Ανεπάρκεια Αορτής - Ανευρύσματα

Εφη Γ.Ρούσκα, MD, PhD

Effie ROUSKA, MD

FUNCTIONAL Aortic Root Anatomy

Aorto ventricular junction

+

Sinotubular junction

↓

Single functional structure

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Aortic Regurgitation

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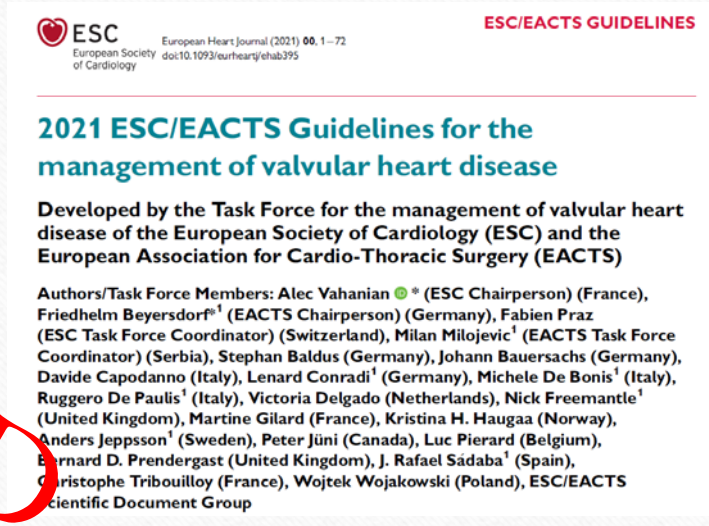
Aortic regurgitation can be caused by primary disease of the aortic valve cusps and/or abnormalities of the aortic root and ascending aortic geometry. Degenerative tricuspid and bicuspid aortic regurgitation are the most common aetiologies in high-income countries, accounting for approximately two-thirds of the underlying aetiology of aortic regurgitation in the EURObservational Registry Programme Valvular Heart Disease II registry.[1]
Other causes include infective and rheumatic endocarditis.

Acute severe aortic regurgitation is mostly caused by infective endocarditis, and less frequently by aortic dissection.



ESC European Society of Cardiology

European Heart Journal (2021) 00, 1–72
doi:10.1093/eurheartj/ehab395

ESC/EACTS GUIDELINES

2021 ESC/EACTS Guidelines for the management of valvular heart disease

Developed by the Task Force for the management of valvular heart disease of the European Society of Cardiology (ESC) and the European Association for Cardio-Thoracic Surgery (EACTS)

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Aetiology of AR

Table 1
Etiology of Aortic Regurgitation

Diseases involving the aortic cusps (leaflets)

- Senile calcific degeneration
- Bicuspid aortic valves
- Rheumatic valvular heart disease
- Endocarditis

Diseases involving the aortic root

- Hypertension
- Bicuspid aortic valve complex
- Collagen disorders, e.g., Marfan and Ehlers Danlos syndromes
- Aortic dissection (Type A)
- Trauma
- Aortitis (vasculitic, infectious)
- Ventricular septal defect

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Evaluation of AR

- **Echocardiography** **is the key examination……**

- **used to describe valve anatomy,**
- **quantify aortic regurgitation,**
- **evaluate its mechanisms,**
- **define the morphology of the aorta, and**
- **determine the feasibility of valve sparing aortic surgery or valve repair**

- **CCT**

- **CMR** (to quantify RF when echo measurements are equivocal or discordant)

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El Khoury functional classification for AR

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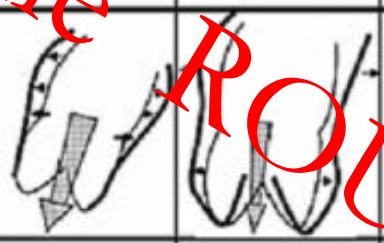
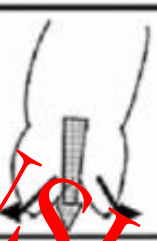
AI Class	Type I: Normal cusp motion with FAA dilatation or cusp perforation				Type II: Cusp Prolapse	Type III: Cusp Restriction
	Ia	Ib	Ic	Id		
Mechanism						
Repair Techniques (Primary)	STJ remodeling *Ascending aortic graft*	Aortic Valve sparing: *Reimplantation or Remodeling with SCA*	SCA	Patch Repair *Autologous or bovine pericardium*	Prolapse Repair *Plication Triangular resection Free margin Resuspension Patch*	Leaflet Repair *Shaving Decalcification Patch*
(Secondary)	SCA		STJ Annuloplasty	SCA	SCA	SCA

El Khoury functional classification for aortic regurgitation (SCA: Subcommissural annuloplasty is used much less frequently now than in the past).

El Khoury

functional classification for AR

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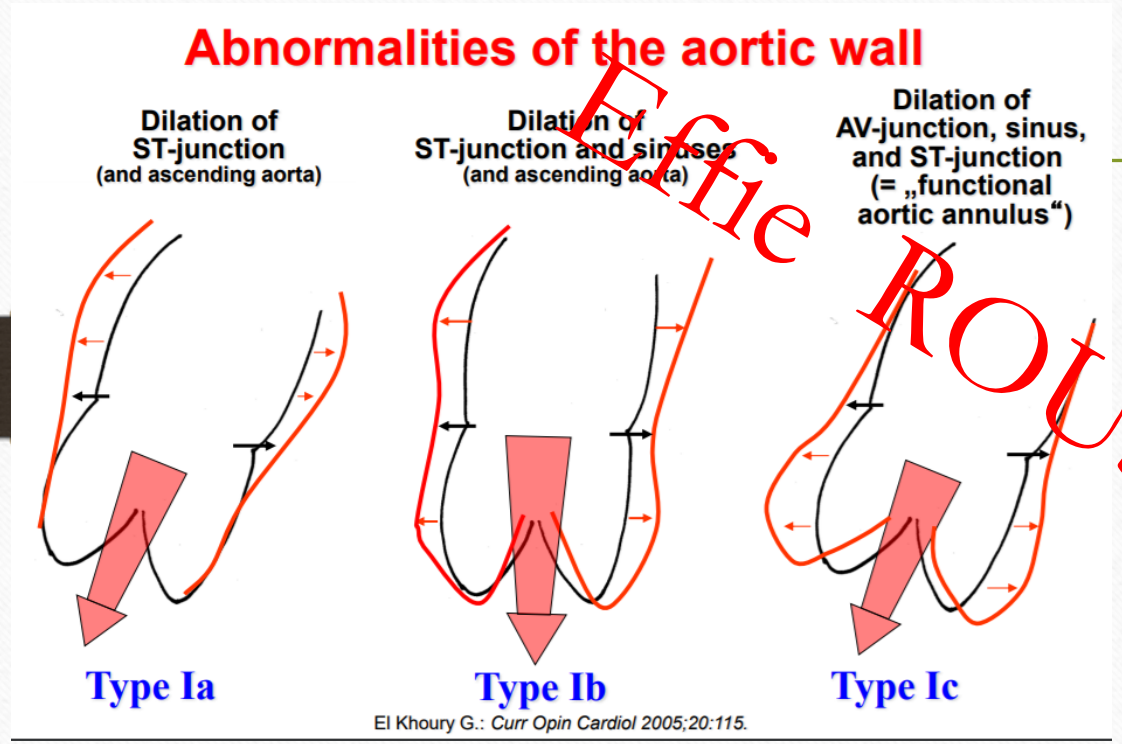
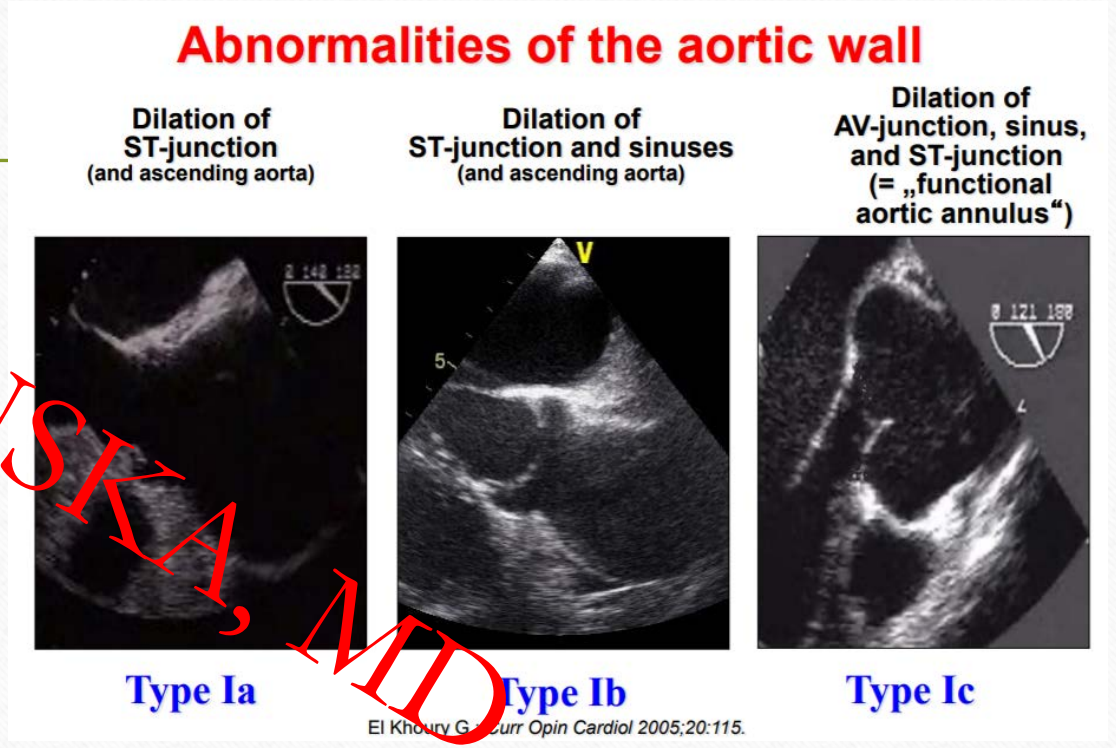
- **Type I dysfunction** was identified when the dimensions of any components of the aortic root, including the aortic annulus, the sinuses of Valsalva, and the sinotubular junction exceeded the upper limits of published normal values[13] and no other cause of AR was identified.

- **Type 2 dysfunction** was considered in the presence of an eccentric AR jet and either a cusp prolapse or a cusp fenestration.

- **Type 3 dysfunction** was considered whenever the quality or quantity of the cusp tissue was judged to be poor.

Echo in AR - Mechanisms





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Central AR jet !

Abnormalities of the aortic valve cusps

Perforation

Paracentral jet

Prolapse

Eccentric jet away from the prolapsing cusp

Restrictive cusp motion

Eccentric jet towards the the more restricted cusp

Type Id

Type II

Type III

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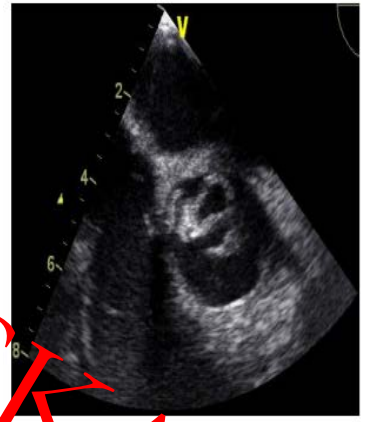
Abnormalities of the aortic valve cusps

Perforation



Prolaps

Restrictive cusp motion

Type Id

Type II

Type III

Abnormalities of the aortic wall

Dilation of ST-junction (and ascending aorta)	Dilation of ST-junction and sinuses (and ascending aorta)	Dilation of AV-junction, sinus, and ST-junction (= „functional aortic annulus“)

Main Etiologies:

- Genetically determined vascular connective tissue disease:
  - Syndromes: Marfan, Ehlers-Danlos, Loeys-Dietz, etc.
  - Bicuspid valve
- Hypertensive aortic root dilatation

Type Ia **Type Ib** **Type Ic**

El Khoury G.: *Curr Opin Cardiol 2005;20:115.*

Abnormalities of the aortic valve cusps

Perforation	Prolaps	Restrictive cusp motion
Endocarditis	- Bicuspid valve - Myxomatous degeneration - Fenestrations - Disrupted commisure: • Dissection • Trauma	- Rheumatic valve disease - Degenerative calcification

Type Id **Type II** **Type III**

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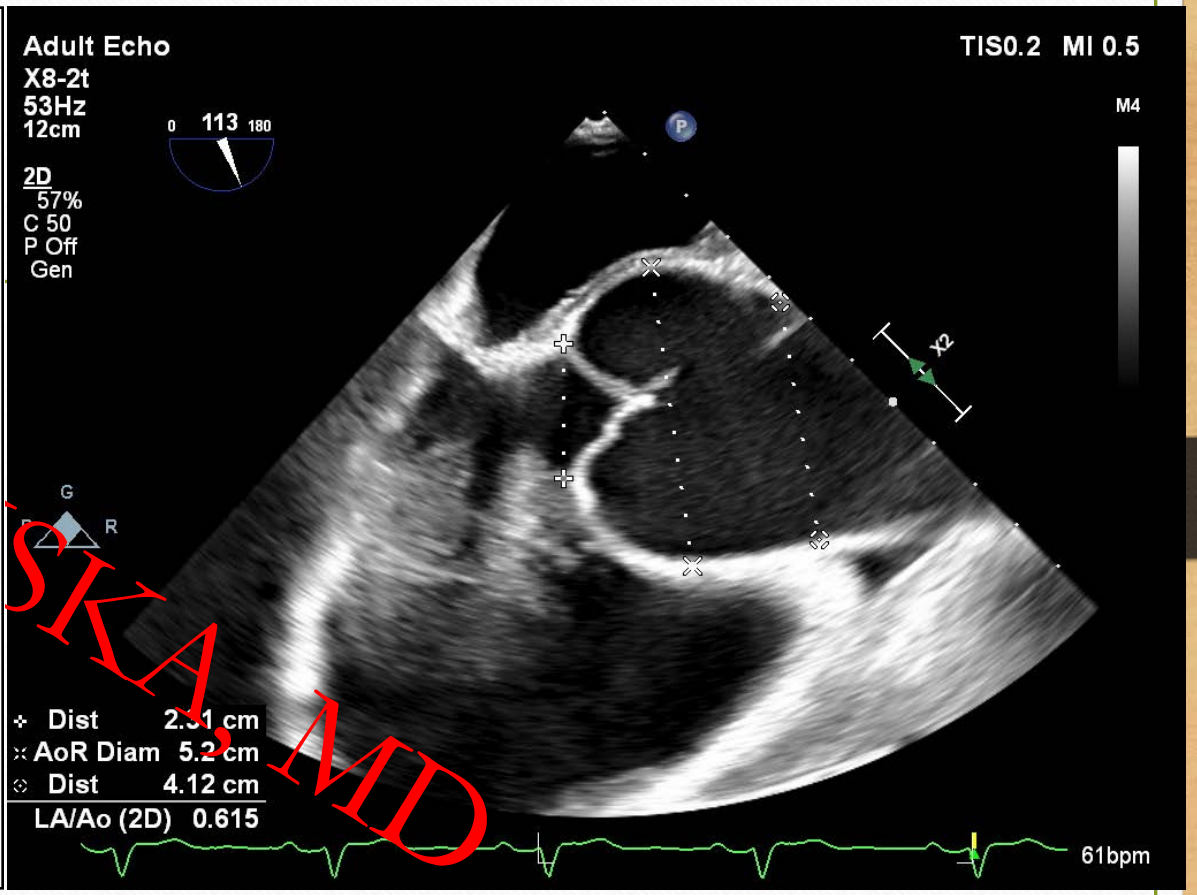
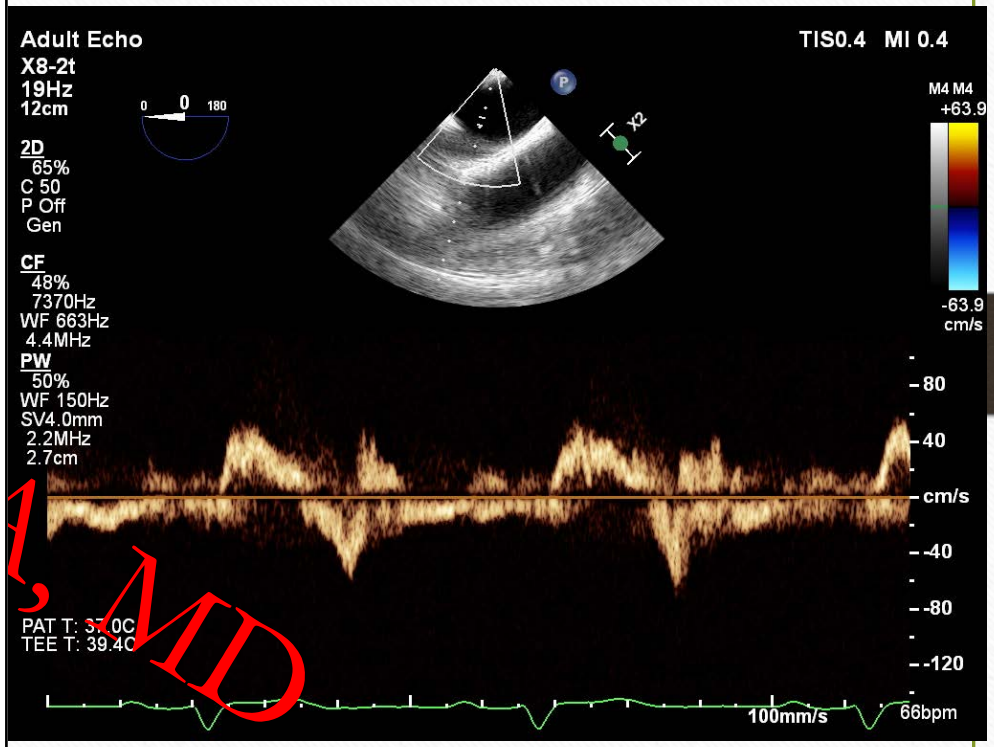

Adult Echo
X8-2t
53Hz
12cm
2D
57%
C 50
P Off
Gen
0 113 180
TIS0.2 MI 0.5
M4
x2
G
R
SKA, MD
Dist 2.31 cm
AoR Diam 5.2 cm
Dist 4.12 cm
LA/Ao (2D) 0.615
61bpm


Adult Echo
X8-2t
19Hz
12cm
2D
65%
C 50
P Off
Gen
CF
48%
7370Hz
WF 663Hz
4.4MHz
PW
50%
WF 150Hz
SV4.0mm
2.2MHz
2.7cm
TIS0.4 MI 0.4
M4 M4
+63.9
-63.9
cm/s
80
40
cm/s
-40
-80
-120
PAT T: 37.0C
TEE T: 39.4C
100mm/s
66bpm

Echo in AR – Number of cusps

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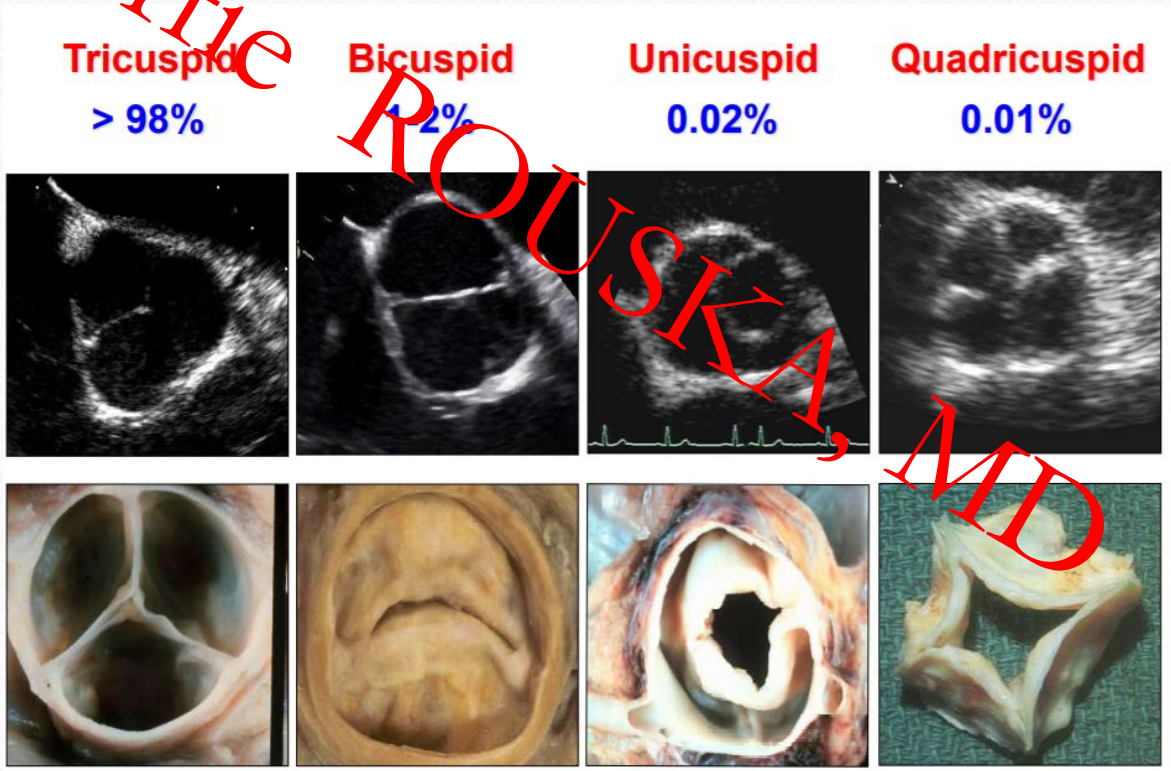

Echo in AR – Useful measurements

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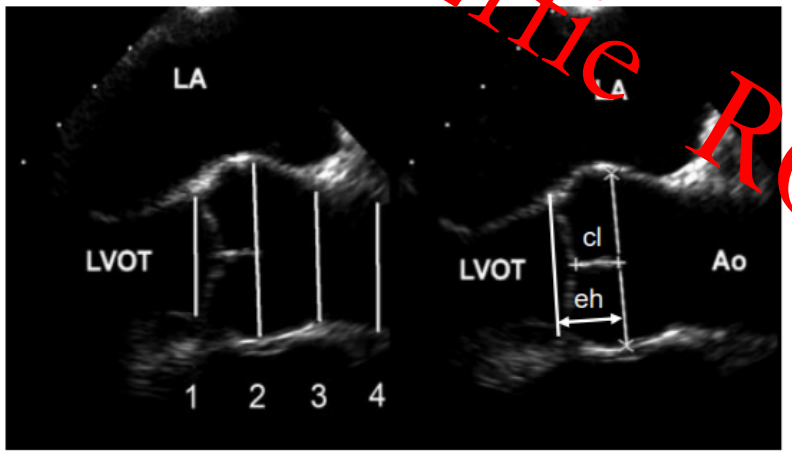
TEE long axis-view:



1: Annulus (≤ 1.6 cm/m²)

2: Sinus (≤ 2.1 cm/m²)

3: ST junction (≤ 1.9 cm/m²)

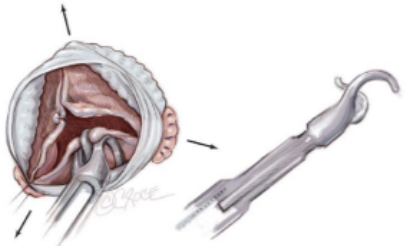
4: Tubular aorta

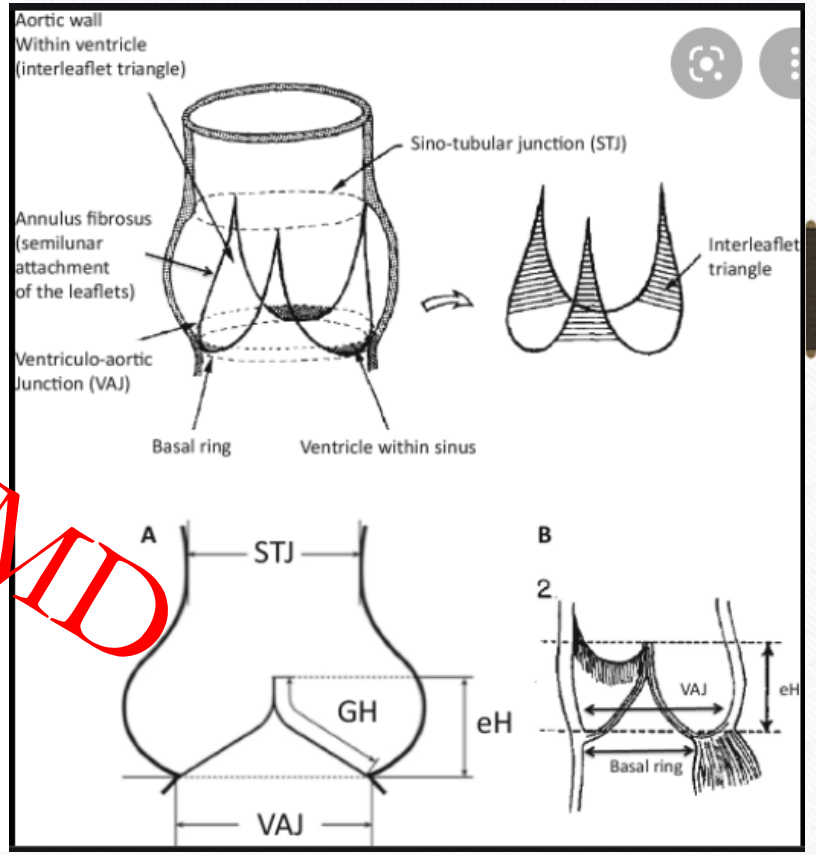
eh: effective height

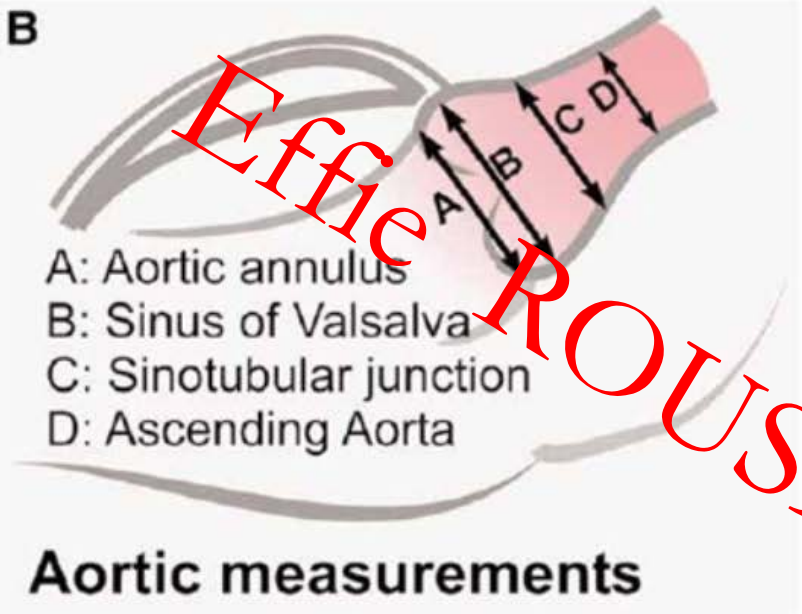
cl: coaptation length

Predictors of a durable repair:

- Absence of more than mild residual AR
- Effective height ≥ 9 mm
- Coaptation length ≥ 4 mm







Aortic dimensions Aortic diameters are proportional to body size and generally increase with aging	
Parasternal long axis view measurements	**Range (cm)**
Aortic annular diameter	1.4–2.6
Sinus of Valsalva	2.1–3.5
Sinotubular junction	1.7–3.4
Ascending aorta	2.1–3.4

Cardiac Ultrasound Laboratory, Massachusetts General Hospita

Effie ROUSKA, MD

Echo in AR – Useful measurements

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- Measurements are performed in the PLAX view(TTE) or in the ME 3ch view (TOE) **from leading edge to leading edge at end-diastole**, except for the Aortic Annulus which is measured in mid-systole.
- It is important to differentiate **three phenotypes of the ascending aorta**:

-AoRoot aneurysms (SOV>45mm)

-Tubular Ascending aneurysm (SOV<45mm)

-Isolated AR (all aortic diameters <40mm)

Echocardiographic criteria for severe AR

Table 5 Echocardiographic criteria for the definition of severe aortic valve regurgitation

Qualitative	
Valve morphology	Abnormal/flail/large coaptation defect
Colour flow regurgitant jet area[a]	Large in central jets, variable in eccentric jets
CW signal of regurgitant jet	Dense
Other	Holodiastolic flow reversal in descending aorta (EDV >20 cm/s)
Semiquantitative	
Vena contracta width (mm)	>6
Pressure half-time[b] (ms)	<200
Quantitative	
EROA (mm^2)	≥30
Regurgitant volume (mL/beat)	≥60
Enlargement of cardiac chambers	LV dilatation

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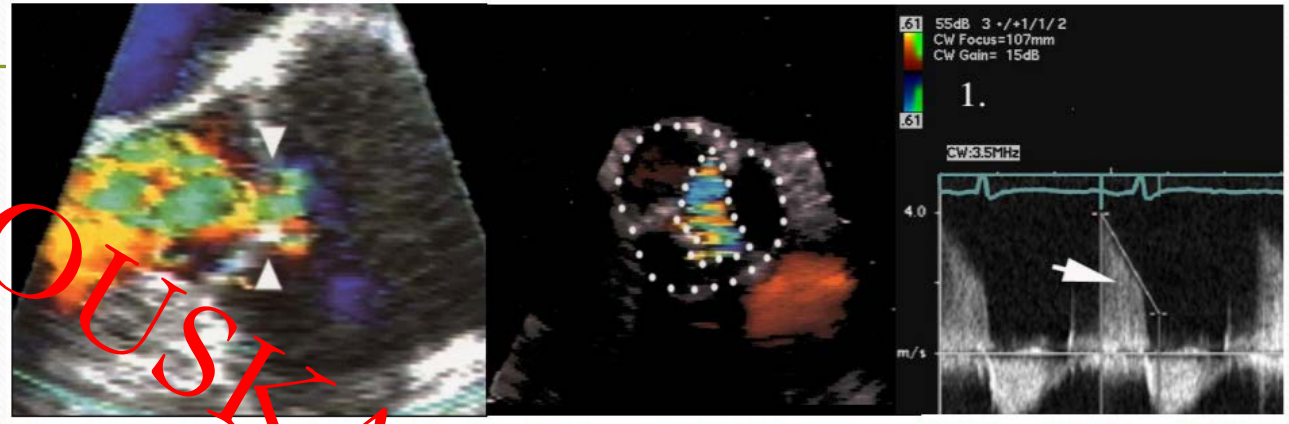

Echocardiographic criteria of severe aortic regurgitation (III.° - IV.°)

- Jet width ≥ 65% of LVOT
- Vena contracta width ≥ 6 mm
- Pressure half time (CW-Doppler) < 200 ms
- Holodiastolic flow reversal in descending aorta
- RV ≥ 60 ml, RF ≥ 50%, ERO ≥ 0.30 cm^2
- Left ventricular enlargement

SEVERITY

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- **1.** Regurgitant **jet width/LVOT diameter** ratio greater than or equal to 60 percent
- **2.** **Vena contracta** greater than 6 mm
- **3.** Regurgitant **jet area/LVOT area** ratio greater than or equal to 60 percent
- **4.** Aortic regurgitation **pressure half-time** less than or equal to 250 ms
- **5.** Holodiastolic **flow reversal** in the descending thoracic or abdominal aorta
- **6.** **Regurgitant volume** greater than or equal to 60 mL
- **7.** **Regurgitant fraction** greater than or equal to 50 percent
- **8.** **Effective regurgitant orifice** greater than or equal to 0.30cm2
- **9.** **Restrictive mitral flow pattern** (usually in acute setting)

- Regurgitant jet height measured as maximal diameter of regurgitant jet just below AV,PLAX view
- LVOT diameter in end diastole

	MILD	MODERATE	SEVERE
Jet width/LVOT diameter	<25%		>/=65%
Vena contracta	<3mm		>/=6mm
Jet area/LVOT area	<5%		>60%
PHT	>500 ms		</= 250ms
Holodiastolic flow reversal			present

	MILD	MODERATE	SEVERE
Reg vol	< 30 ml		>/= 60 ml
Reg fraction	< 30 %		>/= 50%
ERO	< 0.1 cm^2		>/= 0.3 cm^2
Mitral inflow restriction			Present

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Regurgitant jet width/LVOT diameter ratio greater than or equal to 60 percent

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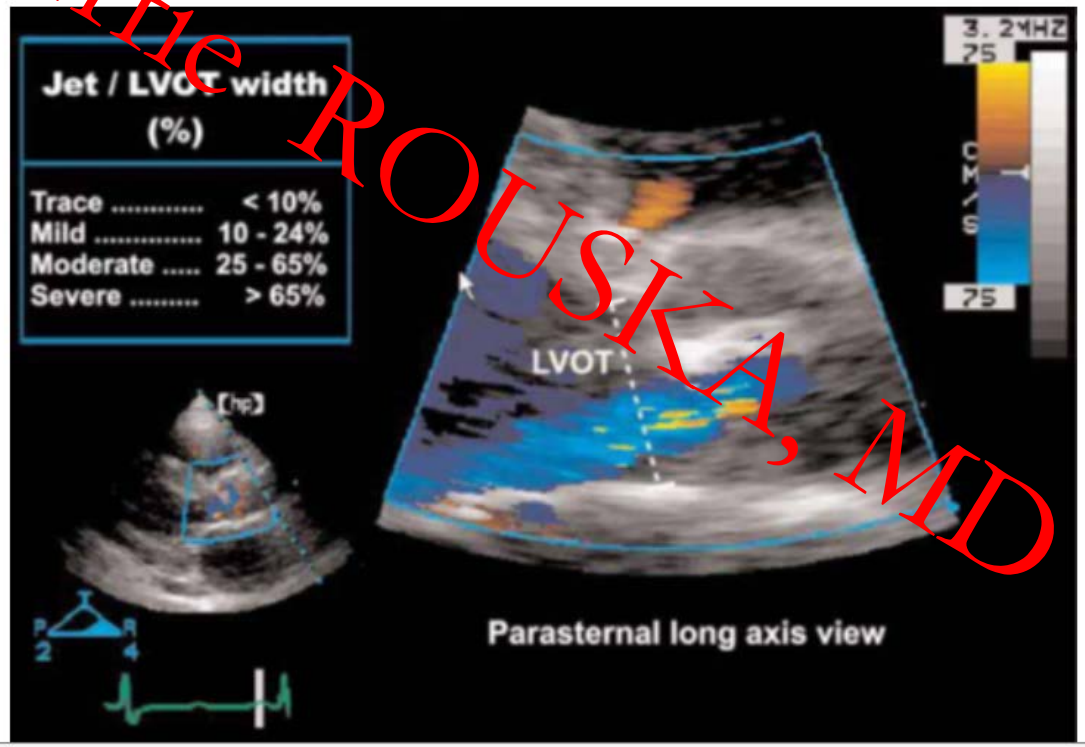
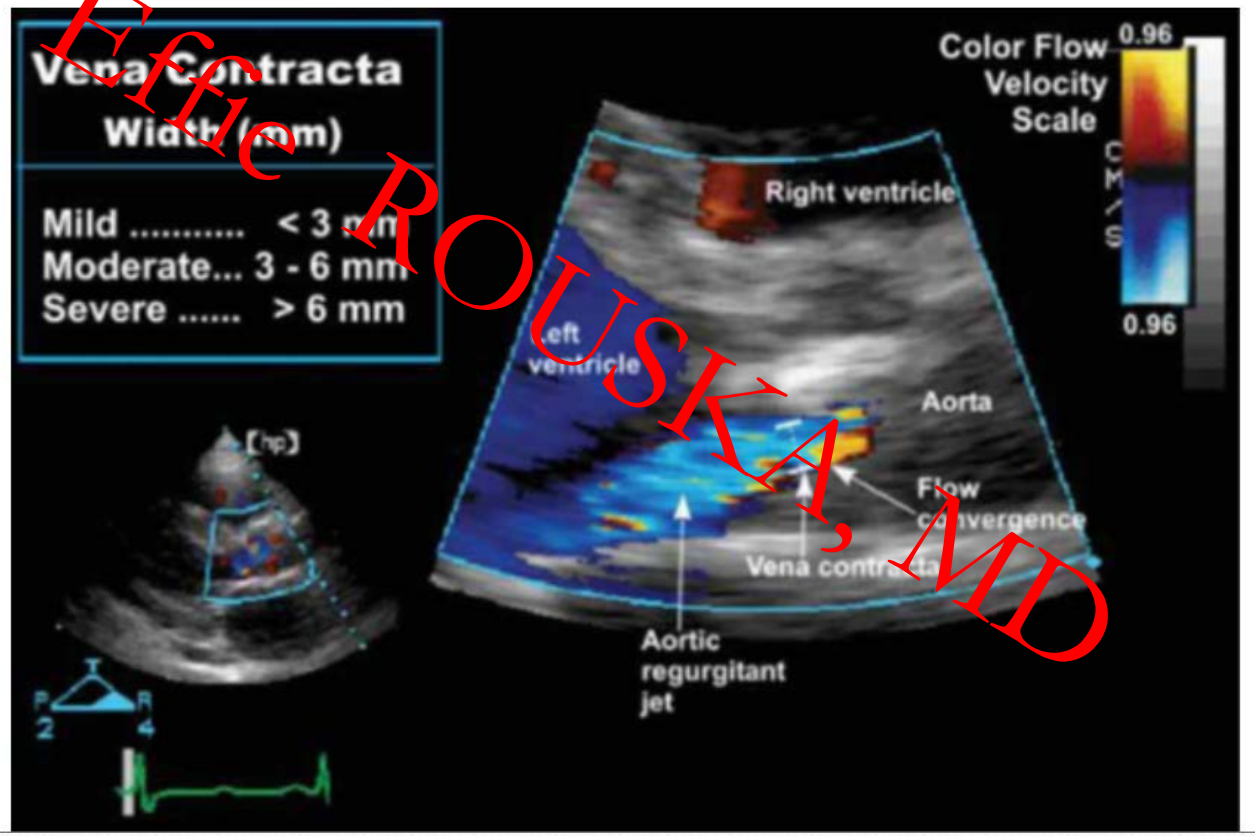


Vena contracta greater than 6 mm



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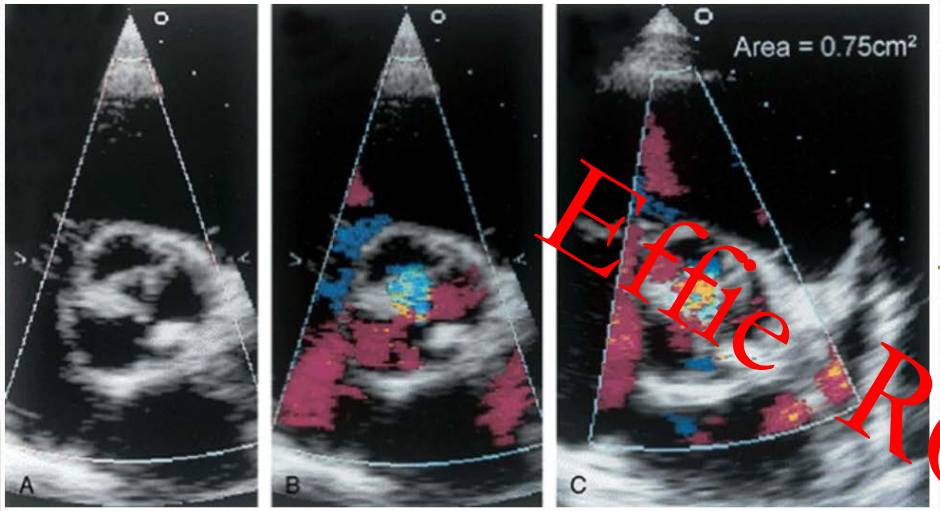

