The Fontan circulation

Folkert Meijboom

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What to expect?

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Why a Fontan-circulation
- *Indications*
- *How does it work*

Types of Fontan circulation
- *Historical overview*

Role of echocardiography

What to expect?

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Why a Fontan-circulation

Indications

How does it work

Types of Fontan circulation

Historical overview

Role of echocardiography

Why a Fontan circulation?

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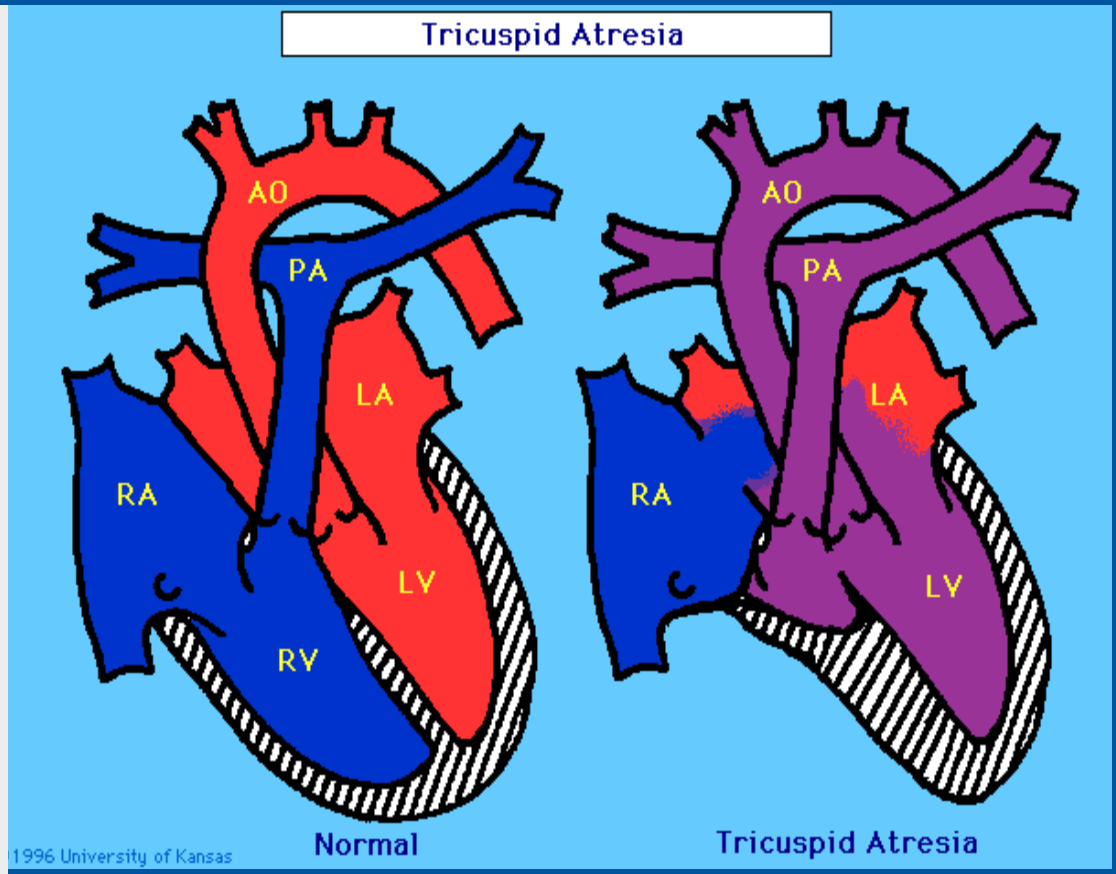
- No functional RV
- Volume overload LV
- systemic venous return plus pulmonary venous return
- Desaturation systemic circulation - cyanosis



- How van this be solved?

Why a Fontan circulation?

