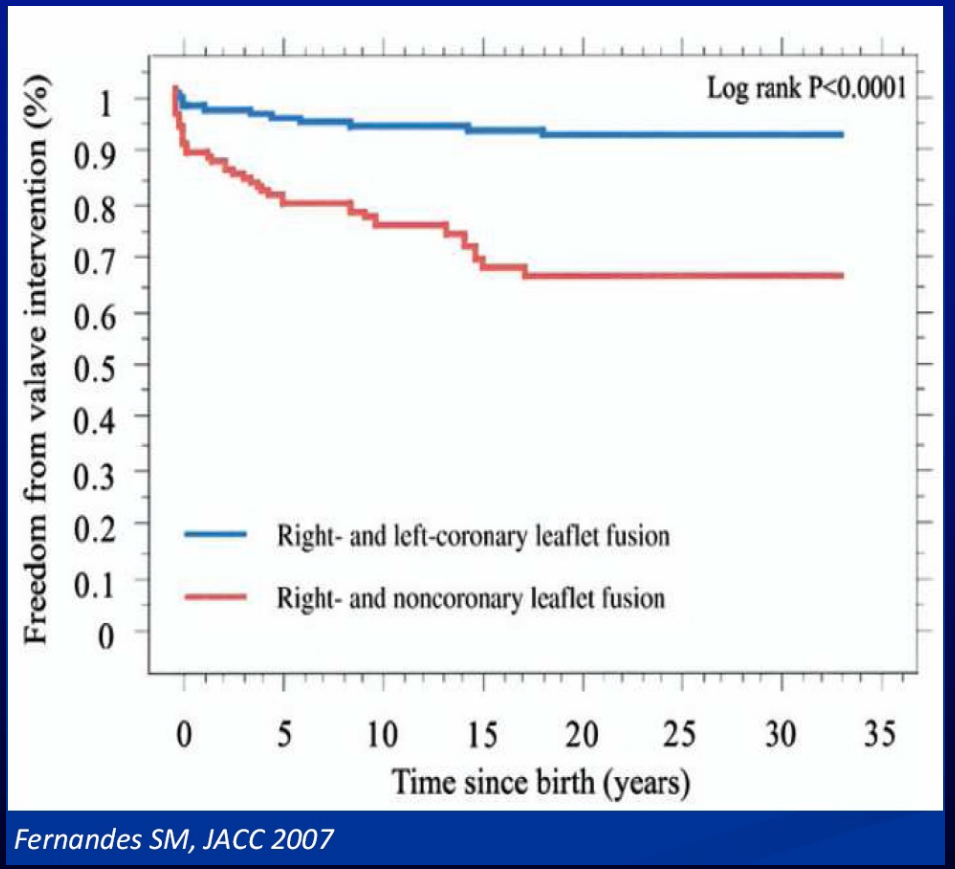
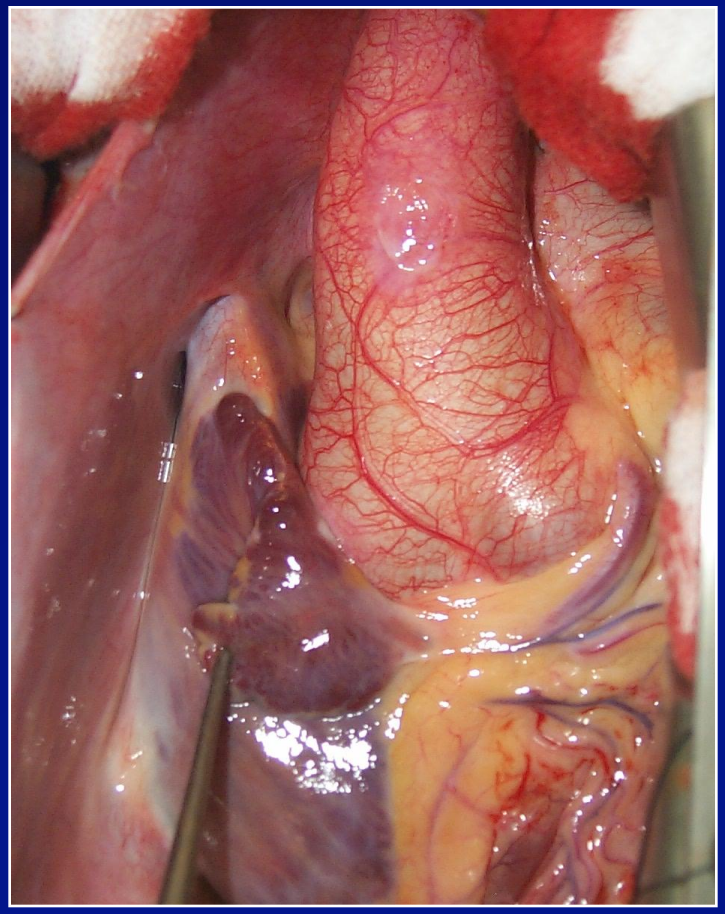


