UNIUNEA EUROPEANĂ | GUVERNUL ROMÂNIEI | Fondul Social European POSDRU 2007-2013 | Instrumente Structurale 2007-2013 | GUVERNUL ROMÂNIEI MINISTERUL MUNCII, FAMILIEI, PROTECȚIEI SOCIALE ȘI PERSOANELOR VÂRSTNICE OIRPOSDRU REGIUNEA CENTRU | UNIVERSITATEA DE MEDICINĂ ȘI FARMACIE "CAROL DAVILA" BUCUREȘTI

AD-COR Program inovativ de formare in domeniul cardiologiei pediatrice
POSDRU/179/3.2/S/152012

Octombrie 2015

MODUL TEORETIC

COR TRIATRIATUM

Continut documentat/ validat/ prezentat de:

⇨ Expert formare medici: NICOLESCU Alin

⇨ Expert formare medici: VEDUTA Alina

*A4 – Planificarea, organizarea si desfasurarea activitatilor de formare a **medicilor** in domeniul cardiologiei pediatrice*

Malformatii congenitale obstructive in inflow-ul VS:

1. Stenoza mitrala congenitala
2. Cor triatriatum stang
3. Stenoze venoase pulmonare
4. Inel supravalvular mitral

Caracteristici:

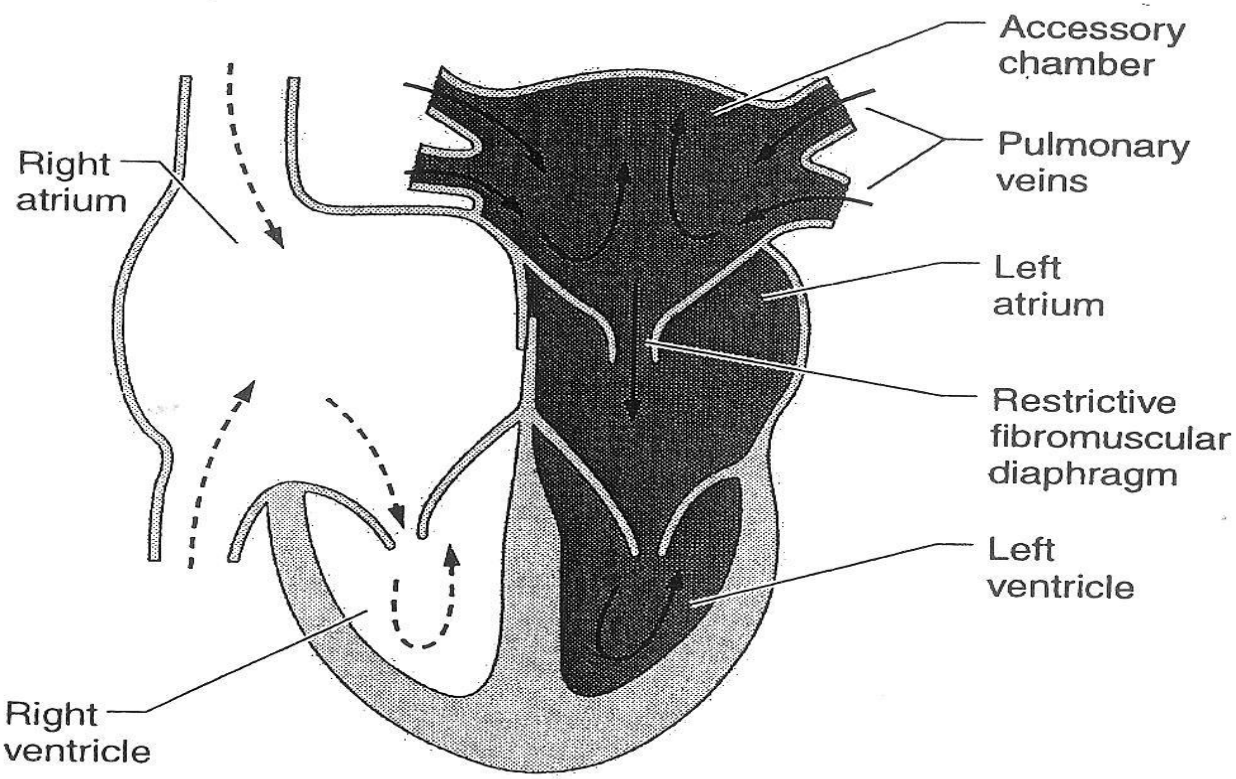
- existenta unei membrane fibromusculare ce imparte AS in 2 portiuni

1.portiunea posterosuperioara proximala ce primeste venele pulmonare

2.portiunea antero-inferioara ce contine auriculul stang si valva pulmonara

- Cor triatriatum stang,
- Cor triatriatum drept
- Cor stang polyatriatum

Drawing of Cor Triatriatum



Istoric

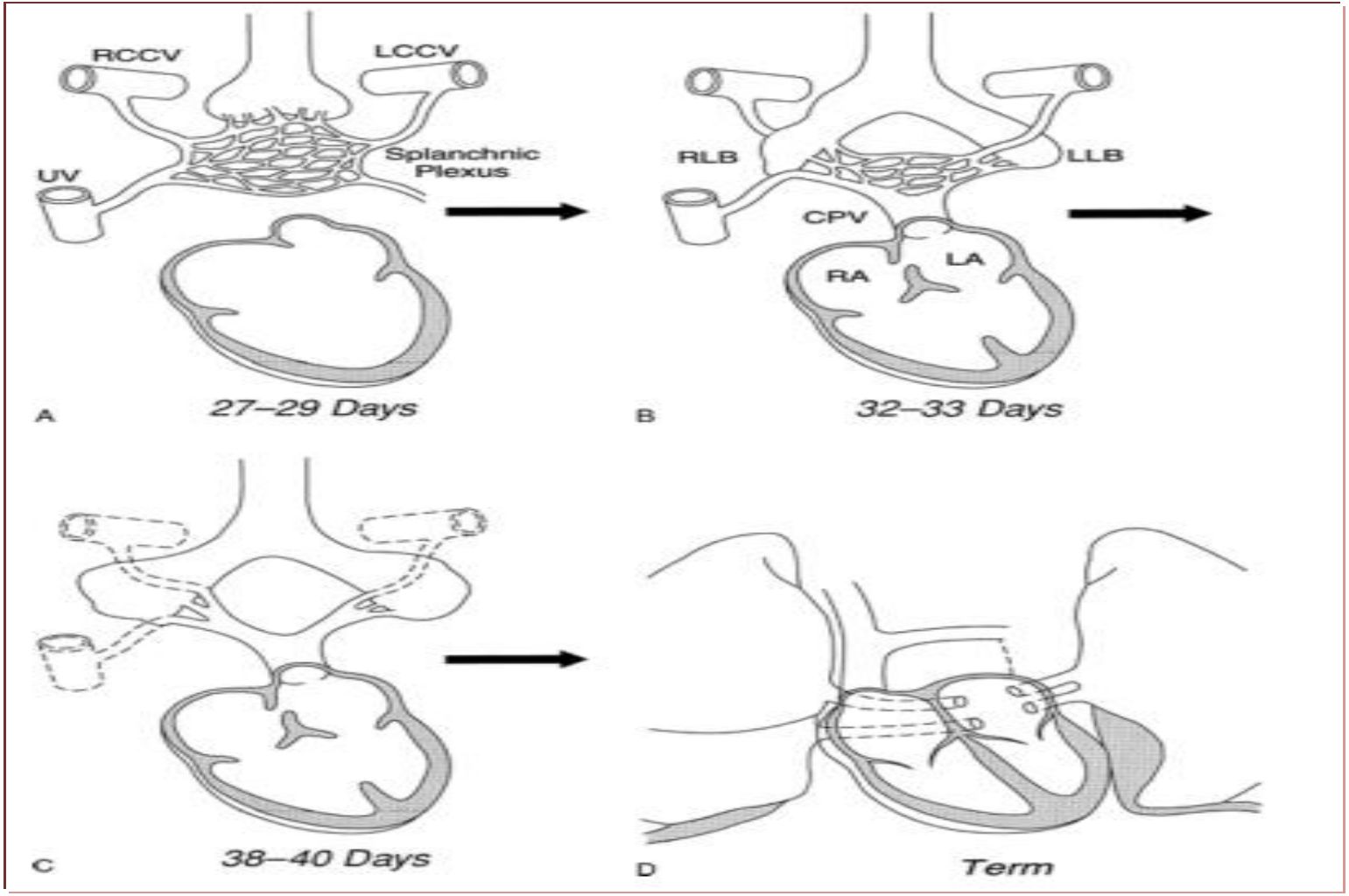
- Church : 1 Descrierea în 1868
- Borst: termenul de cor triatriatum în 1905
- Miller et al: diagnosticul angiografic în 1964
- Ostman-Smith: Descriere ecografica in 1984
- Vineberg & Gialloreto: 1st corecţia chirurgicală în 1956

COR TRIATRIATUM

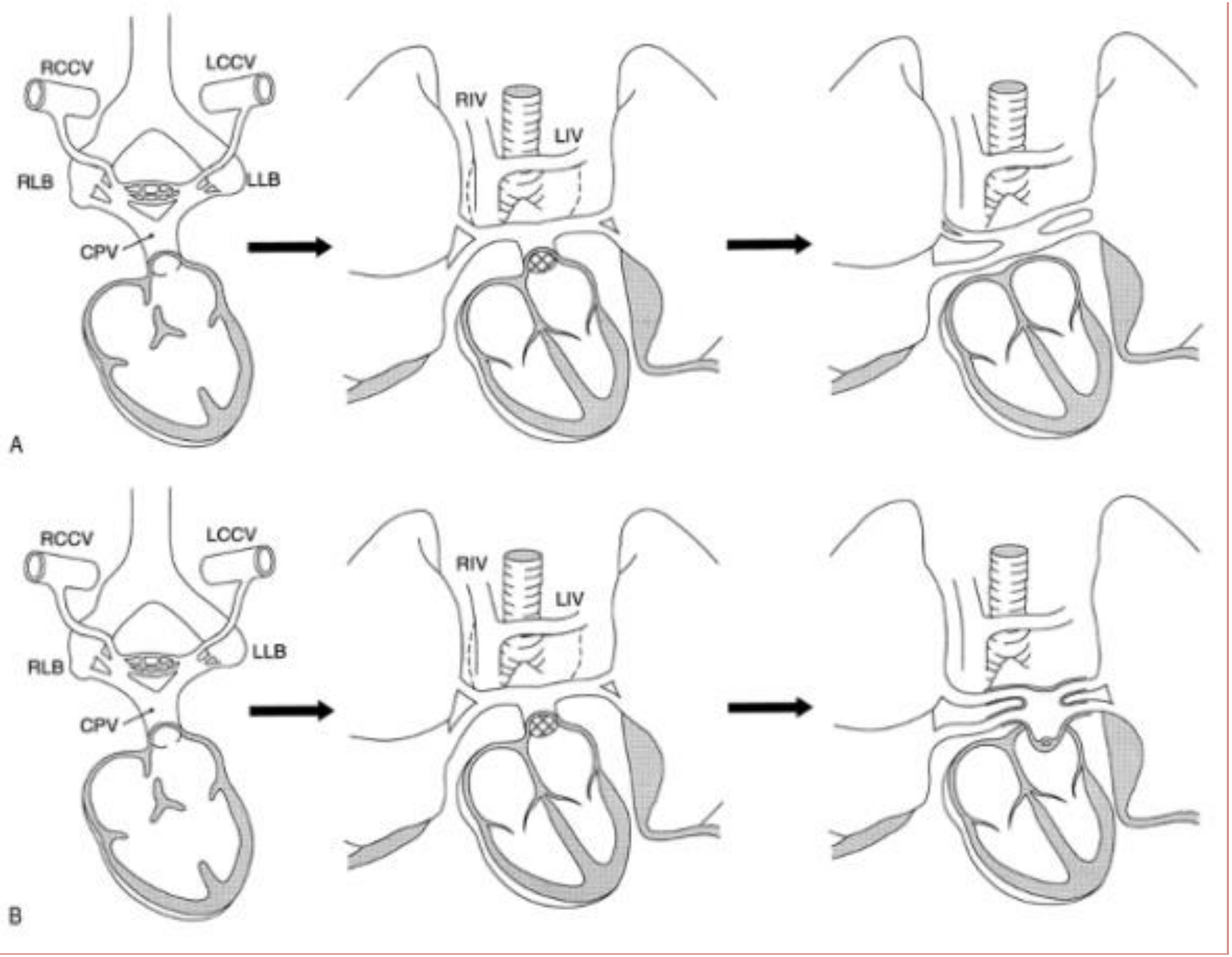
- Prevalenta

0,1-0,4 din MCC

Izolata 30-50 %din cazuri



venous systems. A small evagination can be seen in the posterior wall of the left atrium to the left of the developing septum secundum. **B:** By the end of the first month of gestation, the common pulmonary vein establishes a connection between the pulmonary venous plexus and the sinoatrial portion of the heart. At this time, the connections between the pulmonary venous plexus and the splanchnic venous plexus are still patent. **C:** Next, the connections between the pulmonary venous plexus and the splanchnic venous plexus involute. **D:** The common pulmonary vein (CPV) incorporates into the left atrium so that the individual pulmonary veins connect separately and directly to the left atrium. LA, left atrium; LCCV, left common cardinal vein; LLB, left lung bud; RA, right atrium; RCCV, right common cardinal vein; RLB, right lung bud; UV, umbilical vein. (Adapted from Lucas RV Jr, Anderson RC, Amplatz K, et al. Congenital causes of pulmonary venous obstruction. *Pediatr Clin North Am* 1963;10:781–836, with permission.)


RCCV
LCCV
RLB
LLB
CPV
RIV
LIV
A
RCCV
LCCV
RLB
LLB
CPV
RIV
LIV
B

- TIPURI:

1.Tip A

Fara DSA

A1-DSA in camera proximala

A2-DSA in camera distala

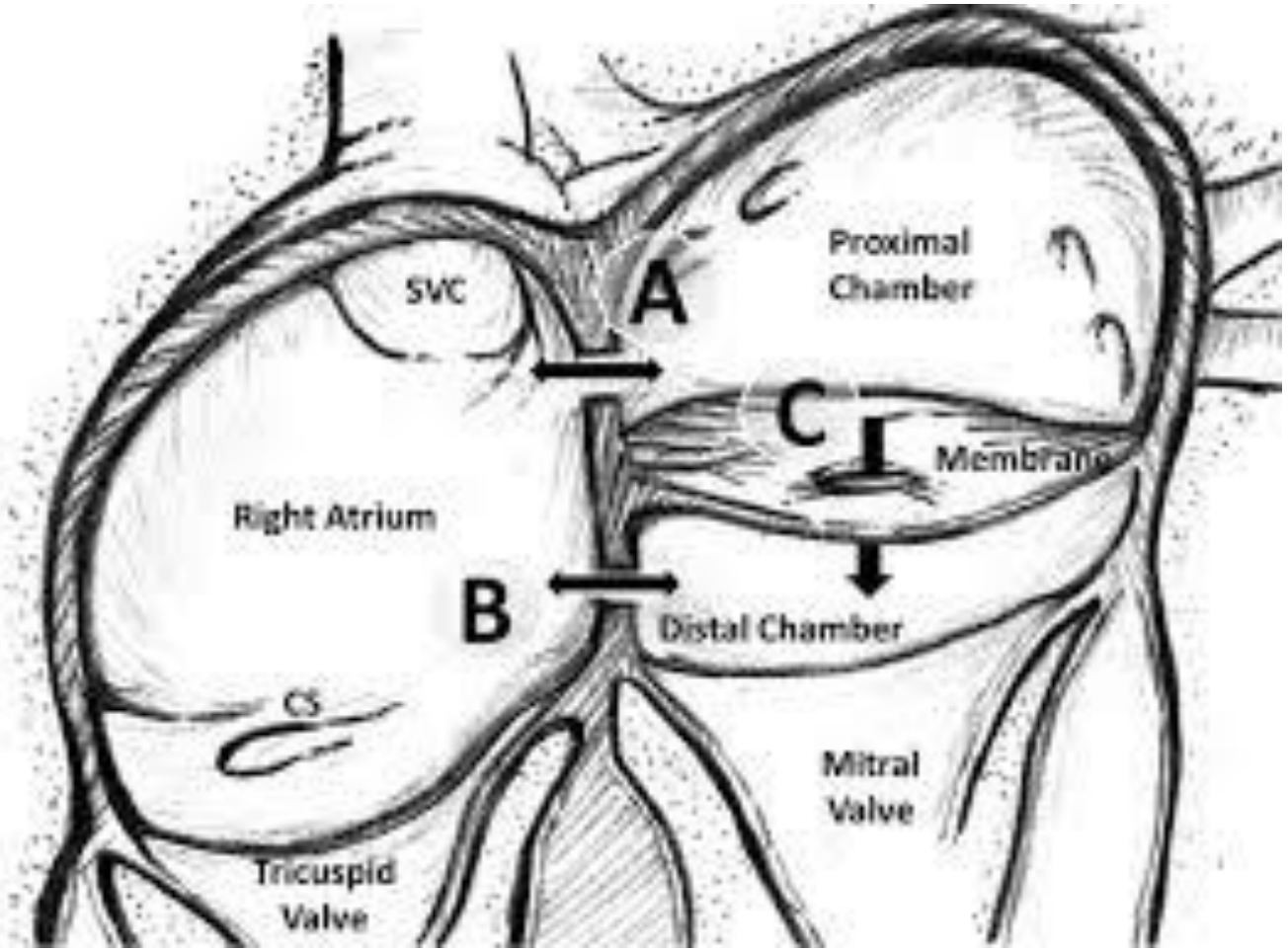
2.Tip B

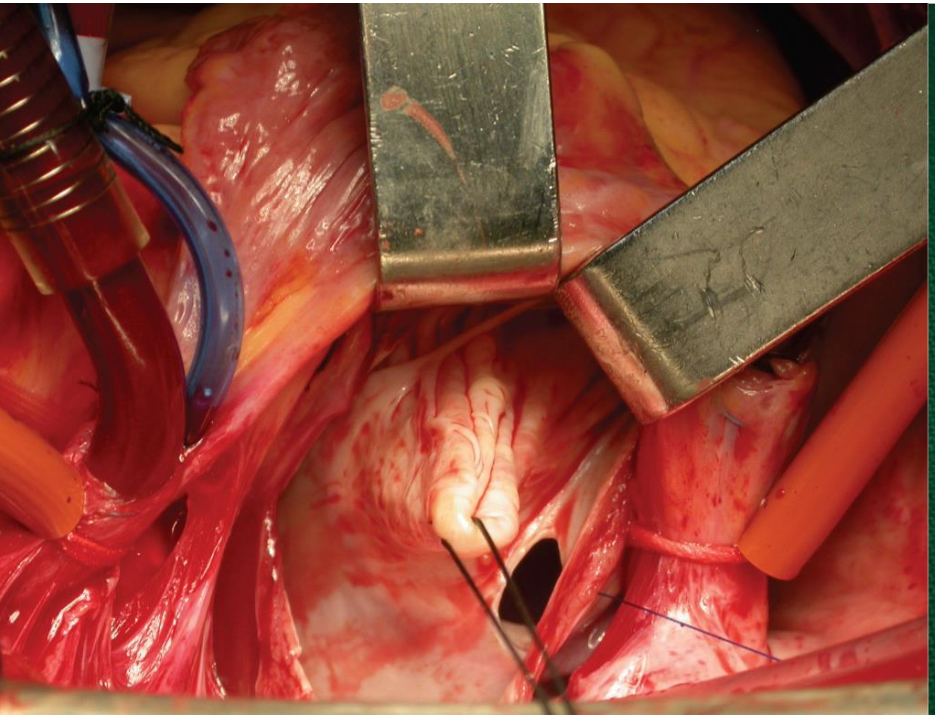
Vene pulmonare drenaj in sinusul coronar

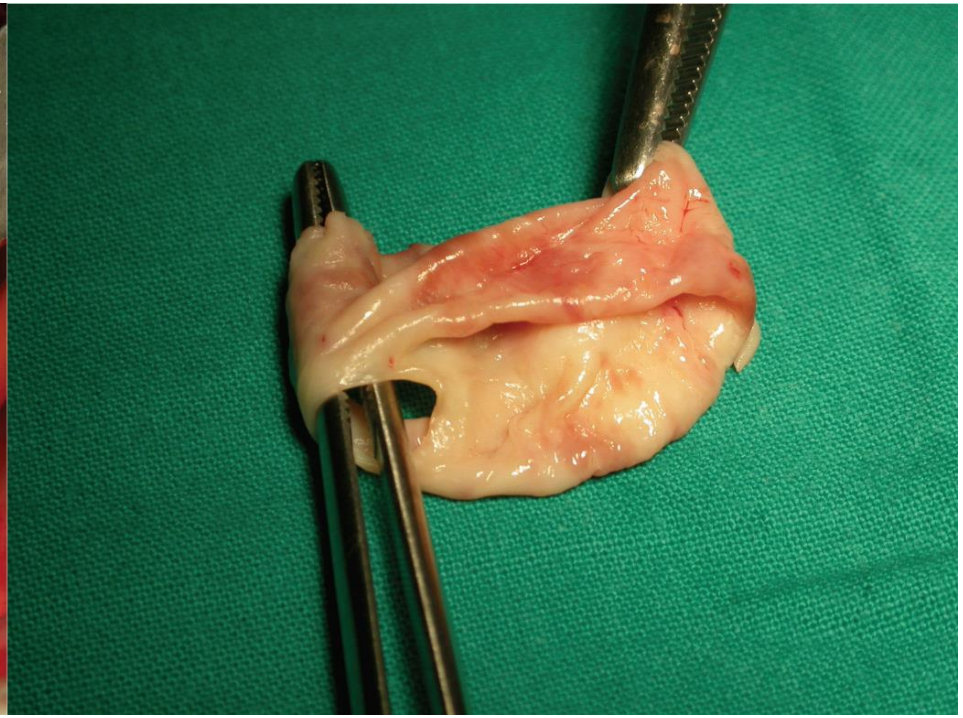
3.Tip C

Venele pulmonare nu dreneaza in camera proximala

4.Diafragm imperforat


SVC
A
Proximal
Chamber
C
Membrane
Right Atrium
B
Distal Chamber
CS
Mitral
Valve
Tricuspid
Valve





MORFOLOGIE

- Camera proximala mai ingrosata iar camera distala este mai subtire
- una sau mai multe deschideri in diafragm
- AD,VD marite din cauza şuntului stg-dpt interatrial,
- VS de obicei mai mic
- Foramen ovale, de obicei patent
- mai frecvent decat alte tipuri de MCC, exista VCS stânga

- Anomaliile asociate

Intoarcere venoasa pulmonara partiala/total aberanta

Sinus coronar unroof

DSV,CoAo,CAVC,TOF

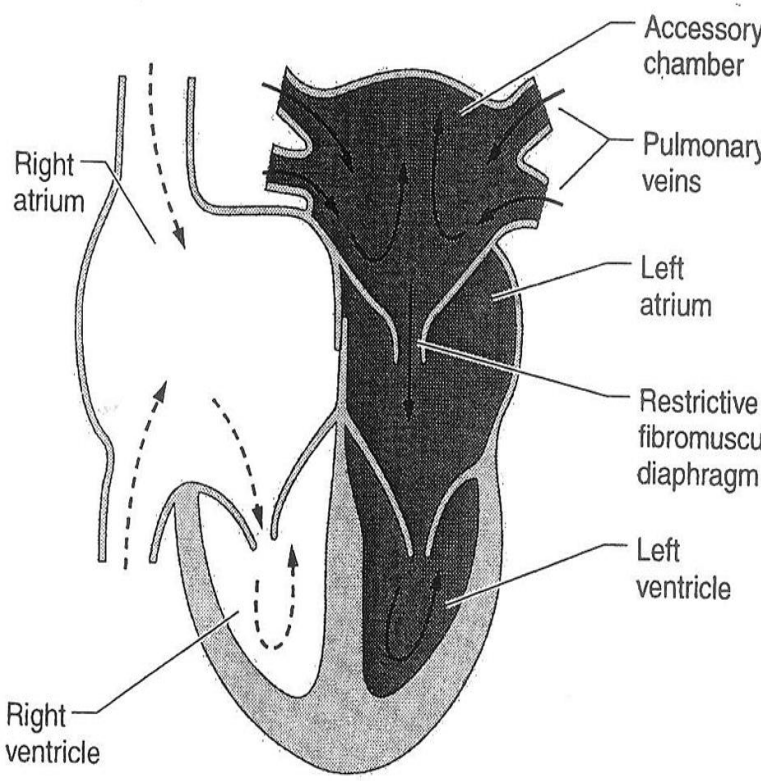
Fiziopatologie

- Camera atriala proximala comunică cu atriul stang adevărat prin diafragma restrictiva fibromusculară.
- Întoarcerea venoasă pulmonară este restrictionata, rezultand congestie venoasă pulmonară şi hipertensiune arterială pulmonara, hipertrofie ventriculară dreaptă şi insuficienţă cardiacă congestivă.
- Severitatea simptomelor clinice depinde de :