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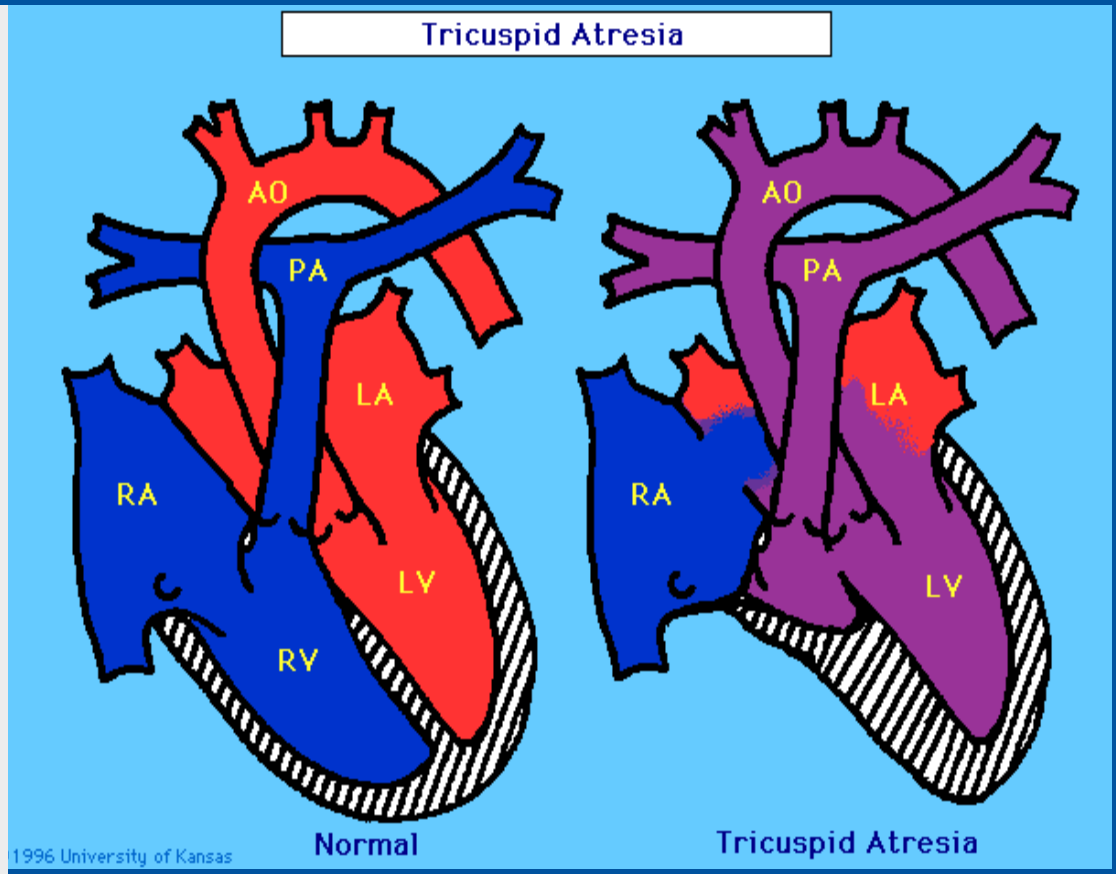

Not by a biventricular repair

→ Univentricular repair

Indication for a Fontan

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situations for which a biventricular is not possible

Indication for a Fontan

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situations for which a biventricular is not possible

- HLHS
- TA
- MA
- DILV
- Unbalanced AVSD
- Ebstein
- PA – intact IVS
- etc

The essence of the Fontan circulation

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- only 1 functional ventricle → systemic ventricle

The essence of the Fontan circulation

Universitair Medisch Centrum Utrecht

- only 1 functional ventricle → systemic ventricle
- no subpulmonary ventricle

The essence of the Fontan circulation

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The essence of the Fontan circulation

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- no subpulmonary ventricle
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- no desaturated blood in the systemic circulation

The essence of the Fontan circulation

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- only 1 functional ventricle → systemic ventricle
- no subpulmonary ventricle
- Systemic venous return flows, without a pumping ventricle, into the lungs
- no desaturated blood in the systemic circulation
- No longer a volume-overloaded LV

How does it work?