Figure 7. Proportion of an aneurysm of the aortic root or ascending aorta in relation to the type of bicuspid aortic valve. A bicuspid aortic valve type 2 (valve with two raphes) was associated with a significantly (P = .022) higher proportion of aneurysms.

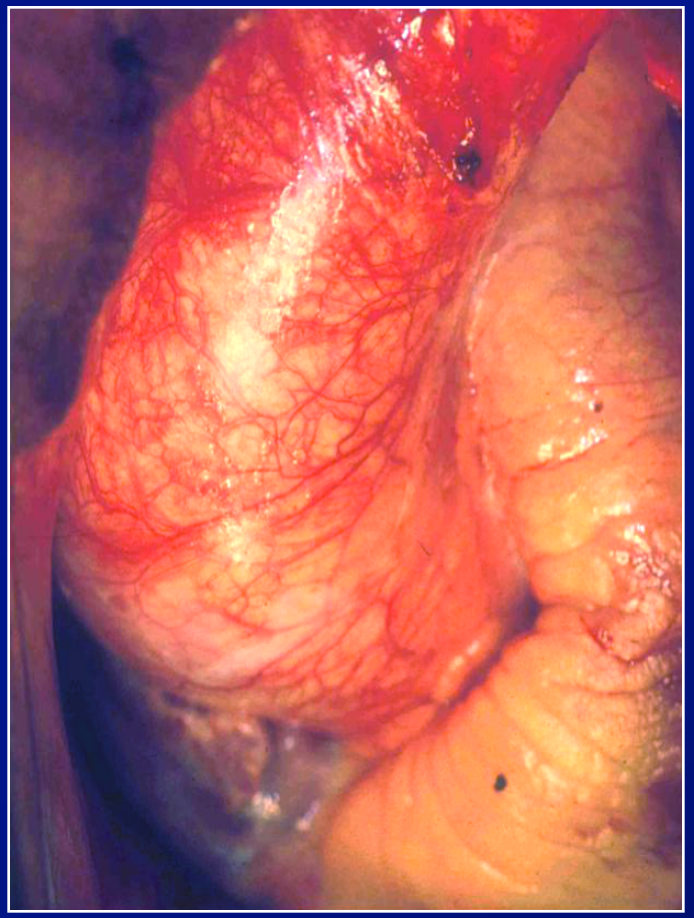