➢ marimea si numarul orificiilor din diafragmul fibromuscular

➢ Prezenta DSA

➢ Alte malformatii asociate



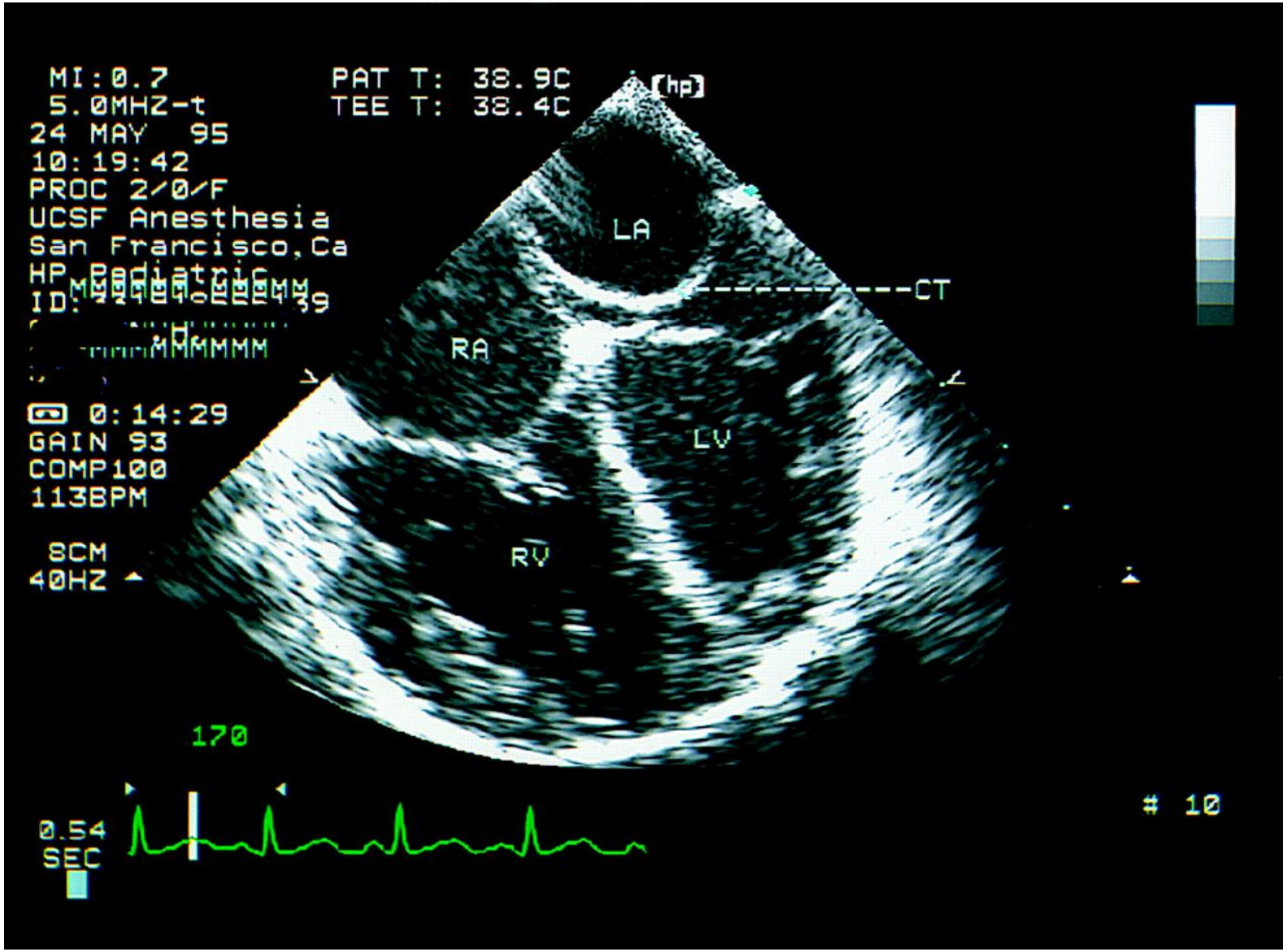
Clinic

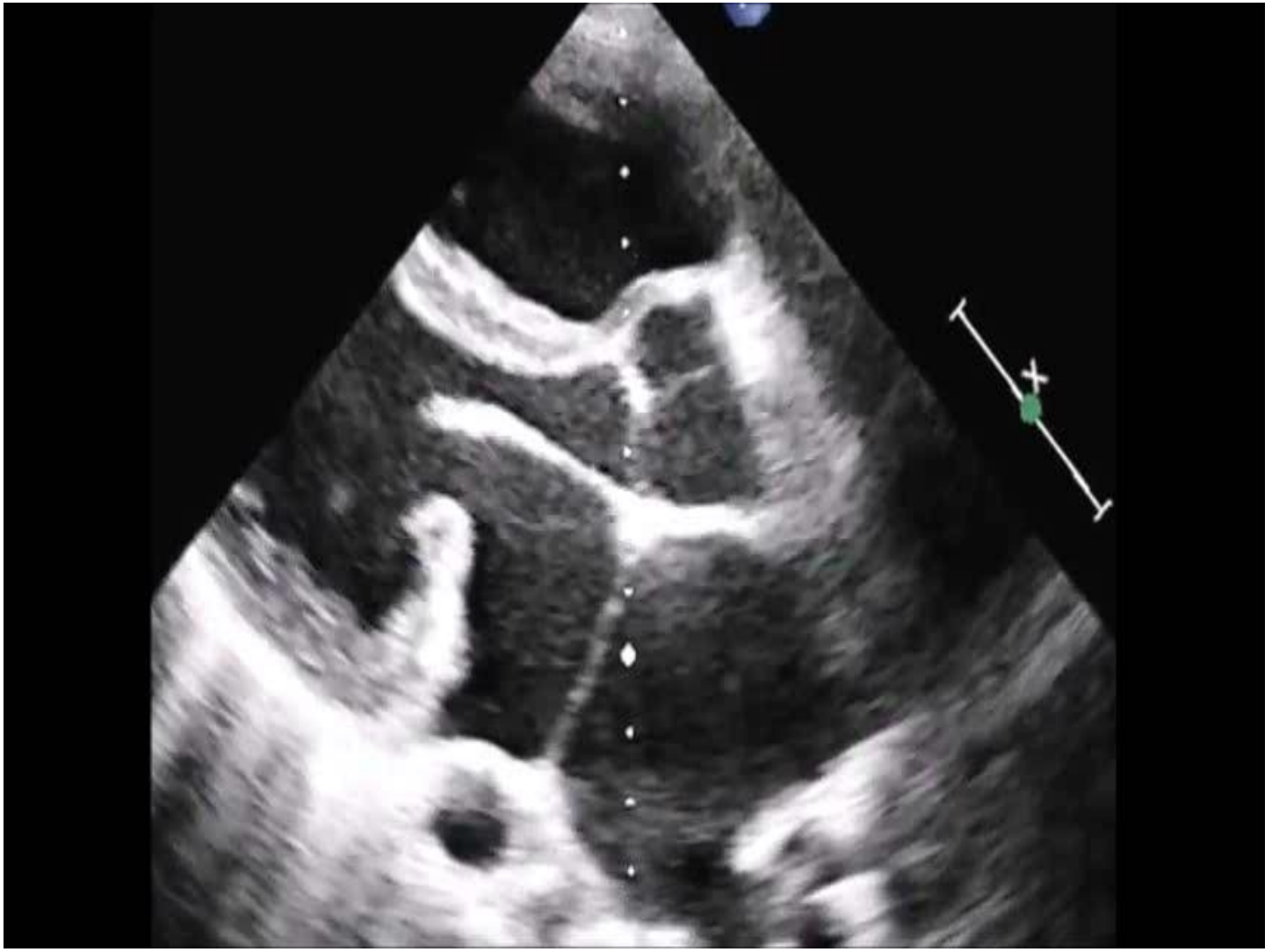
1. Pacienţii cu orificiu mic in diafragm
- paloare,
- tachypnea,
- puls periferic slab,
- deficit de creştere
- Atunci când există asociat sunt stg-dpt,

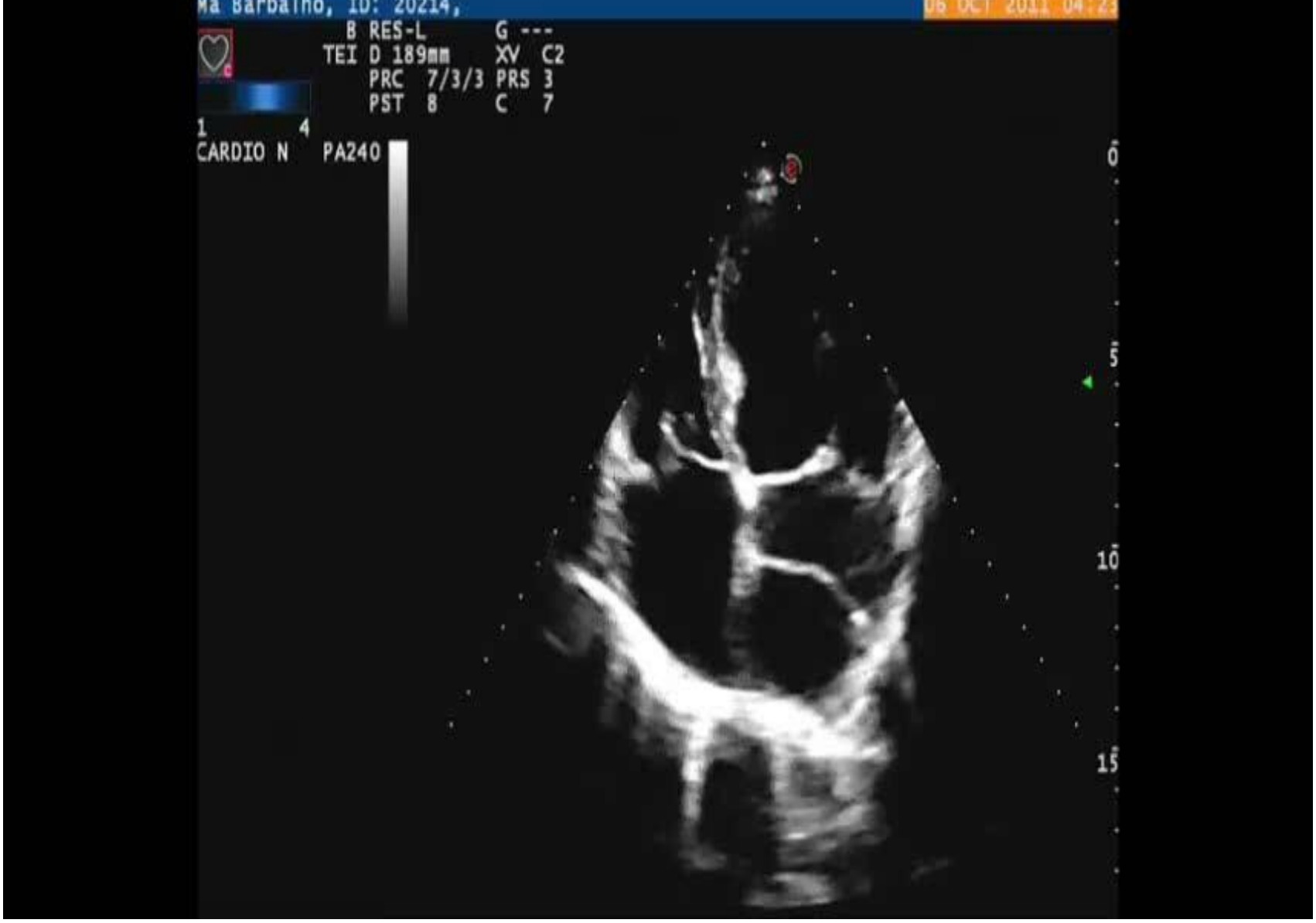
manifestari supraincarcare pulmonara pot fi prezente

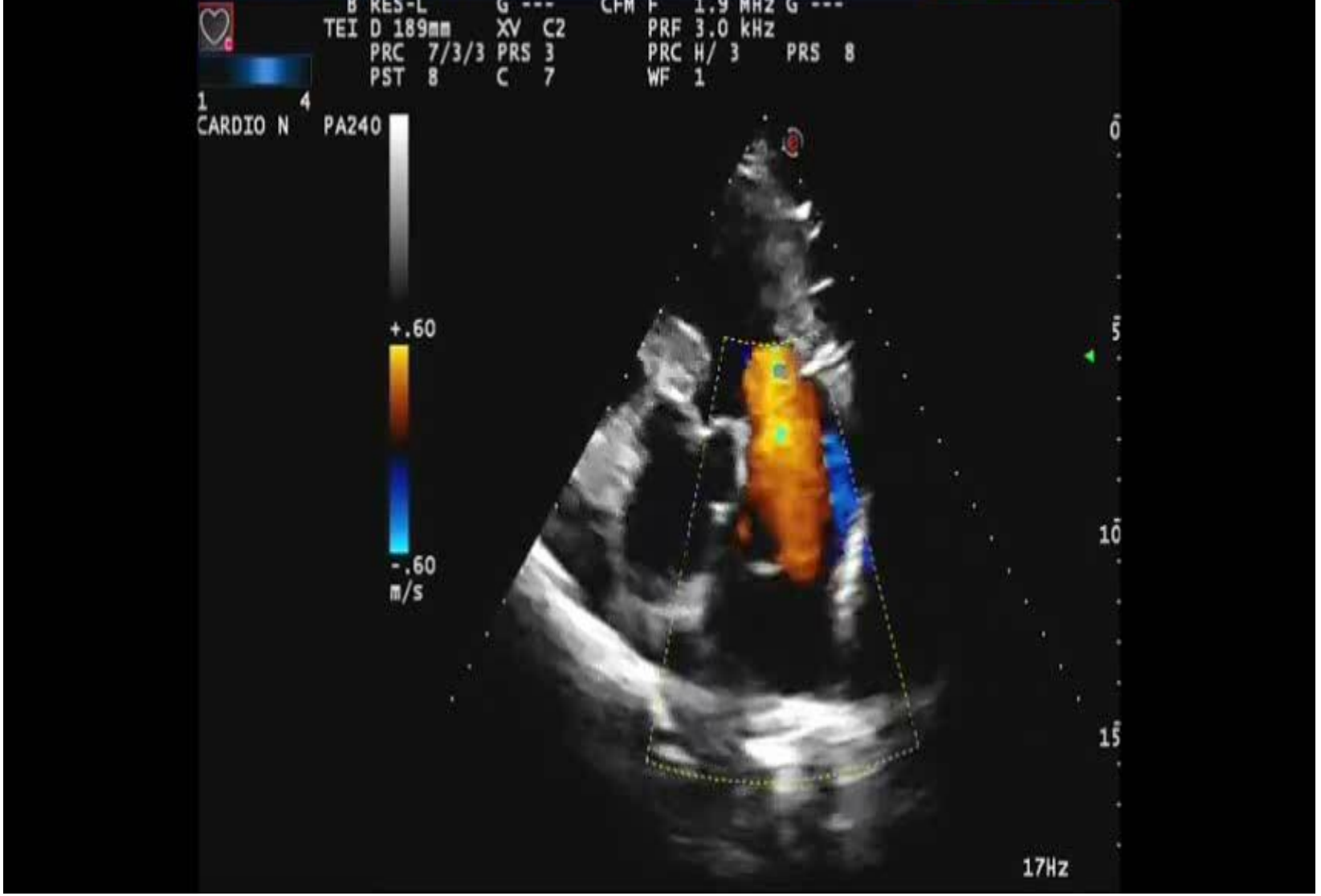
Istorie naturală

1. Anomalie congenitale cardiaca rara
2. Istorie naturala depinde de marimea efectiva a comunicarii prin diafragm
3. De cele mai multe ori , orificiul este sever restrictiv şi aproximativ 75% mor în copilărie fără tratament
4. Atunci când comunică cu atriul drept prin DSA, prognosticul este mai bun (depinde de mărimea DSA).

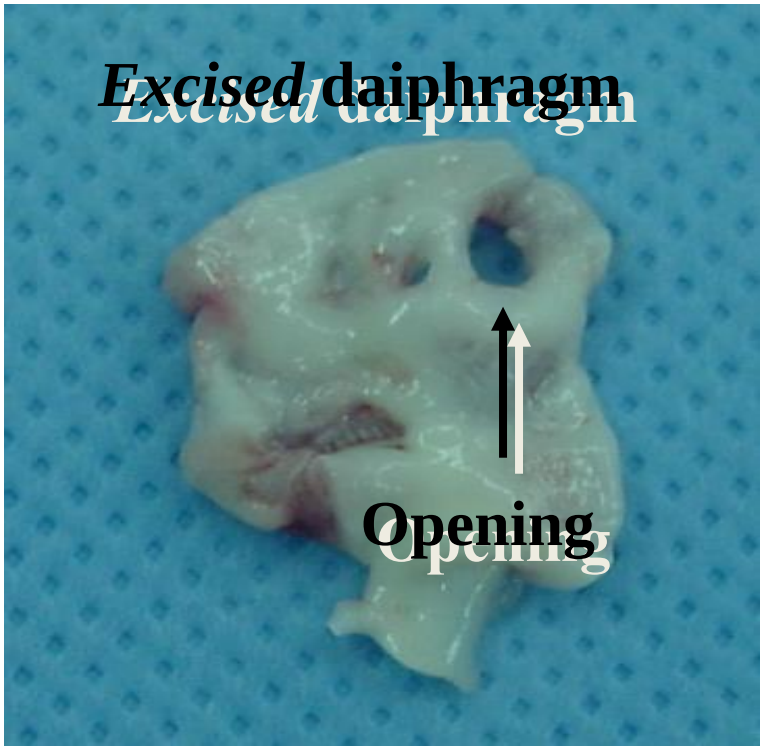

MI:0.7
5.0MHZ-t
24 MAY 95
10:19:42
PROC 2/0/F
UCSF Anesthesia
San Francisco,Ca
HP Pediatric
PAT T: 38.9C
TEE T: 38.4C
(hp)
LA
CT
RA
LV
RV
0:14:29
GAIN 93
COMP100
113BPM
8CM
40HZ
170
0.54
SEC
10




Ma Barbatho, ID: 20214,
06 OCT 2011 04:23
B RES-L G ---
TEI D 189mm XV C2
PRC 7/3/3 PRS 3
PST 8 C 7
1 4
CARDIO N PA240
0
5
10
15


B RES-L G --- CFM F 1.9 MHz G ---
TEI D 189mm XV C2 PRF 3.0 kHz
PRC 7/3/3 PRS 3 PRC H/ 3 PRS 8
PST 8 C 7 WF 1
1 4
CARDIO N PA240
+.60
-.60
m/s
0
5
10
15
17Hz

Operative View of Cor Triatriatum



Rezultate postchirugical

- Supravietuirea se apropie de cea a populaţiei generale şi cu bun rezultat funcţional
- Moarte timpurie -neobişnuit, dar apare în bolnav grav cu managementul miocardic inadecvat

- Complicaţii

1. Restenoza datorită rezectiei inadecvate
2. stenoză vena pulmonară

COR TRIATRIARUM DEXTER

- Extrem de rar
- In marea majoritate a cazurilor asimptomatic
- Poate determina tahicardii supraventriculare recurente
- Poate determinacazuri severe IVD

Concluzii

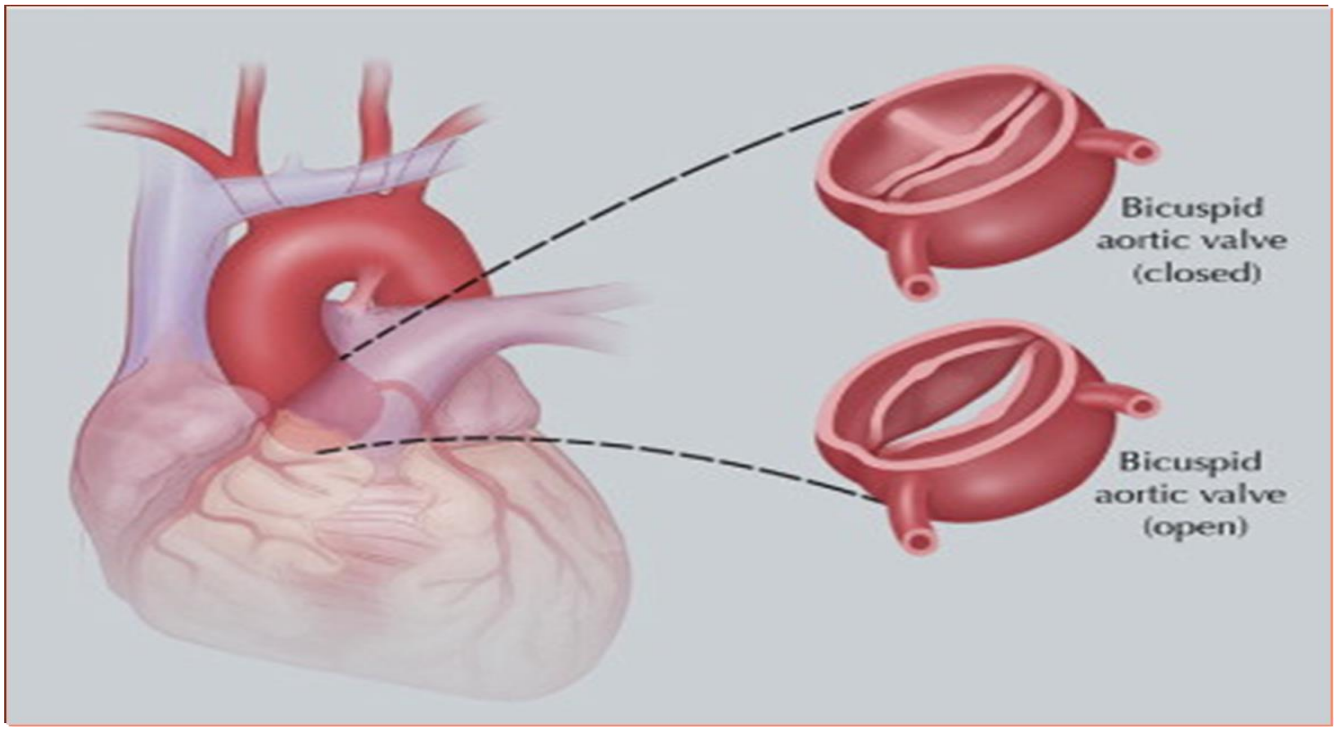
- Face parte din leziunile obstructive in fluxul anterograd VS
- Frecvent asociat cu