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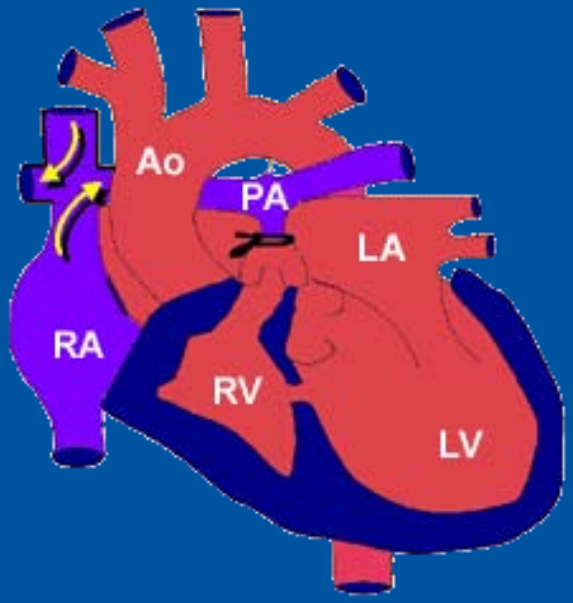
How can an adequate pulmonary blood flow be achieved?



1. elevated systemic venous pressure
2. low pulmonary vascular resistance
3. Low LA pressures

How does it work?

