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"

Iulie 2015

MODUL TEORETIC

MCC CIANOGENE

Continut documentat/ validat/ prezentat de:

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- Expert formare medici: VEDUTA Alina

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

Cianoza

- Definitie: colorarea albastruie a pielii si mucoaselor datorita cresterii cc de Hb redusa>5g/100 ml sange

- Cianoza centrala: reducerea cc O2 in sangele arterial

- Cianoza periferica: creste extractia de O2 de catre tesutul periferic,cc O2 in sangele arterial este normala.

Apare in soc,hipovolemie,vasocontrictie la rece

- Box 11-1. CAUSES OF CYANOSIS
- Reduced arterial oxygen saturation (i.e., central cyanosis)
- Inadequate alveolar ventilation
- Central nervous system depression
- Inadequate ventilatory drive (e.g., obesity, pickwickian syndrome)
- Obstruction of the airway, congenital or acquired
- Structural changes in the lungs and/or ventilation-perfusion mismatch (e.g., pneumonia, cystic fibrosis, hyaline membrane disease, pulmonary edema, congestive heart failure)
- Weakness of the respiratory muscles
- Desaturated blood bypassing effective alveolar units
- **Intracardiac right-to-left shunt (i.e., cyanotic congenital heart defect)**
- Intrapulmonary shunt (e.g., pulmonary atrioventicular fistula, chronic hepatic disease resulting in multiple microvascular fistulas in the lungs)
- **Pulmonary hypertension with the resulting right-to-left shunt at the atrial, ventricular, or ductal levels (e.g., Eisenmenger's syndrome, persistent pulmonary hypertension of the newborn)**
- Increased deoxygenation in the capillaries (i.e., peripheral cyanosis)
- Circulatory shock
- Congestive heart failure
- Acrocyanosis of newborns
- Abnormal hemoglobin (unrelated to the degree of oxygenation)
- Methemoglobinemia (e.g., well water ingestion, aniline dye, congenital methemoglobinemia)
- Carbon monoxide poisoning

Detectia cianozei:

- Cea de origine cardiaca trebuie rapid diagnosticata pentru un management corect
- Nn normal:-acrocianoza

  -policitemie

PULSOXIMETRIE

PO2-GAZE SANGVINE

Hyperoxic test
cyanosed or not

- Pulse oximeter - not always reliable

"a random number generator"

- Rt radial ABG ***in air*** and ***after 5-10 min O2***

paO2 > 250mmHg -excludes CCHD

paO2 > 160 -CCHD unlikely

(UO TAPVR False negative !)

paO2 < 100 -CCHD likely *(usually lower)*

(severe Lung disease *(high paCo2)*, PPHN/PFC)

- "Radial ABG more useful than ECG or CXR in detection of cyanotic heart disease"

Warburton 1981

Consecintele si complicatiile cianozei

1.Policitemia

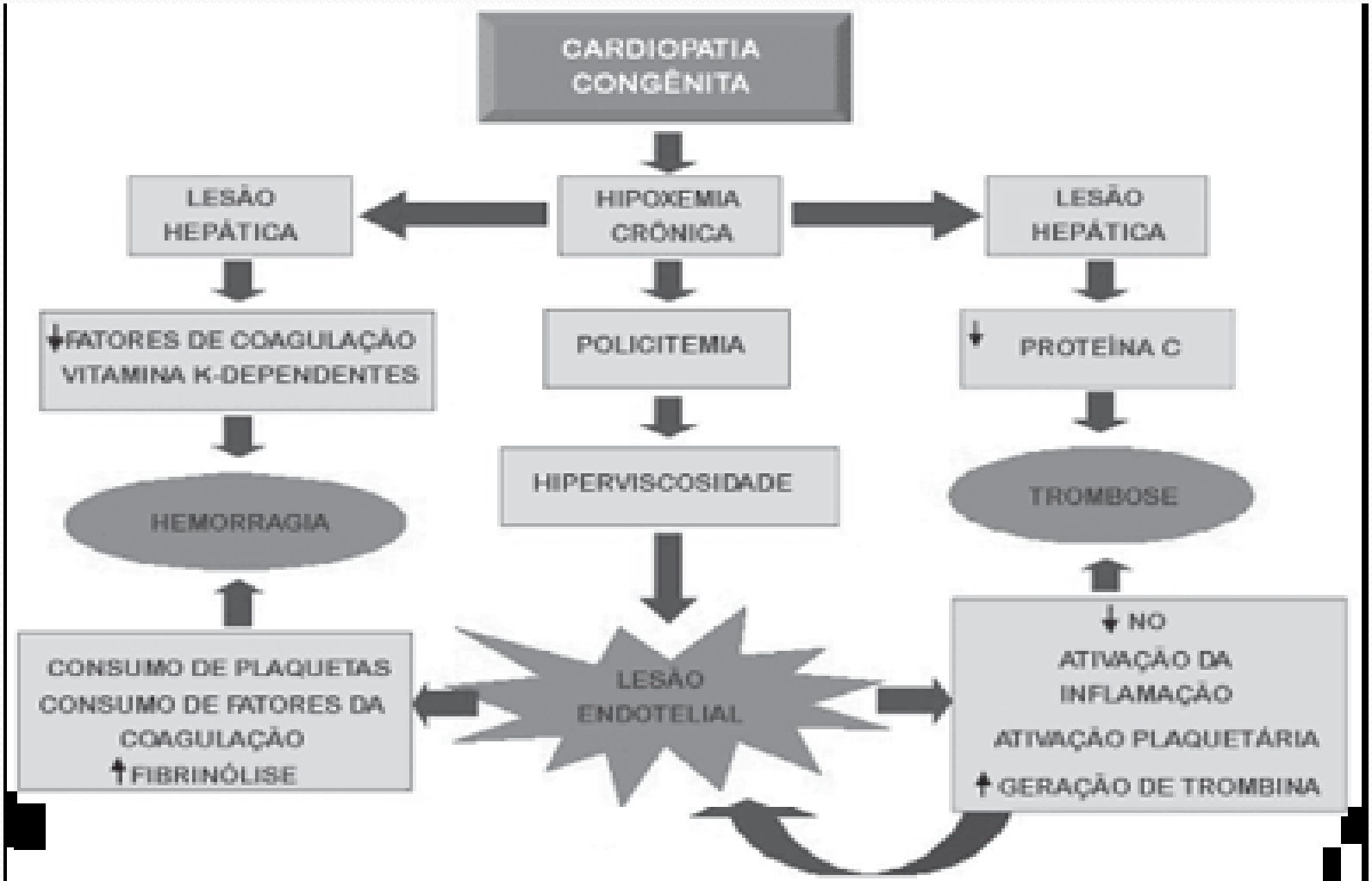


Figura 3. Mecanismos envolvidos na trombogênese e distúrbios hemorrágicos nas cardiopatias congênitas cianogênicas

2.Tulburari de coagulare

- Trombocitopenie
- Niveluri scazute de :Fg,fV,fVII
- Petesii,gingivoragii,epistaxis

3.Hipocratism digital

- Mecanism:megacariocitele patrund in circ sistemica si la nivelul capilarelor degetelor si datorita incetinirii fluxului descardca factori de crestere
- Apare dupa varsta de 6 l
- Poate exista si in

-boli hepatice

-endocardita subacuta

-ereditar

Cianoza (coloratia albastra) buzelor si a extremitatiilor

Degete hipocratice

4.Crize hipoxice

- -perioada de plans extrem de agitat
- -hiperpnee
- -accentuarea cianozei
- -uneori convulsii
- -in faza extrema –deces
- **squatting**

5.Consecinte SNC

- Abces cerebral:febra,cefalee,focar neurologic

-cresterea viscozitatii-microinfarcte-colonizare bacteriana

- AVC

-emboli: cavitati cardiace/sistem venos

Differential Diagnosis of Cyanosis in the Neonate

- **Primary cardiac lesions**
- **Decreased pulmonary blood flow, intracardiac right-to-left shunt**
- Critical pulmonary stenosis
- Tricuspid atresia
- Pulmonary atresia/intact ventricular septum
- Tetralogy of Fallot
- Ebstein anomaly
- Total anomalous pulmonary venous connection with obstruction
- **Normal or increased pulmonary blood flow, intracardiac mixing**
- Hypoplastic left heart syndrome
- Transposition of the great arteries
- Truncus arteriosus
- Complete common atrioventricular canal
- Total anomalous pulmonary venous connection without obstruction
- Other single-ventricle complexes

MCC cianogene cu debit pulmonar scazut sunt intracardiac dreapta-stanga

- TETRALOGIA FALLOT
- ATREZIA PULMONARA
- ATREZIA DE VALVA TRICUSPA
- BOALA EBSTEIN

TETRALOGIA FALLOT

Tetralogy of Fallot

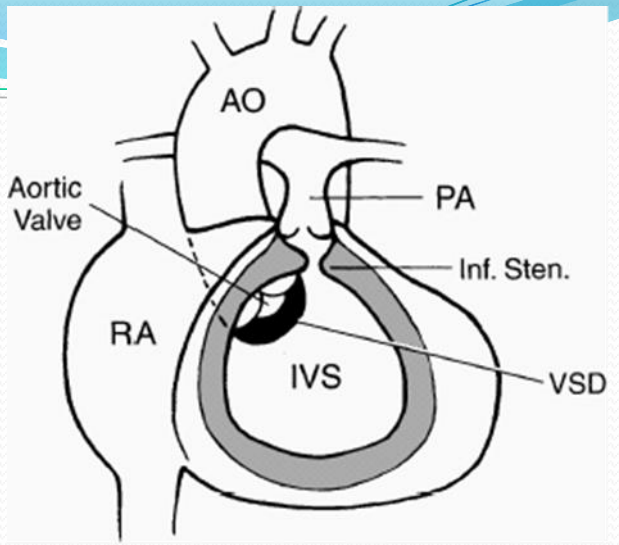
- Etienne- Louis Arthur Fallot made the first published bedside diagnosis that was proven at post-mortem in 1888 and called the condition " maladie bleue"

www.escardio.org/EAE

EUROPEAN ASSOCIATION OF Echocardiography

EUROPEAN SOCIETY OF CARDIOLOGY

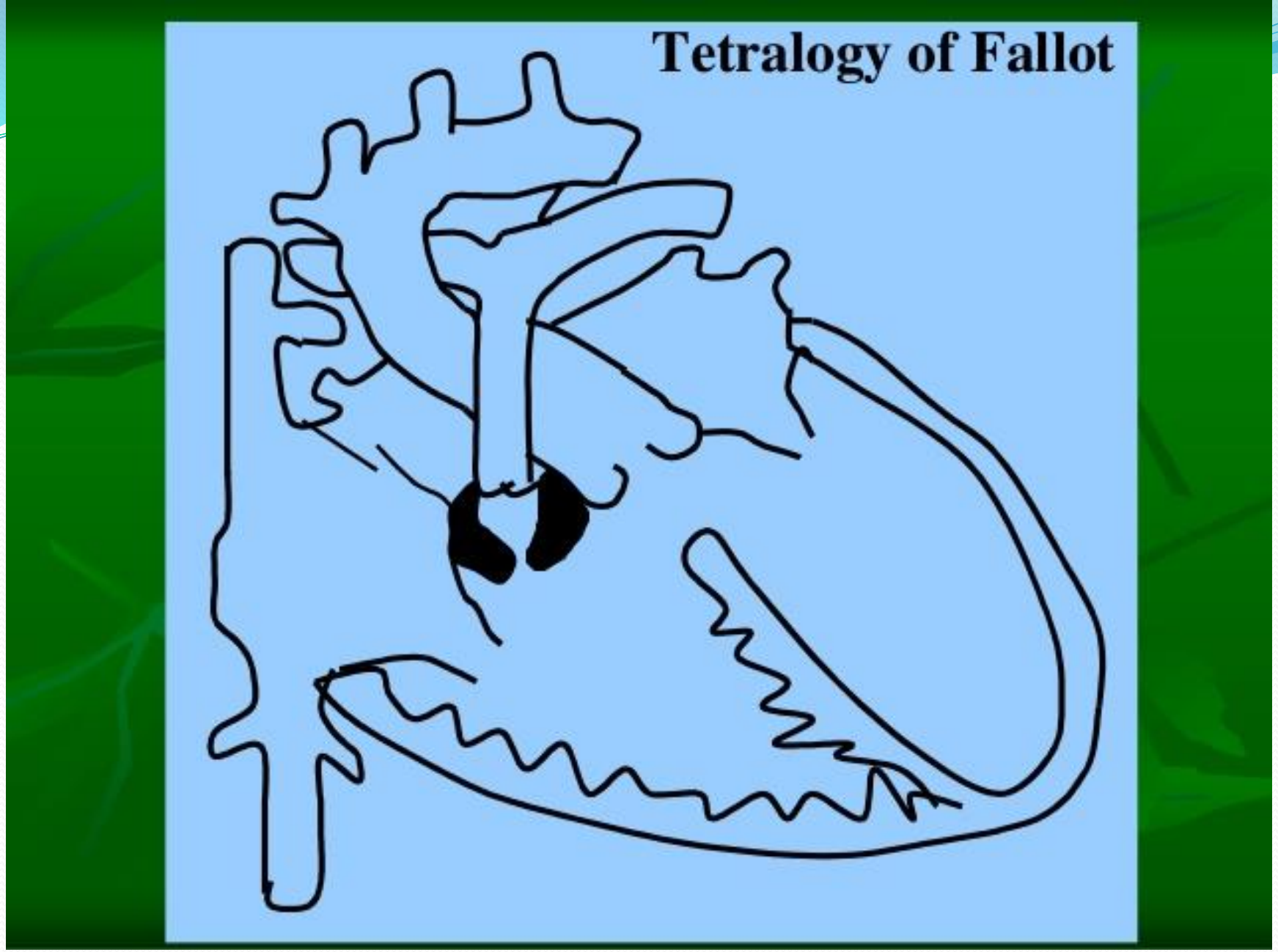
Tetralogia Fallot



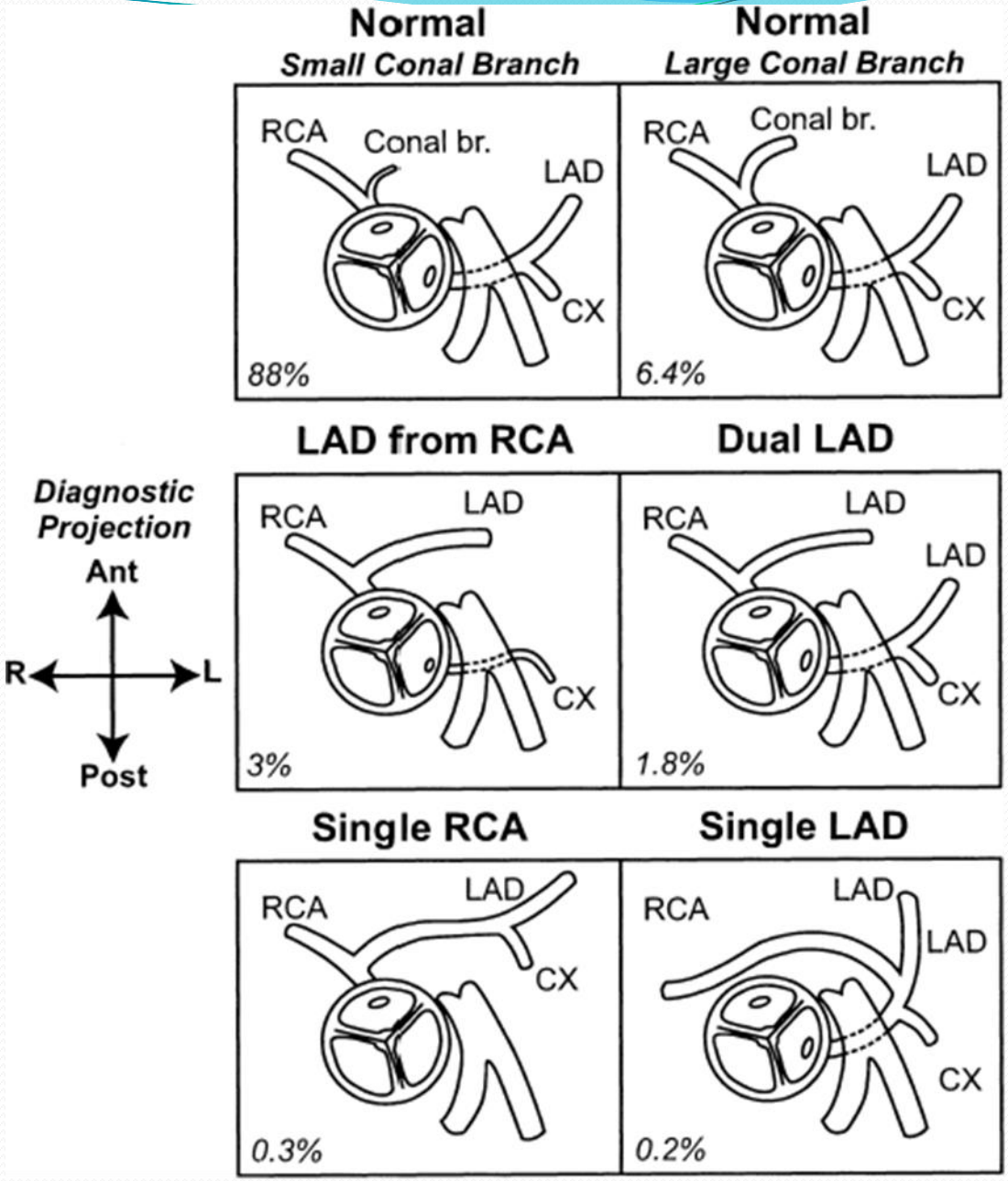
- 10 % din MCC
- Cea mai frecventa MCC cianogena
  - DSV larg: perimembranos cu extindere in zona subpulmonara
  - Obstructie in CEVD:
    - -45% infundibular, 10% valvular, impreuna 30%,15% atrezie VP
  - Hipertrofie VD
  - Ao calare pe SIV

- Functional doar 2 elemente sunt necesare

-DSV larg
-obstructie in CEVD
-HVD e sec., iar Ao poate fi dextropusa variabil


Tetralogy of Fallot

- Anomalii coronariene



Variante si leziuni asociate

- **Dubla cale de iesire VD**
- **TF cu CAVC**
  - Cel mai frecvent Rastelli C

- **TF cu absenta VP**
  - Exista valva rudimentara realizand stenoza si insuficienta severa
  - De obicei se asociaza cu absenta CAP
  - Dilatare de ramuri AP cu compresia bronsiilor si arteriopatie pulmonara intraparenchimatoasa
- **Anomalii de arc aortic**
  - Arc aortic drept -25 %
  - Dublu arc aortic

Variante si leziuni asociate

- **TF cu atrezie pulmonara si/sau hipoplazie severa pulmonara**

➢ Atrezie VP: short segment atrezia

➢ Atrezie AP: long segment atrezia

A fost clasificat oarecum gresit ca si Tunchi tip IV (Collet si Edwards).De asemenea descris ca si Pseudotruncus

Comitetul de Nomenclatura al Societatii de Chirurgie a malformatiilor cardiace clasifica aceasta asociere sub umbrela : Atrezie pulmonara cu DSV

TF cu atrezie pulmonara si/sau hipoplazie severa pulmonara

- Exista 3 categorii

A.

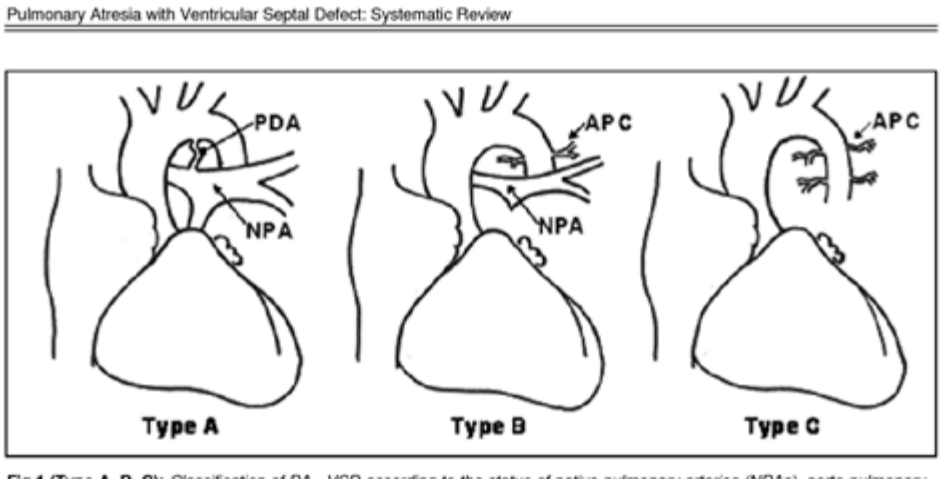
1. intreruperea fluxului pulmonar anterograd la nivelul VP/AP
2. exista ramuri AP confluente
3. CAP

B.

1. De cele mai multe ori atrezie AP
2. Exista ramuri pulmonare si colaterale aortopulmonare/CAP cu varsare in ramuri AP,care iriga ambii lobi pulmonari
3. Poate exista discontinuitate de ramuri AP

C.

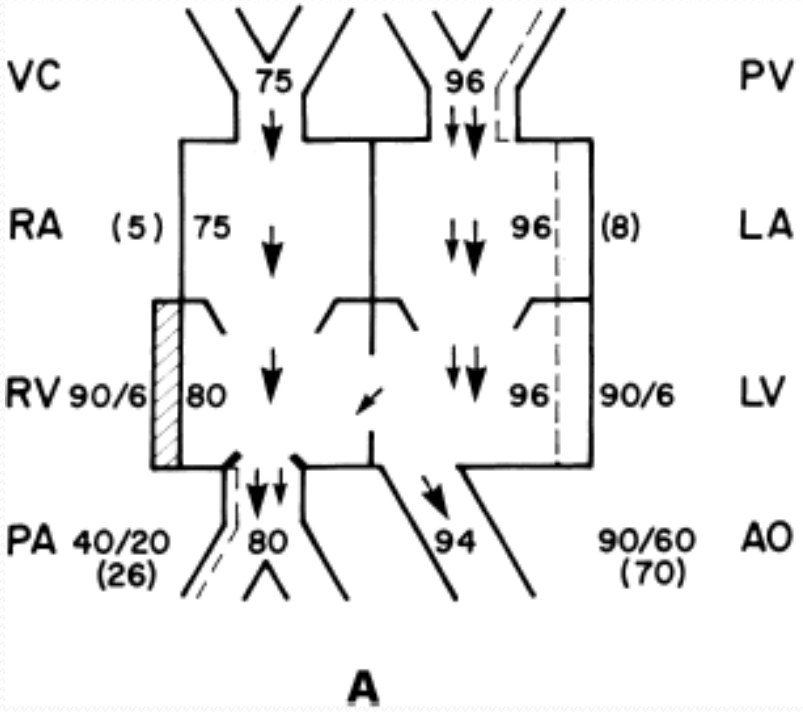
1. Nu exista ramuri AP.
2. Colateralele aortopulmonare iriga direct ambii lobi pulmonari