➢ persistenta de VCS stg

➢ Anomalii de intoarcere venoasa pulmonara

- Aprecierea evolutiei depinde de aprecierea fluxului anterograd catre VM

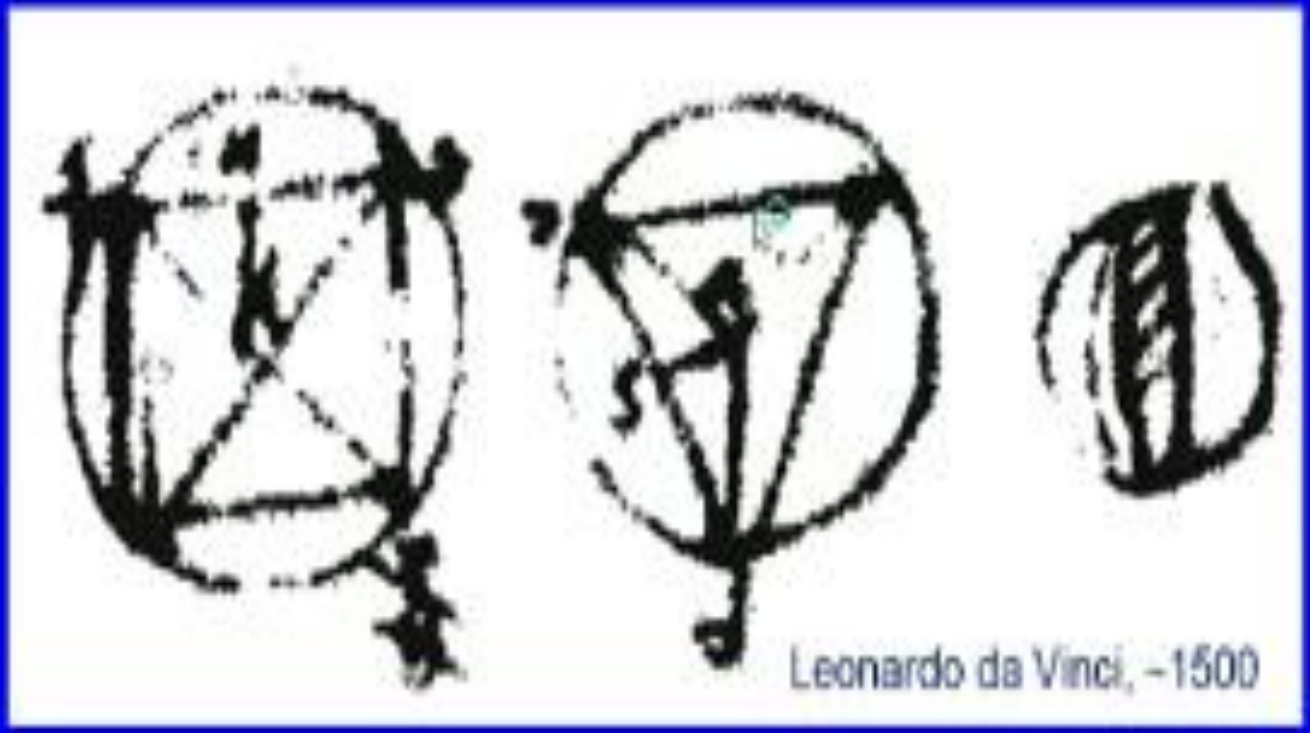
BICUSPIDE AORTICA



Dr Alin Nicolescu
Departament Cardiologie pediatrica
SCUC “M.S.Curie”

Bicuspid Aortic Valve Disease

Prevalence, Genetics and Natural History



Bicuspid Aortic Valve Disease
PREVALENCE (clinical studies)

- Echo screening in neonates (n=1075)
  - BAV 4.6/1000 live births
  - 7.1/1000 boys; 1.9/1000 girls

    Tutar E et al. Am Heart J 2005

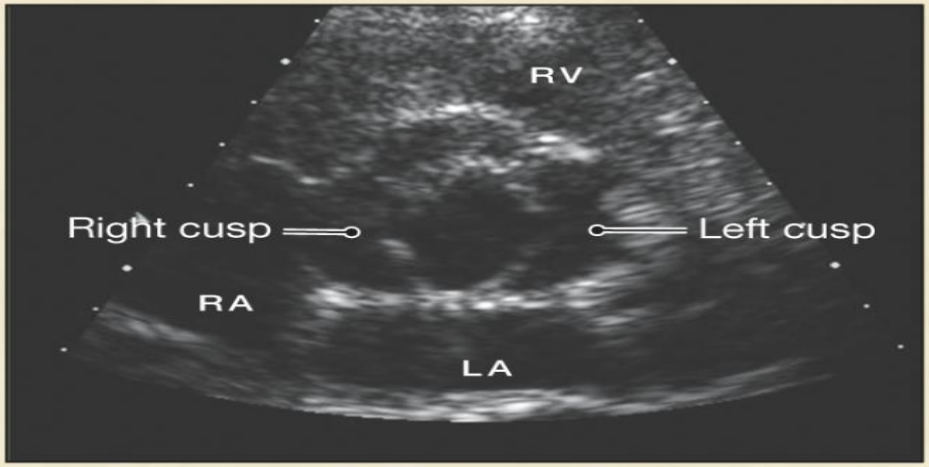
- Echo screening in 20,946 military recruits (Italy)
  - BAV in 0.8% (possibly underestimate)

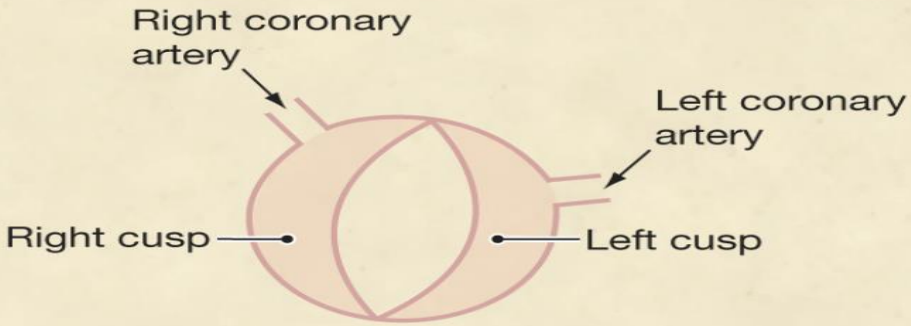
    Nistri S et al. Am J Cardiol 2005

Overall estimate 1-2% of population
Male: female 2:1 ratio
No geographic differences identified

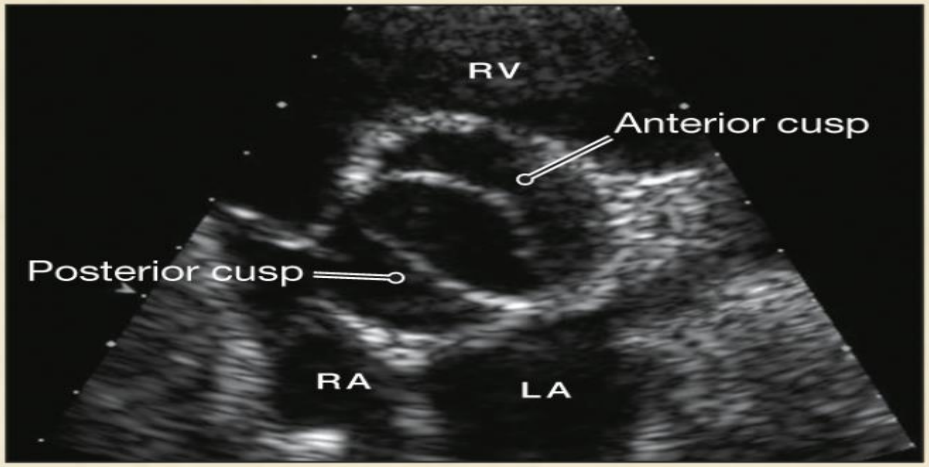
C Phenotypic variations in bicuspid aortic valve anatomy

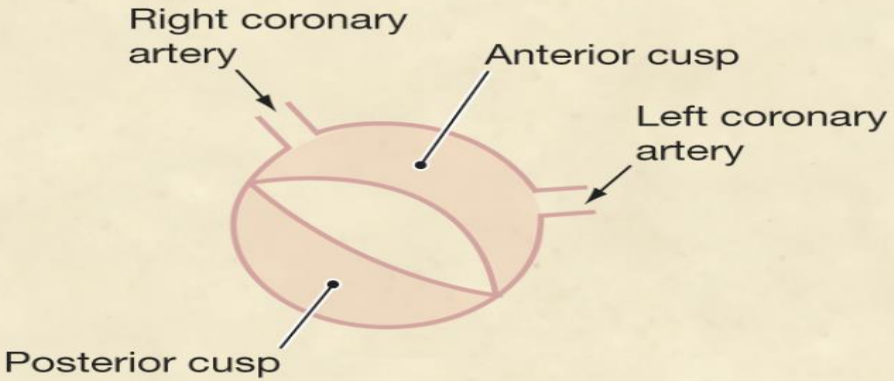
Right-left orientation





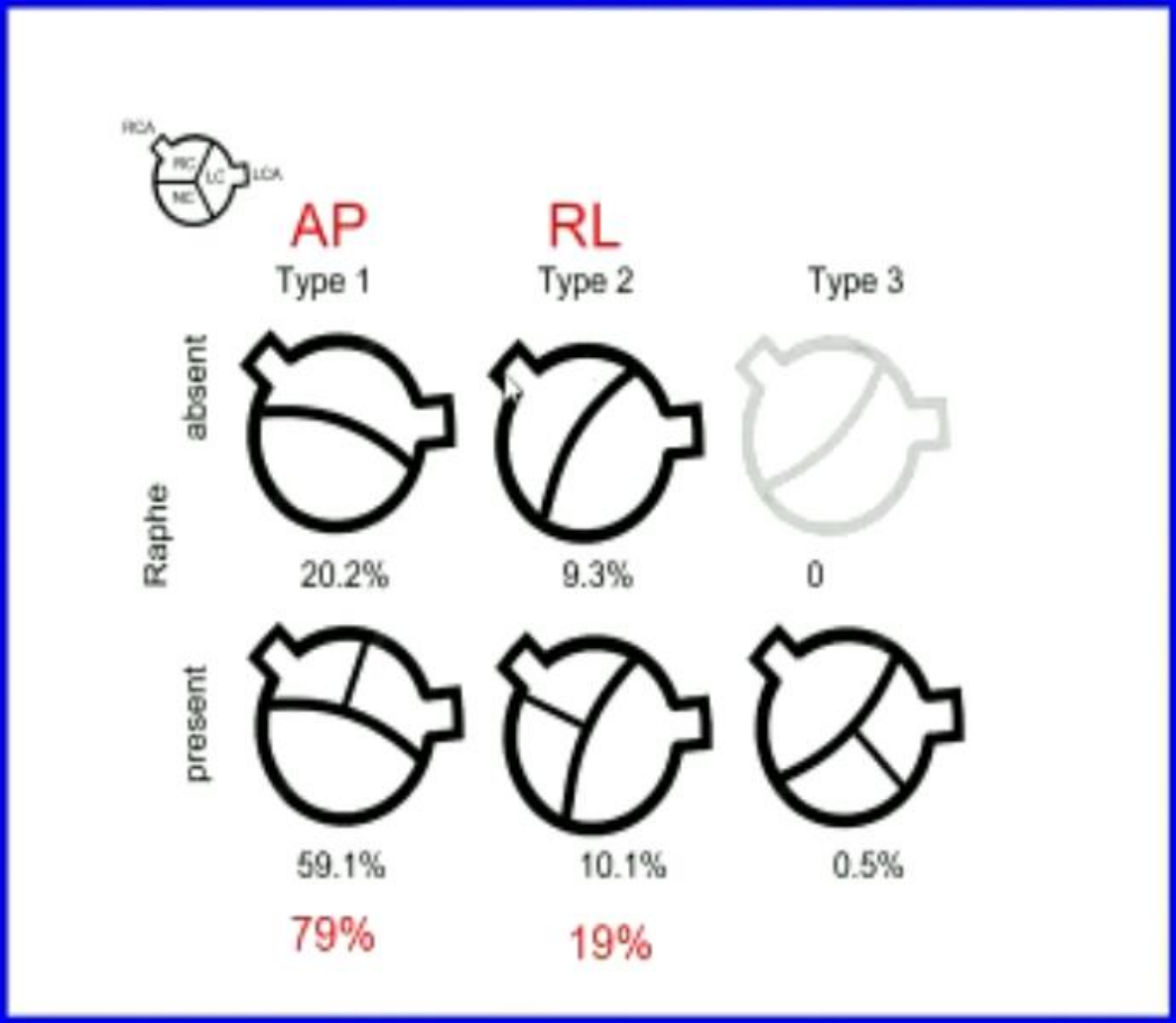
Anterior-posterior orientation





Phenotypic Classification: BAV and Aortic Root

Seattle 2002-05, 192 subjects



Schaefer, et al, Heart 2008

Bicuspidie aortica

- Genetica

Bicuspid Aortic Valve Disease

GENETICS

- Sporadic
  - Most cases have no affected relatives
  - Affects about 1% of the population
- Autosomal Dominant
  - BAV in about 9% of 1st degree relatives
  - Incomplete penetrance

- Se accepta ca 10 % sansa ca o ruda de grd I sa aiba Bic Ao la pacientii cu Bic Ao

- Mutatii gena NOTCH1 au fost evidentiate la 2 familii cu B Ao.Au mai fost descrise loci din cromosomii 18 q, 5 q,13 q

- Recomandarile ACC/AHA suntsa fie efectuat screening pentru toate rudele de gradul I

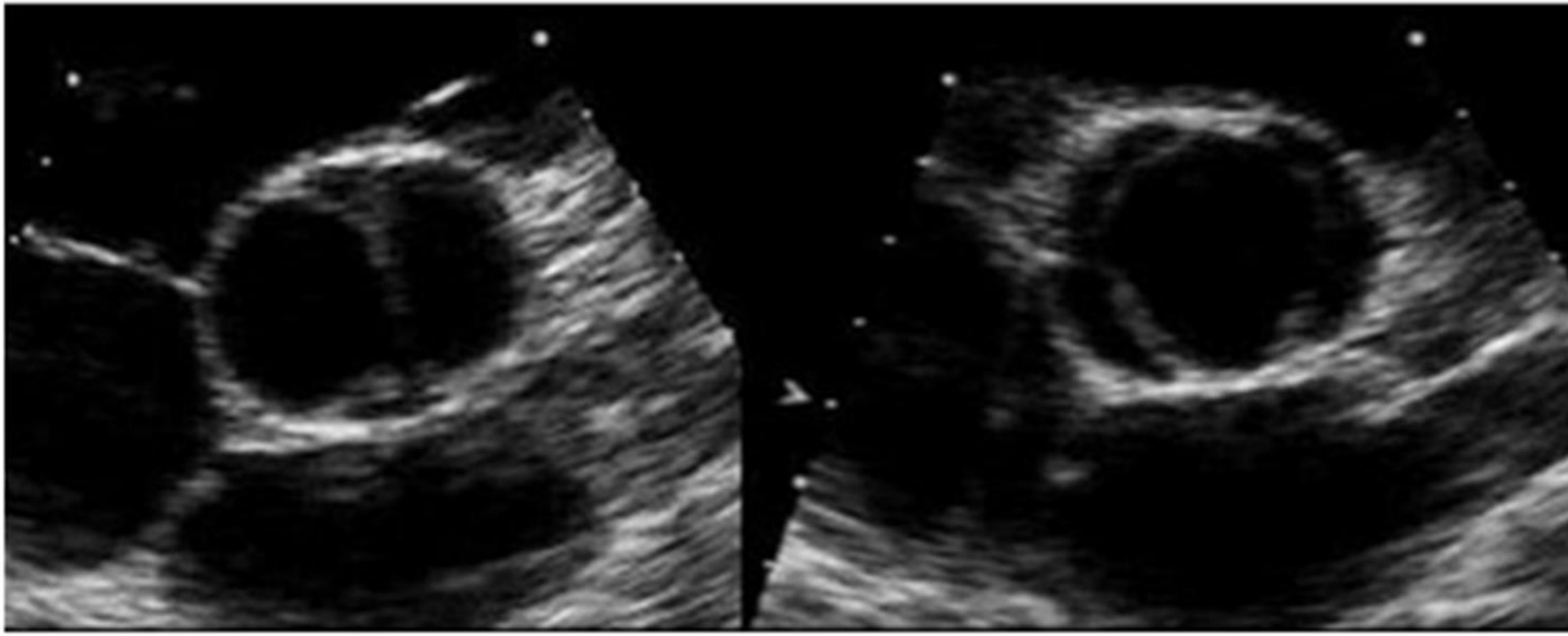
Bicuspidie Aortica

- Diagnostic

- Auscultatie
  - ➢ Clic de ejectie (valve in dom)
  - ➢ Suflu de ejectie (St Ao)
  - ➢ Suflu diastolic (I Ao)
- Ecg
- Normal
- Semne de HVS

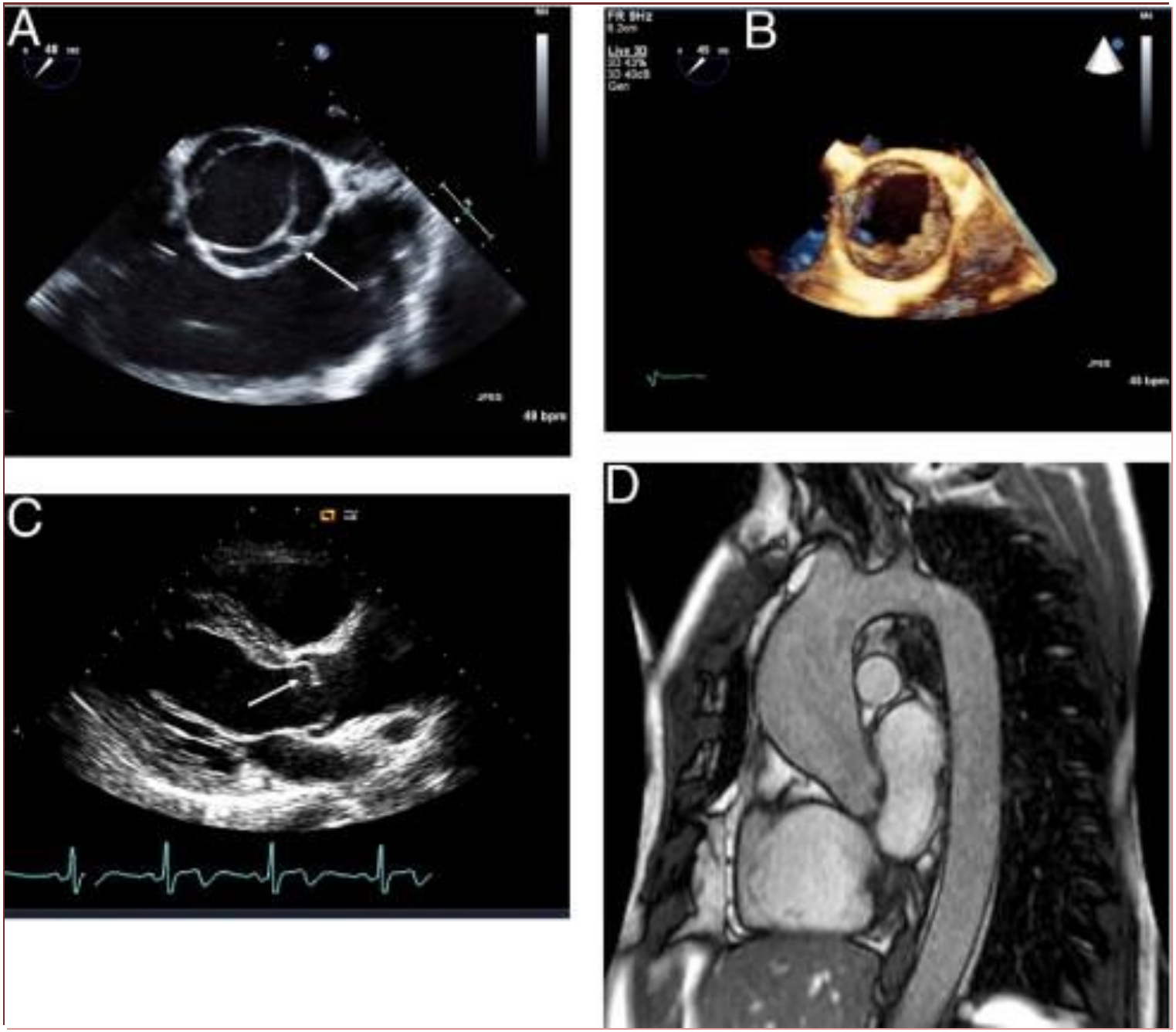
Ecografia

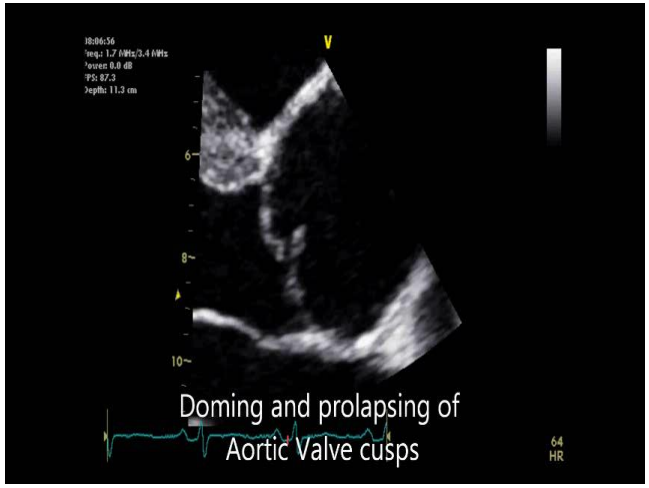
Bicuspid aortic valve-anterior-posterior commissure