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Flow

depends on

pressure difference

between

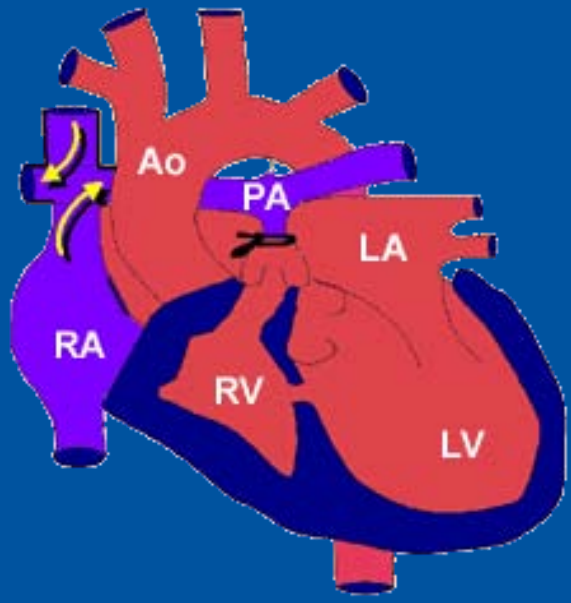
systemic venous pressure

and

LA pressure

and

pulmonary vascular resistance



Pressure difference VCS - LA

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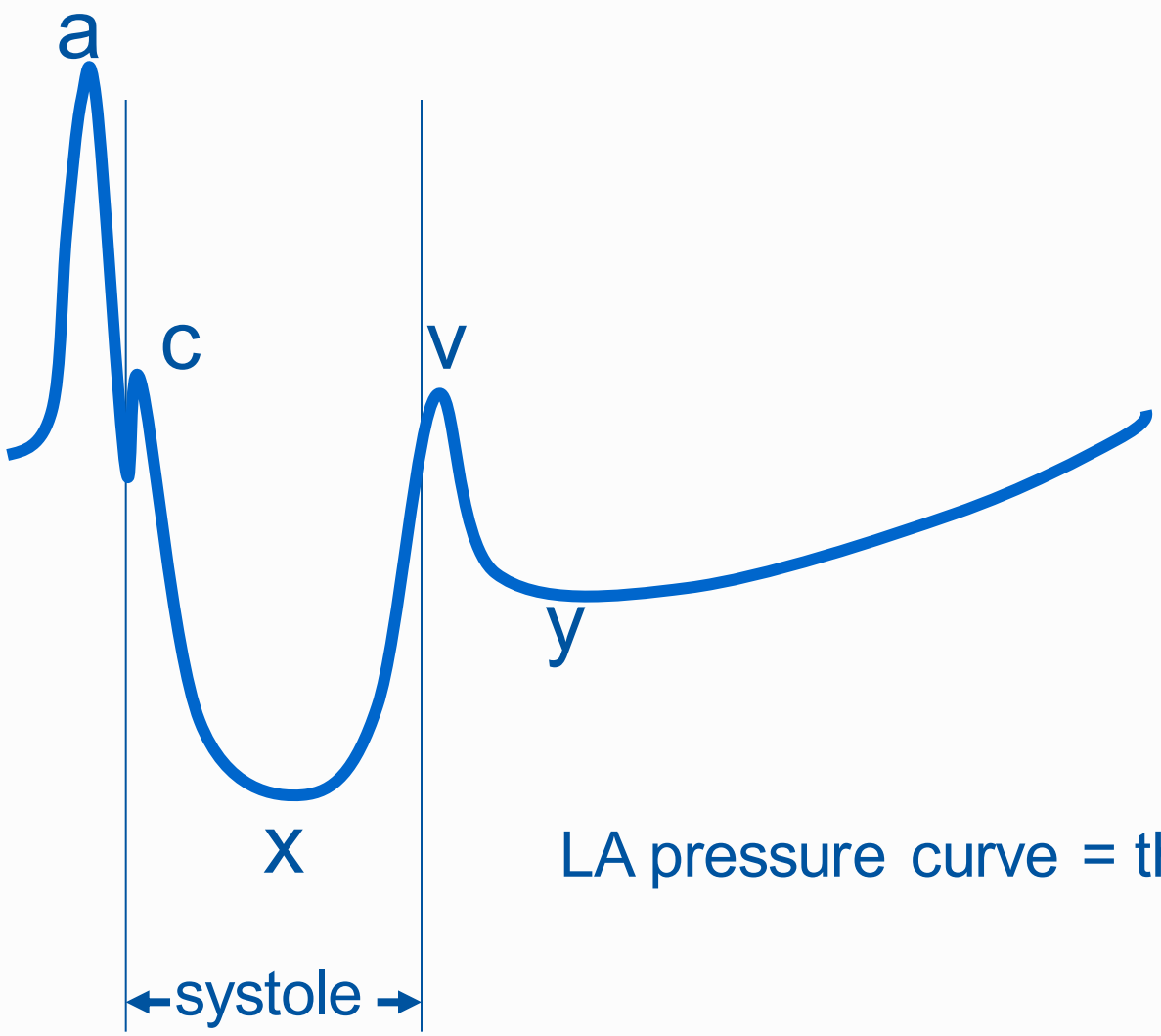
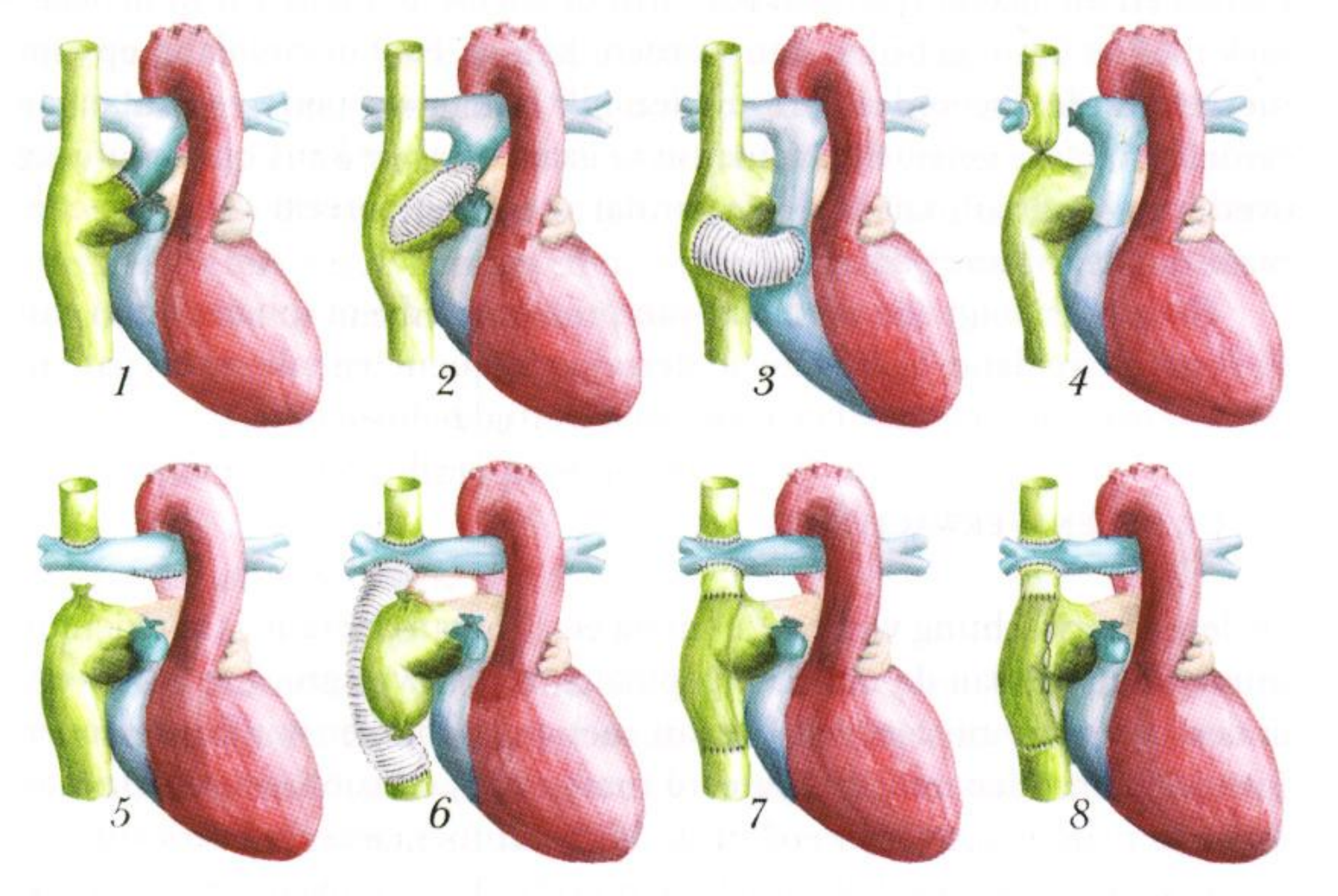
- **Push & pull** circulation:
  - *Elevated systemic venous pressure = push*
  - *Periodic lowering of LA pressure* } pull
    - During systole
    - During rapid filling of ventricle