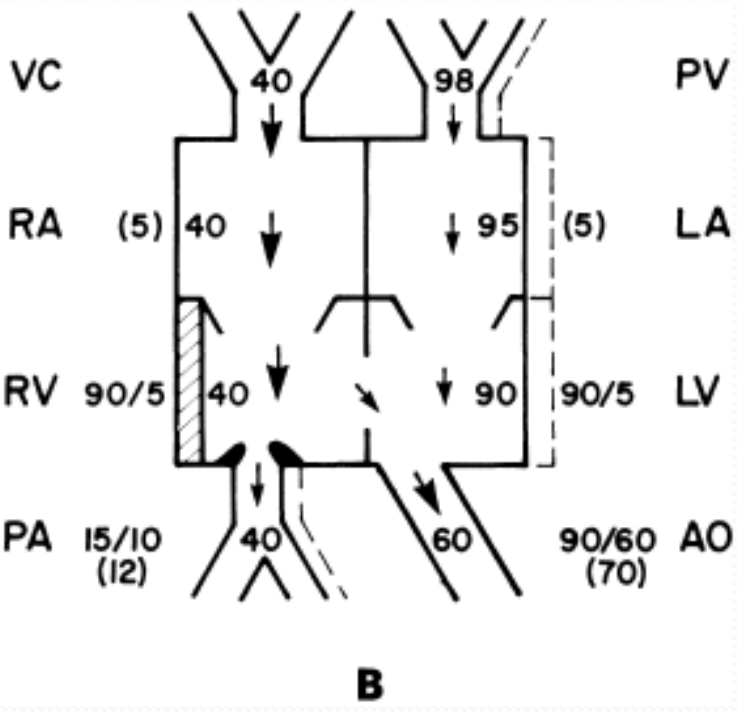


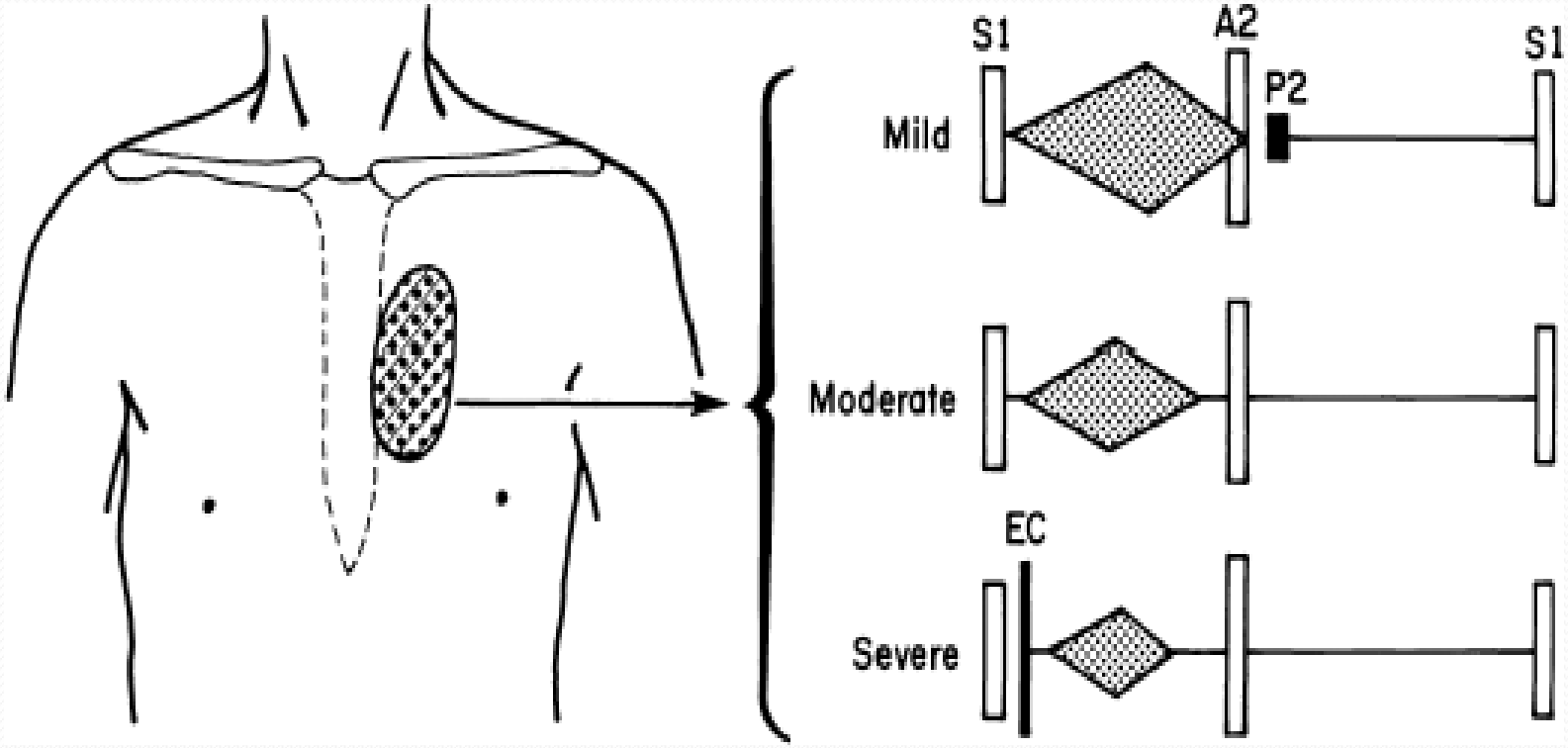
Manifestari clinice

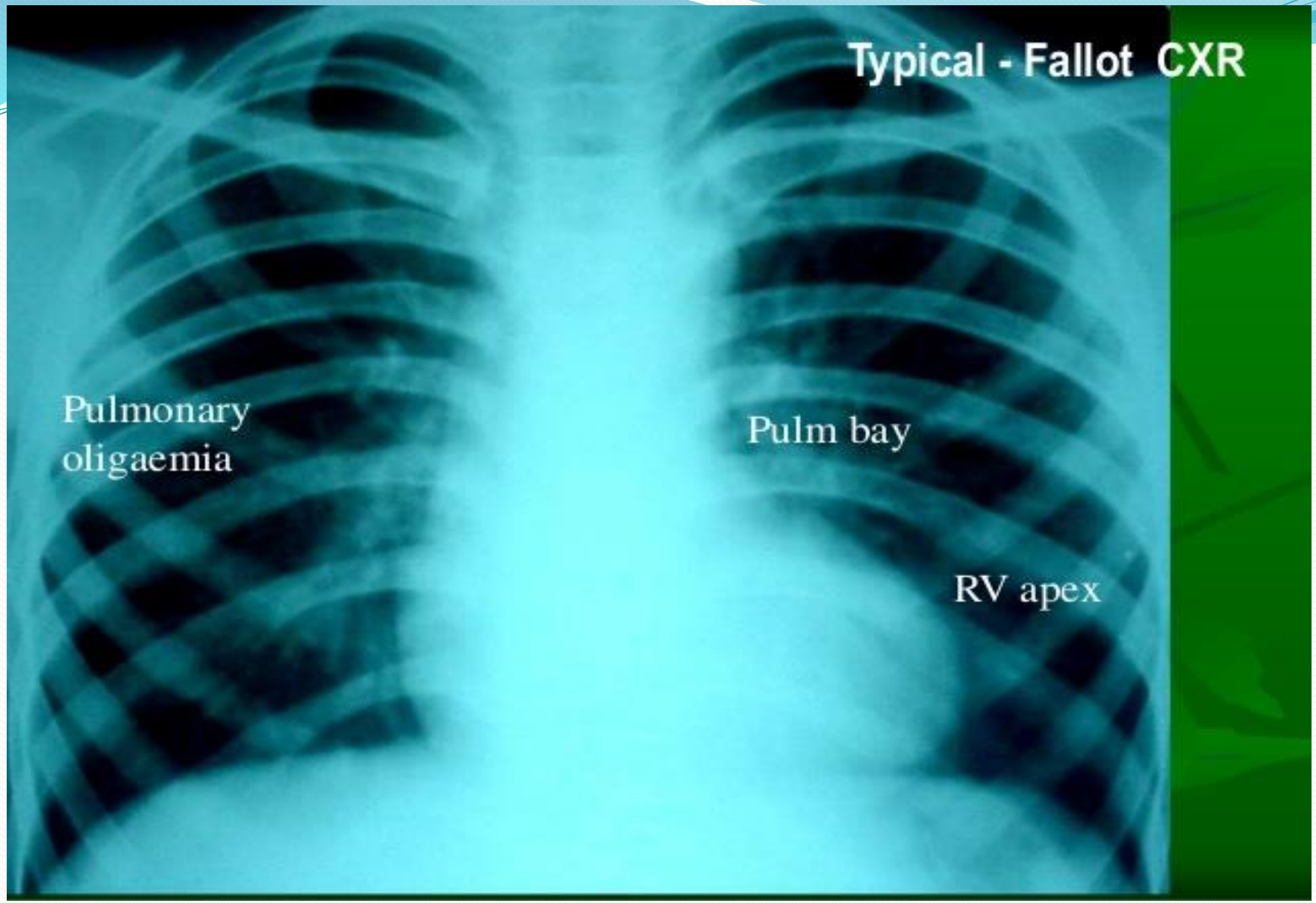
- Suflul se aude de la nastere: este dat de stenoza pulmonara (DSV este larg nerestrictiv)
- Cianoza apare de la nastere sau la scurt timp dupa aceea
- Dispneea la efort,squatting,crizele hipoxice apar mai tarziu in evolutie
- Exista forma de TF acianotica-pink Fallot care poate prezenta semne de ICC secundara unui sunt larg stg-dpt

Pink fallot / cyanotic FAllot

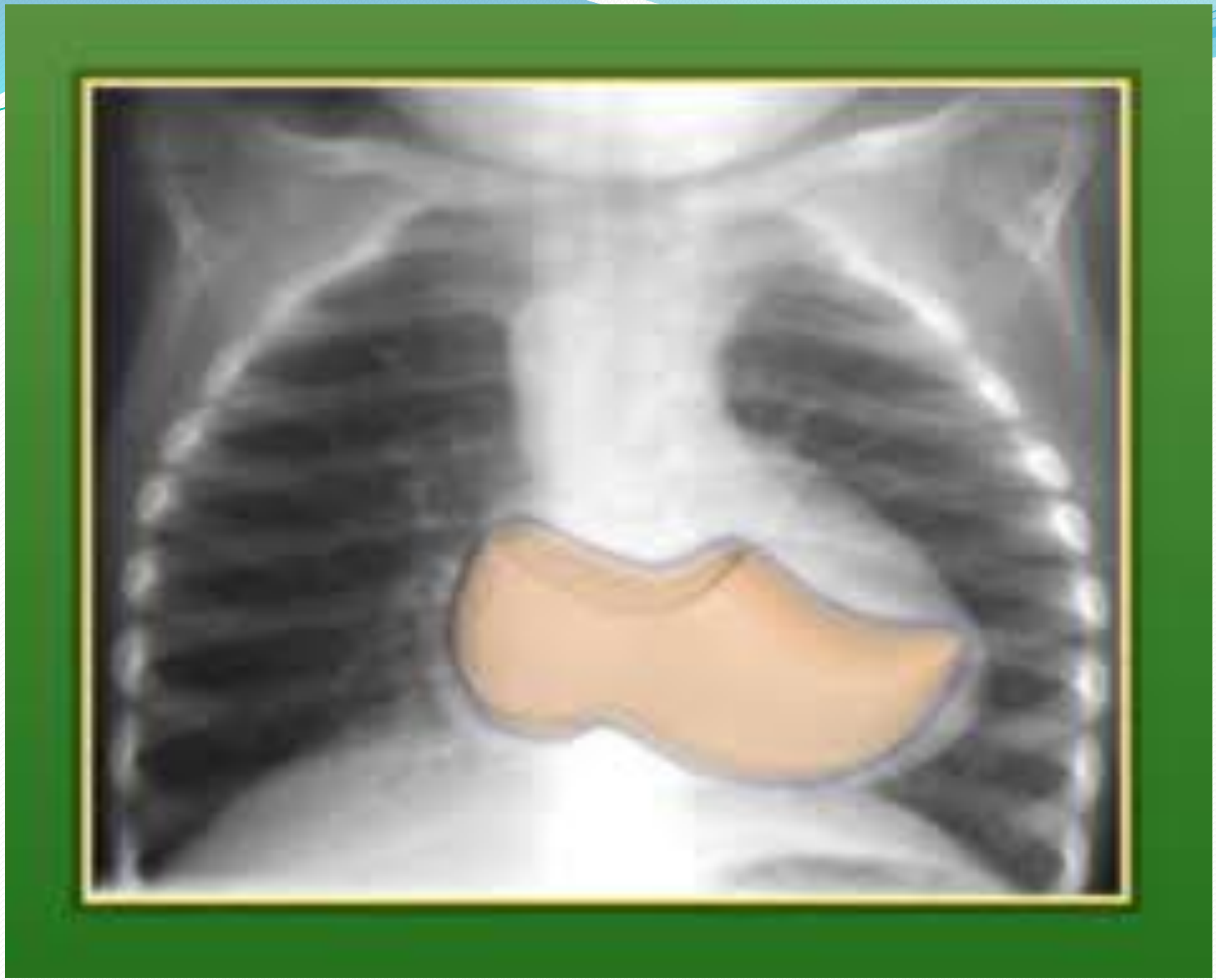




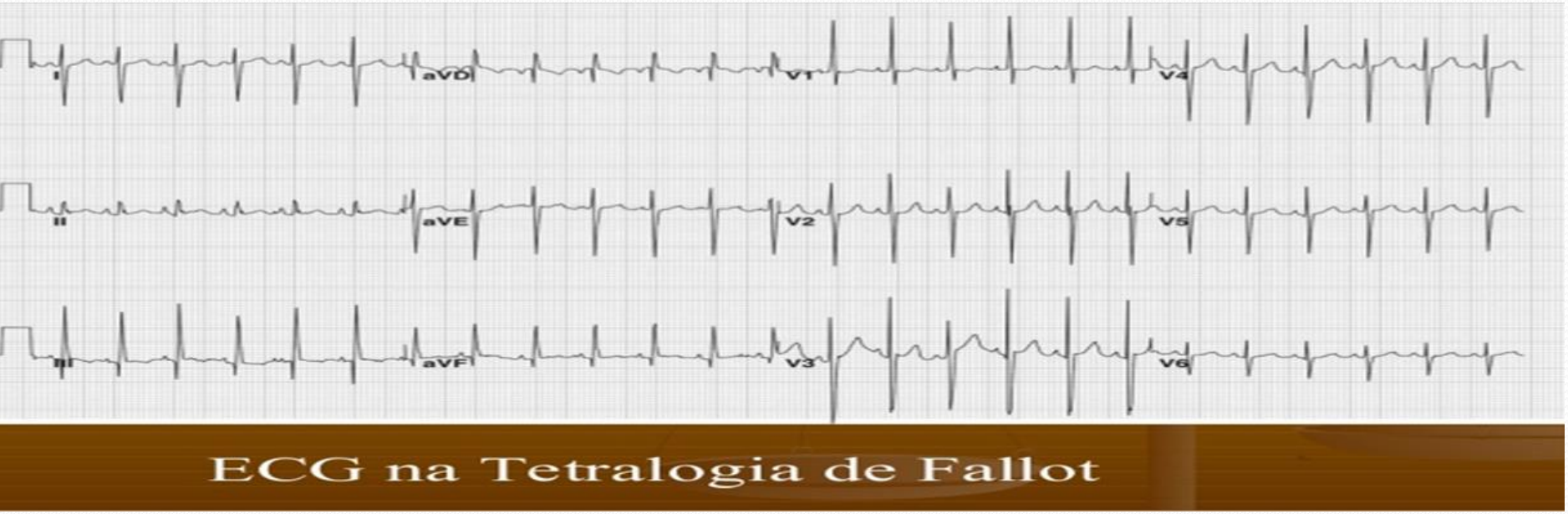

S1
A2
S1
P2
Mild
Moderate
EC
Severe



Rx

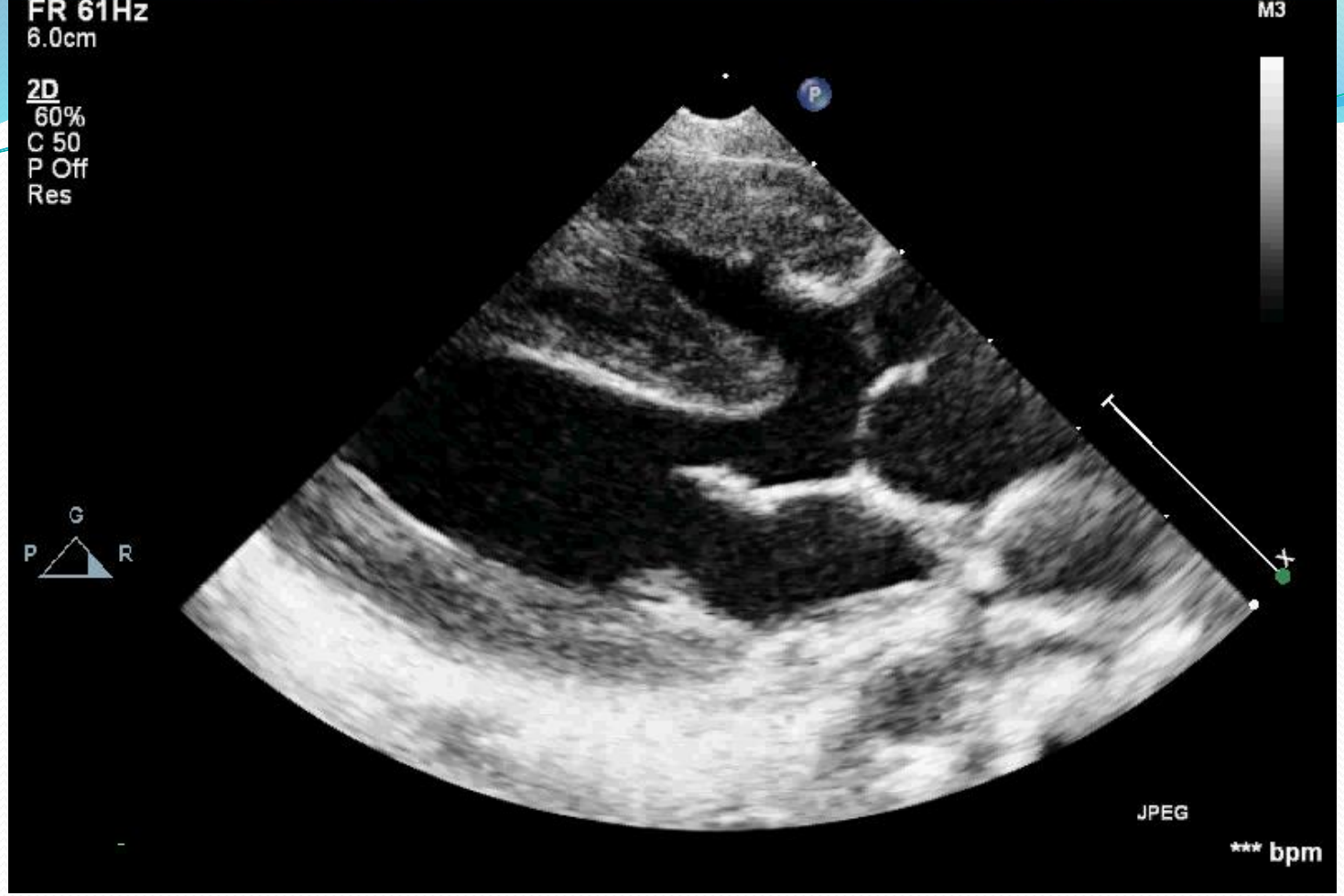


ECG


I
aVD
V1
V4
II
aVE
V2
V5
III
aVF
V3
V6
ECG na Tetralogia de Fallot

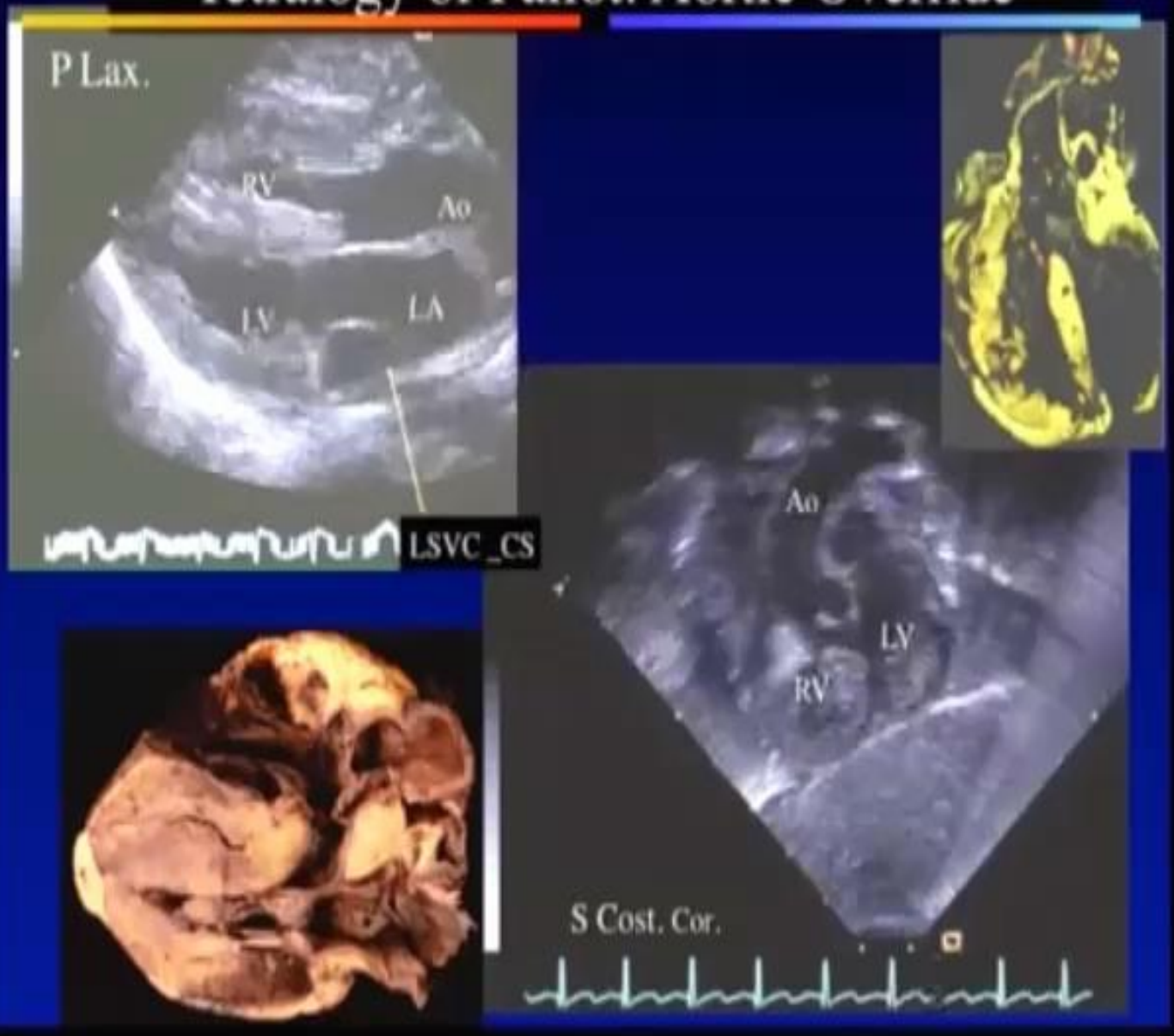
Ecografia

- Pozitia cordului,situs atrial,conex venoase,concordanta A-V
- Evaluarea SIA
- Evaluarea valvelor AV
- DSV: malaliniere/inlet (AV canal)/doubly committed
  existenta DSV musculare
  restrictiv/nonrestrictiv;
  directia fluxului prin DSV
- Gradul si morfologia obstructiei in CEVD
- Inelul VP si morfologia VP
- Dimensiunile AP si ramuri
- Evaluarea Vao
- CAP/MAPCA
- Evaluarea arcului aortic
- Evaluarea arterelor coronare