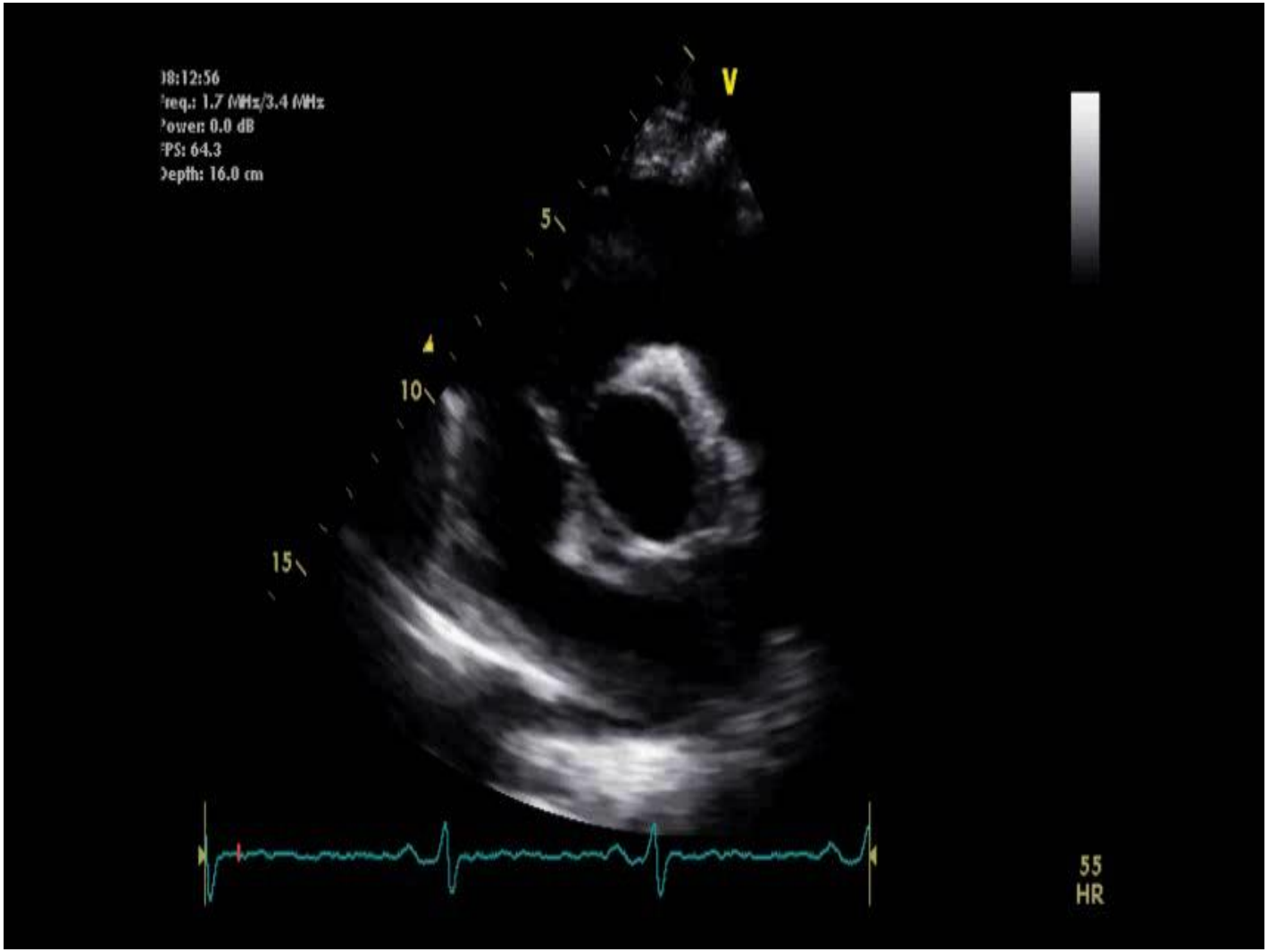


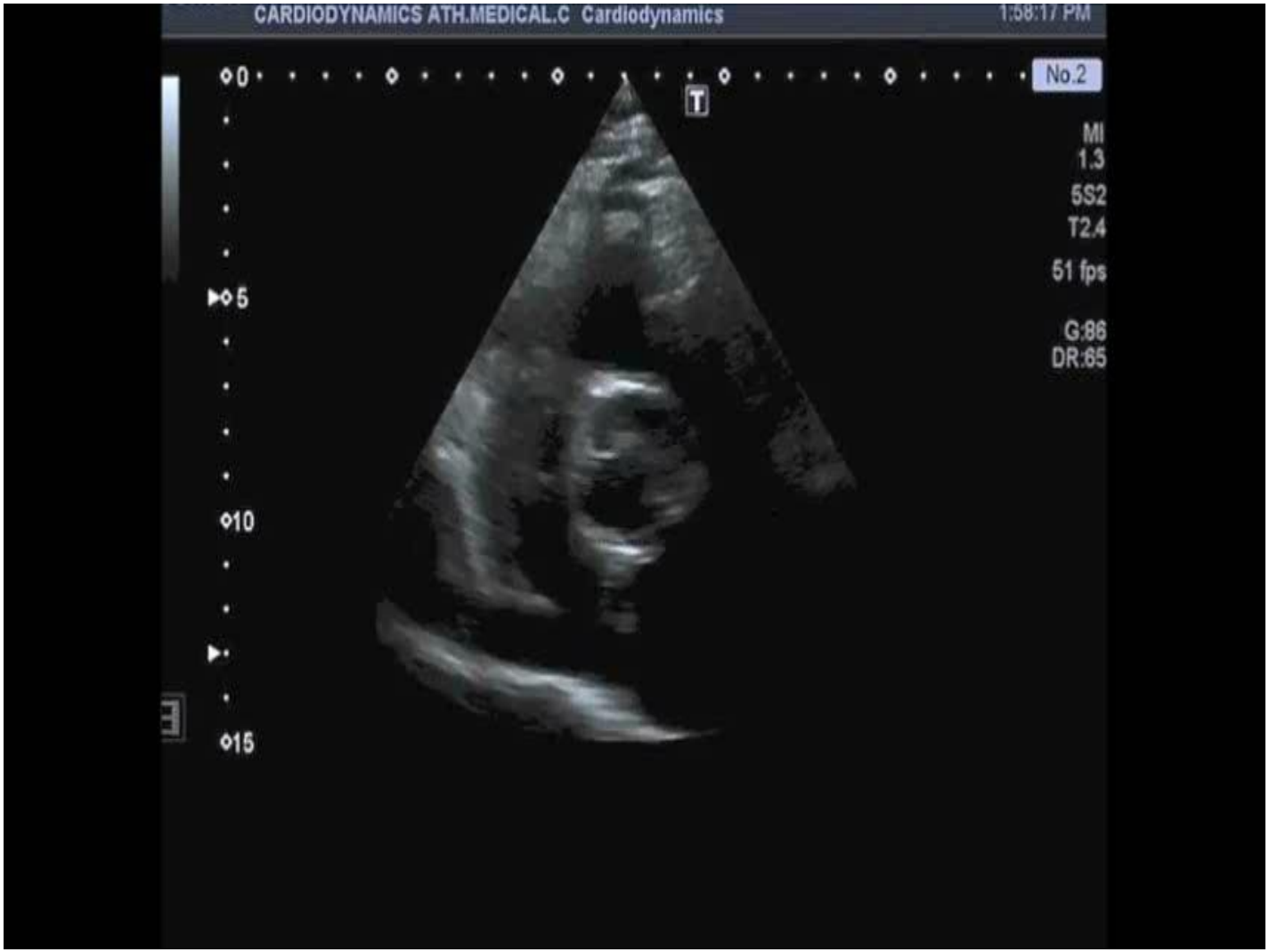
Diastole

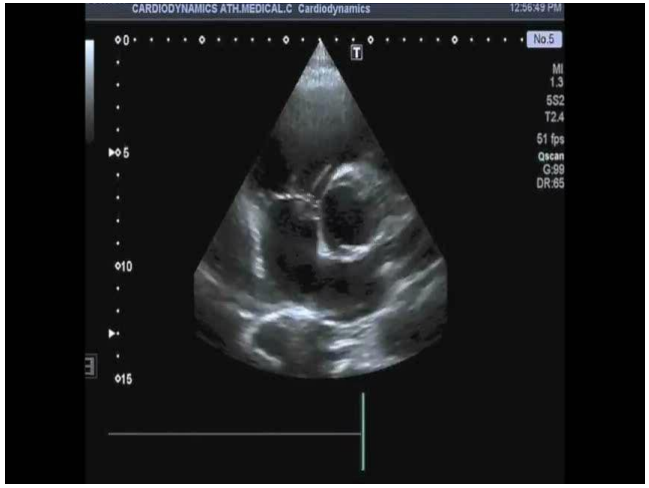
Systole

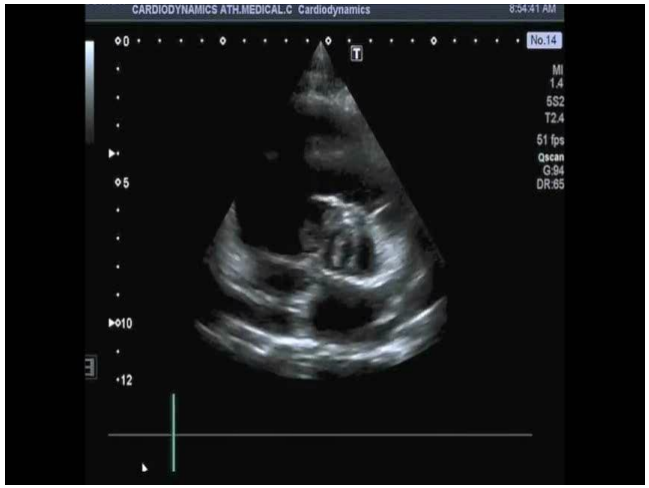

A
B
C
D

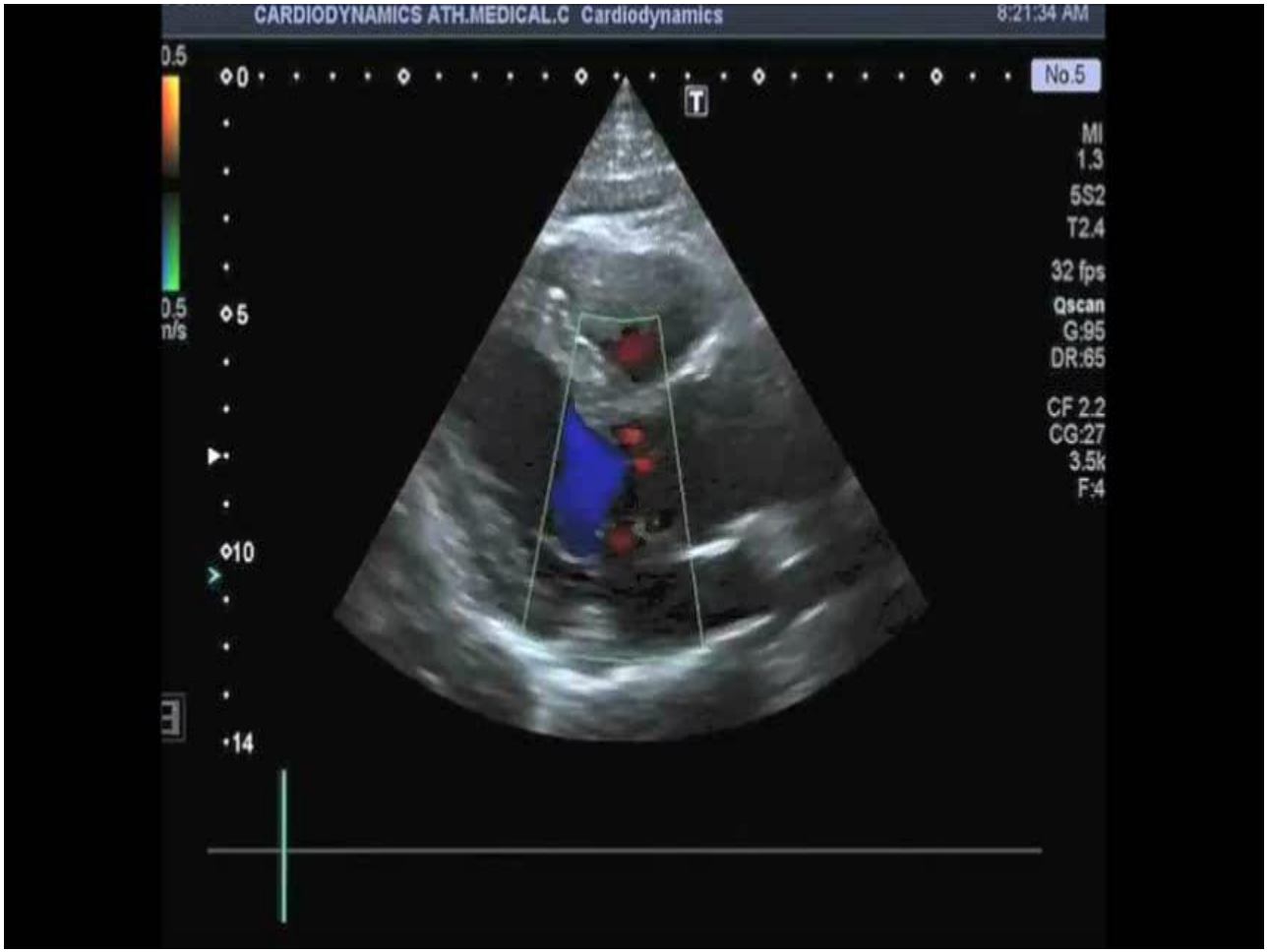

Doming and prolapsing of
Aortic Valve cusps
64
HR


)8:12:56
Freq.: 1.7 MHz/3.4 MHz
Power: 0.0 dB
FPS: 64.3
Depth: 16.0 cm
V
5
10
15
55
HR

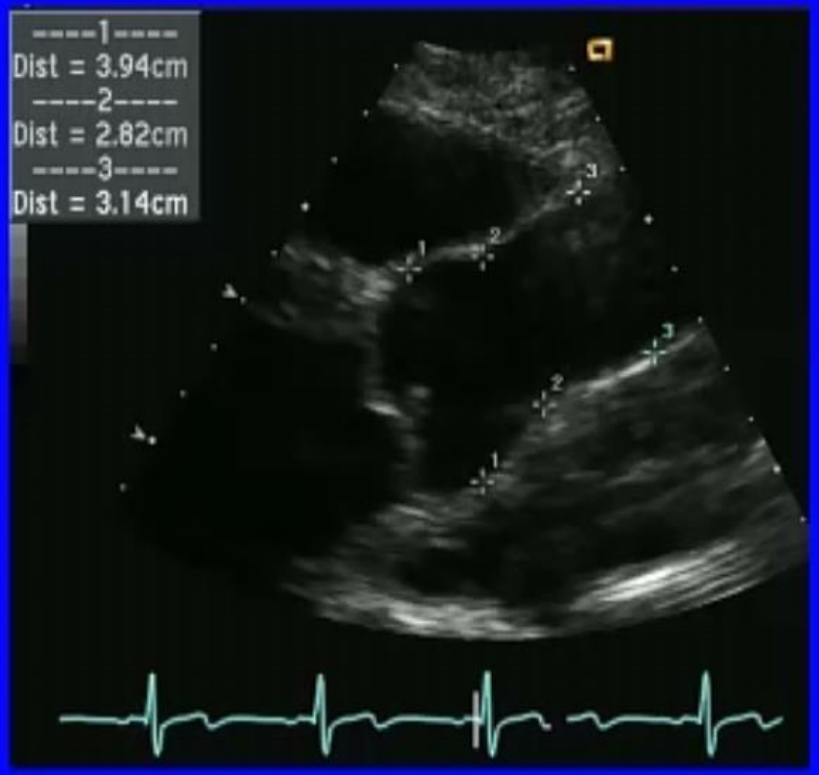

CARDIODYNAMICS ATH.MEDICAL.C Cardiodynamics
1:58:17 PM
No.2
MI
1.3
5S2
T2.4
51 fps
G:86
DR:65
0
5
10
15


CARDIODYNAMICS ATH.MEDICAL.C Cardiodynamics
12:56:49 PM
No.5
MI
1.3
5S2
T2.4
51 fps
Qscan
G:99
DR:65
0
5
10
15


CARDIODYNAMICS ATH.MEDICAL.C Cardiodynamics
8:54:41 AM
No.14
MI
1.4
5S2
T2.4
51 fps
Qscan
G:94
DR:65
0
5
10
12


CARDIODYNAMICS ATH.MEDICAL.C Cardiodynamics
8:21:34 AM
0.5
0
No.5
MI
1.3
5S2
T2.4
32 fps
Qscan
G:95
DR:65
CF 2.2
CG:27
3.5k
F:4
0.5
m/s
5
10
14

33M with BAV and aortic regurgitation



Aortic measurements

Evolutie clinica

- La copil ,in cele mai multe cazuri este asimptomatic
- Se estimeaza ca 1 din 50 de copii cu B Ao are boala valvulara severa pana la adolescenta
- B Ao poate fi simptomatica la copil prin:

❑ St Ao severa prin dimensiune mica a orificiului valvular

❑ I Ao severa

- In perioada de adult

➢ Calcificare valvulara

➢ Aortopatie

STENOZA AORTICA IN B AO

Stenoza severa nn

- Poate fi dgn fetal
- In viata fetala ,supravietuirea nu reprezinta o problema deoarece cordul drept poate asigura tot debitul cardiac gratie PFO si CAP
- St Ao severa la nn asociaza frecvent fibroza miocardica ameliorata partial daca obstructia este inlaturata

STENOZA AORTICA IN B AO

- Copiii care prezinta st Ao severa nn ,chiar daca se realizeaza procedura chirurgicala sau interventionala ,au un prognostic sever
- Copiii care prezinta gradient maxim >50 mmHg au un risc de 1,25 %/an de eveniment cardiovascular
- Copiii cu grd<25 mm Hg,20% au necesitat interventie in evolutia lor
- Studiu UK:

Copiii cu st Ao usoara , <20% au boala valvulara usoara dupa 30 ani de urmarire

PROGRESIA STENOZEI ESTE MAI RAPIDA IN TIPUL 1 (FUZIUNEA COR STG/COR DPT)

INSUFICIENTA AO IN B AO

- Frecventa mai mica decat stenoza aortica
- Mecanism

✓ Prolaps de cuspe

✓ Dilatarea jonctiunii sinotubulare

✓ endocardita

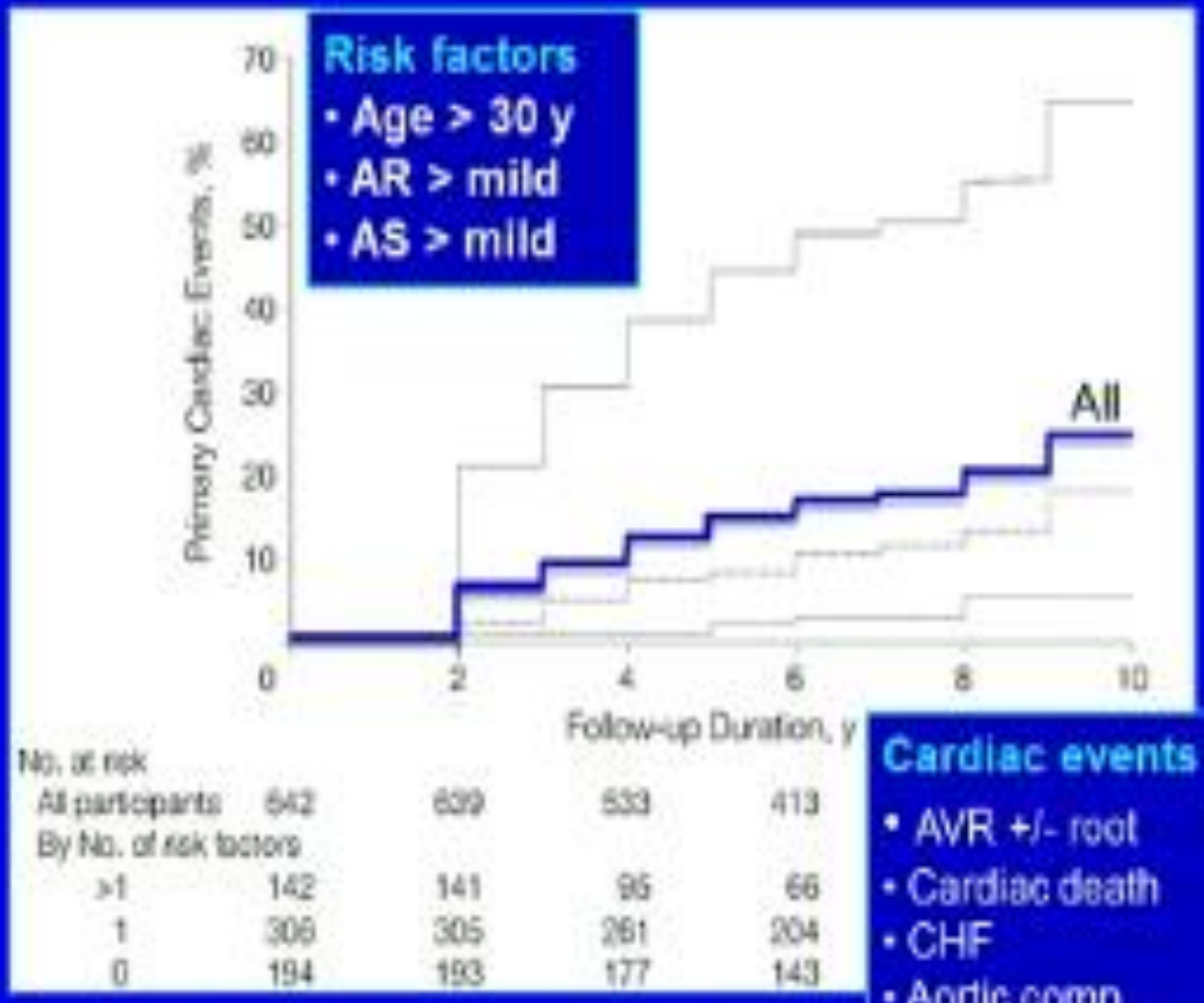
INSUFICIENTA AORTICA IN B AO

- 164 children with BAV and serial echocardiographic examinations
- •Progression of > 1 grade of AR in 44% of patients with R-N (tip 2) fusion vs. 27% with R-L fusion (tip 1)
- (Fernandes et al. J Am Coll Cardiol 2007;22:2211-4)

- La adulti evolutie depinde de

➢ Varsta
➢ Gradul stenozei aortice
➢ Gradul insuficientei aortice
➢ Factori de risc aditionali
❖ HTA
❖ Hipercolesterolemie

Outcomes in adults with bicuspid aortic valves

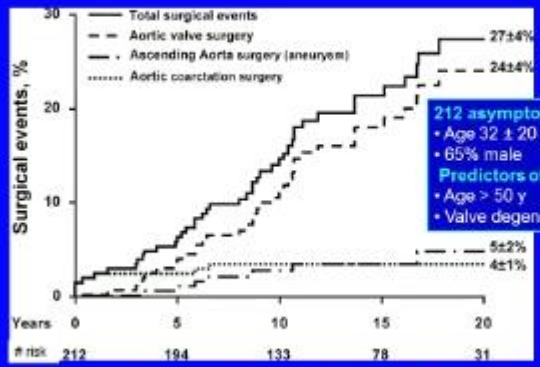


Cardiac events 161 (25%)

- AVR +/- root 142 (22%)
- Cardiac death 17 (3%)
- CHF 16 (2%)
- Aortic comp. 11 (2%)

Tzemos, N. et al. JAMA 2008;300:1317

Outcomes in adults with bicuspid aortic valves

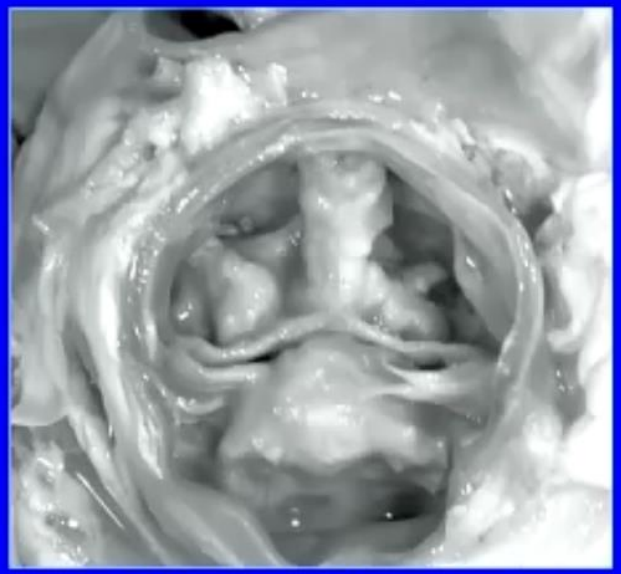


212 asymptomatic BAV pts
- Age 32 ± 20 y
- 65% male

Predictors of outcome
- Age > 50 y
- Valve degeneration

Michelena HT et al. Circulation 2008;117: 2776

The Bicuspid Aortic Valve
Secondary calcification and stenosis



Otto CM Valvular Heart Disease 2nd Ed 2004

AVR for AS (n=933)
53% due to BAV

< 70 years = 60% BAV

> 70 years = 40% BAV

Ko and Roberts 2005

Calcificarea este accelerata de

- modificarea geometriei valvei
- Turbulenta jetului

La care se adauga la fel ca la valva tricuspa ‘mecanismul clasic’

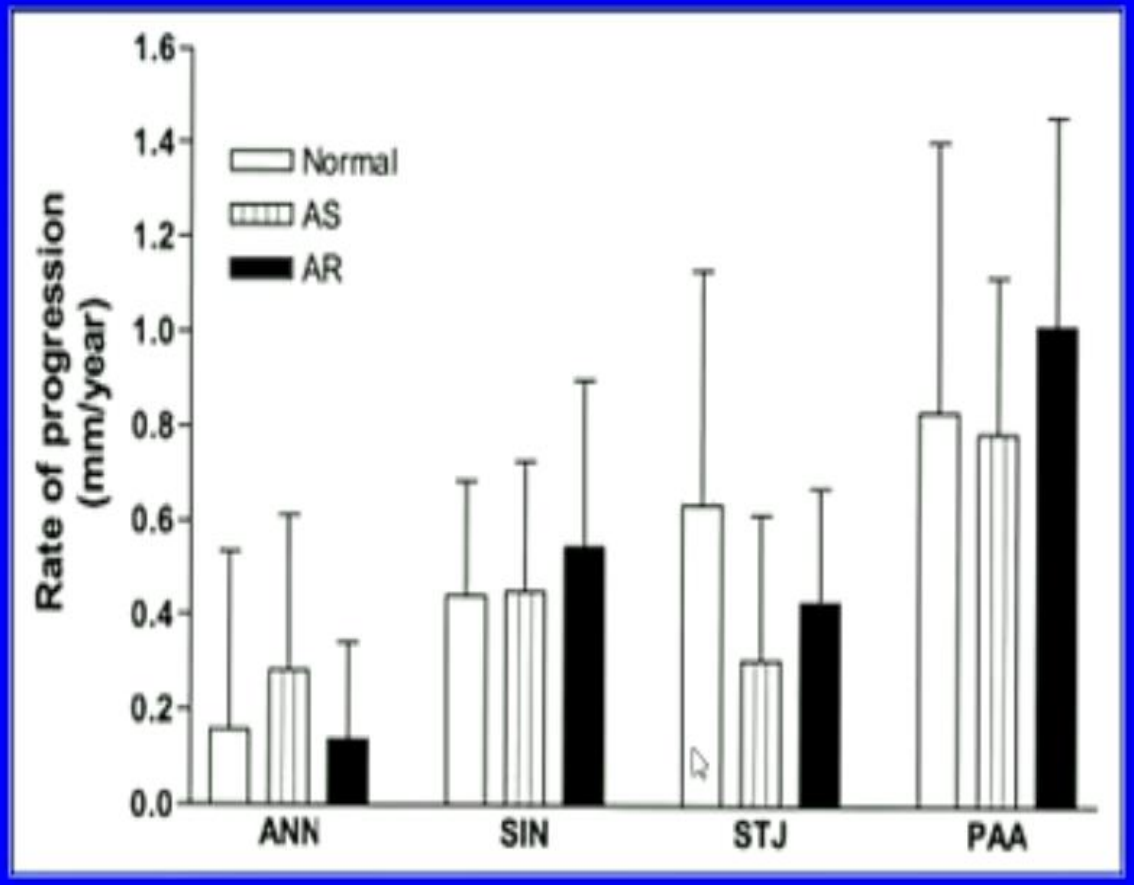
- Leziune endoteliala
- Proces inflamator
- Depozit lipoproteine