LA pressure curve = the “pull”

Anatomy (history) of the Fontan-operation



“the” patient with a Fontan circulation

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- Does not exist

“the” Fontan circulation

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- Mixed bag
- many different types of
  - *Intracardiac anatomy*
  - *Type of surgical repair*

Echocardiography & Fontan

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- Know what the underlying cardiac defect is
- Know what type of Fontan repair is done
- Understand the Fontan circulation

Echocardiography & Fontan

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- Know what the underlying cardiac defect is
- Know what type of Fontan repair is done
- Understand the Fontan circulation

- Read the surgical report!!

Echocardiography & Fontan

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- Know what the underlying cardiac defect is
- Know what type of Fontan repair is done
- Understand the Fontan circulation

- If you do not know where you are looking for, you may not find it

Echocardiography & Fontan

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- in univentricular hearts – **before surgery** – echo is important for:
  - *Intracardiac anatomy*
  - *Ventricular function*
  - *AV valve function*
  - *Biventricular - univentricular repair*

Echocardiography & Fontan

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- Univentricular hearts **after** Fontan
  - *(Intracardiac anatomy)*
  - *Ventricular function*
  - *AV valve function*

Echocardiography & Fontan

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Anatomy & function of the Fontan circulation

Echocardiography & Fontan

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Anatomy & function of the Fontan circulation

ventricular function & Fontan

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ventricular & AV valve function