Fernandes SM, JACC 2007

Localizzazione della malattia

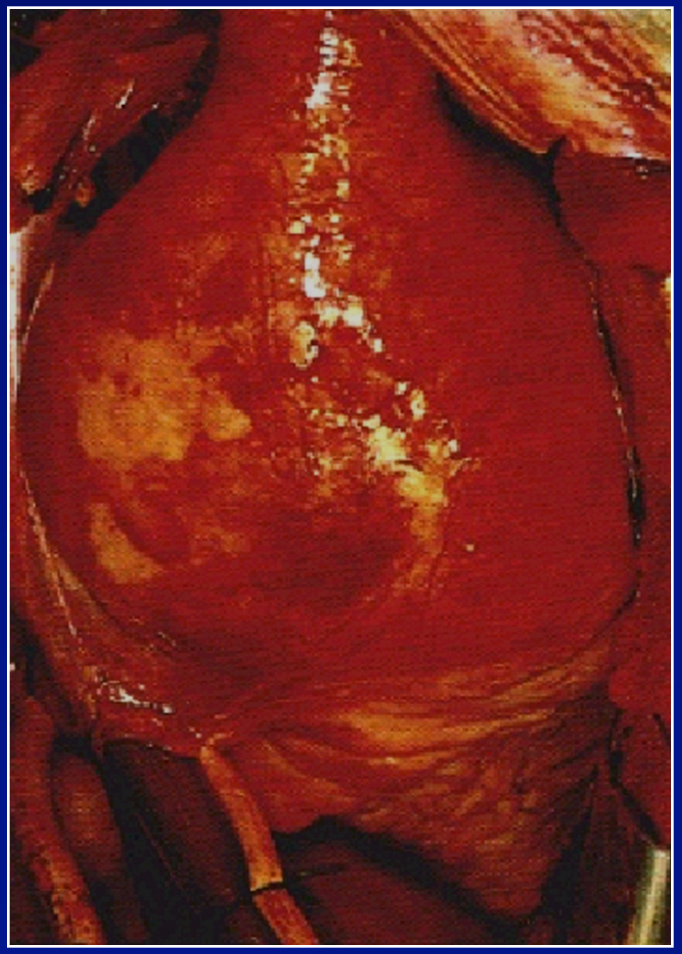


Dilatazione tratto sinusale

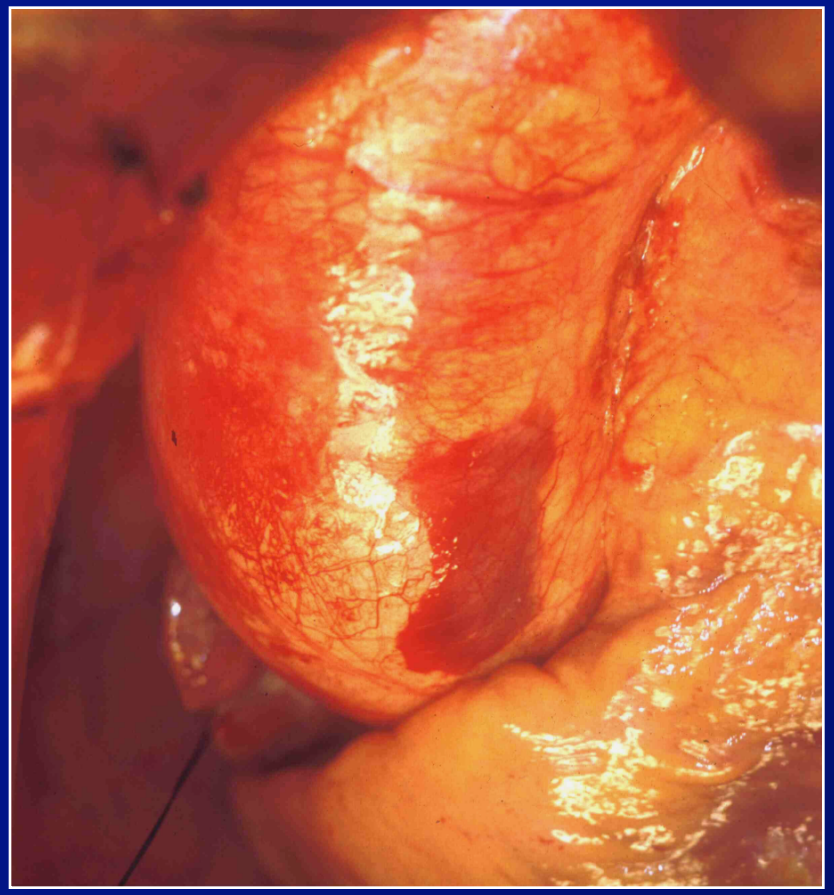


Dilatazione tratto tubulare

Localizzazione della malattia



Aneurisma “a fiasco”



Aneurisma “fusiforme”