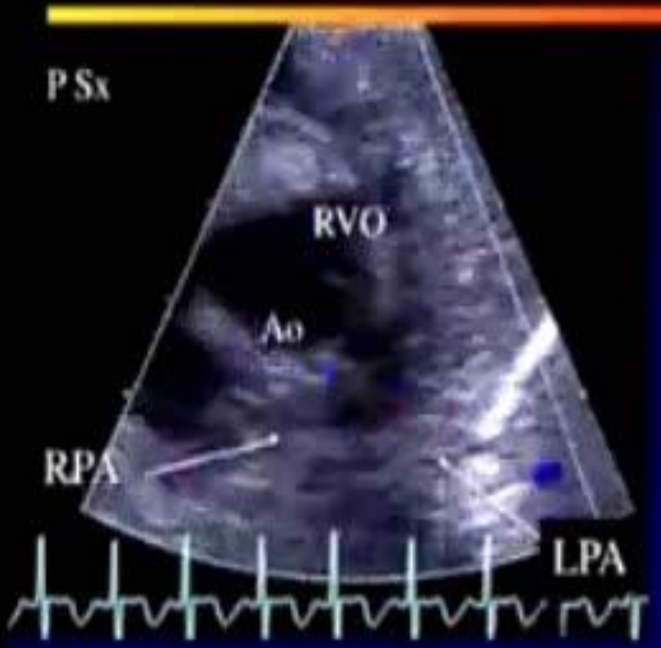


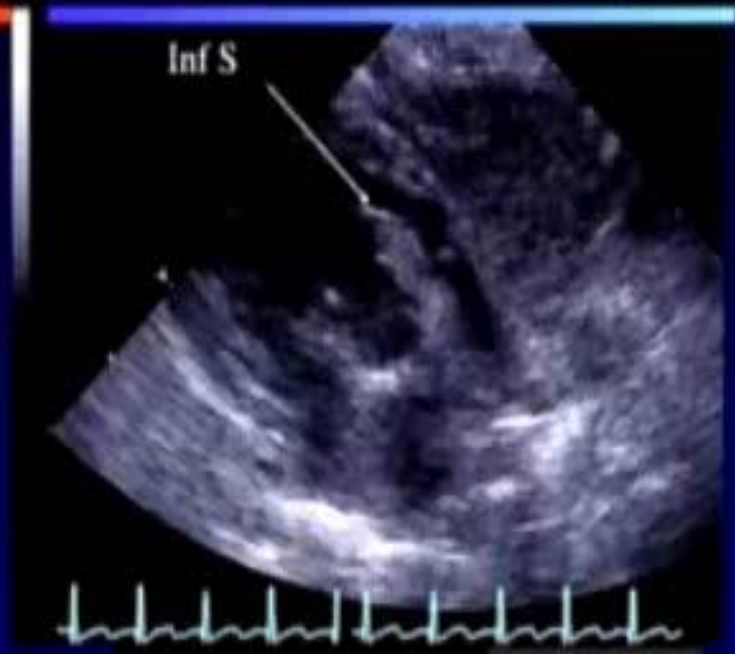
ecografia

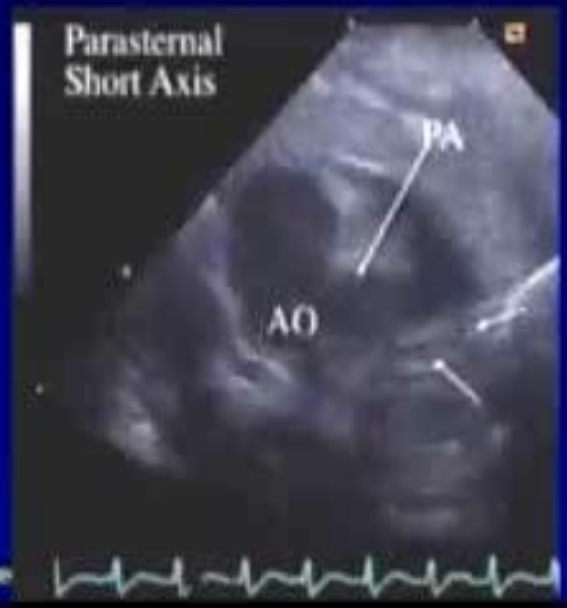
Tetralogy of Fallot: Aortic Override

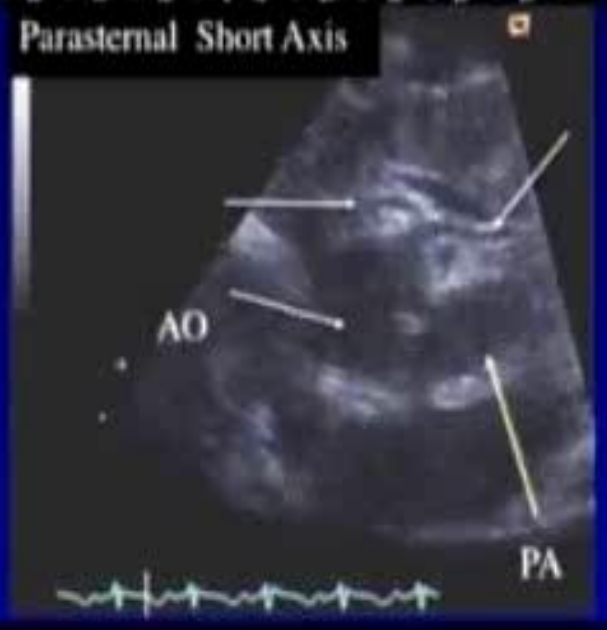


Short Axis View: The Monology of Stensen & Coronary Arts

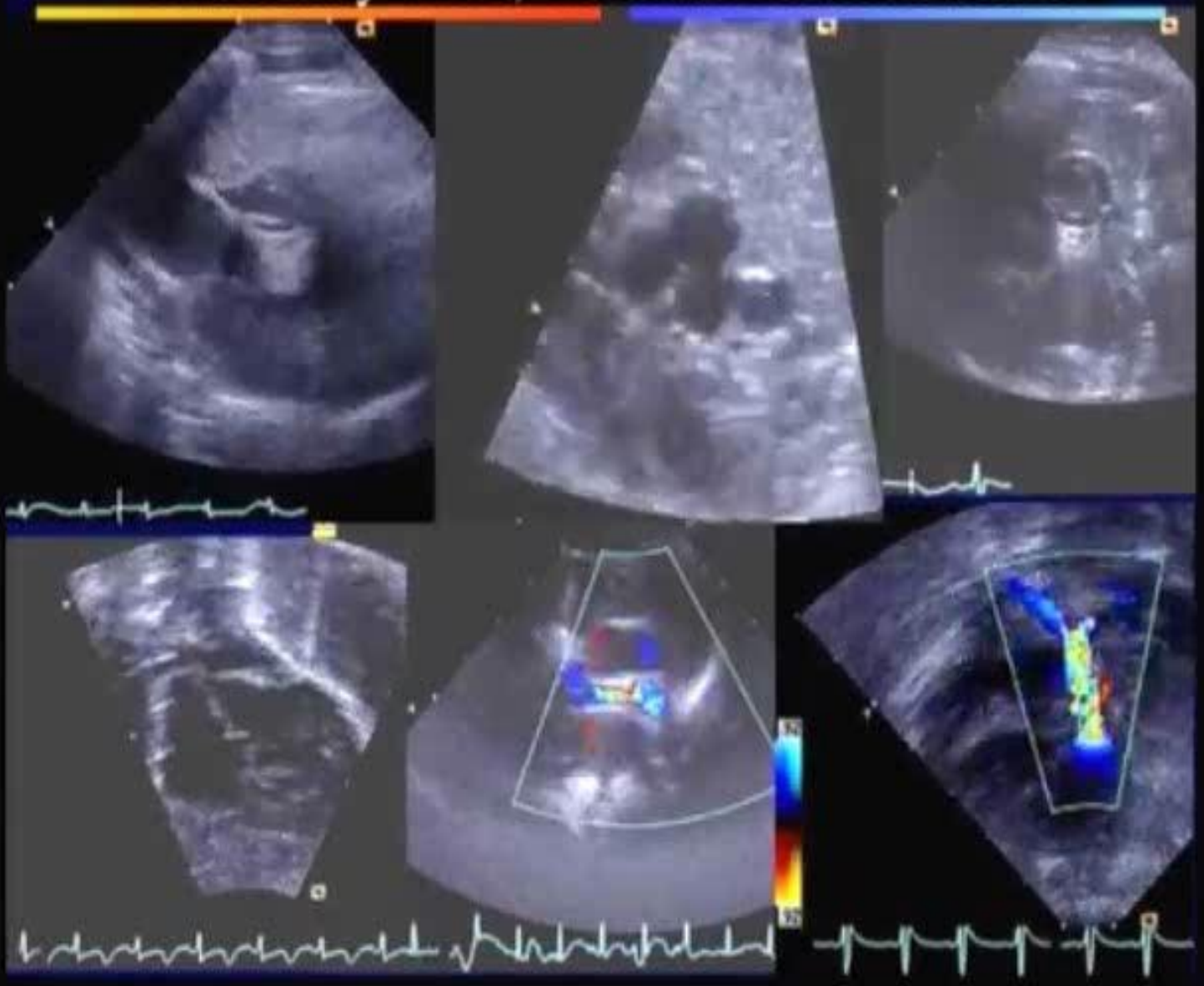




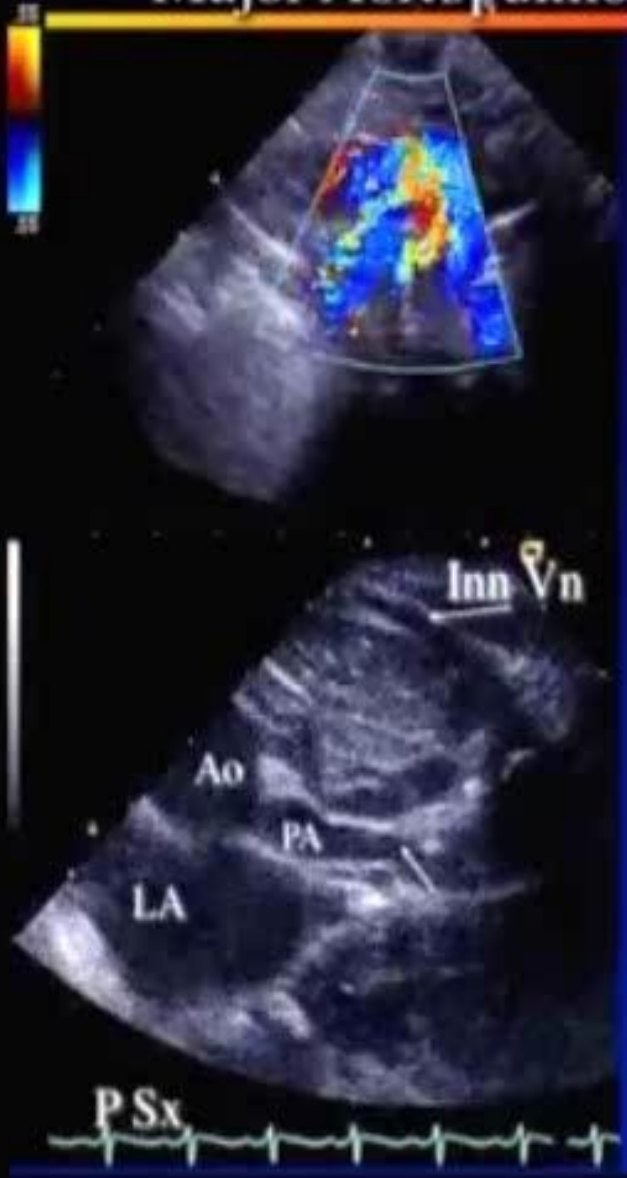


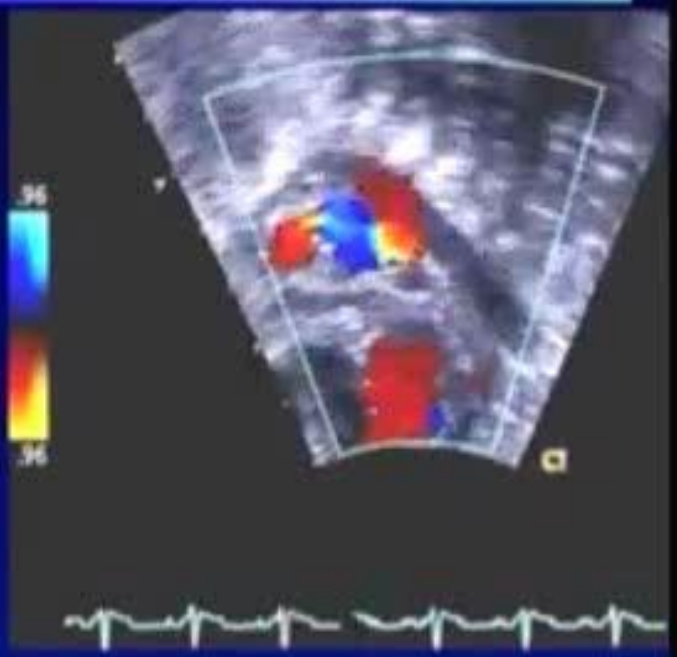


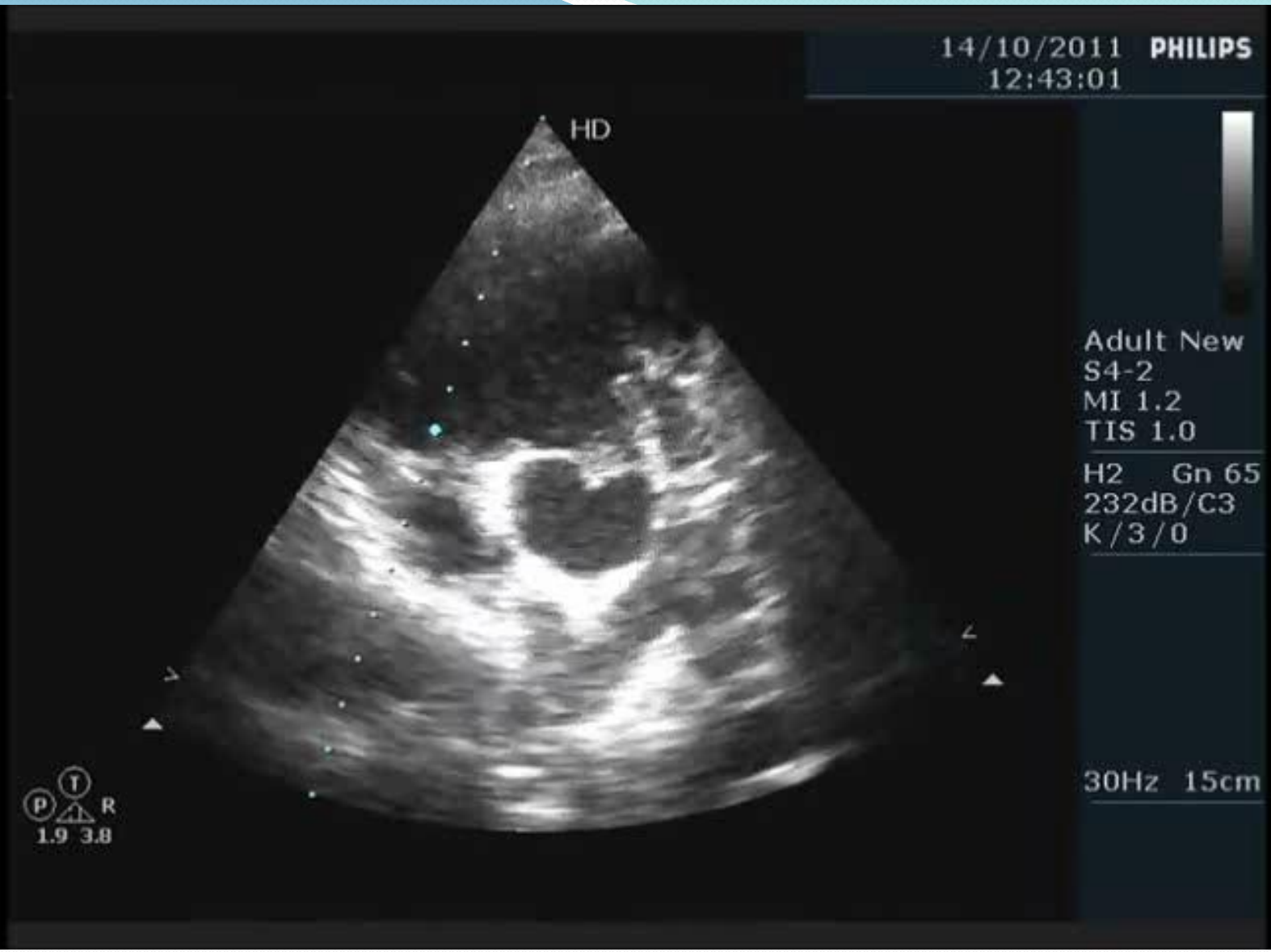
Pulmonary Valve, Main and Branch Arteries

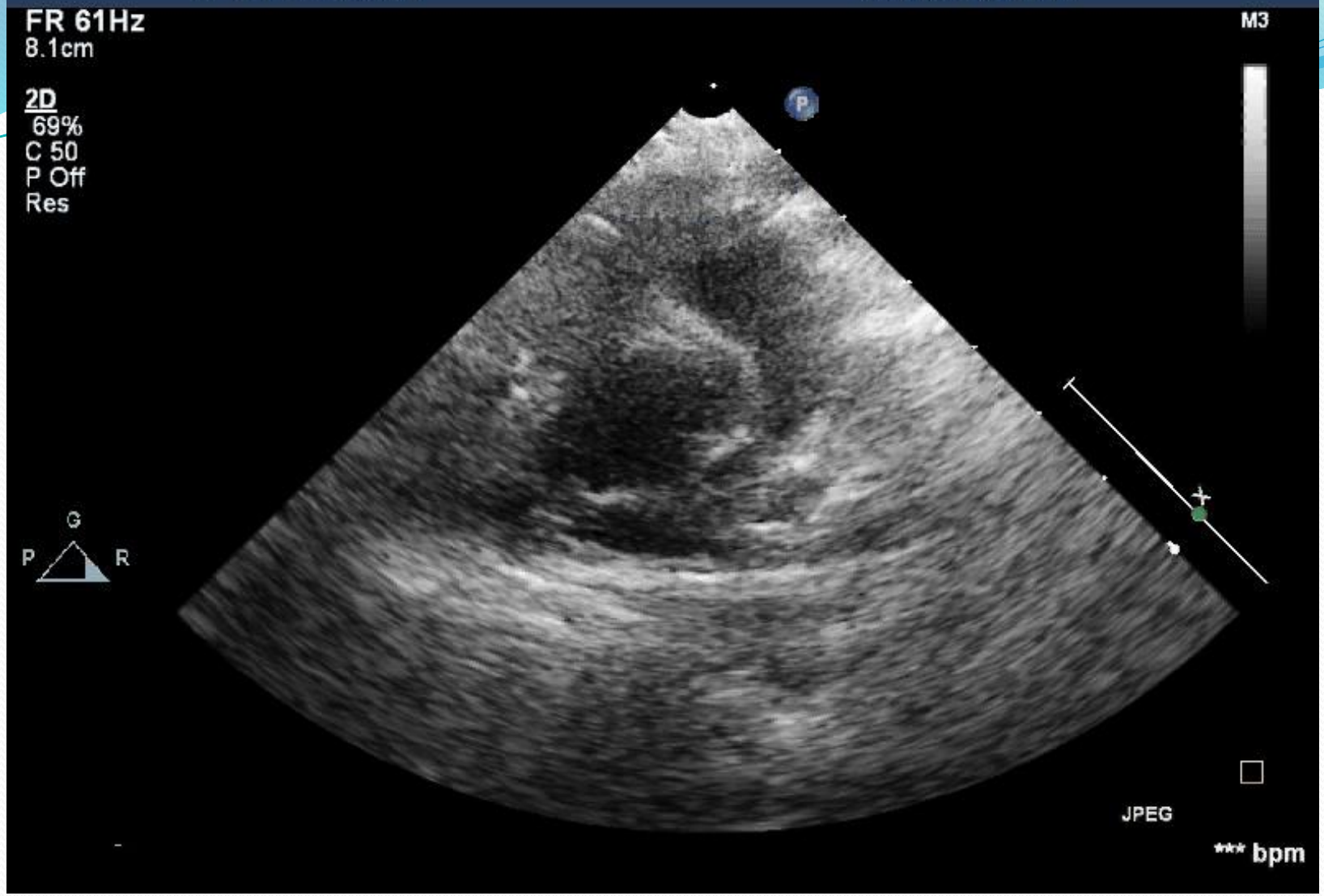


Major Aortopulmonary Collaterals (MAPCA's)






14/10/2011
12:43:01
PHILIPS
HD
Adult New
S4-2
MI 1.2
TIS 1.0
H2 Gn 65
232dB/C3
K/3/0
30Hz 15cm
P
T
R
1.9 3.8


FR 61Hz
8.1cm
2D
69%
C 50
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Res
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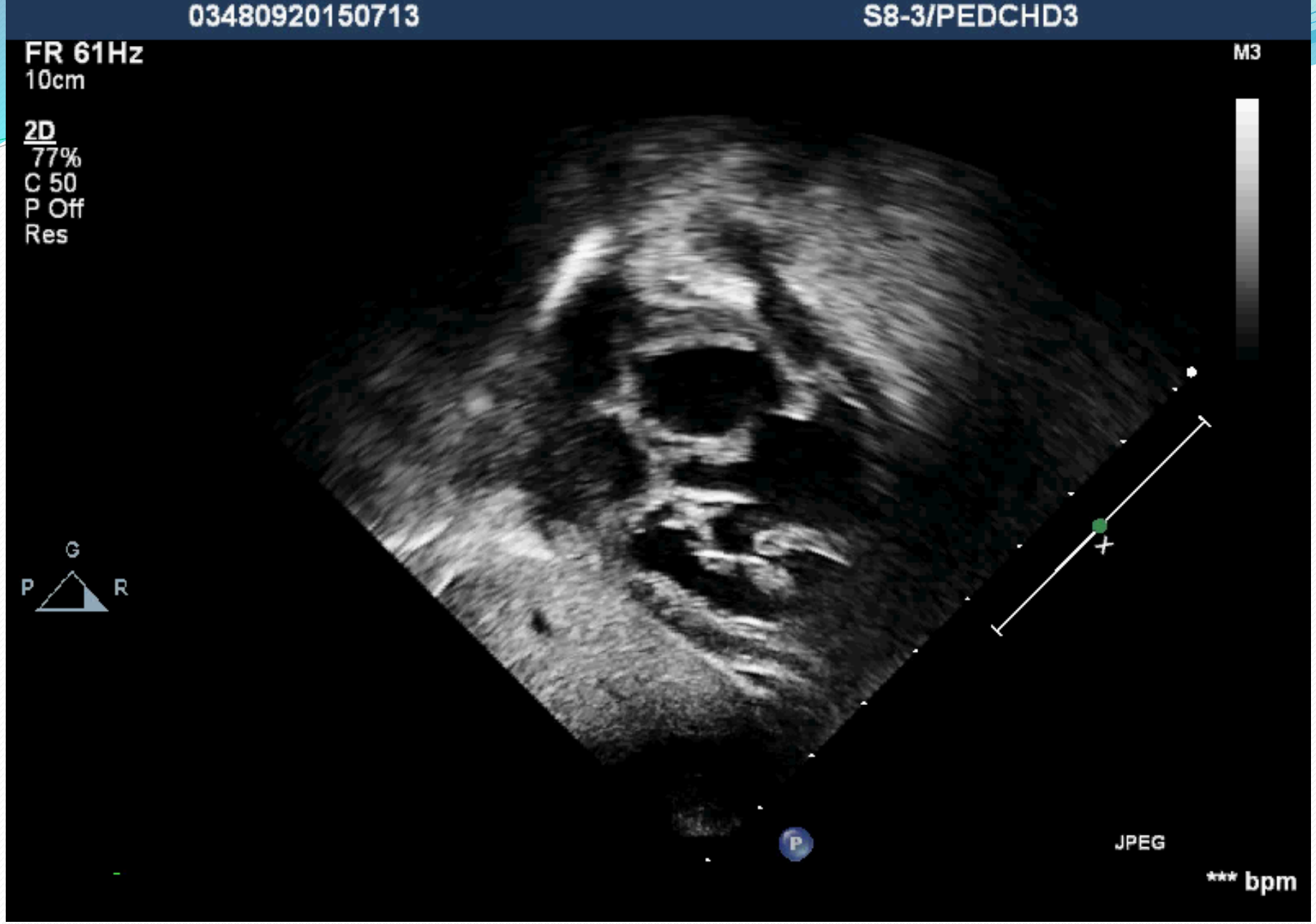
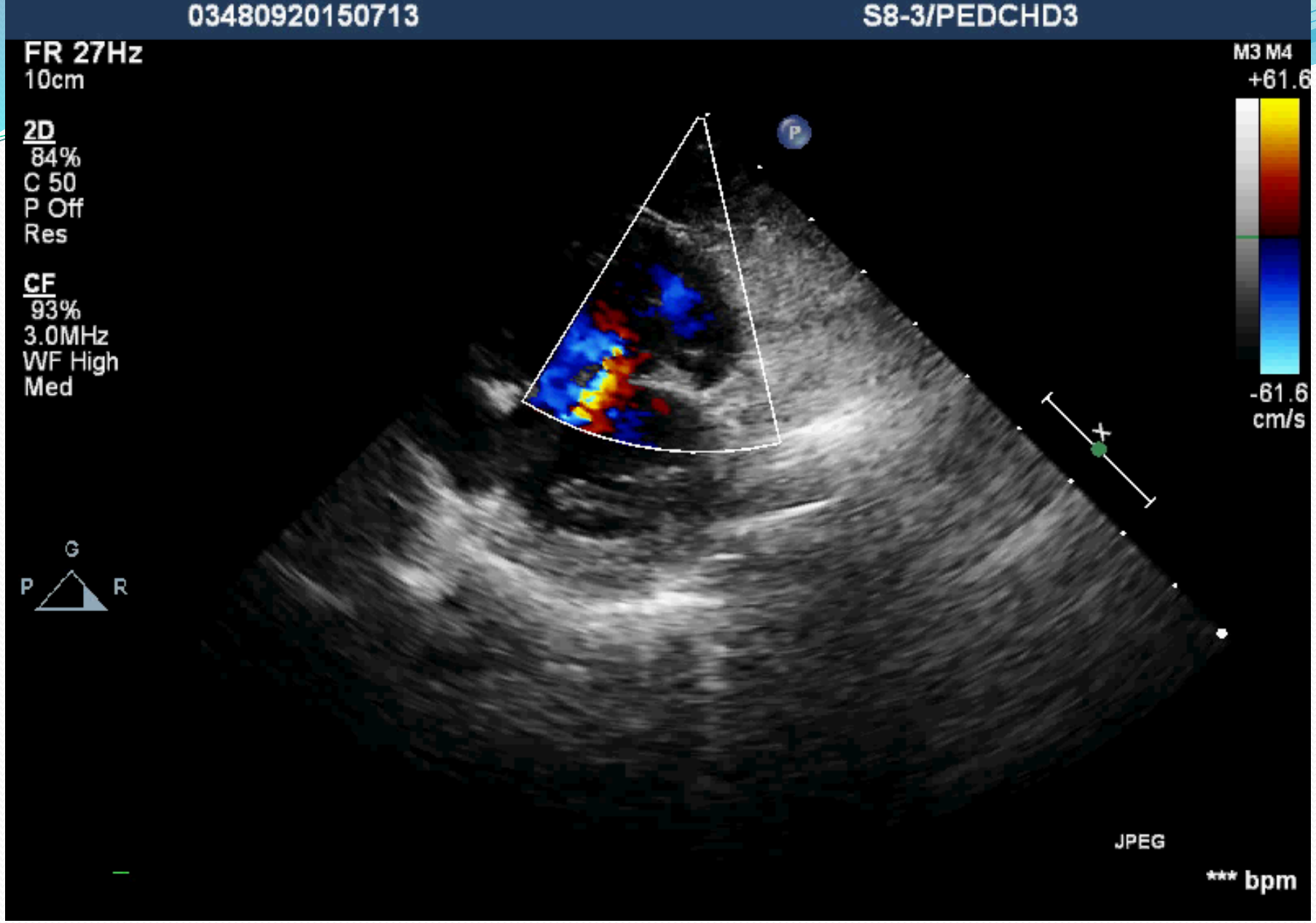

03480920150713
S8-3/PEDCHD3
FR 61Hz
10cm
M3
2D
77%
C 50
P Off
Res
G
P
R
JPEG
*** bpm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S8-3/PEDCHD3
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10cm
2D
84%
C 50
P Off
Res
CF
93%
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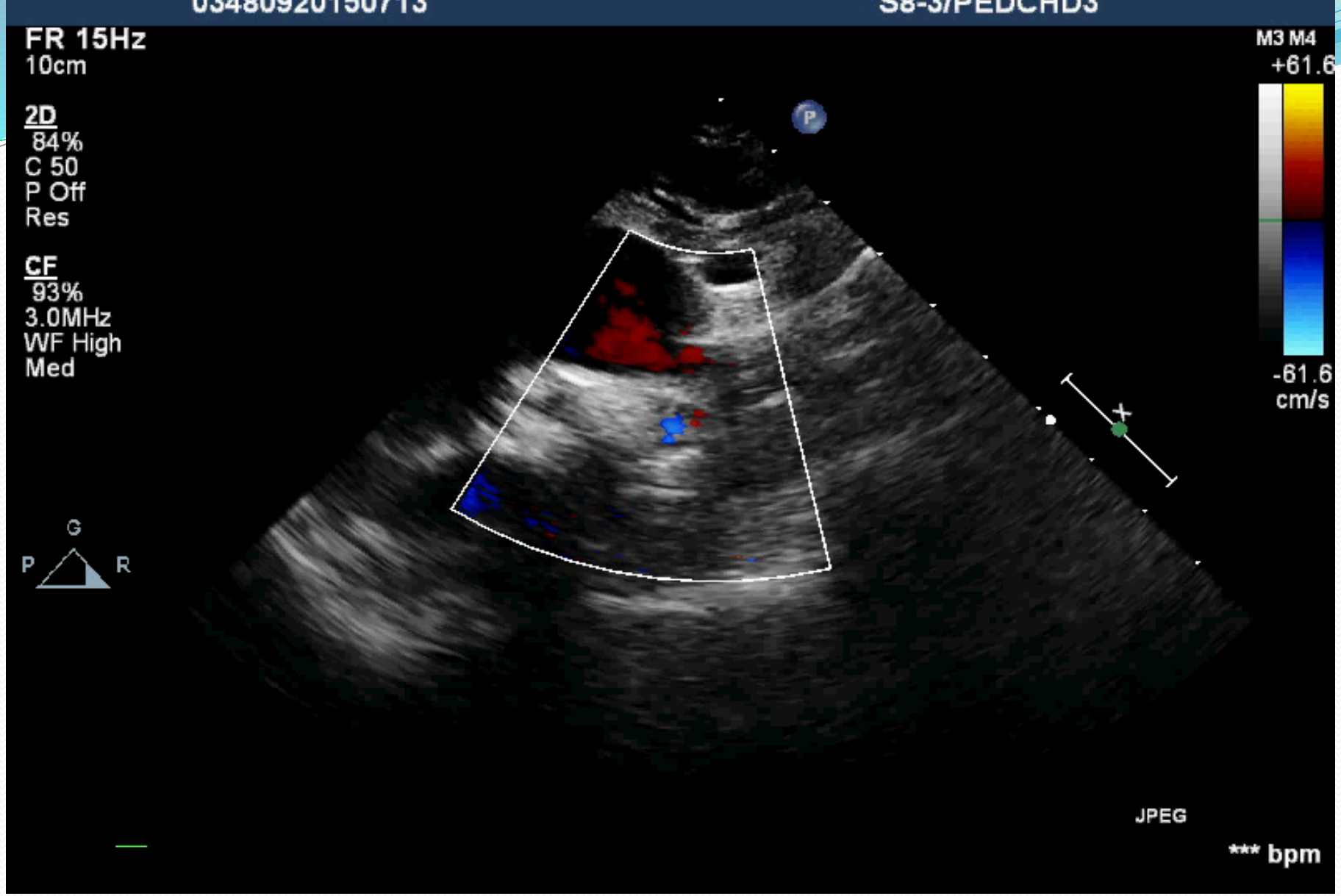
WF High
Med
M3 M4
+61.6
-61.6
cm/s
G
P
R
JPEG
*** bpm


0348092015071 3
S8-3/PEDCHD3
FR 15Hz
10cm
2D
84%
C 50
P Off
Res
CF
93%
3.0MHz
WF High
Med
M3 M4
+61.6
-61.6
cm/s
G
P
R
JPEG
*** bpm

Evolutie naturala-TF

- Cianoza se accentueaza gradat

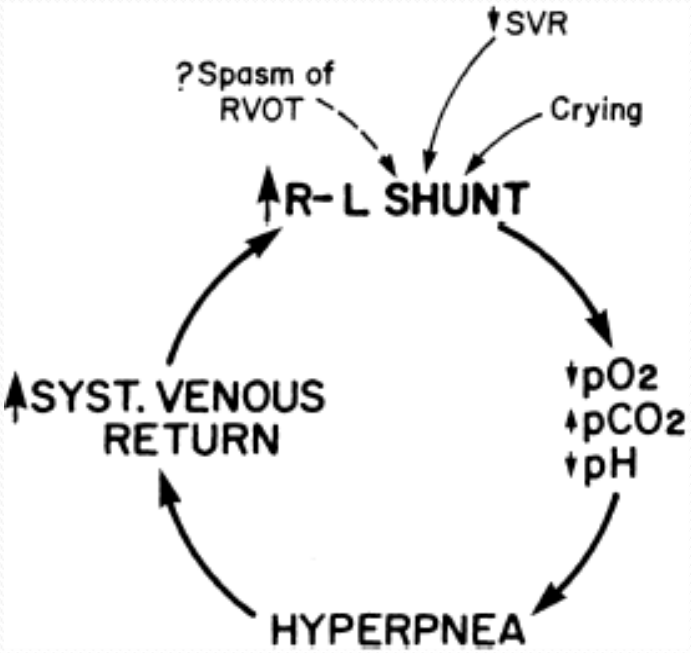
- Policitemie secundara
- Crize hipoxice

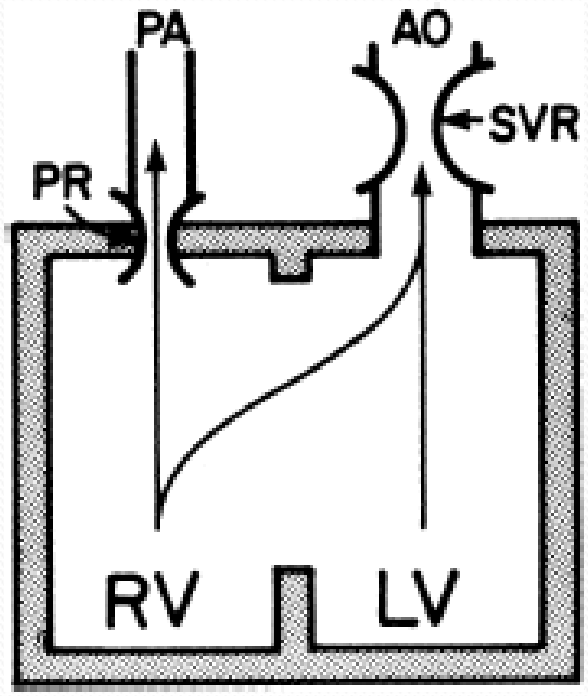
- De obicei cresterea este relativ buna,daca cianoza este severa,cresterea este deficitara