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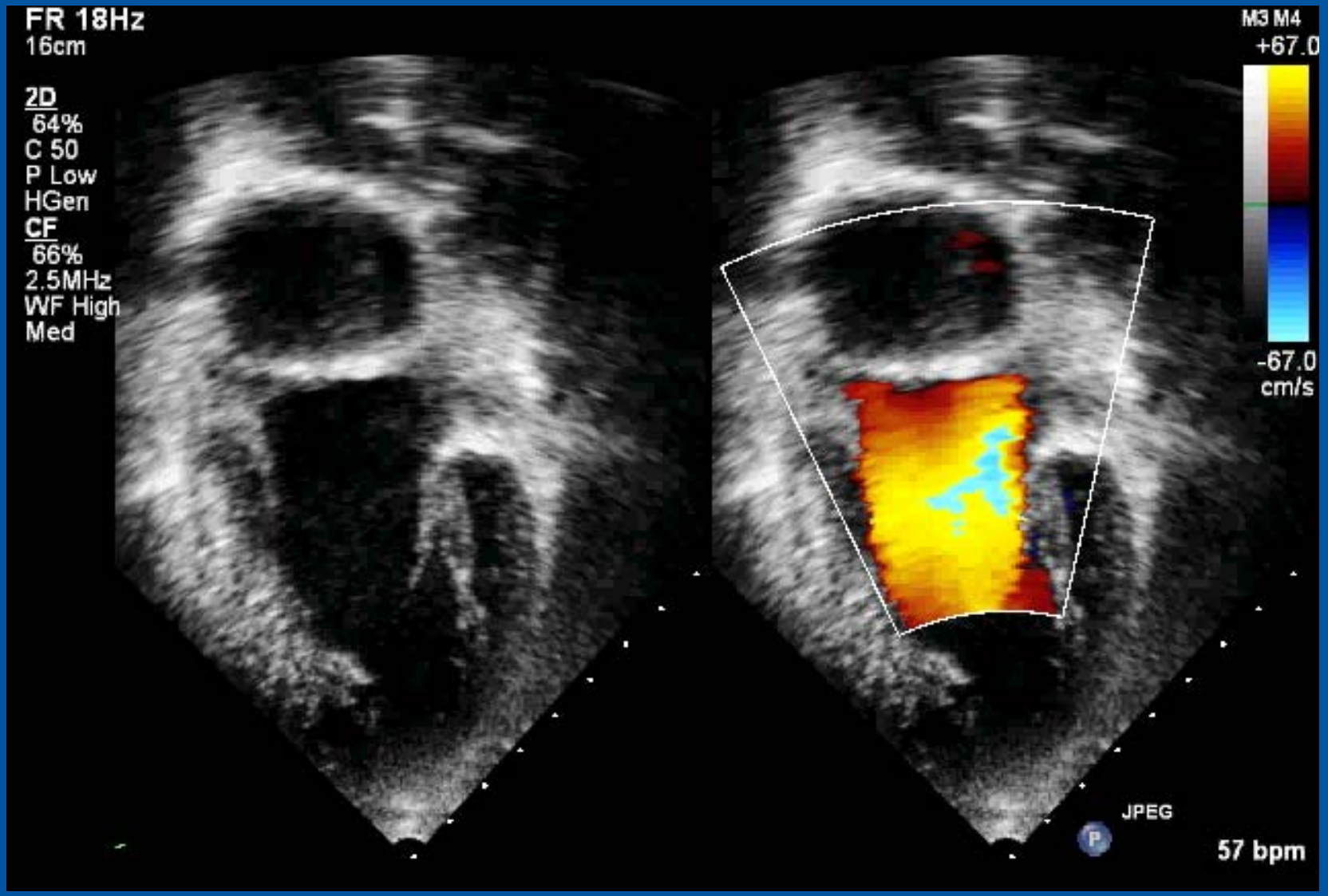