Bicuspid Aortic Valve Disease

Aortopathy

- ↑Aortic diameters
  - Sinuses
  - Ascending aorta
- ↑Aortic stiffness
- ↑ Rate of aortic enlargement
- ↑ Risk of aortic dissection

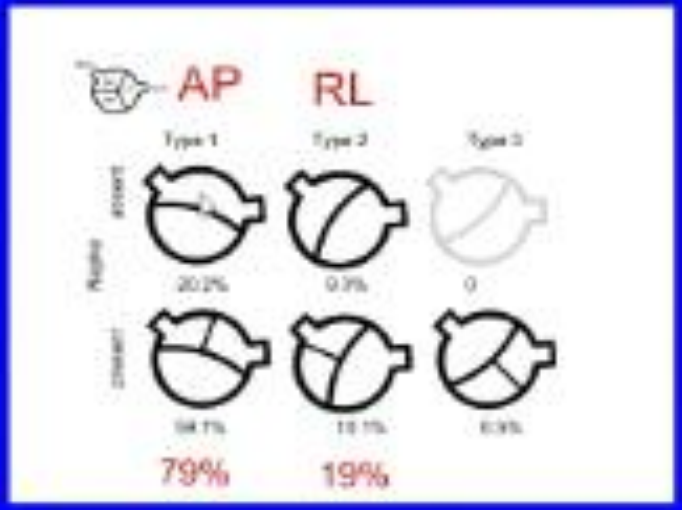
Progression of aortic dilation is independent of valve function in BAV pts

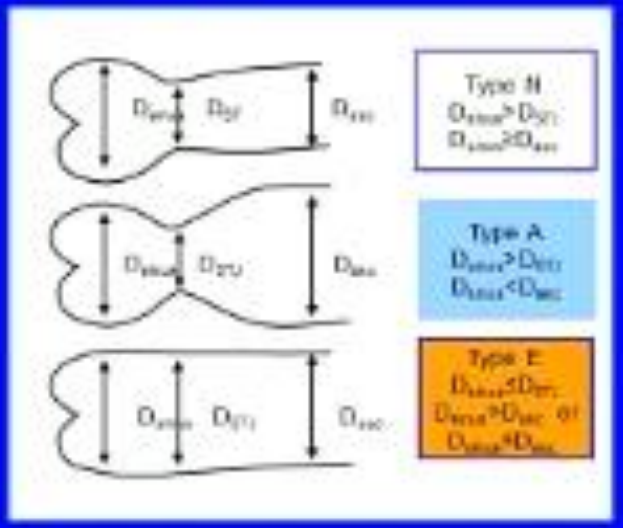


Ferencik and Pape, Am J Cardiol 2003; 92: 43

Phenotypic Classification: BAV and Aortic Root

Seattle 2002-05, 192 subjects





Leaflets

Type 1 = fusion of R+L cusps
Type 2 = fusion of R+N cusps
Type 3 = fusion of L+N cusps

Schaefer, et al, Heart 2008

Type 1 --- Dilated Sinuses

Type 2 ---- ↑Asc Ao + MVP

Phenotypic Classification: BAV and Aortic Root

Seattle 2002-05, 158 subjects

Table 2

Elastic properties of sinus of Valsalva, ascending aorta, and aortic arch leaflet orientation

	A-P	R-L	p Value
Sinus of Valsalva			
Dimension (cm, diastole)	3.48 ± 0.49	3.06 ± 0.59	<0.01
Stiffness index	12.98 (2.78–42.07)	6.41 (2.75–59.72)	<0.01
Distensibility (kPa^{-1} 10^{-3})	12.71 (4.24–71.29)	27.36 (2.70–59.66)	<0.01
Ascending aorta			
Dimension (cm, diastole)	3.22 ± 0.51	3.18 ± 0.64	0.78
Stiffness index	6.40 (2.65–83.81)	9.63 (2.67–60.83)	0.26
Distensibility (kPa^{-1} 10^{-3})	26.33 (1.93–66.29)	16.01 (2.51–62.82)	0.40
Aortic arch			
Dimension (cm, diastole)	2.34 ± 0.40	2.83 ± 0.45	<0.001
Stiffness index	4.75 (1.65–41.80)	6.98 (1.57–23.91)	0.37
Distensibility (kPa^{-1} 10^{-3})	33.11 (2.87–130.72)	23.75 (5.80–158.65)	0.38

Values expressed as mean ± 1 SD or median (range).

Schaefer, et al, AJC 2007

- The elastic laminae of the aortic media provides structural support and elasticity to the aorta. In normal tricuspid valve patients (A), fibrillin-1 microfibrils tether smooth muscle cells to adjacent elastin and collagen matrix components.

In patients with BAV (B), deficient microfibrillar elements result in smooth muscle cell detachment, MMP release, matrix disruption,cell death, and a loss of structural support and elasticity..

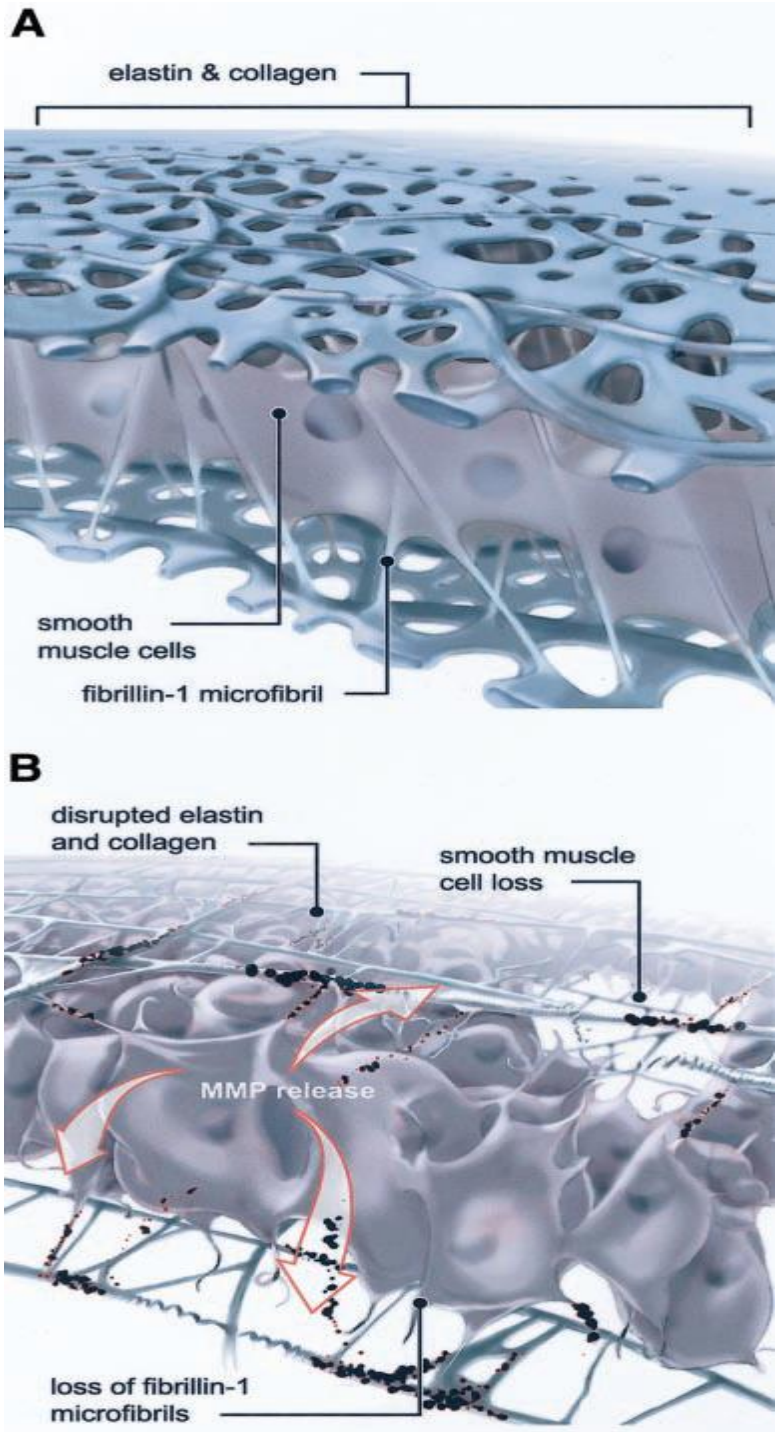
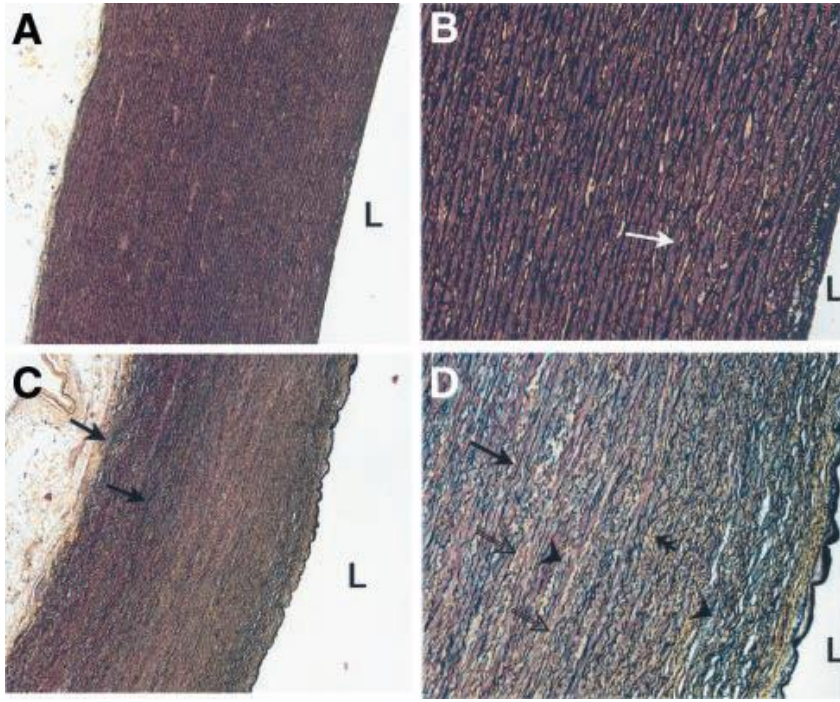
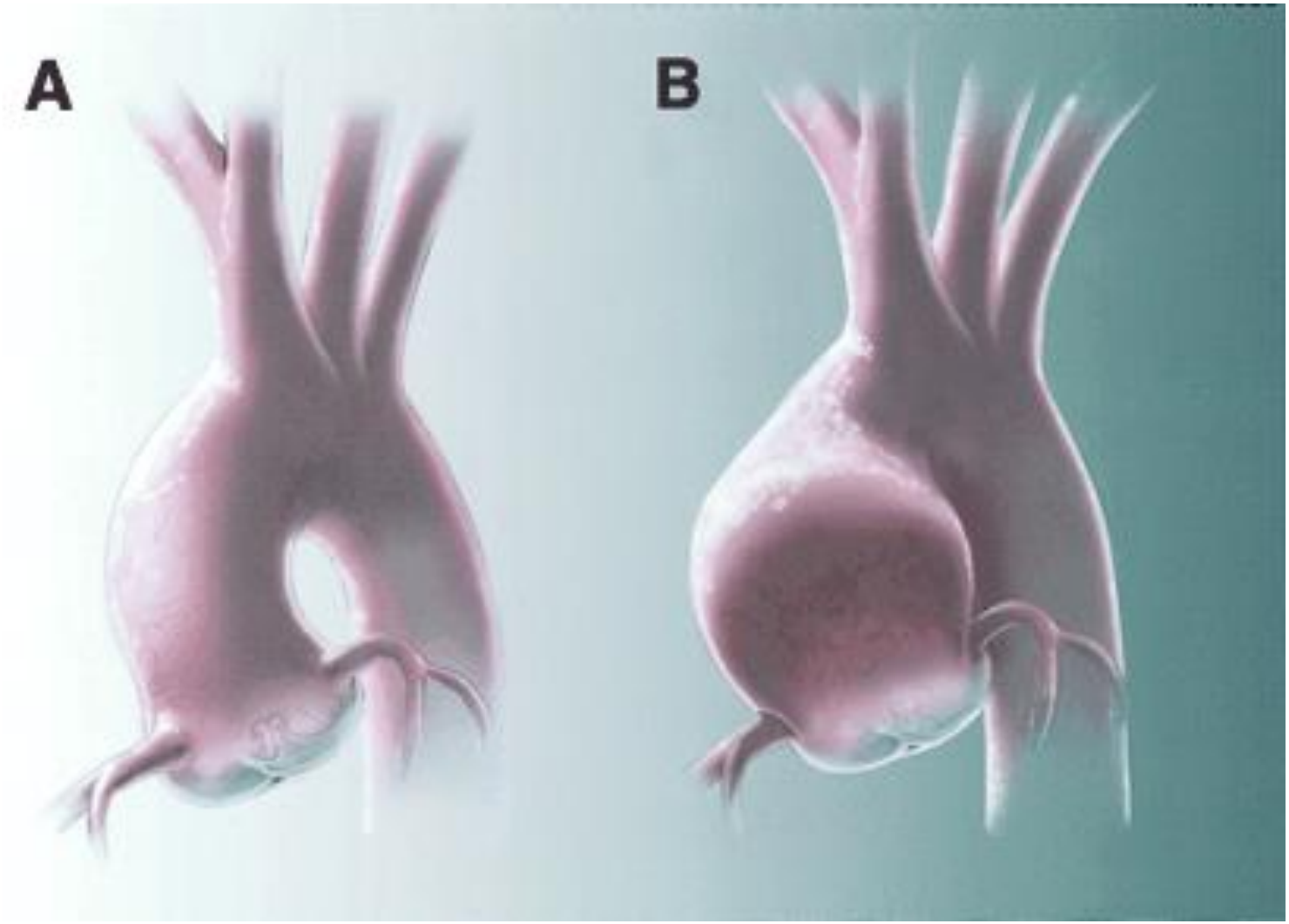
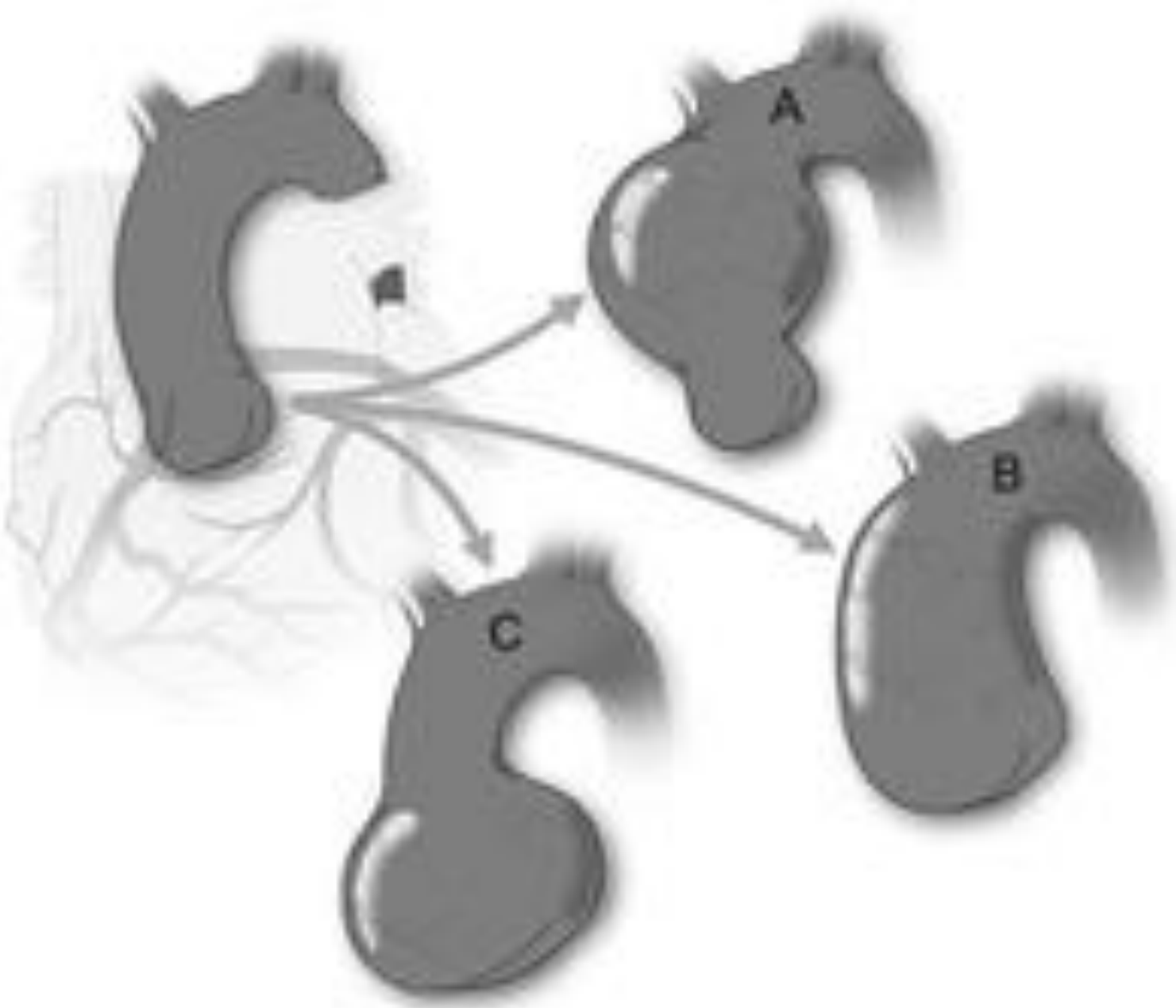
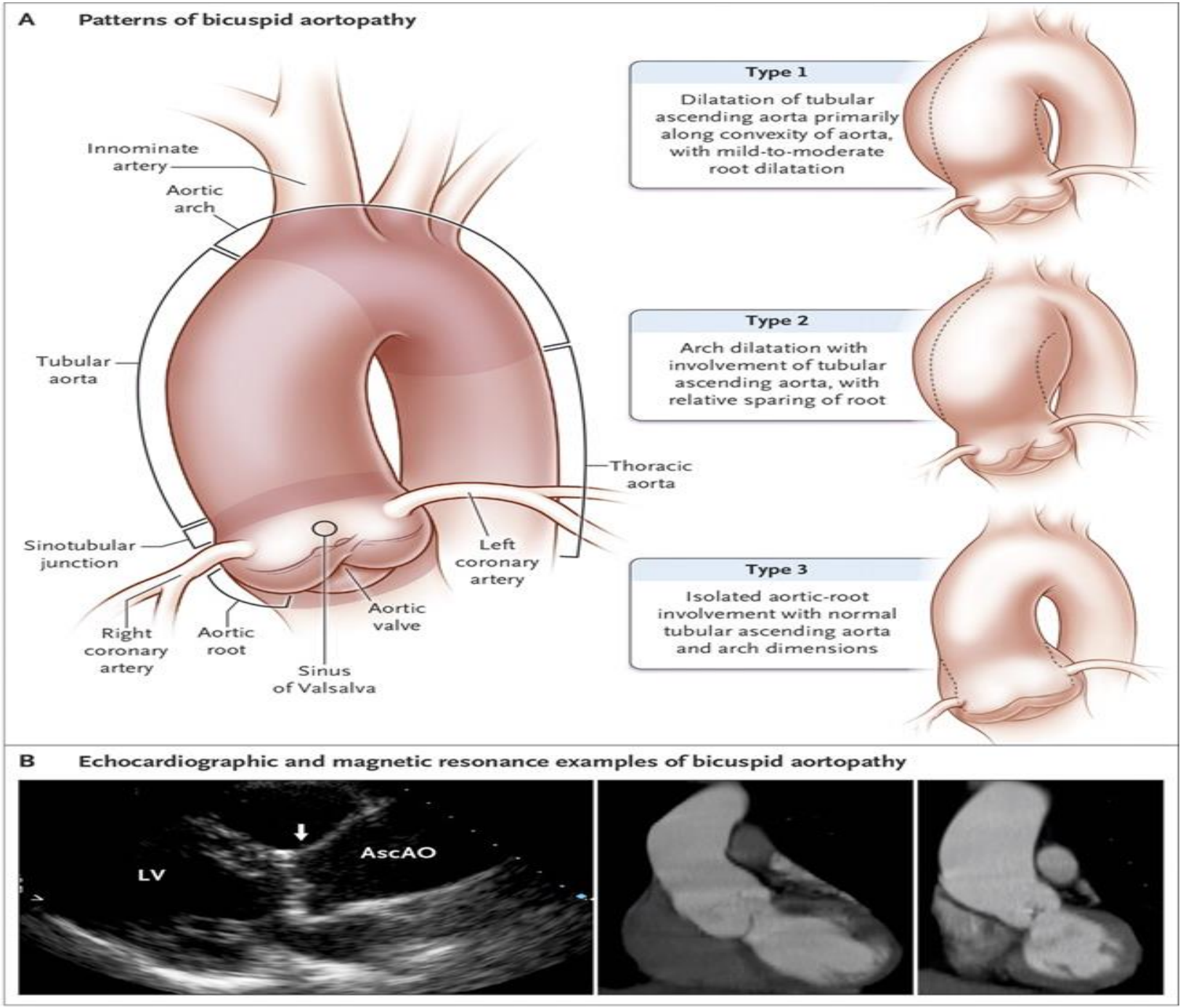


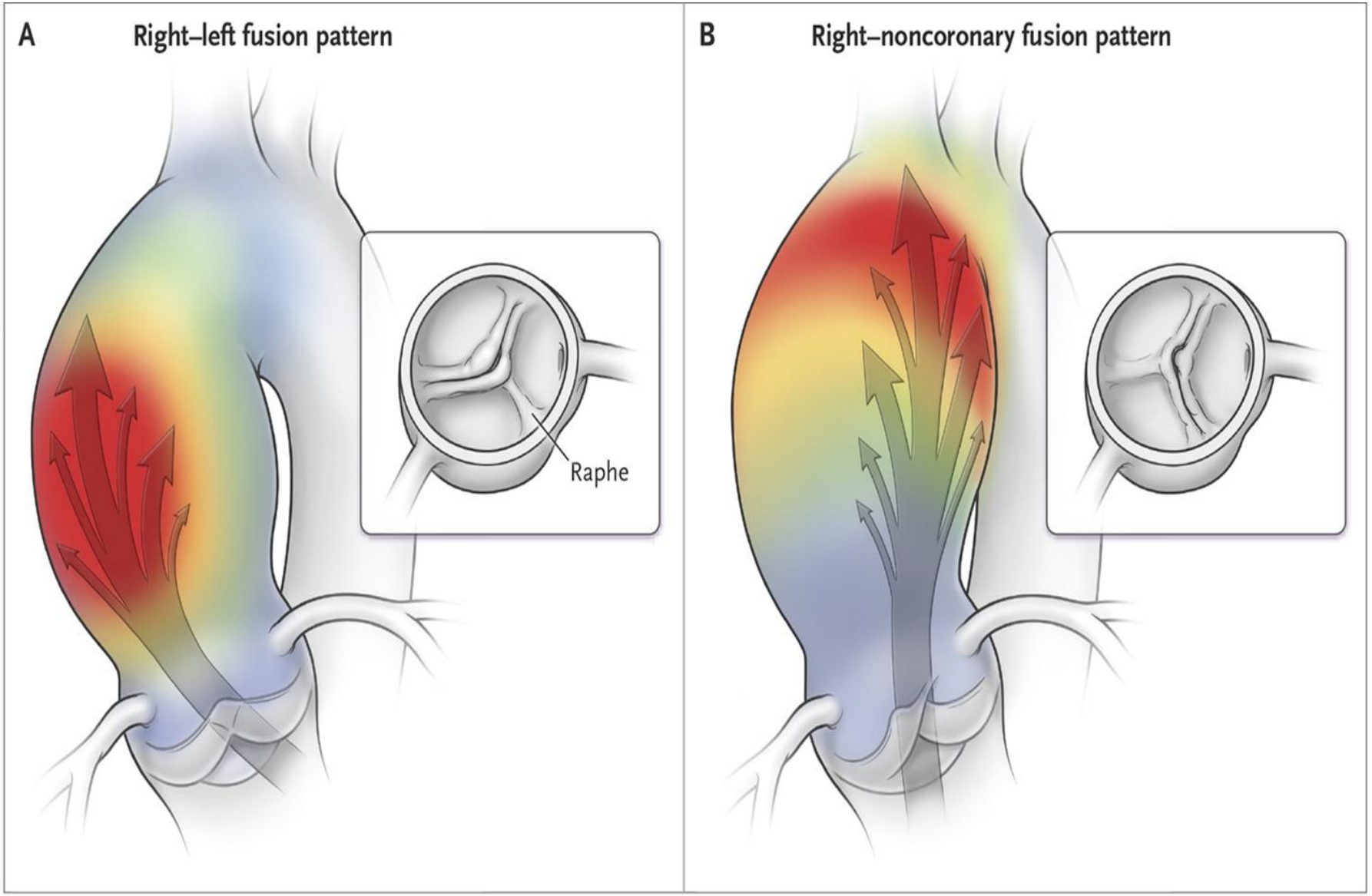
Figure 5. The healthy aorta of a patient with a tricuspid aortic valve (A and B) has a highly organized media of smooth muscle cells (stained red, white arrow), elastic plates (stained black), and collagen (stained yellow). In patients with BAV malformations (C and D), the aorta shows significant changes. Loss of smooth muscle and fibrosis (open arrows) causes collapse of the elastic plates (arrows). Mucopolysaccharides (stained greenish blue) and disarray of the elastic plates (double arrow) are evident in the media.