- Abcesele cerebrale apar mai tarziu in evolutie ,la copilul prescolar/scolar

Crizele hipoxice

- Varf de incidenta 2-4 l
- Nu se coreleaza cu gradul de cianoza in repaus
- Apar la plans,alimentatie,scaun





Crize hipoxice -tratament

- Pozitie genupectorala
- Morfina 0,2 mg/kg

-deprima centrul respirator

-opreste hiperpneea

- O2 se adm de rutina-s-a demonstrat ca are efect minim pe cresterea sat O2
- Bic de Na-corectarea acidozei 1mEq/kg iv,repetat la 10-15 min
- Adrenalina
- Propranolol iv 0,01-0,25 mg/kg
- Ketamina 1-3 mg/kg adm in 60 s,creste rezist sistemica,sedativ

Management

- Medical:

- Propranolol 0,5-1,5 mg/kg c la 6 ore

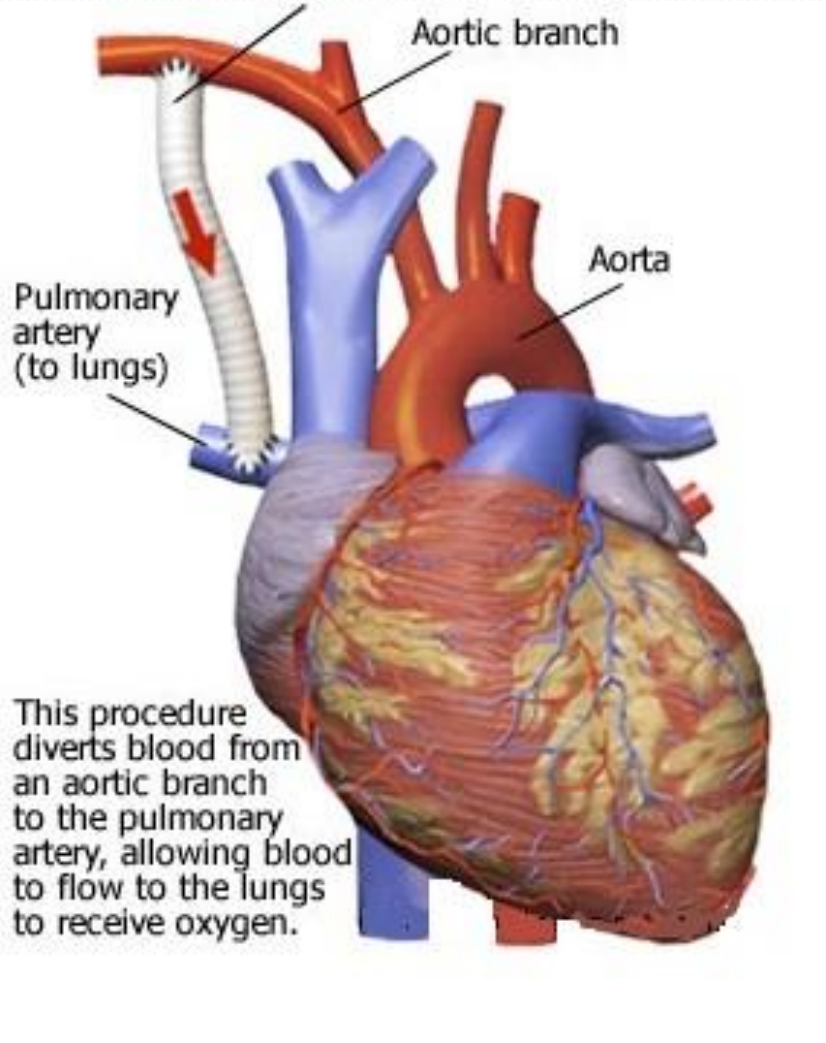
- Tratamentul crizelor hipoxice

- Tratamentul anemiei

Management

- Chirurgical

1.Paleativ- sunt BT



Management

- Chirurgical

2. Corectie totala

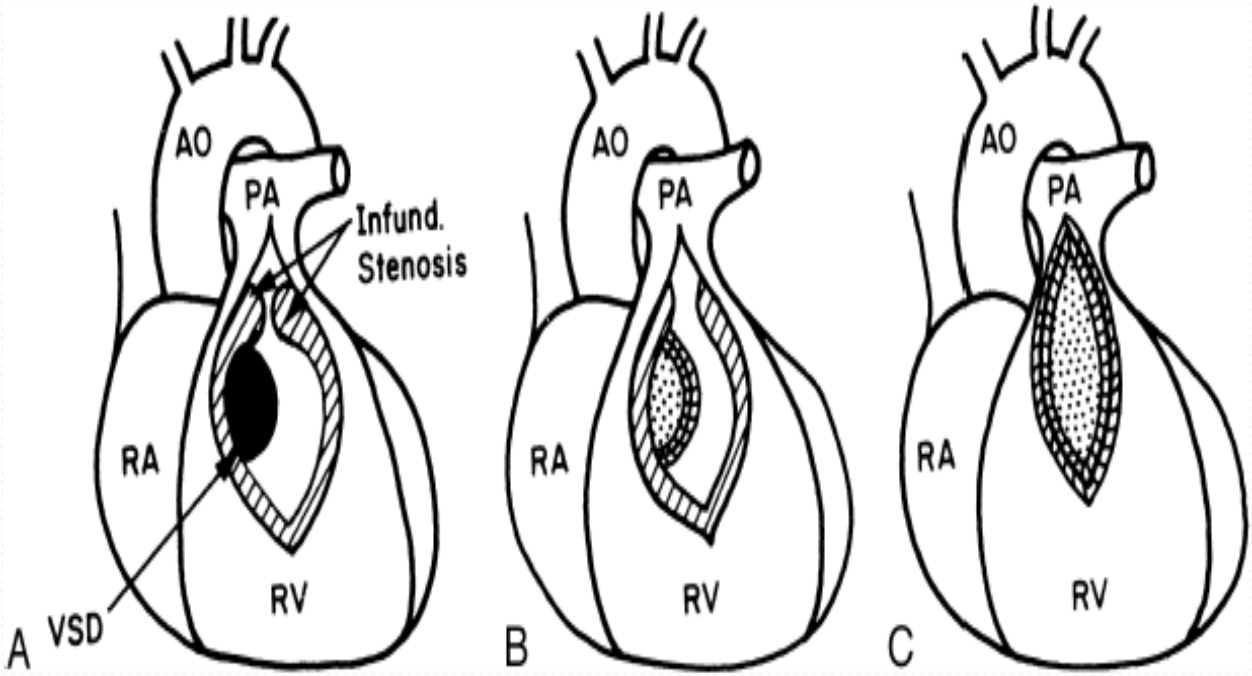
-Simptomatici:

3-4 l

-Asimptomatici:

4-24 l

Mortalitate 2-3 %

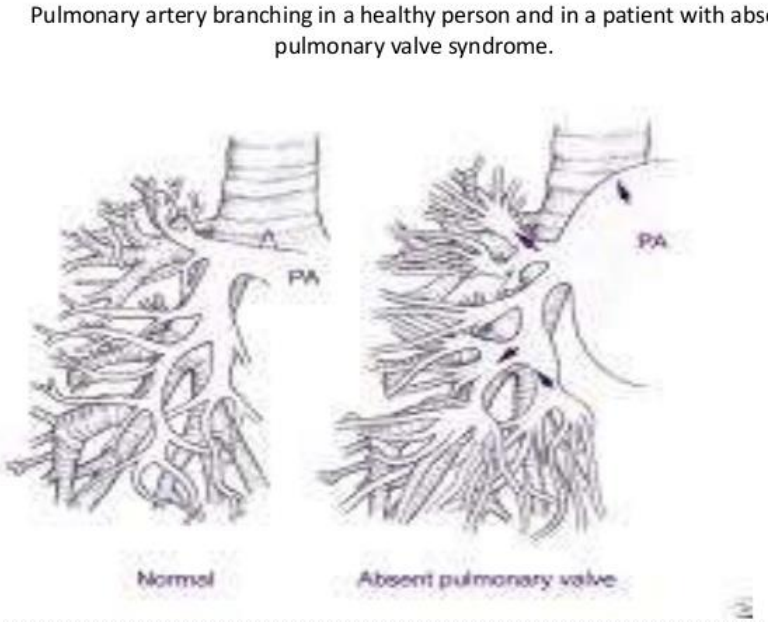


Complicatii postoperatorii

- Insuficienta pulmonara
- BRD
- BAV total
- Aritmii ventriculare

Follow- Up of Repaired ToF

- Significant PR- Following transannular patch
- Residual RVOT obst.
- RV Dilatation , TR, RV Failure
- Residual VSD
- Progressive AI with or without root dilation
- LV Dysfunction
- Infective Endocarditis
- Electrical Complications: RBBB, Bifasicular block, 3° AV block, A Fl. , AF, NSVT, VT
- Sudden Cardiac Death



TF cu absenta de VP

-3-6% din TF

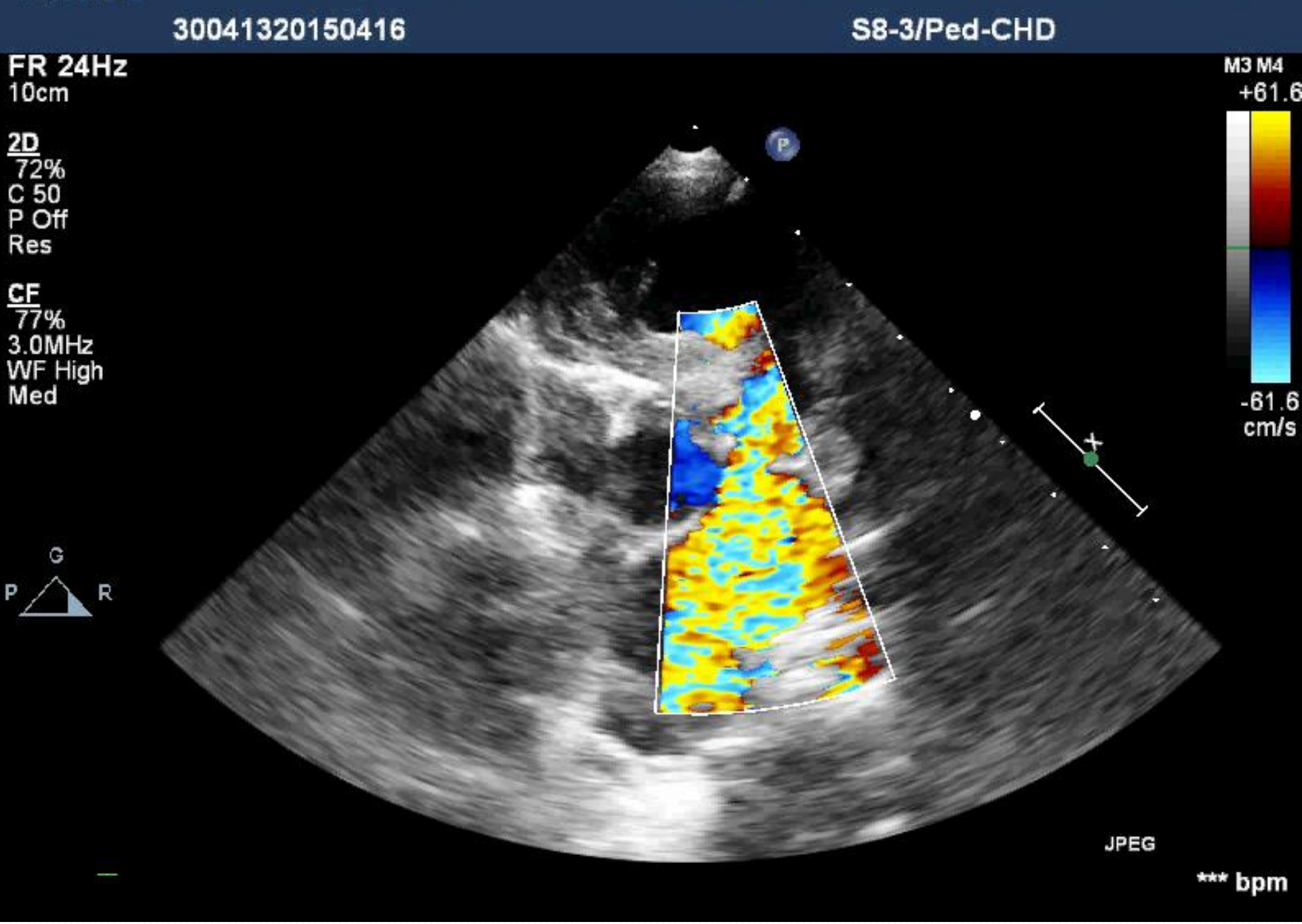
-regurgitare pulmonara severa

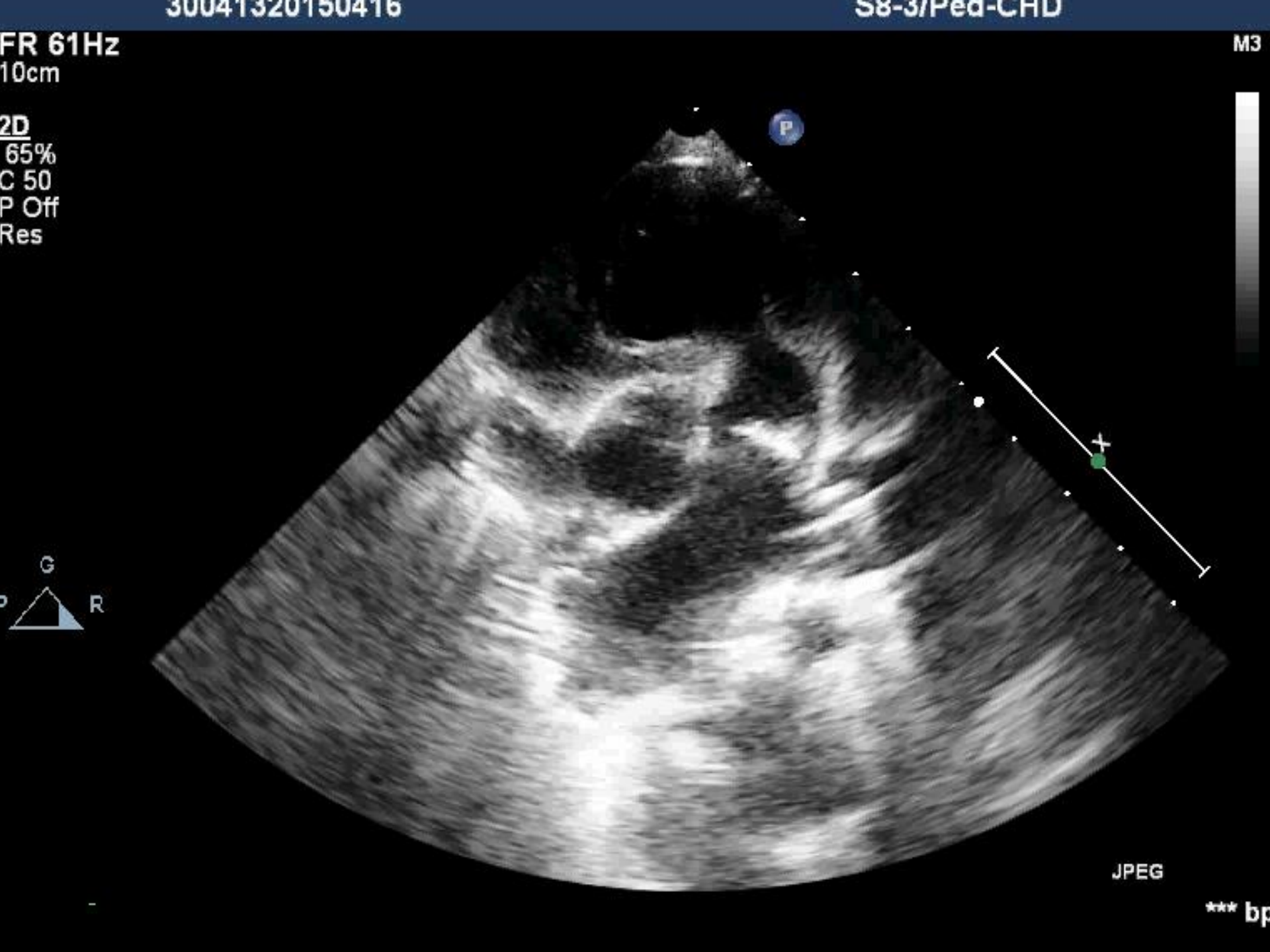
--dilatatie anevrismala tr AP si ramuri

-compresie traheobronsica

cu dificultati severe de respiratie imediat dupa nastere sau din prima saptamana

-exista forme cu simptomatologie usoara


30041320150416
S8-3/Ped-CHD
FR 24Hz
10cm
2D
72%
C 50
P Off
Res
CF
77%
3.0MHz
WF High
Med
M3 M4
+61.6
-61.6
cm/s
G
P
R
JPEG
*** bpm


3004132015041 6
S8-3/Ped-CHD
FR 61Hz
10cm
2D
65%
C 50
P Off
Res
M3
G
P
R
JPEG
*** bp

Atrezie pulmonara cu SIV intact

- < 1% din MCC

-VP atretica

-infundibulul pulmonar atretic

-hipoplazie tr AP

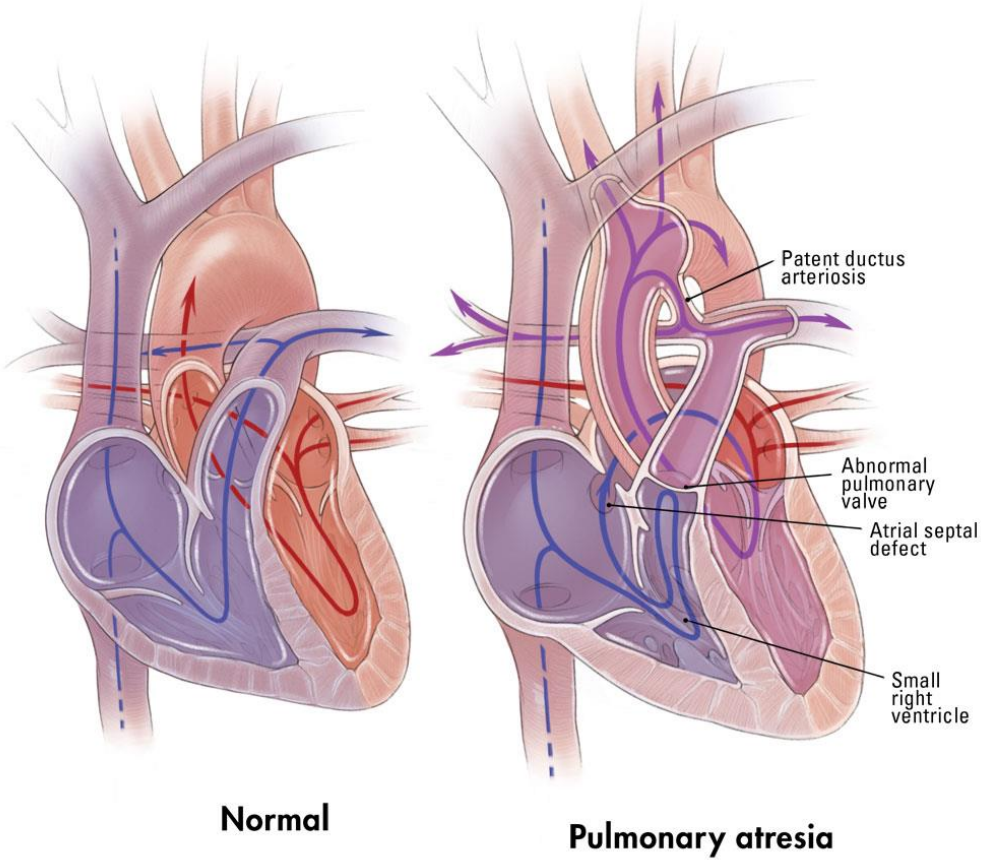
-VD variabil

-VT poate fi ebsteinoida/stenotica

-SIV intact

-fistule coronariene-VD.Circ coronariana dependenta de PTDVI

Necesita DSA/CAP pt supravietuire.(CAP este inversat, vertical)



Tipul I

- Atrezie pulmonara
- Hipoplazie VD

➢ Asociaza

- patologie de VT(hipoplazie,Ebstein)
-hipertrofie severa VD
-Fistule coronariene

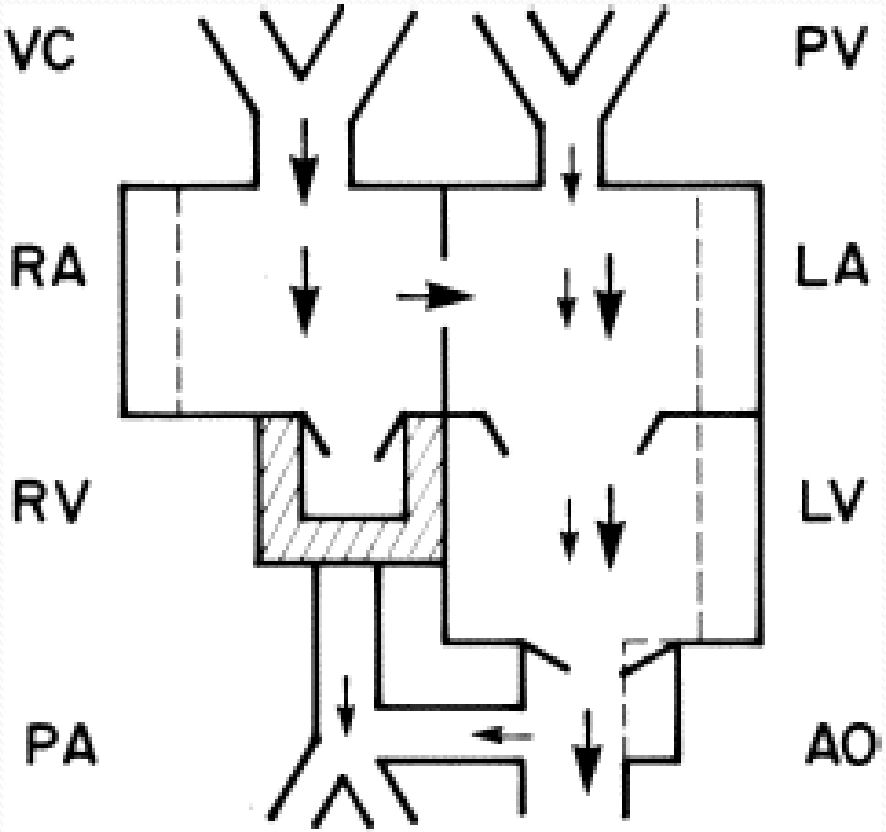
Tipul II

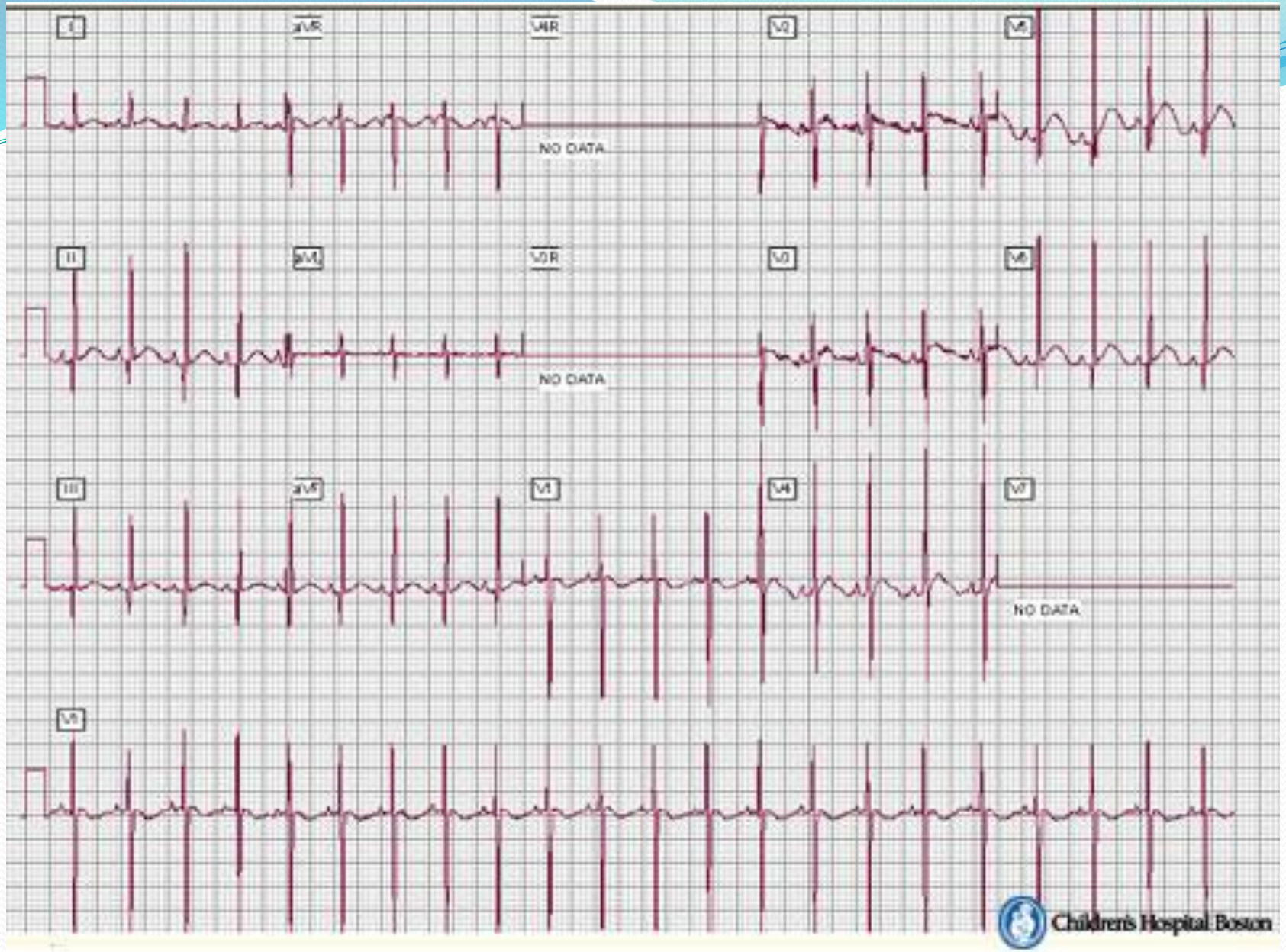
- Atrezie pulmonara
- Cavitati drepte largite sau normale

- Asociaza
- -IT semnificativa
- AD dilatat
- -Fara fistule aorto-coronariene

ISTORIE NATURALA

- 50% din nn mor in primele 2 s
- 85% mor in primele 6 l


VC
PV
RA
LA
RV
LV
PA
AO



Hipertrofie atriala dreapta
Hipertrofie stanga
ax la stg (anormal pt varsta nn)

