Simposio: **LE MALATTIE CRONICHE DELL'AORTA TORACICA.**
Tra anatomia,diagnostica, timing operatorio, tecniche riparative e follow-up: non sempre è agevole orientarsi in una patologia nella quale le indicazioni della letteratura vengono spesso superate e messe in discussione nella pratica di tutti i giorni

ECOCARDIOCHIRURGIA

VI CONGRESSO NAZIONALE
Milano – Atahotel Executive 14-16 marzo 2012

**Milano 15 ottobre 2012**

# La dilatazione isolata dell'aorta ascendente con e senza valvulopatia aortica associata.

## Storia naturale, timing chirurgico e tecnica operatoria

**Claudio Grossi**
**Antonio Capo**

Dipartimento Cardiovascolare
Osp. Santa Croce e Carle - Cuneo

# Fattori di rischio per dissezione aortica

**Conditions associated with increased aortic wall stress**

- Hypertension, particularly if uncontrolled
- Pheochromocytoma
- Cocaine or other stimulant use
- Weight lifting or other Valsalva maneuver
- Trauma
- Deceleration or torsional injury (eg, motor vehicle crash, fall)
- Coarctation of the aorta

**Conditions associated with aortic media abnormalities**

- Inflammatory vasculitides
  - Takayasu arteritis
  - Giant cell arteritis
  - Behçet arteritis
- Other
  - Pregnancy
  - Polycystic kidney disease
  - Chronic corticosteroid or immunosuppression agent administration
- Infections involving the aortic wall either from bacteremia or extension of adjacent infection

**L' indicazione chirurgica nasce dalla provata capacità di prevenire la dissezione aortica**

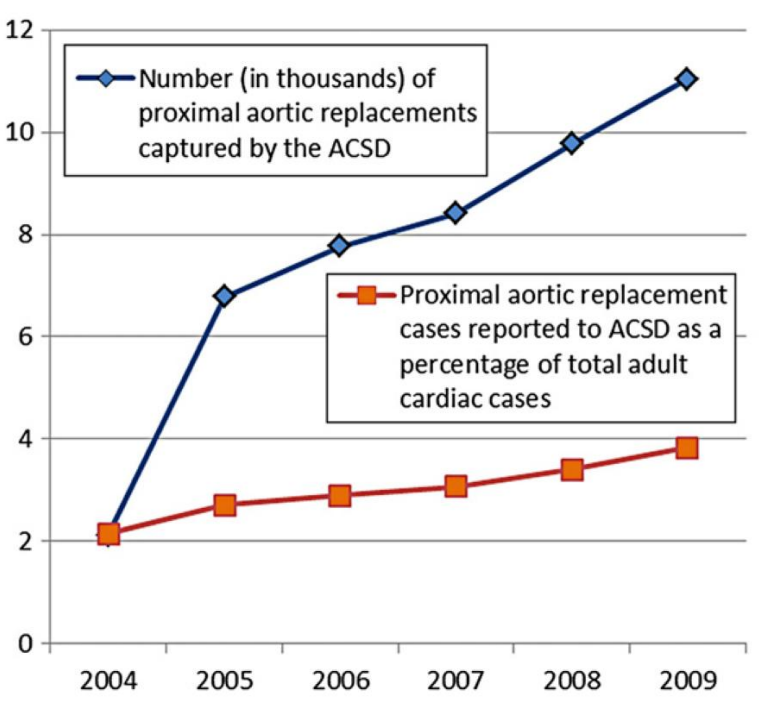
## Contemporary Results for Proximal Aortic Replacement in North America

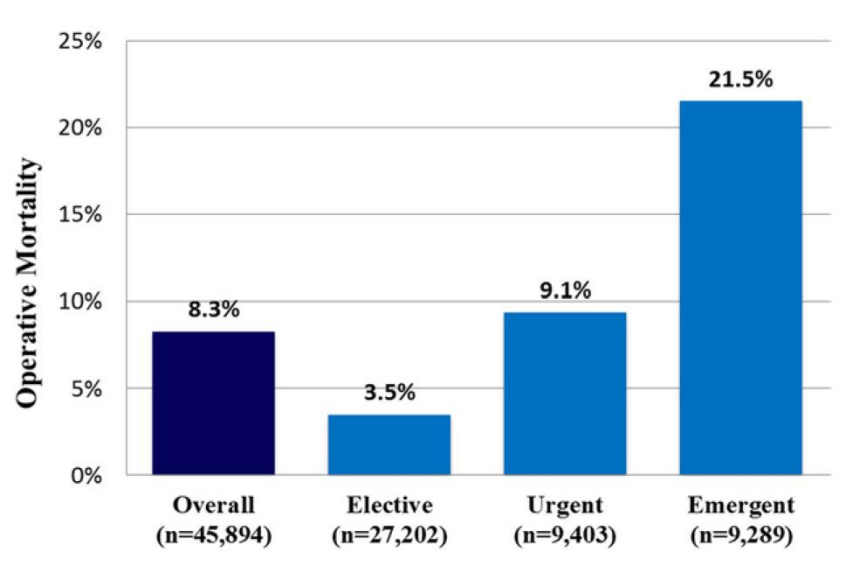
(J Am Coll Cardiol 2012;60:1156–62)

Judson B. Williams, MD, MHS,*† Eric D. Peterson, MD, MPH,*‡ Yue Zhao, PHD,*
Sean M. O'Brien, PHD,* Nicholas D. Andersen, MD,† D. Craig Miller, MD,§ Edward P. Chen, MD,‖
G. Chad Hughes, MD†

*Durham, North Carolina; Stanford, California; and Atlanta, Georgia*

- *Society of Thoracic Surgeons Database* per pazienti operati sostituzione aorta ascendente (+/- radice +/- arco) dal 2004 al 2009



# Chirurgia aorta ascendente

***Cuneo: maggio '00 – febbraio'12***

## 463 PAZIENTI / DRG chir. totali (6,8 %)

**Elettivi** **384**

euroscore add. 7,4 / log 12,0 mortalità – 1,8%

**Urgenze** **15**

euroscore add. 7,5 / log 12,7 mortalità – 6,7%

**Emergenze** **64**

euroscore add. 9,2 / log 21,6 mortalità – 28,1%

## Come vanno le cose:

- **La parete aortica è un ambiente biologicamente attivo**.
- Nell'aorta **ascendente il contenuto in elastina è alto**, diminuisce procedendo verso l'aorta toracica ed addominale.
- **La frammentazione delle fibre elastiche è un normale processo dell'invecchiamento**, ma è accelerato in alcuni individui per motivi non chiari
- **Il diametro dell'aorta tipicamente aumenta con l'età.**

## Istopatologia Aneurisma aortico: *come si presenta?*

- L'aorta aneurismatica è caratterizzata da **alterazione e degradazione delle fibre elastiche con aumento di deposizione di proteoglicani**
- Lo sviluppo dell'aneurisma nell'aorta ascendente **non è associato ad un indebolimento meccanico ma ad un irrigidimento della parete**
- La recente letteratura evidenzia **il ruolo delle cellule dell'infiammazione** nel contesto della dilatazione
- La malattia **non è associata a necrosi**, il termine medionecrosi è confondente

- Come vanno le cose:

**Increased Tissue Microarray Matrix Metalloproteinase Expression Favors Proteolysis in Thoracic Aortic Aneurysms and Dissections**

George J. Koullias, MD, PhD, Pars Ravichandran, MD, Dimitris P. Korkolis, MD, PhD, David L. Rimm, MD, and John A. Elefteriades, MD

Section of Cardiothoracic Surgery, Department of Surgical Pathology, and Yale Microarray Facility, Yale New Haven Hospital, New Haven, Connecticut

(Ann Thorac Surg 2004;78:2106–11)

- **Matrix metalloproteinases (MMPs)** sono un gruppo di proteasi che giocano un importante ruolo nell'equilibrio lisi-sintesi proteica nella parete aortica
- L'aumento del valore di MMP esprime una modifica del metabolismo della parete aortica con un **aumento della proteolisi rispetto alla normalità**, questo gioca un ruolo fisiopatologico nella genesi dell'aneurisma

## Aterosclerosi

- In passato l'aterosclerosi era riportata essere il secondo più comune fattore causale della degenerazione aneurismatica nell'aorta ascendente
- Il ruolo preciso dell'aterosclerosi è controverso
- **Ci possono essere lesioni aterosclerotiche che però si ritiene siano fattore comitante e non causale verso l'aneurima**.

STATE-OF-THE-ART PAPER

# Thoracic Aortic Aneurysm

## Clinically Pertinent Controversies and Uncertainties

JACC Vol. 55, No. 9, 2010
March 2, 2010:841–57

John A. Elefteriades, MD,* Emily A. Farkas, MD†
*New Haven, Connecticut; and St. Louis, Missouri*

"Silver lining" della malattia aortica

- La malattia dell'aorta prossimale sembra avere un **effetto protettivo verso l'aterosclerosi**
- Il contenuto in **Calcio è significativamente più basso nei pazienti con aneurisma aorta ascendente** rispetto ai controlli per età e sesso
- **Un alto tasso di MMP potrebbe favorire una riduzione del carico aterosclerotico,** in parte come lisi delle placche ad opera delle proteasi,

# Genetica

European Journal of Cardio-thoracic Surgery 35 (2009) 931–940

Review

## Inherited diseases and syndromes leading to aortic aneurysms and dissections

Ahmet Okay Caglayan [a,*], Munis Dundar [b]

[a] Kayseri Education and Research Hospital, Department of Medical Genetics, 38010, Kayseri, Turkey
[b] Erciyes University, Medical Faculty, Department of Medical Genetics, Kayseri, Turkey

- Il 15% dei portatori di aneurisma aortico hanno un **parente di primo grado con vari gradi della stessa malattia**
- Perciò dopo il riscontro diagnostico è importante ottenere un anamnesi famigliare comprendente le morti improvvise ed eseguire **controlli sui parenti stretti**
- Le forme famigliari portano ad una diagnosi più precoce (56.8 a.) rispetto alle forme sporadiche (64.3 a.) ma più tardiva rispetto ai Marfan (24.8 a.)