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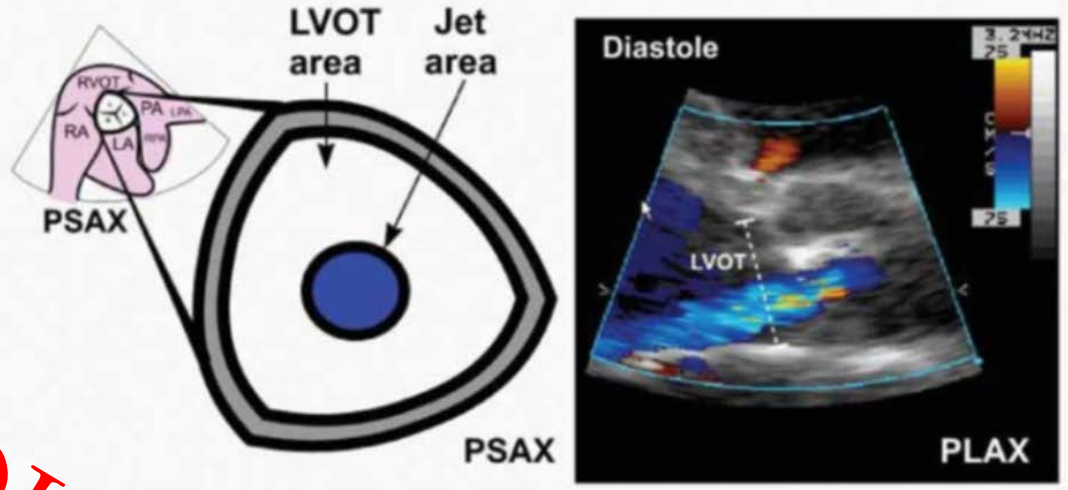
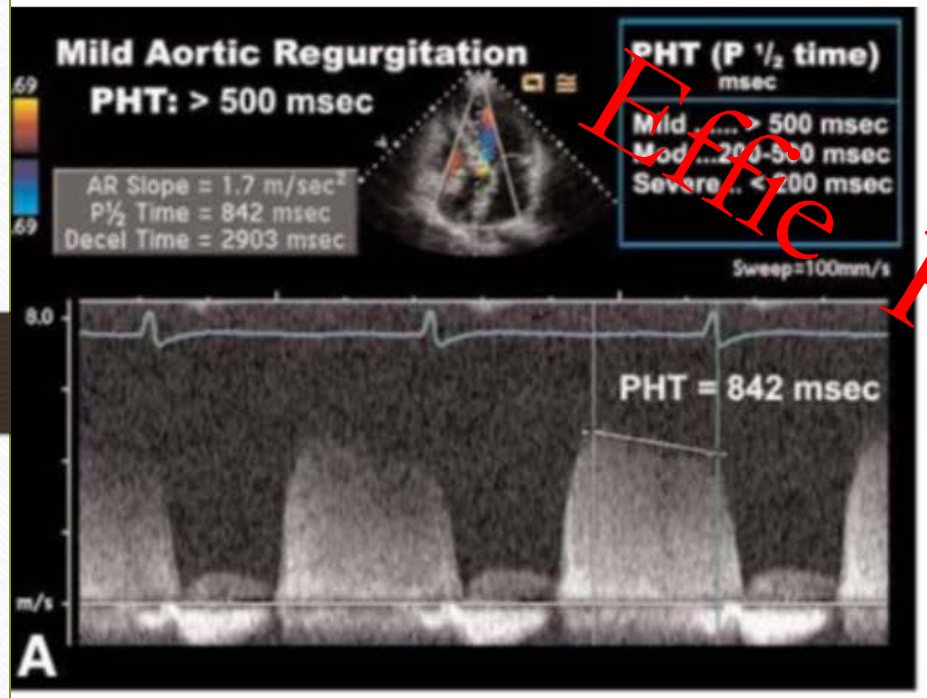


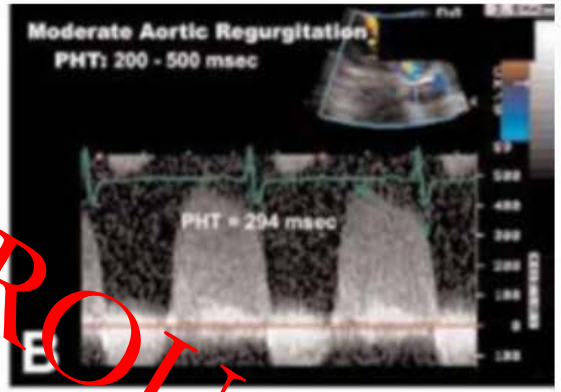
Jet / LVOT Area Ratio = $\frac{\text{Jet Area}}{(0.785 \times \text{LVOT diameter})^2}$

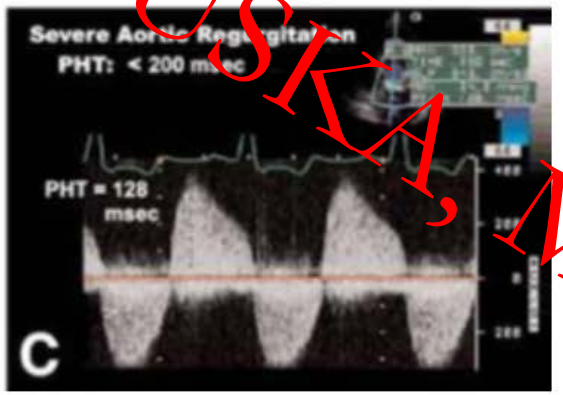
Simplified

Mild < 5%
Moderate 5–60%
Severe > 60%

- Regurgitant jet area measured from PSAX view at level of LVOT
- LVOA measured at end diastole at same site
- Ratio calculated







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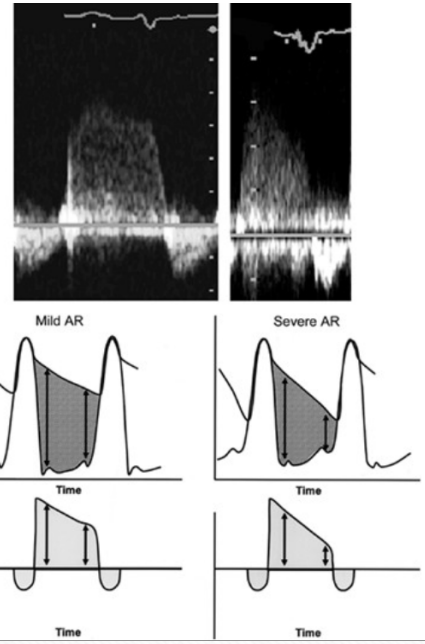
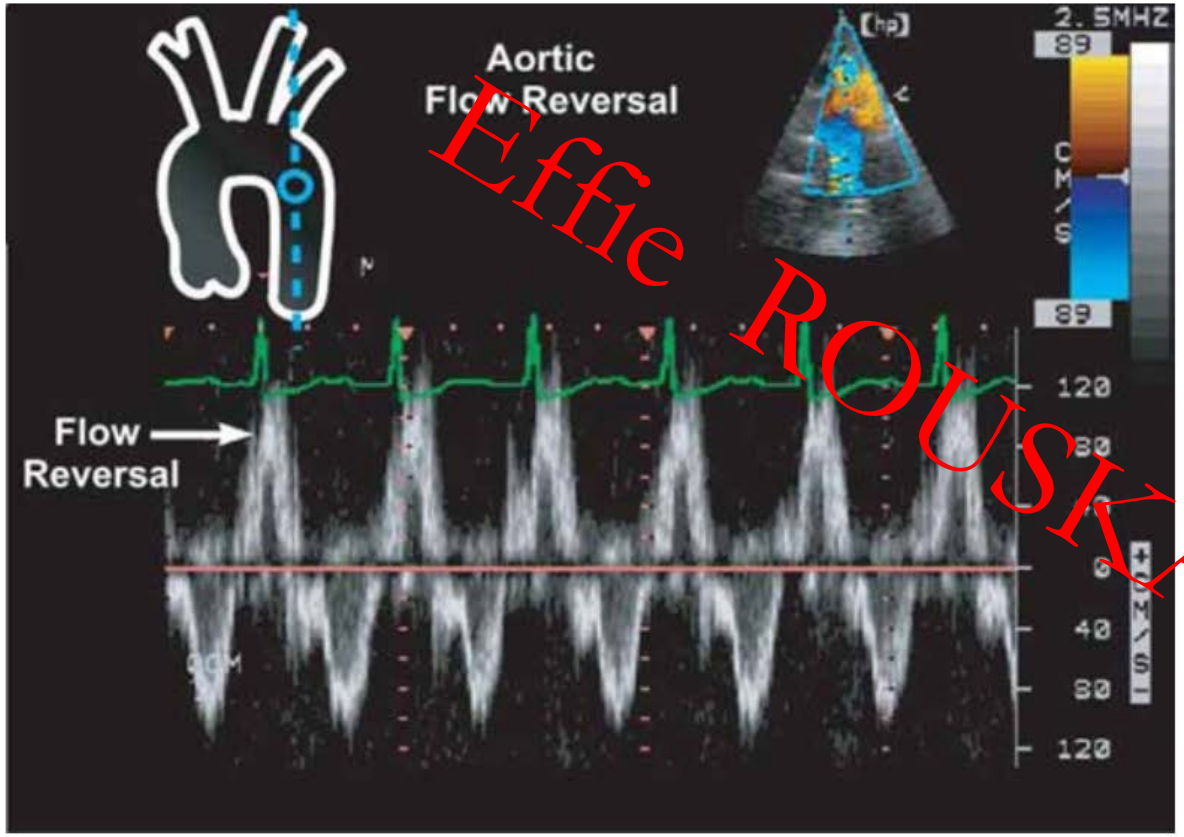
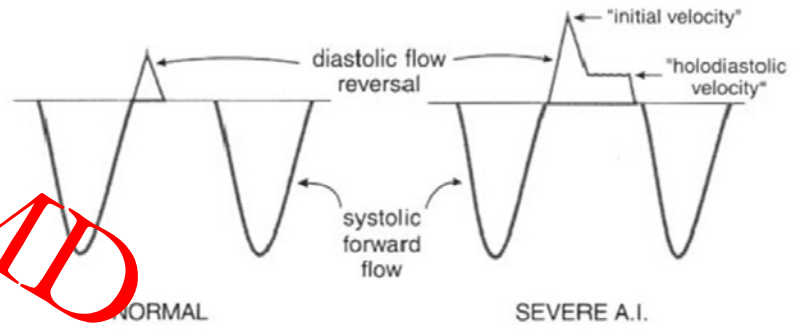
- Regurgitant doppler signal is a function of pressure gradient between aorta and LV
- Mild AR –small increase in LVEDP-gradual decline and flat deceleration slope
- Severe AR –LVEDP rises rapidly-rapid decline





- Suprasternal window-descending aortic flow profile
- Short period of low velocity flow reversal-normal
- Pan diastolic flow reversal with end diastolic velocity>20cm/s



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Calculation of R.Volume and R.fraction

- SV=CSAxVTI
- R.Volume=SV[lvot]-SV[mv]
- RF=R.Volume/SV[lvot]
- ERO=R.Volume/VTI[AR jet]
- R.V>60ml,RF>50%,ERO>0.3cm² indicate severe AR

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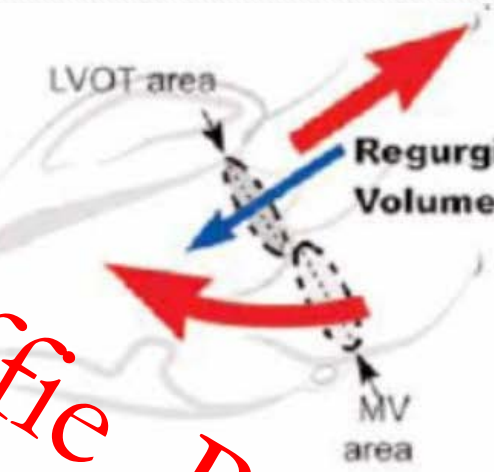
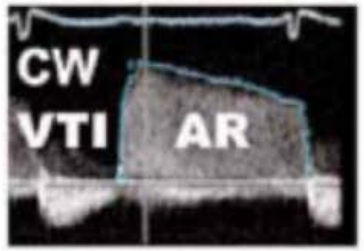


Regurgitant Volume (RV) = $SV_{AV} - SV_{MV}$



Effective Regurgitant Orifice Area (EROA) = Regurgitant Volume / VTI AR *CW Doppler*

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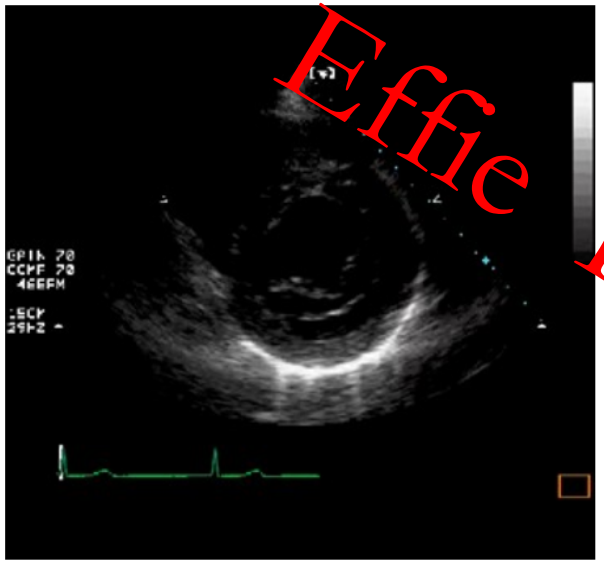
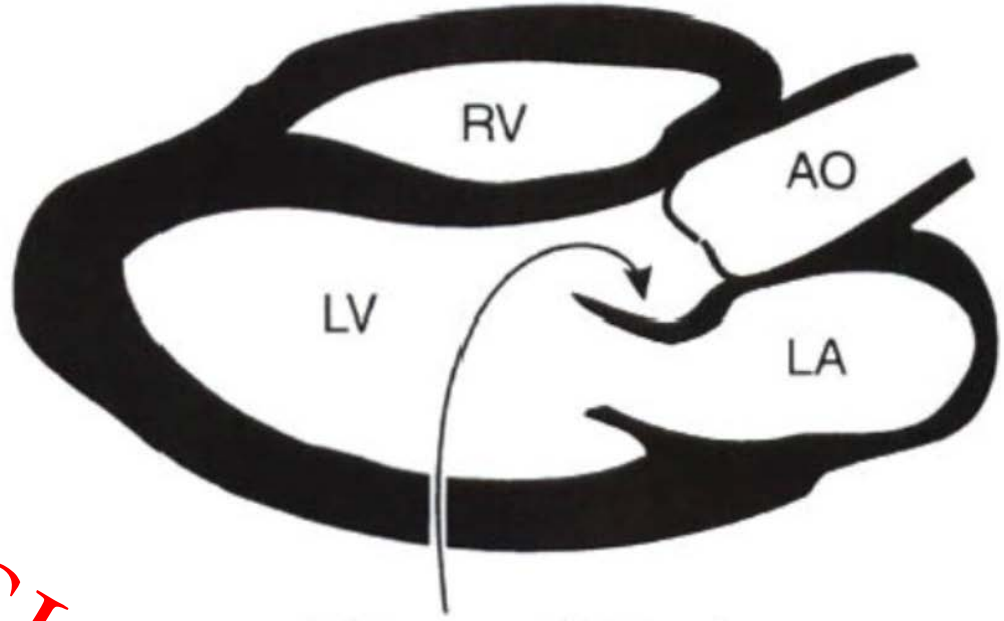
Regurgitation Severity (by EROA) mm^2

Mild	< 0.10 cm^2
Moderate	0.10 – 0.19 cm^2
Moderate - Severe	0.20 – 0.29 cm^2
Severe	> 0.30 cm^2

Restrictive mitral flow pattern





"Reversed" Doming

Fig. 7.26 Diagram of a 2-D image of a patient with AR. The regurgitant jet strikes the anterior leaflet of the mitral valve, not allowing it to open fully, giving the appearance of "*reversed doming*."

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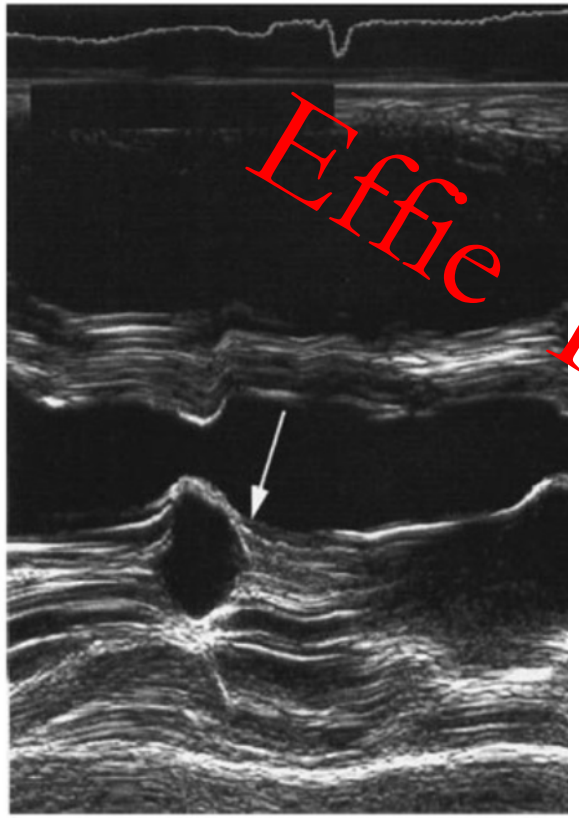

Fig. 7.24 M-mode of the mitral valve in a patient with severe AR demonstrates early mitral valve closure (arro secondary to a markedly elevated LVEDP.

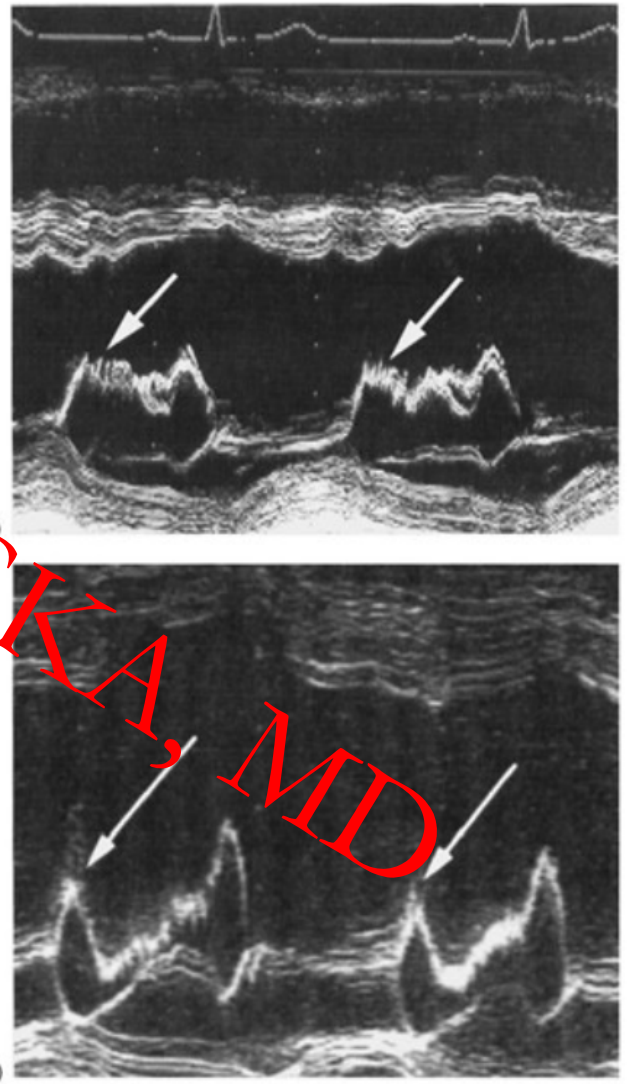

Fig. 7.25 Depressed E wave (arrows) noted by M-mode of the mitral valve (a & b) in two different patients with severe AR. Diastolic fluttering is also noted on the mitral valve leaflet.

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Extent of jet

- *Trivial*—just below aortic leaflets.
- *Mild*—LVOT.
- *Moderate*—AR extends to mitral leaflet level.
- *Severe*—body of the LV.

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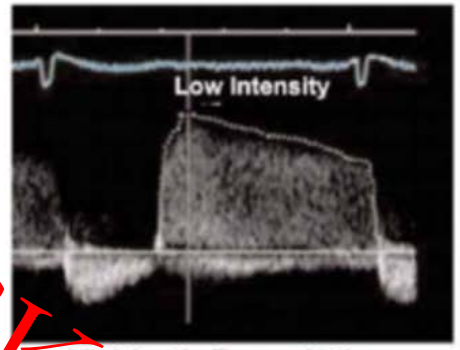
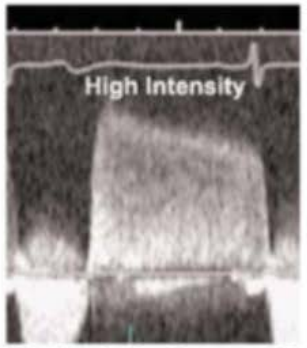
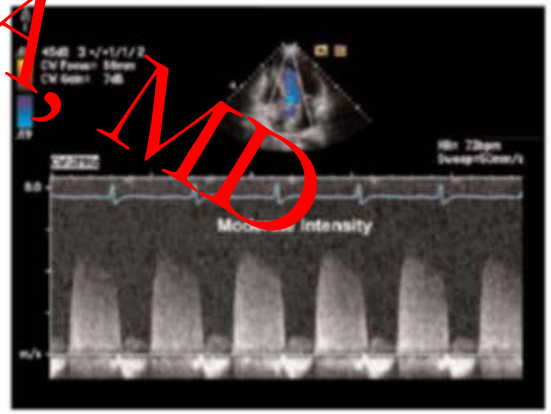
Signal intensity



Mild Aortic Regurgitation



Moderate to Severe AR



Acute vs Chronic AR

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ACUTE VS CHRONIC

- Shape of the envelope CW doppler
- Rate of deceleration of flow
- Premature mitral valve closure
- Endocarditis,dissection
- Normal lv dimensions

Recommendations on indications for surgery

in

(A)severe aortic regurgitation

&

(B) Aortic root or Tubular ascending aortic aneurysm

(*irrespective of the severity of aortic regurgitation*)

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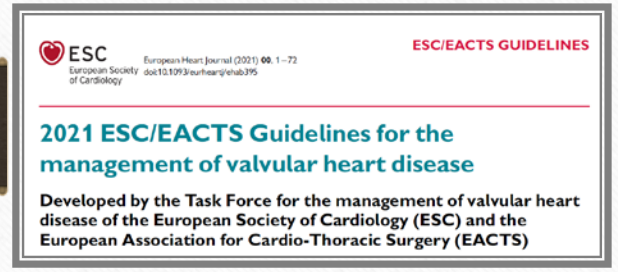

Indications for surgery	Class[a]	Level[b]
A) Severe aortic regurgitation		
Surgery is recommended in symptomatic patients regardless of LV function.[105–109]	I	B
Surgery is recommended in asymptomatic patients with LVESD >50 mm or LVESD >25 mm/m^2 BSA (in patients with small body size) or resting LVEF ≤50%.[107,108,112,114,115]	I	B
Surgery may be considered in asymptomatic patients with LVESD >20 mm/m^2 BSA (especially in patients with small body size) or resting LVEF ≤55%, if surgery is at low risk.	IIb	C
Surgery is recommended in symptomatic and asymptomatic patients with severe aortic regurgitation undergoing CABG or surgery of the ascending aorta or of another valve.	I	C
Aortic valve repair may be considered in selected patients at experienced centres when durable results are expected.	IIb	C
B) Aortic root or tubular ascending aortic aneurysm[c] (irrespective of the severity of aortic regurgitation)		
Valve-sparing aortic root replacement is recommended in young patients with aortic root dilation, if performed in experienced centres and durable results are expected.[133–136,140]	I	B
Ascending aortic surgery is recommended in patients with Marfan syndrome who have aortic root disease with a maximal ascending aortic diameter ≥50 mm.	I	C
Ascending aortic surgery should be considered in patients who have aortic root disease with maximal ascending aortic diameter: • ≥55 mm in all patients. • ≥45 mm in the presence of Marfan syndrome and additional risk factors[d] or patients with a *TGFBR1* or *TGFBR2* mutation (including Loeys–Dietz syndrome).[e] • ≥50 mm in the presence of a bicuspid valve with additional risk factors[d] or coarctation.	IIa	C
When surgery is primarily indicated for the aortic valve, replacement of the aortic root or tubular ascending aorta should be considered when ≥45 mm.[f]	IIa	C

Indications for surgery	Class[a]	Level[b]
A) Severe aortic regurgitation		
Surgery is recommended in symptomatic patients regardless of LV function.[105–109]	I	B
Surgery is recommended in asymptomatic patients with LVESD >50 mm or LVESD >25 mm/m^2 BSA (in patients with small body size) or resting LVEF ≤50%.[107,108,112,114,115]	I	B
Surgery may be considered in asymptomatic patients with LVESD >20 mm/m^2 BSA (especially in patients with small body size) or resting LVEF ≤55%, if surgery is at low risk.	IIb	C
Surgery is recommended in symptomatic and asymptomatic patients with severe aortic regurgitation undergoing CABG or surgery of the ascending aorta or of another valve.	I	C
Aortic valve repair may be considered in selected patients at experienced centres when durable results are expected.	IIb	C

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B) Aortic root or tubular ascending aortic aneurysm[c] (irrespective of the severity of aortic regurgitation)		
Valve-sparing aortic root replacement is recommended in young patients with aortic root dilation, if performed in experienced centres and durable results are expected.[133–136,140]	I	B
Ascending aortic surgery is recommended in patients with Marfan syndrome who have aortic root disease with a maximal ascending aortic diameter ≥50 mm.	I	C
Ascending aortic surgery should be considered in patients who have aortic root disease with maximal ascending aortic diameter: • ≥55 mm in all patients. • ≥45 mm in the presence of Marfan syndrome and additional risk factors[d] or patients with a *TGFBR1* or *TGFBR2* mutation (including Loeys–Dietz syndrome).[e] • ≥50 mm in the presence of a bicuspid valve with additional risk factors[d] or coarctation.	IIa	C
When surgery is primarily indicated for the aortic valve, replacement of the aortic root or tubular ascending aorta should be considered when ≥45 mm.[f]	IIa	C

Effie ROUSKA, MD

Pre-op ITEE evaluation of AR

- *Repairability of AR*
  - **Criteria for repair**
    - Bicuspid valves with prolapse
    - Tricuspid valves with one cusp prolapse
    - Dilatation of aortic annulus / root
    - Perforation of aortic cusps
    - Aortic dissection
  - **Exclusion criteria for repair**
    - Fibrosis, thickening, calcification

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Grim RA, Stewart WJ. Cardiology Clinics 1998;16:477

Repairability of AR

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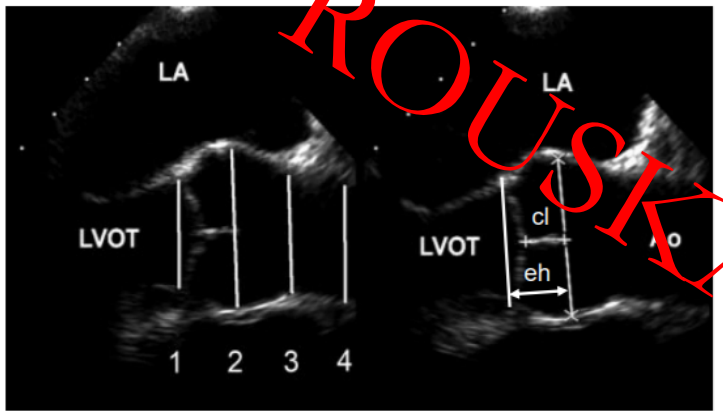
TEE long axis-view:



1: Annulus (≤ 1.6 cm/m²)

2: Sinus (≤ 2.1 cm/m²)

3: ST junction (≤ 1.9 cm/m²)

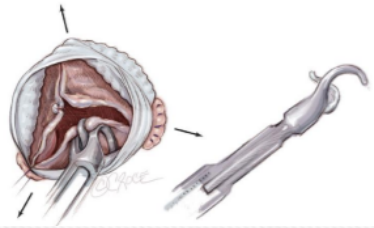
4: Tubular aorta

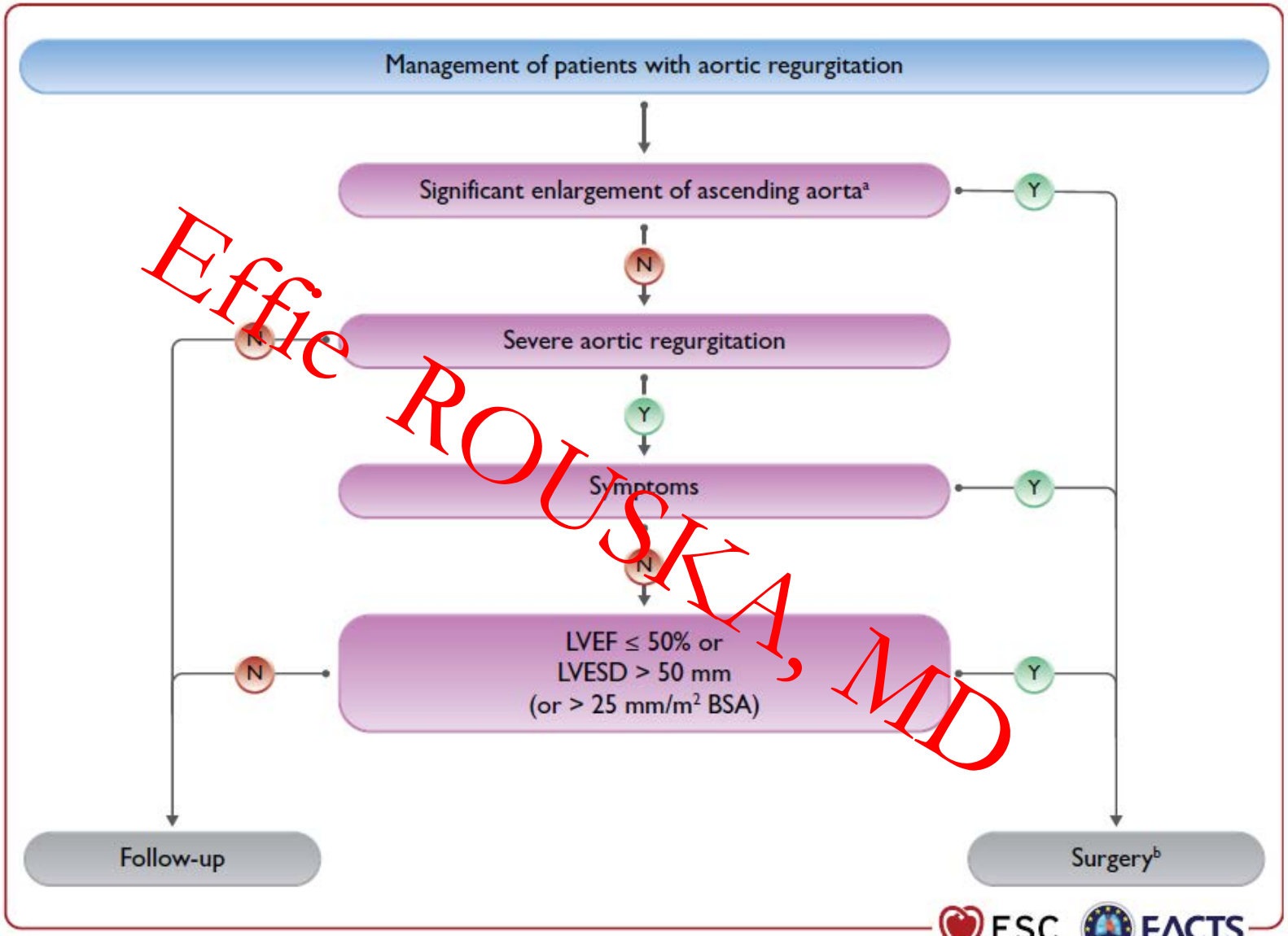
eh: effective height

cl: coaptation length

Predictors of a durable repair:

- Absence of more than mild residual AR
- Effective height ≥ 9 mm
- Coaptation length ≥ 4 mm





Effie ROUSKA, MD

Medical Therapy

Effie ROUSKA, MD

4.3 Medical therapy

Medical therapy, especially angiotensin-converting enzyme inhibitors (ACEI) or dihydropiridines, may provide symptomatic improvement in individuals with chronic severe aortic regurgitation in whom surgery is not feasible. The value of ACEI or dihydropiridine in delaying surgery in the presence of moderate or severe aortic regurgitation in asymptomatic patients has not been established and their use is not recommended for this indication.

In patients who undergo surgery but continue to suffer from heart failure or hypertension, ACEI, angiotensin receptor blockers (ARBs), and beta-blockers are useful.[141,142]

In patients with Marfan syndrome, beta-blockers remain the mainstay for medical treatment and reducing shear stress and aortic growth rate and should be considered before and after surgery.[143–145] While ARBs did not prove to have a superior effect when compared to beta-blockers, they may be considered as an alternative in patients intolerant to beta-blockers.[146–148] By analogy, while there are no studies that provide supporting evidence, it is common clinical practice to advise beta-blocker or ARBs in patients with bicuspid aortic valve if the aortic root and/or ascending aorta is dilated. Management of aortic regurgitation during pregnancy is discussed in section 13.

Serial Testing

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- **AR** (asymptomatic pts / severe AR / normal LV – FU every year)
  (pts with 1st diagnosis / or LV compromise – FU every 3-6 months)

- **Asc Aorta dilatation (>40mm)** – any increase >3mm should be validated by CCT angiography/CMR & compared to baseline

- After repair, **Marfan pts** remain at risk for dissection of the residual Aorta
  (lifelong regular FU at an expert centre is required)

Special patient populations

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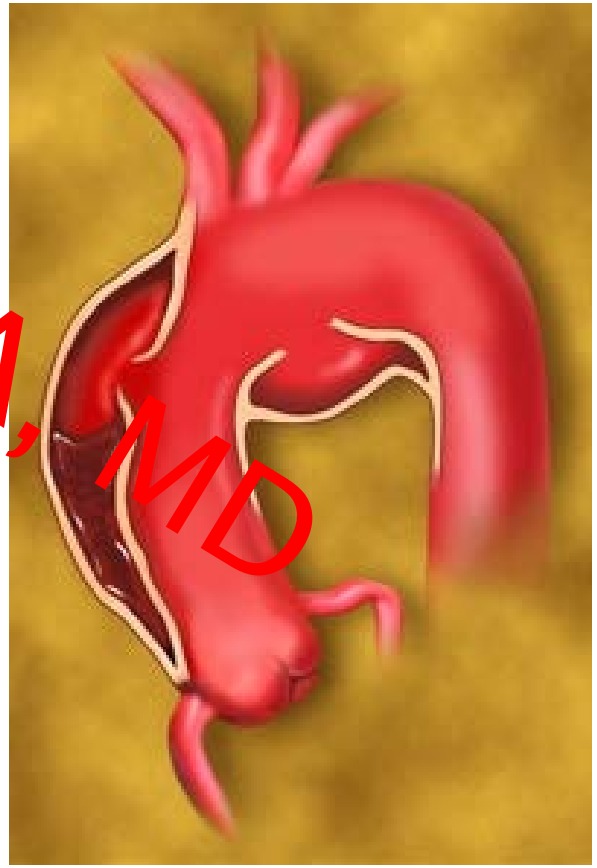
- In pts with **moderate AR who undergo CABG or MV surgery,** the decision to treat the AoV is controversial (the Heart Team should decide based on the aetiology, clinical factors, life expectancy & pt's operative risk).
- The level of **physical & sports activity in the presence of a dilated aorta** remains a matter of clinical judgement in the absence of evidence.
- Given the familial risk of **thoracic Aortic Aneurysms, screening and referral for genetic testing of the pt's 1st degree relatives** with appropriate imaging is indicated in pts with connective tissue disease.
- For pts with **BAV, pt's 1st degree relatives** appropriate to have an Echo screening.