Ecografia

- **Debitul prin CAP**

-mentine debitul pulmonar

- **Suntul prin DSA**

Decomprima AD si permit suntul din sistemul venos sistemic catre AS

- **Marimea VD**

-prezenta sau absenta celor 3 segmente:inlet,trabecular,outlet

- **Evaluarea CEVD**

-prezenta lumenului infundibular,atrezia musculara/membranoasa

- **Prezenta unei deschideri in VP**

-datorita jetului din CAP greu vizibila (jetul inchide valva),se poate vizualiza IP

Evaluarea VT

-morfologia valvelor ,

-marimea inelului

Z score< -3 ,corectie univentriculara;se asociaza cu sinusoide

Z score>-3,poate fi corectat biventricular

Inel T/inel Mi >0,5 prognostic biventricular bun

Existenta de fistule coronariene –VD/sinusoide

- **Evaluarea VT**

-morfologia valvelor ,

-marimea inelului

Z score< -3 ,corectie univentriculara;se asociaza cu sinusoide

Z score>-3,poate fi corectat biventricular

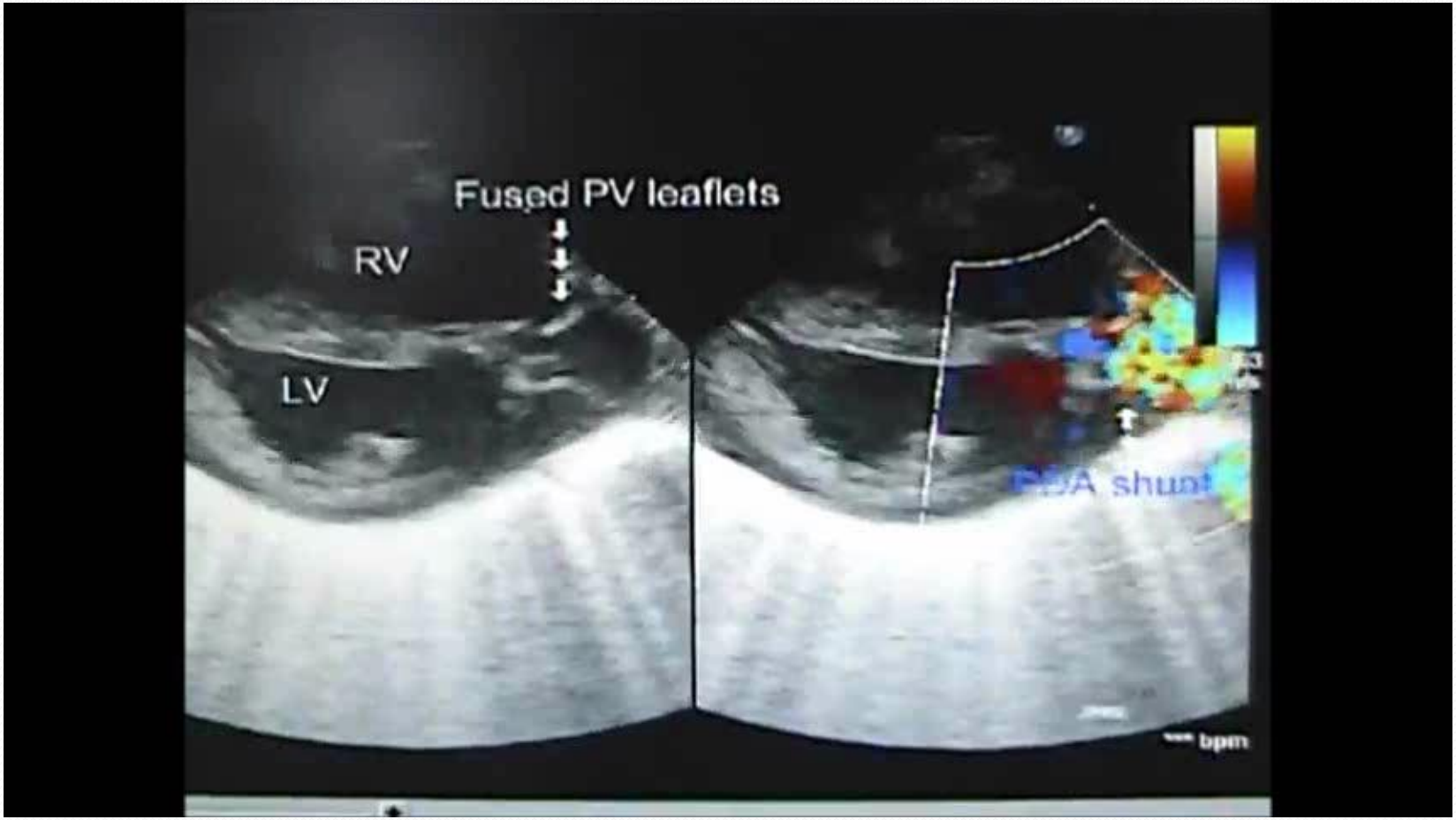
Inel T/inel Mi >0,5 prognostic biventricular bun

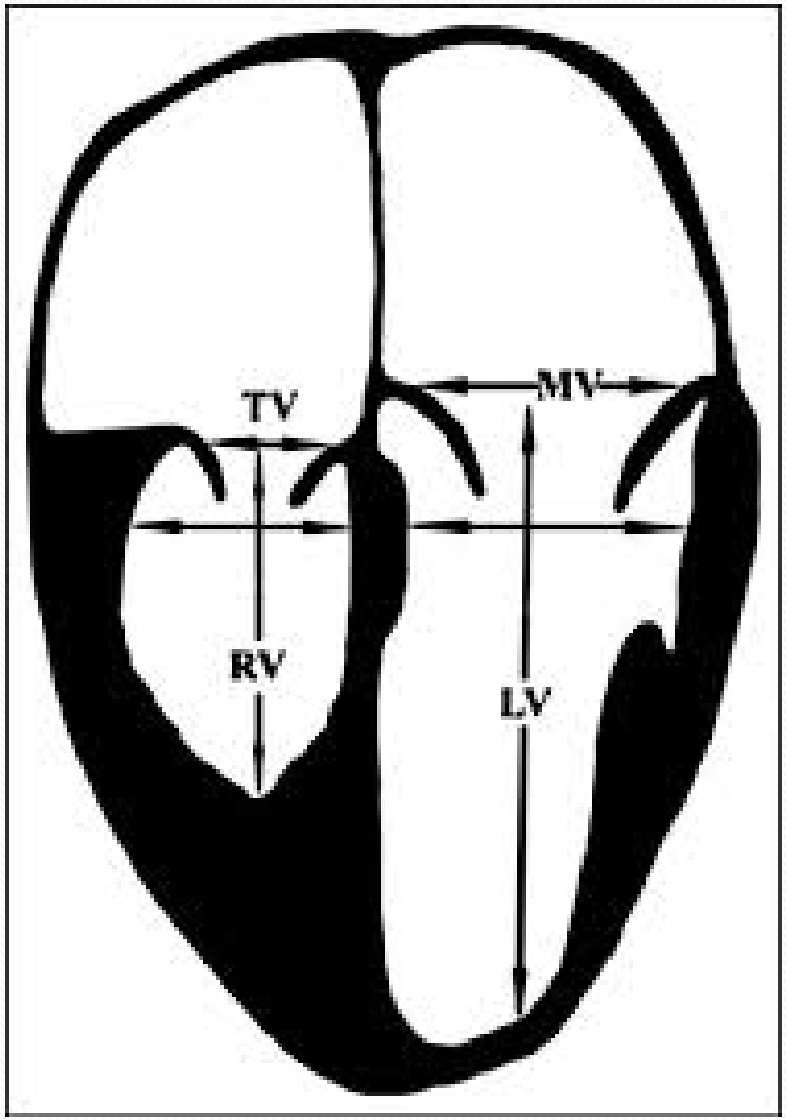
- **Existenta de fistule coronariene –VD/sinusoide**

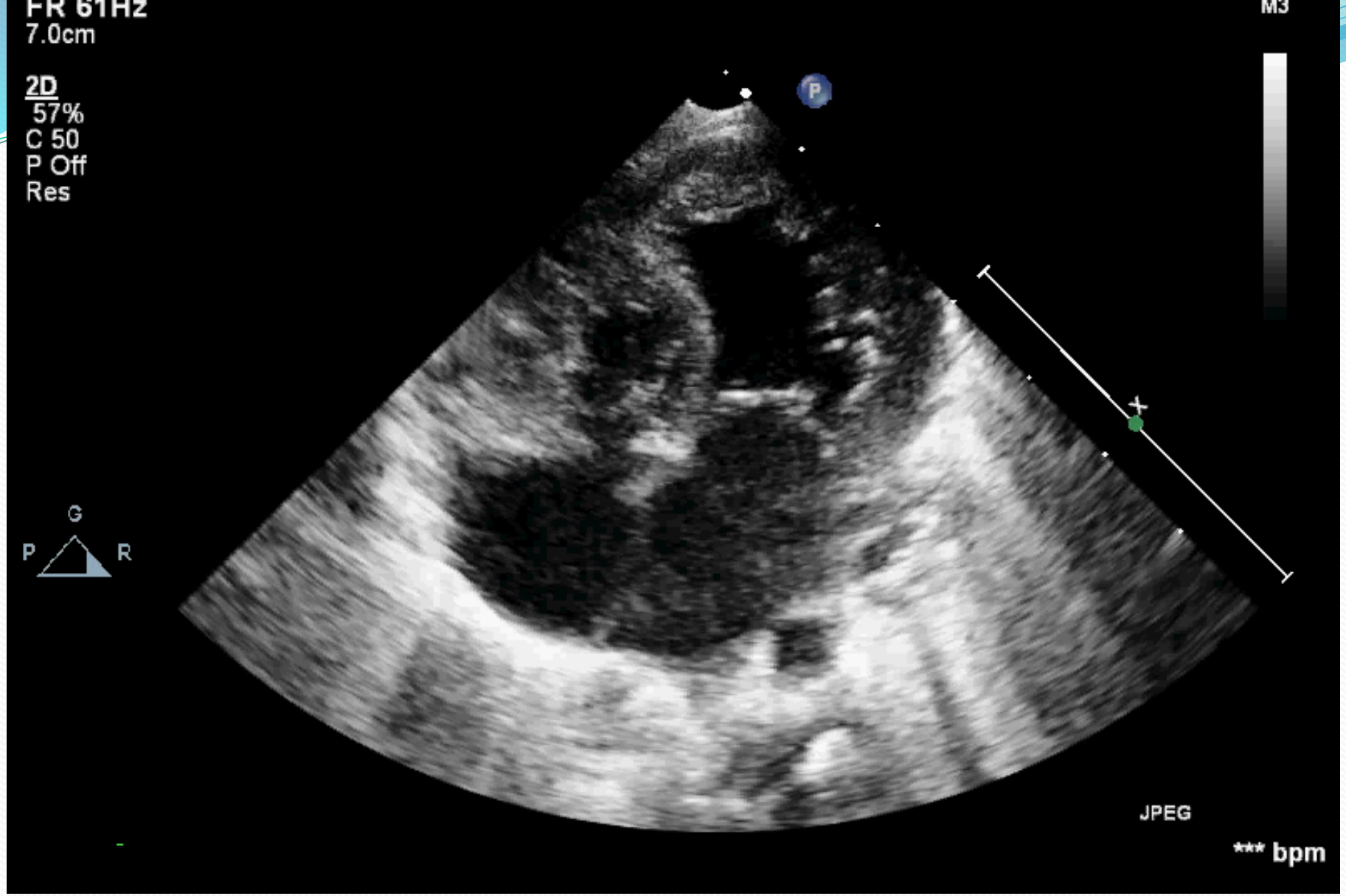
Sinusoidele VD sunt canale endoteliale in miocard care comunica cu cavitatea VD.

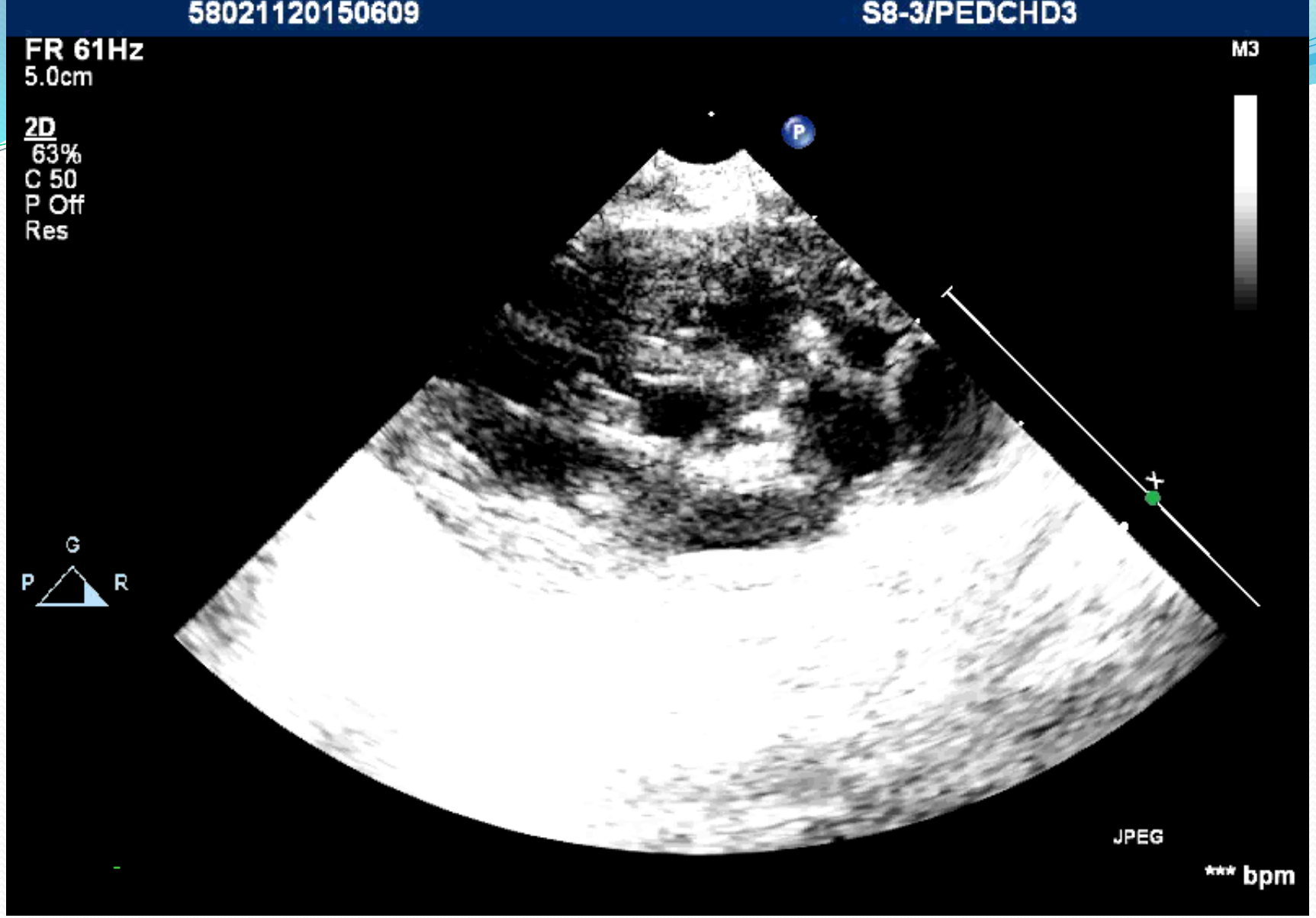
Exista in viata fetala timpurie spatii sinusoide care hranesc miocardul.Acestea pot persista in viata postnatala si determina comunicare VD-artere coronare

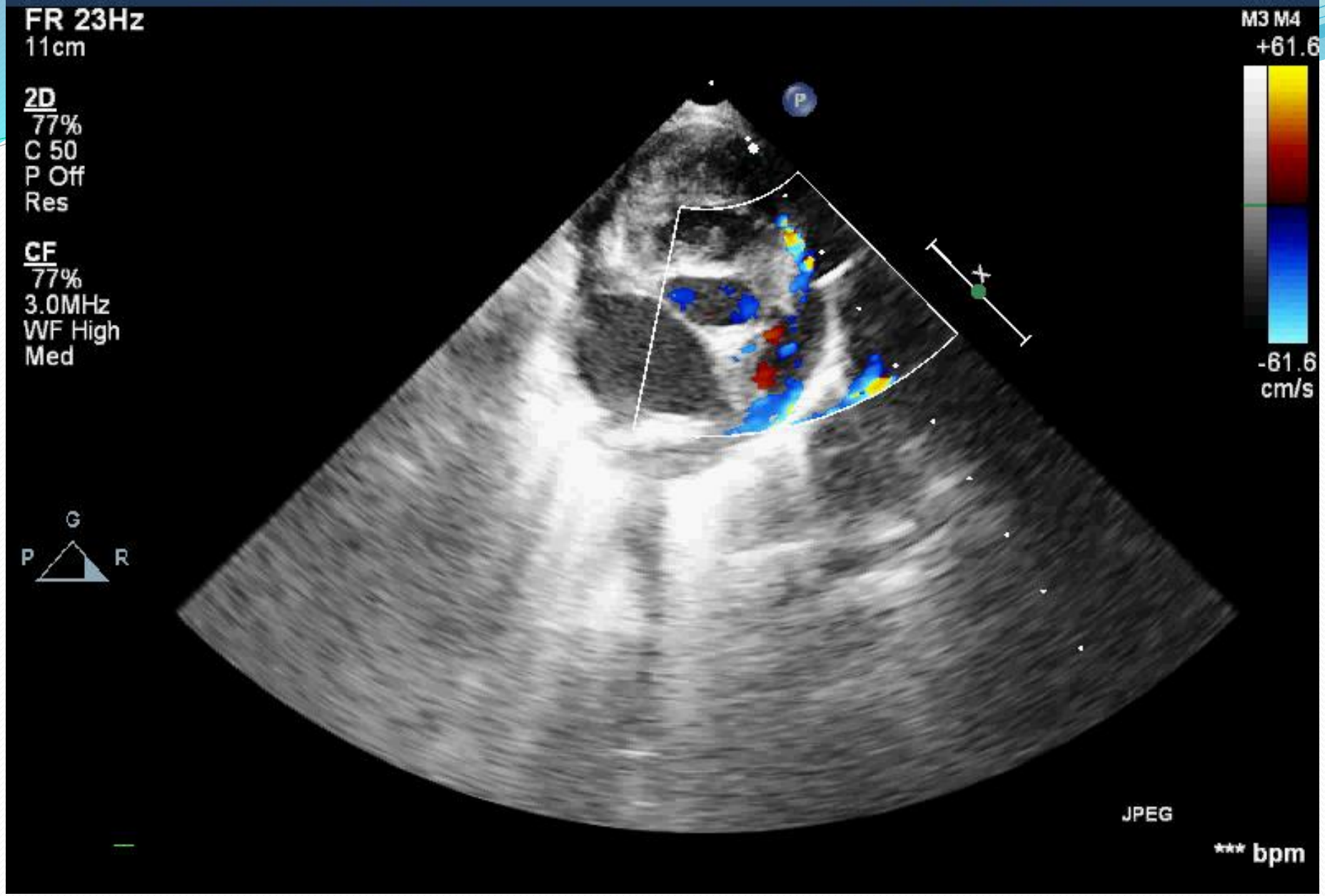
Anormalitati artere coronariene:stenoze severe sau atrezia ostiumului coronar-ca urmare mioccardul nu primeste flux anterograd din a coronara ci retrograd din cavitatea VD.Circulatie coronariana dependenta de VD

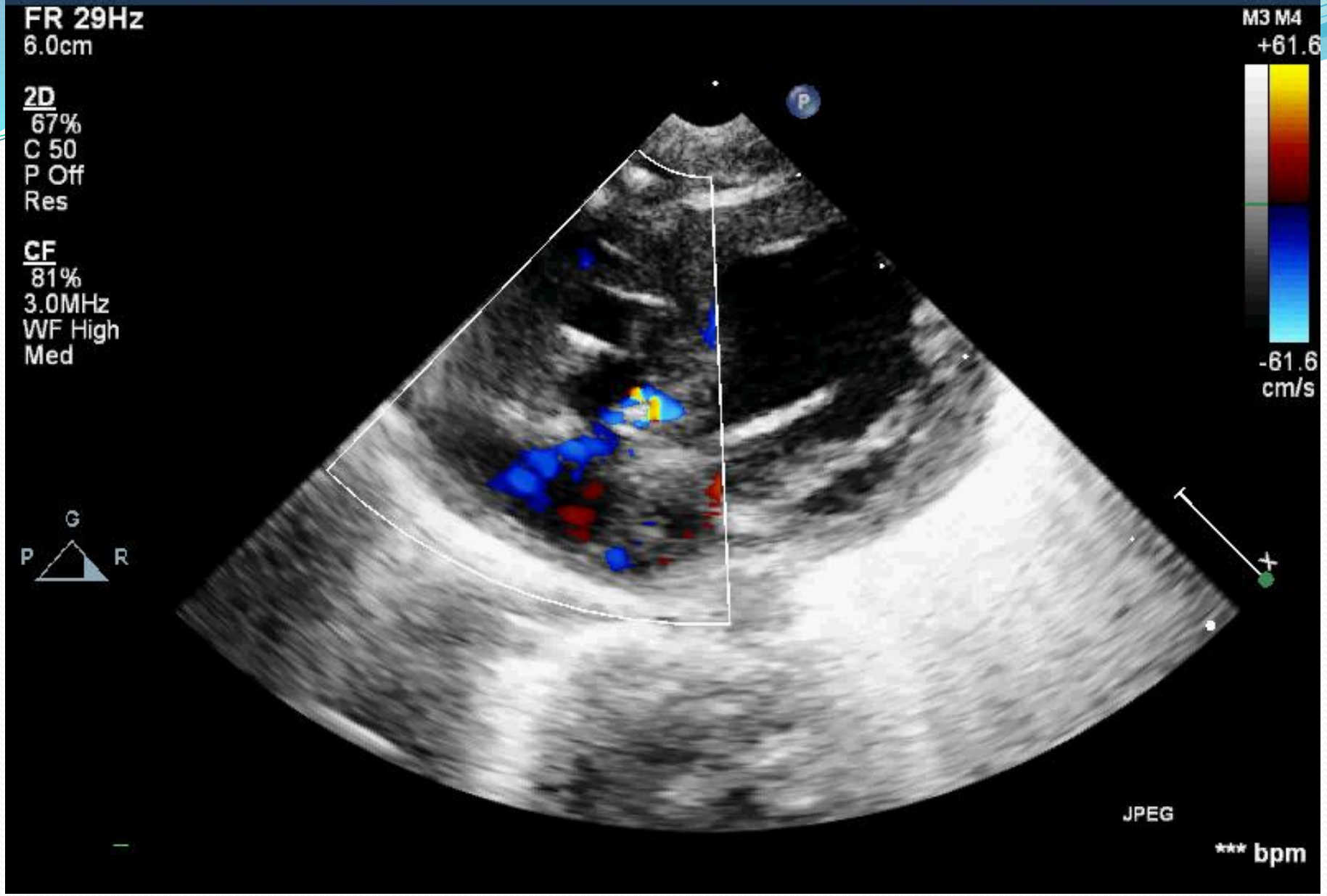

Fused PV leaflets
RV
LV
bpm


TV
MV
RV
LV


FR 61Hz
7.0cm
2D
57%
C 50
P Off
Res
M3
P
G
P
R
JPEG
*** bpm


580211201506O9
S8-3/PEDCHD3
FR 61Hz
5.0cm
2D
63%
C 50
P Off
Res
M3
G
P
R
JPEG
*** bpm


FR 23Hz
11cm
2D
77%
C 50
P Off
Res
CF
77%
3.0MHz
WF High
Med
M3 M4
+61.6
-61.6
cm/s
G
P
R
JPEG
*** bpm


FR 29Hz
6.0cm
2D
67%
C 50
P Off
Res
CF
81%
3.0MHz
WF High
Med
G
P
R
M3 M4
+61.6
-61.6
cm/s
JPEG
*** bpm

Cateterism

- Aprecierea circulatiei coronariene: stenoze,fistule
- Aprecierea dependentei circ coronariene de VD
- Aprecierea VD

Management

- Medical

-PG E1

-corectarea acidozei metabolice

-mentinerea unui Ht>40 pentru o perfuzie tisulara adecvata

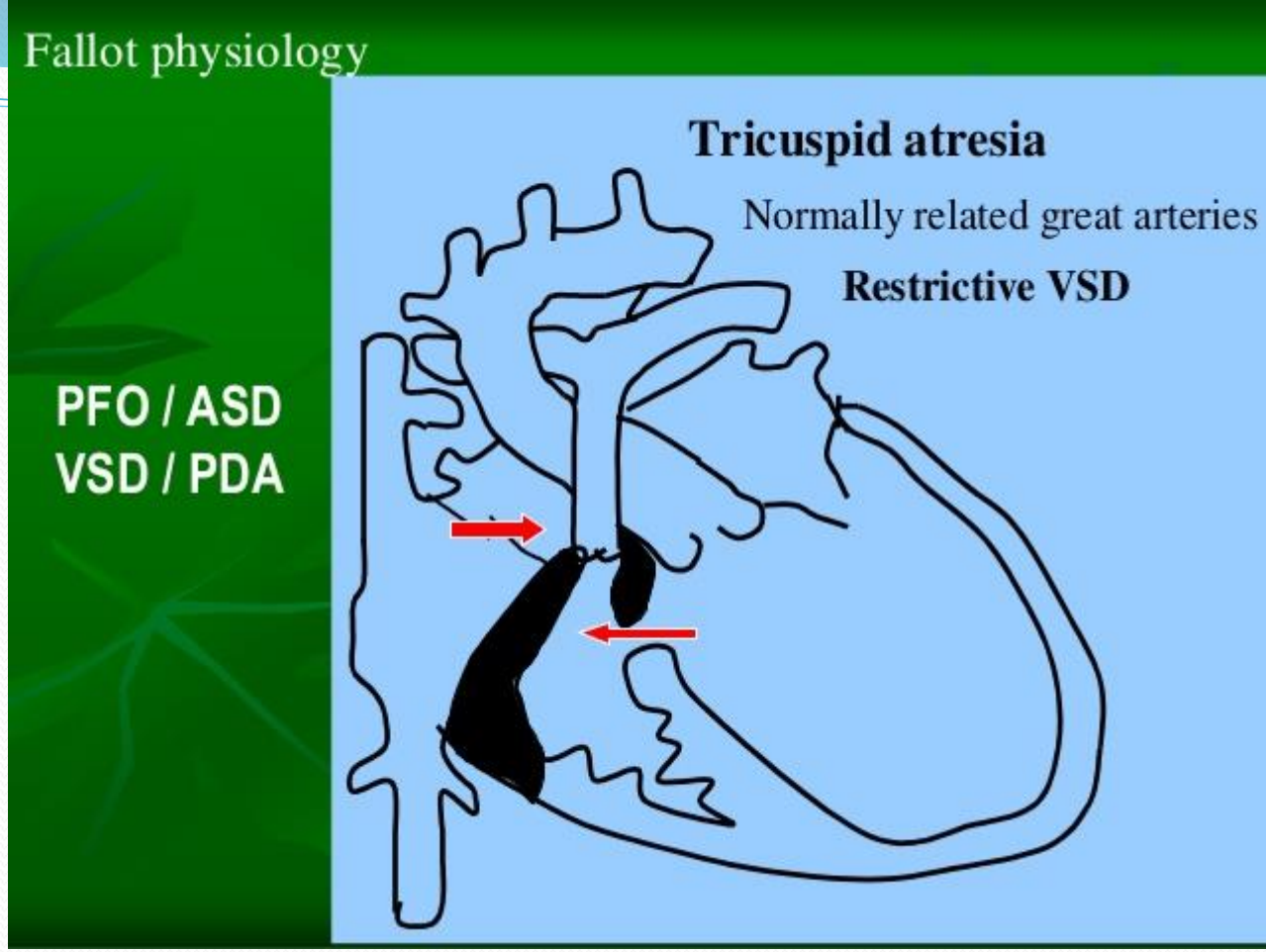
-suport inotropic daca perfuzia este inadecvata