Review

# Inherited diseases and syndromes leading to aortic aneurysms and dissections

Ahmet Okay Caglayan [a,*], Munis Dundar [b]

[a] Kayseri Education and Research Hospital, Department of Medical Genetics, 38010, Kayseri, Turkey
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European Journal of Cardio-thoracic Surgery 35 (2009) 931–940

- Genetica

- Geni responsabili fper **forme sindromiche** di aneurisma e dissezione aortica

| Syndrome (inheritance) | Gene symbol | Chromosome localization | Protein name | Gene function | Affected aortic segments |
|---|---|---|---|---|---|
| Marfan (autosomal dominant) | *FBN1* | 15q21.1 | Fibrillin-1 | *FBN1* encodes fibrillin-1, a large glycoprotein that is a component of extracellular matrix structures called microfibrils | Dilatation of the ascending aorta involving the sinuses of Valsalva; dissection of the ascending aorta |
| | *TGFβR2*[a] | 3p24–25 | Transforming growth factor-beta receptor type II | TGF signaling plays an important role in cellular proliferation, differentiation and extracellular matrix production | Predominantly ascending aortic disease; however, significant descending aortic disease and aneurysms of other vessels also occurred in affected family members |
| Ehlers–Danlos syndrome type IV (autosomal dominant) | *COL3A1* | 2q31 | Collagen alpha-1(III) chain | The *COL3A1* gene encodes the chains of type III procollagen, a major structural component of skin, blood vessels, and hollow organs | Proximal branches of the aortic arch, the descending thoracic aorta and the abdominal aorta. The distal branches of the aorta, especially the renal, mesenteric, iliac and femoral arteries, are also particularly affected [102] |
| Turner syndrome (chromosomal) | | 45X | | | Aortic root dilatation with or without dissection has been incidentally noted in 6%-9% of patients with Turner syndrome [103,104] |

Review

# Inherited diseases and syndromes leading to aortic aneurysms and dissections

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European Journal of Cardio-thoracic Surgery 35 (2009) 931–940

- Genetica

# 2

| | | | | | |
|---|---|---|---|---|---|
| Noonan syndrome (autosomal dominant) | *PTPN11* | 12q24.1 | Tyrosine-protein phosphatase non-receptor type 11 (SHP-2) | The protein is expressed throughout the body and it is an important player in cellular response to growth factors, hormones, cytokines and cell adhesion molecules | Coarctation of aorta |
| | *KRAS* | 12p12.1 | GTPase KRas | Their proteins regulate cell fates and they are key regulators of the RAS-RAF-MEK-ERK pathway, which is important for proliferation, growth and death of cells | |
| | *RAF1* | 3p25 | RAF proto-oncogene serine/threonine-protein kinase | | |
| | *SOS1* | 2p22–p21 | Son of sevenless homolog 1 | | |
| Osteogenesis imperfecta (autosomal dominant) | *COL1A1* | 17q21.3–q22 | Collagen alpha-1(I) chain | They encode the chains of type I procollagen, the major protein in bone and most other connective tissues | Ascending aorta |
| | *COL1A2* | 7q22.1 | Collagen alpha-2(I) chain | | |
| Homocystinuria (autosomal recessive) | *CBS* | 21q22.3 | Cystathionine beta-synthase | CBS is a pyridoxal 50-phosphate (PLP)–dependent enzyme and condenses homocysteine and serine to cystathionine, an irreversible step in transsulfuration | Abdominal aorta (especially elderly patients) |
| Autosomal dominant polycystic kidney disease (autosomal dominant) | *PKD1* | 16p13.3 | Polycystin 1 | Cell cycle regulation and intracellular calcium transport | Thoracic aortic aneurysms |
| | *PKD2* | 4q21–q22 | Polycystin 2 | Member of the family of voltage-activated calcium channels | |
| Pseudo xanthoma elasticum (autosomal recessive) | *ABCC6 (MRP6)* | 16p13.1 | ATP-binding cassette transporter C6 (multidrug resistance associated protein 6) | Cellular transport protein | Especially abdominal, sometimes arch and thoracic aortic aneurysms |
| Hurler syndrome (autosomal recessive) | IDUA | 4p16.3 | Alpha-L-iduronidase | Lysosomal degradation of glycosaminoglycans (heparan and dermatan sulphate) | Coarctation of aorta |
| Loeys–Dietz syndrome (autosomal dominant) | *TGFBR1* and *TGFBR2* | 9q33–q34 and 3p24–p25 | Transforming growth factor $\beta$ receptors 1 and 2 | TGF signaling plays an important role in cellular proliferation, differentiation and extracellular matrix production | Affected patients have a high risk of aortic dissection or rupture at an early age and at aortic diameters that ordinarily would not be predictive of these events |

Review

# Inherited diseases and syndromes leading to aortic aneurysms and dissections

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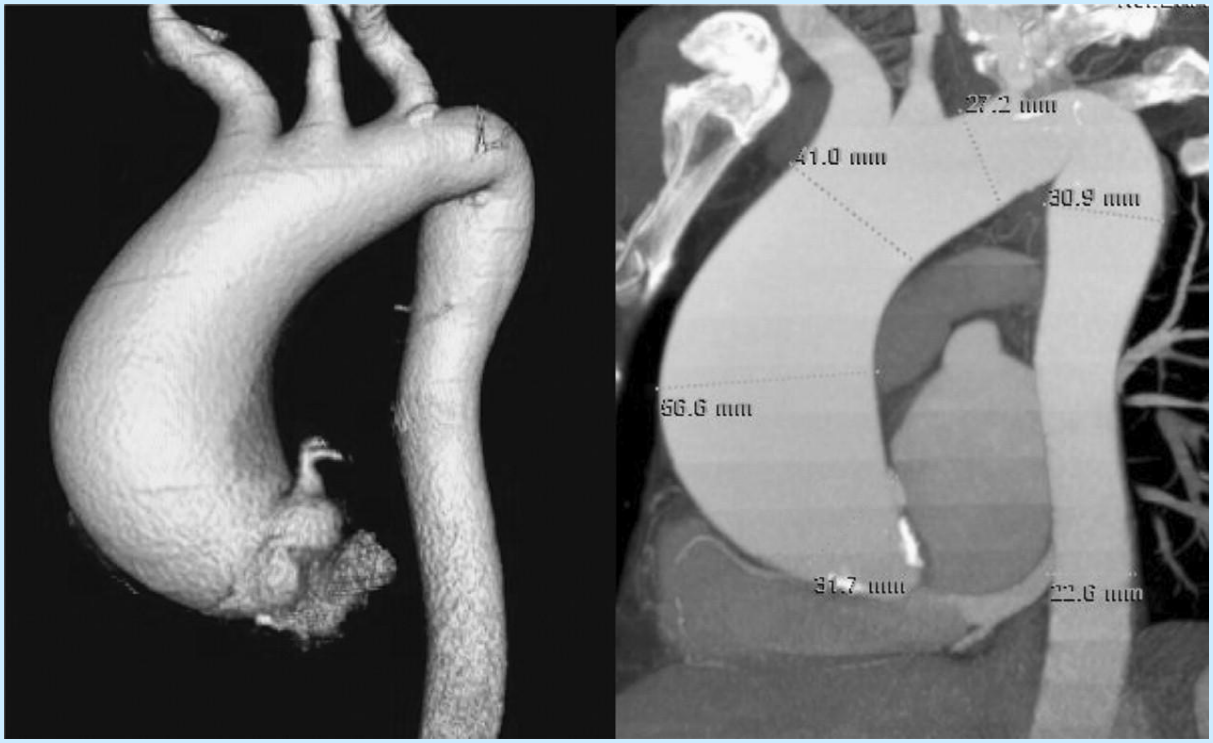
- Genetica

- Geni responsabili per forme **"non sindromiche" di** aneurismi e dissezione aortica

| | Locus name (gene cards) | Gene symbol | Chromosomal localization | Protein name |
|---|---|---|---|---|
| Affected genes | TAAD1 (AAT2) | Unknown | 5q13–14 | Unknown |
| | FAA1 (AAT1) | Unknown | 11q23.3–24 | Unknown |
| | TAAD2 (AAT3) | *TGFBR2* | 3p24–25 | TGF-beta receptor type-2 |
| | MYH11 | MYH11 | 16p13.13–p13.12 | Myosin-11 |
| Positional candidate genes | FBLN2 | FBLN2 | 3p25.1 | Fibulin-2 |
| | TIMP4 | TIMP4 | 3p25 | Tissue inhibitor of metalloproteinases 4 |
| Candidate genes | MMP3 | MMP3 | 11q22.3 | Matrix metalloprotease 3 |
| | COL1A1 | COL1A1 | 17q21.3–q22 | Collagen alpha-1(I) chain |
| | *COL1A2* | COL1A2 | 7q22.1 | Collagen alpha-2(I) chain |

Human MYH11 gene mutations provide the first example of a direct change in a specific SMC protein leading to an inherited arterial disease [92]. The *TAAD2* locus encompasses the *MFS2* locus, raising the possibility that these conditions are allelic. Data are compiled from the following standard references: gene symbol from HUGO; chromosomal locus, locus name, critical region, complementation group from OMIM; protein name from Swiss-Prot.

- Definizione




- **Aneurisma** (vero): dilatazione permanente localizzata di un'arteria, con aumeno dei diametri **di almeno 50%** ripetto ai valori normali.
- **Ectasia**: dilatazione di un'arteria **di meno del 50%** rispetto al normale.

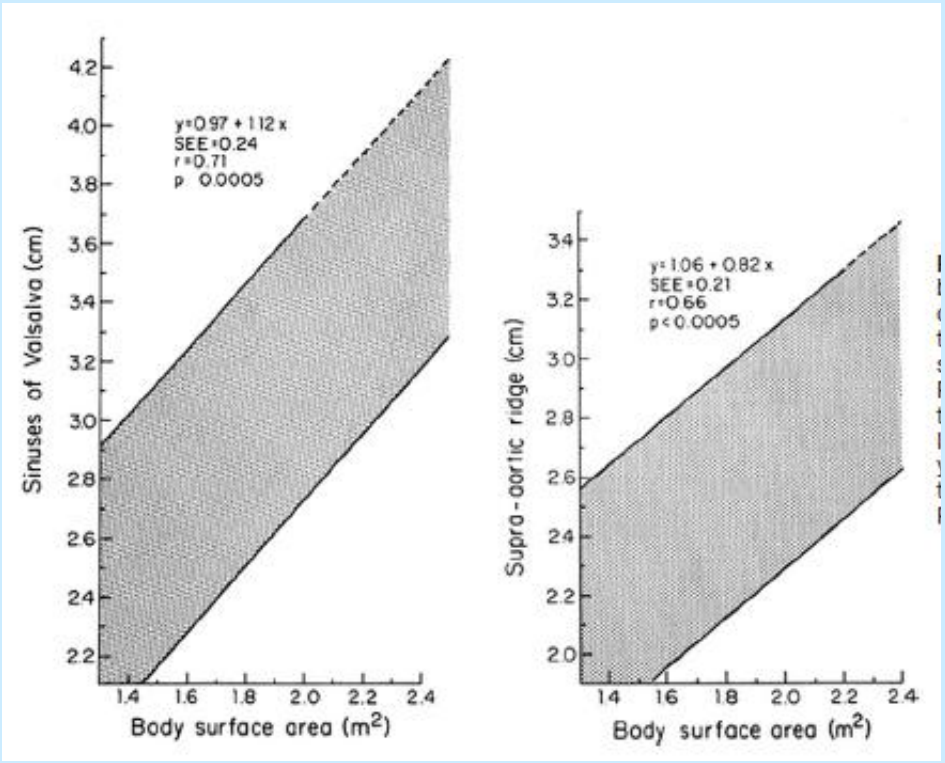
- Che cosa è "normale"?