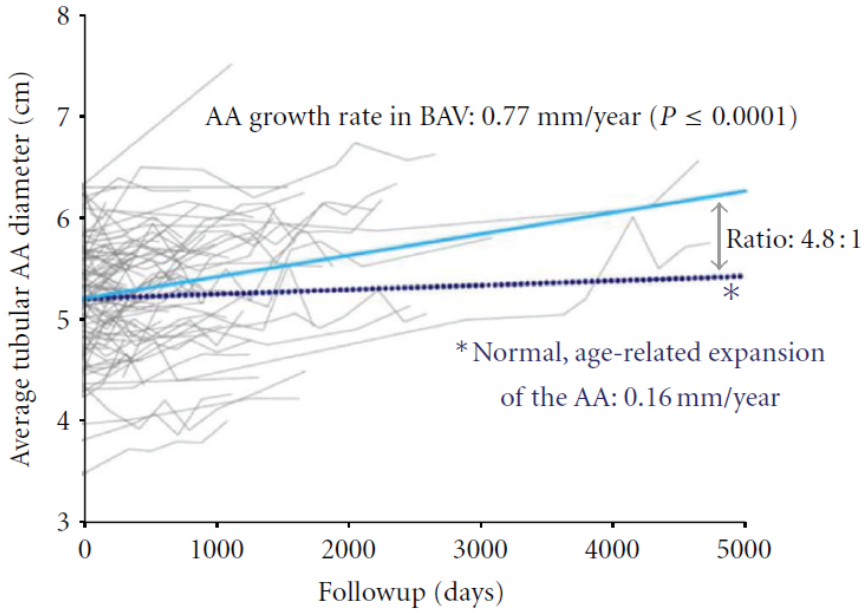


FIGURE 3: From Etz et al. [8]. Average growth of the ascending aorta in patients ($n = 116$) with normally functioning bicuspid aortic valve versus normal, age-related expansion. (Data for dotted line in this figure are derived from Hannuksela et al. [14].

TIMING CHIRURGICO

Rischio operatorio

Rischio di rottura/dissezione

ACC-AHA 2010

9.2.2.1.1. Recommendations for Asymptomatic Patients With Ascending Aortic Aneurysm

CLASS I

1. **Asymptomatic patients with degenerative thoracic aneurysm, chronic aortic dissection, intramural hematoma, penetrating atherosclerotic ulcer, mycotic aneurysm, or pseudoaneurysm, who are otherwise suitable candidates and for whom the ascending aorta or aortic sinus diameter is 5.5 cm or greater, should be evaluated for surgical repair. (371) (*Level of Evidence: C*)**
2. **Patients with Marfan syndrome or other genetically mediated disorders (vascular Ehlers-Danlos syndrome, Turner syndrome, bicuspid aortic valve, or familial thoracic aortic aneurysm and dissection) should undergo elective operation at smaller diameters (4.0 to 5.0 cm depending on the condition; see Section 5) to avoid acute dissection or rupture. (81,114,143,371, 436–439) (*Level of Evidence: C*)**

Recommendations for asymptomatic patients with Ascending Aortic Aneurysm

Class I

3. Patients with a growth rate of more than 0.5 cm/y in an aorta that is less than 5.5 cm in diameter should be considered for operation. (*Level of Evidence: C*)
4. Patients undergoing aortic valve repair or replacement and who have an ascending aorta or aortic root of greater than 4.5 cm should be considered for concomitant repair of the aortic root or replacement of the ascending aorta. (*Level of Evidence: C*)

ACC-AHA 2010

2014 ESC Guidelines on the diagnosis and treatment of aortic diseases

Recommendations on interventions on ascending aortic aneurysms

Recommendations	Class[a]	Level[b]
Surgery is indicated in patients who have aortic root aneurysm, with maximal aortic diameter[c] ≥50 mm for patients with Marfan syndrome.	I	C
Surgery should be considered in patients who have aortic root aneurysm, with maximal ascending aortic diameters: • ≥45 mm for patients with Marfan syndrome with risk factors.[d] • ≥50 mm for patients with bicuspid valve with risk factors.[e,f] • ≥55 mm for other patients with no elastopathy.[g,h]	IIa	C
Lower thresholds for intervention may be considered according to body surface area in patients of small stature or in the case of rapid progression, aortic valve regurgitation, planned pregnancy, and patient's preference.	IIb	C

American guidelines recommend prophylactic surgery for patients with Marfan syndrome

1 When the aorta reaches a diameter of 5.0 cm.
2 When the aorta reaches a diameter of 4.5 cm with either a positive family history of complications.
3 When the patient is undergoing aortic valve replacement, if the aorta exceeds 4.5 cm.
4 Associated significant aortic valve regurgitation, if the aorta exceeds 4.5 cm.
5 When the annual rate growth exceeds 0.5 cm.
Women with childbearing potential (see section on pregnancy).