-mentinerea unei sat >70%

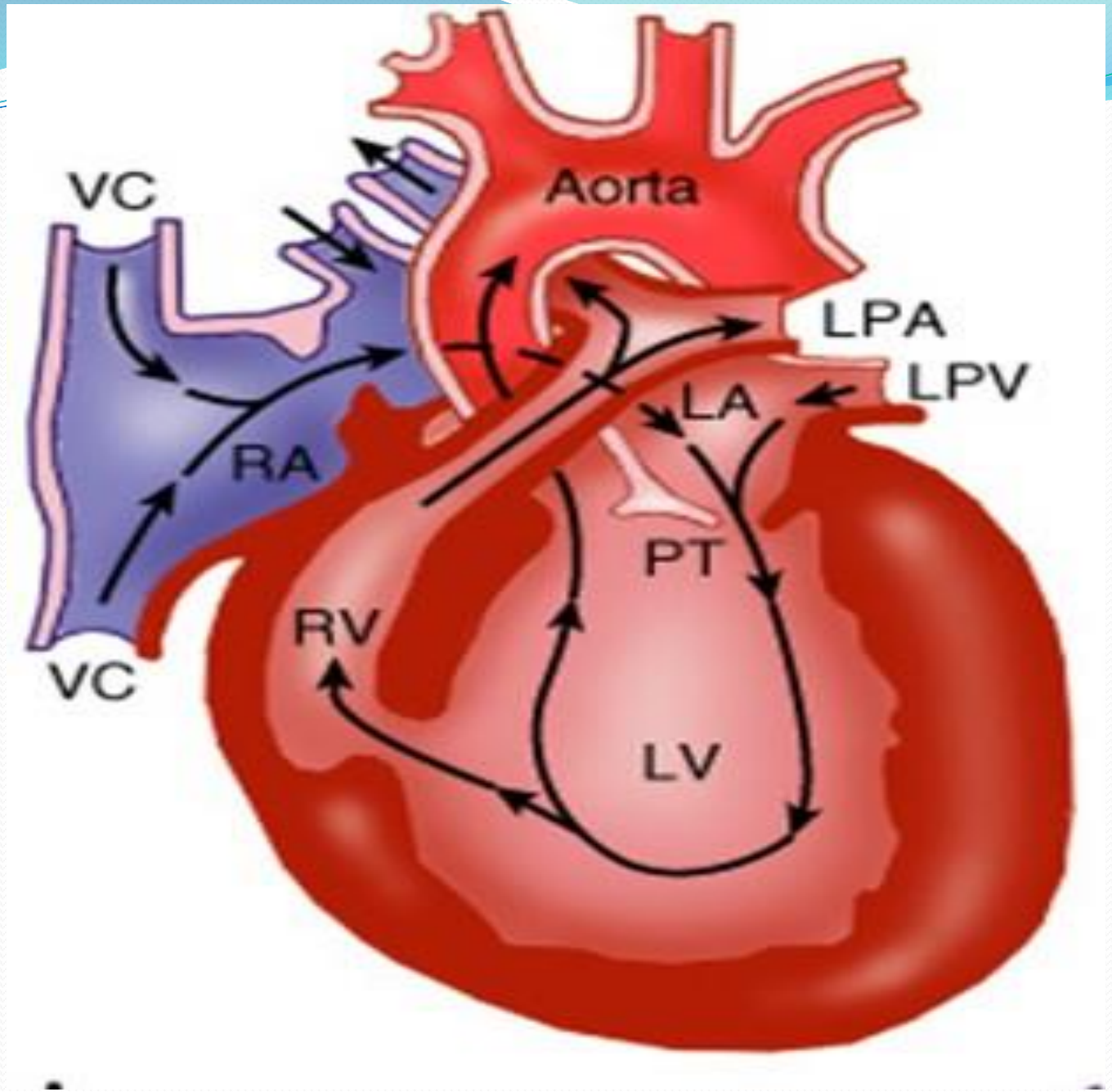
- Chirurgical

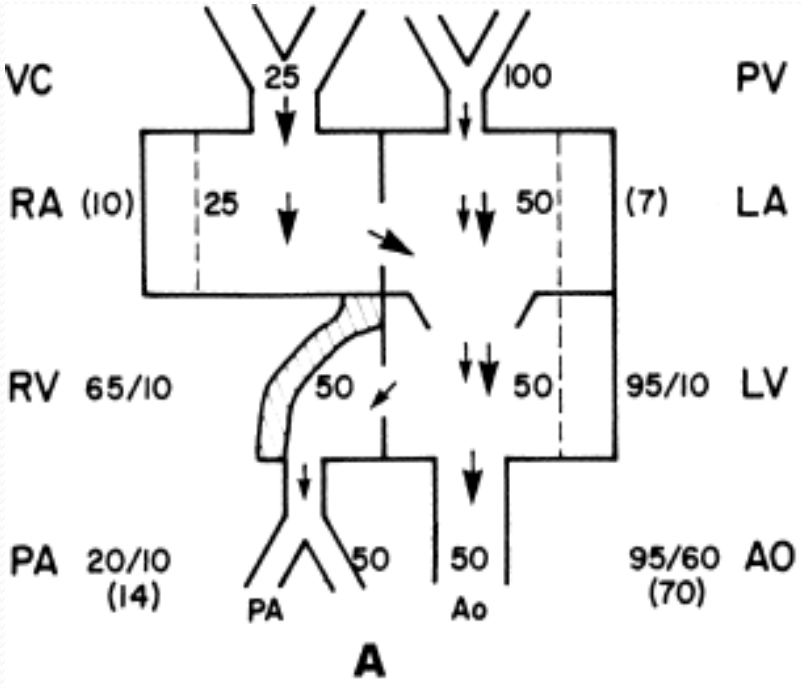
-abordarea poate fi biventriculara sau univentriculara in functie de dimensiunile VD,inelul tricuspidei si prezenta dependentei circulatiei coronariene de VD

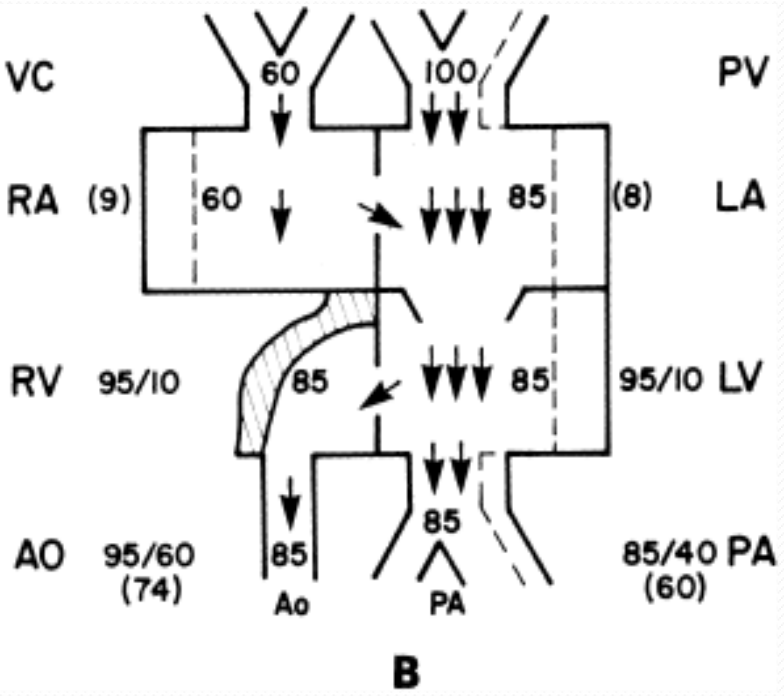
Atrezie VT



- 1-3% din MCC
- VT este absenta
- Hipoplazie VD prin nedezvoltare regiunii inlet
- Necesita DSA/DSV/CAP pentru supravietuire
- Vasele mari normal pozitionate in 70% si transpuse in 30%


VC
Aorta
LPA
LPV
LA
RA
PT
RV
VC
LV

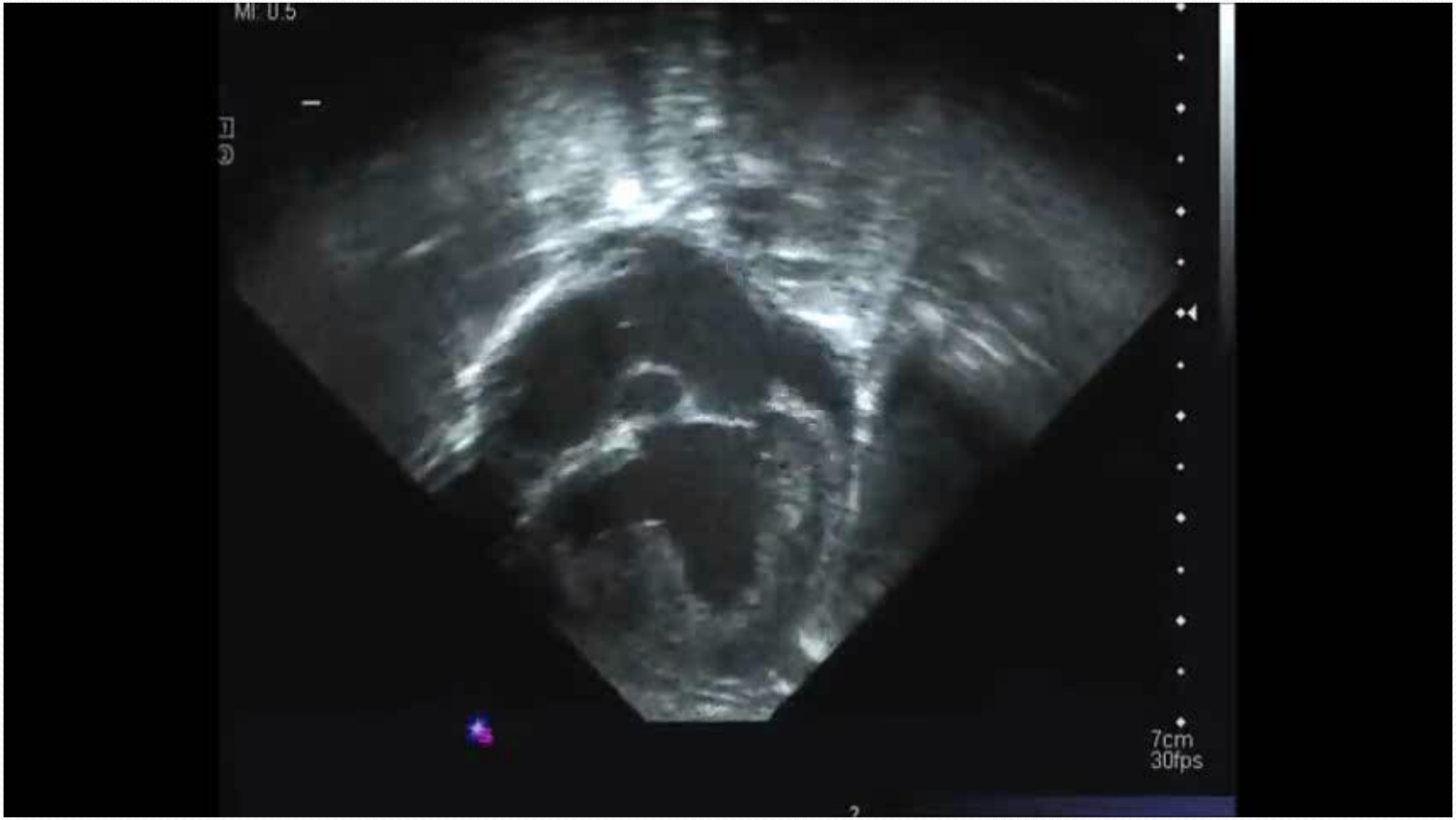

VC
25
100
PV
RA (10)
25
50
(7)
LA
RV 65/10
50
50
95/10
LV
PA 20/10
(14)
50
50
95/60
(70)
AO
PA
Ao
A

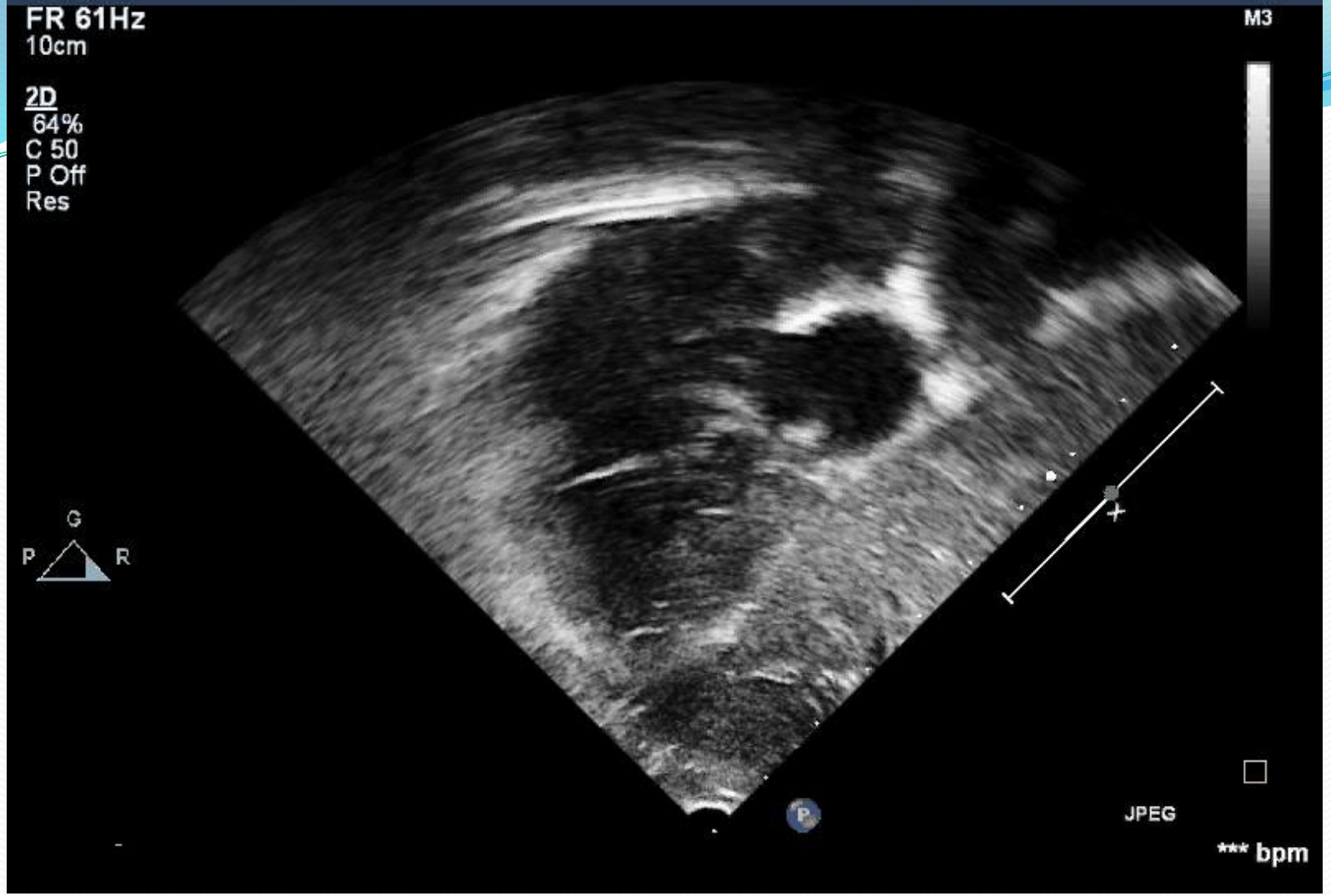

VC
60
100
PV
RA (9)
60
85
(8)
LA
RV 95/10
85
85
95/10
LV
85
AO 95/60
(74)
85
85/40
(60)
PA
Ao
PA
B

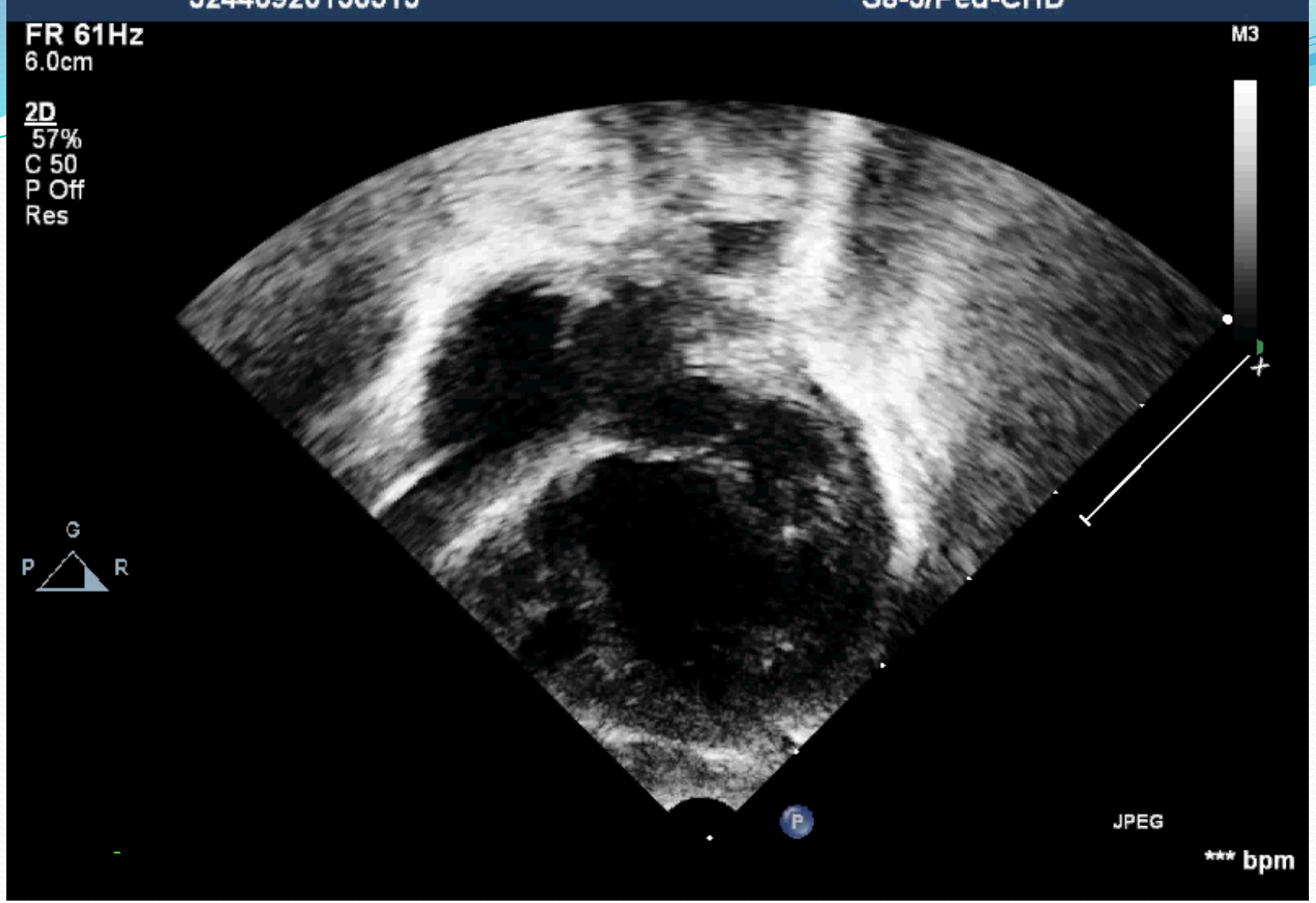
Manifestari clinice

- Cianoza severa apare de la nastere
- Tahipnee
- Dificultati de alimentare

eco




FR 61Hz
10cm
2D
64%
C 50
P Off
Res
M3
G
P
R
JPEG
*** bpm


FR 61Hz
6.0cm
2D
57%
C 50
P Off
Res
M3
G
P
R
JPEG
*** bpm

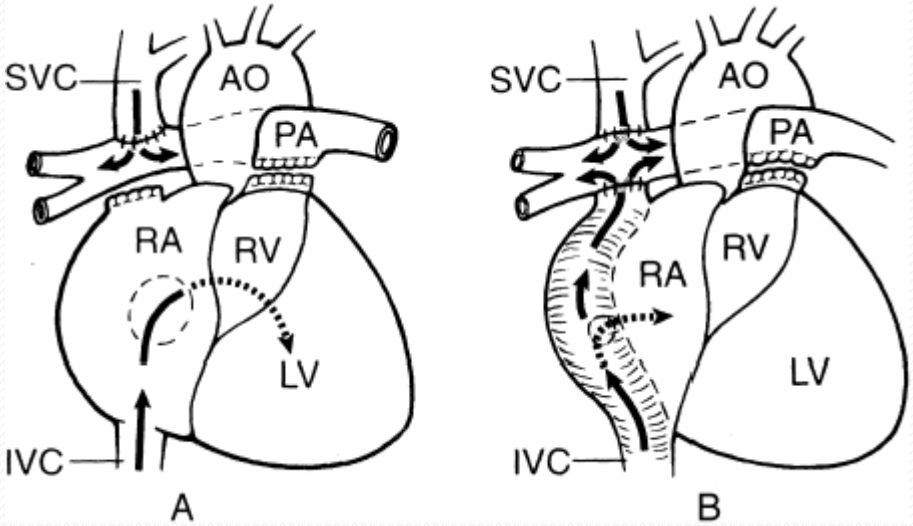
Management

- Medical

-PG E1 pentru mentinerea CAP

-Rareori ICC ce necesita tratament

Chirurgical:proceduri paleative-Fontan



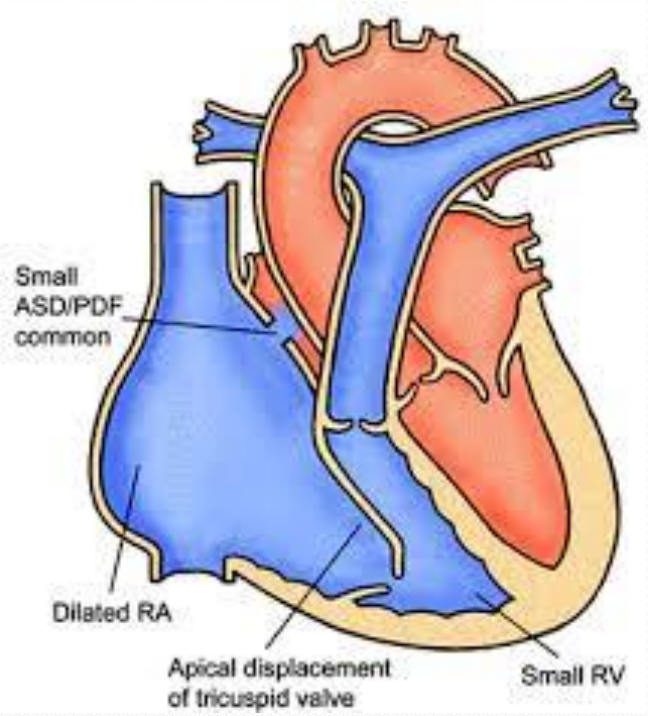
EBSTEIN

- <1 % din MCC
- Malformatie VT cu insertie apicalizata a VT septala si posterioara
- Atrializare VD
- AD dilatat
- PFO/DSA
- Regurgitare tricuspidiana variabila
- Asociere cu alte MCC:

-atrezie pulmonara,T.Fallot,DSV

- Asociere frecenta cu WPW

-Tulburari de ritm supraventriculare



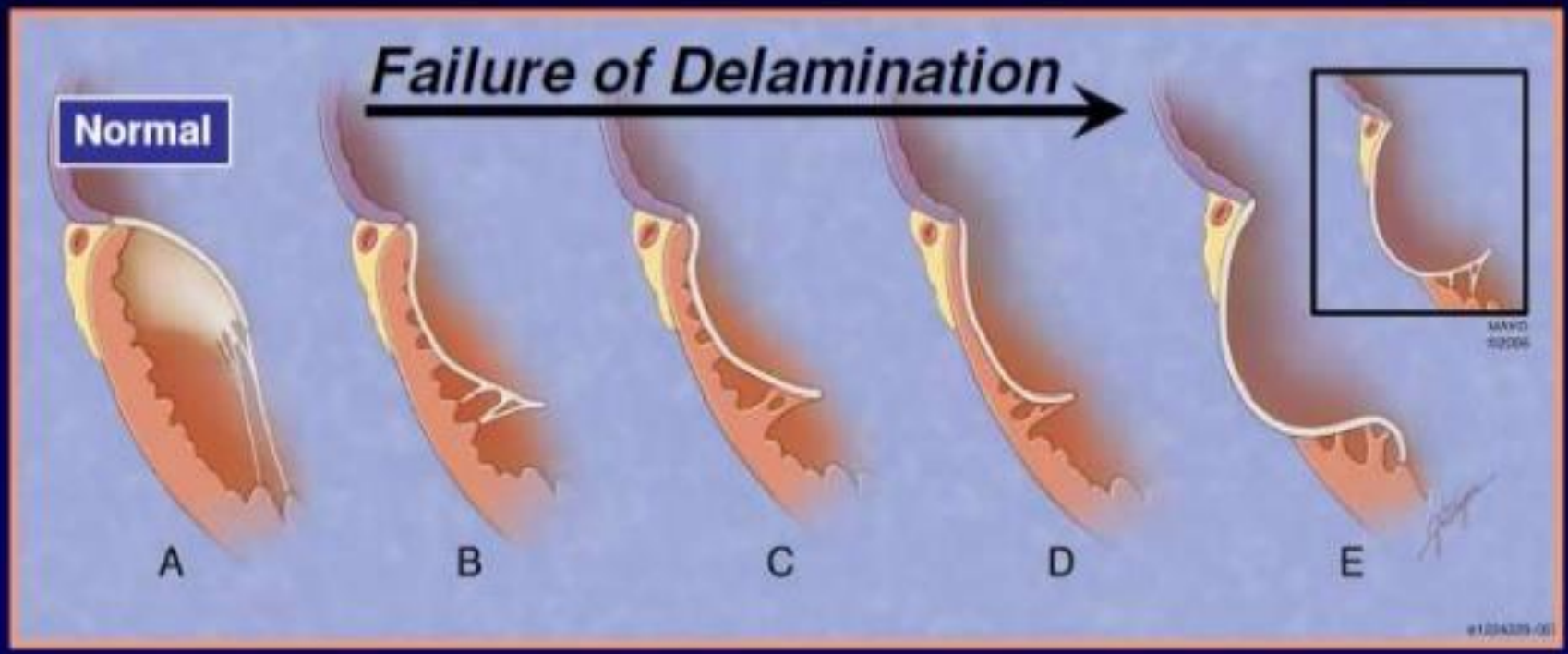
A pencil sketch of Wilhelm Ebstein published in the Festschrift celebrating Ebstein's 70th birthday. Published by permission of the Mayo Clinic Proceedings, where it was published by Mann RJ, Lie JT. The life story of Wilhelm Ebstein (1836–1912) and his almost overlooked description of a congenital heart disease.