Aortic Size Assessment by Noncontrast Cardiac Computed Tomography: Normal Limits by Age, Gender, and Body Surface Area

Arik Wolak, MD,* Heidi Gransar, MS,* Louise E. J. Thomson, MB ChB,*
John D. Friedman, MD, FACC,* Rory Hachamovitch, MD, FACC,* Ariel Gutstein, MD,*
Leslee J. Shaw, PhD, FACC,* Donna Polk, MD, MPH,* Nathan D. Wong, PhD,†
Rola Saouaf, MD,* Sean W. Hayes, MD,* Alan Rozanski, MD, FACC,*
Piotr J. Slomka, PhD,* Guido Germano, PhD, FACC,* Daniel S. Berman, MD, FACC*
*Los Angeles and Irvine, California*

JACC: CARDIOVASCULAR IMAGING, VOL. 1, NO. 2, 2008
MARCH 2008:200–9

- **Il diametro aortico correla meglio con la BSA che con il BMI**
- **Il diametro medio dell'aorta ascendente è 33 ± 4 mm**
- **Il limite superiore dei normali diametri aortici è 41 mm**



- Che cosa è “normale”?

STATE-OF-THE-ART PAPER

**Thoracic Aortic Aneurysm**

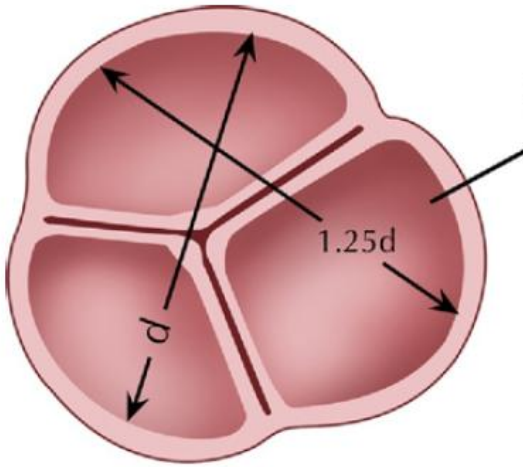
Clinically Pertinent Controversies and Uncertainties

JACC Vol. 55, No. 9, 2010
March 2, 2010:841–57

John A. Elefteriades, MD,* Emily A. Farkas, MD†
*New Haven, Connecticut; and St. Louis, Missouri*

- La sezione trasversa dell’aorta **non è un cerchio.**
- questo è vero anche in TEE di buona qualità
- **La dilatazione asimmetrica di un seno può sfuggire**

## Which Is the True “Diameter?”




**Aorta in cross-section just above aortic valve**

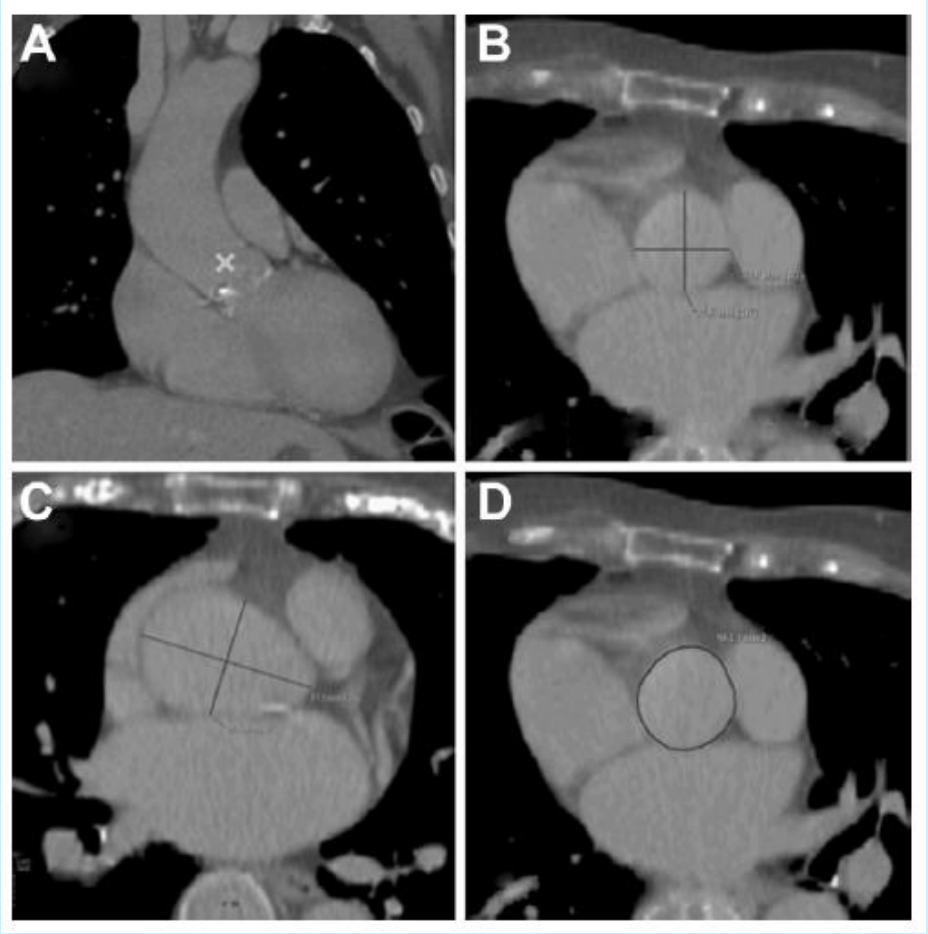
- “measuring”..

**Impact of Image Analysis Methodology on Diagnostic and Surgical Classification of Patients With Thoracic Aortic Aneurysms**

Dorinna D. Mendoza, MD, Minisha Kochar, MD, Richard B. Devereux, MD,

(Ann Thorac Surg 2011;92:904–13)

Occorre attenzione nel modo di misurare




- (A) Coronal view of ascending thoracic aorta
- (B) Double oblique
- (C) Axial diameter.
- (D) Enface planimetry

- “measuring”..

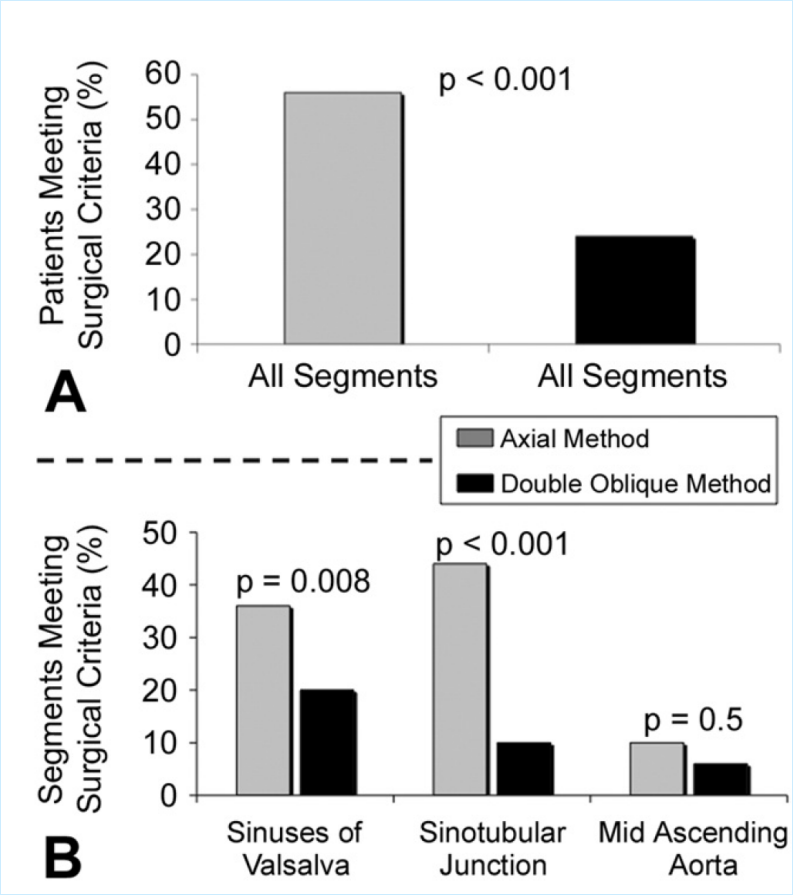
**Impact of Image Analysis Methodology on Diagnostic and Surgical Classification of Patients With Thoracic Aortic Aneurysms**

Dorinna D. Mendoza, MD, Minisha Kochar, MD, Richard B. Devereux, MD,

(Ann Thorac Surg 2011;92:904–13)

**il modo di misurare influenza pesantemente i criteri di indicazione chirurgica**

- (A). Patients meeting established surgical criterion (>5 cm) for prophylactic thoracic aortic aneurysms repair based on measurements yielded by axial (grey bars) versus double oblique (black bars) methods.
- (B) Ascending aortic segments meeting surgical criterion stratified by each method.



- “measuring”..

**2010 ACCF/AHA/AATS/ACR/ASA/SCA/SCAI/SIR/STS/SVM Guidelines for the Diagnosis and Management of Patients With Thoracic Aortic Disease: Executive Summary**

Recommendations for Aortic Imaging Techniques to Determine the Presence and Progression of Thoracic Aortic Disease

CLASS I

1. Le misure devono essere effettuate **con riferimenti anatomici riproducibili, perpendicolarmente all’asse del flusso ematico**. (C)
2. Per **CT e MRI deve essere misurato il diametro esterno** (*C*)
3. Per **ecocardiografia, deve essere misurato il diametro interno**. (*C*)