A
B


A
B
C


A Patterns of bicuspid aortopathy
Innominate artery
Aortic arch
Tubular aorta
Thoracic aorta
Sinotubular junction
Left coronary artery
Aortic valve
Right coronary artery
Aortic root
Sinus of Valsalva
Type 1
Dilatation of tubular ascending aorta primarily along convexity of aorta, with mild-to-moderate root dilatation
Type 2
Arch dilatation with involvement of tubular ascending aorta, with relative sparing of root
Type 3
Isolated aortic-root involvement with normal tubular ascending aorta and arch dimensions
B Echocardiographic and magnetic resonance examples of bicuspid aortopathy
LV
AscAO


A
Right–left fusion pattern
Raphe
B
Right–noncoronary fusion pattern

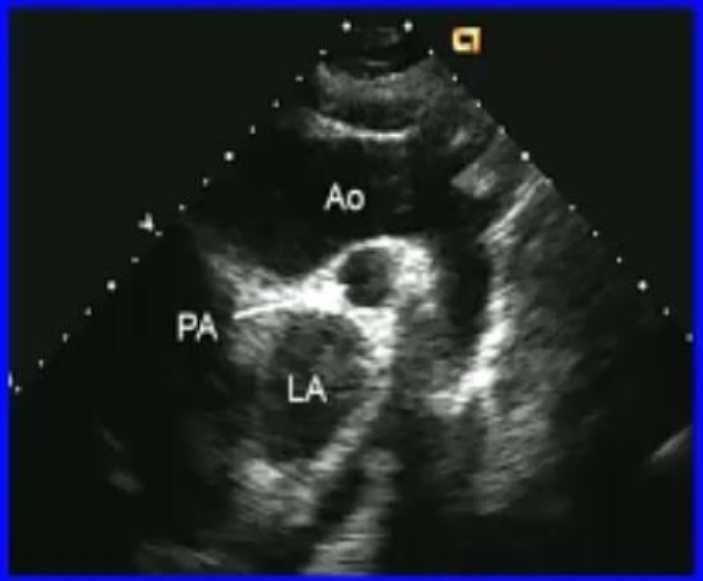
Bicuspid Aortic Valve Disease

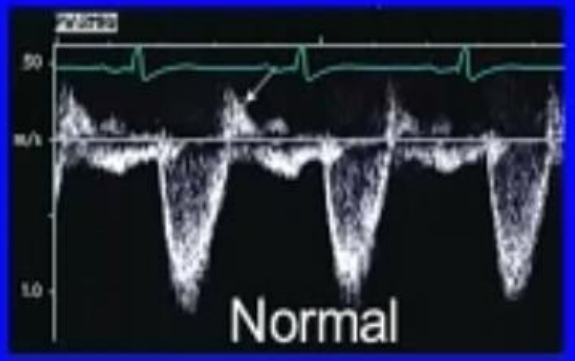
ASSOCIATED CONDITIONS

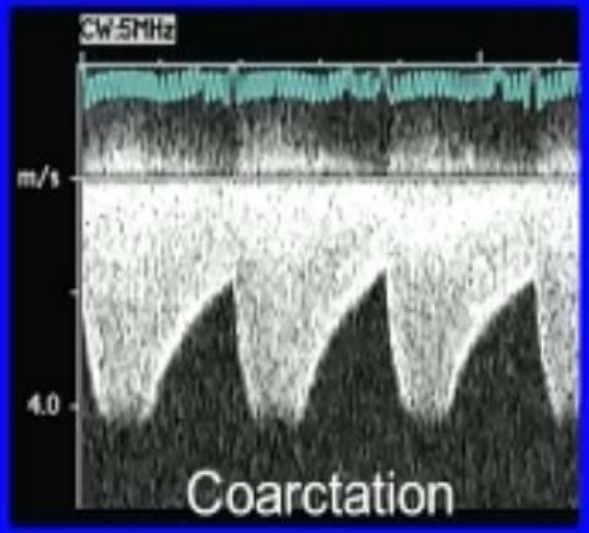
Condition	Incidence of BAV (%)	Comments
Coarctation of the aorta	50	BAV confers increased risk of aortic complications
Turner syndrome	30	Most frequent cardiac abnormality; right-left cusp fusion most common
Supravalvular aortic stenosis	30	Usually part of William syndrome
Subvalvular aortic stenosis	23	May result in significant aortic regurgitation
Patent ductus arteriosus	Unknown	Usually diagnosed in childhood/infancy
Sinus of Valsalva aneurysm	15-20	Frequently asymptomatic; most commonly involves right coronary sinus
Ventricular septal defect	30	May result in significant aortic regurgitation
Shone complex	60-85	Series of left-sided obstructive lesion
Ascending aortic dilatation	Common	BAV is one of the most common associates of a dilated ascending aorta

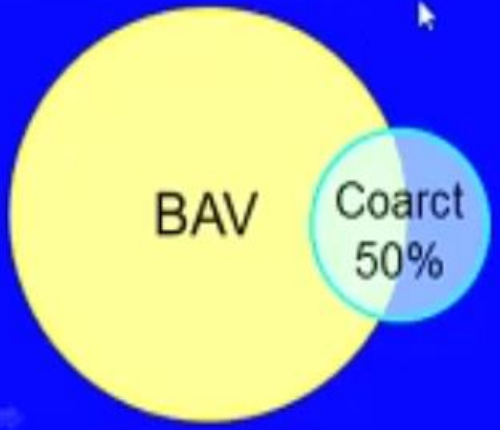
Braverman, In Otto and Bonow, Valvular Heart Disease, 4th Ed, 2013

Bicuspid Aortic Valve Disease
Associated with aortic coarctation









Bicuspid Aortic Valve Disease

Risk of Endocarditis

- Risk is higher than general population
  - Difficult to quantitate
  - Estimated lifetime risk 271/100,000
  - Comparative risk 5/100,000
  - Endocarditis may be initial presentation
- Antibiotic prophylaxis no longer recommended
- Emphasis now on:
  - Optimal dental care
  - Oral health
  - Education about signs and symptoms

MANAGEMENT

- Medicamentos
- beta blocant (cu exceptia cazurilor cu insuficienta severa)

-monitorizarea TA

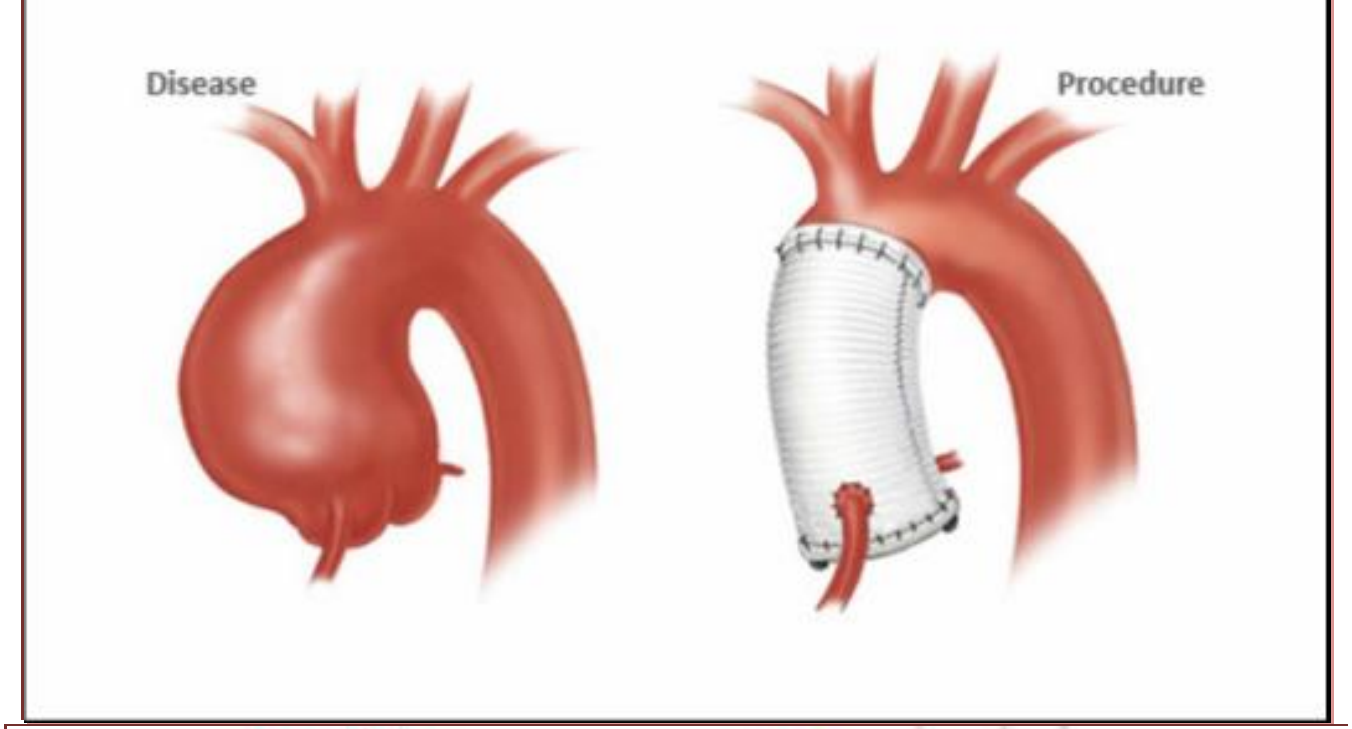
- Blocant de receptor angiotensina

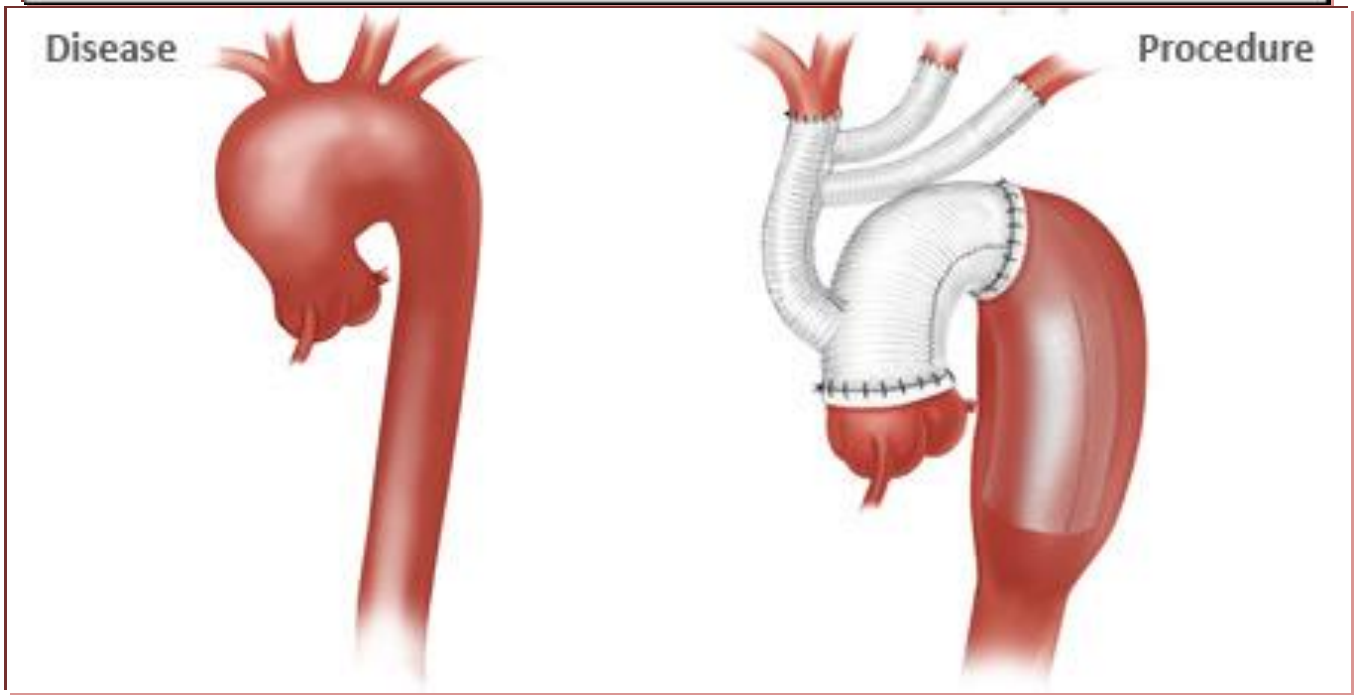
Losartan

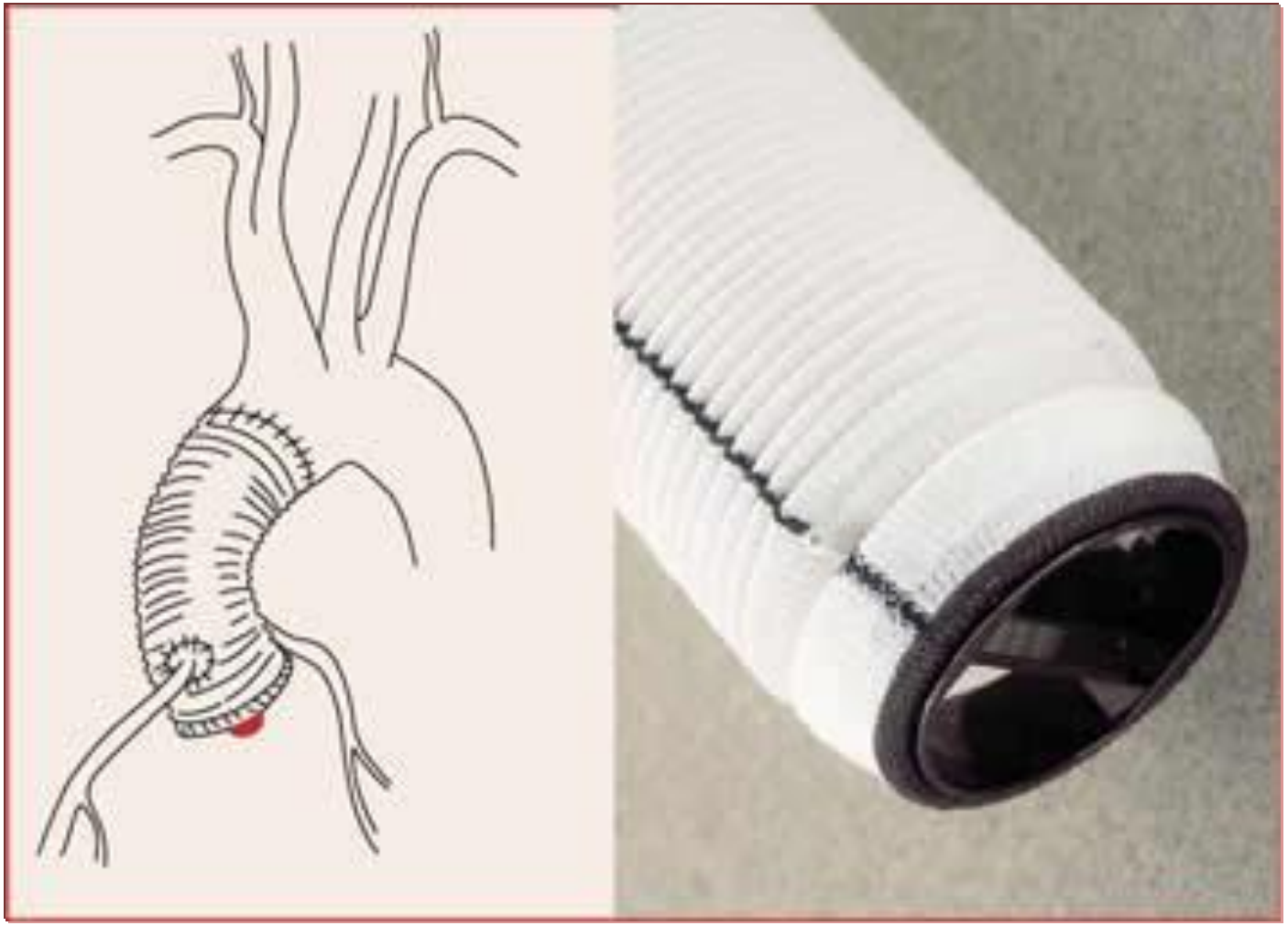
MANAGEMENT

Chirurgical

- Valvuloplastie nn
- Ross
- Inlocuire valvulara
- Inlocuire valvulara si Ao ascendenta
- Bental


Disease
Procedure


Disease
Procedure

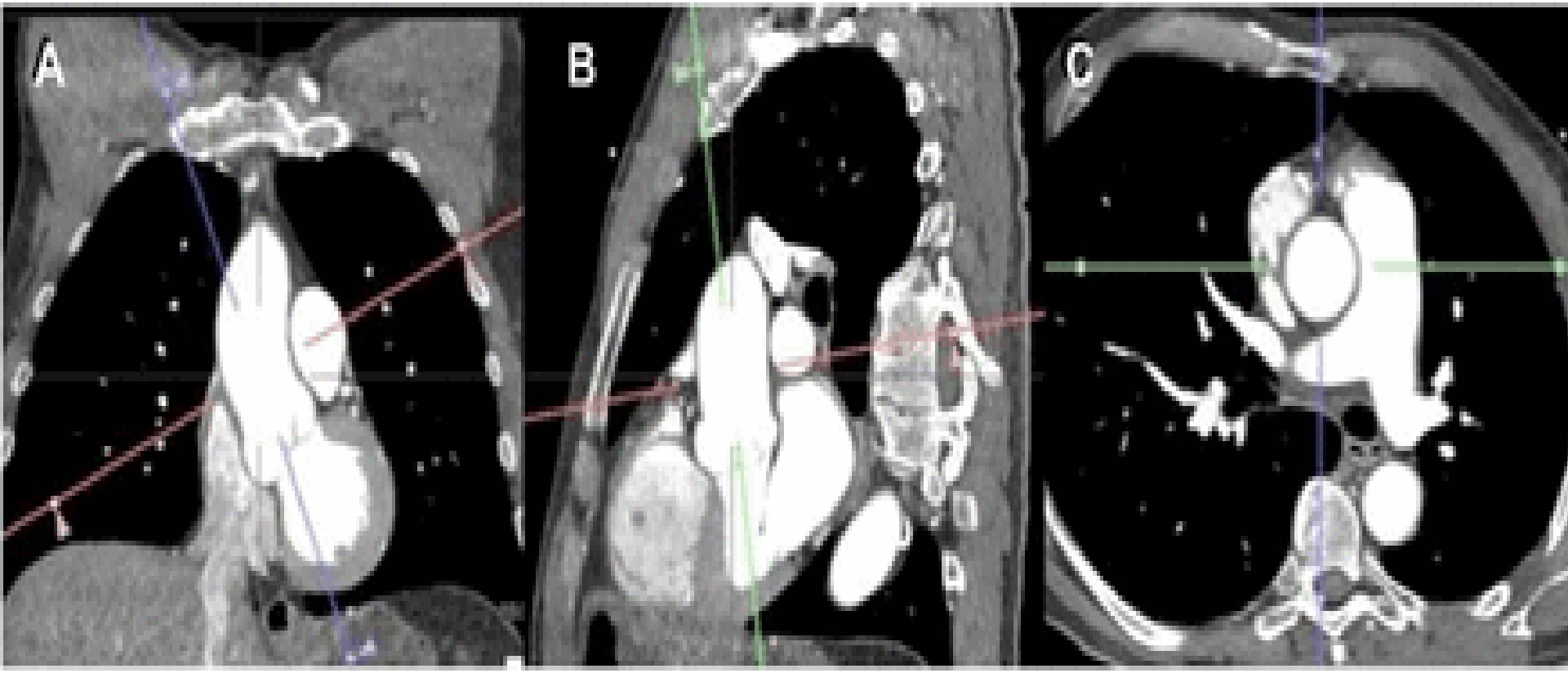


2014
AHA/ACC Guideline

- Class I 2. Aortic magnetic resonance angiography or CT angiography is indicated in patients with a bicuspid aortic valve when morphology of the aortic sinuses, sinotubular junction, or ascending aorta cannot be assessed accurately or fully by echocardiography. (Level of Evidence: C)
- TTE can provide accurate assessment of the presence and severity of aortic dilation in most patients. However, in some patients, only the aortic sinuses can be visualized, because the ascending aorta is obscured by intervening lung tissue. When echocardiographic images do not provide adequate images of the ascending aorta to a distance ≥4.0 cm from the valve plane, additional imaging is needed

2014
AHA/ACC Guideline

- Class I 1. Operative intervention to repair the aortic sinuses or replace the ascending aorta is indicated in patients with a bicuspid aortic valve if the diameter of the aortic sinuses or ascending aorta is greater than 5.5 cm (113, 268, 269). (Level of Evidence: B)


A
B
C

2014
AHA/ACC Guideline

- Surgery is recommended with aortic dilation of 5.1 cm to 5.5 cm only if there is a family history of aortic dissection or rapid progression of dilation. In all other patients, operation is indicated if there is more severe dilation (5.5 cm). The writing committee also does not recommend the application of formulas to adjust the aortic diameter for body size

2014
AHA/ACC Guideline

- Class IIa 2. Replacement of the ascending aorta is reasonable in patients with a bicuspid aortic valve who are undergoing aortic valve surgery because of severe AS or AR (Sections 3.2.3 and 4.3.3) if the diameter of the ascending aorta is greater than 4.5 cm. (Level of Evidence: C

Concluzii

- Bicuspidia aortica este frecventa in populatie
- Reprezinta principala cauza de disfunctie valvulara aortica la copil
- Disfunctia valvulara progreseaza spre varsta de adult in special stenoza