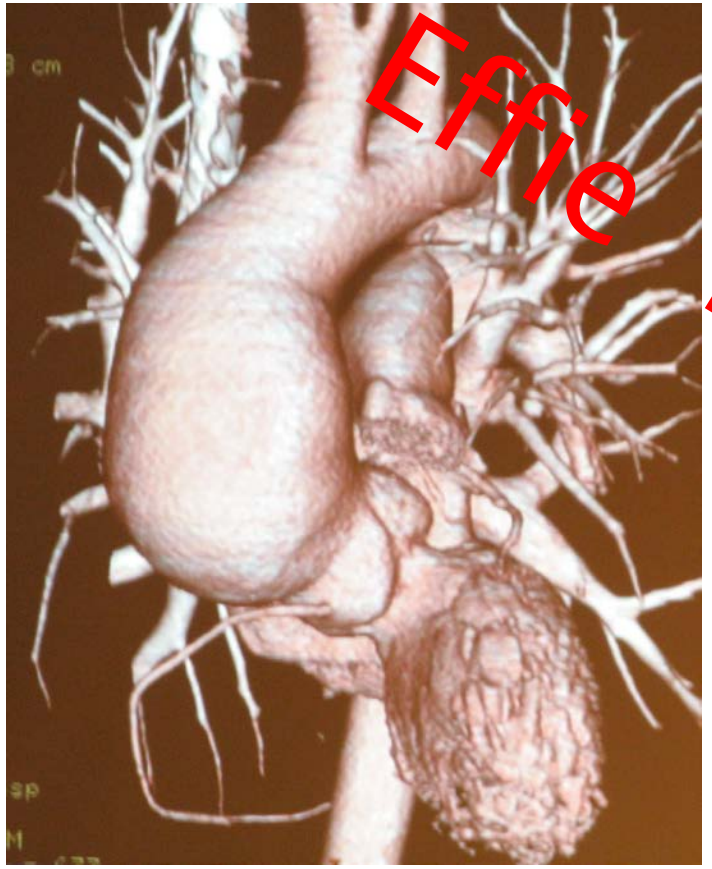
ΑΝΕΥΡΥΣΜΑ – ΔΙΑΧΩΡΙΣΜΟΣ

- Δυο διαφορετικές νοσολογικές οντότητες
  - Παθοφυσιολογία
  - Θεραπευτική προσέγγιση

Effie ROUSKA, MD



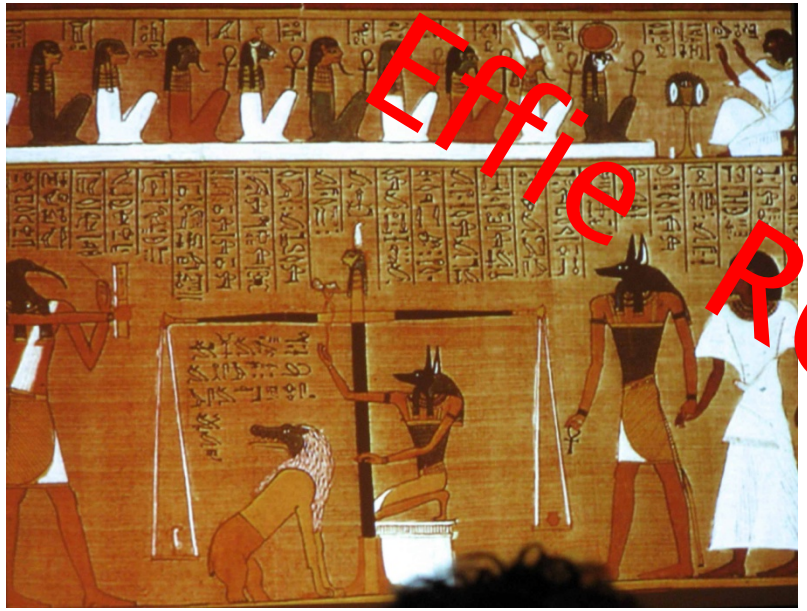
ΑΝΕΥΡΥΣΜΑ ΑΝΙΟΥΣΗΣ ΑΟΡΤΗΣ



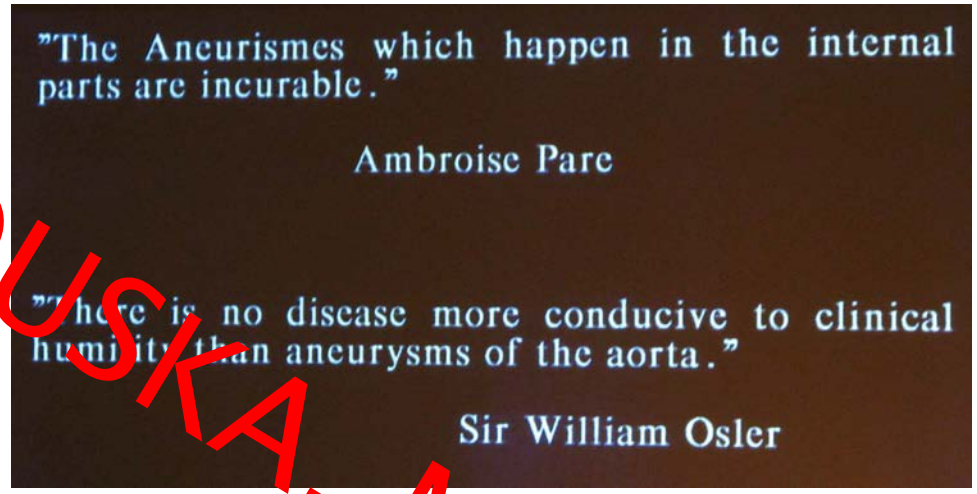
Effie ROUSKA, MD

- *Ιστορική αναδρομή*
- *Ορισμός*
- *Εμφάνιση*
- *Παράγοντες κινδύνου*
- *Παθογένεια*
- *Αιτιολογία*
- *Φυσική εξέλιξη*
- *Κλινική Διάγνωση*
- *Απεικονιστικές μέθοδοι*
- *Συντηρητική Θεραπευτική προσέγγιση*
- *Παρακολούθηση*

Ιστορική αναδρομή



Αιγυπτιακός ιατρικός
Πάπυρος Ebers (1552 πΧ)



Γάλλος χειρουργός
Ambroise Paré (1510–1590 μΧ)

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Ορισμός

- Μόνιμη τοπική διάταση της αορτής, διαμέτρου ≥ 50% της κανονικής

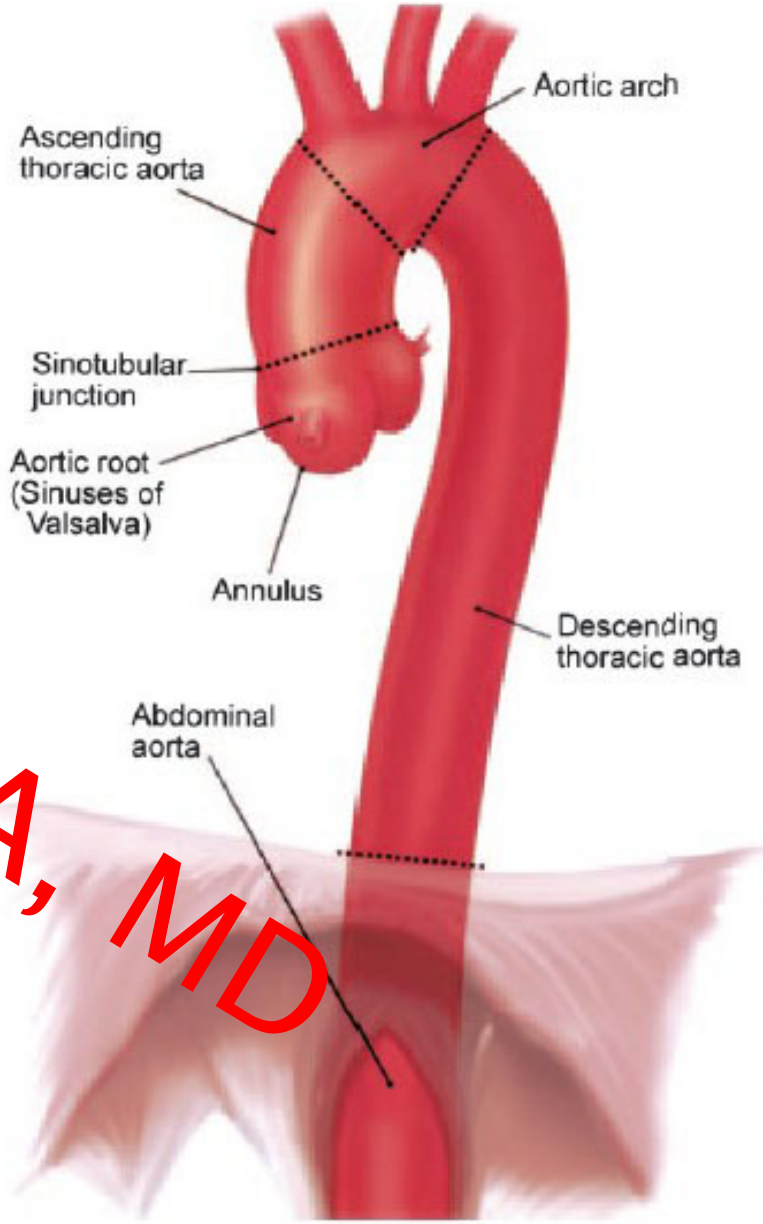
- Το αορτικό τοίχωμα πρέπει να αποτελείται και από τους τρεις χιτώνες του

Effie ROUSKA, MD

Τοπογραφική κατανομή

- το 60% αφορά την αορτική ρίζα ή/και την ανιούσα αορτή
- το 40% αφορά την κατιούσα θωρακική αορτή
- το 10% αφορά το αορτικό τόξο
- το 10% αφορά όλη την αορτή

Effie ROUSKA, MD



Anatomy of thoracic and proximal abdominal aorta.

Ατρακτοειδή – Σακοειδή

- *ατρακτοειδή* εκτείνονται σε όλη την περίμετρο
- *σακοειδή* σε μέρος της περιμέτρου

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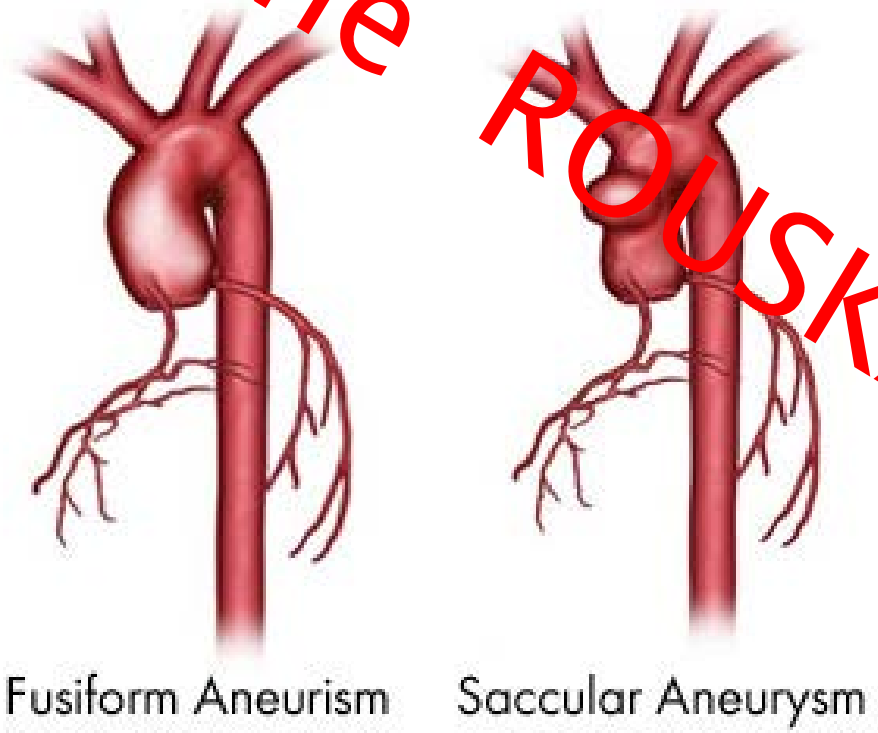
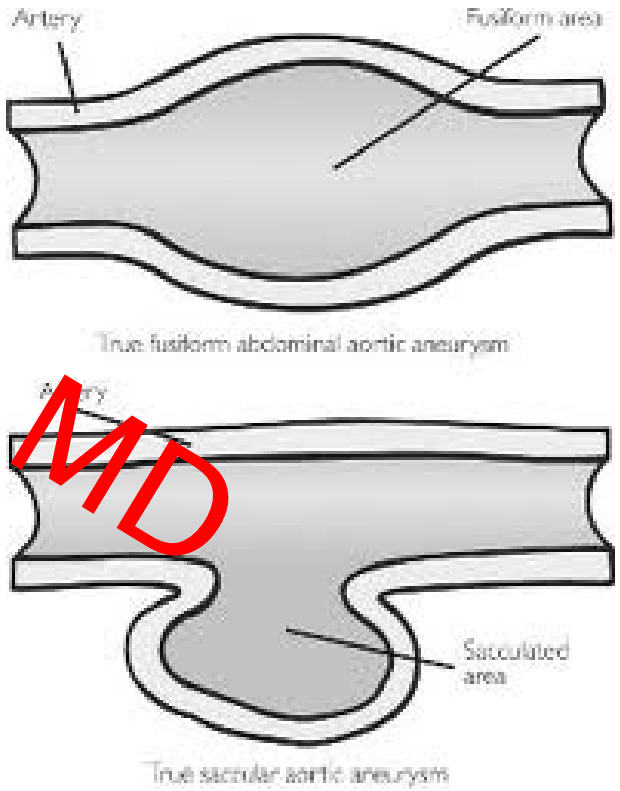




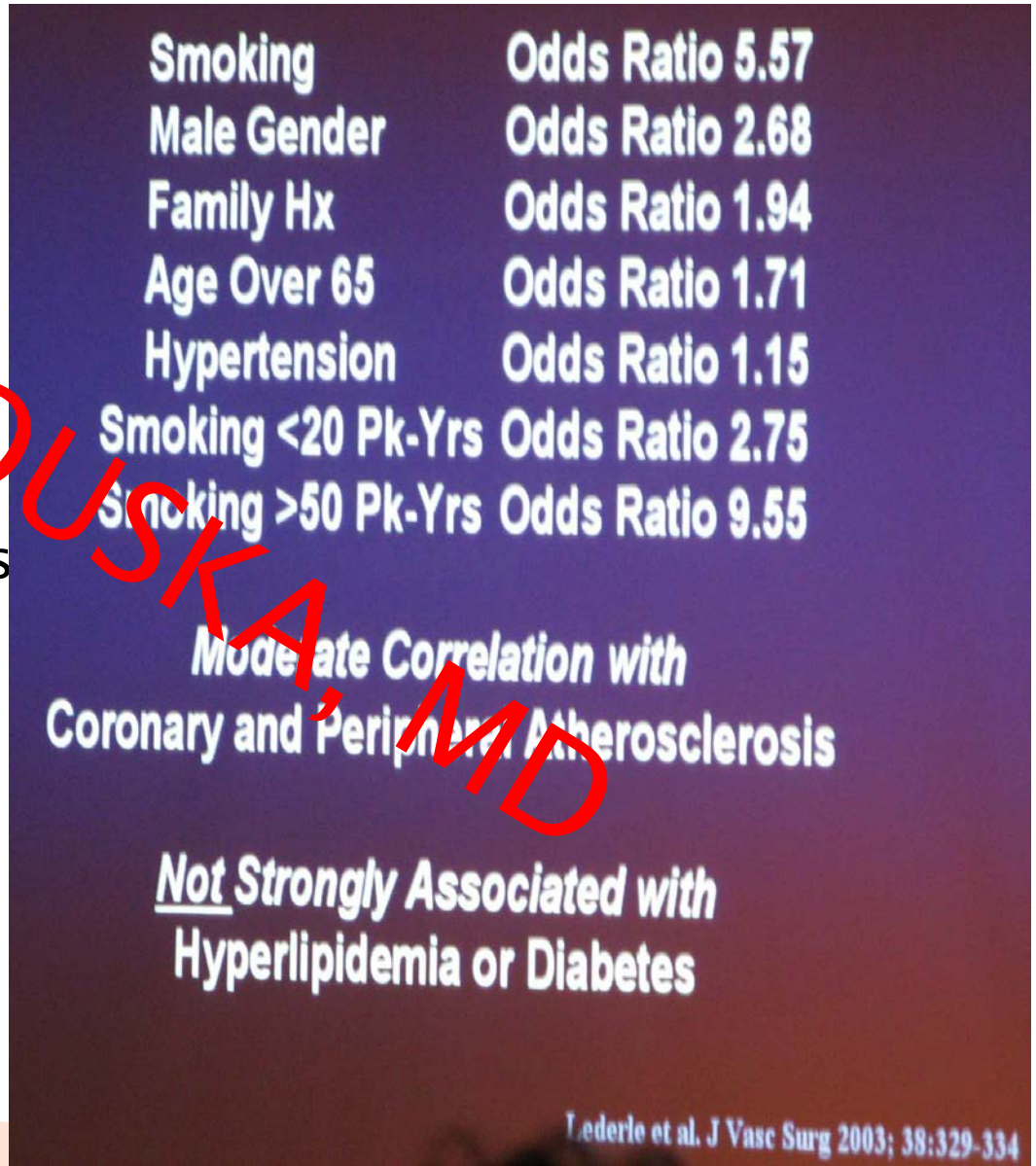
Εμφάνιση

- *13η αιτία θανάτου στις ΗΠΑ*
- 5.9 νέες περιπτώσεις ανά 100.000 ατόμων ανά έτος
- μέση ηλικία εμφάνισης κυμαίνεται μεταξύ του 59 και 69 έτους της ηλικίας
- Στους άνδρες εμφανίζεται σε νεαρότερη ηλικία καθώς και συχνότερα από τις γυναίκες (2:1 – 4:1)

Effie ROUSKA, MD

Παράγοντες κινδύνου

- Κάπνισμα
- Υπέρταση
- Αθηροσκλήρωση
- Γενετικές ανωμαλίες
  - σύνδρομο Marfan
  - σύνδρομο Ehlers-Danlos
- Δίπτυχη AoV
- Σύφιλη



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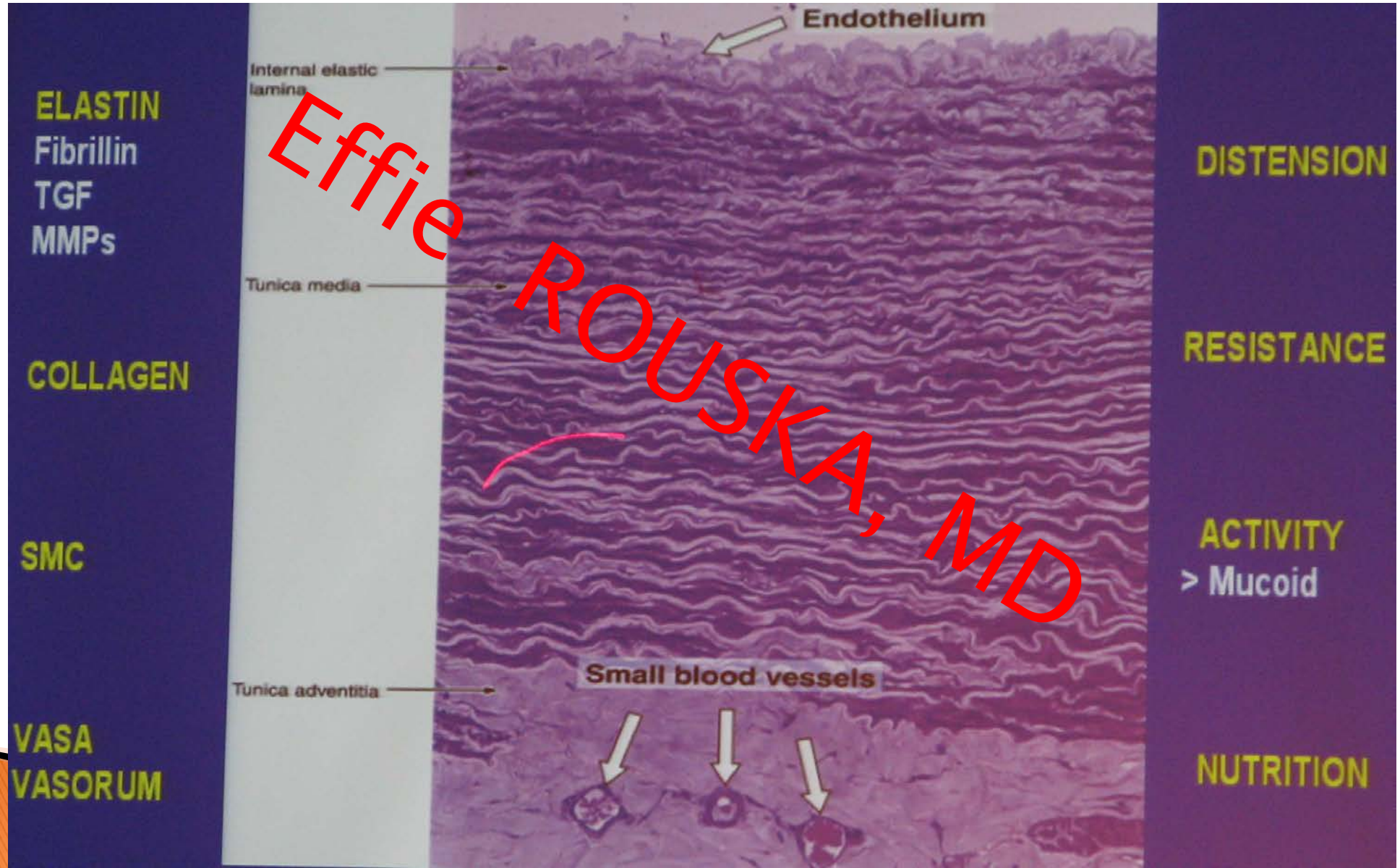
STRUCTURE

NORMAL AORTA

FUNCTION



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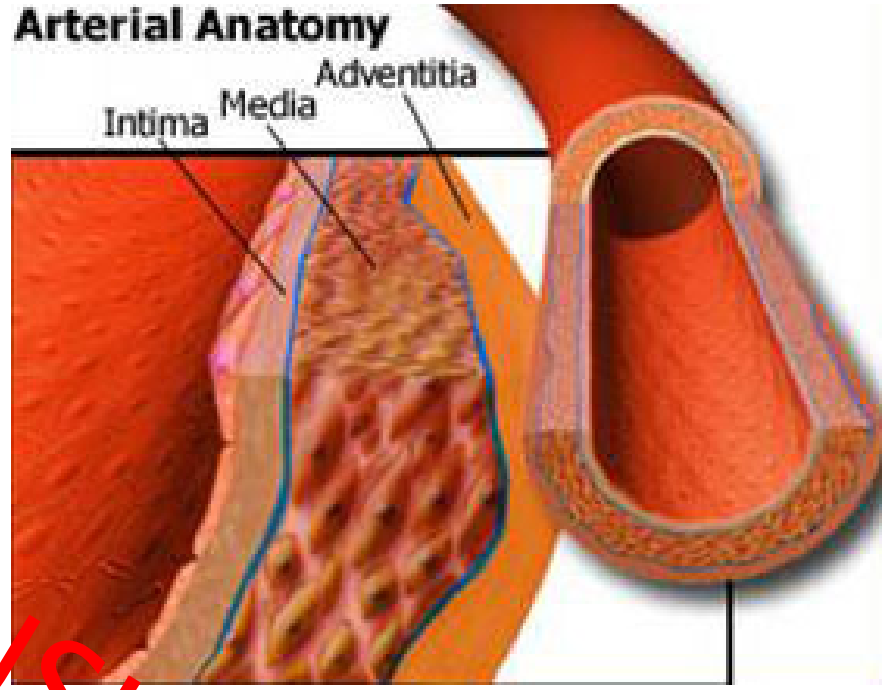
- ***Μέσος χιτώνας***
  - Αποτελείται από:
    - ελαστίνη,
    - κολλαγόνο
    - λείες μυϊκές ίνες



Effie ROUSKA, MD

- ***Ιδιότητες***
  - Ελαστικότητα (elasticity)
  - Αντοχή εφελκυσμού (tensile strength)

Παθογένεια

- ***Κυστική εκφύλιση μέσου χιτώνα***
  - εκφύλιση των ελαστικών ινών και νέκρωση των λείων μυϊκών ινών
  - αντικατάσταση τους από βασεόφιλο βλεννώδες άμορφο υλικό,
  - εικόνα κύστης του μέσου χιτώνα

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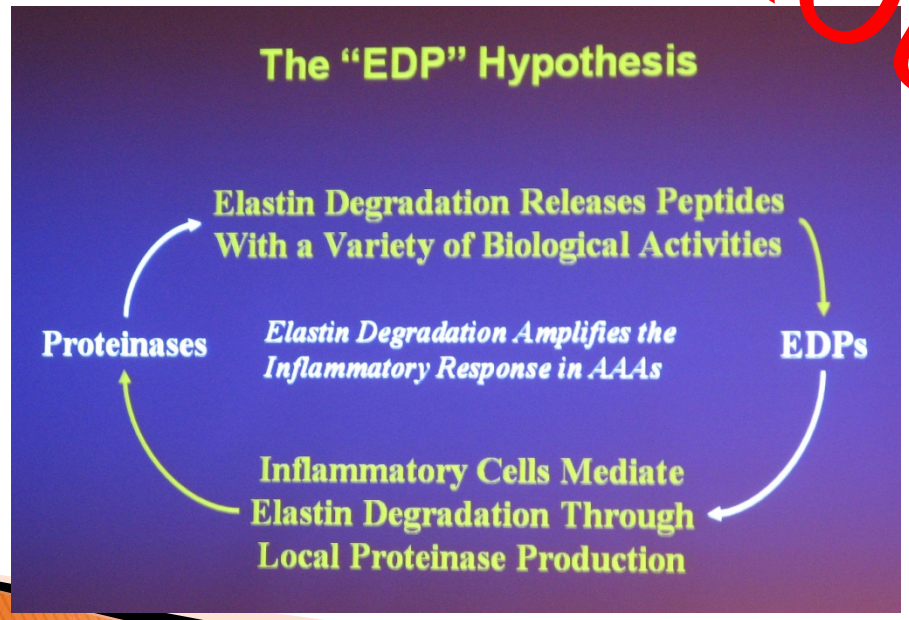

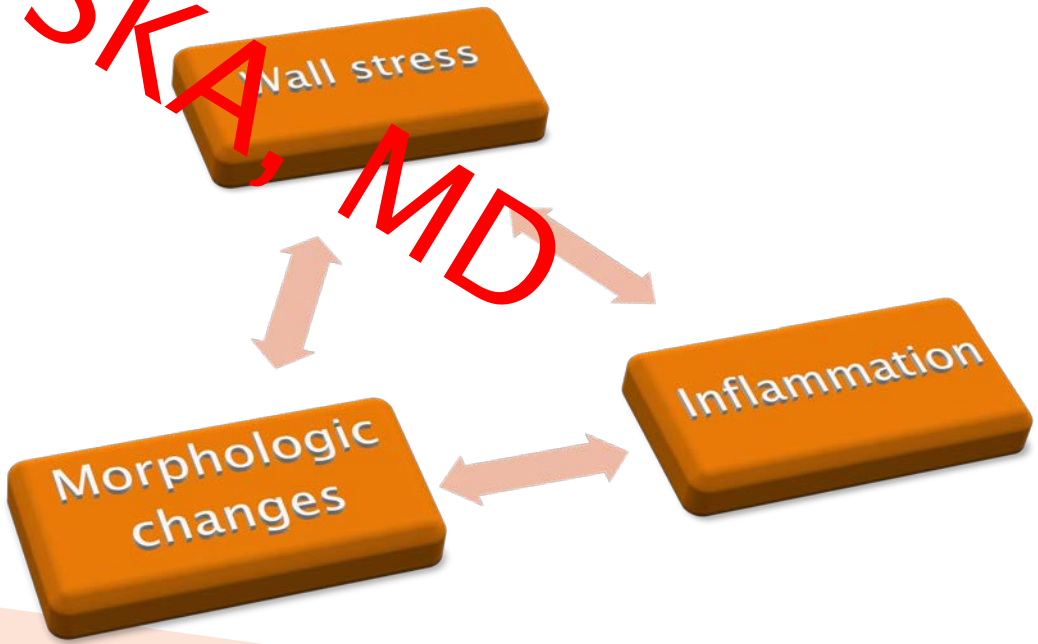

- ***Μηχανικές λειτουργίες της αορτής***
  - Απορροφά και αποθηκεύει ενέργεια κατά την συστολή
  - Αποδίδει ξανά την ενέργεια κατά τη διαστολή
  - Εξισορροπεί την πίεση παλμού
  - Αποφορτίζει την αριστερή κοιλία

- ***Μορφολογικές αλλαγές αορτής σε ανεύρυσμα***
  - Μικρότερη ενδοτικότητα
  - Λιγότερη ελαστικότητα
  - Λέπτυνση του τοιχώματος
  - Αύξηση της τοιχωματικής τάσης

Effie ROUSKA, MD

Αιτιολογία

- Ιδιοπαθής κυστική εκφύλιση μέσου χιτώνα
- Κληρονομικές διαταραχές
  - *σύνδρομο Marfan*
  - *σύνδρομο Ehlers-Danlos*
  - *οικογενής κατανομή*
  - *δίπτυχη αορτική βαλβίδα*
- Αθηροσκλήρυνση
- Ανευρύσματα σχετιζόμενα με Ao Διαχωρισμό
- Ανευρύσματα σχετιζόμενα με στένωση της AoV
- Ανευρύσματα σχετιζόμενα με λοιμώξεις
  - *μυκωτικά ανευρύσματα*
  - *συφιλιδικά ανευρύσματα*
- Ανευρύσματα σχετιζόμενα με αρτηρίτιδες
- Ανευρύσματα σχετιζόμενα με τραύμα

Effie ROUSKA, MD

Φυσική εξέλιξη

- Η φυσική εξέλιξη των ανευρυσμάτων που δεν έχουν αντιμετωπιστεί χειρουργικά, είναι ο θάνατος λόγω ρήξης
- Ένα ραγέν ανεύρυσμα θωρακικής αορτής εκδηλώνεται με ξαφνικό θάνατο σε ποσοστό 25%

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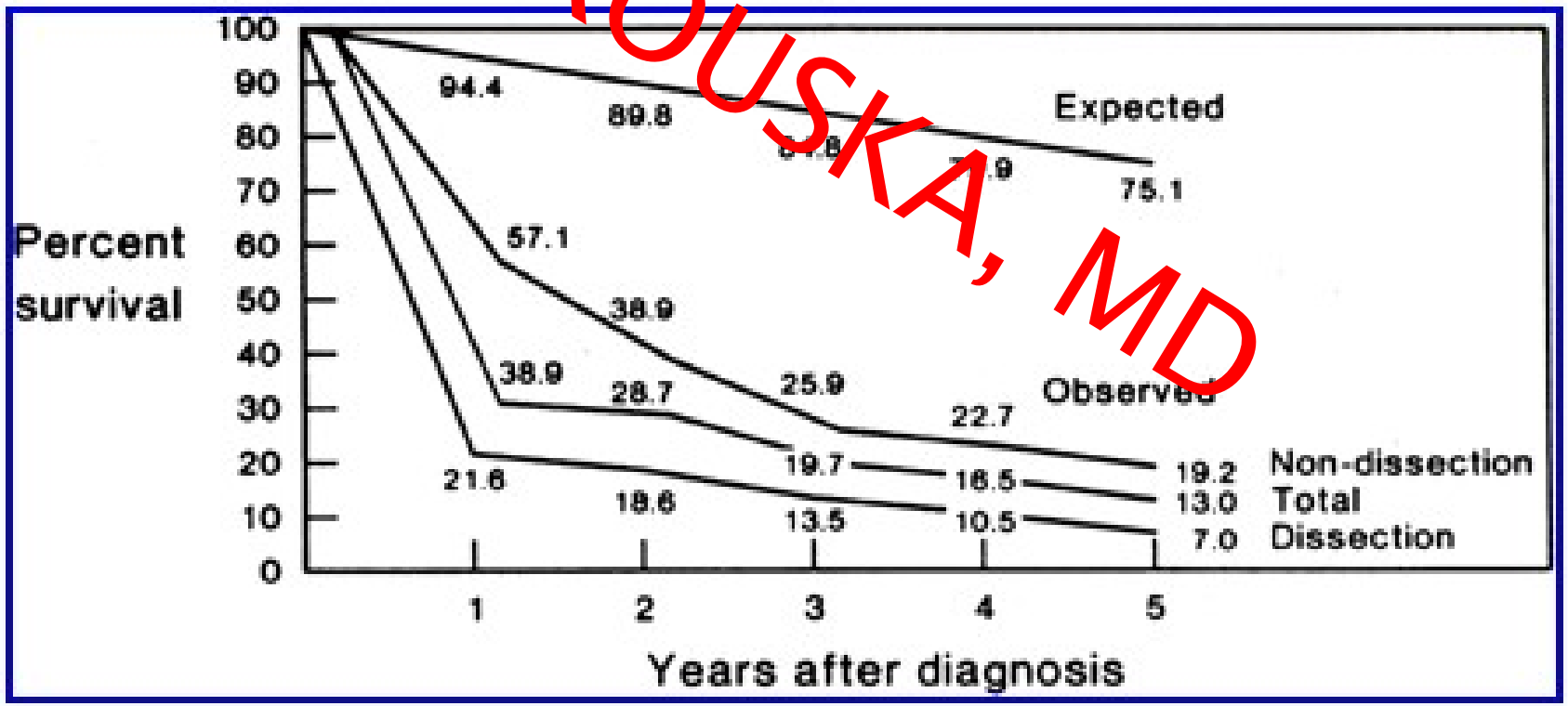

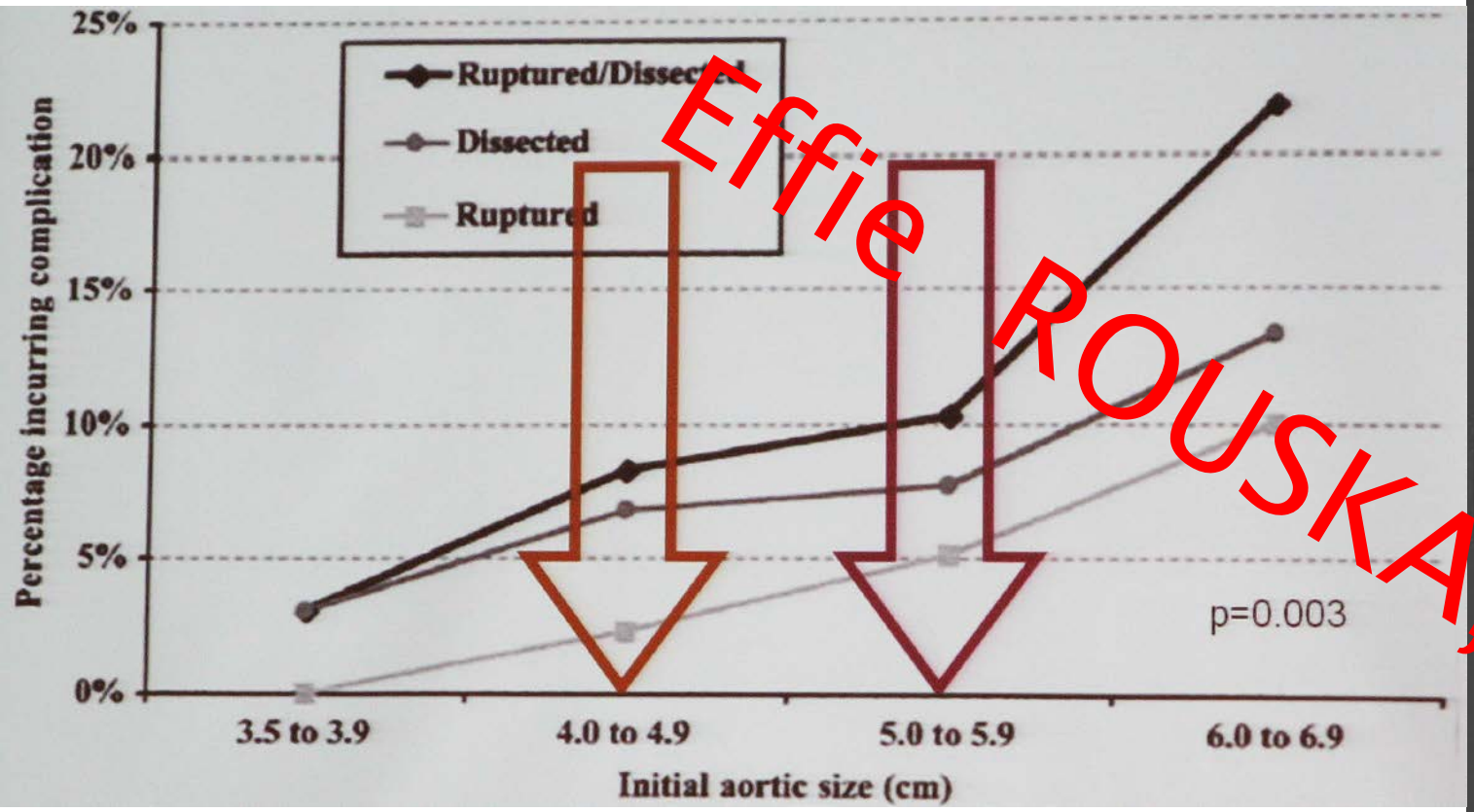

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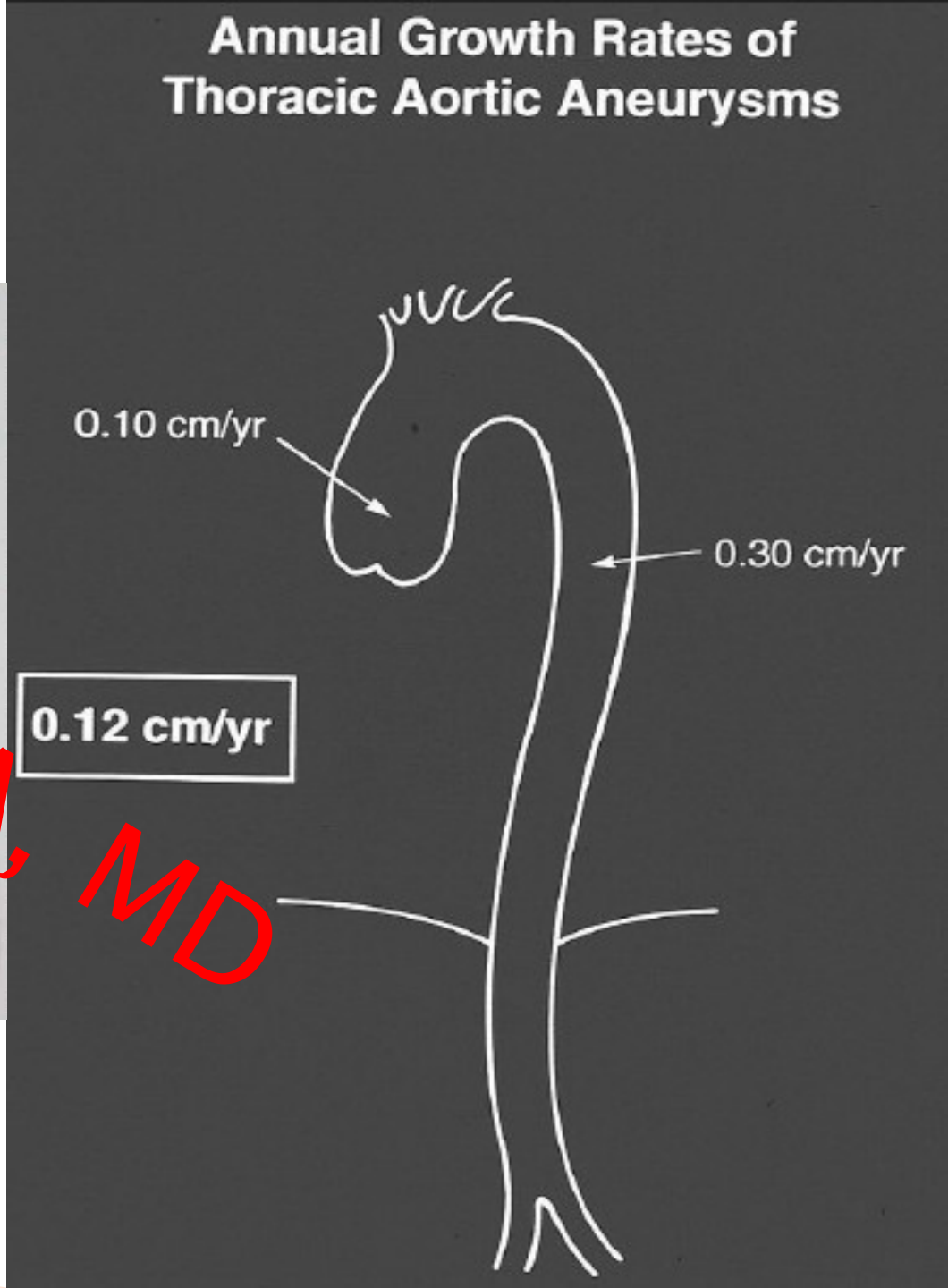


Κλινική Διάγνωση

- συνήθως χωρίς συμπτώματα, τυχαία (CXR, echo)
- χρόνιο πρόσθιο θωρακικό άλγος
  (25-75% πρώτο σύμπτωμα)
- σημεία καρδιακής ανεπάρκειας από ανεπάρκεια AoV
  (διάταση αορτικής ρίζας)
- συμπτώματα πίεσης (άλγος, βήχας, συριγμός, αιμόπτυση, βράγχος, σύνδρομο άνω κοίλης φλέβας)
- σπάνια ρήξη → RA, SVC (HOCF)
- εξωτερικά γνωρίσματα νόσων συνδετικού ιστού