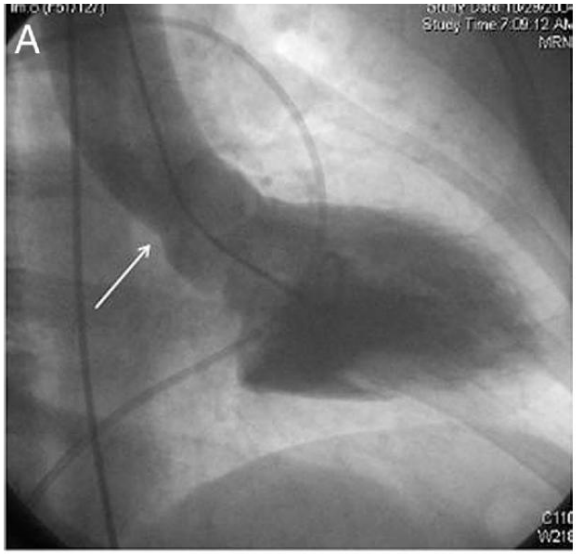
STATE-OF-THE-ART PAPER

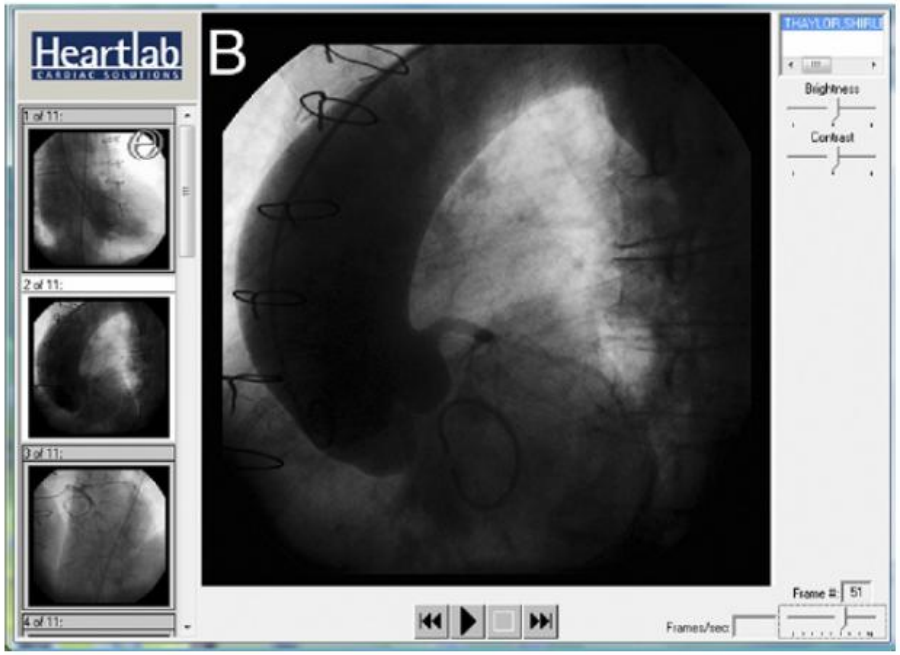
# Thoracic Aortic Aneurysm

## Clinically Pertinent Controversies and Uncertainties

John A. Elefteriades, MD,* Emily A. Farkas, MD†
*New Haven, Connecticut; and St. Louis, Missouri*

JACC Vol. 55, No. 9, 2010
March 2, 2010:841–57




- E' importante ricordare che **la forma è importante come le dimensioni**

**Loss of Normal "Waist" at Sinotubular Junction: A Sign of Intrinsic Aortic Disease**

STATE-OF-THE-ART PAPER

# Thoracic Aortic Aneurysm

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- **Quale è la storia naturale dell'aneurisma?**

- L'aorta toracica ascendente cresce molto lentamente, **circa 0.1 cm per anno.**
- L'aorta discendente cresce più velocemente
- Per ragioni non completamente chiare la meccanica dell'aorta discendente tollera dilatazioni maggiori.

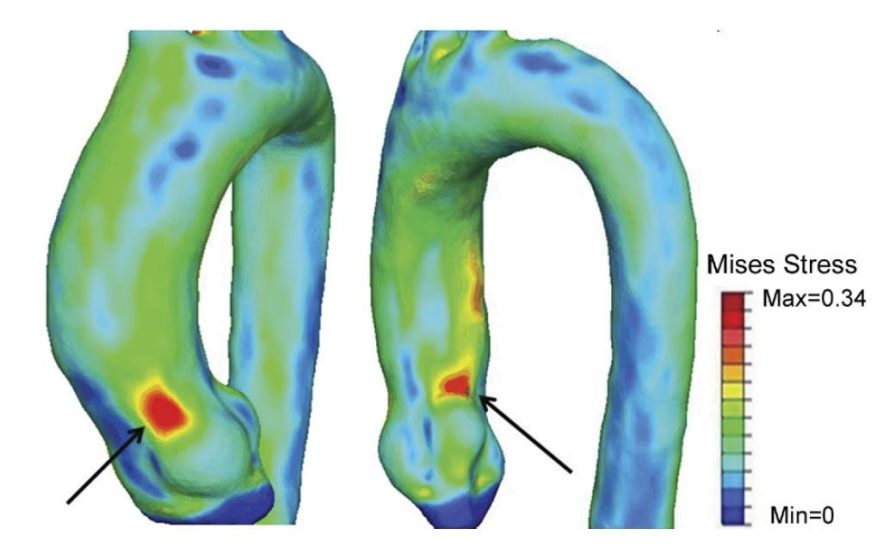
## ▪ “Imaging” del rischio

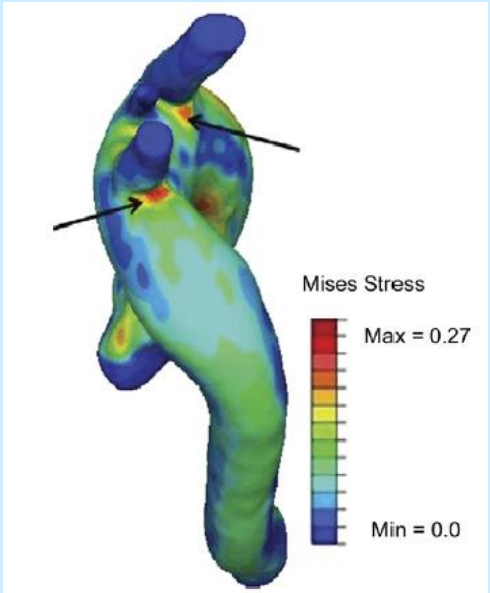
**Pathogenesis of Acute Aortic Dissection: A Finite Element Stress Analysis** (Ann Thorac Surg 2011;91:458–64)

Derek P. Nathan, MD, Chun Xu, PhD, Joseph H. Gorman III, MD, Ron M. Fairman, MD, Joseph E. Bavaria, MD, Robert C. Gorman, MD, Krishnan B. Chandran, PhD, and Benjamin M. Jackson, MD

Gorman Cardiovascular Research Group, Division of Cardiac Surgery, and Division of Vascular Surgery and Endovascular Therapy, University of Pennsylvania, Philadelphia, Pennsylvania; and Department of Biomedical Engineering, University of Iowa, Iowa City, Iowa

- angio CT-scan ecg-gated di 47 individui con normale aorta
- Nell’aorta normale ci sono **picchi di “wall stress” parietale sopra la giunzione seno-tubolare e sopra l’ostio coronarica sinistro**.



STATE-OF-THE-ART PAPER

# Thoracic Aortic Aneurysm

## Clinically Pertinent Controversies and Uncertainties

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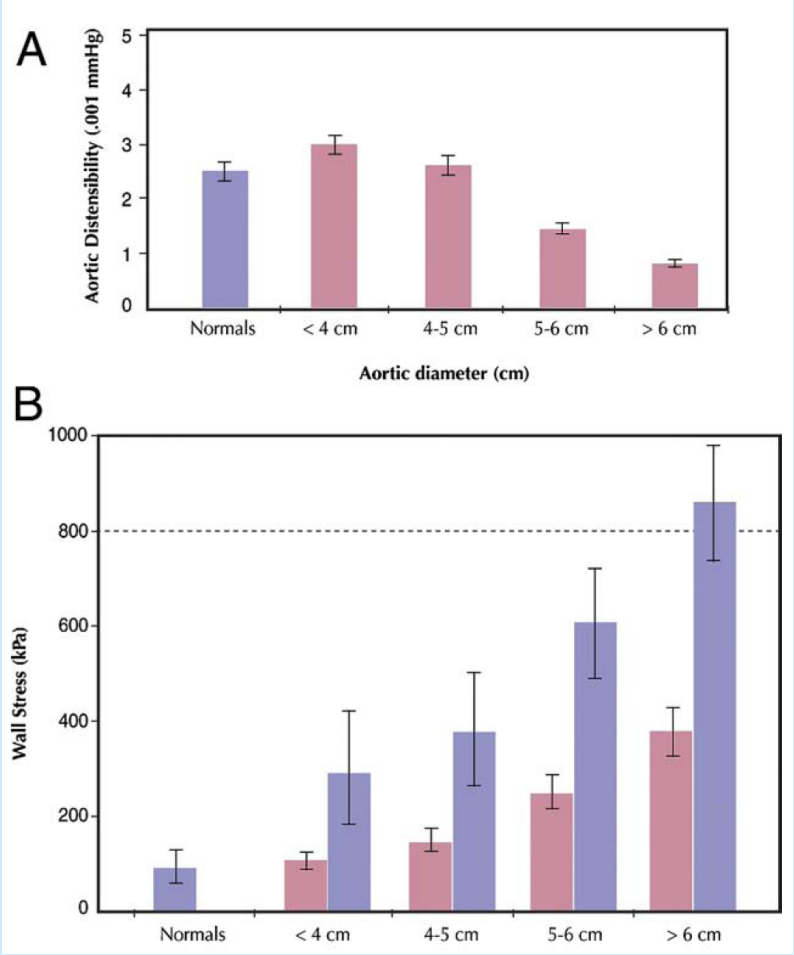
JACC Vol. 55, No. 9, 2010
March 2, 2010:841–57

- **Distensibilità**

- Più si allarga l'aorta, più diminuisce la distensibilità, a **circa 6 cm l'aorta ascendente diviene un tubo rigido.**
- Ne consegue che l'aorta **non si dilata in sistole** e tutta l'energia della contrazione cardiaca e della pressione diviene "wall stress"**.**



- **Rischio Clinico**

STATE-OF-THE-ART PAPER

JACC Vol. 55, No. 9, 2010
March 2, 2010:841–57

## Thoracic Aortic Aneurysm

### Clinically Pertinent Controversies and Uncertainties

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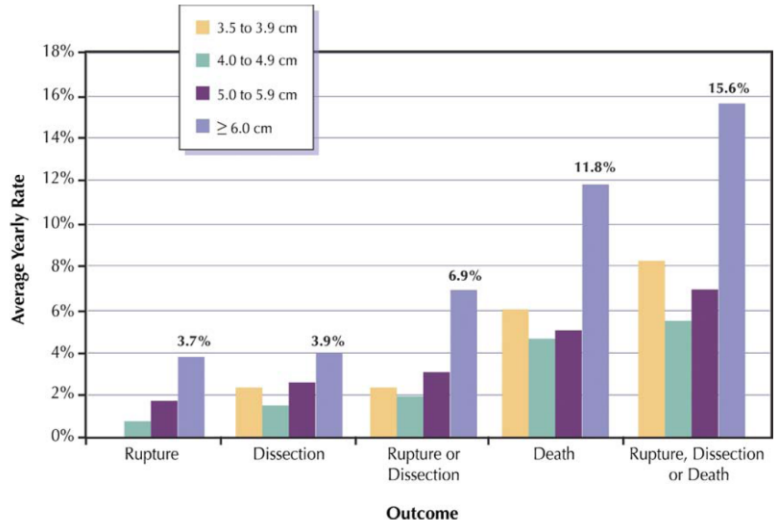
*New Haven, Connecticut; and St. Louis, Missouri*