ventricular & AV valve function

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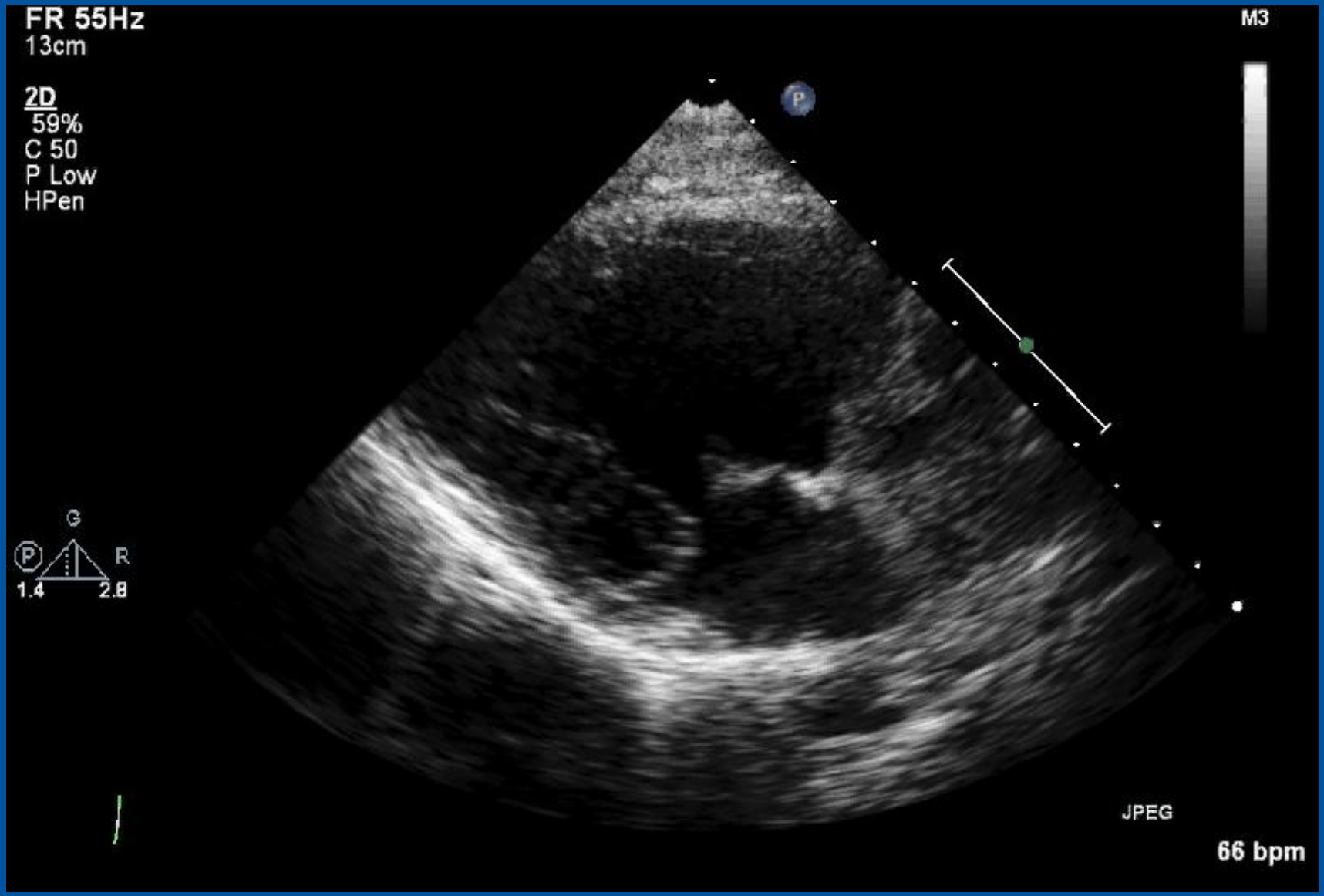