Effie ROUSKA, MD

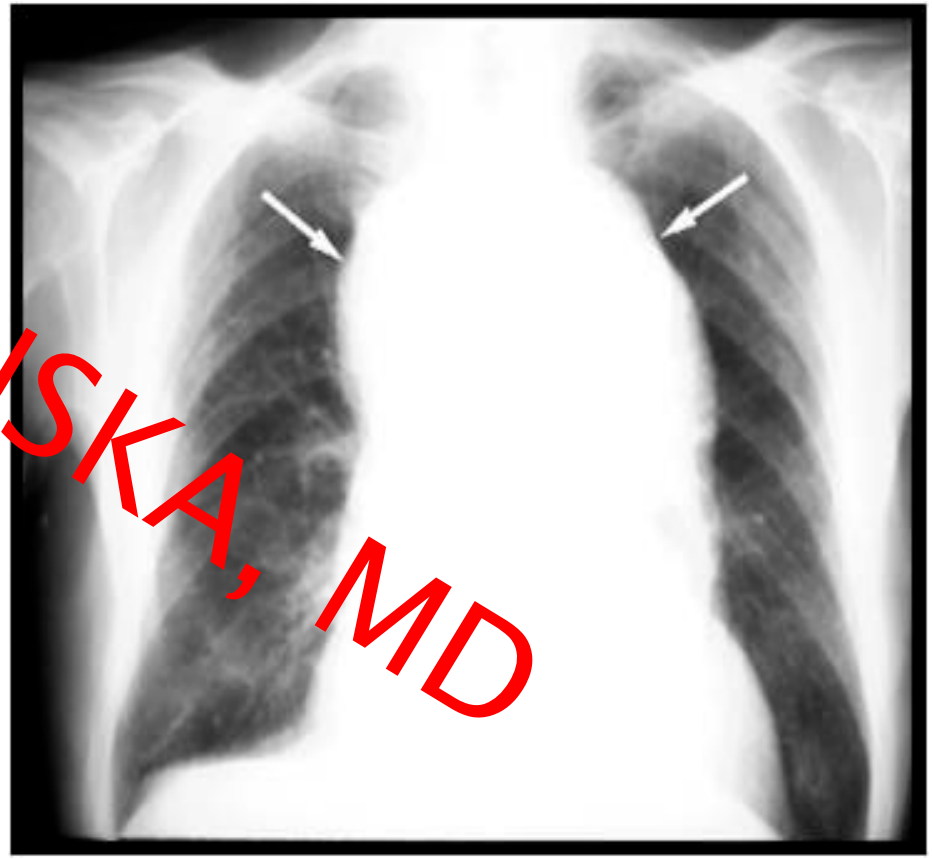
Απεικονιστικές μέθοδοι

- Ακτινογραφία θώρακος
- Ηχωκαρδιογραφία
- Αορτογραφία (CA)
- Αξονική τομογραφία / αγγειογραφία (CT/CTA)
- Μαγνητική αγγειογραφία (MRA)

Effie ROUSKA MD

Ακτινογραφία θώρακος

- αύξηση του μεσοθωρακίου
- μεγέθυνση του αορτικού knob
- παρεκτόπιση της τραχείας



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Ηχωκαρδιογραφία

Διαθωρακική

- ασφαλής
- γρήγορα διαθέσιμη
- μη επεμβατική
- εύκολη επαναληψιμότητα
- εξαρτώμενη από τον ενεργούντα την εξέταση

Διοισοφάγειος

- ακριβέστερη αξιολόγηση του μεγέθους της Ao
- απεικόνιση μεγαλύτερου μέρος της Ao
- μορφολογία της AoV (δίπτυχη βαλβίδα)
- λειτουργικότητα της AoV (ανεπάρκεια)

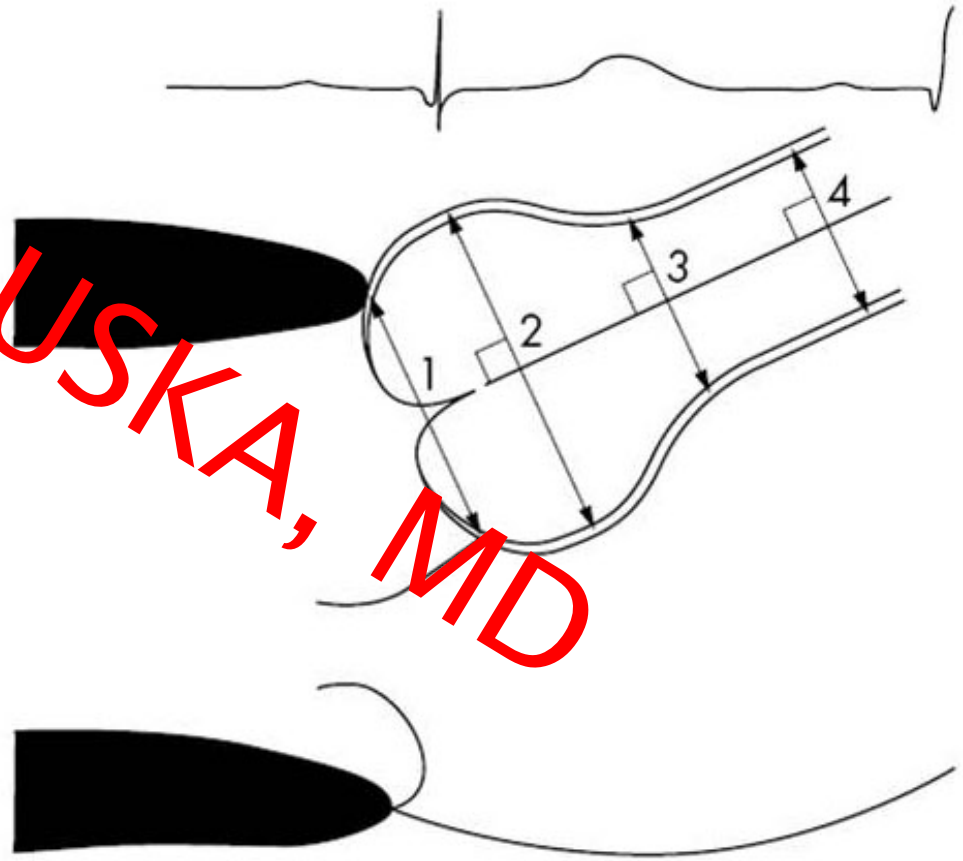
Effie ROUSKA, MD

Aortic root dm's

- *end diastole*
- *parasternal long-axis*

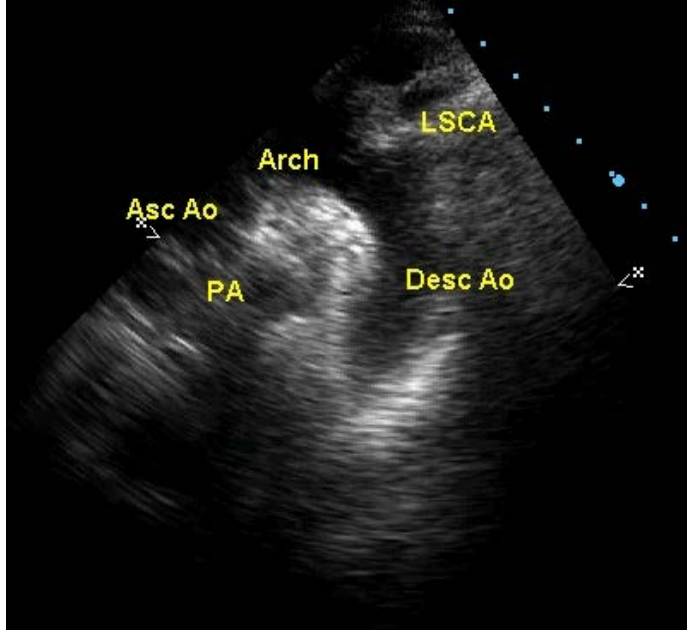
Effie ROUSKA, MD

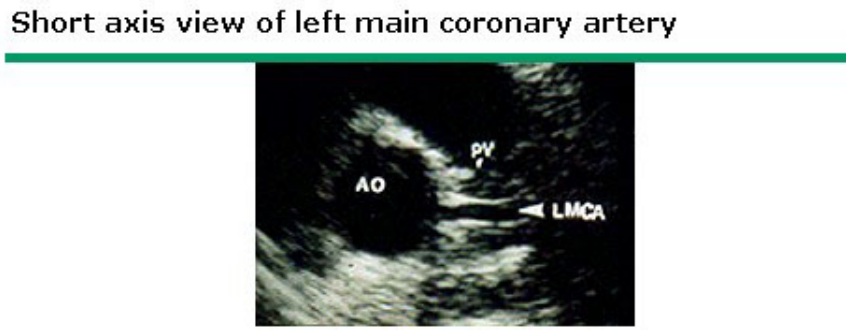
- 4 levels:
  - (a) annulus,
  - (b) sinuses,
  - (c) ST junction,
  - (d) AscAo

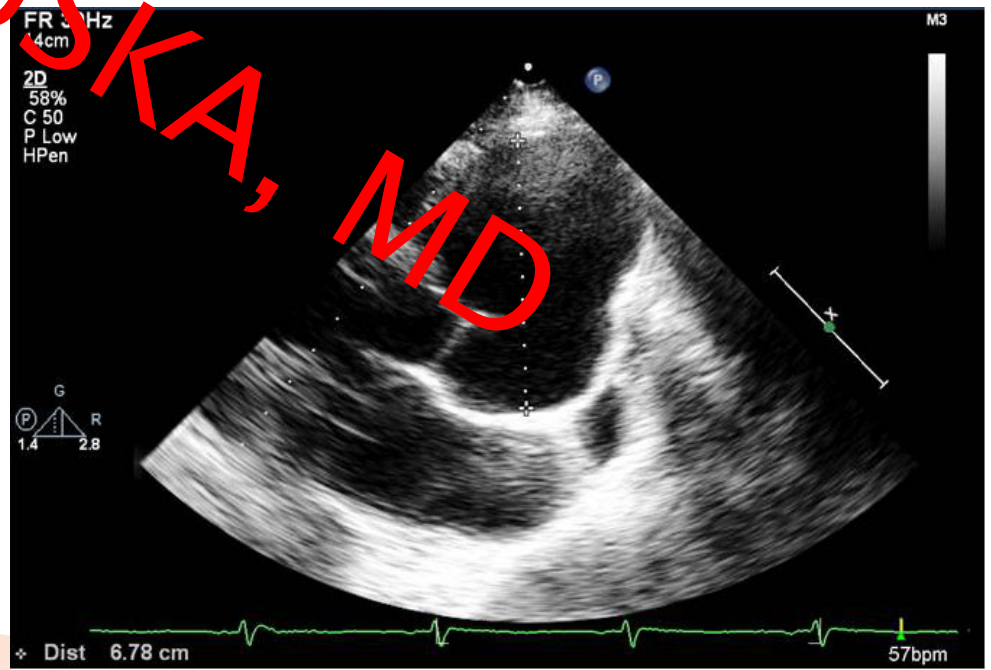


TTE

- *υπεροστερνική λήψη*
  - απεικόνιση του αορτικού τόξου
- *αρ. παραστερνική λήψη*
  - αορτική ρίζα και η ανιούσα αορτή



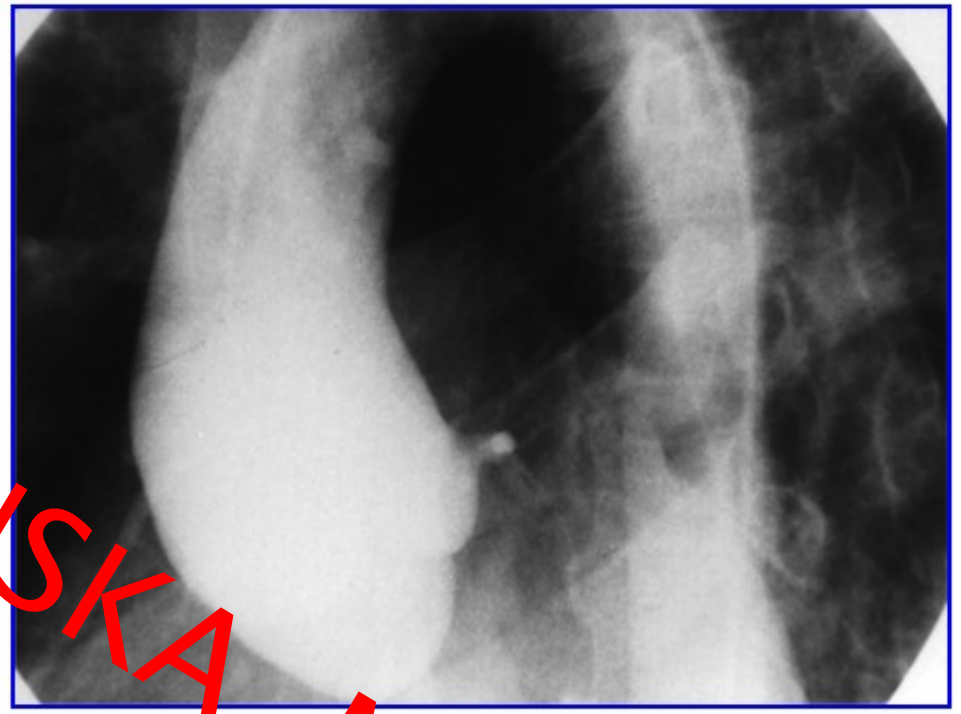




Effie ROUSKA, MD

Αορτογραφία (CA)

- μεγάλη ευκρίνεια
- χαρακτηριστικές εικόνες
- ελέγχος
  - στεφανιαίων αγγείων
  - λειτουργία της LV



Effie ROUSKA, MD

Μειονεκτήματα

- επιπλοκές στο σημείο παρακέντησης
- υποεκτίμηση ΑΑ σε παρουσία θρόμβου

Αξονική τομογραφία /αγγειογραφία (CT/CTA)

- μη επεμβατική
- ευρέως διαθέσιμη
- γρήγορη και ακριβή εκτίμηση
  - αυλού
  - τοιχώματος
  - περιαορτικής περιοχής
- εκτίμηση μεγέθους και έκτασης νόσου

Effie ROUSKA, MD

Μειονεκτήματα

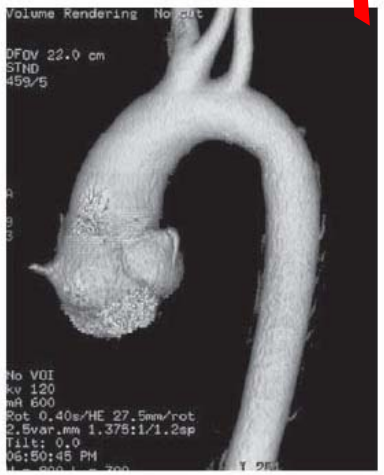
- χρήση του σκιαγραφικού
- δεν δίνει πληροφορίες για AoV
- ακατάλληλη για παρακολούθηση ασθενών με δίπτυχη AoV

Σύγχρονες τεχνικές CT

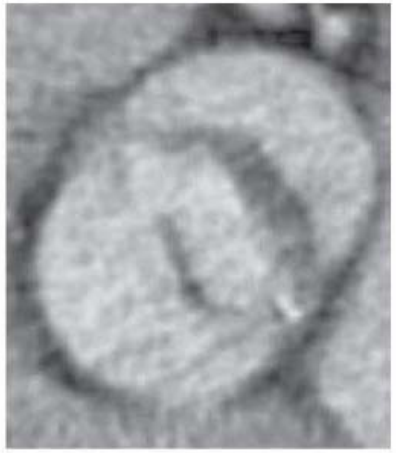
- ελικοειδης CT
- η χρήση πολυτομικών τομογράφων
- η χρήση της 3D ανασύστασης

Effie ROUSKA, MD

δίνουν εξαιρετικές εικόνες
με ευαισθησία περί το 100%
και ειδικότητα περί το 99%



Marfan syndrome and
annuloaortic ectasia
in a 40-year-old man



Ascending aortic
aneurysm and
bicuspid aortic valve
in a 40-year-old woman

Μαγνητική αγγειογραφία (MRA)

- ακριβής και αποτελεσματική
- ανασύσταση και δημιουργία 3D εικόνων
- παροχή λήψεων πολλαπλών τομών
- απεικόνιση ολόκληρης της Ao και των κλάδων της
- η αποφυγή έκθεσης στην ακτινοβολία

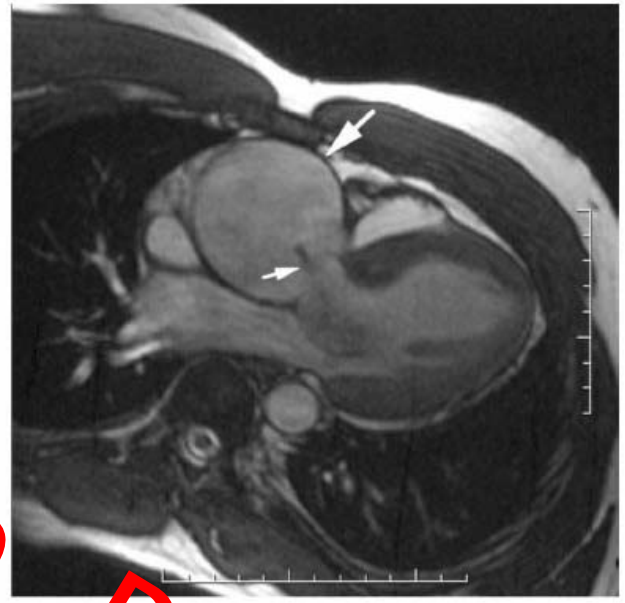
Effie ROUSKA, MD

Μειονεκτήματα

- δεν είναι παντού διαθέσιμη
- η διάρκεια εξέτασης είναι μεγάλη
- το γαδολινίου αντενδείκνυται σε ασθενείς με νεφρική ανεπάρκεια
- δεν υπάρχει πρόσβαση για παρακολούθηση του ασθενή

Αντενδείξεις

- ασθενείς που πάσχουν από κλειστοφοβία
- ασθενείς με εμφυτεύσιμα υλικά και βηματοδότες



Ascending aortic aneurysm
in a 34-year-old man

Effie ROUSKA, MD

Saccular atherosclerotic aneurysm
in a 75-year-old man

Επισημάνσεις επί των απεικονιστικών μεθόδων

- εκτίμηση απολύτου μεγέθους της Ao
- συγκριτική εκτίμηση αύξησης της Ao

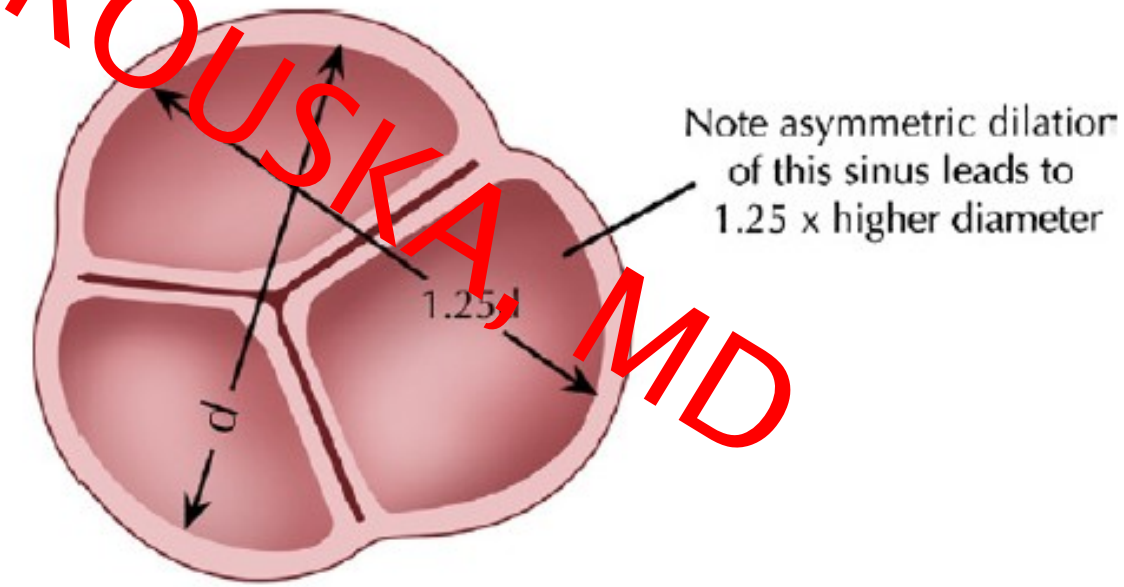
- *Το αορτικό τοίχωμα πρέπει να περιλαμβάνεται η όχι στην μέτρηση??*
  - CT χωρίς σκιαγραφικό
    είναι ακριβέστερο να μετράται *όλο το τοίχωμα*,
  - CT με χρήση του σκιαγραφικού
    είναι ευκολότερο να μετράται *μόνο ο αυλός*

Effie ROUSKA. MD

Επισημάνσεις επί των απεικονιστικών μεθόδων

- σε αξονικές τομές η αορτή δεν εμφανίζεται ως πλήρης κύκλος
- η αορτική διάμετρος διαφέρει ανάλογα με ποια διάμετρο θα επιλέξουμε

Effie ROUSKA, MD

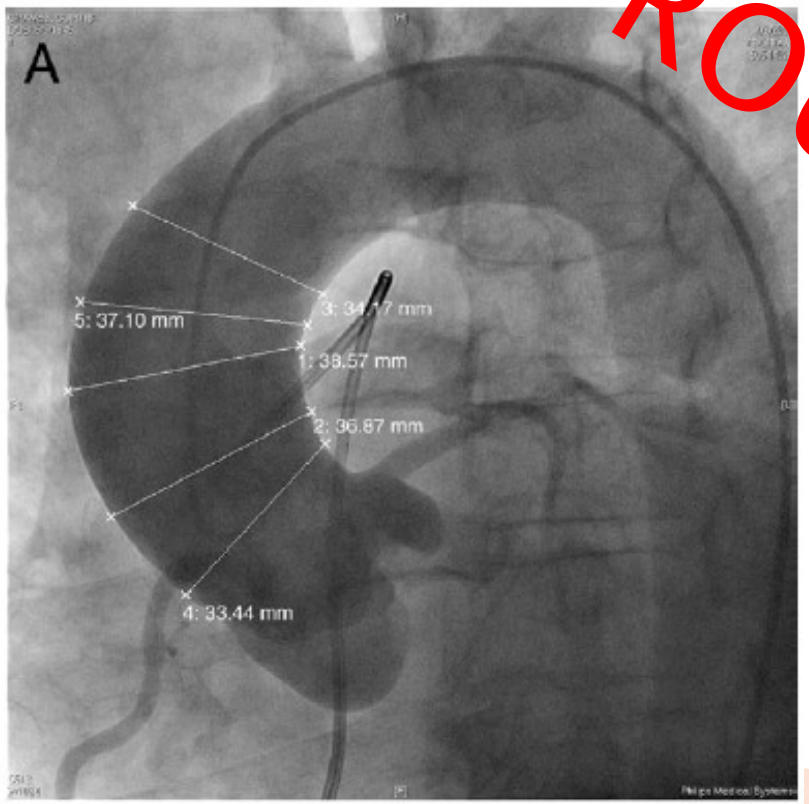


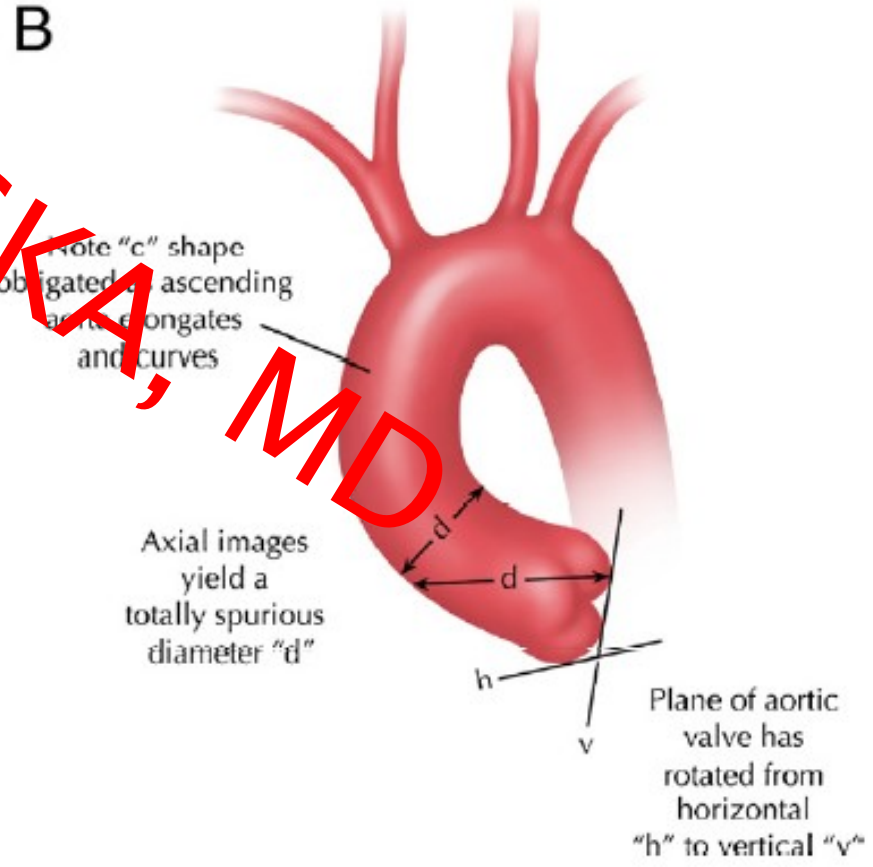
Aorta in cross-section just above aortic valve

Επισημάνσεις επί των απεικονιστικών μεθόδων

- Η πολυπλοκότητα της γεωμετρίας της αορτής πρέπει να λαμβάνεται υπόψη κατά τις μετρήσεις
- Η ανιούσα αορτή δεν είναι κάθετη
- Το αορτικό τόξο είναι "C" σχήματος παρά μια τέλεια καμπύλη

Effie ROUSKA, MD



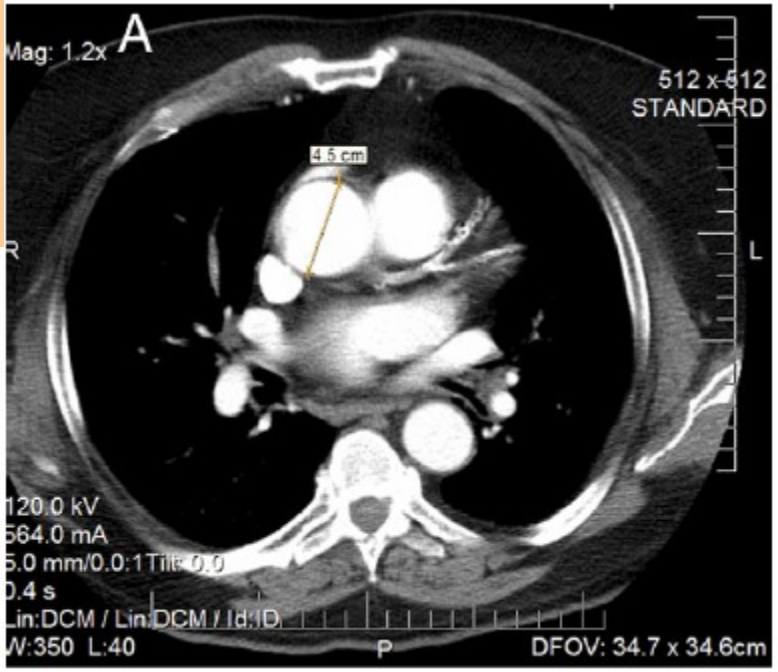


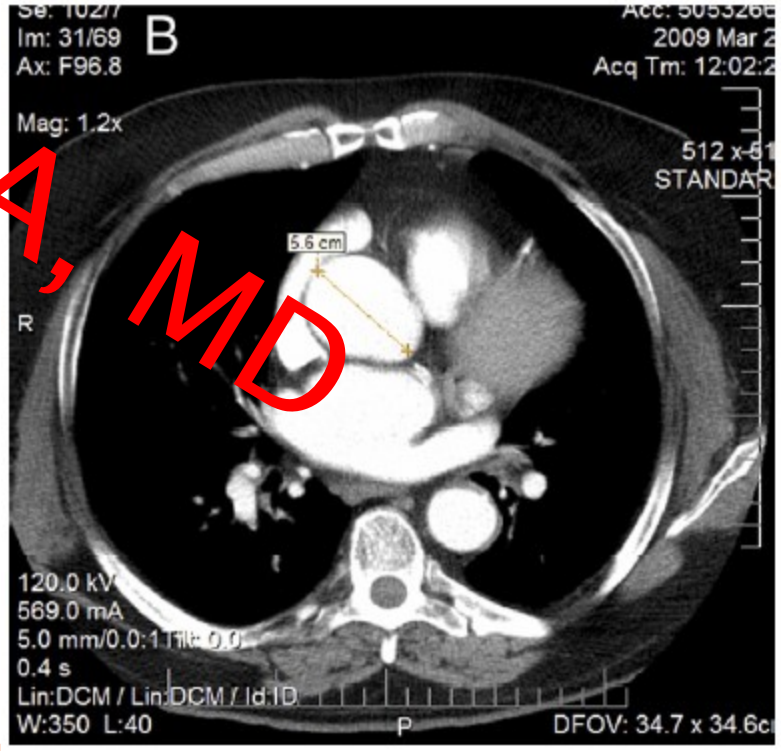
Επισημάνσεις επί των απεικονιστικών μεθόδων

CT vs Echo

Effie ROUSKA, MD

- CT :
  - μη ακριβής αξιολόγηση του μεγέθους της ρίζας και του κεντρικού τμήματος της ανιούσης αορτής
- Echo:
  - ακριβής αξιολόγηση του μεγέθους της ρίζας και του κεντρικού τμήματος της ανιούσης αορτής
  - περιφερικότερα η παρεμβολή των τραχειοβρογχικού δένδρου δημιουργεί πολλές δυσκολίες





Συντηρητική Θεραπευτική προσέγγιση

- επιθετικός έλεγχος της ΑΥ
  - ≤ 140/90 mmHg για τους μη διαβητικούς
  - ≤ 130/80mmHg για τους διαβητικούς
- βελτιστοποίηση των λιπιδαιμικού προφίλ
  - LDL 70mg/dl
- διακοπή του καπνίσματος
- βελτίωση της γενικής κατάστασης

[Class I, Evidence C]

Effie ROUSKA, MD

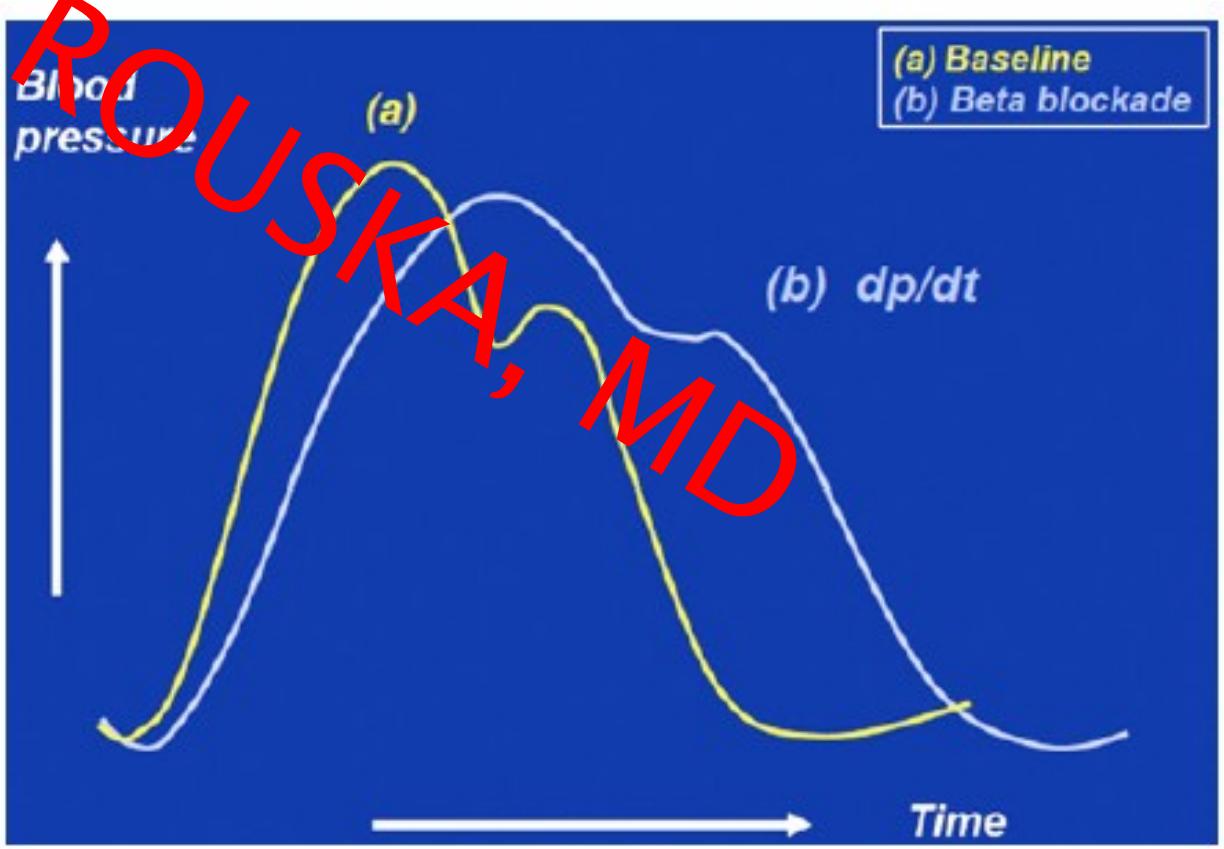
Effie ROUSKA, MD

Β-αποκλειστές

- ελαττώνουν τον ρυθμό ανάπτυξης του ανευρύσματος λόγω της μείωσης του dp/dt

[Class I, Evidence B]

Effie ROUSKA, MD



A MEA – AT1

- μείωσης της ενεργοποίησης του προφλεγμονώδους μεταγραφικού παράγοντα NF-κB *[Class IIa, Evidence B]*

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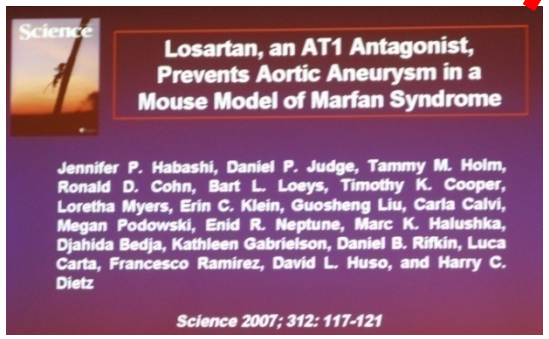
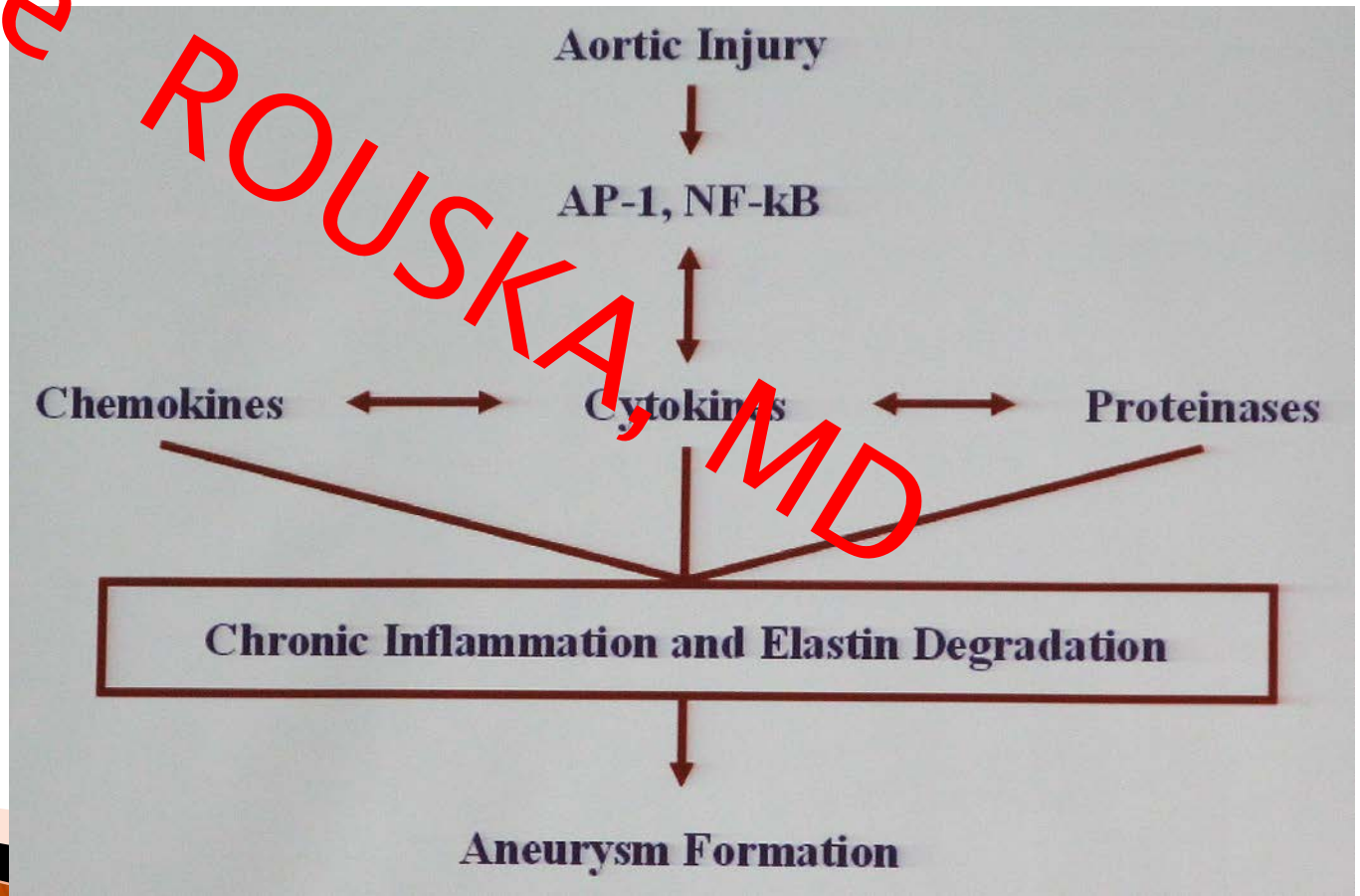
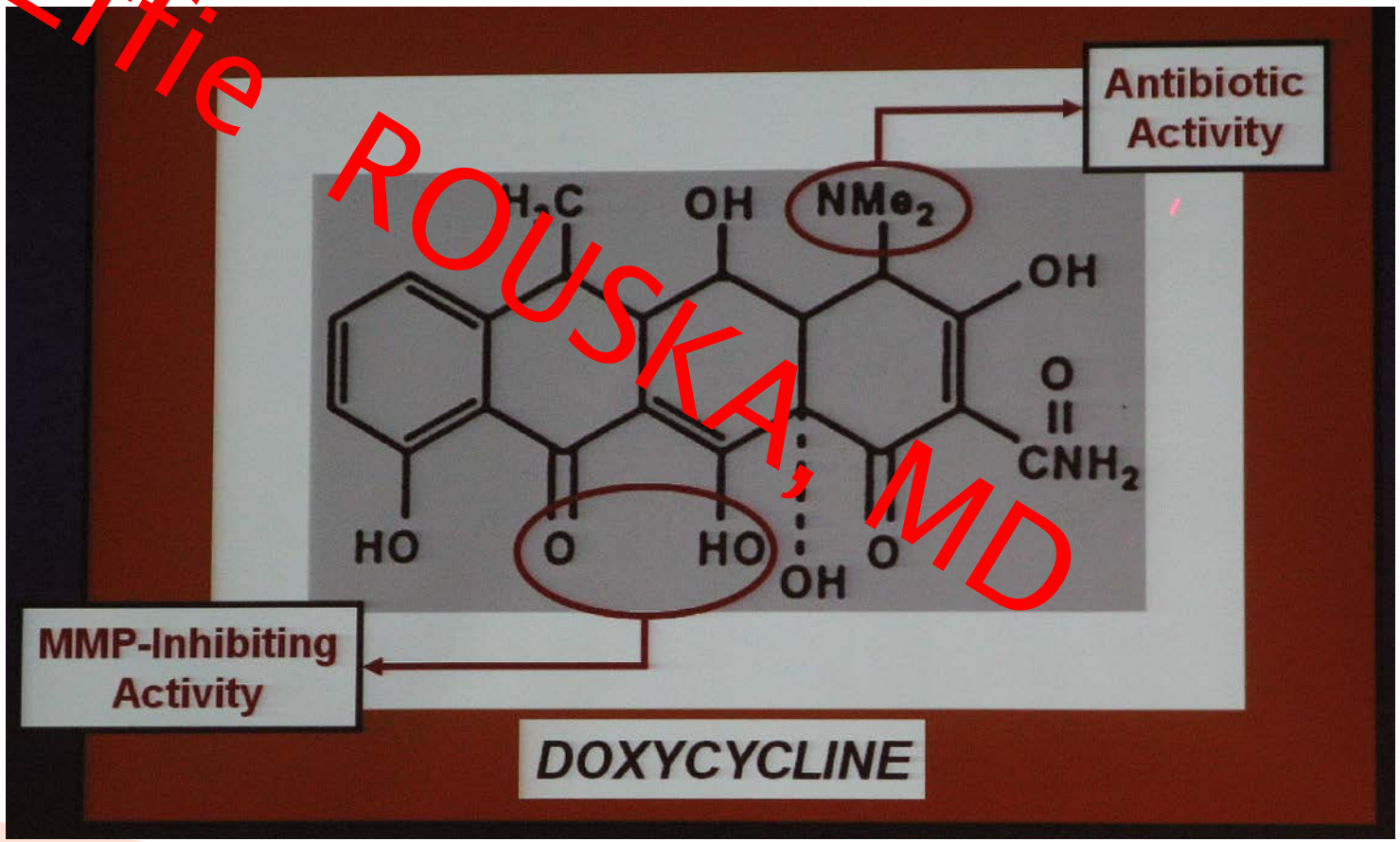