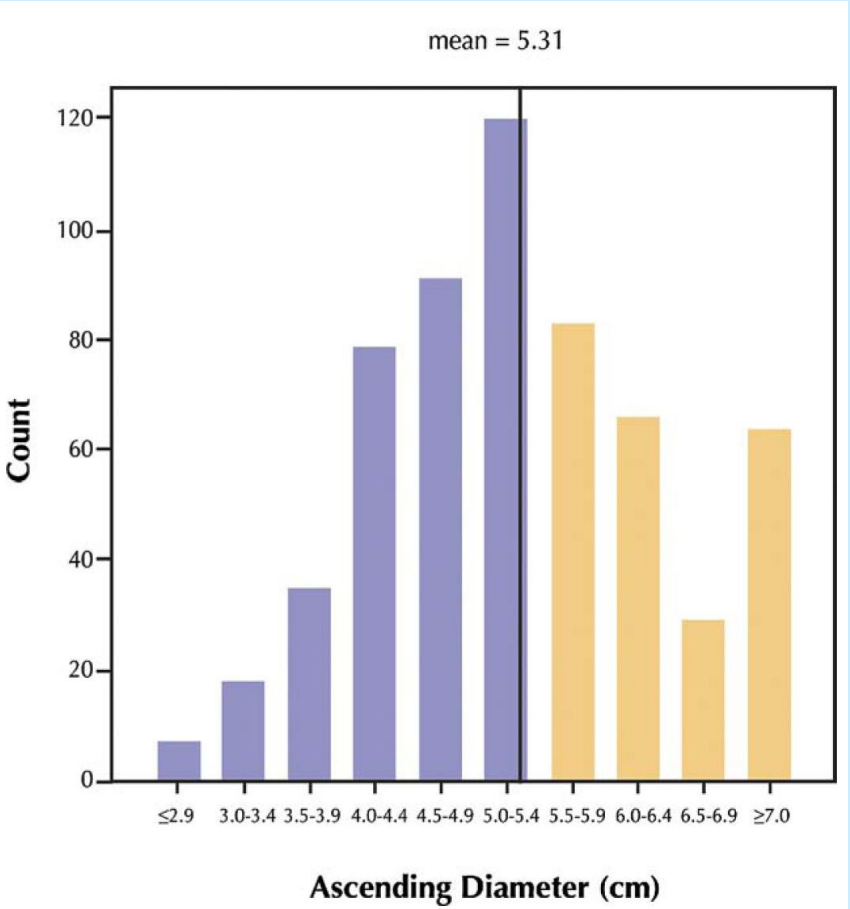

Yearly Rates of Rupture, Dissection, or Death Related to Aortic Size

- Intorno alle dimensioni di **6 cm accadono cose "speciali"** nelle caratteristiche meccaniche di parete **che si traducono in eventi clinici.**

- Questi dati sono alla base della **formulazione delle lineeguida** che indicano l'intervento chirurgico come cura profilattica degli aneurismi aortici per misure superiori a 5,5 cm..

- **IRAD registry**

- **Noi sappiamo però che le dissezioni avvengono anche a diametri inferiori ai 6 cm. con frequenza altrettanto significativa**




Distribution of aortic size at the time of presentation with acute type A

➢ ***Questi individui rappresentano un problema clinico che ci investe quotidianamente:***

- La questione diventa la possibilità di **identificare i pazienti in cui il rischio chirurgico è comunque giustificato, nonostante le misure aortiche non raggiungano i valori critici**
- Sono in valutazione numerosi **parametri con capacità di valutazione prognostica**

- **“Biomarkers” sindrome aortica acuta**

## Biomarkers in Acute Aortic Dissection and Other Aortic Syndromes

(J Am Coll Cardiol 2010;56:1535–41)

Aaron M. Ranasinghe, MD, Robert S. Bonser, MD
*Birmingham, United Kingdom*

- Matrix metalloproteinases
- sm MHC (smooth muscle myosin heavy chain)
- Calponin (counterpart of troponin in smooth muscle cell)
- sELAF (elastin degradation products)
- C-reactive protein
- D-dimer

- **Attualmente non esiste alcun singolo biomarker capace di identificare positivamente la presenza e l’evoluzione delle sindromi aortiche**

Ascending aortic curvature as an independent risk factor for type A dissection, and ascending aortic aneurysm formation: a mathematical model☆

European Journal of Cardio-thoracic Surgery 33 (2008) 995–1001

Michael P. Poullis [a,*], Richard Warwick [a], Aung Oo [a], Robert J. Poole [b]

[a] Department of Cardiothoracic Surgery, Thomas Drive, Liverpool L14 3PE, United Kingdom
[b] Department of Engineering, University of Liverpool, United Kingdom

- **La curvatura dell'arco aortico è relativamente più importante dei diametri aortici, della pressione aortica, della portata, dell'uso di beta-bloccanti e della BSA rispetto alla meccanica delle forze che agiscono sulla parete aortica,**
- Inoltre la curvatura aortica può anche spiegare perchè alcune dissezioni avvengono sopra la giunzione seno-tubolare ed altre prima dell'origine del tronco anonimo

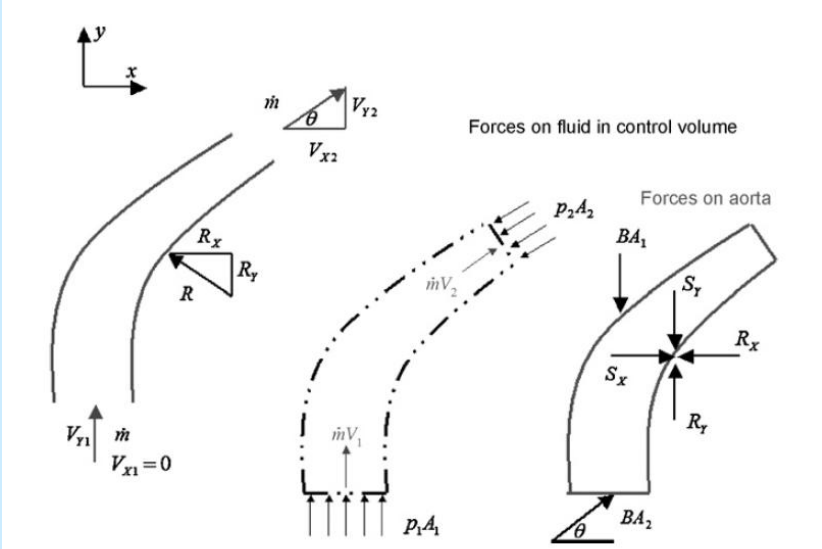



Fig. 7. Diagrammatic representation of mathematical model, and the variables utilised.

Ascending aortic curvature as an independent risk factor for type A dissection, and ascending aortic aneurysm formation: a mathematical model☆

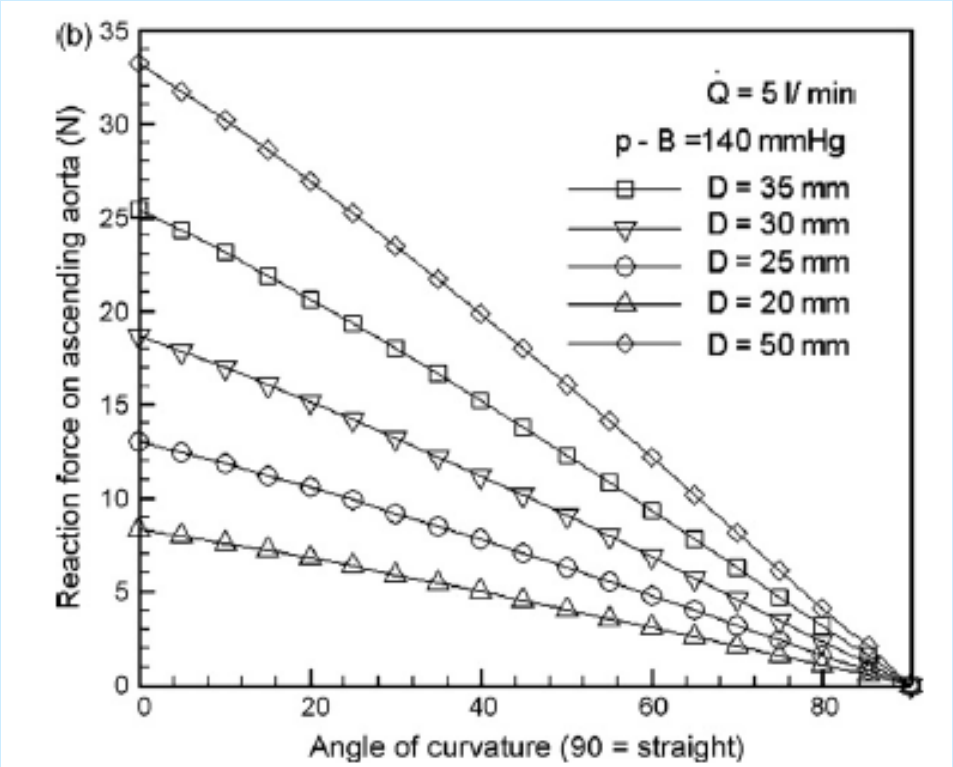
European Journal of Cardio-thoracic Surgery 33 (2008) 995–1001

Michael P. Poullis [a,*], Richard Warwick [a], Aung Oo [a], Robert J. Poole [b]

[a] Department of Cardiothoracic Surgery, Thomas Drive, Liverpool L14 3PE, United Kingdom
[b] Department of Engineering, University of Liverpool, United Kingdom

- Con l'aumento del diametro aortico da 3.5 a 5.5 cm lo stress di parete aumenta di circa **quattro volte**

- Con l'aumento della curvatura dell'arco aortico lo stress di parete può aumentare di circa **10 volte passando da un modello rettilineo ad uno perpendicolare**



- Valvole Bicuspidi

# 2010 ACCF/AHA/AATS/ACR/ASA/SCA/SCAI/SIR/STS/SVM Guidelines for the Diagnosis and Management of Patients With Thoracic Aortic Disease: Executive Summary

JACC Vol. 55, No. 14, 2010
April 6, 2010:1509–44

Recommendations for Bicuspid Aortic Valve and Associated Congenital Variants in Adults

CLASS I

1. **First-degree relatives of patients** with a **bicuspid aortic valve, premature onset of thoracic aortic disease with minimal risk factors, and/or a familial form of thoracic aortic aneurysm and dissection should be evaluated for the presence of a bicuspid aortic valve and asymptomatic thoracic aortic disease**. (*C*)
2. **All patients with a bicuspid aortic valve should have both the aortic root and ascending thoracic aorta evaluated for evidence of aortic dilatation** (*B*)

# Increased Ascending Aortic Wall Stress in Patients With Bicuspid Aortic Valves

(Ann Thorac Surg 2011;92:1384–90)

Derek P. Nathan, MD, Chun Xu, PhD, Ted Plappert, CVT, Benoit Desjardins, MD, Joseph H. Gorman III, MD, Joseph E. Bavaria, MD, Robert C. Gorman, MD, Krishnan B. Chandran, PhD, and Benjamin M. Jackson, MD

Departments of Surgery and Radiology, Divisions of Cardiac Surgery and Vascular Surgery, and Gorman Cardiovascular Research Group, Hospital of the University of Pennsylvania, Philadelphia, Pennsylvania; and Department of Biomedical Engineering, University of Iowa, Iowa City, Iowa