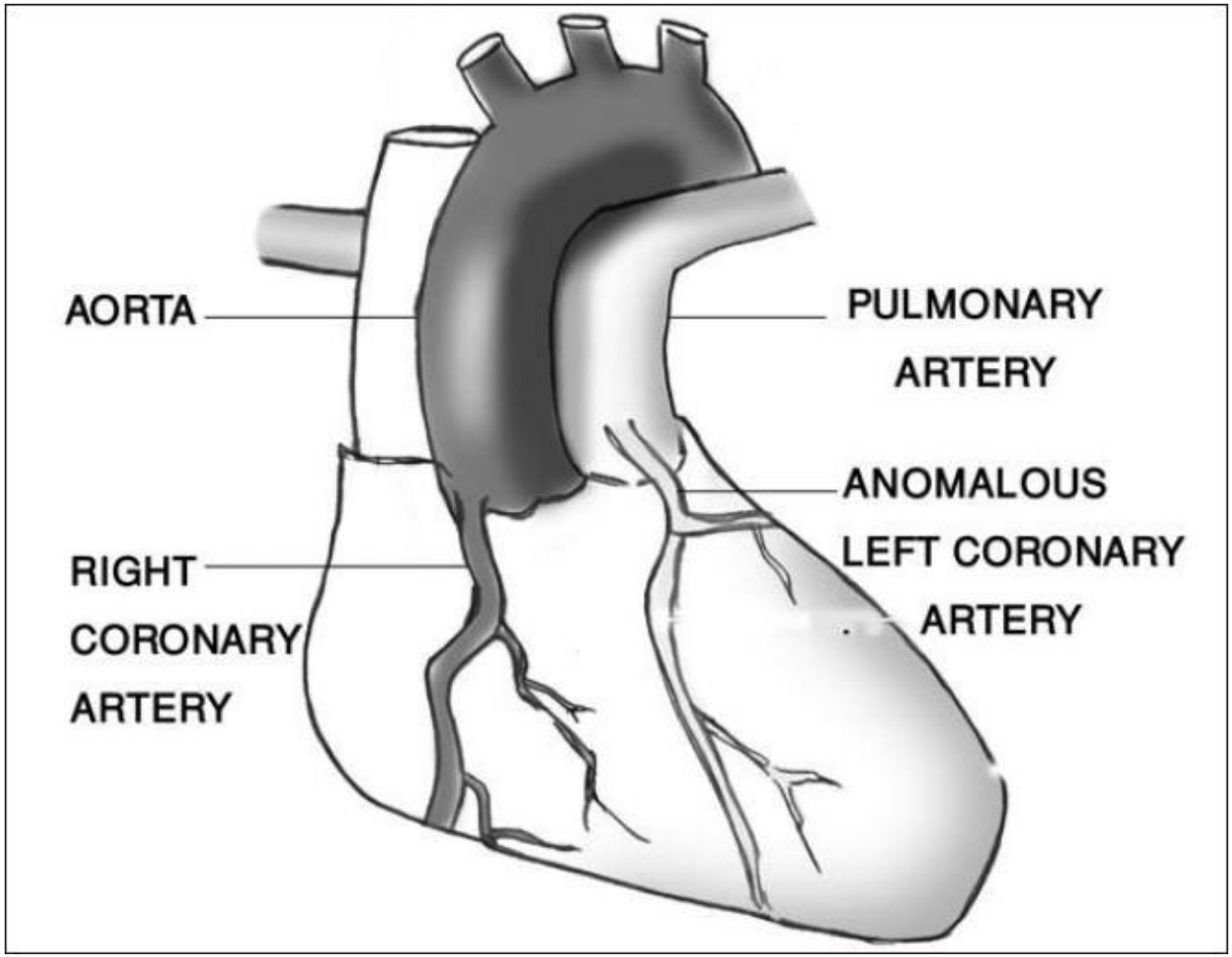
Concluzii

- Dilatarea aortica:
  - Este determinata de structura peretelui arterial si morfologia valvei
  - Nu depinde de functia valvei

ALCAPA

DR .Alin Nicolescu
Departament de Cardiologie pediatrica
SCUC “M.S.Curie”

- Anomalie de origine a arterei coronare stangi- origine din AP
- Sd Bland-White –Garland
- 1/300 000 nn
- 0,25-0,5% din MCC


AORTA
PULMONARY ARTERY
ANOMALOUS LEFT CORONARY ARTERY
RIGHT CORONARY ARTERY

- Reprezinta o anomalie congenitala rara
- Exista 2 tipuri de sd ALCAPA

➢ Tipul infantil

❖ IMA

❖ ICC

❖ 90% mor in primul an de viata

➢ Tipul adult

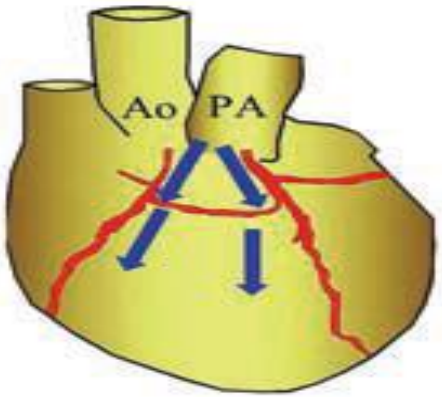
➢ Fara simptomatologie precoce

➢ Ischemie micardica cr silentioasa

➢ I M

➢ IVS

➢ Poate reprezenta o cauza de moarte subita



Neonatal Period

Pressure PA = Ao
Antegrade flow in LCA

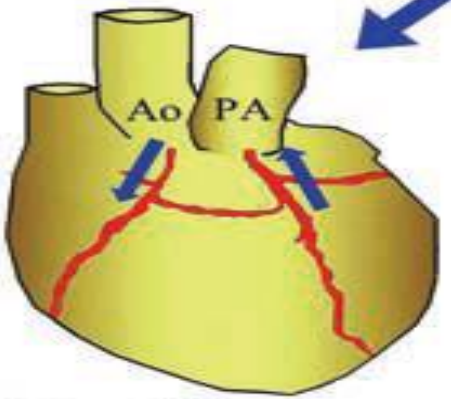
↓

No symptoms

Decrease in PA pressure
Ability to develop collaterals between RCA and LCA

2 TYPES OF ALCAPA

Infant Type



Pressure PA < Ao
Retrograde flow LCA
No collaterals between RCA and LCA
RCA and LCA normal in size

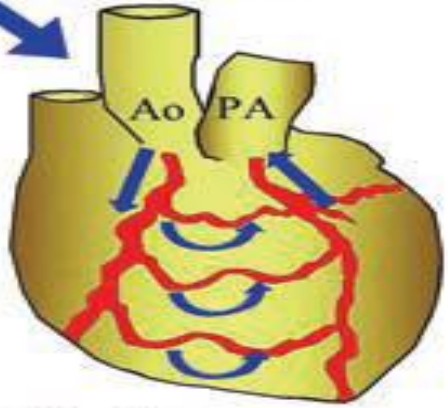
↓

Ischemic cardiomyophathy due to infarction

↓

90% death first year

Adult Type



Pressure PA < Ao
Retrograde flow LCA
Collaterals between RCA and LCA
Marked dilatation of the LCA and RCA due to the increased longstanding volume

↓

Chronic myocardial ischema
Dysrhythmias

↓

Sudden death

ALCAPA-tipul infantil

- In viata fetala e bine tolerata
- ➢ Pres Ao=pres AP
- Dupa nastere pres in AP scade---fluxul in ACS devine treptat retrograd
- Simptomatologia infantil/adult depinde de colateralele existente

ALCAPA-tipul infantil

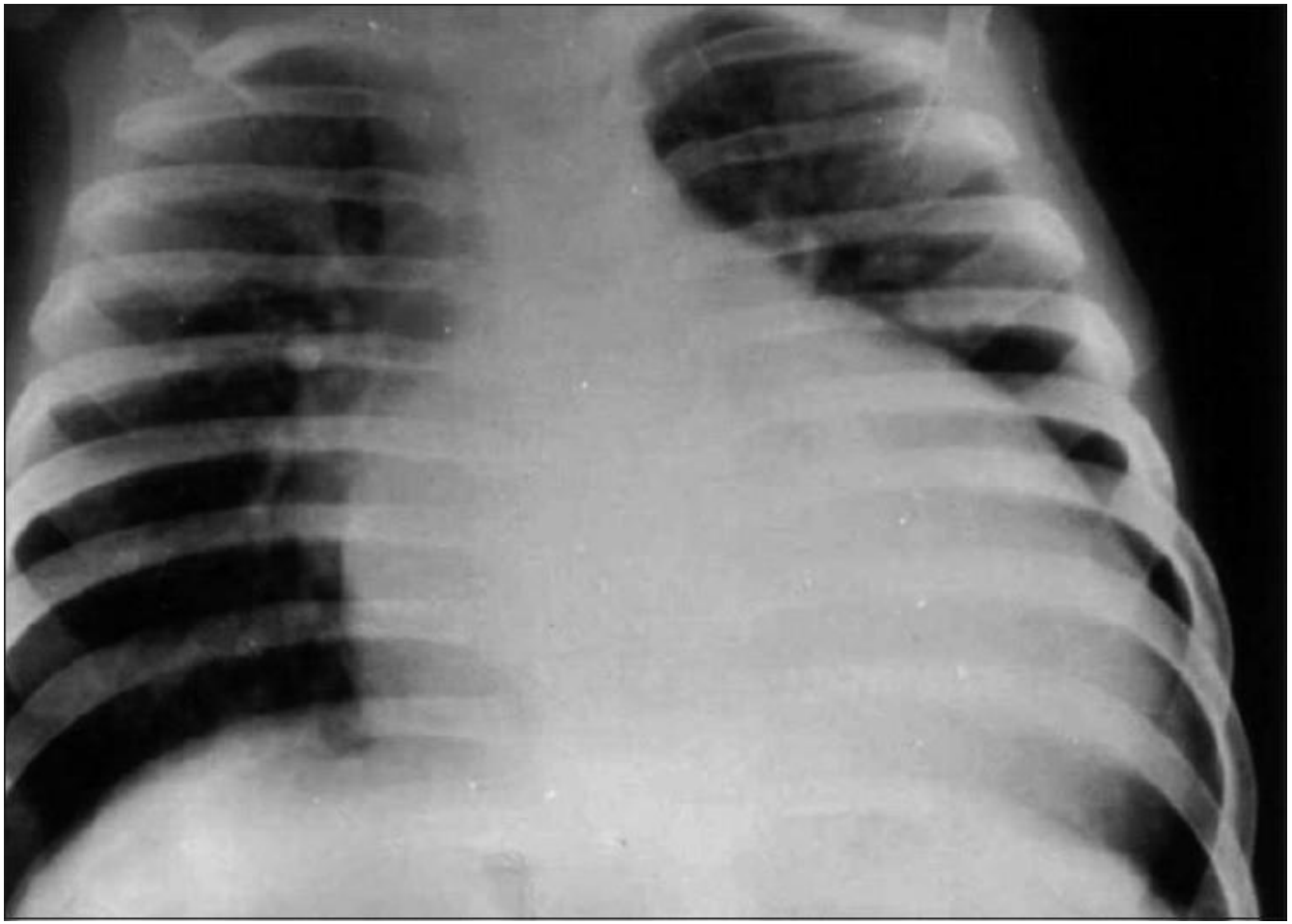
- Debutul simptomatologiei –la aprox 8 s de viata
- Nu exista circulatie colaterala eficienta
- Flux revers la niv ACS-----ischemie VS
- ------IC/IM si IMA
- DGN diferential cu CMD

ALCAPA-tipul infantil

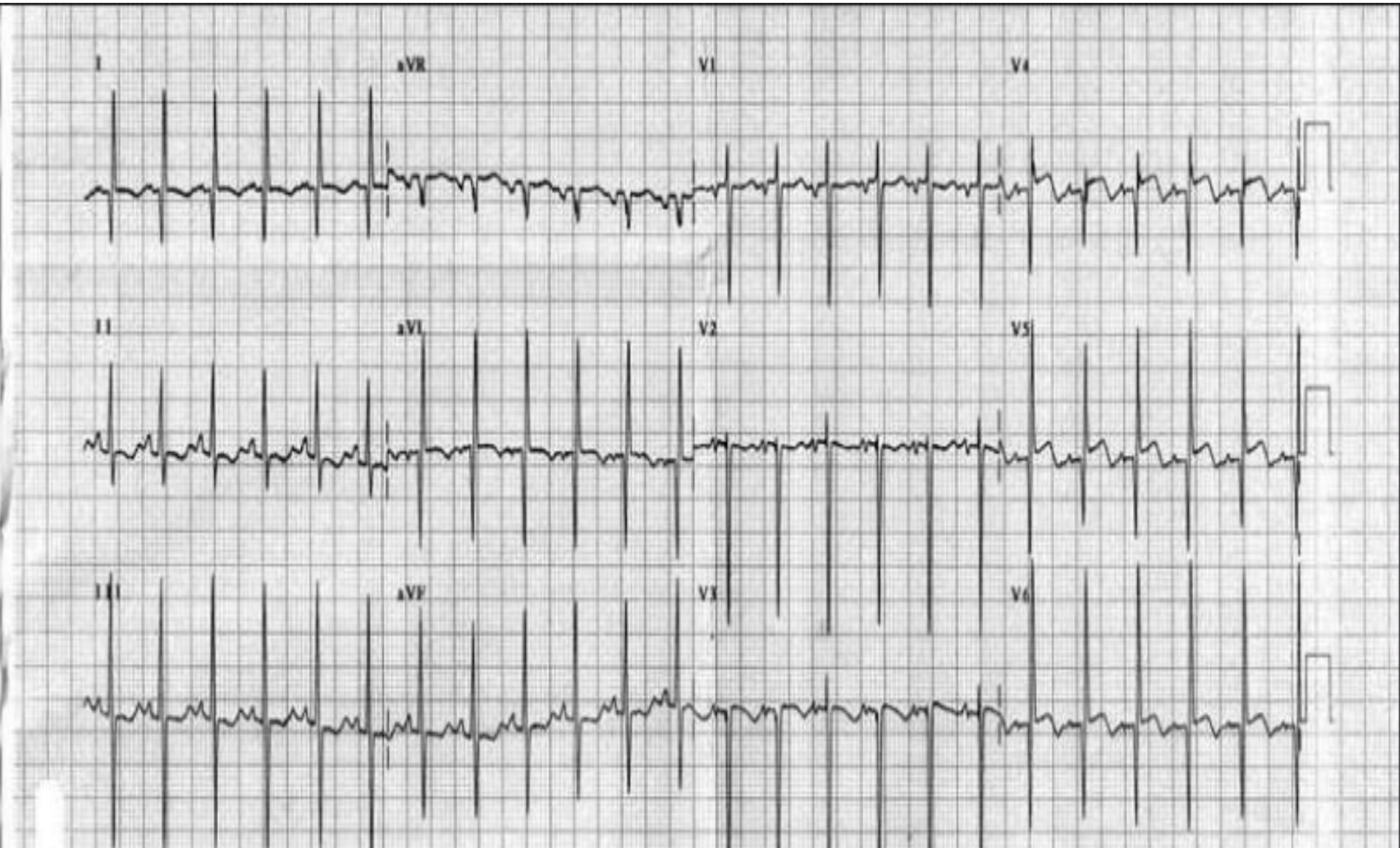
- Clinic
- Falimentul cresterii
- Transpiratie profuza
- Paloare
- Dispnee
- Angina la plans sau la efortul de alimentare

ALCAPA-tipul infantil

- Fara corectie chirurgical ,deces la saptamani sau luni de la nastere in 90% din cazuri