Doxycycline και Roxithromycin

- περιορίζουν την ενεργοποίηση των μεταλλοπρωτεασών στο αορτικό τοίχωμα



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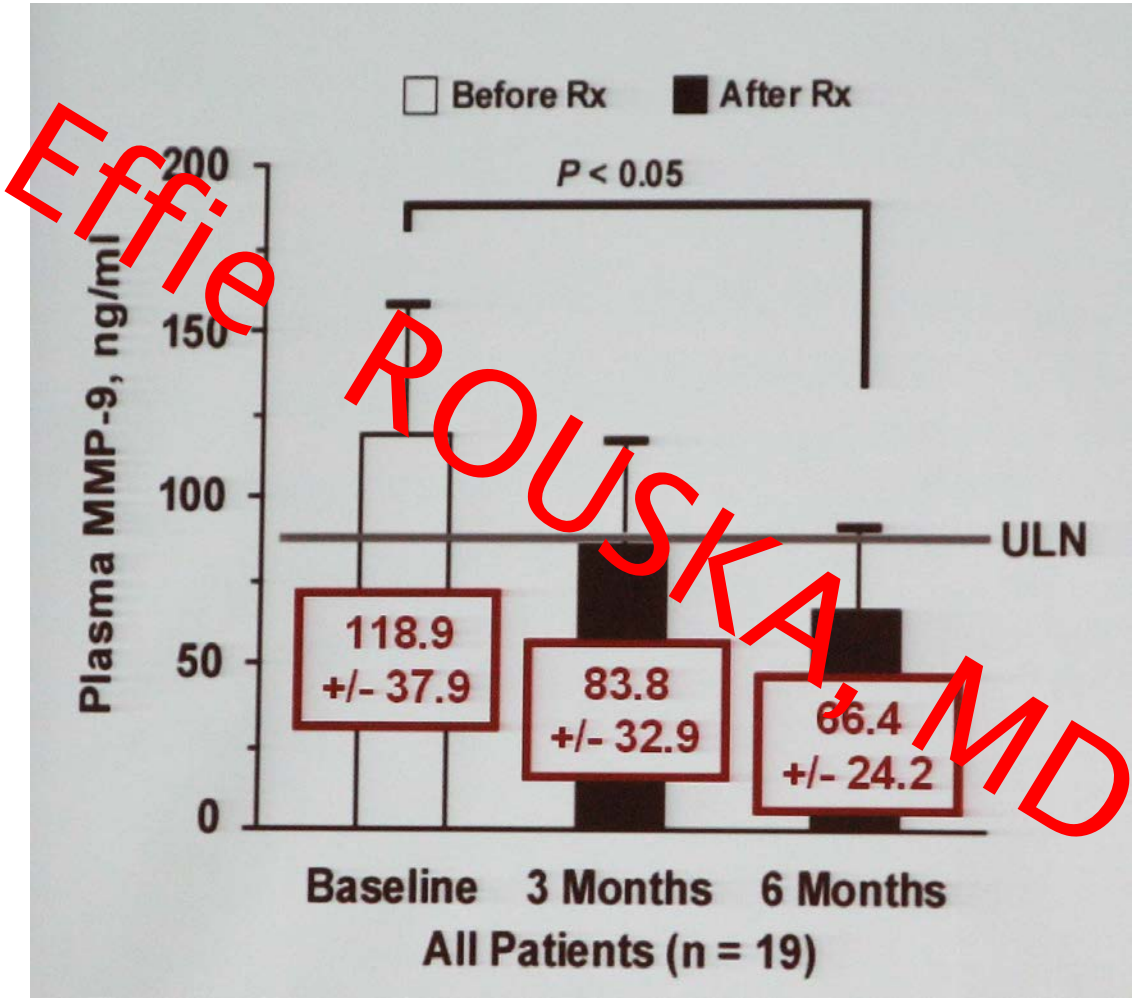
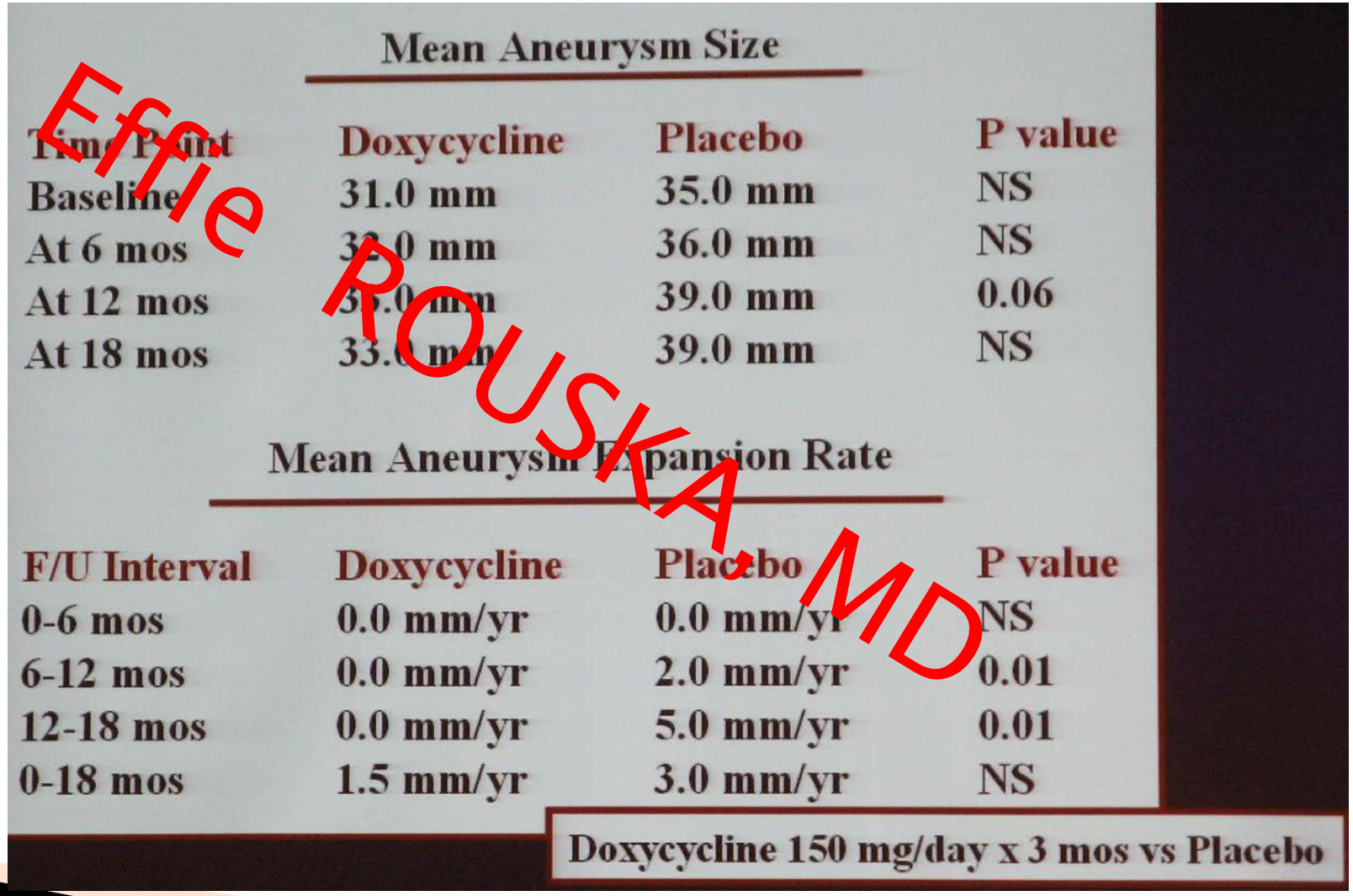
Doxycycline reduces plasma MMP-9 in Pts with small asymptomatic AAAs



Effie ROUSKA, MD

Baxter et al. J Vasc Surg 2002; 36: 1-12

Doxycycline suppresses aneurysm expansion in Pts with small asymptomatic AAAs



Mean Aneurysm Size

Time Point	Doxycycline	Placebo	P value
Baseline	31.0 mm	35.0 mm	NS
At 6 mos	32.0 mm	36.0 mm	NS
At 12 mos	35.0 mm	39.0 mm	0.06
At 18 mos	33.0 mm	39.0 mm	NS

Mean Aneurysm Expansion Rate

F/U Interval	Doxycycline	Placebo	P value
0-6 mos	0.0 mm/yr	0.0 mm/yr	NS
6-12 mos	0.0 mm/yr	2.0 mm/yr	0.01
12-18 mos	0.0 mm/yr	5.0 mm/yr	0.01
0-18 mos	1.5 mm/yr	3.0 mm/yr	NS

Doxycycline 150 mg/day x 3 mos vs Placebo

Effie ROUSKA, MD

Mosorin et al. J Vasc Surg 2001; 34: 606

"Let your food be your medicine"
Hippocrates

Effie ROUSKA, MD

Proposed Therapeutic Purposes

Anti-Oxidant
Anti-Inflammatory
Pulmonary Diseases
Gastrointestinal Diseases
Diabetes & Metabolic Diseases
Female Reproduction
Suppression of Malignancies
Alzheimer's Disease

Παρακολούθηση

- ασυμπτωματικοί ασθενείς με ανεύρυσμα της ανιούσης αορτής
- άτομα με οικογενειακό ιστορικό κληρονομικών διαταραχών και οικογενούς κατανομής
- άτομα με δίπτυχη ΑΥ
- Αθλητές (ειδικά άρσης βαρών)

Effie ROUSKA, MD

- Perform serial imaging studies to follow aneurysm growth
  - Repeat imaging study 6 months after diagnosis
  - If stable, annually thereafter
- In general, use same modality each time
  - But when possible, and if anatomy is stable, switch from CTA to MRA or echo to reduce cumulative radiation

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MASSACHUSETTS GENERAL HOSPITAL
HEART CENTER

- Both aneurysm size and rate of growth are important
- Interpretation must include measurements
  - "No significant change" may really mean aneurysm is up to 3 mm larger
  - Can add up over years
- Inter-reader variability = 2-5 mm
  - Don't just rely on the report to determine if significant growth
  - Compare serial exams side-by-side and measure comparable images

Effie ROUSKA, MD

- Most importantly, screening should be recommended
  - Internists often tell the relatives it's not necessary
- Modality should be chosen to match suspected anatomy
  - Aortic root (e.g., Marfan syndrome)
    - Echocardiogram or MRA
    - CT unreliable
  - Ascending thoracic aorta
    - CT or MRI
    - Echocardiogram unreliable
  - Arch or descending thoracic aorta
    - CTA or MRA

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Indications for Surgery

Indications for intervention on thoracic aortic aneurysms

Indication for intervention on aortic root aneurysms	
≥ 40mm	Consider surgery in females with low body surface area harboring a TGFBR2 mutation and presenting with extra-aortic features
≥ 45mm	Marfan Syndrome with risk factors Family history of aortic dissection, aortic size increase >3mm/yr, severe AR , severe MR, desire for pregnancy Indication for surgery on the aortic valve TGFBR1 or TGFBR2 mutation (including Loeys-Dietz Syndrome)
≥ 50mm	Bicuspid aortic valve with risk factors Coarctation of the aorta, systemic hypertension, family history of dissection, aortic size increase >3mm/yr Marfan Syndrome without risk factors
≥ 55mm	Other patients with no elastopathy

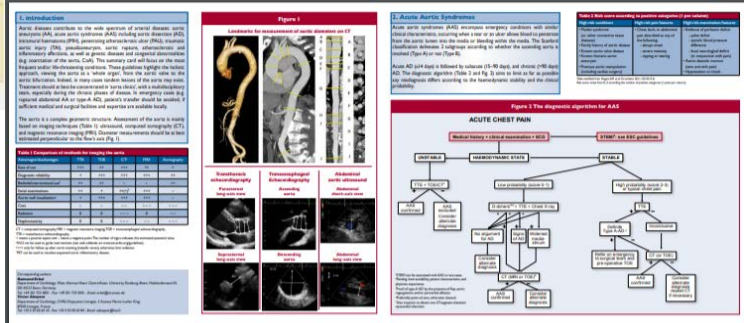
Indication for intervention on aortic arch aneurysms	
≥ 55mm	Patients with isolated aortic arch aneurysm

Indication for intervention on descending aortic aneurysms	
≥ 55mm	If TEVAR possible
≥ 60mm	If TEVAR not possible

Effie ROUSKA, MD

udsonUK

s – Aortic Diseases; ESC Clinical Practice Guidelines –
Jondeau, G. et al. International Registry of Patients Carrying
of the Montalcino Aortic Consortium. Circ Cardiovasc Genet.



Interesting Case

- 51-year-old Marfan female patient
- with a history of surgery for type A aortic dissection

Effie ROUSKA, MD

On admission

- presented with uncontrolled hypertension
- known hypertensive on medical treatment
  - *B-Blocker*
  - *ACE-inhibitor*
- medical treatment became not efficient recently
- admitted neglect in her appointments over the last 3 yrs

PMH

Marfan patient – Ghent Criteria

System	Major criterion	Involvement
Skeletal	At least 4 of the following features:	2 of the major features, or 1 major feature and 2 of the following:
	● Pectus carinatum	● Pectus excavatum
	● Pectus excavatum requiring surgery	● Joint hypermobility
	● ULSR <0.86 or span:height >1.05	● High palate with dental
	● Wrist and thumb signs	● Crowding
	● Scoliosis >20° or spondylolisthesis	● Characteristic face
	● Reduced elbow extension (<170°)	
	● Pes plenus	
	● Protrusio acetabulae	
Ocular	Lens dislocation (ectopia lentis)	Flat cornea
		Increased axial length of globe (causing myopia)
		Hypoplastic iris or ciliary muscle (causing decreased miosis)
Cardiovascular	Dilatation of the aortic root	Mitral valve prolapse
	Dissection of the ascending aorta	Dilatation of the pulmonary artery, below age 40
		Calcified mitral annulus, below age 40
		Other dilatation or dissection of the aorta
Pulmonary	None	Spontaneous pneumothorax
		Apical blebs
Skin/Integument	None	Striae atrophicae
		Recurrent or incisional hernia
Dura	Lumbosacral dural ectasia	None
Genetic findings	Parent, child or sibling meets these criteria independently	None
	Fibrillin 1 mutation known to cause Marfan syndrome	
	Inheritance of DNA marker haplotype linked to Marfan syndrome in the family	

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PMH

- Type –A aortic dissection 11 years ago

- confusional state
- bruises on both carotid arteries
- no palpable pulses in the right upper and the left lower extremity

CT angiography *demonstrated Type –A aortic dissection extended from aortic root to iliac arteries*

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PMH

- **Operation:**
  - Supracoronary ascending aorta replacement with a tubular graft (Vascutec, 25mm) and resuspension of the incompetent aortic valve
- **Recovery:**
  - Very good
- **Histopathological examination:**
  - media of the aortic wall with a profound decrease in the amount of elastin and loss of the highly aligned and ordered lamellar arrangement
  - extensive deposits of mucopolysaccharides

Effie ROUSKA, MD

Recent admission – *O/E*

- HR 76bpm, SR
- BP 175/95mmHg
- <u>On auscultation</u>
  - parasternal diastolic murmur
    (third left intercostal space
    radiating widely along the left sternal border)

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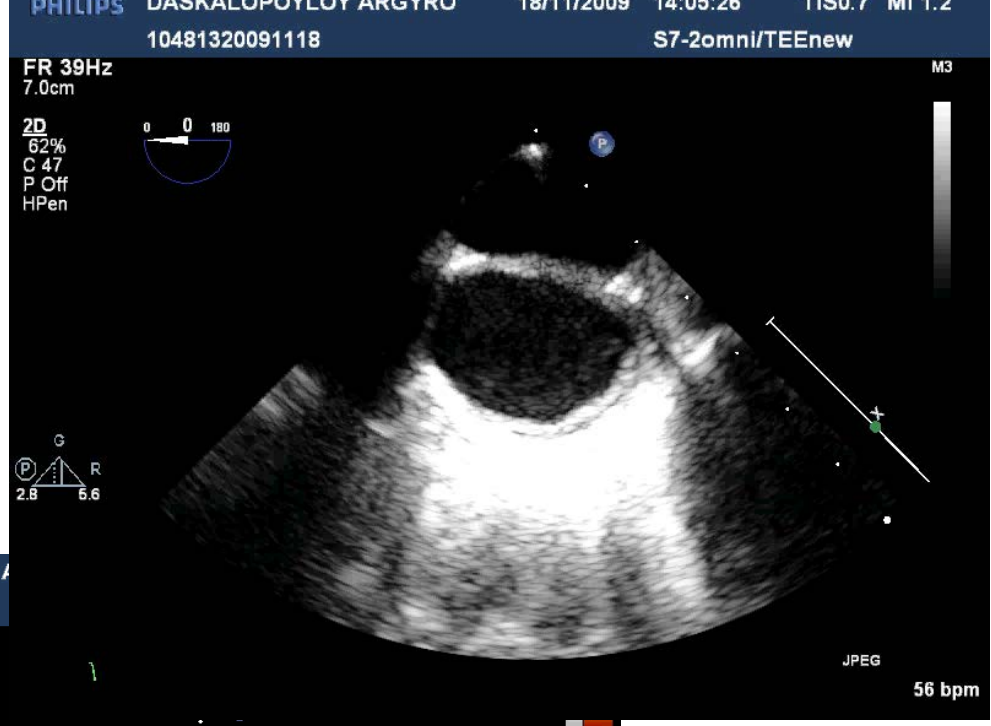
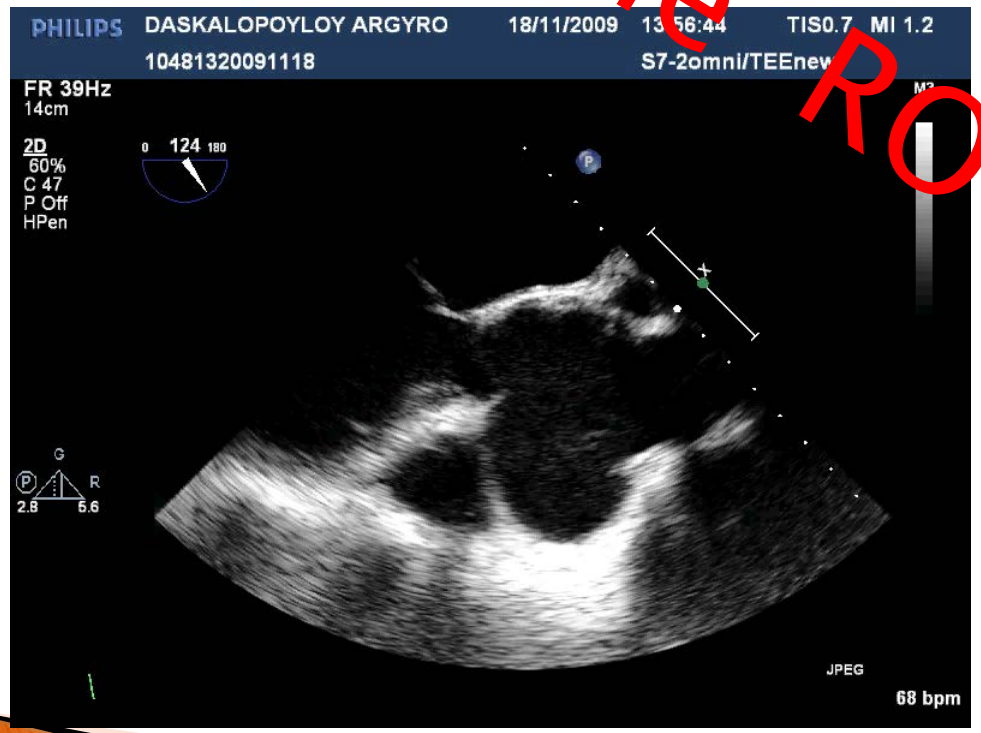
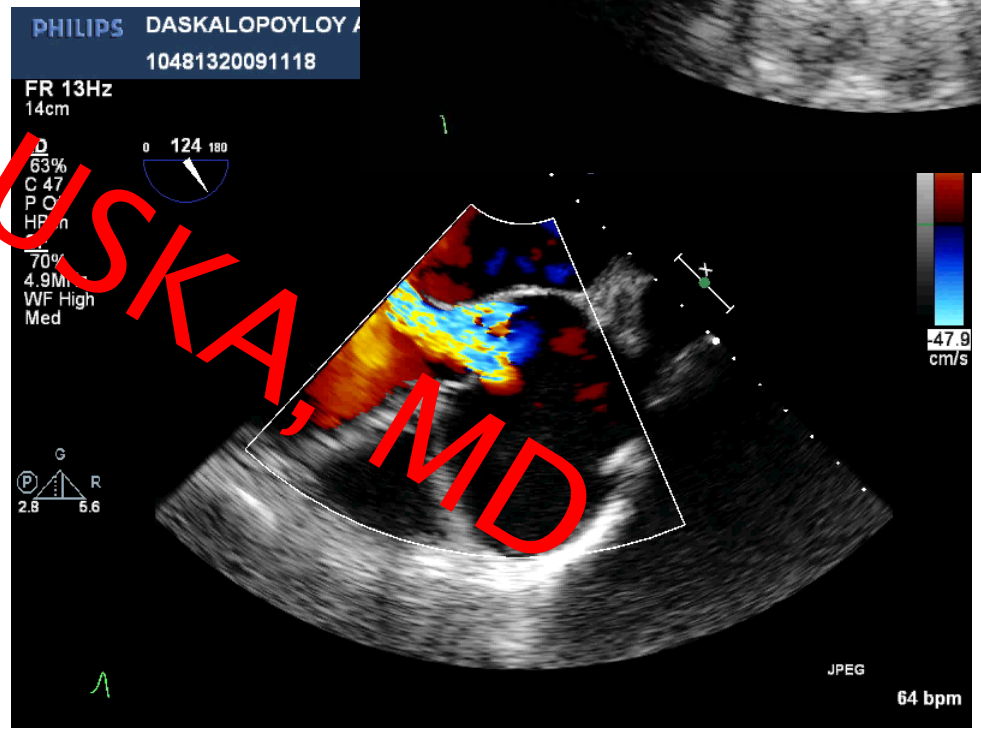
TOE images







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Coronary Angiogram –Aortography

- Normal coronaries
- Aneurysm of Valsalva

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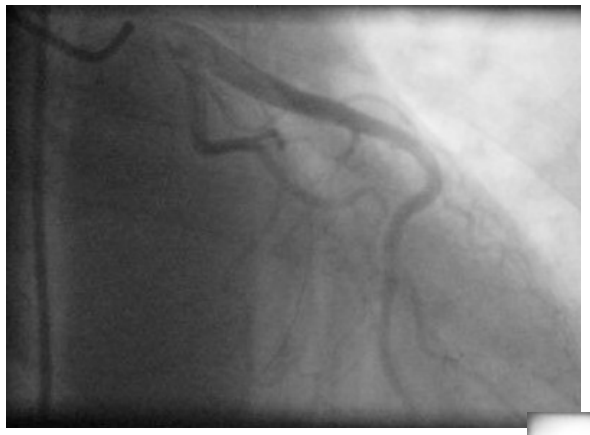

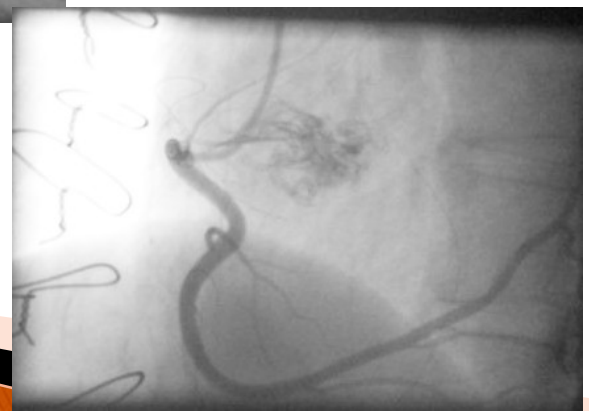

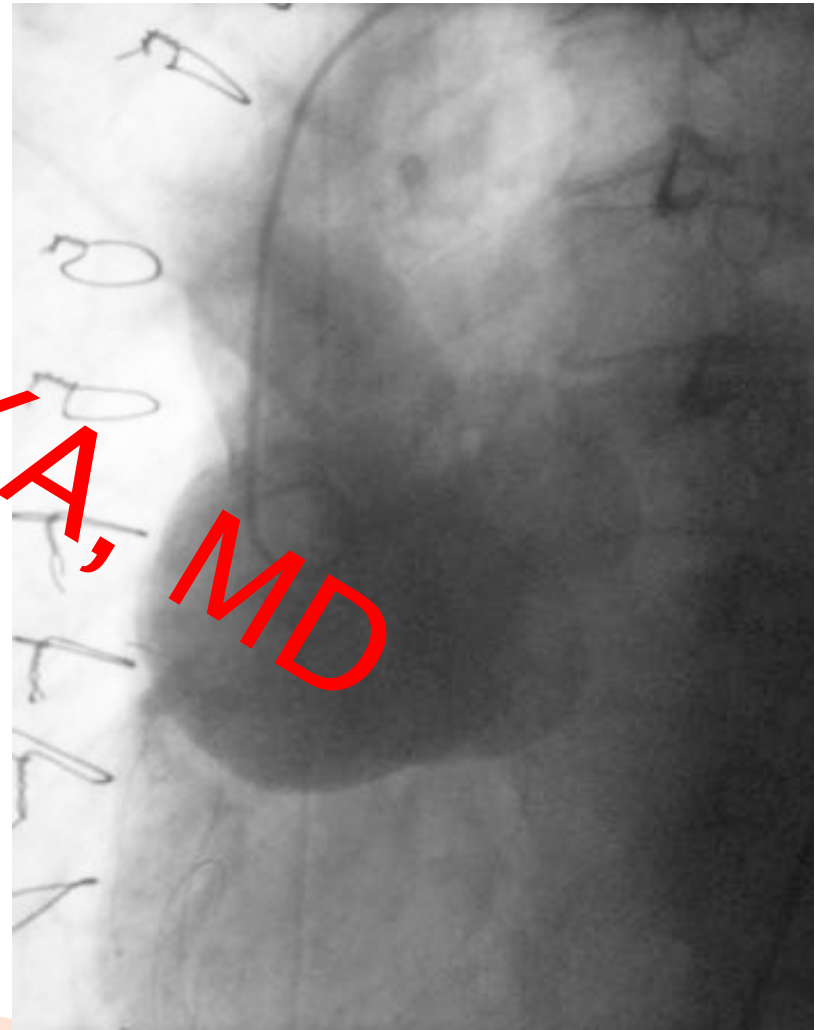

CT angiography

- severe Aortic root dilatation and findings consistent with the previous operation

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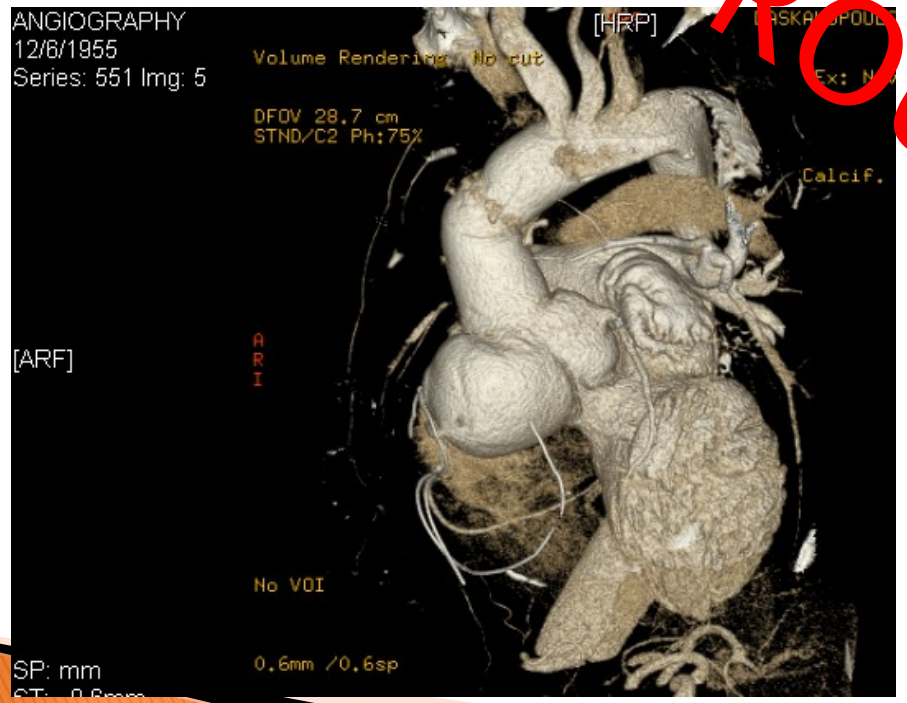
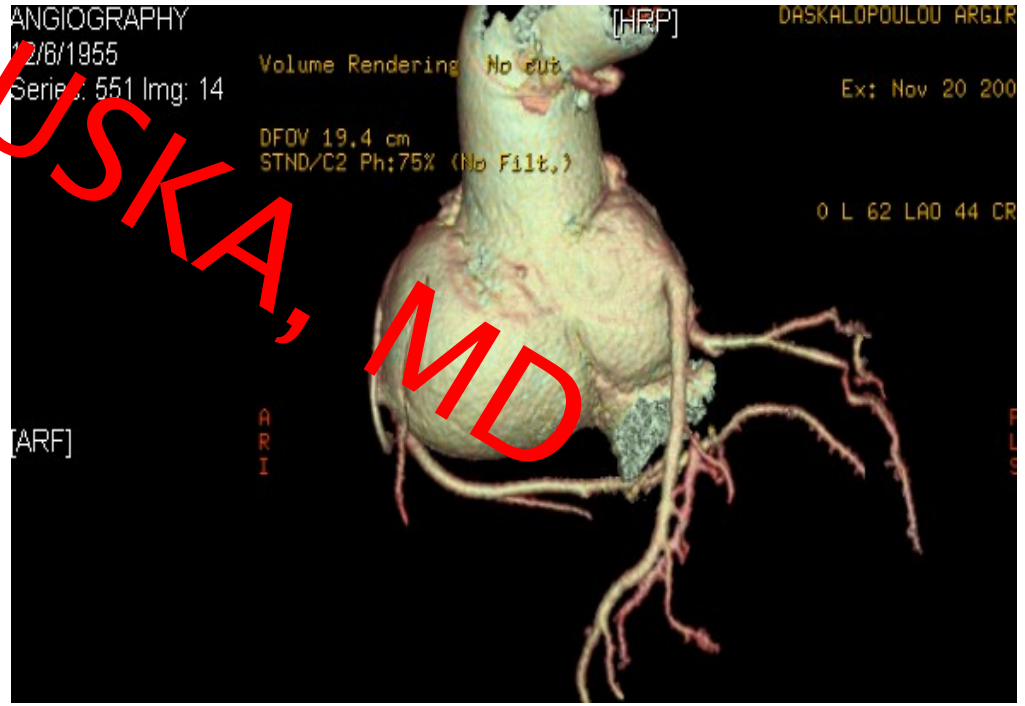
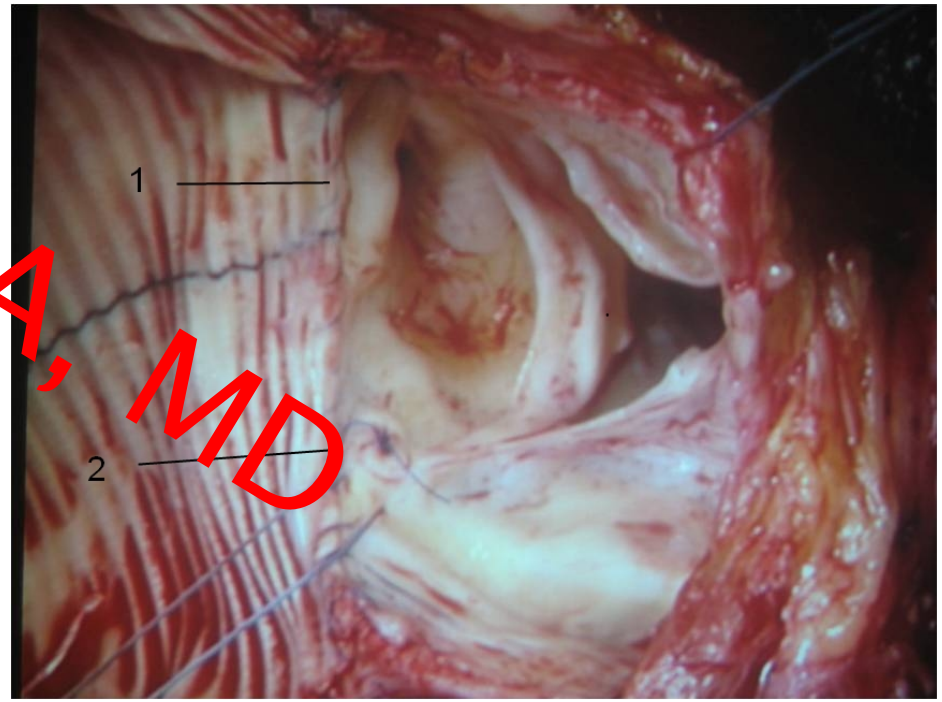




Urgent Redo-operation

Effie ROUSKA, MD

- Bentall procedure
- Post-op complications
- Long hospital stay
- Discharged the 55th day in a good condition



Discussion

- Marfan syndrome (MfS) is an autosomal dominant inherited connective tissue disorder with variable phenotypic expression of cardiovascular, ocular and musculoskeletal manifestations
- Usually associated with mutation in fibrillin-1 (FBN1) gene on chromosome 15, which encodes for the glycoprotein fibrillin.
- Estimated prevalence of MfS is 1 in 10,000
- 26% of cases have no family history (new mutation)

Effie ROUSKA, MD

- Dean JC. Heart. 2002 ;88:97
- Westaby S. Ann Thorac Surg. 1999;67:1861
- Detter C, et al . Eur J Cardiothorac Surg. 1998 ;13:416

Natural History

- Life expectancy is determined by the severity of cardiovascular involvement
- Without surgical intervention, many patients die in the third decade of their lives from complications of aortic root aneurysm *(rupture, dissection, insufficiency)*
- Life expectancy improved in the past 30years as a result of improved surgical management

Effie ROUSKA, MD

•David TE, et al J Thorac Cardiovasc Surg. 2009 ;138:859

Surgery in MfS:

1. *Bentall procedure* (golden standard
   - Excellent early and late postoperative
   - Complications related to long-term anticoagulation.