- **La misura dello stress di parete nell'aorta ascendente di pazienti con BAV è più grande che nei pazienti con TAV.**
- Ci sono **differenze nella localizzazione** dello stress di parete nelle aorte con BAV rispetto alle TAV.
- I pazienti con BAV tendono ad avere uno **stress di parete più alto nel lato convesso dell'aorta ascendnte sopra la coronaria dx**

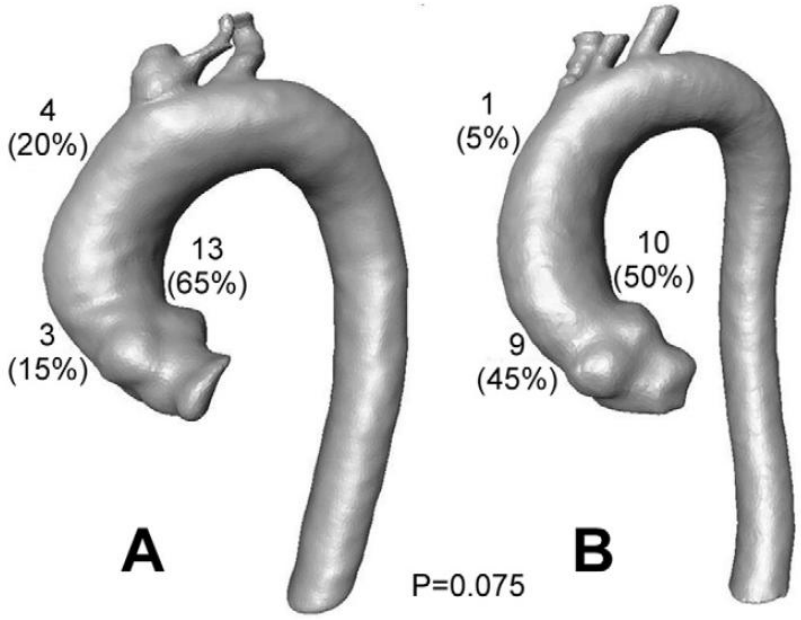



*Fig 2. Location of 99th percentile wall stress in the ascending aorta in patients with (A) tricuspid and (B) bicuspid aortic valves. This differing distribution of peak wall stress approached but did not reach statistical significance (p = 0.075).*

*Radiology:* Volume 255: Number 1—April 2010

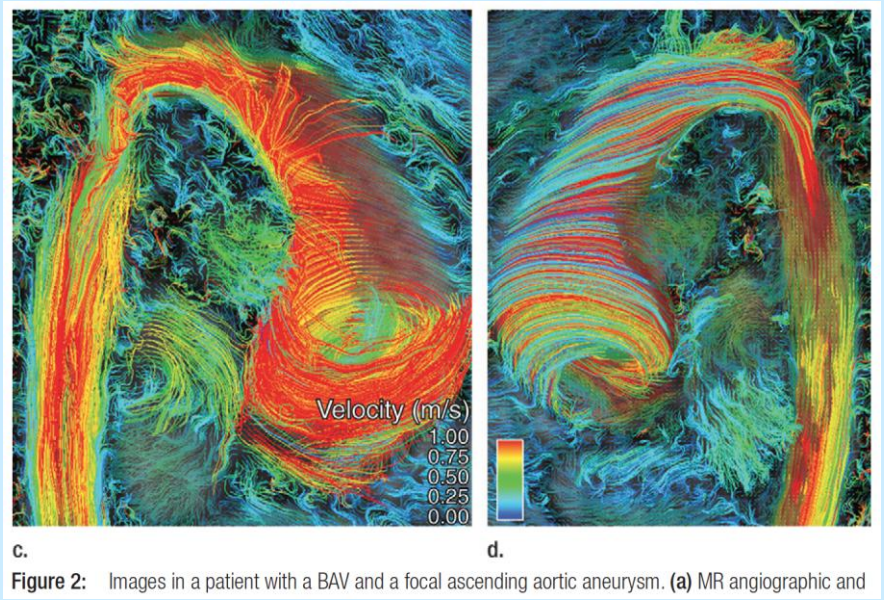
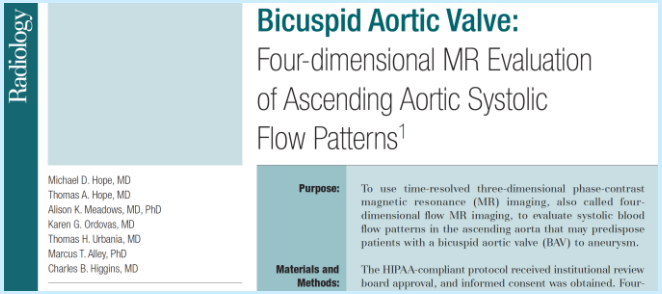



**Figure 2:** Images in a patient with a BAV and a focal ascending aortic aneurysm. **(a)** MR angiographic and




- Con la RNM 4D si possono evidenziare le **differenze di flusso tra BAV e TAV** con formazione di anormali vortici non laminari anche in assenza di stenosi valvolari.

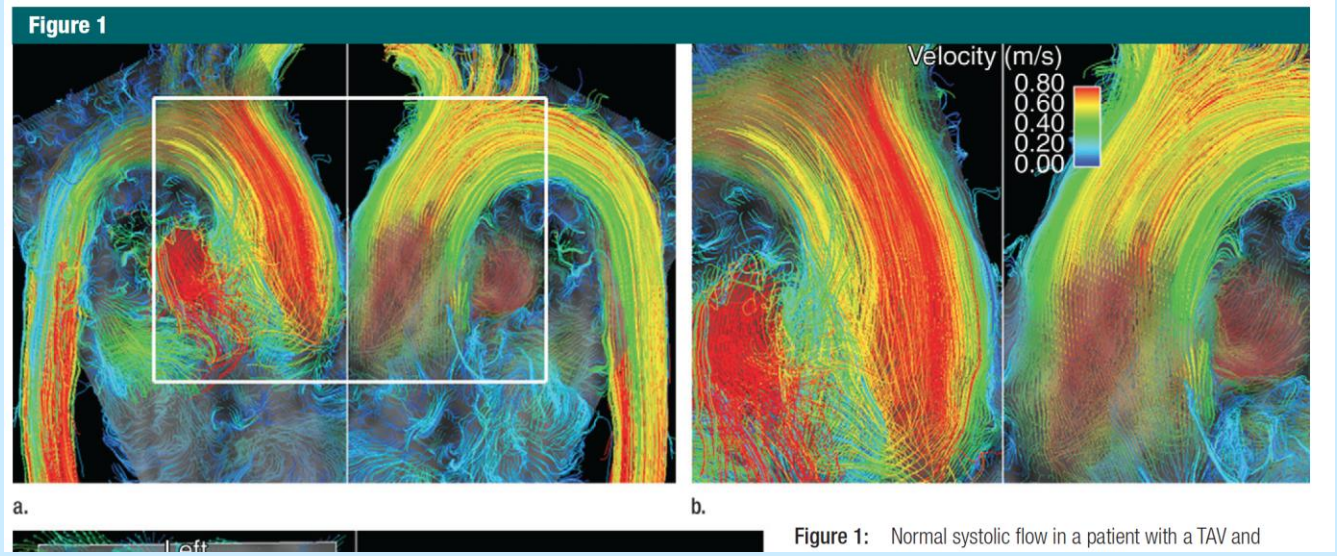



**Figure 1:** Normal systolic flow in a patient with a TAV and

# Restricted cusp motion in right-left type of bicuspid aortic valves: A new risk marker for aortopathy

(J Thorac Cardiovasc Surg 2012;144:360-9)

Alessandro Della Corte, MD, PhD,[a] Ciro Bancone, MD,[a] Carlo A. Conti, PhD,[b] Emiliano Votta, PhD,[b] Alberto Redaelli, PhD,[b] Luca Del Viscovo, MD,[c] and Maurizio Cotrufo, MD[d]

## "cusp opening angle" (COA)

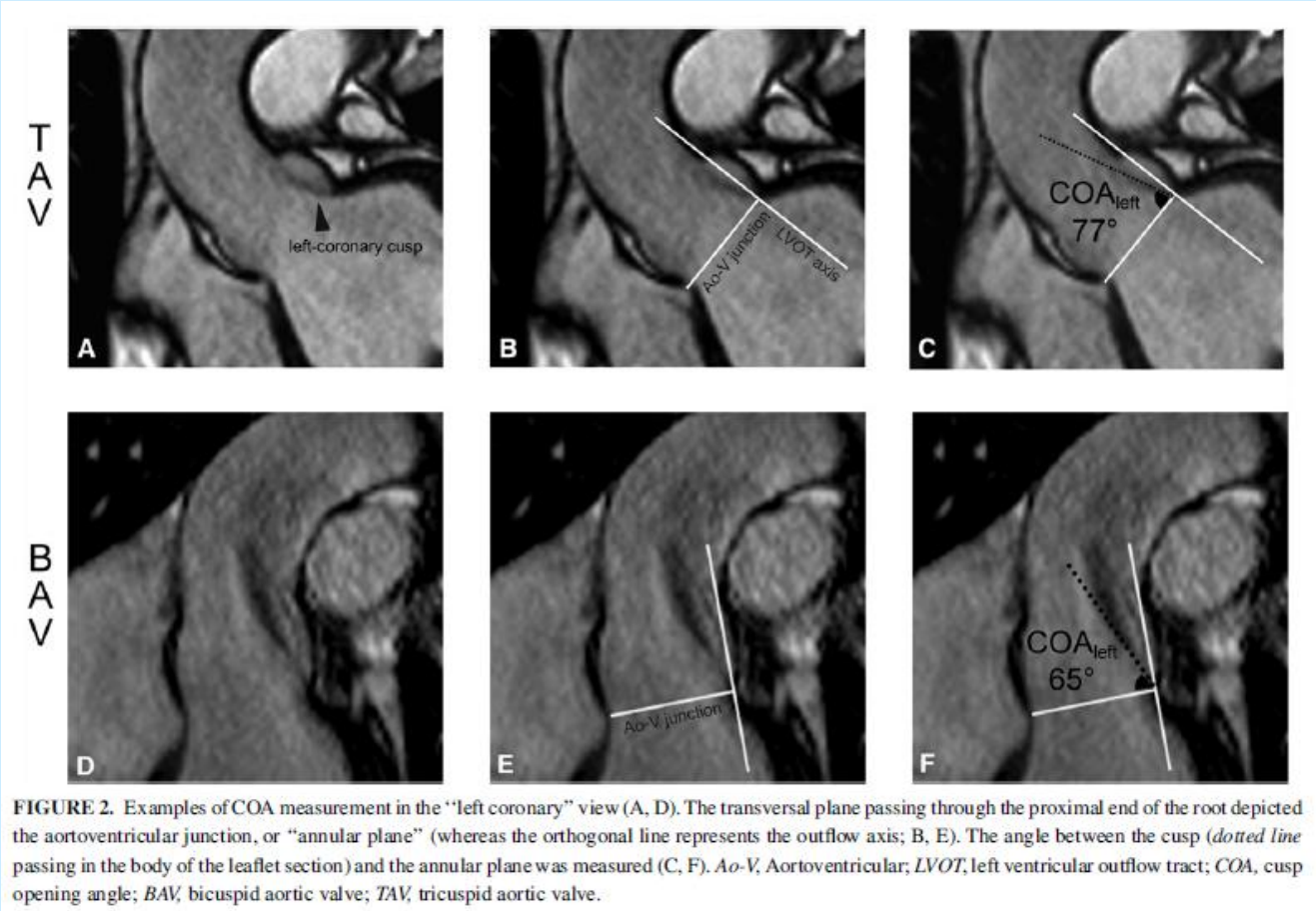



**FIGURE 2.** Examples of COA measurement in the "left coronary" view (A, D). The transversal plane passing through the proximal end of the root depicted the aortoventricular junction, or "annular plane" (whereas the orthogonal line represents the outflow axis; B, E). The angle between the cusp (*dotted line* passing in the body of the leaflet section) and the annular plane was measured (C, F). *Ao-V,* Aortoventricular; *LVOT,* left ventricular outflow tract; *COA,* cusp opening angle; *BAV,* bicuspid aortic valve; *TAV,* tricuspid aortic valve.