Anatomy

- Maldevelopment of Tricuspid Valve
- *Embryologically*, TV is normally formed by delamination or exfoliation of inner layer of RV myocardium up towards the tricuspid annulus
- If this process is arrested or incomplete, the attachments of the leaflets are apically displaced

Ebstein's Anomaly

Right Ventricle & Tricuspid Valve



Spectrum with Infinite Variability

MAYO CLINIC

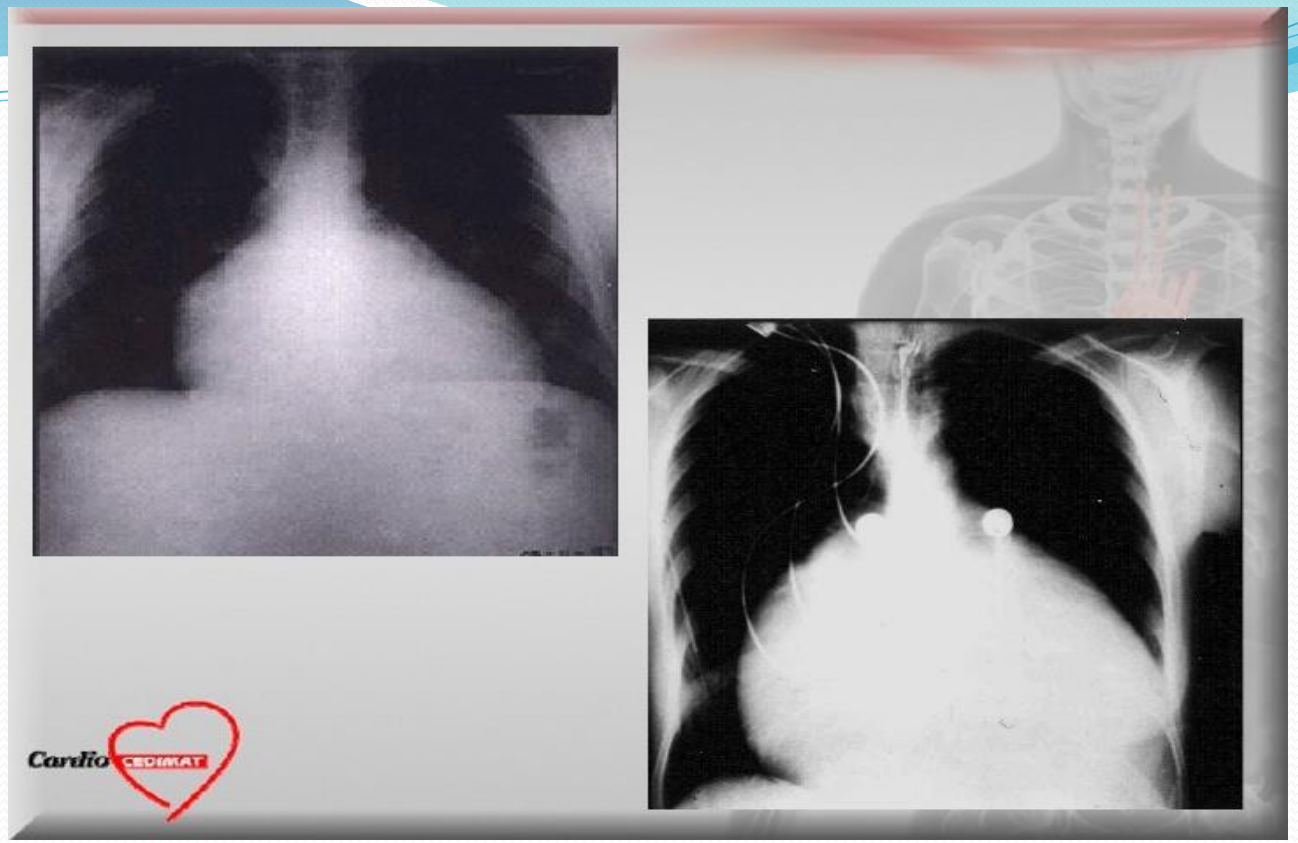
Manifestari clinice neonatale

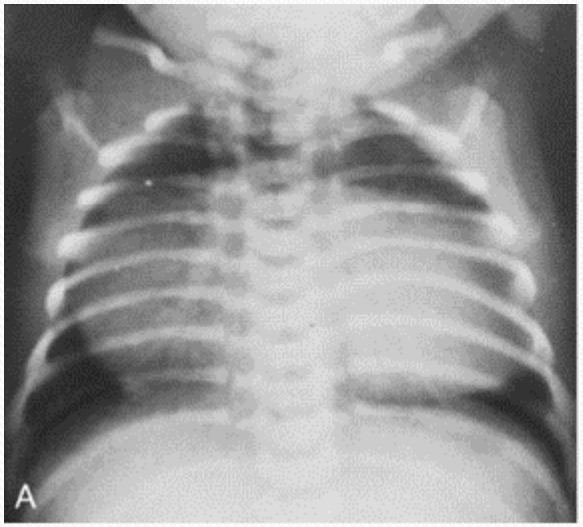
Debit pulmonar scazut datorita

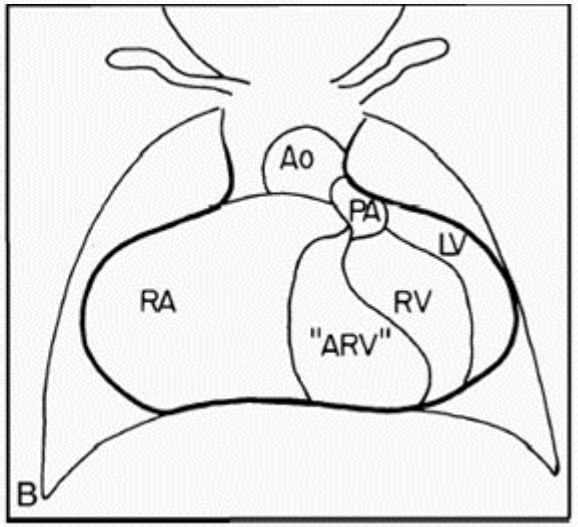
- IT severe
- Disfunctiei de VD
- RVP crescute postnatal

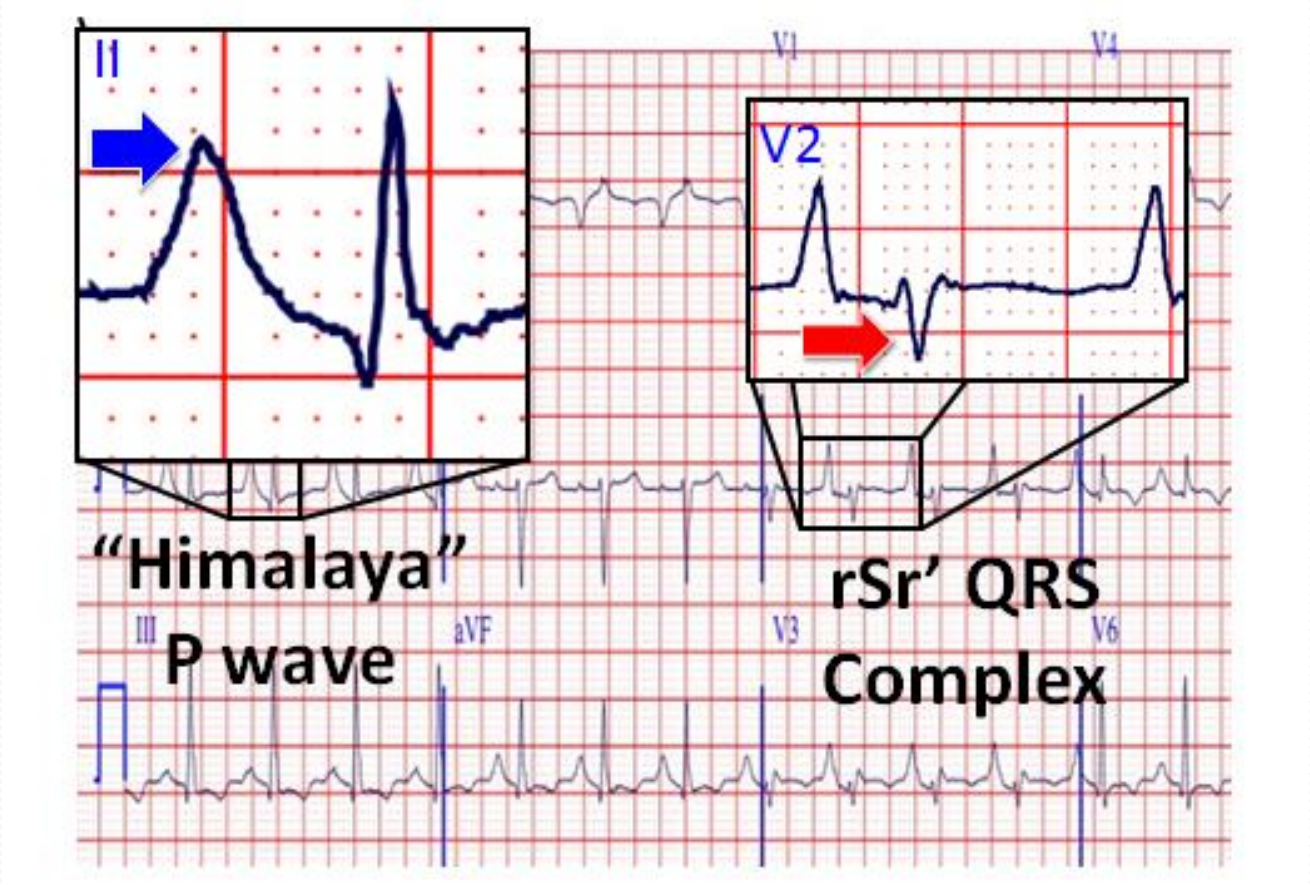
Cianoza
- datorita suntului dr-stg prin DSA

Evolutia se amelioreaza odata cu scaderea RVP


Cardio
CEDIMAT


A


Ao
PA
LV
RA
RV
"ARV"
B


II
V1
V4
V2
"Himalaya"
P wave
rSr' QRS
Complex
III
aVF
V3
V6

Ecografia

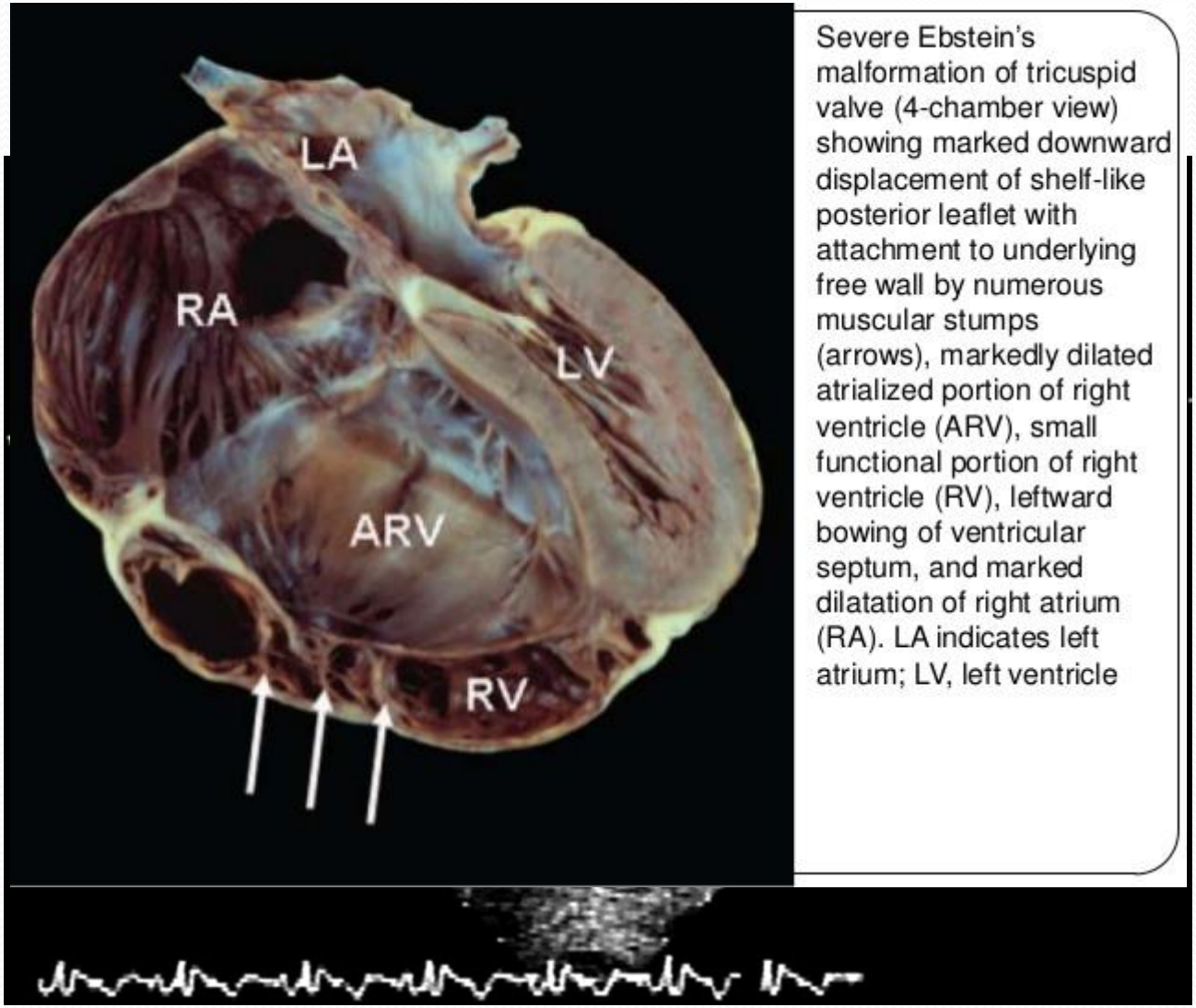
- Insertie apicalizata VT septala >8 mm/m2
- Morfologia si atasarea de sept a VT septala si anterioara (tethering)
- Coaptarea VT/gradul de IT
- Dilatarea AD
- Prezenta DSA/PFO
- Aprecierea functionala VD
- Asocierea cu alte malformatii

Echo for Ebstein's

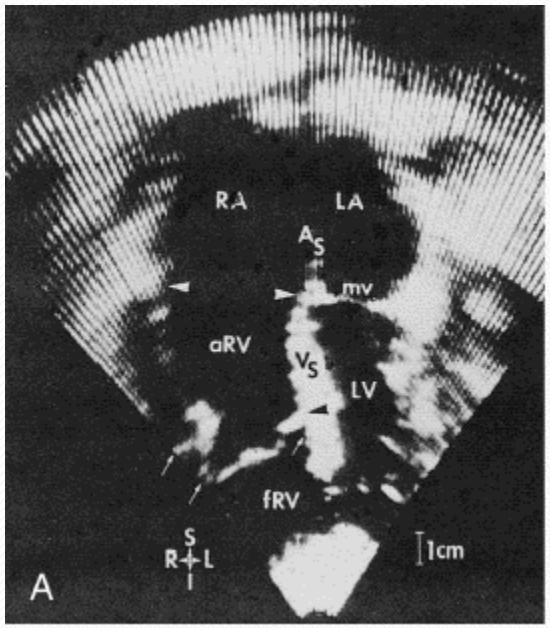
What the Surgeon Wants to Know?

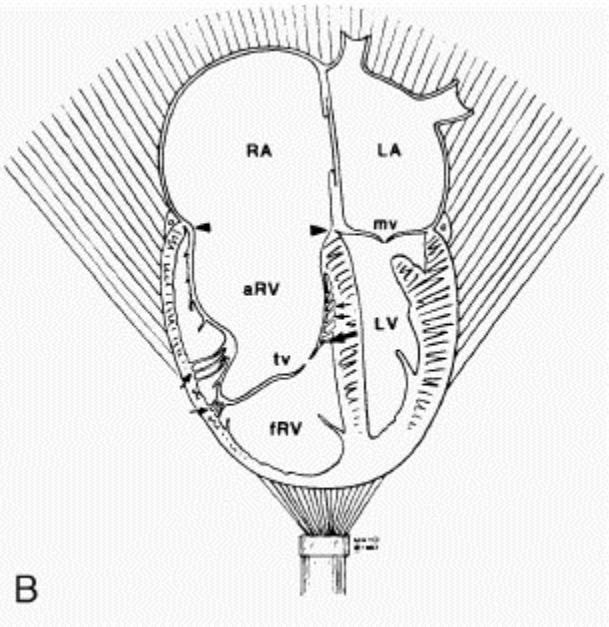
- Mechanism of TR - # of jets, location
- Delamination & tethering of each leaflet
- Status of leaflet edges
- How much annular dilatation
- Unsupported segments, fenestrations
- Size & function of RV & atrialized RV
- Ventricular septum & LV function

MAYO CLINIC

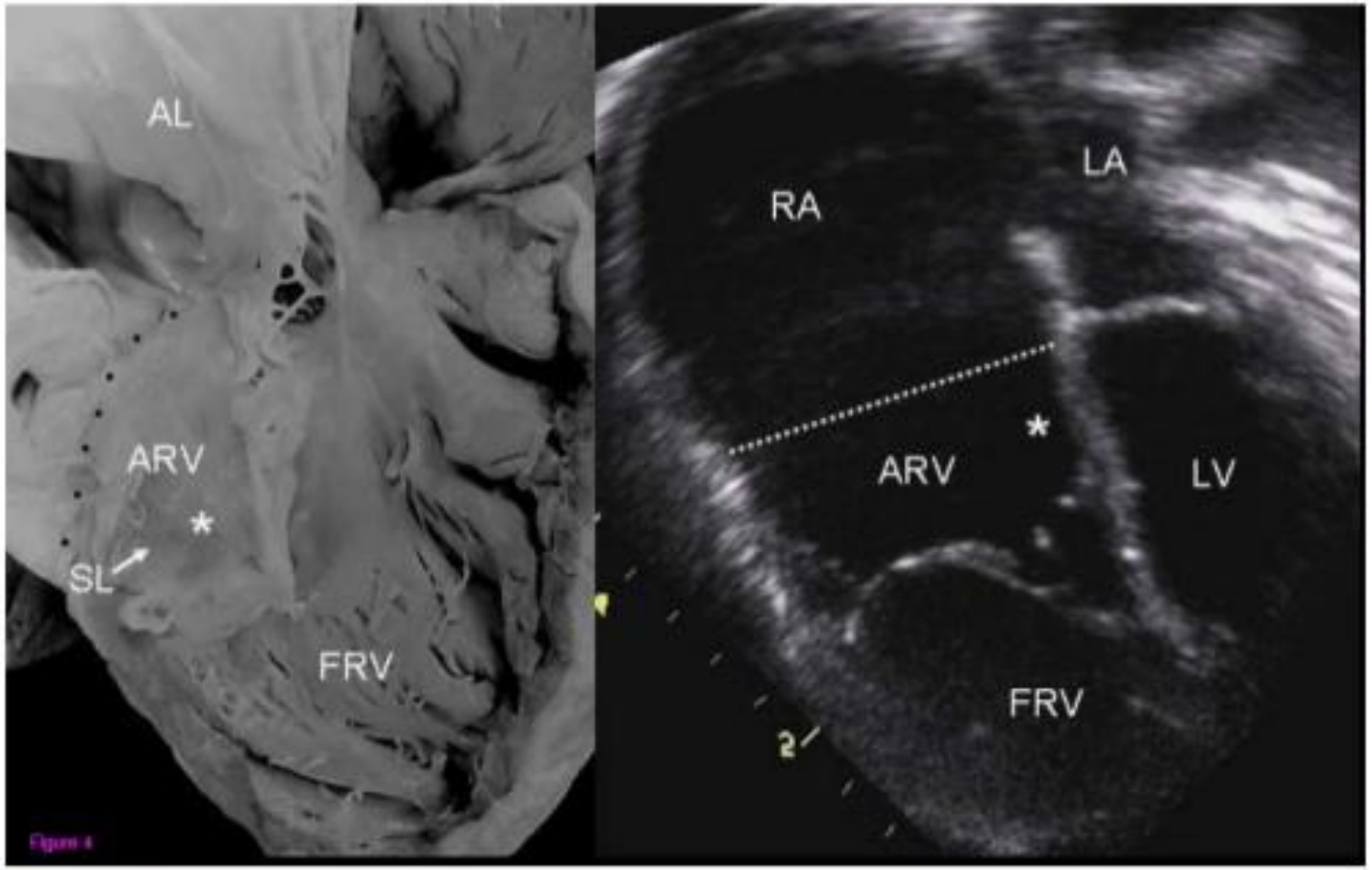


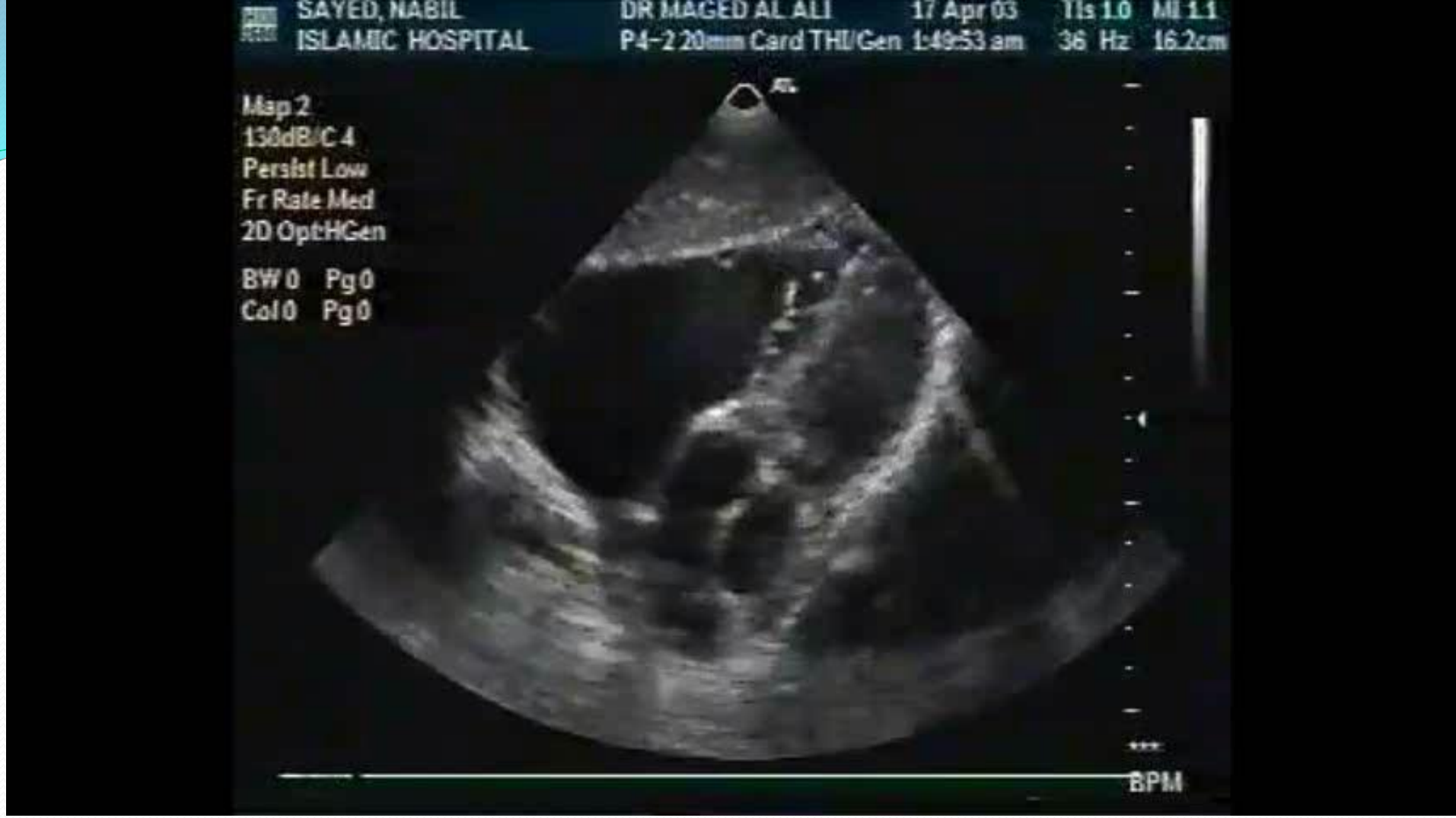
Severe Ebstein's malformation of tricuspid valve (4-chamber view) showing marked downward displacement of shelf-like posterior leaflet with attachment to underlying free wall by numerous muscular stumps (arrows), markedly dilated atrialized portion of right ventricle (ARV), small functional portion of right ventricle (RV), leftward bowing of ventricular septum, and marked dilatation of right atrium (RA). LA indicates left atrium; LV, left ventricle