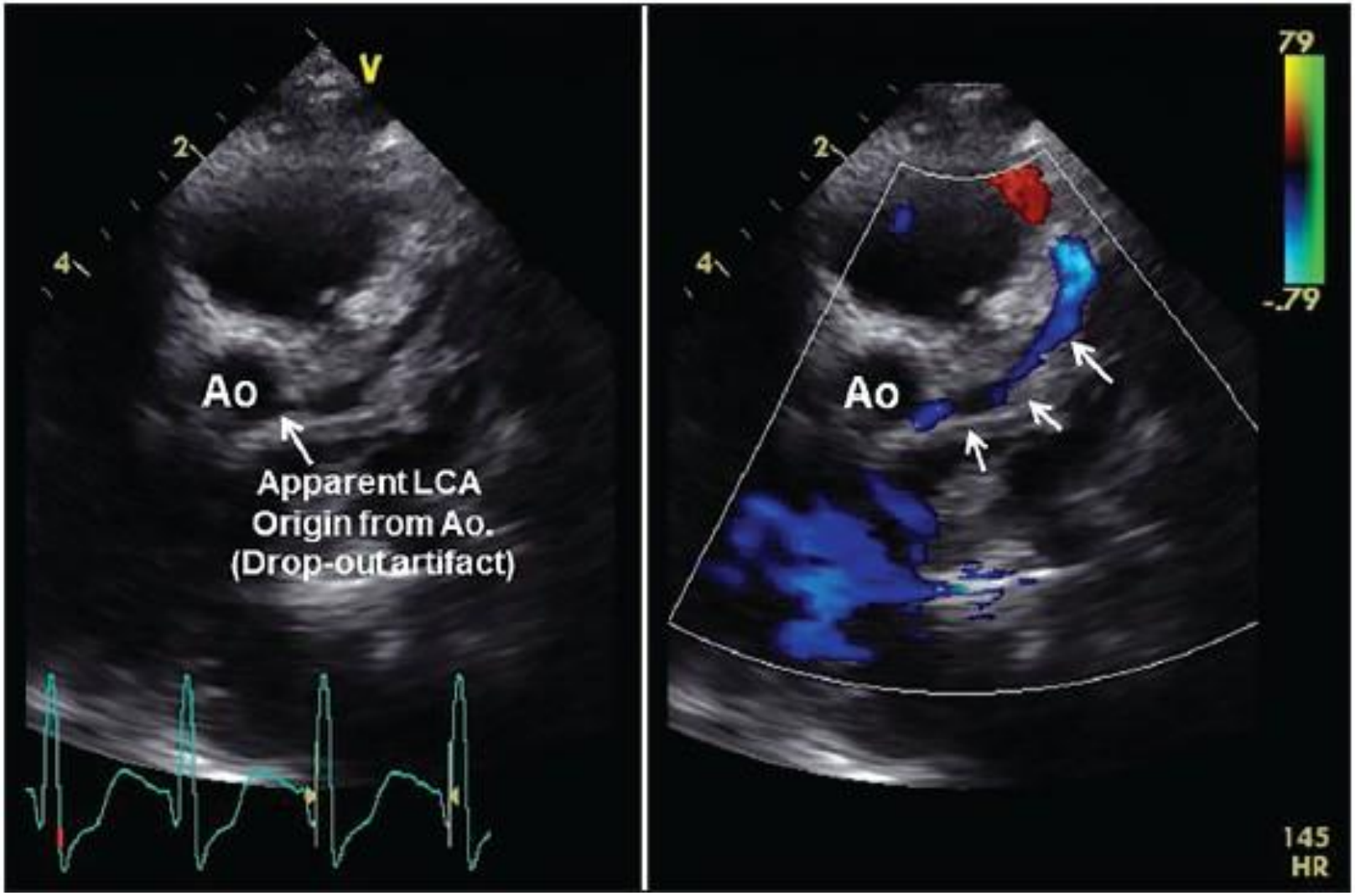


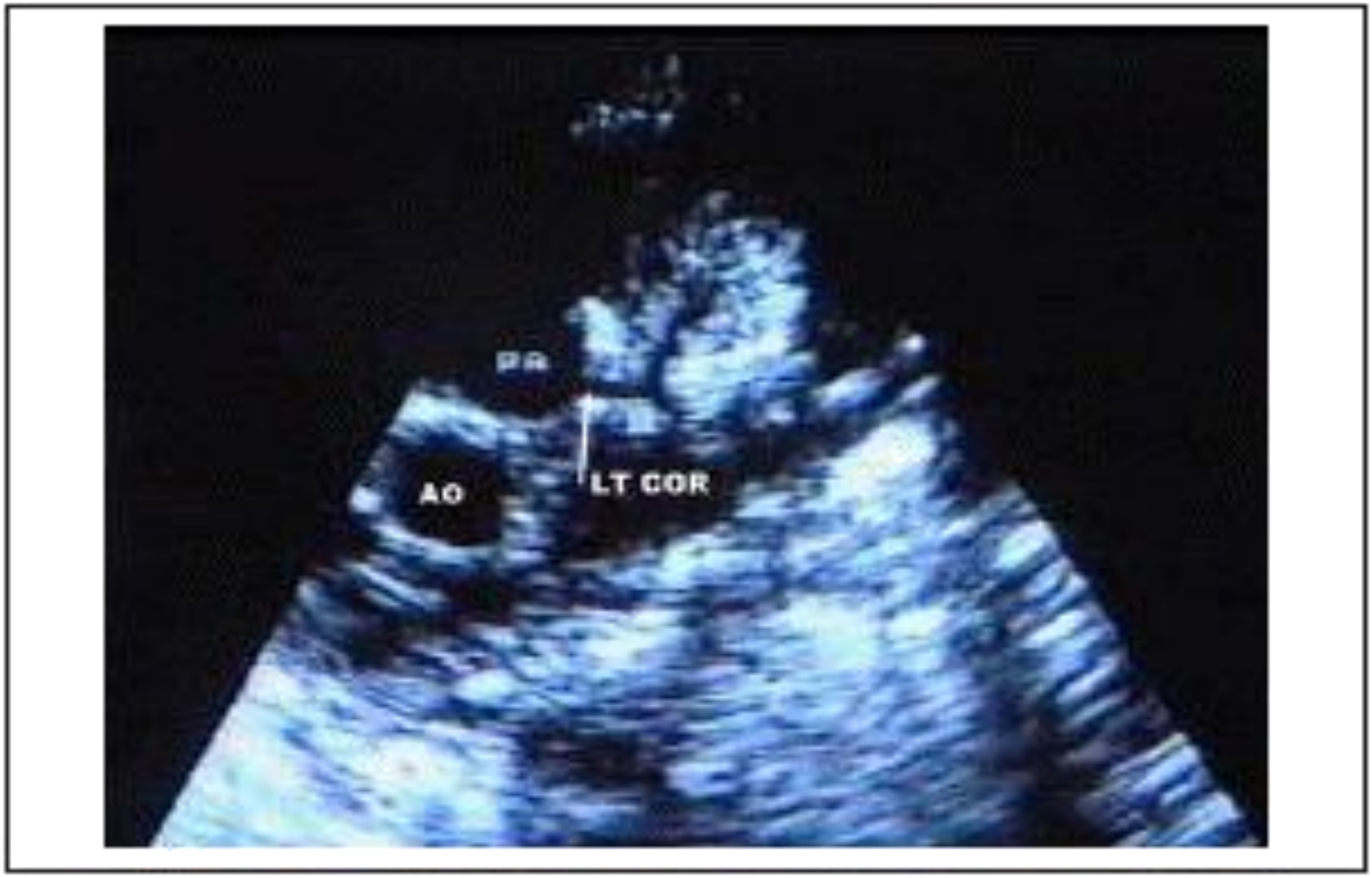
Q in DI,avL,V5-V6
T neg DI,avL,V5-V6

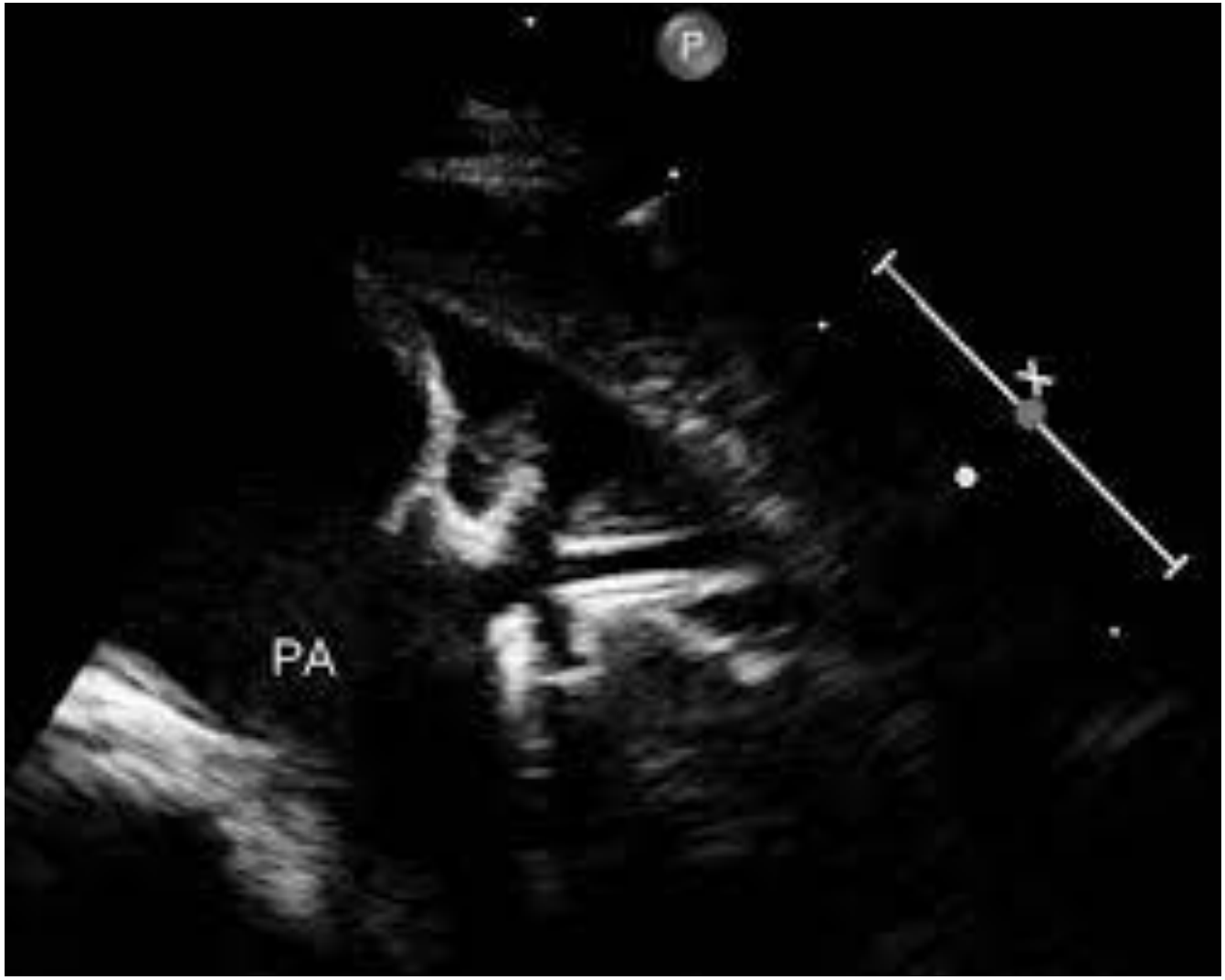


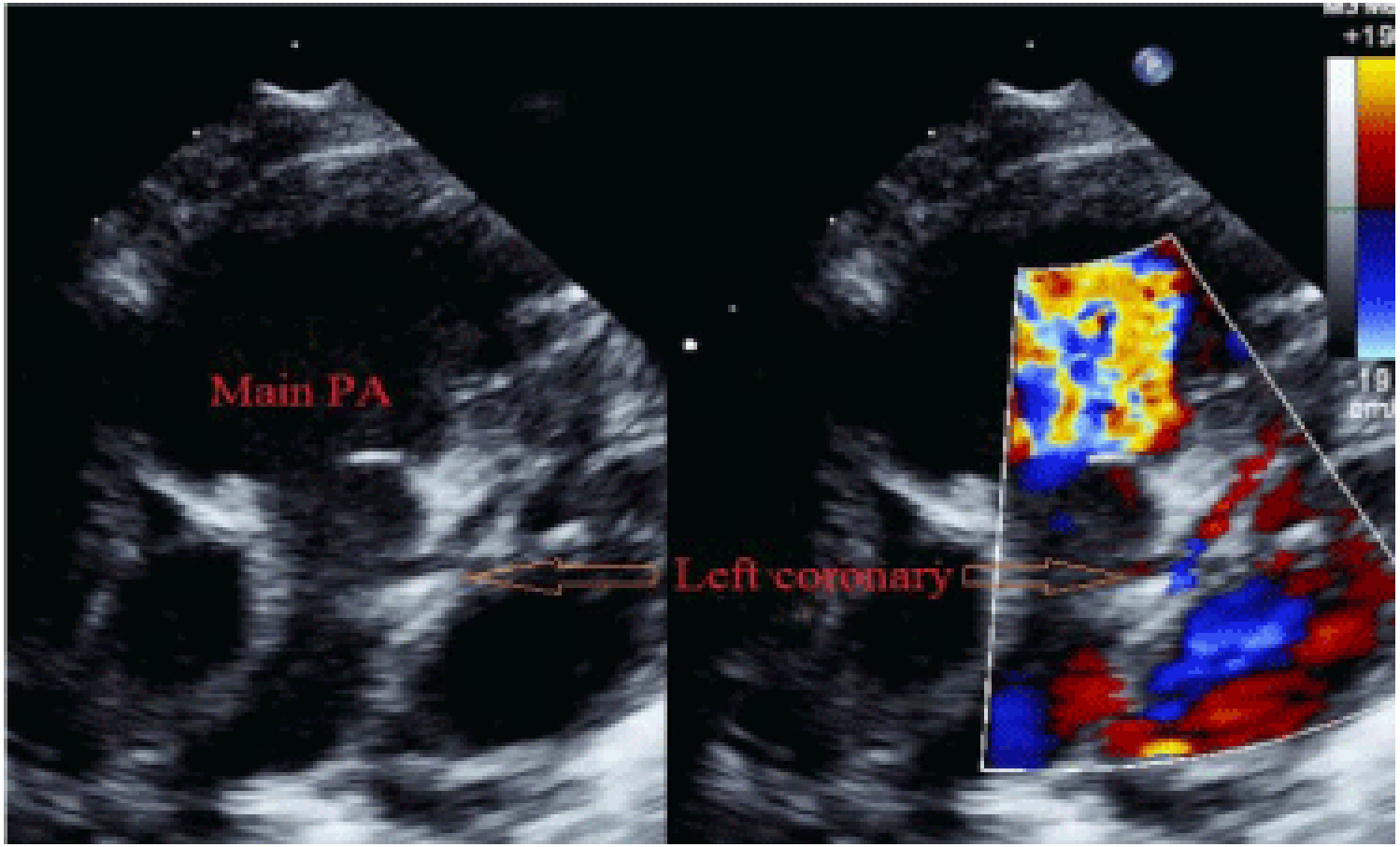
ALCAPA tipul adult

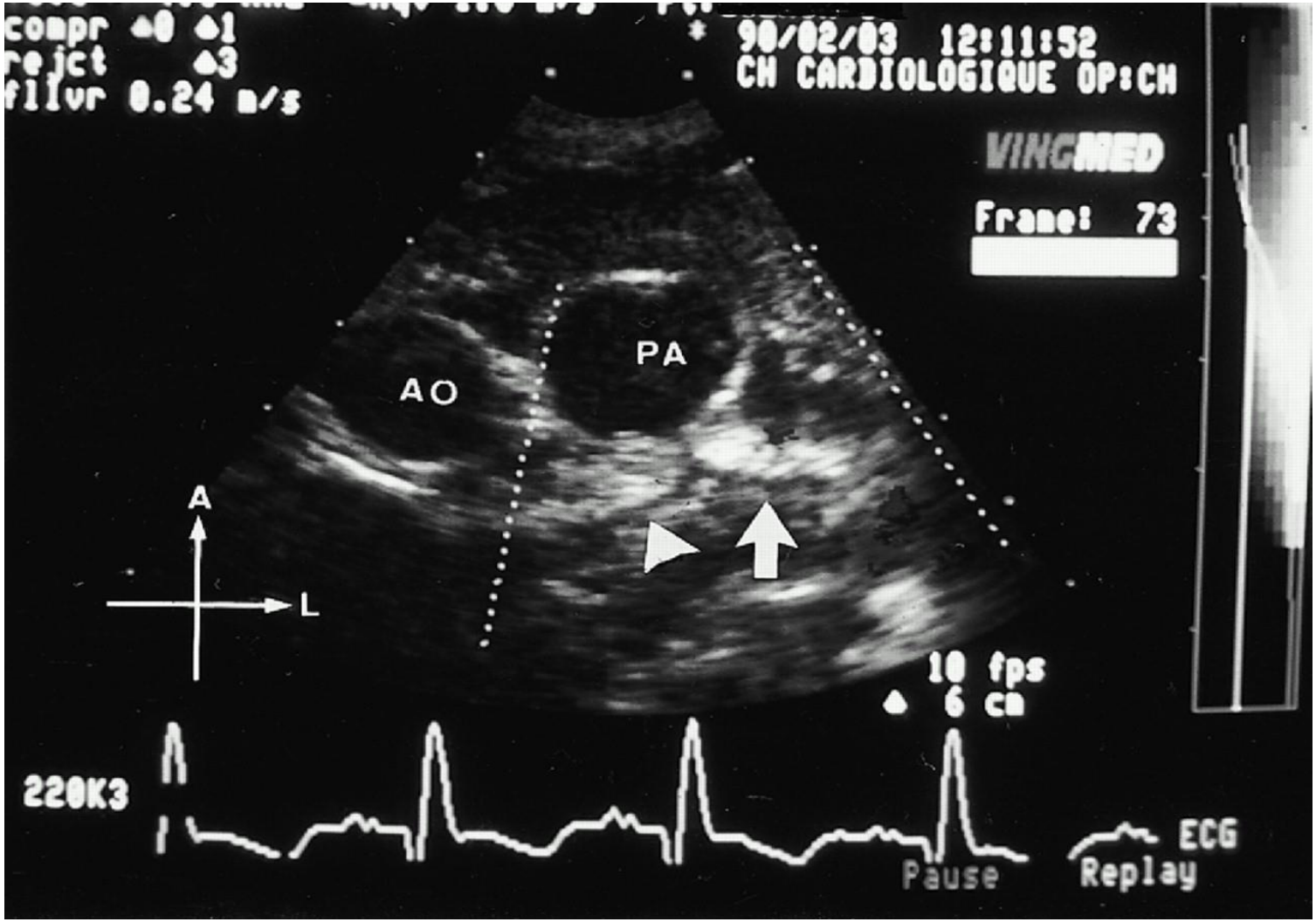
- Colateralele intre ACS si ACD reusesc sa aduca flux coronarian in VS insa:
- Nu si in zona subendocardica------ischemie-----aritmii maligne in evolutie

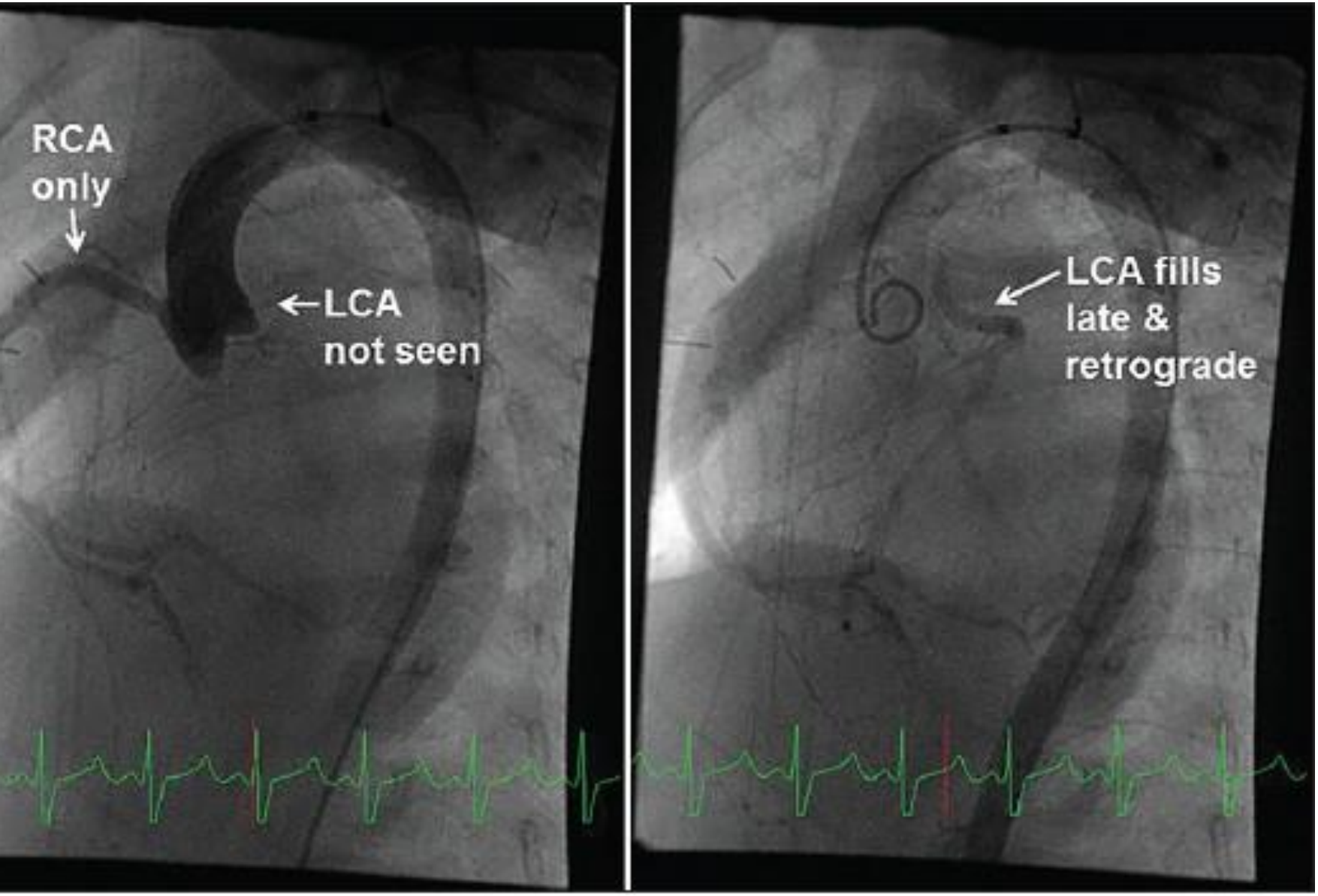

V
2
4
Ao
Apparent LCA
Origin from Ao.
(Drop-out artifact)
79
-.79
145
HR

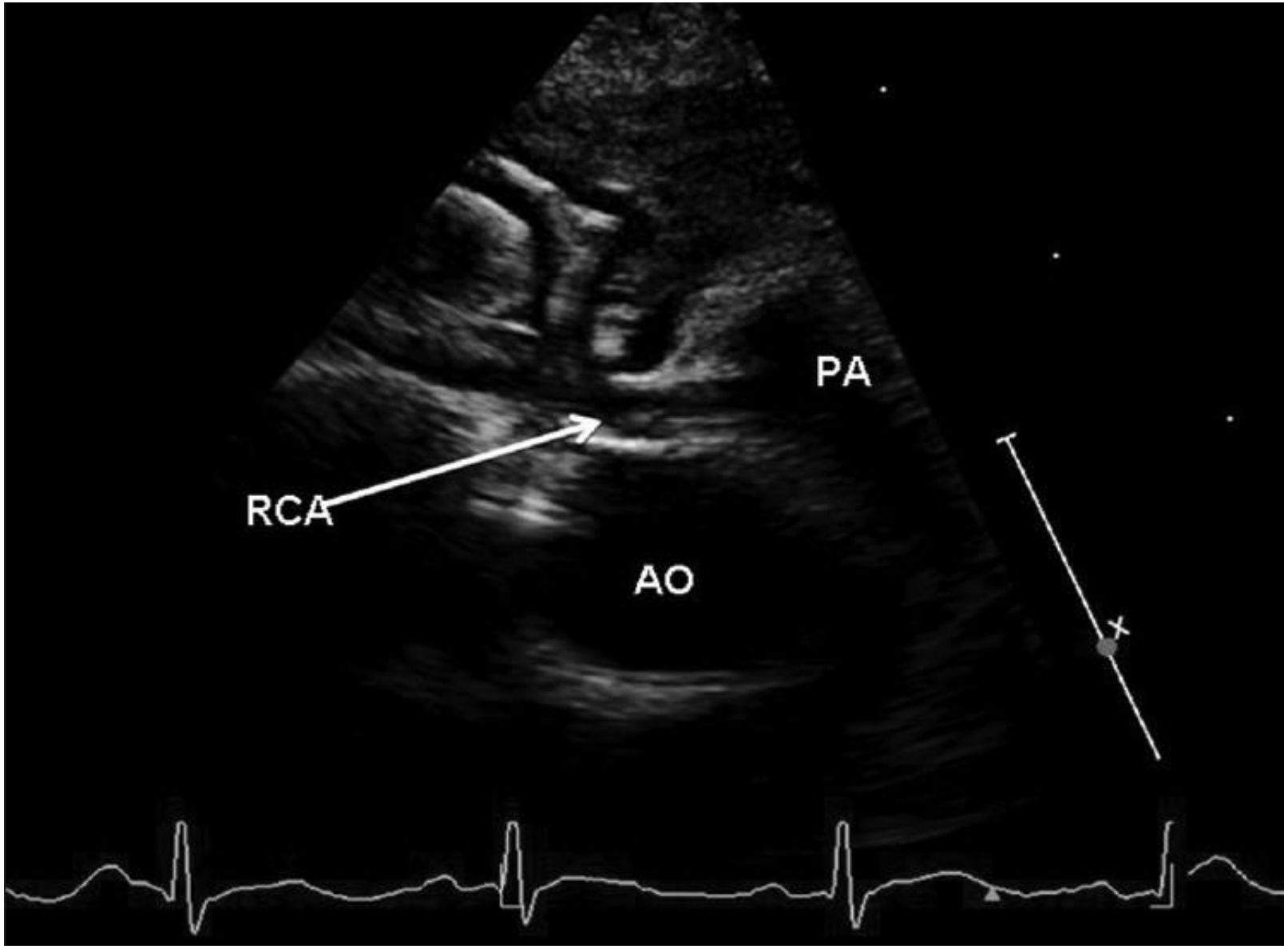

PA
AO
LT COR

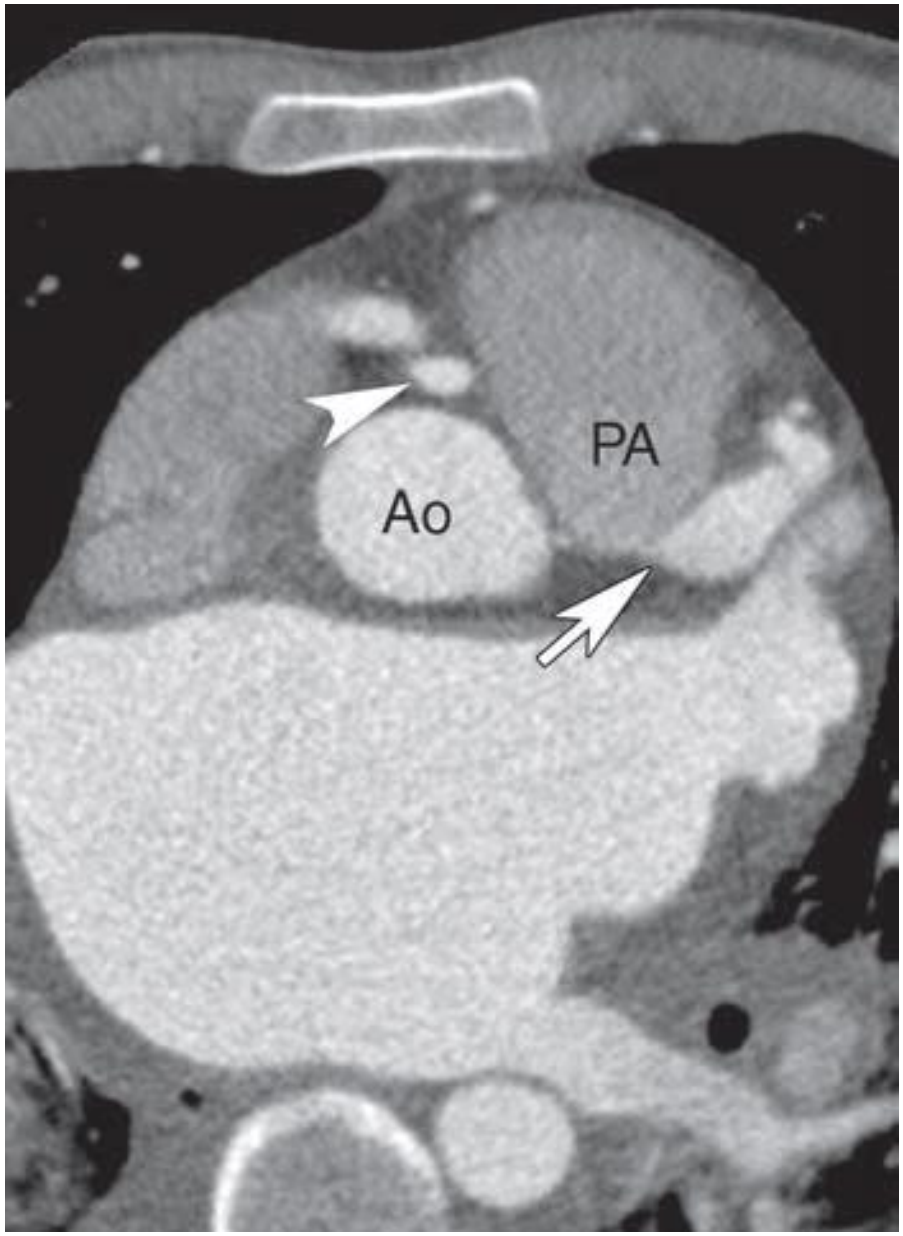

P
PA

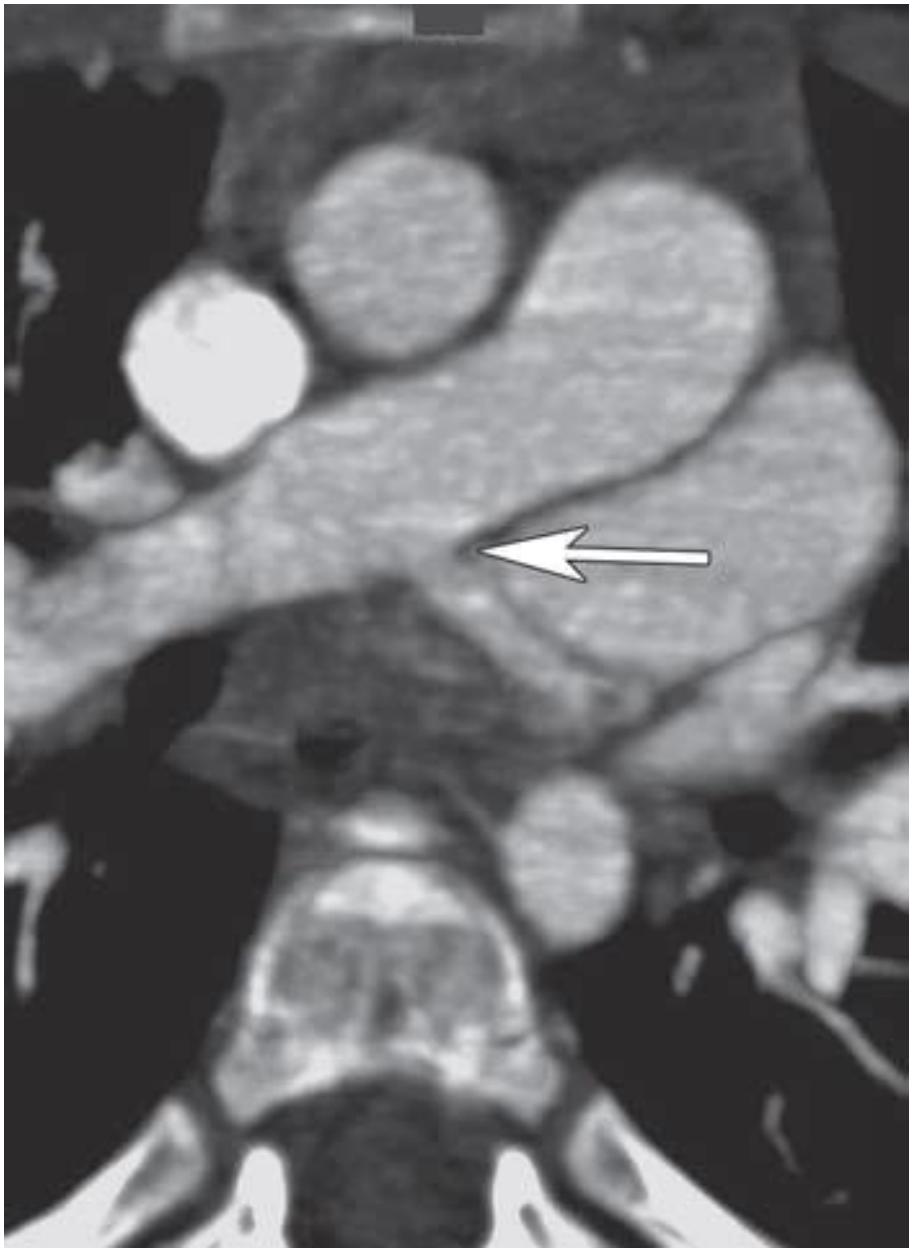

Main PA
Left coronary


compr ▲0 ▲1
reject ▲3
filvr 0.24 m/s
90/02/03 12:11:52
CH CARDIOLOGIQUE OP:CH
VINGMED
Frame: 73
PA
AO
A
L
10 fps
6 cm
220K3
ECG
Pause
Replay


RCA
only
LCA
not seen
LCA fills
late &
retrograde


PA
RCA
AO


PA
Ao



Management

Chirurgical

- Reimplantare ACS in Ao
- Tunelizare ACS in AO (cand distanta intre Ao si ACS este mare)
- Frecvent postoperator necesita ECMO

ALCAPA-concluzie

- Este una din cauzele importante de cardiomegalie si disfunctie ventriculara stanga la nn
- Necesita un diagnostic rapid