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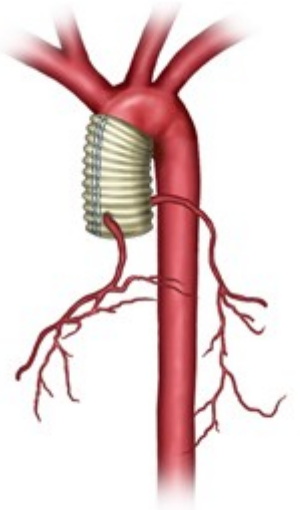


Button Bentall Procedure

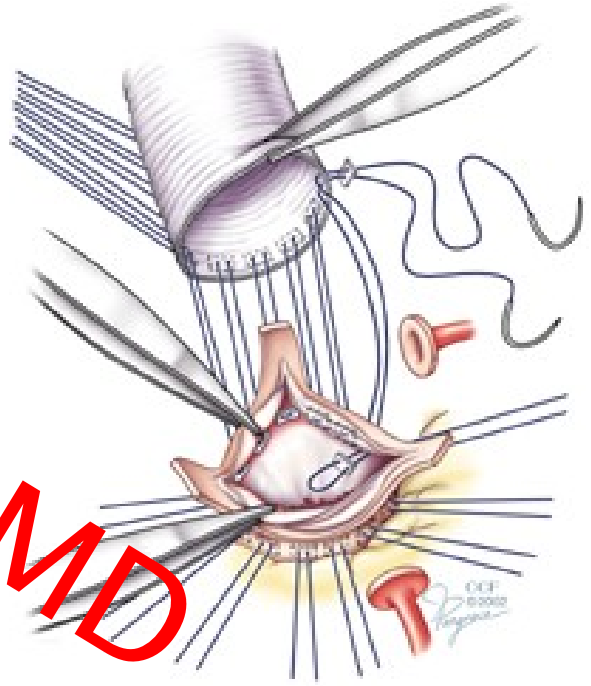
- Karck M, et al J Thorac Cardiovasc Surg. 2004;127:391
- Braverman AC. Curr Opin Cardiol. 2004 ;19:549
- *Elefteriades J .J Thorac Cardiovasc Surg 2002;123:201*

Surgery in MfS:

2. *Valve-sparing root replacement (David / Yacoub)*

Effie ROUSKA, MD

- Operative results similar to Bentall procedure
- Time-consuming
- Durability has to be proven



- Cameron DE, et al Ann Thorac Surg. 2009;87:1344
- Patel ND, et al Ann Thorac Surg. 2008;85:2003
- Volguina IV, et al J Thorac Cardiovasc Surg. 2009 ;137:1124
- Kallenbach K, et al Ann Thorac Surg. 2007;83:S764
- *Elefteriades J. J Thorac Cardiovasc Surg 2002;123:201*

Surgery in MfS:

3. Conservative procedures

(replacement of the dissected aorta with tube graft

–

with/without resuspension of AV)

- Saves time
- High risk Reoperation usually needed!
- **Close follow-up of patients is mandatory!**

Effie ROUSKA, MD

- Westaby S. Ann Thorac Surg. 1999;67:1861
- De Paulis R, et al Eur J Cardiothorac Surg.2005; 27: 86
- Treasure T. Heart. 2000;84:674

Ευχαριστώ

Effie ROUSKA, MD

Το μόνο πράγμα που μας διδάσκει ο θάνατος
είναι πως είναι επείγον να αγαπήσουμε.
-Eric-Emmanuel Schmit

ΔΙΑΧΩΡΙΣΜΟΣ ΑΝΙΟΥΣΗΣ ΑΟΡΤΗΣ

- *Ιστορική αναδρομή*
- *Ονοματολογία*
- *Εμφάνιση*
- *Ανατομική Ταξινόμηση*
- *Παθογένεια*
- *Λειτουργική Ταξινόμηση*
- *Αιτιολογία – Παράγοντες κινδύνου*
- *Φυσική Εξέλιξη*
- *Κλινικά Χαρακτηριστικά*
- *Διάγνωση*
- *Αρχική Θεραπευτική προσέγγιση*

Ιστορική αναδρομή

- Sennertus (16ο αιώνα) — 1η αναγνώριση
- Gurin (1935) — 1η περιγραφή
- Laennec(1826) — ορισμός ως νόσος
- Shennan (1934) — παθογενετικός μηχανισμός
- Maunoir (τέλη του 17ου)) — 1η χειρουργ. αντιμετώπιση
- De Bakey και Cooley — αρχές χειρουργικές

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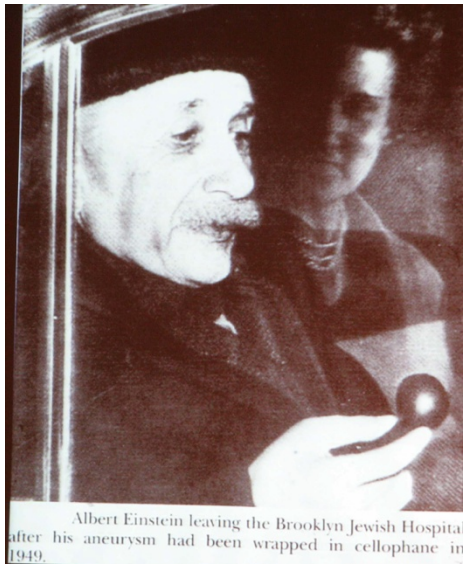
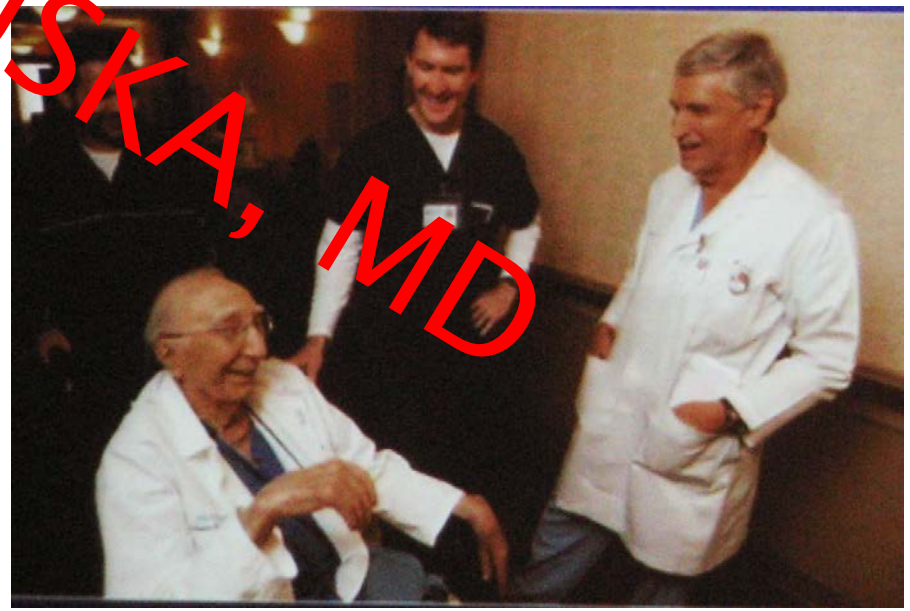
Διάσημη νόσος – Διάσημοι ασθενείς

George II
King of Great Britain and Ireland

A. Einstein





De Bakey

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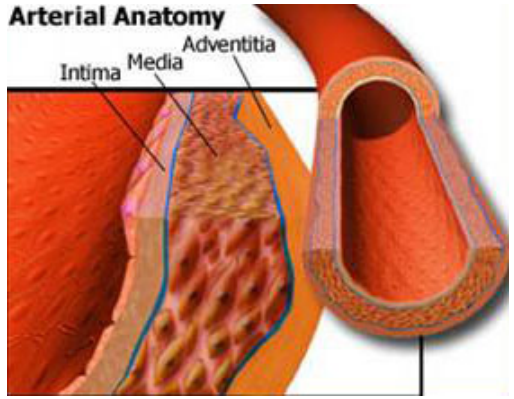
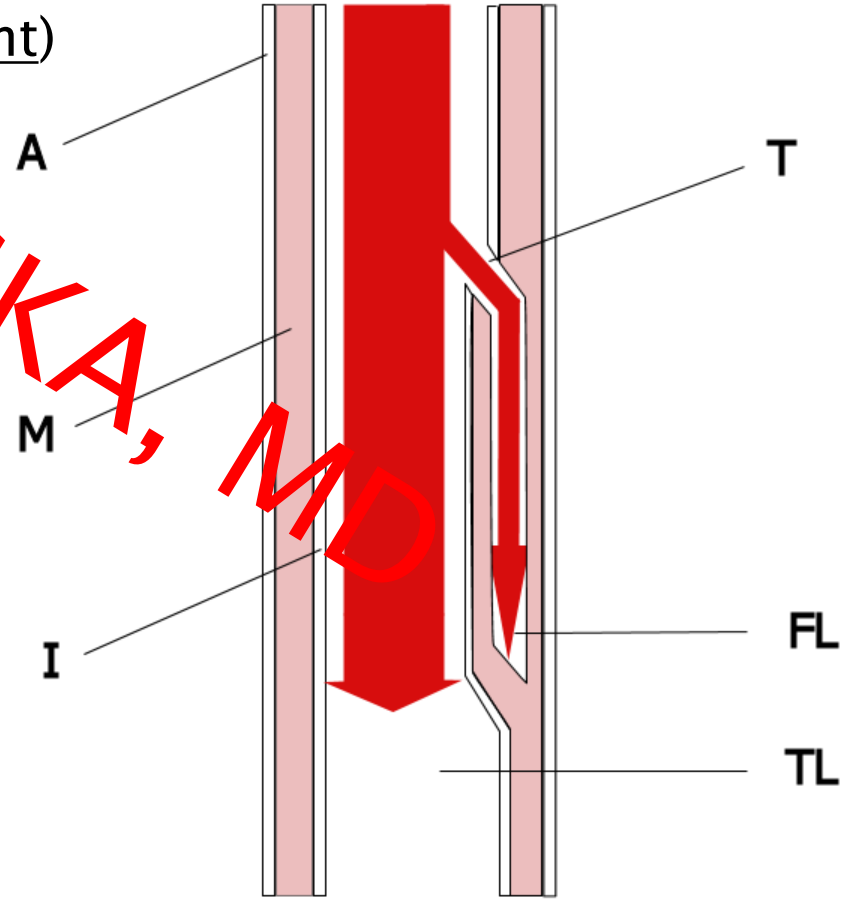
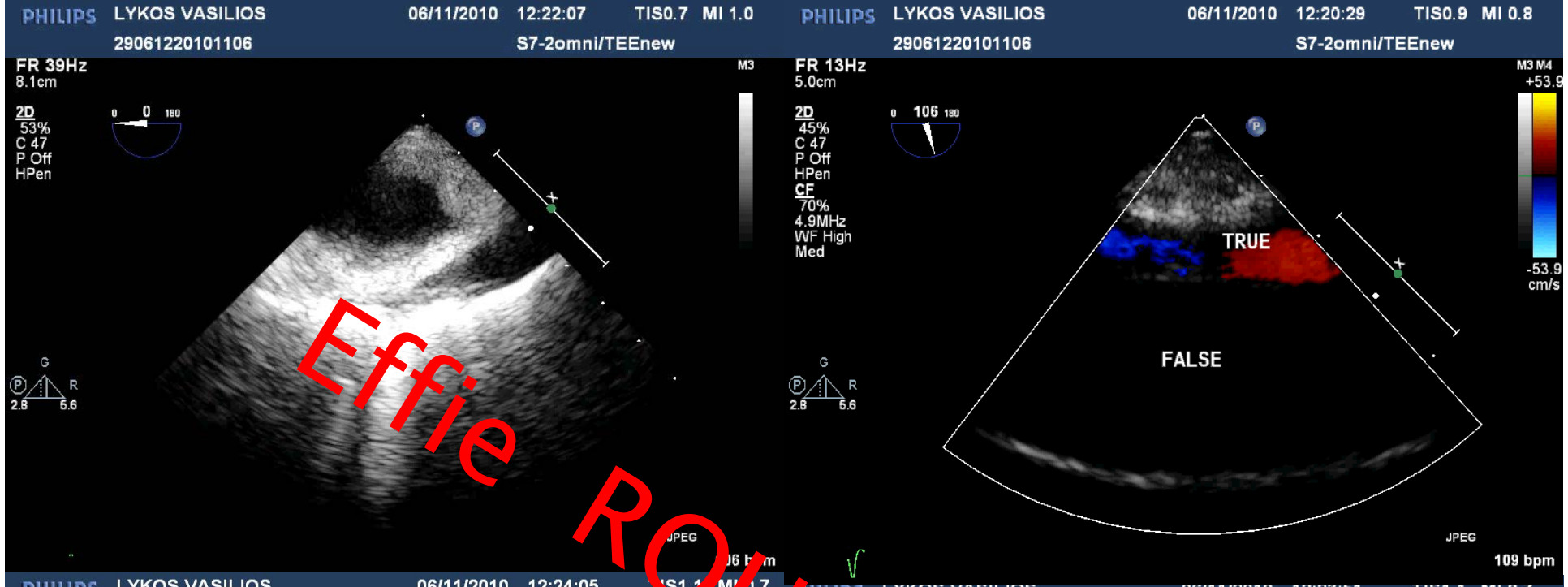
Ονοματολογία

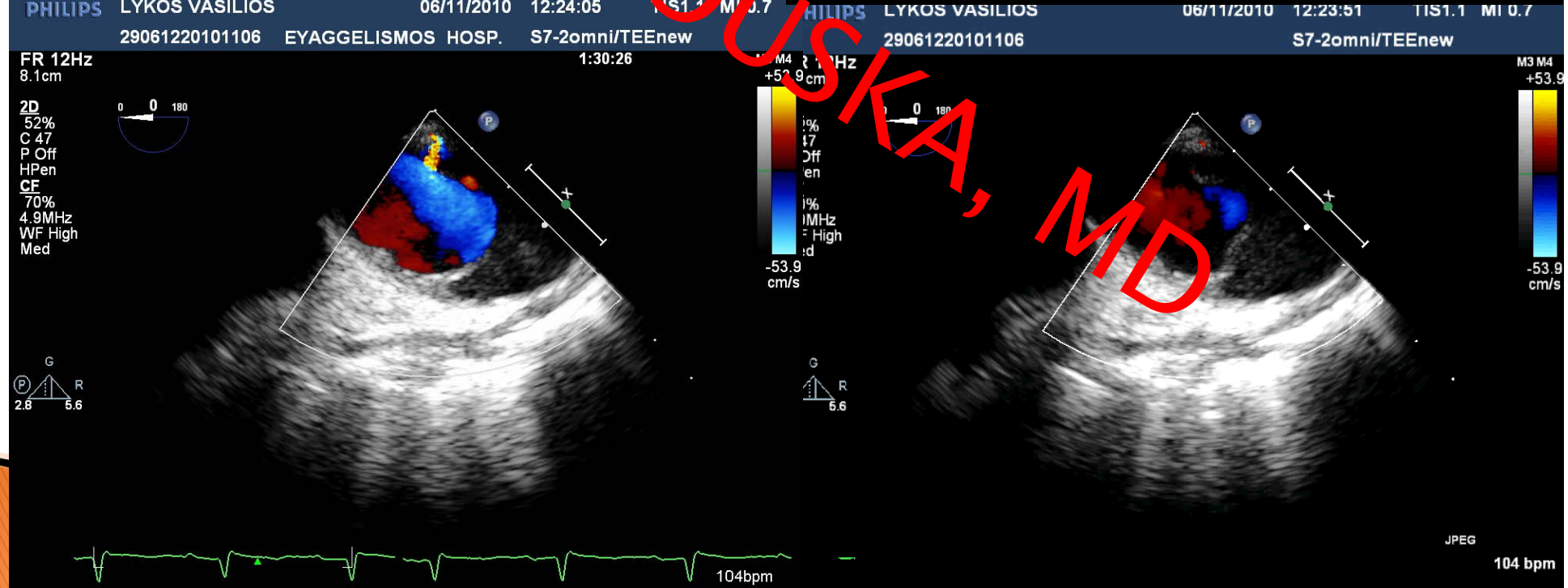


- Ιστίο (flap)
- Σημείο εισόδου (tear–entry point)
- Αληθής αυλός (True lumen)
- Ψευδής αυλός (False lumen)

Effie ROUSKA, MD







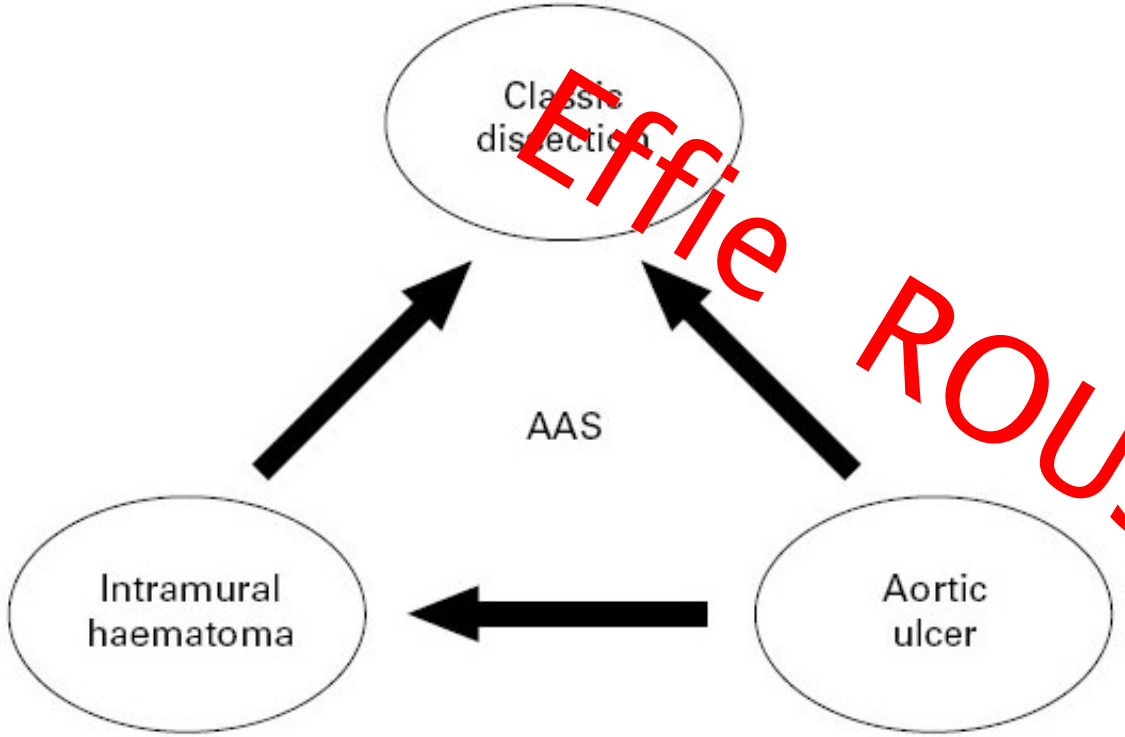
Effie ROUSKA, MD

Μορφές

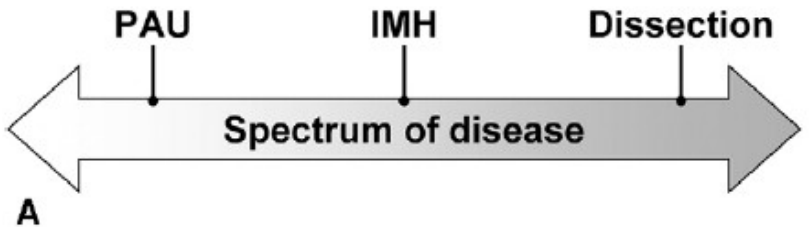
- *Οξύς διαχωρισμός*
  - *1 – 2 εβδομάδες*
- *Χρόνιος* διαχωρισμός
  - ≥ 2 εβδομάδες
- *Υποξεία μορφή διαχωρισμού*
  - *2 εβδομάδες – 2 μήνες*

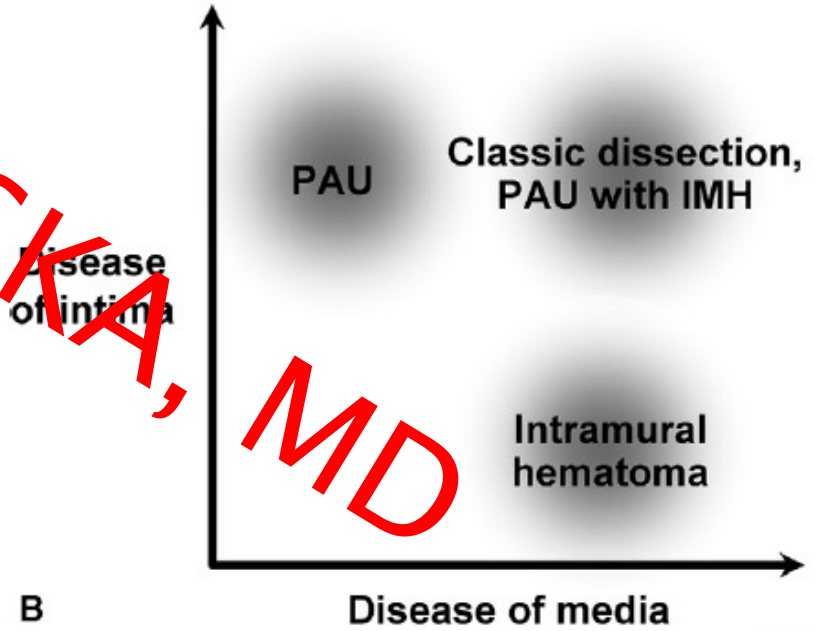
Effie ROUSKA, MD

Οξύ αορτικό σύνδρομο



Acute aortic syndrome (AAS). Arrows indicate the possible progression of each of these aortic lesions.





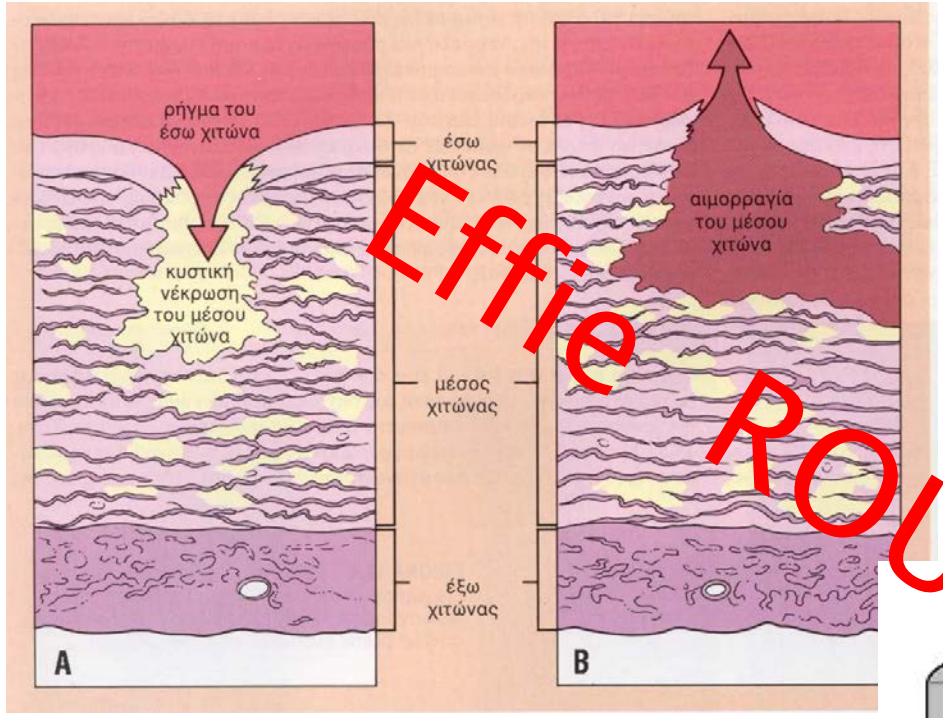
Effie ROUSKA, MD

Εμφάνιση

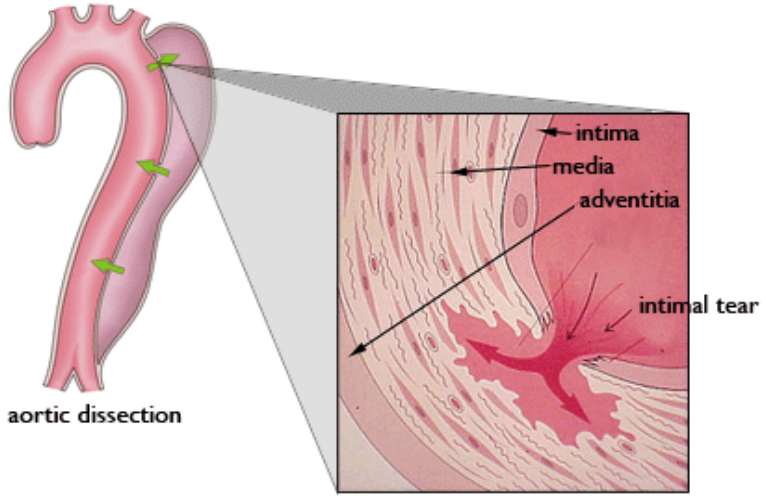
- Η πιο συχνή θανατηφόρος κατάσταση που αφορά την αορτή
- 0.5-2.95 % ανά 100000 ανά έτος
- 5000-7000 νέες περιπτώσεις ανά έτος (ΗΠΑ)
- 3χ πιο συχνός από τη ρήξη Ao Κοιλιακού ανευρύσματος
- Συχνότερα στους άνδρες (1.7-2.6:1)
- 5η με 6η δεκαετία της ζωής

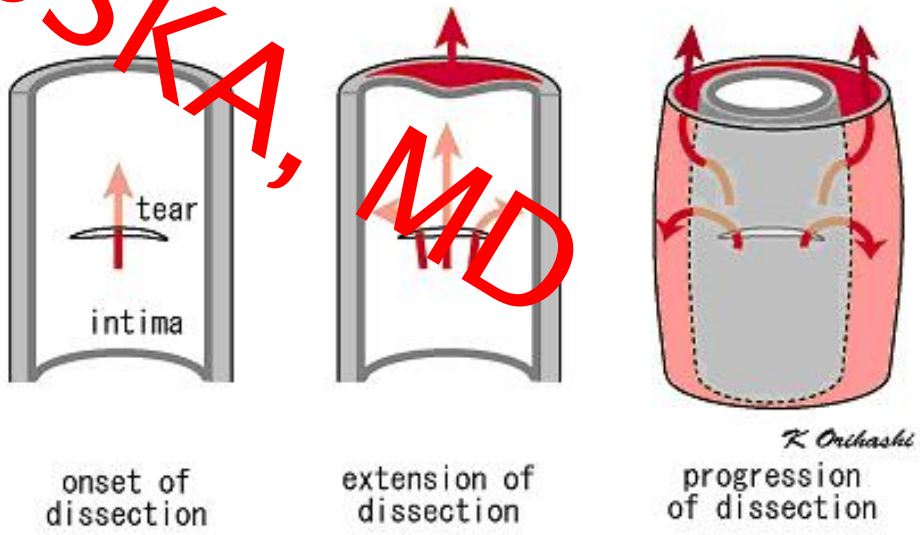
Effie ROUSKA, MD

Παθογένεια



primary tear *secondary tear*





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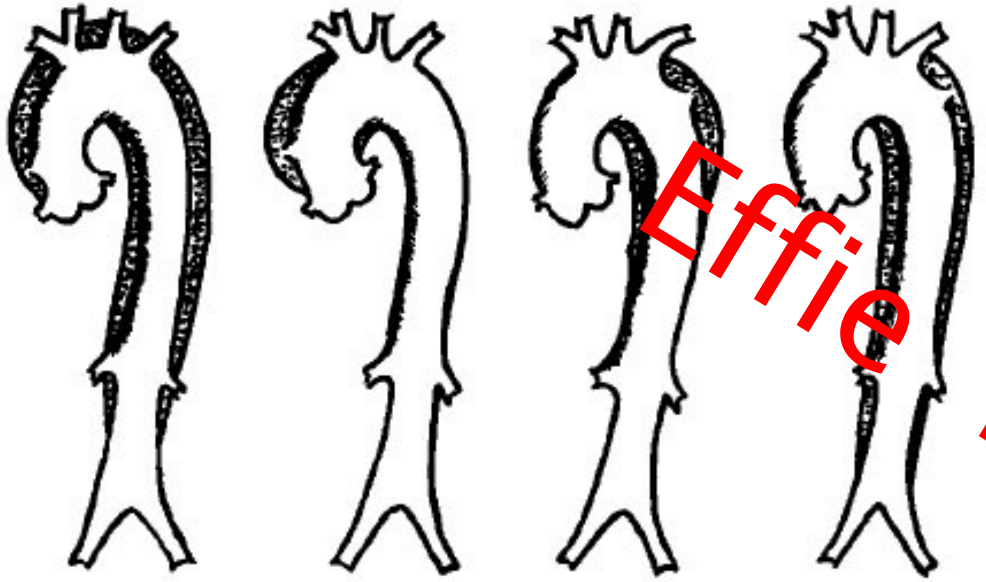
Ανατομική Ταξινόμηση

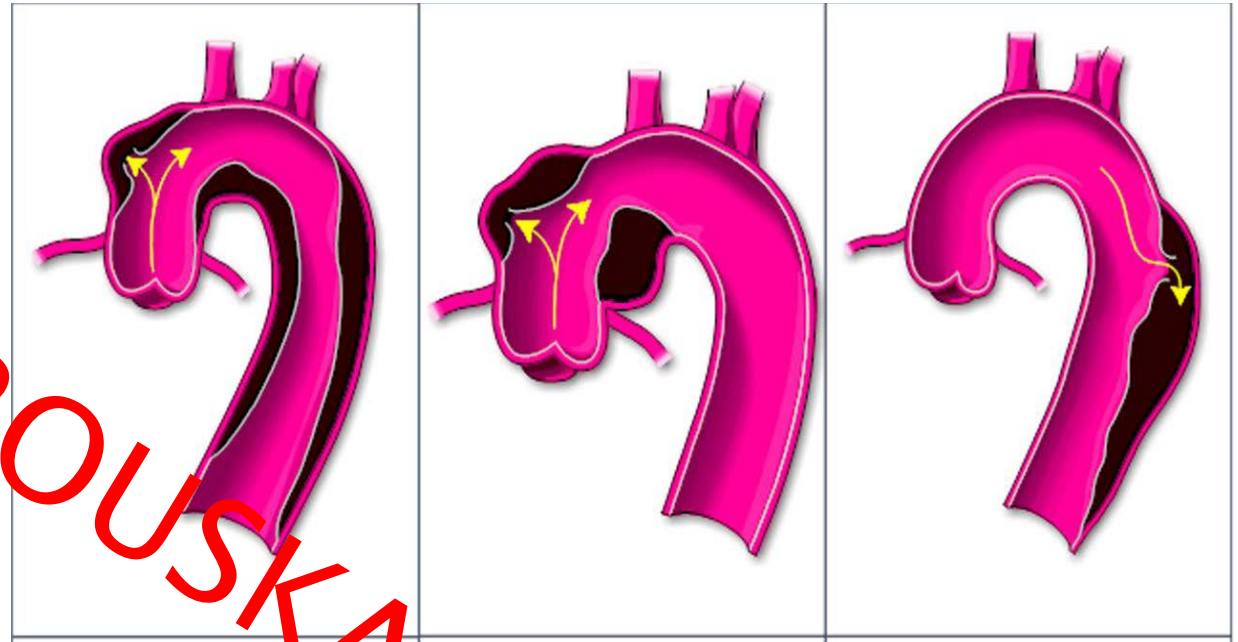


DeBakey Type I | DeBakey Type II | DeBakey Type IIIa | DeBakey Type IIIb

Stanford Type A | Stanford Type A | Stanford Type B | Stanford Type B

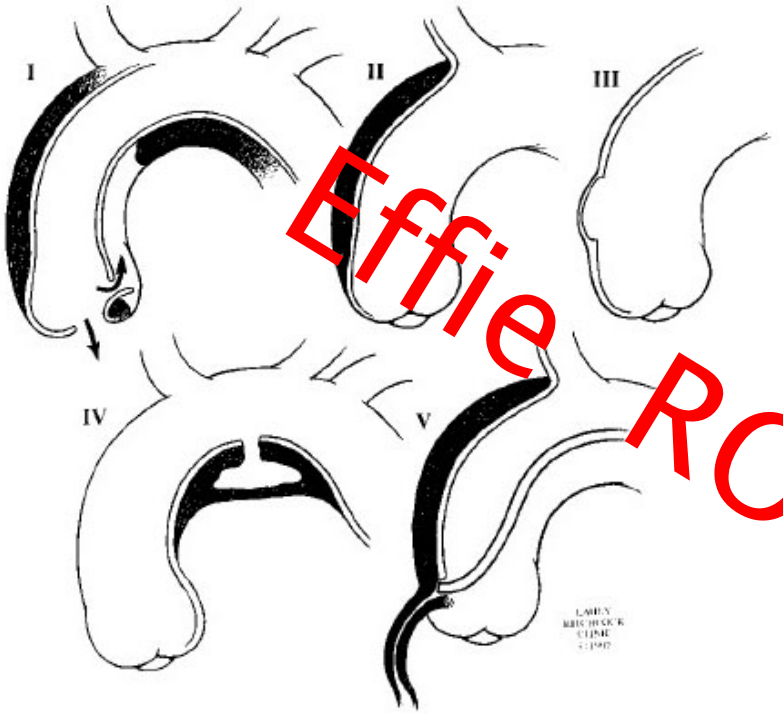
Types of aortic dissection.

Effie ROUSKA, MD

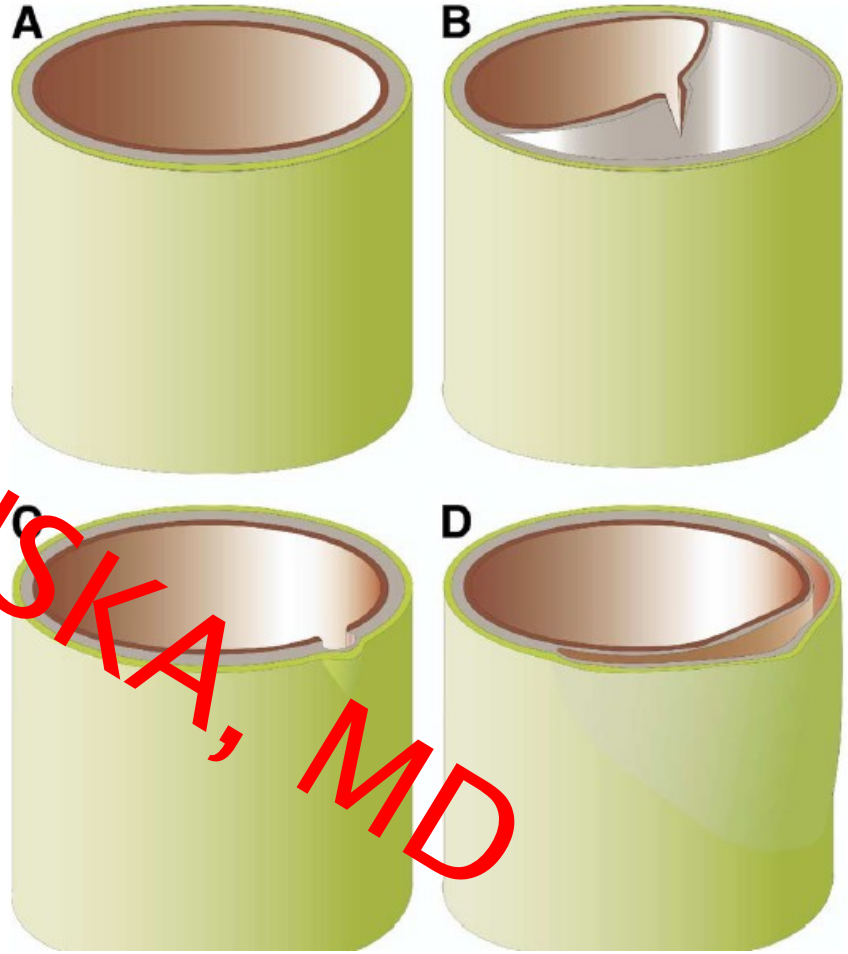


De Bakey Type I	De Bakey Type II	DeBakey Type III
Stanford Type A (Ascending aorta involved)		Stanford Type B
60%	10-15%	25-30%

Λειτουργική Ταξινόμηση



Classes of aortic dissection: class 1, classic dissection with flap between true and false aneurysm and clot in false lumen; 2, intramural hematoma; 3, limited intimal tear with eccentric bulge at tear site; 4, penetrating atherosclerotic ulcer with surrounding hematoma, usually subadventitial; 5, iatrogenic or traumatic dissection illustrated by coronary catheter causing dissection.