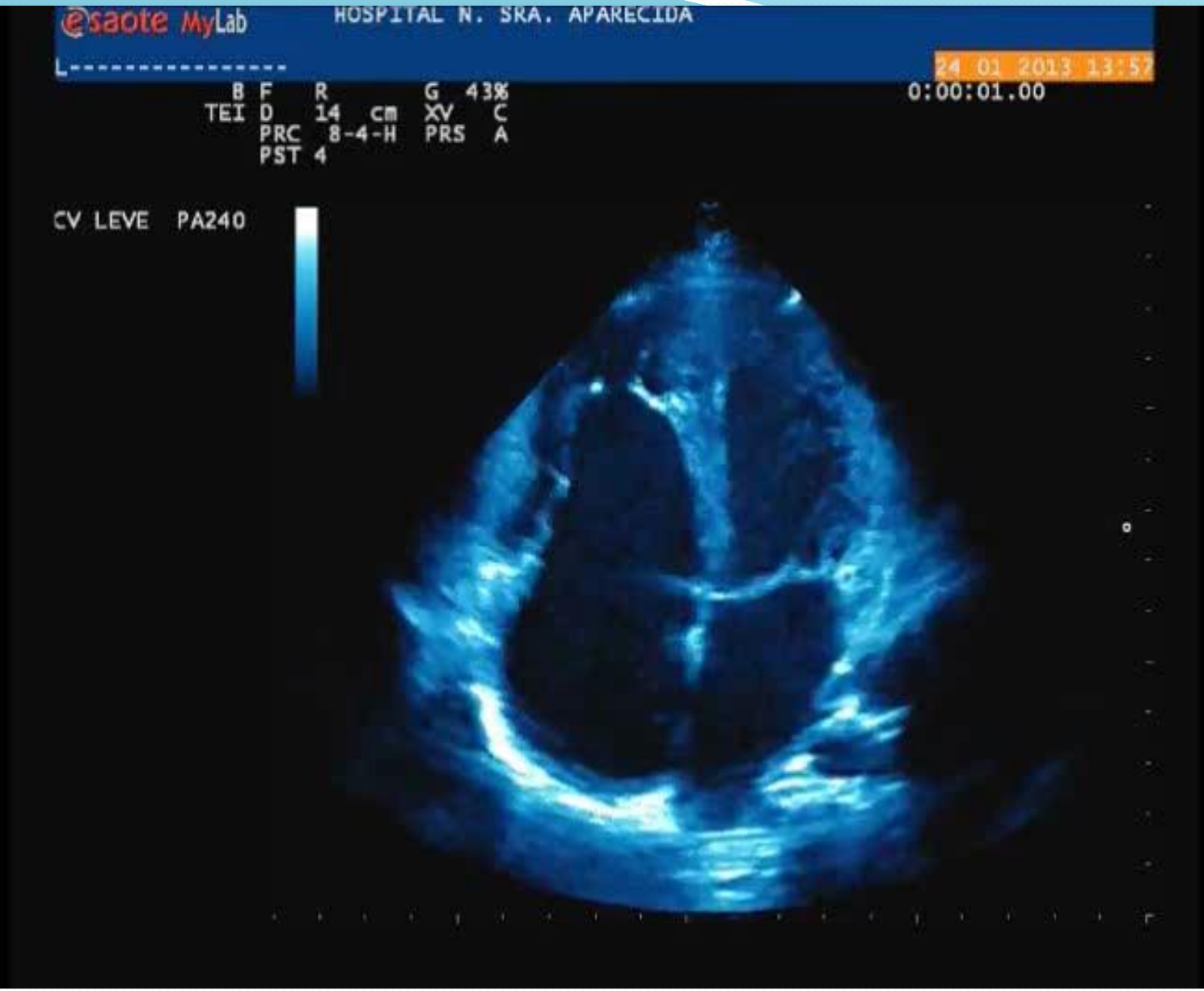

RA
LA
A_S
mv
aRV
V_S
LV
fRV
S
R+L
I
1cm
A

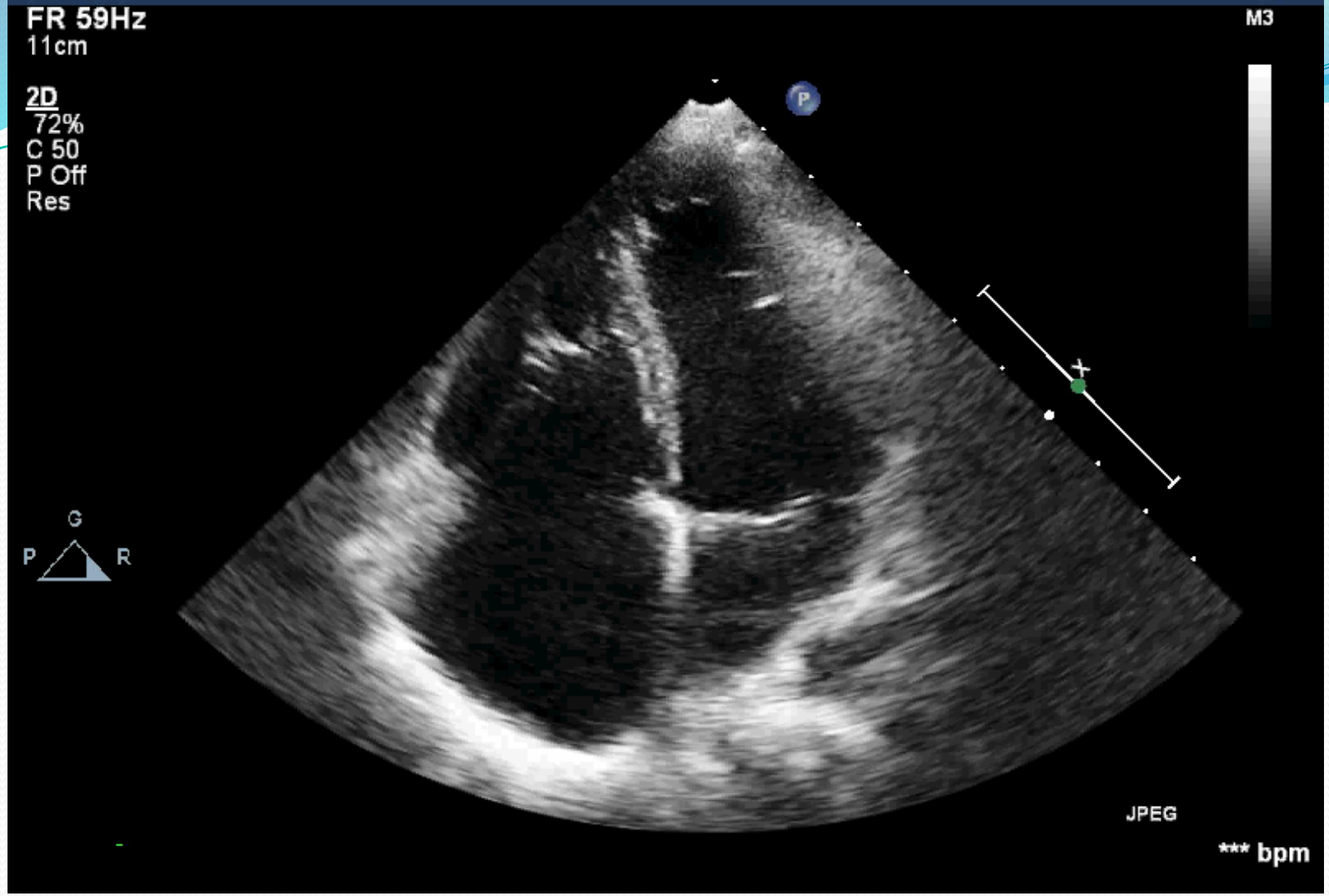

RA
LA
mv
aRV
LV
tv
fRV
B

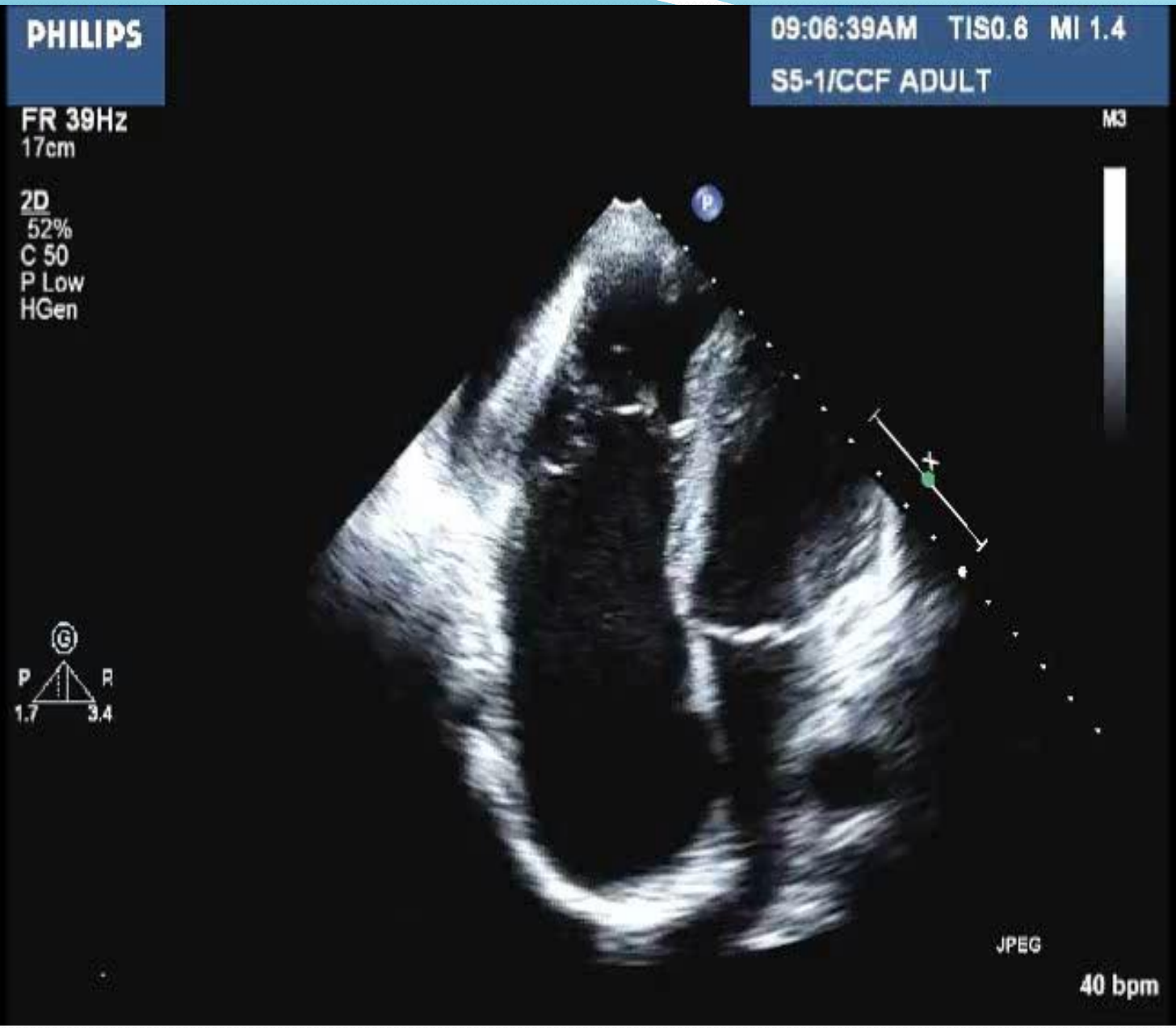
Anatomy

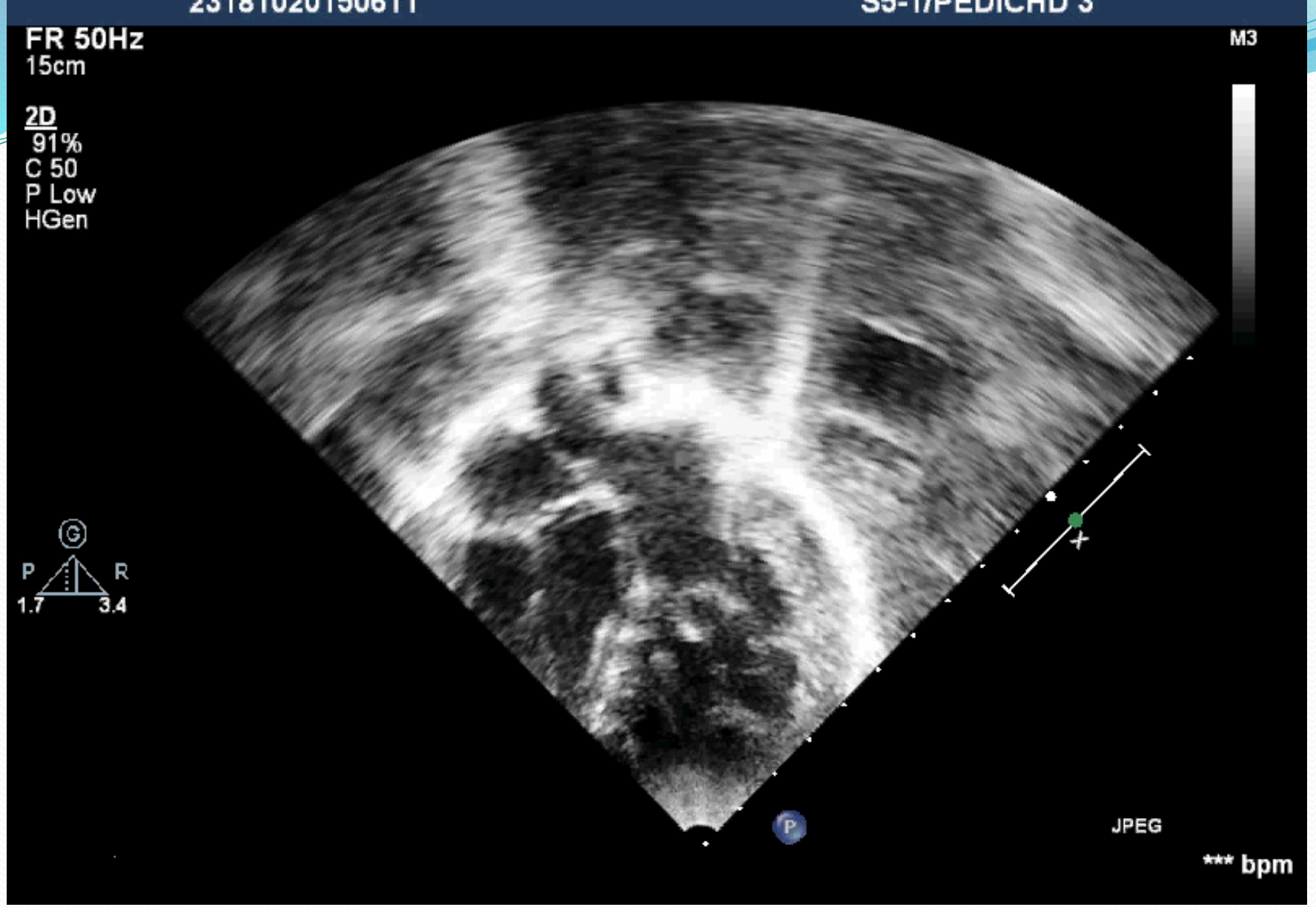



SAYED, NABIL
ISLAMIC HOSPITAL
DR MAGED AL ALI
P4-2 20mm Card THI/Gen
17 Apr 03
1:49:53 am
TIs 1.0
MI 1.1
36 Hz
16.2cm
Map 2
130dB/C 4
Persist Low
Fr Rate Med
2D Opt:HGen
BW 0 Pg 0
Col 0 Pg 0
BPM


esaote MyLab
HOSPITAL N. SRA. APARECIDA
24 01 2013 13:52
B F R G 43%
TEI D 14 cm XV C
PRC 8-4-H PRS A
PST 4
0:00:01.00
CV LEVE PA240


FR 59Hz
11cm
2D
72%
C 50
P Off
Res
M3
G
P
R
JPEG
*** bpm


PHILIPS
09:06:39AM TIS0.6 MI 1.4
S5-1/CCF ADULT
FR 39Hz
17cm
2D
52%
C 50
P Low
HGen
M3
P
R
1.7
3.4
JPEG
40 bpm


FR 50Hz
15cm
2D
91%
C 50
P Low
HGen
M3
G
P
R
1.7
3.4
JPEG
*** bpm

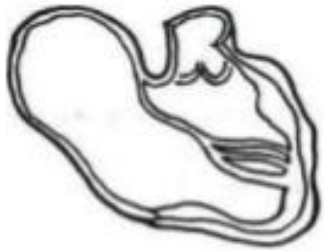
Carpentier's classification

- In 1988, Carpentier et al. proposed the following classification of Ebstein's anomaly -
- **Type A:** The volume of the true RV is adequate
- **Type B:** A large atrialized component of the RV exists, but the anterior leaflet of the TV moves freely
- **Type C:** The anterior leaflet is severely restricted in its movement and may cause significant obstruction of the RVOT
- **Type D:** Almost complete atrialization of the ventricle except for a small infundibular component.

EBSTEIN'S ANOMALY
(Carpentier's classification)

TYPE A (M+ C+)

TYPE B (M+ C -)



TYPE C (M- C -)

TYPE D ("tricuspid suck")

M – mobility *C - contractility*

Celermajer Index

- Celermajer et al. described an echocardiographic grading score for neonates with Ebstein's anomaly, the Great Ormond Street Echocardiography (GOSE) score, with grades 1 to 4.
- The ratio of the combined area of the RA and atrialized RV is compared to the functional RV and left heart. This classification is particularly helpful with neonatal Ebstein's anomaly.

- Grade 1: ratio <0.5
- Grade 2: ratio of 0.5 to 0.99
- Grade 3: ratio of 1.0 to 1.49
- Grade 4: ratio ≥ 1.5

Celermajer DS, Bull C, Till JA, et al. Ebstein's anomaly: presentation and outcome from fetus to adult J Am Coll Cardiol 1994;23:170–176

Evolutie naturala

- 18 % din nn simptomatici ,deces in perioada neonatala
- -30% deces pana la varsta de 10 a

- Cianoza,ICC,disfunctie VS,tulburari de ritm

Management

- Medical

-nn cianotici sever:

- PGE1,
- suport inotrop,
- vasodilatatoare pulmonare,OXID NITRIC
- corectia acidozei metabolice

Preop Mx of Critically ill neonates & infants

- *Hypoxia & Acidosis cause pulmonary vasoconstriction !!*

while

- *Hyperoxia & Alkosis causes pulmonary vasodilatation !!*

Preop Mx of Critically ill neonates & infants

- **Measures to reduce PVR**
  -Mechanical Ventilation -High FiO2/ Hyperventillation /PEEP
  -Sedation +Paralysis –avoids reflex ↑ in PVR secondary to noxious stimuli
  -Alkalosis-pulmonary vasodilator /pH7.50-7.60/ sodabicarb or THAM
  -Inhaled NO-starting dose 5-40ppm

- **PGE1 infusion-** to keep the ductus arteriosus patent & maintain the pulmonary blood flow
- **Correction of Metabolic Acidosis** –improves myocardial function
- **Inotropes**- Avoid Adrenaline as far as possible as it ↑es PVR
- **Avoid Overzealous Volume Infusions**-can cause further annular dilatation –worsen the TR

Management

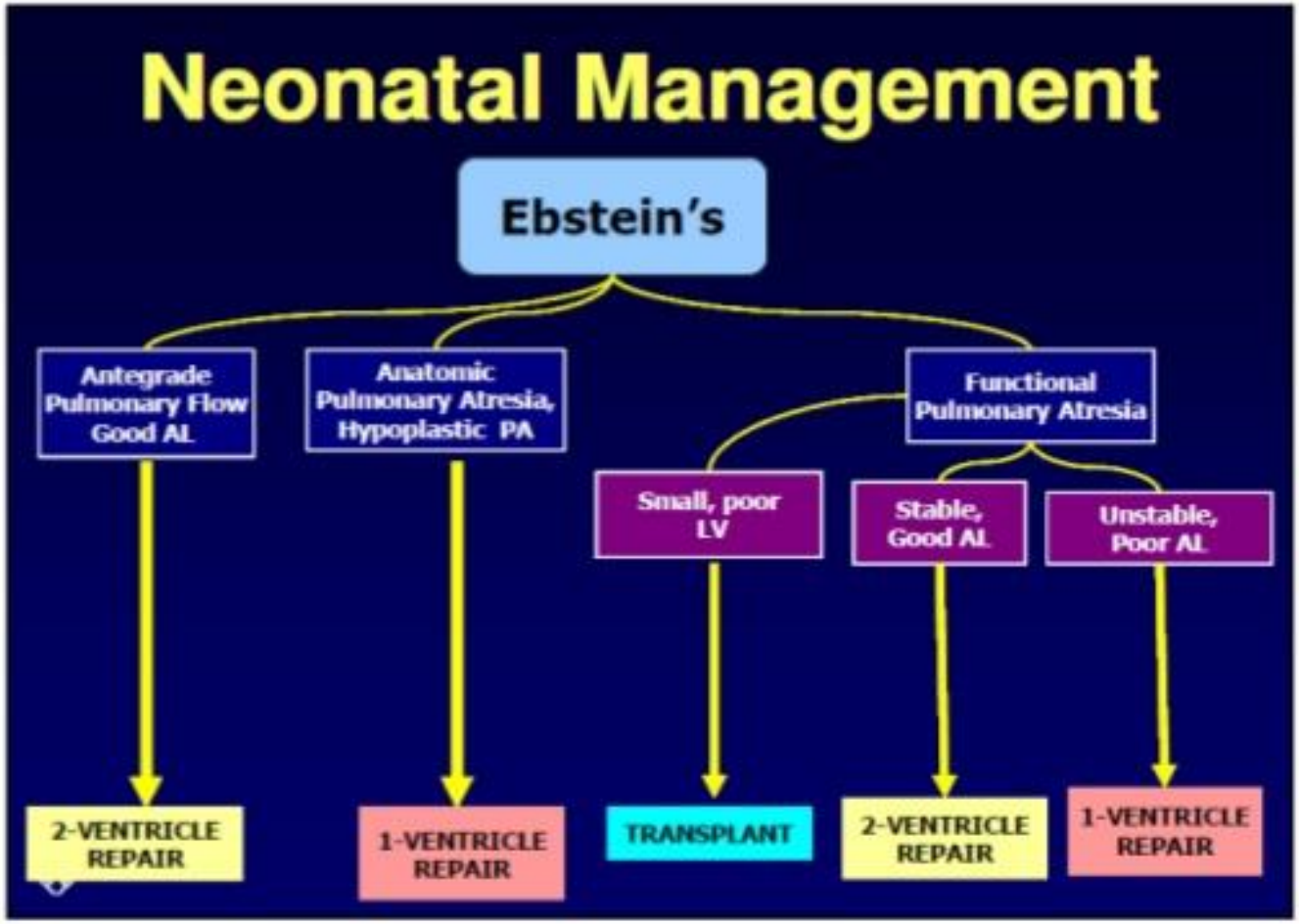
- Chirurgical

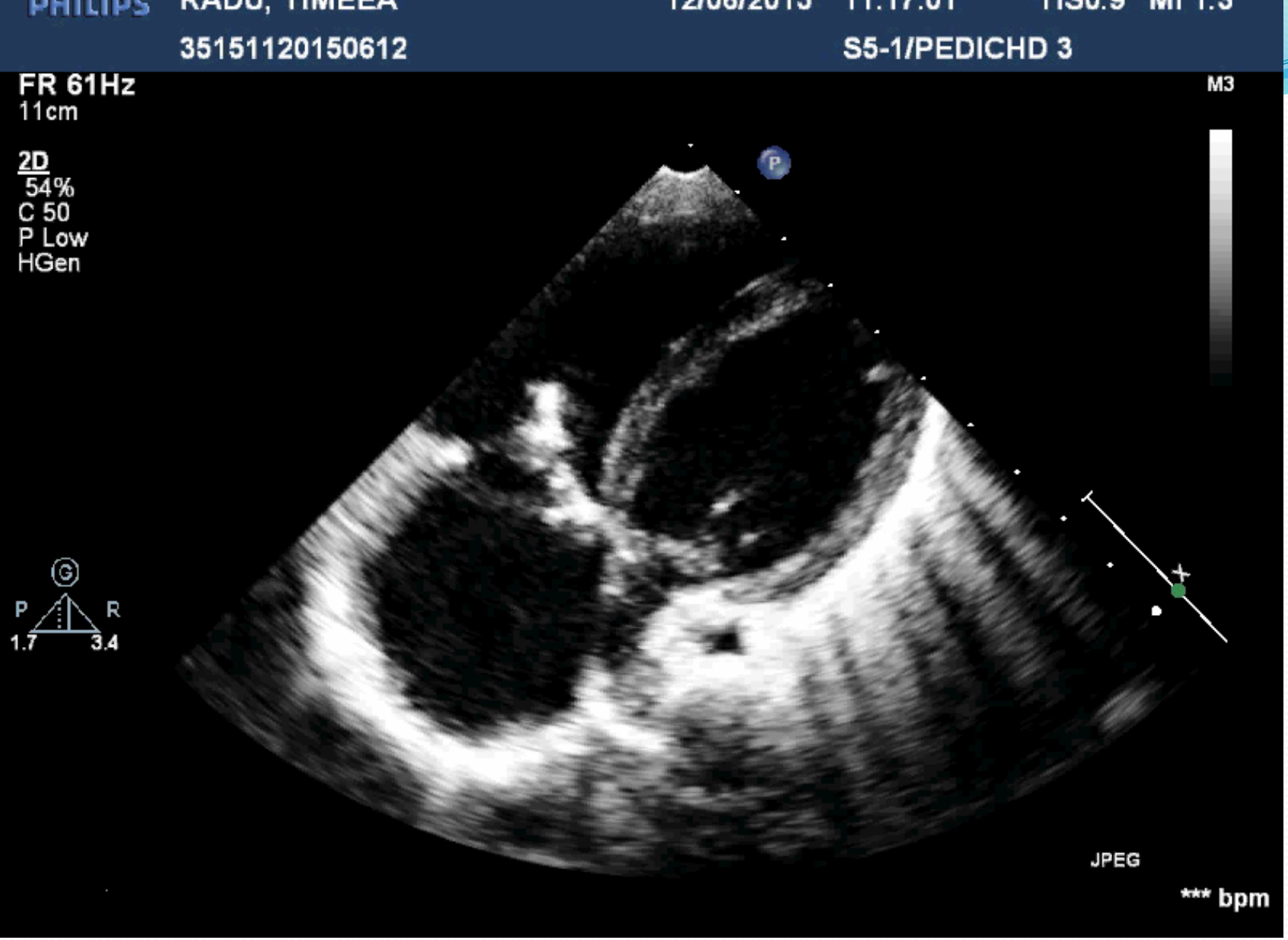
- ***Indicatie***:

1. Nn critici care in ciuda trat medical simptomatologia persista
2. Restrictia severa a activitatii fizice,clasa functionala III,IV
3. Cianoza severa,ICC
4. Aritmii severe-ablatia cu RF

- ***Abordare***

1. -inchidere DSA/chirurgia VT
2. -protezare VT
3. Proceduri Fontan


Neonatal Management
Ebstein's
Antegrade Pulmonary Flow Good AL
Anatomic Pulmonary Atresia, Hypoplastic PA
Functional Pulmonary Atresia
Small, poor LV
Stable, Good AL
Unstable, Poor AL
2-VENTRICLE REPAIR
1-VENTRICLE REPAIR
TRANSPLANT
2-VENTRICLE REPAIR
1-VENTRICLE REPAIR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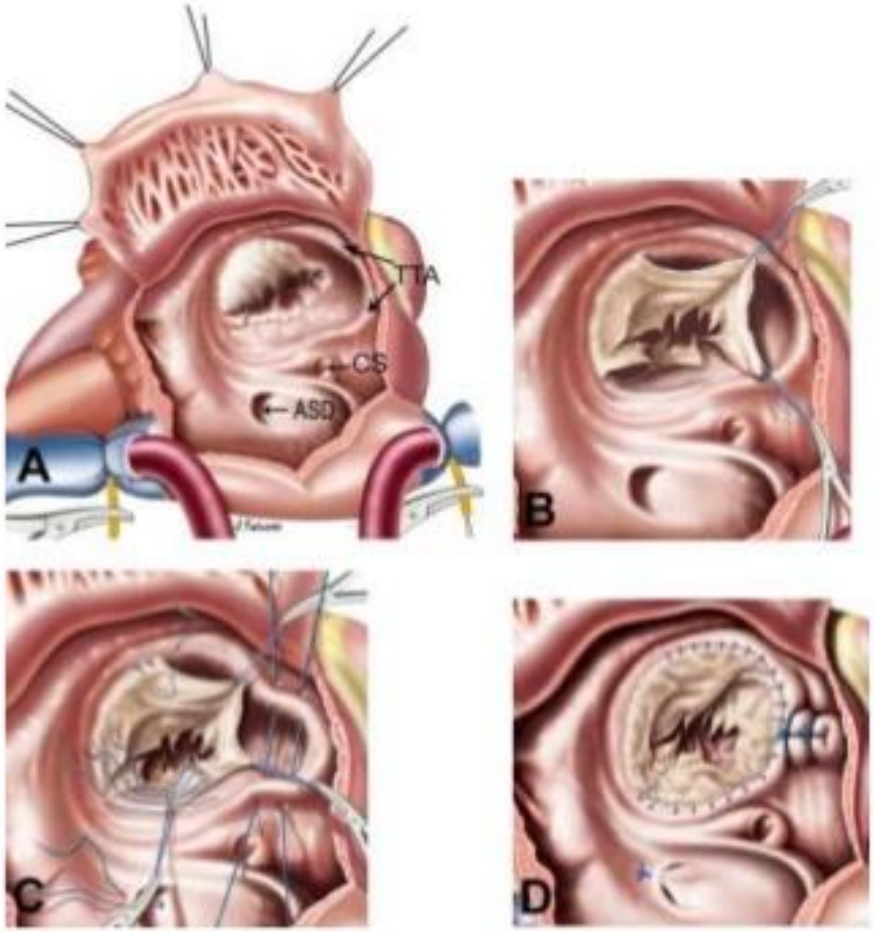
Operative steps for Ebstein's anomaly repair



da Silva J. P. et al.; J Thorac Cardiovasc Surg 2007;133:215-223

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