**Restricted cusp motion in right-left type of bicuspid aortic valves: A new risk marker for aortopathy**

(J Thorac Cardiovasc Surg 2012;144:360-9)

Alessandro Della Corte, MD, PhD,[a] Ciro Bancone, MD,[a] Carlo A. Conti, PhD,[b] Emiliano Votta, PhD,[b] Alberto Redaelli, PhD,[b] Luca Del Viscovo, MD,[c] and Maurizio Cotrufo, MD[d]

- **Una riduzione dl movimento della cuspide è sufficiente per causare una modifica del flusso aortico come osservata alla MRI**
- COA è una misura quantitativa dell'apertura abnorme delle BAV con fusione delle cuspidi destra e sinistra (la più comune)
- **Nell'analisi multivariata il "cusp opening angle" (COA) predice indipendentemente il diametro dell'aorta ascendente ed la sua crescita**

**Replacement of the ascending aorta in bicuspid aortic valve disease: Where do we draw the line?**

Thoralf M. Sundt III, MD

J Thorac Cardiovasc Surg 2010;140:S41-4

***"Dove tiriamo una riga ?"***

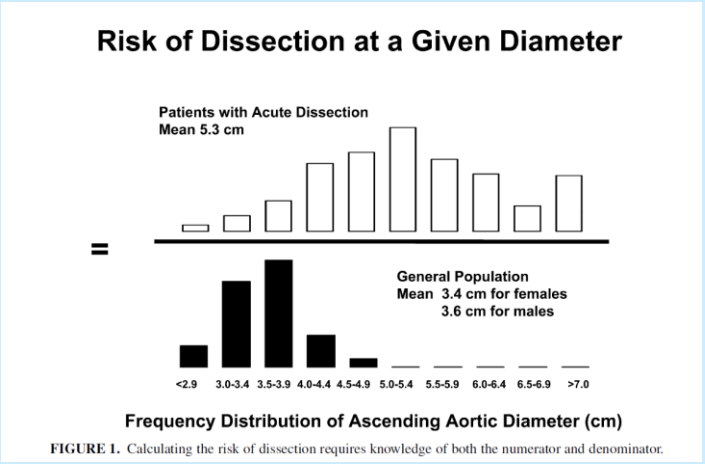



FIGURE 1. Calculating the risk of dissection requires knowledge of both the numerator and denominator.

- BAV come Marfan ?

- L'evidenza che la valvola bicuspide è un fattore di rischio indipendente dalle dimensioni per dissezione è povera
- **La bicuspidia non è analoga alla sindrome di Marfan**

Bicuspid aortic valve leaflet morphology in relation to aortic root morphology: a study of 300 patients undergoing open-heart surgery☆

Veronica Jackson[a,*], Johan Petrini[b], Kenneth Caidahl[b], Maria J. Eriksson[b], Jan Liska[a], Per Eriksson[c], Anders Franco-Cereceda[a]

[a] Cardiothoracic Surgery Unit at the Department of Molecular Medicine and Surgery at Karolinska Institutet and Karolinska University Hospital
[b] The Clinical Physiology Unit at the Department of Molecular Medicine and Surgery at Karolinska Institutet and Karolinska University Hospital
[c] The Atherosclerosis Research Unit at the Center for Molecular Medicine at the Department of Medicine at Karolinska Institutet

European Journal of Cardio-thoracic Surgery 40 (2011) e118—e124

- Epidemiologia degli operati aneurisma aorta ascendente
- **70—80% of BAV patients have a fused right- and left coronary cusp**

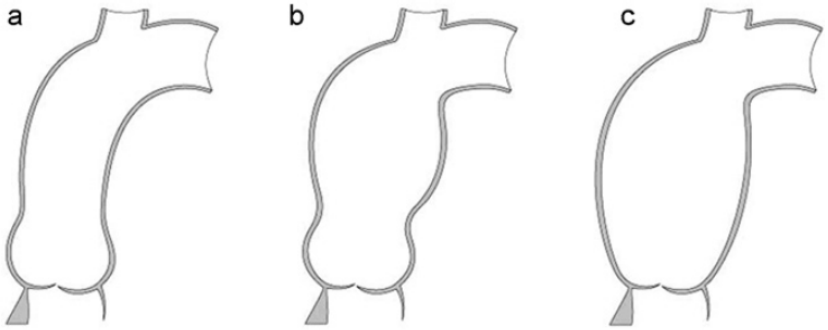



Fig. 1. Aortic root morphology. Schematic illustrations of the three distinct morphological patterns of the aorta observed in this study. Normal aorta configuration (a); aneurysm of the aorta (b); ectasia of the aorta (c).

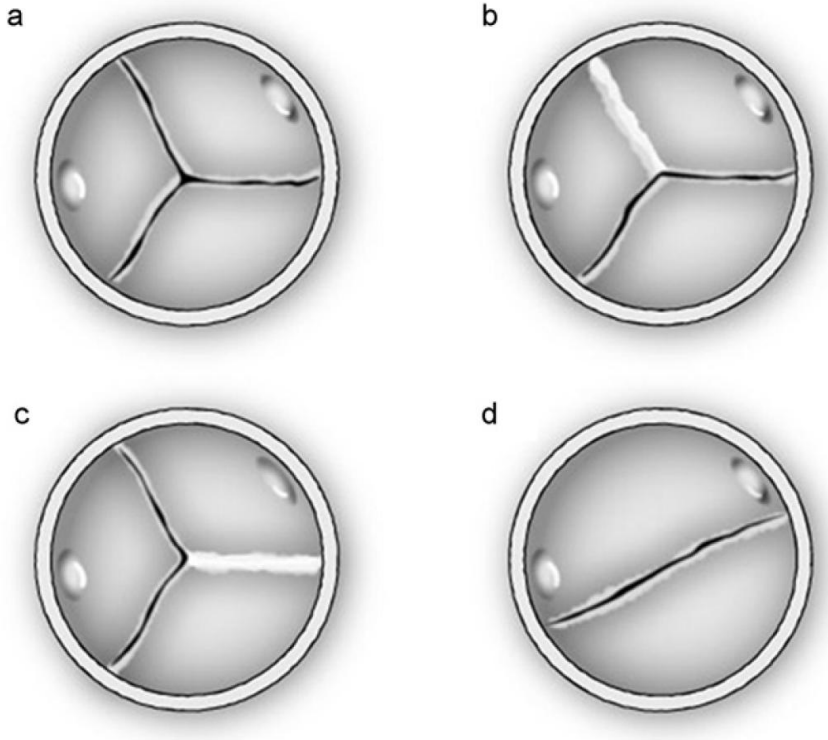



Fig. 2. Schematic illustration of aortic valve leaflet morphology observed in this study. Normal tricuspid aortic valve (a); fusion of the right- and left-coronary cusp with remnant raphe (b); fusion of the left- and non-coronary cusps (c); true bicuspid valve without any remnant raphe (d).

Bicuspid aortic valve leaflet morphology in relation to aortic root morphology: a study of 300 patients undergoing open-heart surgery☆

Veronica Jackson[a,*], Johan Petrini[b], Kenneth Caidahl[b], Maria J. Eriksson[b], Jan Liska[a], Per Eriksson[c], Anders Franco-Cereceda[a]

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European Journal of Cardio-thoracic Surgery 40 (2011) e118—e124

- I nostri risultati confermano che la **BAV è più comunemente associata ad aneurisma aorta ascendente** rispetto alla TAV ed hanno un anulus più grande
- **La morfologia dell'aorta ascendente non correla con la presenza e la tipologia** di BAV.

- **La presenza di aneurisma nell'aorta ascendente non è correlata con una particolare morfologia di bicuspidia**
- **La morfologia della bicuspidia non può essere usata per predire aneurismi presenti o futuri**

# Individualized Thoracic Aortic Replacement for the Aortopathy of Bicuspid Aortic Valve Disease

Brian Lima[1], Judson B. Williams[1], S. Dave Bhattacharya[1], Asad A. Shah[1], Nicholas Andersen[1], Andrew Wang[2], J. Kevin Harrison[2], G. Chad Hughes[1]

*[1]Division of Thoracic and Cardiovascular Surgery, Department of Surgery, [2]Division of Cardiovascular Medicine, Department of Medicine, Duke University Medical Center, Durham, NC, USA*

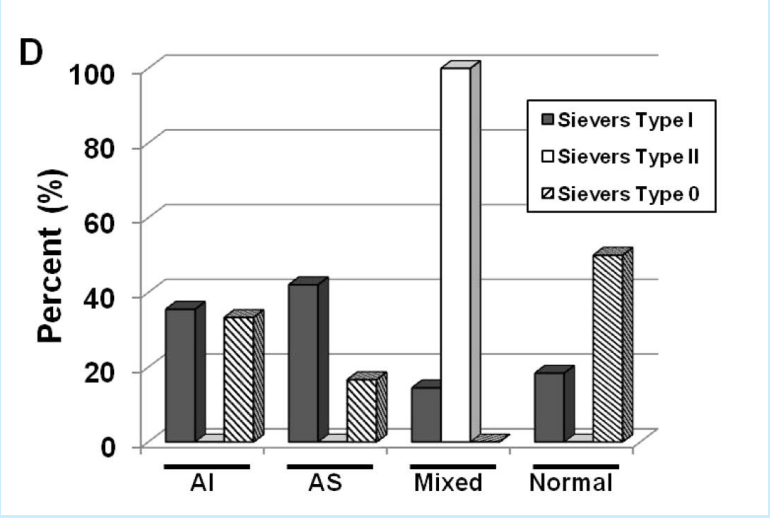
The Journal of Heart Valve Disease 2011;20:387-395

- **L'anatomia patologica dell'aorta ascendente non è prevista dalla morfologia della bicuspidia della valvola aortica**
- La dilatazione dell'aorta ascendente è più comune nei pazienti con quadri predominanti di rigurgito valvolare aortico

*Table II: Bicuspid aortic valve morphology (Sievers classification).*

| Sievers class | No. of patients (n = 89) |
|---|---|
| Type I | 77 (86) |
| R/L | 67 (75) |
| R/N | 8 (9) |
| L/N | 2 (2) |
| Type II | 6 (7) |
| Type 0 | 6 (7) |

Values in parentheses are percentages.
Fusion of right/left (R/L), right/non- (R/N), or left/non- (L/N) coronary cusps.
Type I: one raphe present; Type II: two raphe present; Type 0: no raphe present ('true bicuspid'). Based on the classification system described by Sievers (23).