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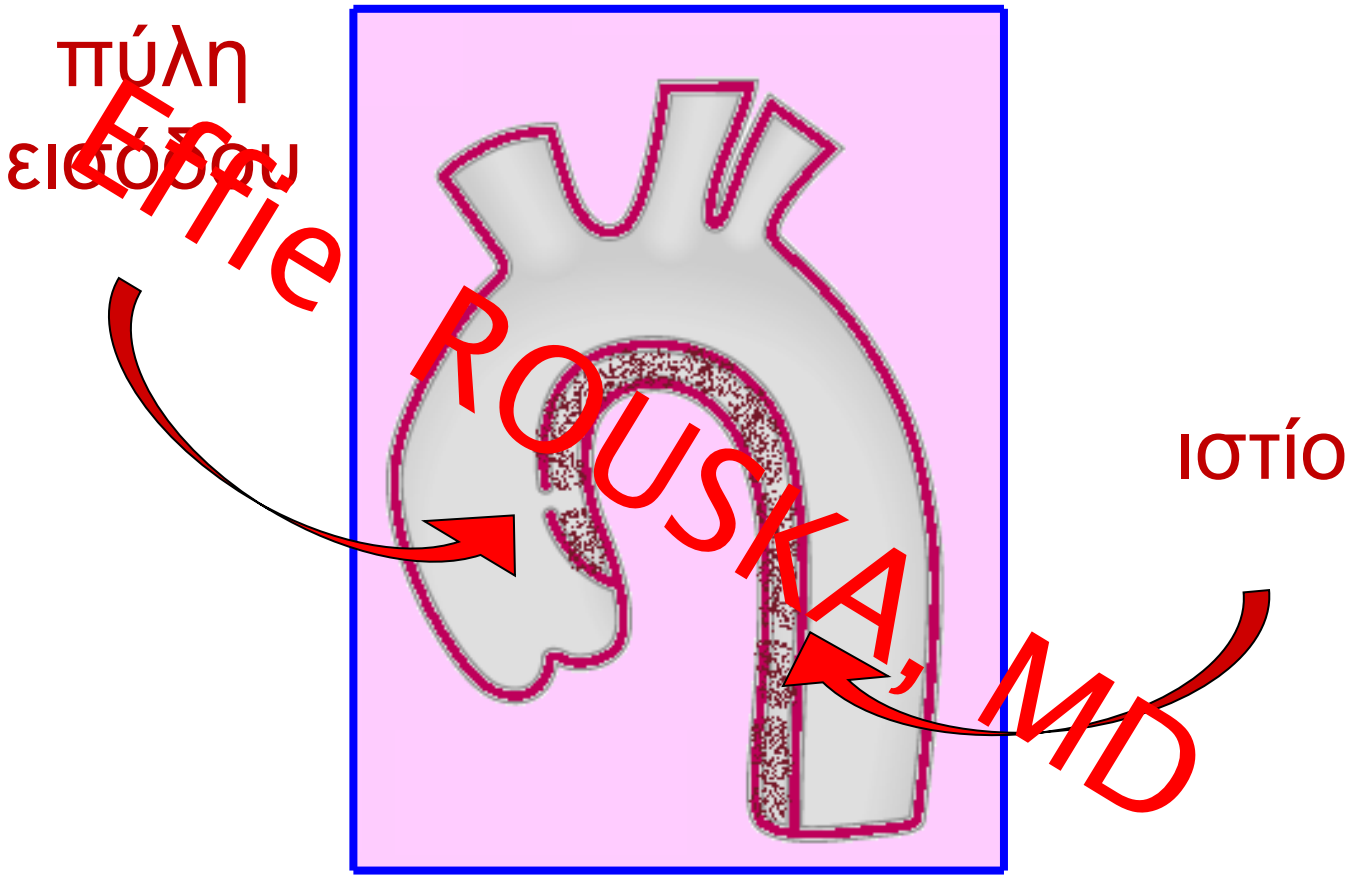
Svensson LG, et al, Circulation 1999;99:1331

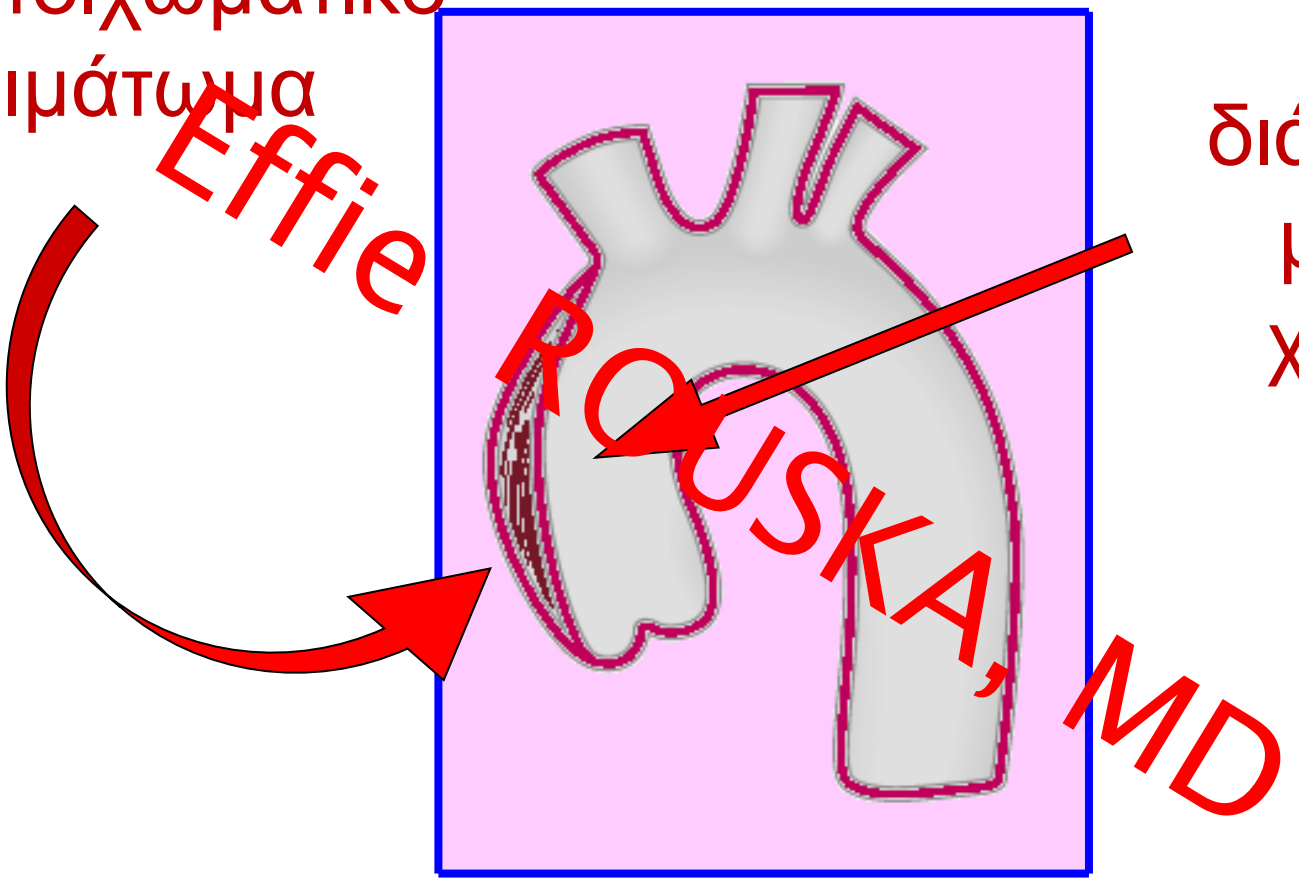
Class 1



κλασσικός αορτικός διαχωρισμός

Class 2

ενδοτοιχωματικό αιμάτωμα

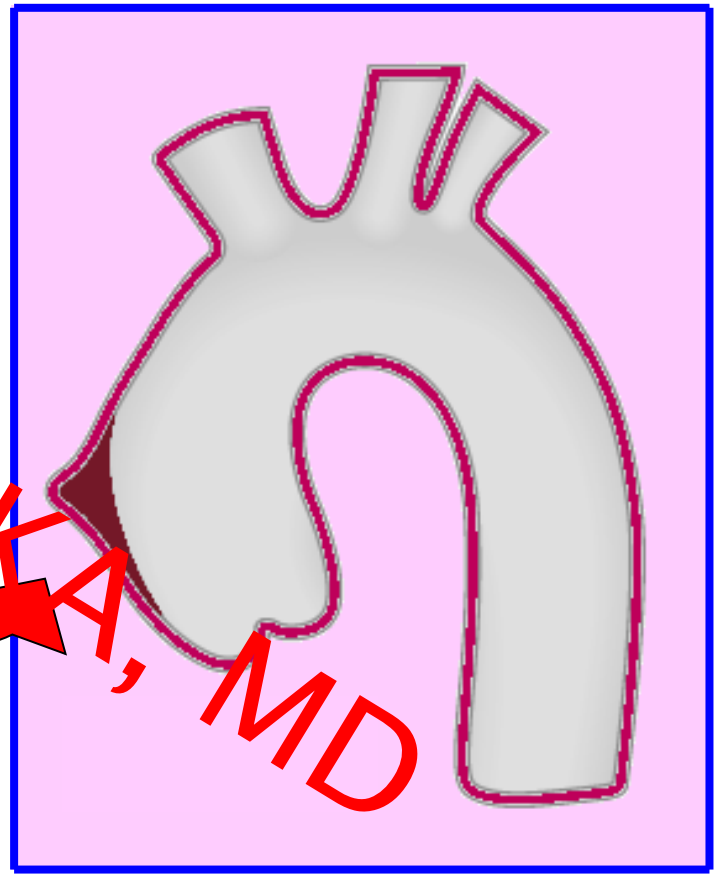


διάσπαση μέσου χιτώνα

ενδοτοιχωματικό αιμάτωμα

Class 3

προσεκβολή αορτικού τοιχώματος



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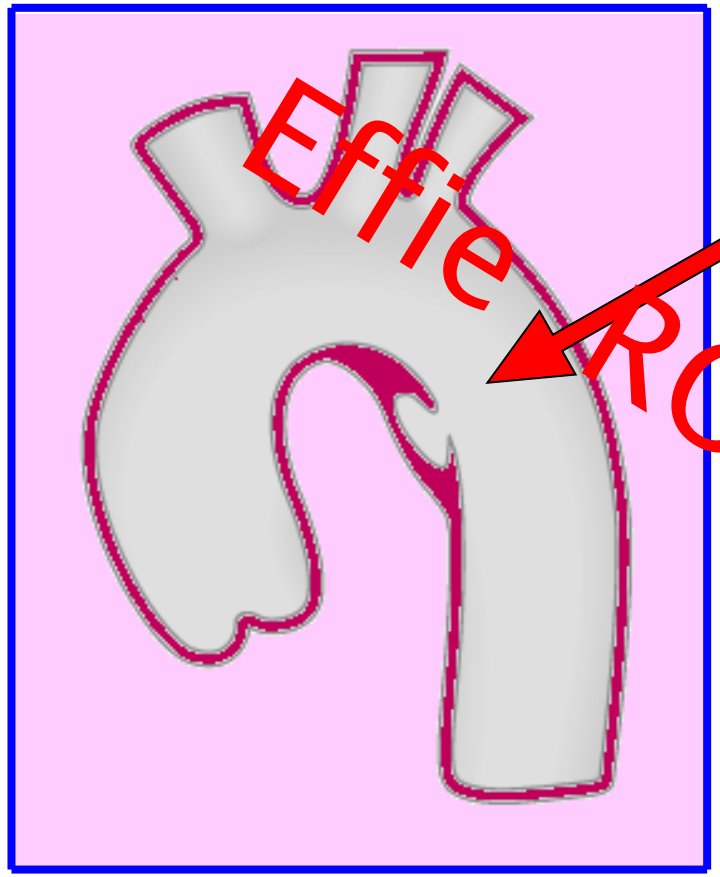
περιορισμένος διαχωρισμός

Class 4



διάτρηση

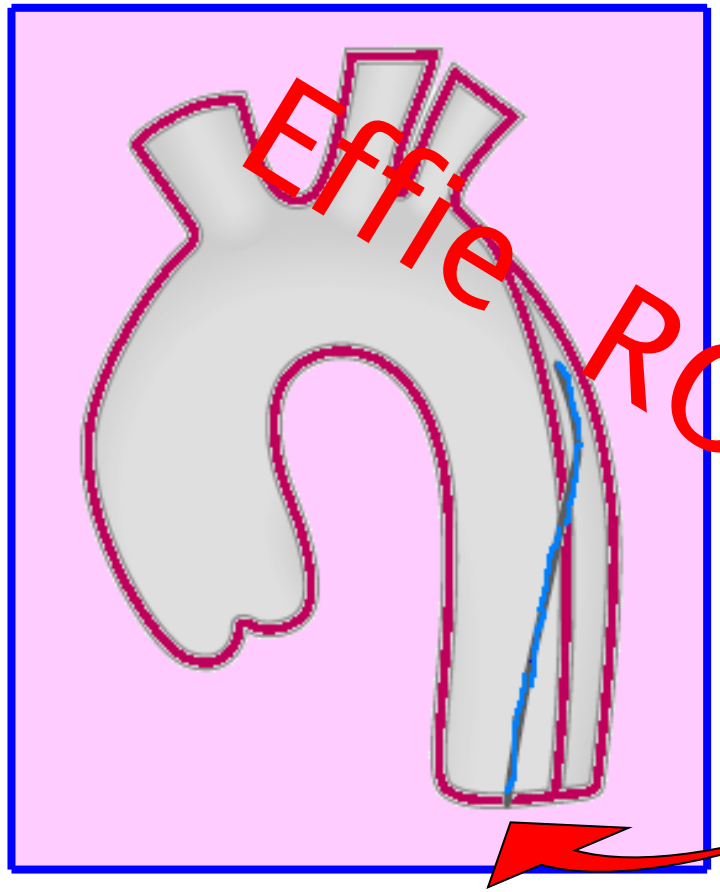
προς τον έξω χιτώνα

με τοπικό αιμάτωμα

Effie ROUSKA, MD

διατιτραίνον αθηροσκληρωτικό έλκος

Class 5



καθετήρας

ιατρογενής / τραυματικός διαχωρισμός

Αιτιολογία – Παράγοντες κινδύνου

Effie ROUSKA, MD

Conditions associated with increased aortic wall stress

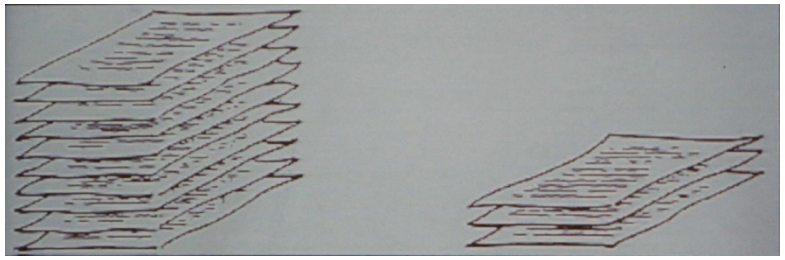
- Hypertension, particularly if uncontrolled
- Pheochromocytoma
- Cocaine or other stimulant use
- Weight lifting or other Valsalva maneuver
- Trauma
- Deceleration or torsional injury (e.g., motor vehicle crash, fall)
- Coarctation of the aorta

Conditions associated with aortic media abnormalities

- Genetic
  - Marfan syndrome
  - Ehlers-Danlos syndrome, vascular form
  - Bicuspid aortic valve (including prior aortic valve replacement)
  - Turner syndrome
  - Loeys-Dietz syndrome
  - Familial thoracic aortic aneurysm and dissection syndrome
- 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

Φυσική Εξέλιξη (θάνατος)

- 40% immediately
- 70% in 24h
- 94% in 1 week
- 100% in 5 weeks



Shennan 1934

- 33% in 24h
- 50% in 48h
- 80% in 1 week
- 95% in 1 month

Lindsay and Hurst 1967

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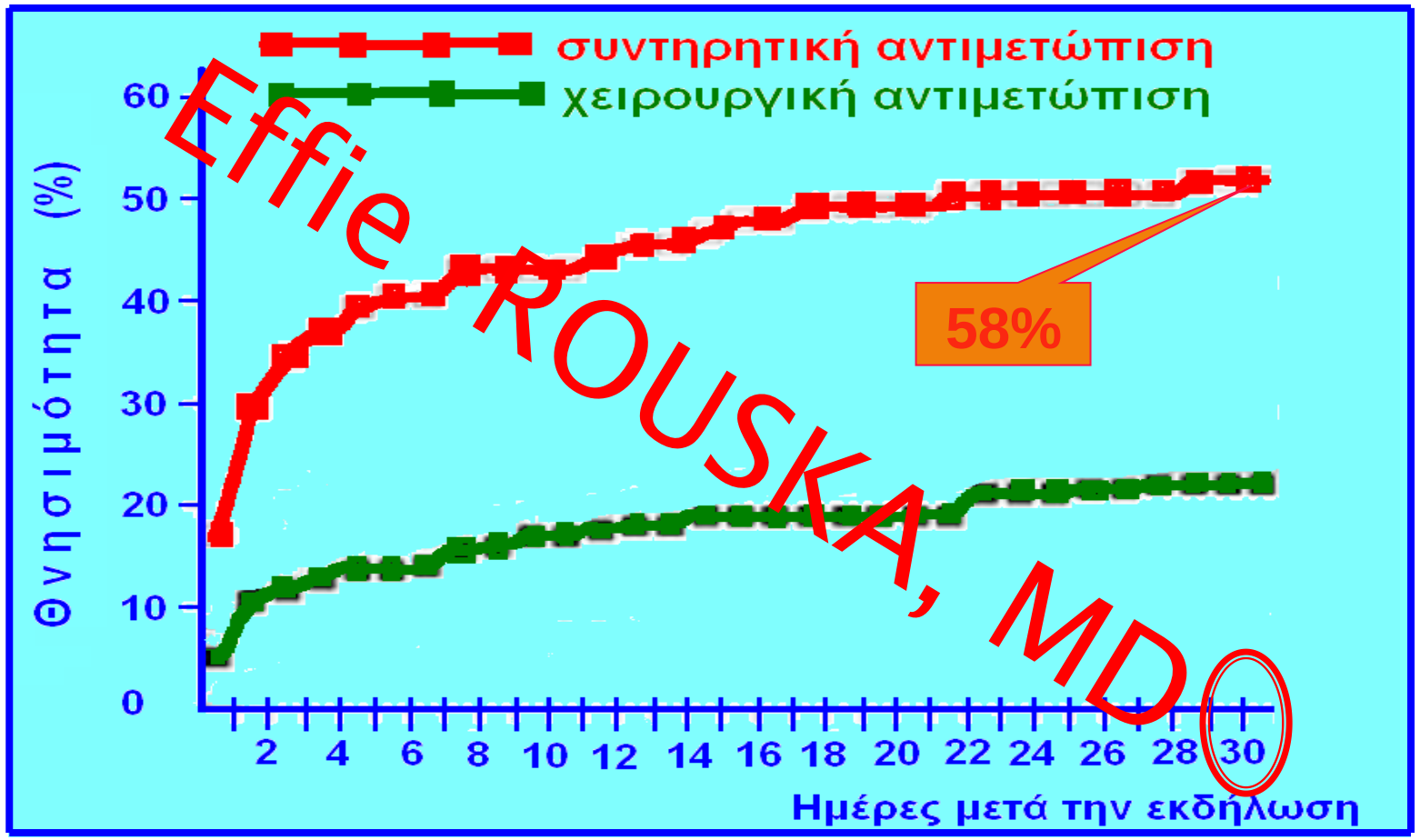
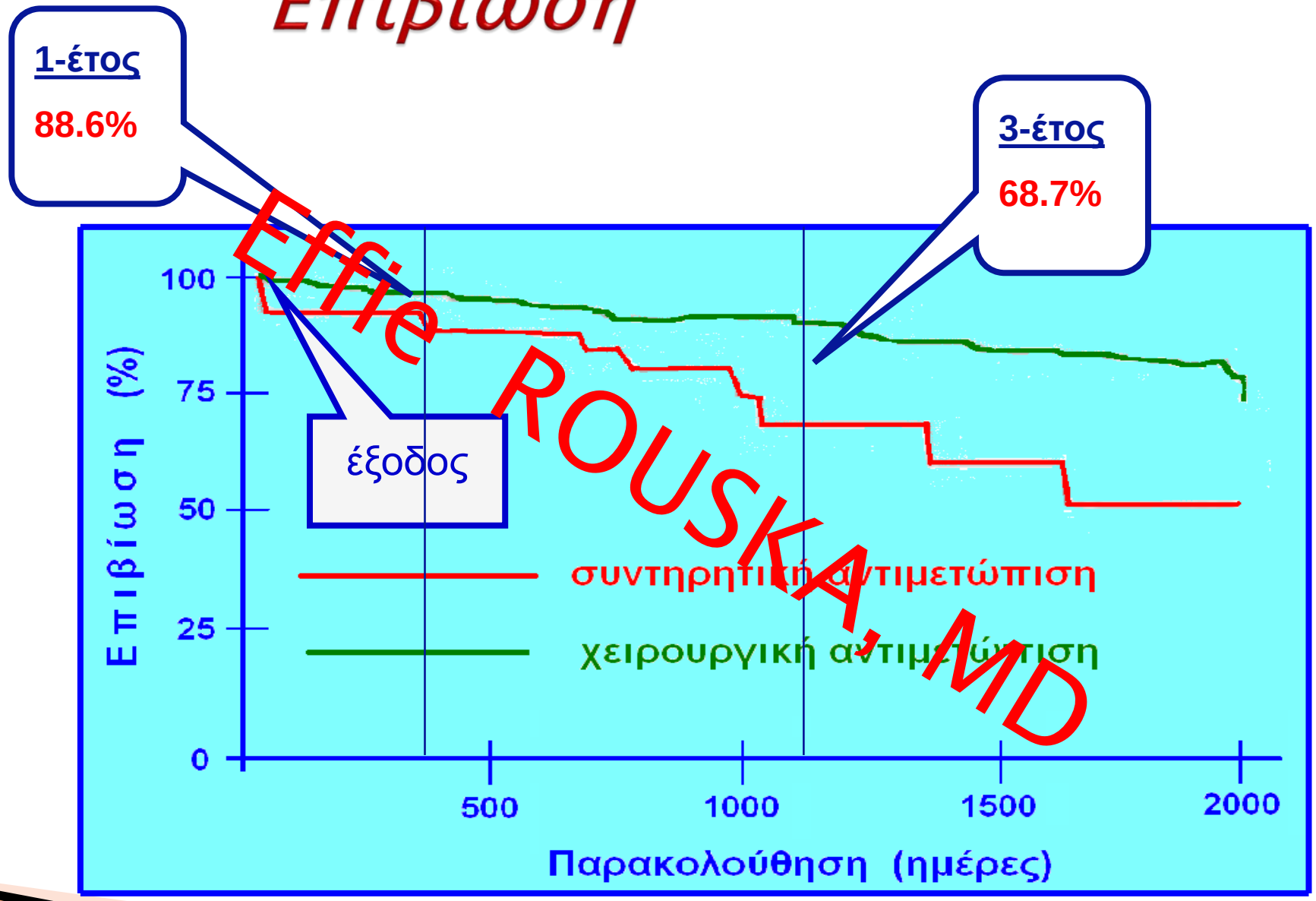
Θνητότητα



IRAD study, JAMA 2000

Επιβίωση



IRAD study, Circulation 2006

Untreated type A AoA

- Intrapericardial rupture → Tamponade
- Acute AR → LV failure
- Coronary compromisation → m. ischemia
- Ao branches occlusion
- Free rupture

Effie ROUSKA, MD

Κλινικά Χαρακτηριστικά

Aortic dissection — common presenting symptoms

Pain
- Pain alone
- Pain with syncope
- Pain with signs of congestive heart failure

Pain with cerebrovascular accident (stroke)
Congestive heart failure without pain
Cerebrovascular accident without pain
Abnormal chest roentgenogram without pain
Pulse loss without pain

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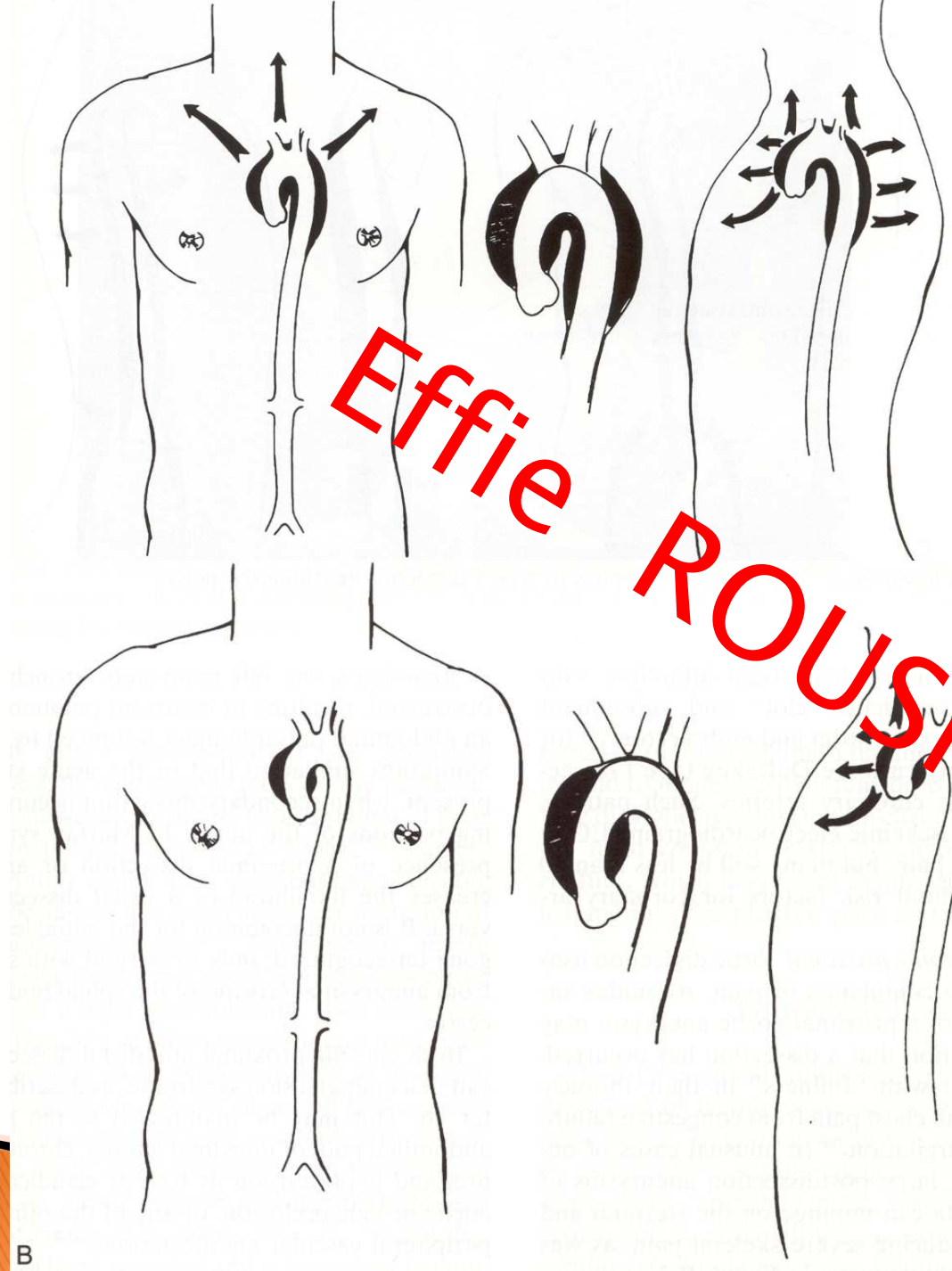
International Registry of Acute Aortic Dissection (IRAD) Physical Findings of 591 Patients With Type A Aortic Dissection

Presenting Hemodynamics and Clinical Findings	Frequency/Finding
Hypertensive	32%
Normotensive	45%
Hypotensive	14%
Shock	13%
Cardiac tamponade	5%
Murmur of aortic insufficiency	45%
Pulse deficits	26%
Pericardial friction rub	2%
Cerebrovascular accident	8%
Ischemic peripheral neuropathy	3%
Ischemic spinal cord damage	2%
Ischemic lower extremity	10%
Compromised consciousness	12%
Congestive heart failure	5%
First blood pressure systolic, mean	130 mm Hg
First blood pressure diastolic, mean	75 mm Hg

Αντανακλάσεις πόνου



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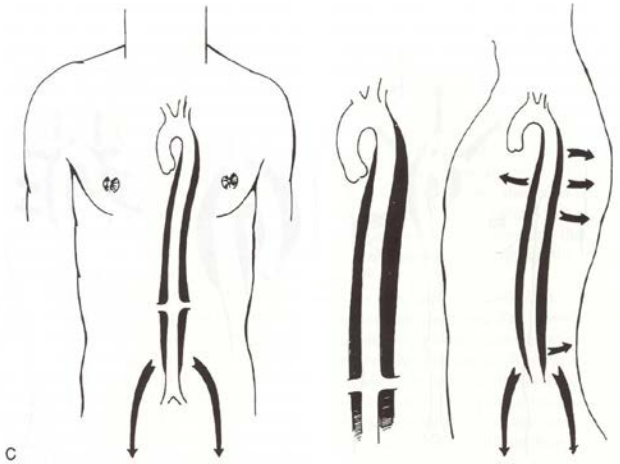
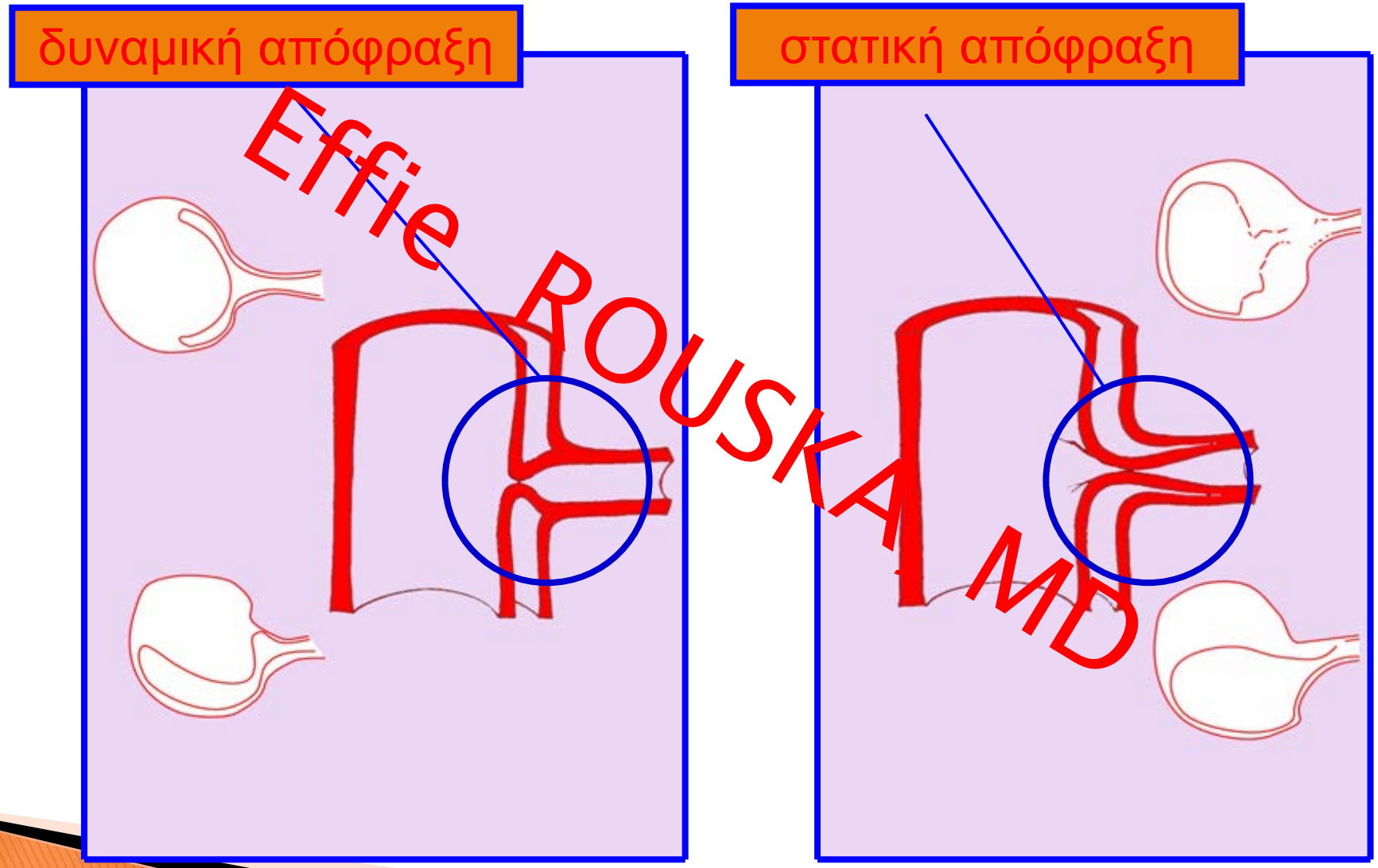


C

B

Μηχανισμοί Αρτηριακής Απόφραξης



Απόφραξη Αρτηριακών Κλάδων

Effie ROUSKA, MD

Hirst AE, Medicine 1955

End-Organ Complications of Acute Aortic Dissection

Type	End-Organ Complication
Cardiovascular	Aortic insufficiency
	Syncope
	Pericardial tamponade
	Myocardial ischemia or infarction
	Congestive heart failure
Neurologic	Ischemic stroke or transient ischemic attack
	Peripheral neuropathy
	Paraplegia/paraparesis
	Spinal ischemia
Pulmonary	Pleural effusion
	Aortopulmonary fistula with hemorrhage
Gastrointestinal	Mesenteric ischemia or infarction
	Aortoenteric fistula with hemorrhage
Renal	Renal failure
	Renal ischemia or infarction
Extremities	Limb ischemia

Διαφορική διάγνωση

Aortic dissection — differential diagnosis

Acute coronary syndrome with and without ST-elevation
Aortic regurgitation without dissection
Aortic aneurysms without dissection
Musculoskeletal pain
Pericarditis
Mediastinal tumours
Pleuritis
Pulmonary embolism
Cholecystitis
Atherosclerotic or cholesterol embolism

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Διάγνωση

- ΗΚΓ
- Εργαστηριακές Εξετάσεις
- Ακτινογραφία Θώρακος
- Άλλες Απεικονιστικές Μέθοδοι

Effie ROUSKA, MD

Ηλεκτροκαρδιογράφημα

Electrocardiography			
Normal	188 (30%)§	113 (31%)	301 (30%)
Left ventricular hypertrophy	139 (23%)§	56 (32%)*	195 (26%)
Myocardial ischaemia or infarction	149 (24%)§	38 (10%)‡	187 (17%)‡

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IRAD study

(%)

- Μεταβολές ST / T — 41.4
- Υπερτροφία αριστεράς κοιλίας — 26.1
- Ισχαιμία — 15.1
- Πρόσφατο Έμφραγμα — 3.2

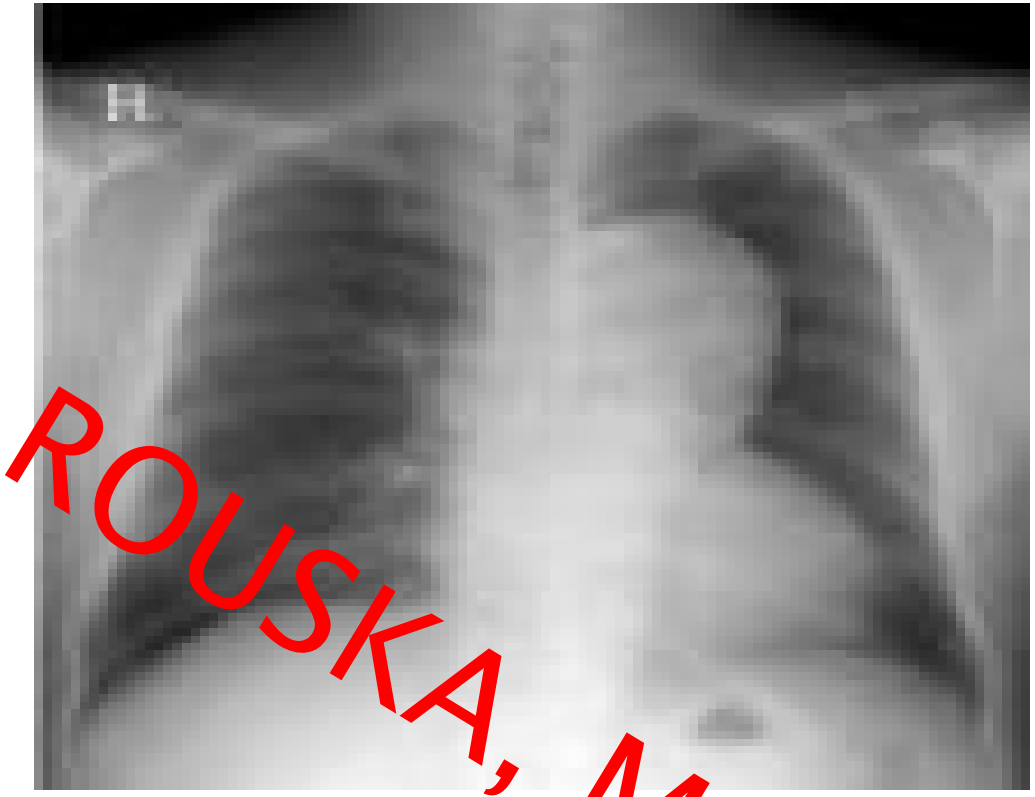
Εργαστηριακές εξετάσεις
Διαγνωστικοί Δείκτες

- Πολλοί διαθέσιμοι αλλά :
  - όχι ειδικοί
  - πολύ ευαίσθητοι
  - κανένας δεν μπορεί να χρησιμοποιηθεί σαν αυτόνομος δείκτης

Effie ROUSKA, MD

- **smooth muscle myosin heavy chain protein**
- **D-dimers**
- **Potentional markers**
  - Circulating cells
    - CD-28 null t-cells
    - CD-3+, CD-4+, CD-16+, CD-56+
  - Plasma/ serum markers
    - Metalloproteases
      - MMP-9
    - Markers of collagen turn over
      - PIII NP
      - EP
    - Inflammation markers
      - IL-1, IL-6
      - IFN-g, TNF-a
    - Other markers
      - Osteoprogerin
      - Osteopontin

Ακτινογραφία Θώρακος



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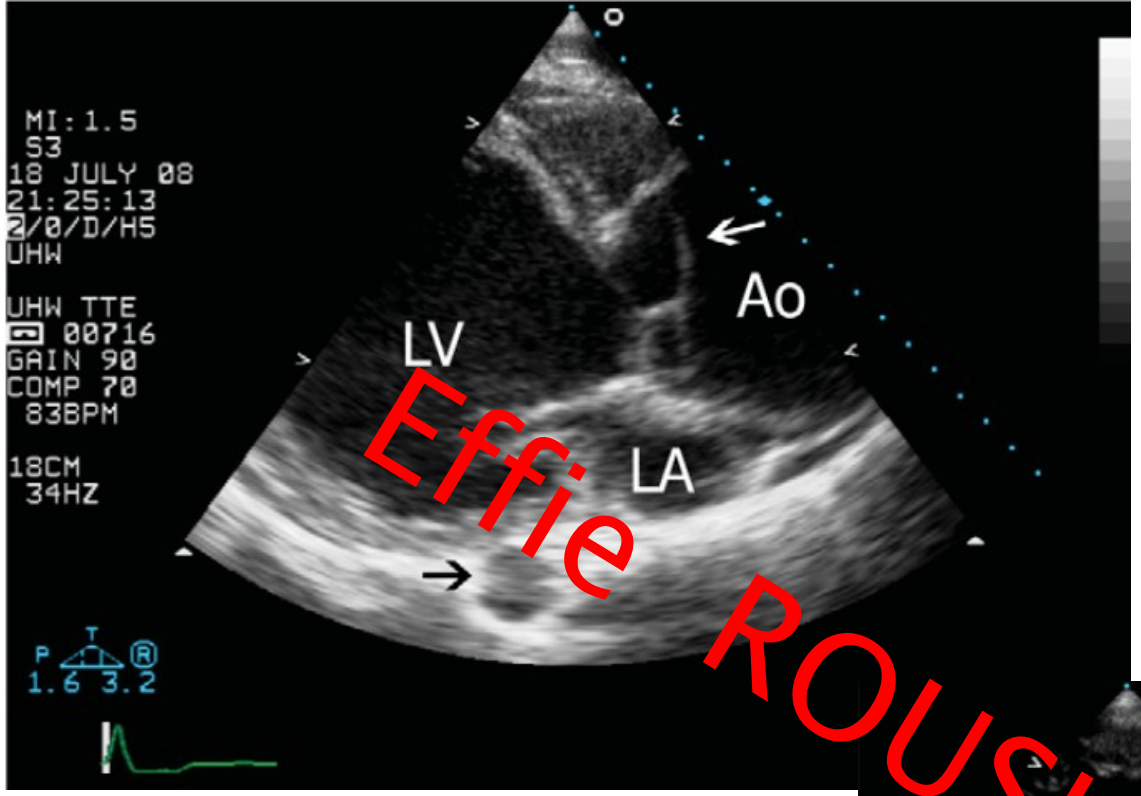
Chest radiography			
Widened mediastinum	331 (63%)	202 (56%)	533 (60%)
Abnormal aortic contour	124 (47%)*	171 (49%)	295 (48%)
Normal	67 (11%)‡	74 (21%)	141 (16%)‡