Recommendations for the management of aortic root dilation in patients with bicuspid aortic valve

Recommendations	Class[a]	Level[b]
Patients with known BAV should undergo an initial TTE to assess the diameters of the aortic root and ascending aorta.	I	C
Cardiac MRI or CT is indicated in patients with BAV when the morphology of the aortic root and the ascending aorta cannot be accurately assessed by TTE.	I	C
Serial measurement of the aortic root and ascending aorta is indicated in every patient with BAV, with an interval depending on aortic size, increase in size and family history	I	C
In the case of a diameter of the aortic root or the ascending aorta >45 mm or an increase >3 mm/year measured by echocardiography, annual measurement of aortic diameter is indicated.	I	C
In the case of aortic diameter >50 mm or an increase >3 mm/year measured by echocardiography, confirmation of the measurement is indicated, using another imaging modality (CT or MRI).	I	C
In cases of BAV, surgery of the ascending aorta is indicated in case of: • aortic root or ascending aortic diameter >55 mm. • aortic root or ascending aortic diameter >50 mm in the presence of other risk factors.[c] • aortic root or ascending aortic diameter >45 mm when surgical aortic valve replacement is scheduled.	I	C

Various subtypes of BAV are associated with different forms of aortic dilation.[462] In patients with an LCC–RCC type BAV, ascending aorta dilation is common, but aortic root dilation is also seen.[463] In the RCC–NCC type, the aortic root is rarely affected and only dilation of the ascending aorta is seen.[313] Aortic dilation is maximal at the level of the tubular aorta, with a mean rate of 0.5 mm/year, similar to that seen in Marfan patients.[316]

Rapid progression of >5 mm/year and larger diameters are associated with increased risk of AD or rupture, with a sharp increase of risk at a diameter >60 mm. A higher gradient across a stenotic BAV and more severe aortic regurgitation (higher stroke volume) are reported to be associated with faster increase in aortic dimensions.[477] In the absence of stenosis or regurgitation, severe dilation also can occur, especially in young adults.[478,479]

2014 ESC Guidelines on the diagnosis and treatment of aortic diseases

Criteria for elective replacement of the ascending aorta in patients with BAV

Table 3: Criteria for elective replacement of the ascending aorta in patients with BAV.

AHA/ACC guidelines
Class I
(1) Aortic diameter >5.0 cm
(Level of evidence: B)
(2) Aneurysm growth rate >0.5 cm/year
(Level of evidence: B)
(3) Aortic diameter >4.5 cm with concomitant indication for elective aortic valve repair/replacement
(Level of evidence: B)
Aortic size ratios and indexes
Aortic diameters >4.5 cm and either of the following:
(1) Ratio of aortic area to body height >10 cm^2/m in asymptomatic patients with well-functioning BAV, or 8-9 cm^2/m in symptomatic patients [91]
(2) Ratio of aortic diameter to body surface area >4.5 cm/m^2 [92]
Other criteria (unvalidated)
Aortic diameters >4.5 cm and any of the following:
(1) Aortic coarctation, corrected or uncorrected [93]
(2) First-degree relative with ascending aortic dissection or rupture
(3) Long smoking history, especially with COPD [94, 95]

Bicuspid Aortic Valve Disease and Ascending Aortic Aneurysms: Gaps in Knowledge

Cardiology Research and Practice
Volume 2012, Article ID 145202, 16 pages

Aortic Valve and Ascending Aorta Guidelines

Class IIa

For patients with Loeys-Dietz syndrome, a threshold of 4.2 cm maybe considered for surgery.
(Level of evidence C)

The cross-sectional area of the root in square centimeters divided by the patient's height in meters and exceeding 10 may be considered an indication for surgery.
(Level of evidence C)

In female patients with a connective tissue disorder who are considering pregnancy, a prophylactic repair may be considered when the aortic root exceeds 4.0 cm.
(Level of evidence C)

Separate valve and ascending aortic replacement are recommended for patients without significant aortic root dilation, for elderly patients, or for young patients with minimal dilation in whom a biological valve is being inserted or a bicuspid valve is being repaired. (Level of evidence B)

Ann Thorac Surg 2013;95:1491–505

CONCLUSIONI

Eziopatogenesi

Fattori di rischio

Linee guida

Le patologie concomitanti

Evoluzione della malattia

IL CHIRURGO

GRAZIE