GUIDELINES

**Clinician Update**

## What Is New in Dilatation of the Ascending Aorta?

### Review of Current Literature and Practical Advice for the Cardiologist

Luc Cozijnsen, MD; Richard L. Braam, MD, PhD; Reinier A. Waalewijn, MD, PhD; Marc A.A.M. Schepens, MD; Bart L. Loeys, MD, PhD; Matthijs F.M. van Oosterhout, MD; Daniela Q.C.M. Barge-Schaapveld, MD, PhD; Barbara J.M. Mulder, MD, PhD

(*Circulation*. 2011;123:924-928.)

# Cosa fare con aorta ascendente > 4.0 cm:

- Consigli per I clinici che hanno in cura Pz con ectasia aortica

**Diametri >4.0 cm**

- Valutare la presenza di connettivopatie e alterazioni geniche
- Iniziare terapia con β-bloccante
- Mantenere uno stretto controllo della pressione arteriosa
- Limitare moderatamente l'attività fisica
- Sconsigliare le gravidanze
- Controllare ogni anno TTE e/o CT / MRI imaging

# Guidelines on the management of valvular heart disease (version 2012)

**The Joint Task Force on the Management of Valvular Heart Disease of the European Society of Cardiology (ESC) and the European Association for Cardio-Thoracic Surgery (EACTS)**

## Indications for surgery in aortic root disease (whatever the severity of AR)

- Surgery is indicated in patients who have aortic root disease with maximal ascending aortic diameter
- ≥50 mm for patients with Marfan syndrome. (I C)
- ≥45 mm for patients with Marfan syndrome with risk factors (a)
- ≥50 mm for patients with bicuspid valve with risk factors (b)
- ≥55 mm for other patients (IIa C)

(a): Risk factors
- Family history of aortic dissection and/or aortic size increase >2 mm/year (on repeated measurements using the same imaging technique, measured at the same aorta level with side-by-side comparison and confirmed by another technique),
- Severe AR or mitral regurgitation,
- Desire of pregnancy.

(b): Risk factors
- Coarctation of the aorta,
- Systemic hypertension,
- Family history of dissection
- Increase in aortic diameter >2 mm/year (on repeated measurements using the same imaging technique, measured at the same aorta level with side-by-side comparison and confirmed by another technique).

# 2010 ACCF/AHA/AATS/ACR/ASA/SCA/SCAI/SIR/STS/SVM Guidelines for the Diagnosis and Management of Patients With Thoracic Aortic Disease: Executive Summary

JACC Vol. 55, No. 14, 2010
April 6, 2010:1509–44

Recommendation for Symptomatic Patients With Thoracic Aortic Aneurysm

CLASS I

1. **Patients with symptoms suggestive of expansion** of a thoracic aneurysm should be evaluated for prompt surgical intervention unless life expectancy from comorbid conditions is limited or quality of life is substantially impaired. (*Level of Evidence: C*)

# 2010 ACCF/AHA/AATS/ACR/ASA/SCA/SCAI/SIR/STS/SVM Guidelines for the Diagnosis and Management of Patients With Thoracic Aortic Disease: Executive Summary

JACC Vol. 55, No. 14, 2010
April 6, 2010:1509–44

## Recommendation for Asymptomatic Patients With Thoracic 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. (*C*)

# 2010 ACCF/AHA/AATS/ACR/ASA/SCA/SCAI/SIR/STS/SVM Guidelines for the Diagnosis and Management of Patients With Thoracic Aortic Disease: Executive Summary

JACC Vol. 55, No. 14, 2010
April 6, 2010:1509–44

Recommendation for Asymptomatic Patients With Thoracic Aortic Aneurysm

CLASS I

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**) to avoid acute dissection or rupture. (*C*)

## Controindicazioni

- La chirurgia dell'aneurisma aortico non ha controindicazioni assolute.
- **Le controindicazioni relative sono individualizzate** (rapporto costo/beneficio) basandosi sui rischi complessivi del paziente:
  - inteventi ad alto rischio includono le persone molto anziane, le insufficienze renali terminali, le insufficienze respiratorie, cirrosi ad altre condizioni di gravi comorbilità

- Tecniche Chirurgiche

# Procedure specifiche e potenziali indicazioni

- **Quale chirurgia**

| | |
|---|---|
| Sostituzione con protesi tubolare semplice | Aneurisma aorta ascendente con normale radice aortica (possibile correzione rigurgito centrale da dilatazione giunzione seno-tubolare) |
| Tubo valvolato composito con protesi meccanica o biologica | Aneurisma aorta ascendente e patologia valvolare non correggibile |
| Sostituzione separata valvolare e aorta ascendente con protesi | Aneurisma aorta ascendente con accettabile radice aortica e patologia valvolare non correggibile |
| Riparazione valvolare aortica associata a sostituzione aorta ascendente | Aneurisma aorta ascendente e patologia valvolare correggibile |
| Allograft aortico | Endocarditi con distruzione radice aortica o infezione di precedenti graft |

# Quali sono i problemi?

- Quando sostituire l'aorta ascendente in corso di sostituzione valvolare aortica?
  - Cosa cambia con la valvola bicuspide?
  - Cosa cambia con altri fattori di rischio?

- Tubo valvolato – composito?
- Quando ricorrere all'arresto di circolo?

- Tecniche alternative:
  Aortoplastica, Wrapping?

- Cosa fare con malattia aterosclerotica dell'ascendente?

- Il reintervento:
  Infezioni protesiche, pseudoaneurismi

## Contemporary Results for Proximal Aortic Replacement in North America

(J Am Coll Cardiol 2012;60:1156–62)

Judson B. Williams, MD, MHS,*† Eric D. Peterson, MD, MPH,*‡ Yue Zhao, PHD,*
Sean M. O'Brien, PHD,* Nicholas D. Andersen, MD,† D. Craig Miller, MD,§ Edward P. Chen, MD,‖
G. Chad Hughes, MD†

*Durham, North Carolina; Stanford, California; and Atlanta, Georgia*

- *Society of Thoracic Surgeons Database* per pazienti operati sostituzione aorta ascendente (+/- radice +/- arco) dal 2004 al 2009

**Table 3** Selected Predictors of Operative Mortality Among All Patients

| Variable | Adjusted OR (95% CI) | p Value |
|---|---|---|
| Status: emergent vs. elective | 5.91 (5.31–6.58) | <0.0001 |
| Pre-operative shock | 2.01 (1.74–2.31) | <0.0001 |
| Status: urgent vs. elective | 2.01 (1.78–2.27) | <0.0001 |
| Concomitant CABG | 2.14 (1.87–2.46) | <0.0001 |
| Concomitant mitral valve procedure | 1.63 (1.36–1.96) | <0.0001 |
| Any reoperation | 1.63 (1.43–1.86) | <0.0001 |
| Cerebrovascular disease | 1.43 (1.28–1.59) | <0.0001 |
| Chronic kidney disease stage 3 or greater, GFR <60 ml/min/1.73 $m^2$ | 1.43 (1.32–1.56) | <0.0001 |
| Moderate or severe chronic lung disease | 1.36 (1.20–1.54) | <0.0001 |
| Arch involvement | 1.23 (1.10–1.37) | 0.0002 |

**Table 5** Selected Predictors of Operative Mortality Among Elective Patients

| Variable | Adjusted OR (95% CI) | p Value |
|---|---|---|
| Pre-operative dialysis | 4.04 (2.56–6.37) | <0.0001 |
| Any reoperation | 2.29 (1.93–2.70) | <0.0001 |
| Concomitant CABG | 1.99 (1.70–2.32) | <0.0001 |
| Moderate or severe chronic lung disease | 1.85 (1.52–2.25) | <0.0001 |
| Congestive heart failure, NYHA class IV | 1.74 (1.28–2.38) | 0.0005 |
| Concomitant mitral valve procedure | 1.69 (1.34–2.14) | <0.0001 |
| Female vs. male | 1.57 (1.36–1.83) | <0.0001 |
| Immunosuppressive treatment | 1.55 (1.10–2.17) | <0.0113 |
| Age >70 yrs, 5-yr increments | 1.44 (1.33–1.56) | <0.0001 |
| Pre-operative atrial fibrillation | 1.32 (1.12–1.56) | 0.0011 |


- Rischio di dissezione aortica
- Definizione
  - Istopatologia
  - Fisiopatologia
  - Studi genetici
- Normalità?
  - Come la misuriamo
- La dilatazione
  - Wall stress
- La Bicuspidia
- Lineeguida
- Tecniche chirurgiche

# SESSIONE PLENARIA

## Ia giornata AUDITORIUM 14 MARZO 2012

**Simposio** **Le malattie croniche dell'aorta toracica**

*Tra anatomia, diagnostica, timing operatorio, tecniche riparative e follow-up: non sempre è agevole orientarsi in una patologia nella quale le indicazioni della letteratura vengono spesso superate e messe in discussione nella pratica di tutti i giorni*

**Moderatori:** **Luigi Oltrona Visconti – Ugolino Livi**

**11.00** **Mauro Pepi –** Valutazione integrata ECO, RM e TC dell'apparato valvolare aortico e del vaso nelle malattie dell'aorta toracica. *Come dare all'emodinamista o al cardiochirurgo tutte le informazioni necessarie all'intervento. Come rilevare problemi a distanza.*

**11.20** **Claudio Grossi –** La dilatazione isolata dell'aorta ascendente con e senza valvulopatia aortica associata.
*Storia naturale, timing chirurgico e tecnica operatoria.*

**11.40** **Fabrizio Settepani –** La tecnica tradizionale di sostituzione dell'aorta ascendente e dell'arco.
*Tecnica e risultati di un intervento complesso ma consolidato.*

**12.00** **Davide Pacini –** Le tecniche ibride di sostituzione dell'aorta ascendente e dell'arco. *Come cambia la tecnica associando le endoprotesi. I risultati.*

**12.20** **Tommaso Donati –** La terapia endovascolare della dissezione cronica di tipo B. *Quando l'uso dell'endoprotesi semplifica la tecnica e riduce l'invasività della procedura. Risultati a confronto con l'approccio tradizionale.*

**12.40** DISCUSSIONE

**13.00** **Pausa lunch**