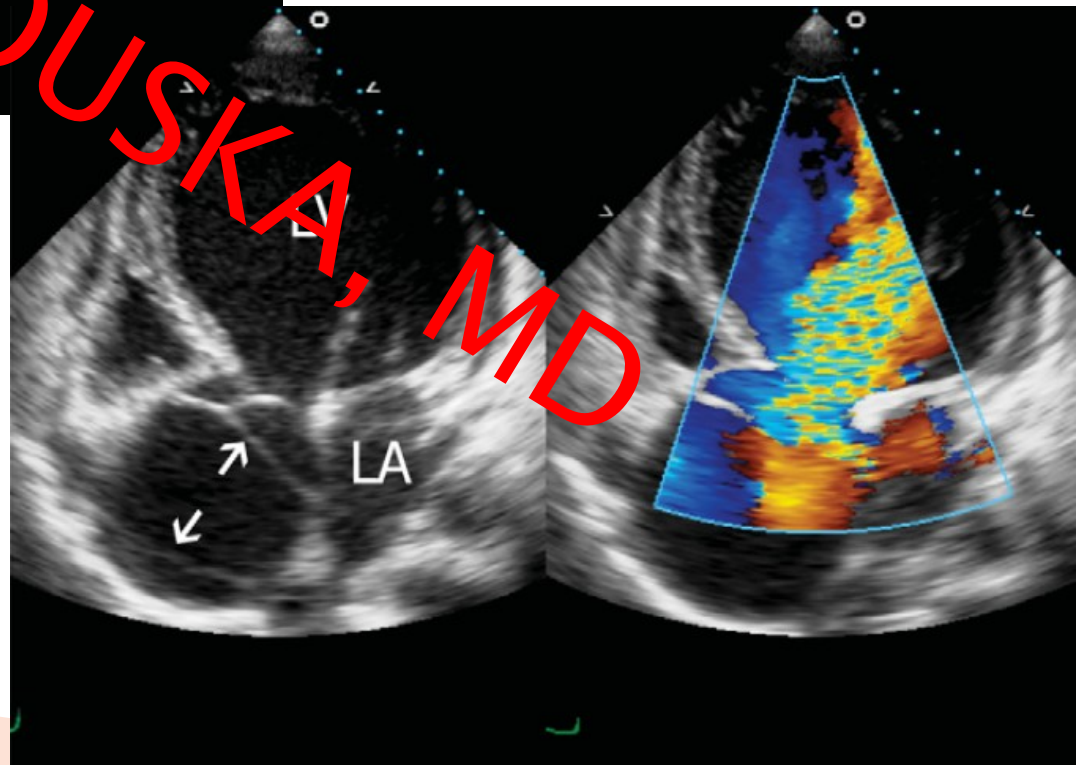
Άλλες Απεικονιστικές Μέθοδοι

- Αορτογραφία
- Ηχωκαρδιογραφία
- Αξονική Τομογραφία
- Μαγνητική Αγγειογραφία

Effie ROUSKA, MD

TTE





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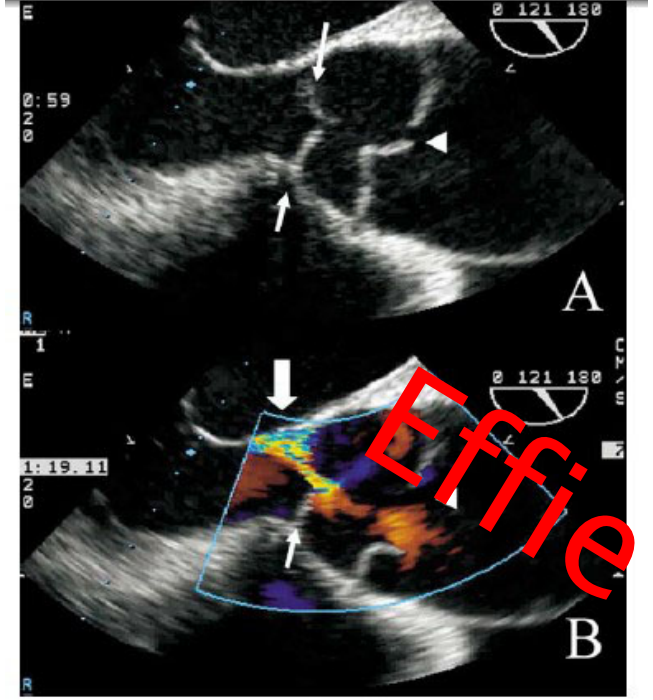
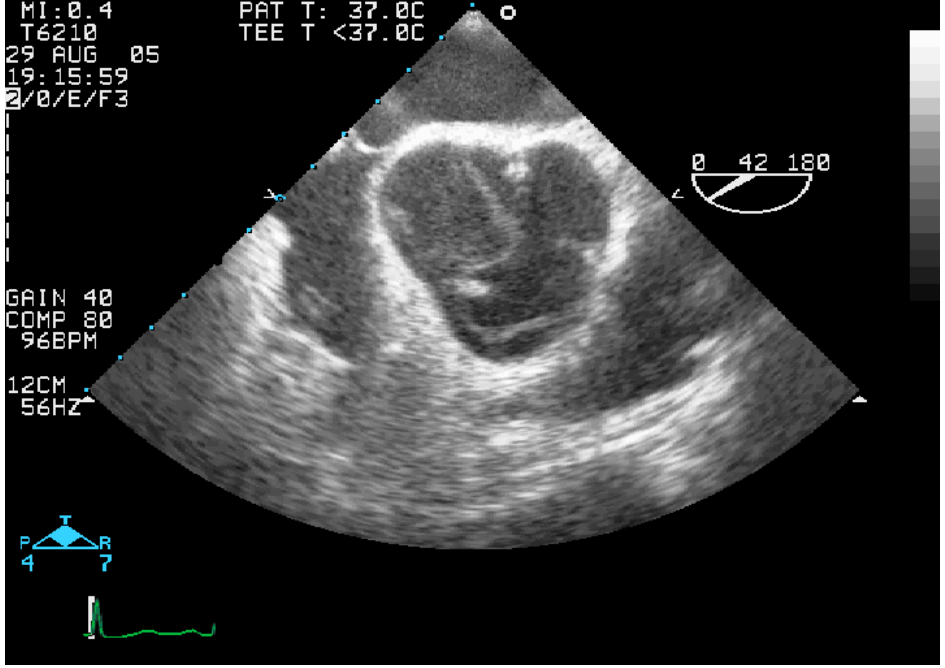
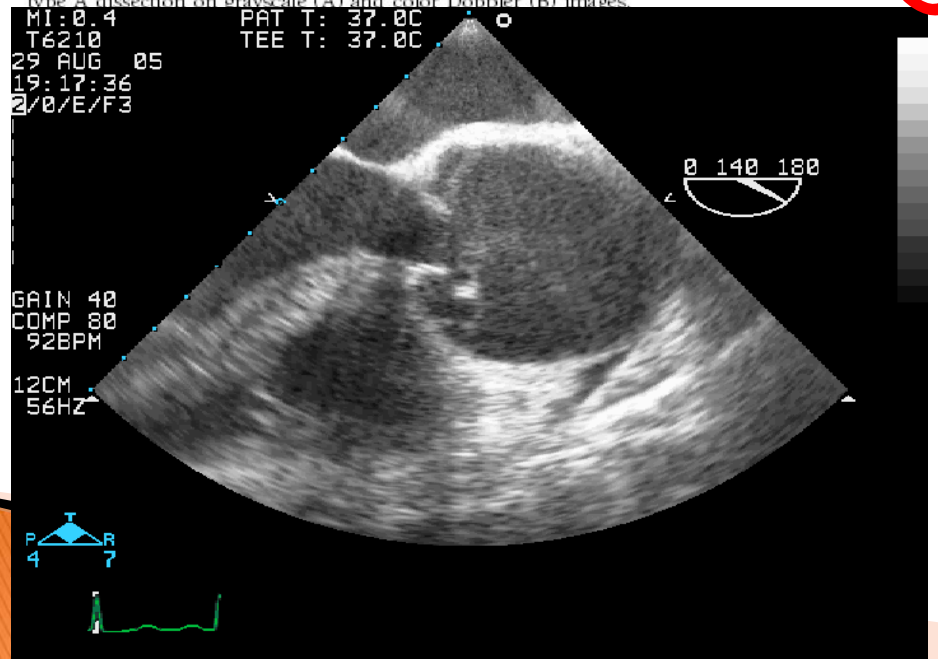
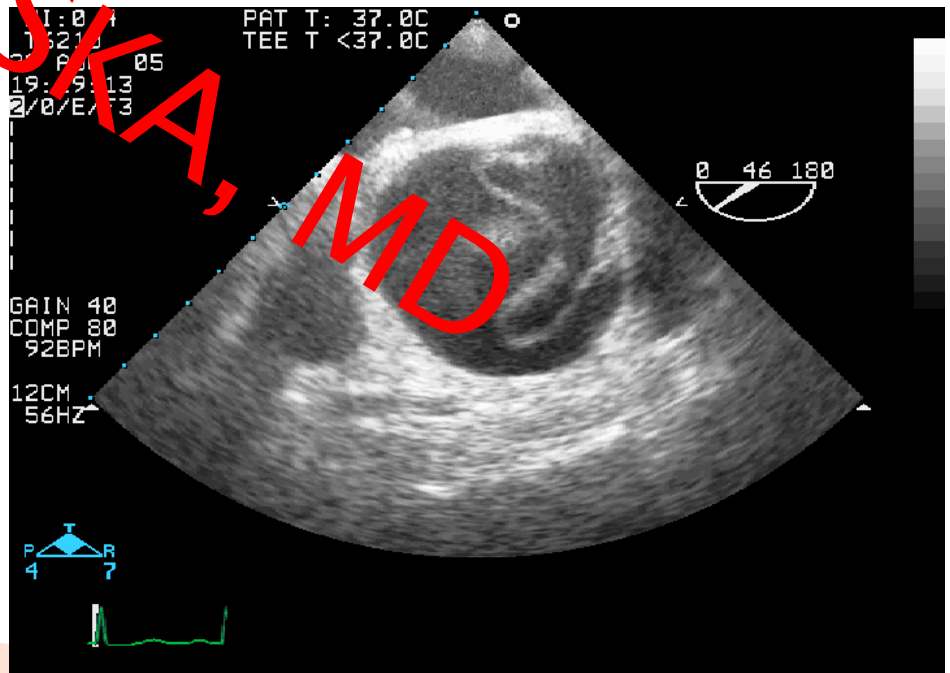
TOE



Images courtesy of Susan E. Wiegers, MD, and Victor A. Ferrari, MD, Hospital of the University of Pennsylvania. TEE reveals type A dissection on grayscale (A) and color Doppler (B) images.







Effie ROUSKA, MD

Ενδαγγειακή ηχωκαρδιογραφία (IVUS)

- εκτίμησης ολόκληρης της αορτής
- υψηλή ευαισθησία
- υψηλή ειδικότητα

- υψηλή επεμβατικότητα
- δυσκολία στον εντοπισμό του intimal tear

Effie ROUSKA, MD

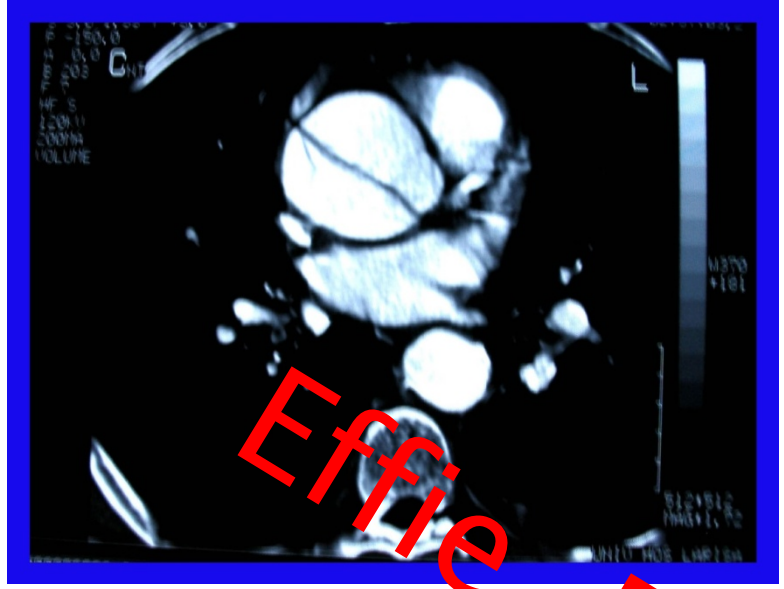
Αξονική Τομογραφία

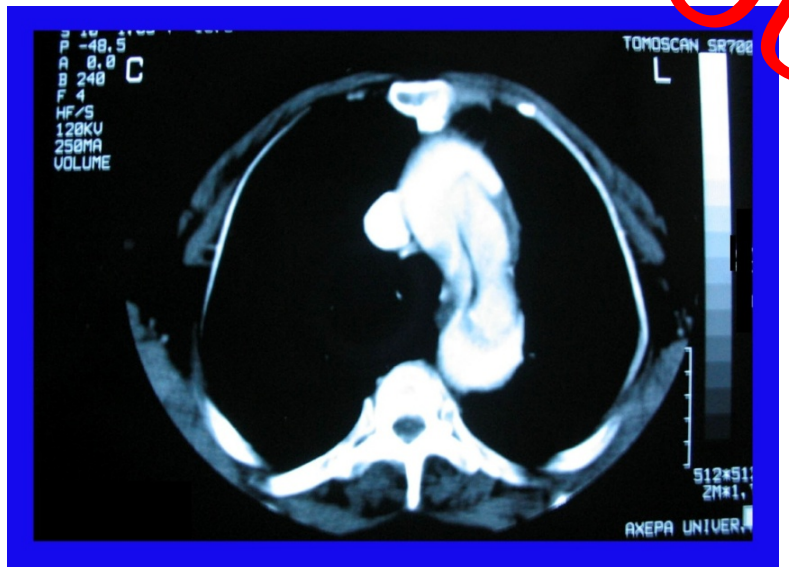
- πιο διαδεδομένη
- υψηλή ευαισθησία και ειδικότητα
- νέες τεχνολογικές εξελίξεις
  - ελικοειδής τεχνική
  - πολυτομικοι τομογράφοι
- πληροφορίες για
  - τη συμμετοχή ή όχι της κοιλιακής κορτής και των κλάδων της
  - ύπαρξη ή μη αιμοπερικαρδίου ή πλευριτικής συλλογής
  - τη συμμετοχή των τραχηλικών κλάδων

- *<u>Μειονεκτήματα</u>*
- η ακτινοβολία και η χορήγηση σκιαστικού
- η απεικονιστική της αορτική ρίζα και αορτικής βαλβίδας

Effie ROUSKA, MD

CTA





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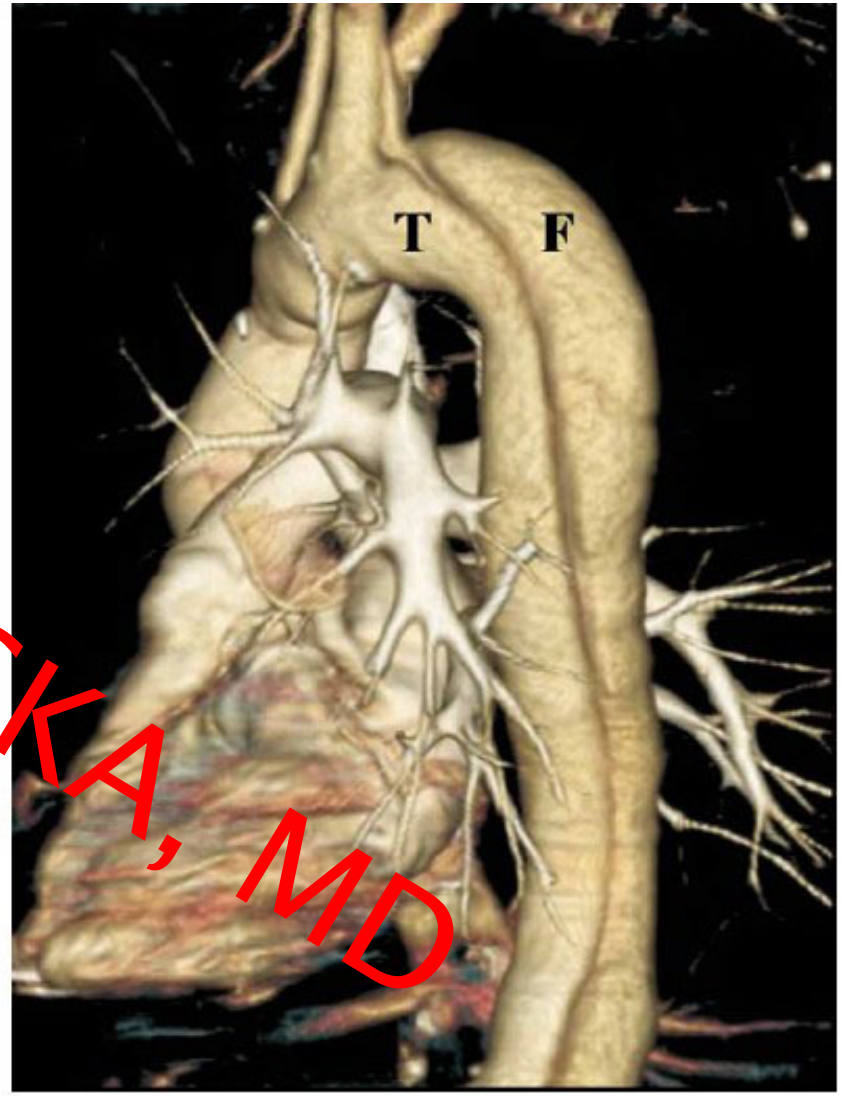
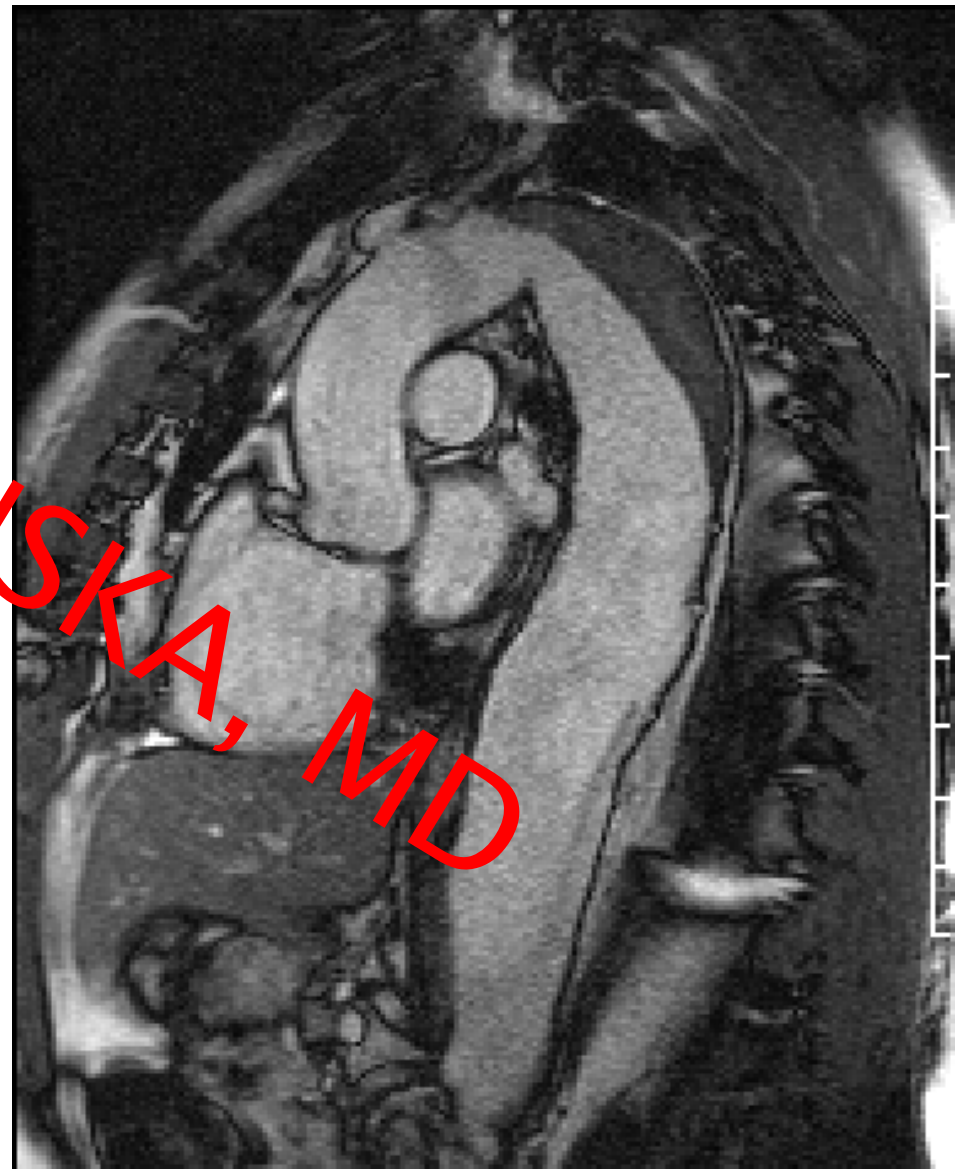


Volume-rendered enhanced CTA reveals true (T) and false (F) lumens of dissection in the descending thoracic aorta.

CTA



Effie ROUSKA, MD

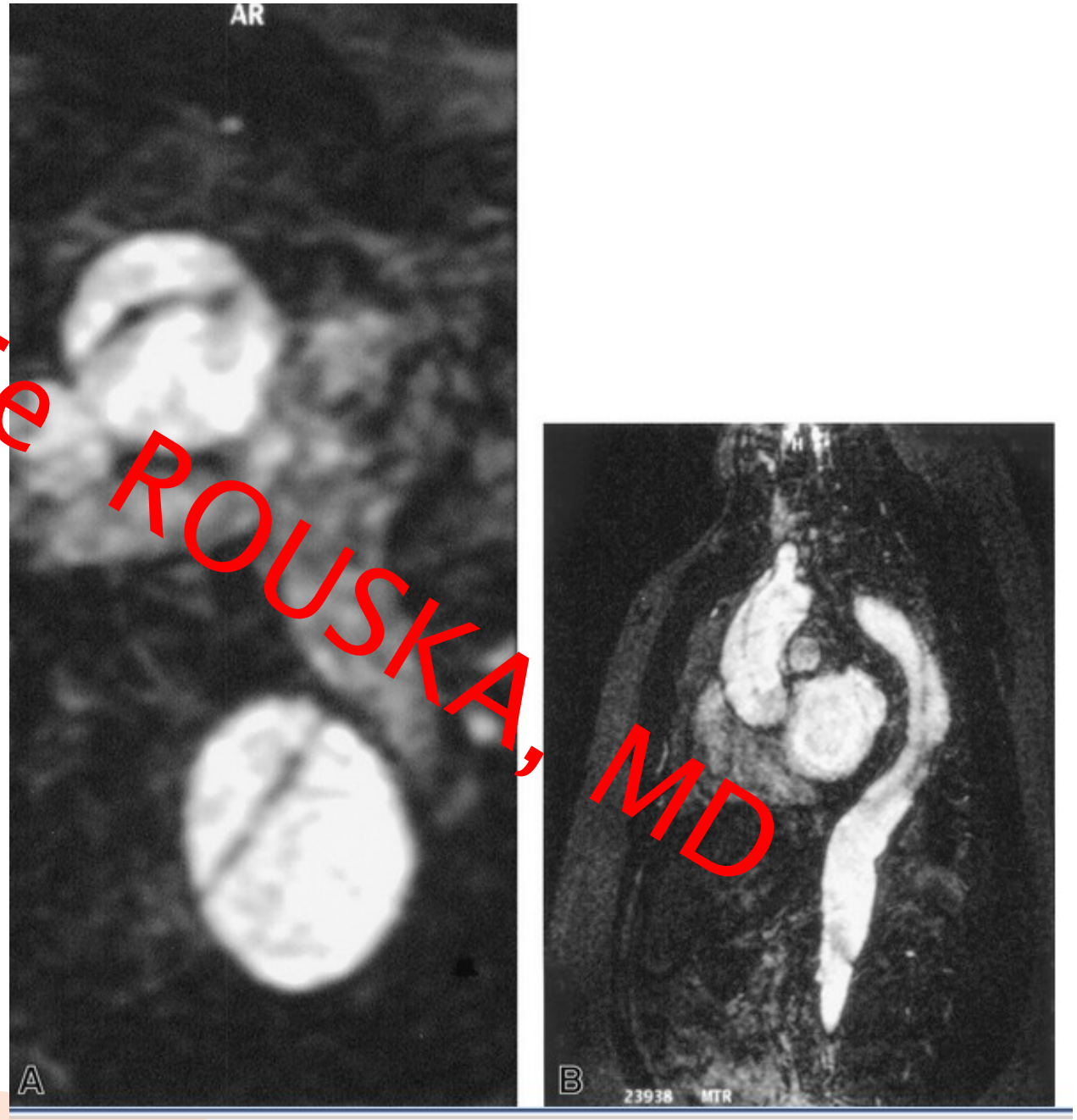
Μαγνητική Αγγειογραφία

- ως πρώτη διαγνωστική τεχνική μόνο στο 2%
- υψηλής ανάλυσης και ευκρίνειας εικόνες με λεπτομέρειες για όλες τις ανατομικές θέσεις
- δεν επιβαρύνει τη νεφρική λειτουργία
-
- *Μειονεκτήματα*
- δεν είναι πάντοτε διαθέσιμη
- αδυναμίας συνεργασίας και ιατρικής παρακολούθησης στη διάρκεια της διενέργειας της μεθόδου

Η MRA δεν έχει θέση στην επείγουσα διαγνωστική

Effie ROUSKA, MD

MRI



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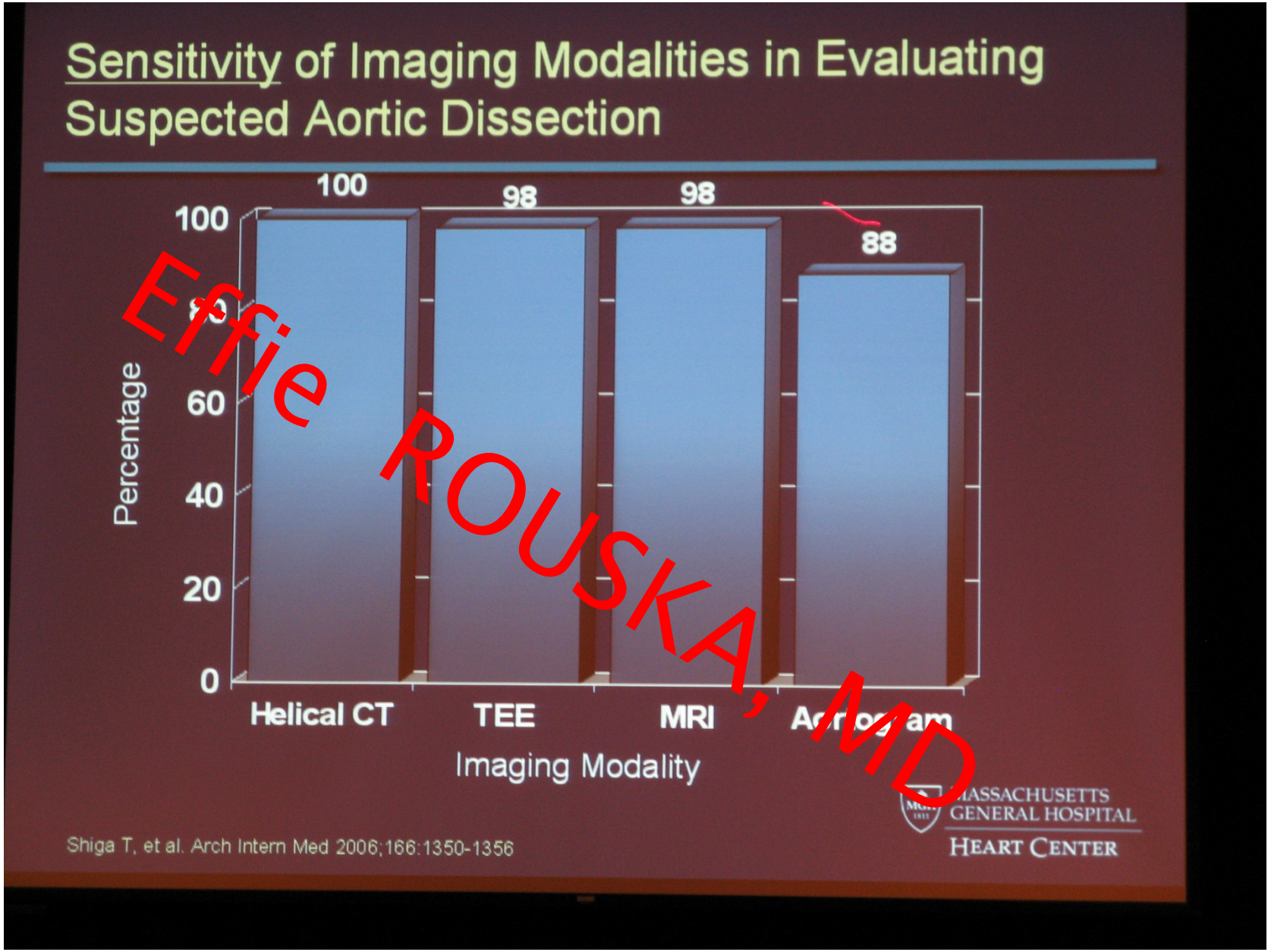
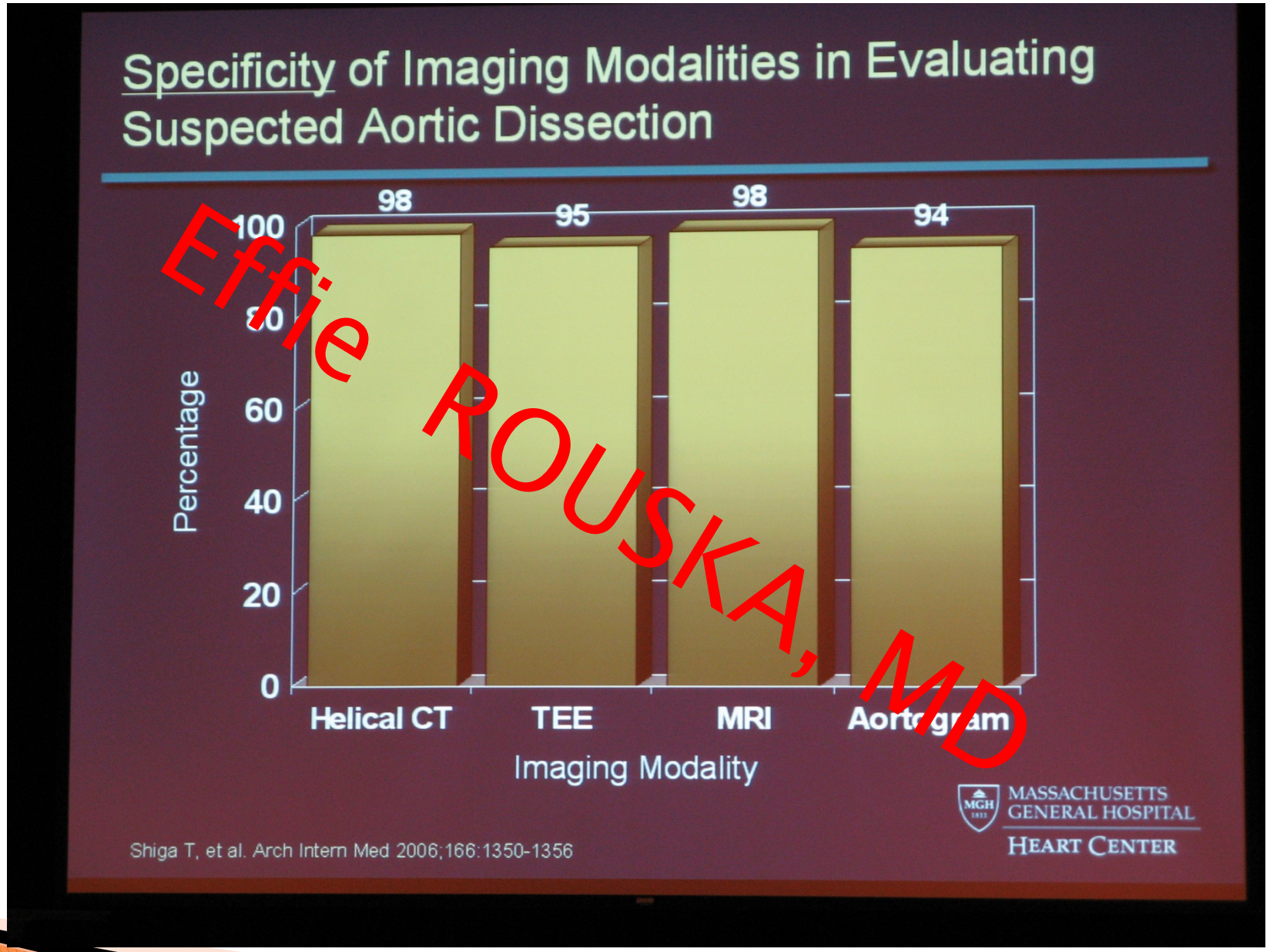

Sensitivity of Imaging Modalities in Evaluating Suspected Aortic Dissection
100
98
98
88
100
60
40
20
0
Percentage
Helical CT
TEE
MRI
Imaging Modality
Shiga T, et al. Arch Intern Med 2006;166:1350-1356
MASSACHUSETTS GENERAL HOSPITAL
HEART CENTER
Effie ROUSKA, MD


Specificity of Imaging Modalities in Evaluating Suspected Aortic Dissection
98
95
98
94
100
60
40
20
0
Percentage
Helical CT
TEE
MRI
Aortogram
Imaging Modality
Shiga T, et al. Arch Intern Med 2006;166:1350-1356
MASSACHUSETTS GENERAL HOSPITAL
HEART CENTER
Effie ROUSKA, MD

Comparison of Modalities

	CT	TEE	MRI
Accuracy with classic dissection	+++	+++	+++
Accuracy with dissection variants	+++	+	+++
Visualization of entire aorta	+++	−	+++
Branch artery involvement	+++	+	+++
Coronary involvement	++	++	++
Aortic valve involvement	+	+++	+
Entry/reentry tear localization	+++	+	+++
Rupture	+++	++	+++
Objectivity/reproducibility	+++	+	+++
Portability	−	+++	−
Availability	+++	++	+
Speed	+++	++	+
Ease of monitoring patients	+++	+++	−
Patient comfort/lack of invasiveness	+++	−	+

Key: +++, excellent; ++, good; +, fair; −, poor.

Effie ROUSKA, MD

Βασικοί στόχοι

- Επιβεβαίωση της διάγνωσης του διαχωρισμού
  - εντοπισμό του ιστίου
- Διευκρίνιση της έκτασης του διαχωρισμού
- Διαφοροδιάγνωση του ψευδούς και του αληθούς αυλού
- Εντοπισμός του πρώτου σημείου ρήξεως
- Διευκρίνηση της επικοινωνίας ή όχι του διαχωρισμού (communicating, non-communicating)
- Εκτίμηση αν κλάδοι της αορτής εμπεριέχονται στον διαχωρισμό
- Εκτίμηση της ανεπάρκειας της αορτικής βαλβίδας
- Εντοπισμός σημείων εξαγγείωσης (περιαορτικό αιμάτωμα, αιμοπερικάρδιο, πλευριτική συλλογή, περικαρδιακή συλλογή)

Effie ROUSKA, MD

Aortic dissection: differentiation between true and false lumen

	True lumen	False lumen
Size	true<false	most often false>true
Pulsation	systolic expansion	systolic compression
Flow direction	systolic antegrade flow	systolic antegrade flow reduced or absent or retrograde flow
Localization within the aortic arch	inner contour	outer contour
Signs of slow flow	rare	frequent; depending on degree of communication
Thrombus	rare	frequent; depending on degree of communication

Effie ROUSKA, MD

Διαγνωστική προσέγγιση

- απόφαση όμως για το ποια μέθοδος θα επιλεγεί εξαρτάται από την διαθεσιμότητα αλλά και την εμπειρία του προσωπικού που θα υποδεχθεί τον ασθενή, καθώς και την σταθερότητα ή μη της γενικής του κατάστασης

Effie ROUSKA, MD

Υποψία ΟΑΣ σε ασταθή ασθενή

- Οδηγείται στο χειρουργείο
- Διασωλήνωση - Γραμμές monitoring
- ΤΟΕ μελέτη
- Αν ΤΟΕ αρνητικό CT σε δεύτερο στάδιο

Effie ROUSKA, MD

Υποψία ΟΑΣ σε σταθερό ασθενή

- Ιστορικό
- Φυσική εξέταση
- Αξονική τομογραφία
- ΤΟΕ

Effie ROUSKA, MD

Αρχική Θεραπευτική προσέγγιση

- Ο βασικός στόχος είναι να περιορίσει την εξέλιξή του ώστε να μην προχωρήσει:
  - σε ρήξη του αορτικού τοιχώματος
  - σε μυοκαρδιακή ισχαιμία
  - ανεπάρκεια της αορτικής βαλβίδας
- Αυτό επιτυγχάνεται με τη χειρουργική σύγκλειση του entry point
- Μέχρι τότε……

Effie ROUSKA, MD

- εισέρχεται στην ΜΕΘ
- monitoring αιμοδυναμικών παραμέτρων
- εισάγονται
  - ευρείες φλεβικές γραμμές,
  - ουροκαθετήρας
  - αρτηριακή γραμμή
- φαρμακευτικής αγωγής
  - αντιμετώπιση του πόνου (μορφίνη)
  - έλεγχος της αρτηριακής πίεσης

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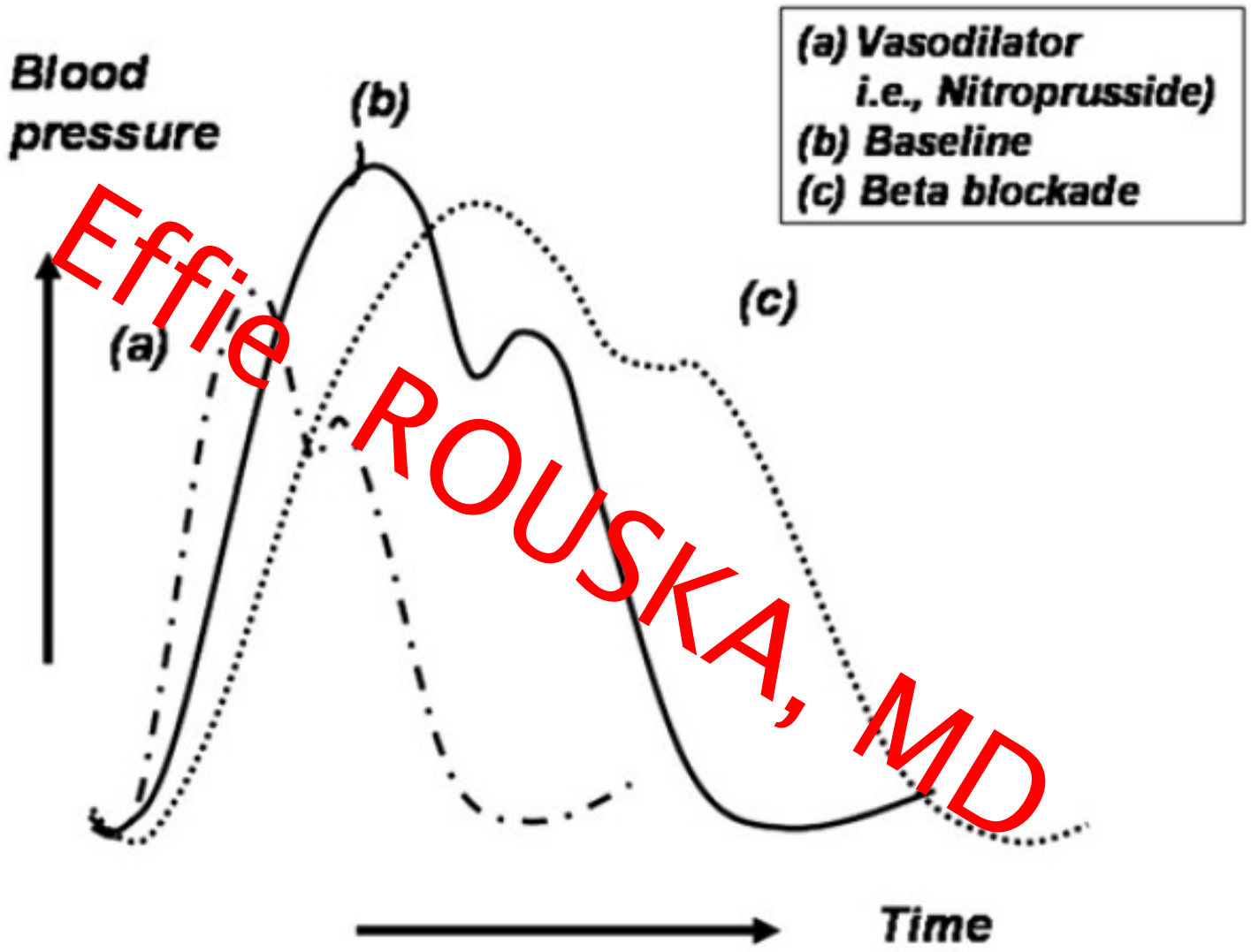

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Ευχαριστώ

Effie ROUSKA, MD

Το μόνο πράγμα που μας διδάσκει ο θάνατος
είναι πως είναι επείγον να αγαπήσουμε.
-Eric-Emmanuel Schmit