ventricular & AV valve function

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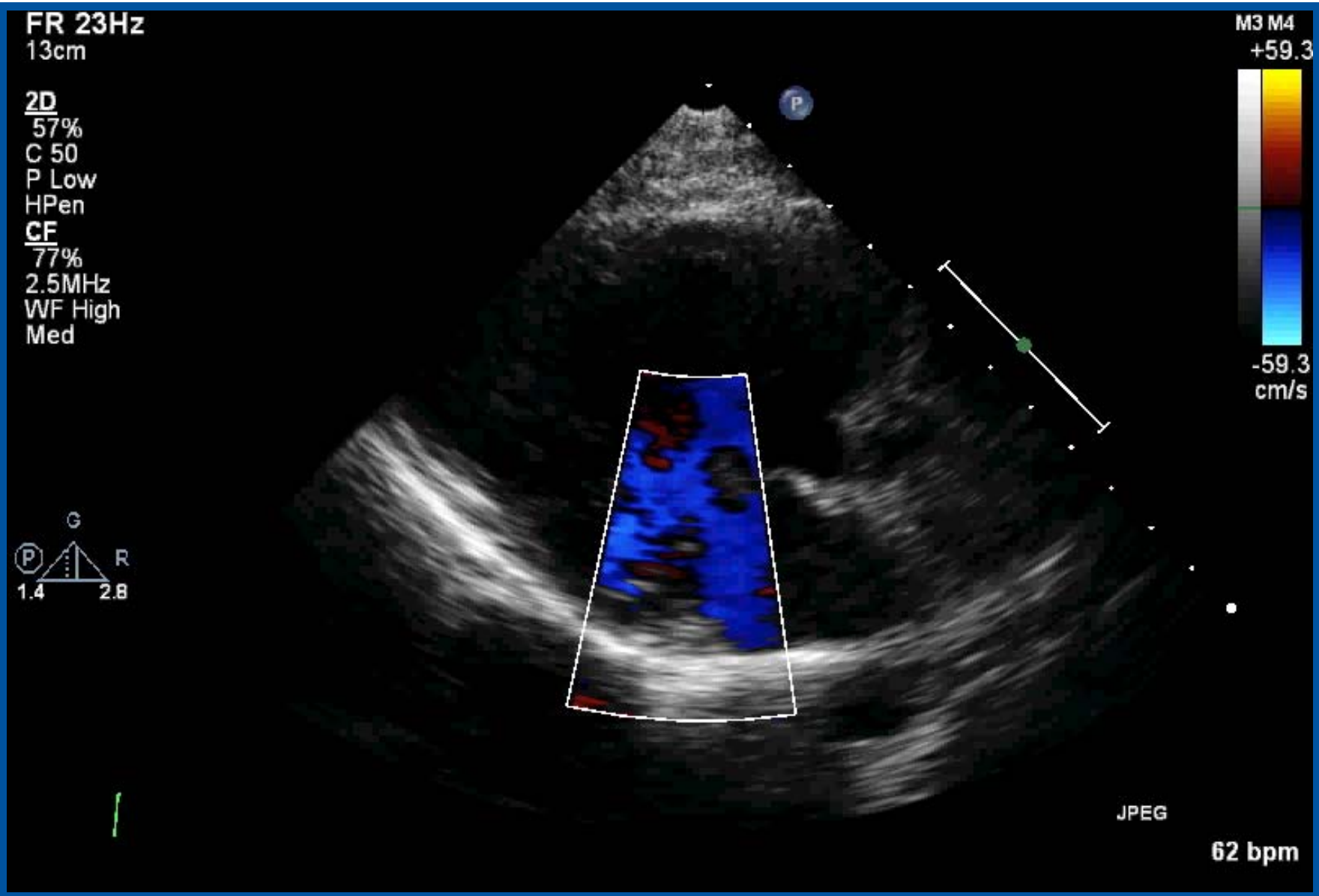

AV valve function

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- Assessment of AV valve regurgitation = very important

  Moderate to severe regurgitation

  - → *elevated LA pressure*
  - → *decrease in transpulmonary gradient*
  - → *decrease in transpulmonary flow*
  - → *decrease in cardiac output*
  - → *compensatory mechanism = fluid retention*
  - → *elevated central venous pressure*
  - → *transpulmonary gradient restored*
  - → *cardiac output restored*

AV valve function

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  - → *decrease in cardiac output*
  - → *compensatory mechanism = fluid retention*
  - → *elevated central venous pressure*
  - → *transpulmonary gradient restored*
  - → *cardiac output restored*
    - To a certain limit → decompensation

Fontan & AV valve function

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- Assessment of AV valve regurgitation = very important
- If one sees development of AV valve regurgitation
  - *Alarm signs*
  - *Worsening ventricular function*
- How to assess AV valve regurgitation: beyond the scope of this talk

Fontan & ventricular function

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How do we assess & measure function of the systemic ventricle?

Fontan & ventricular function

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- EAE & ASE recommendations on chamber quantification
- LV = systemic ventricle
  - *Standardized cross sections*
    - 4Ch view
    - Parasternal long axis
  - *Normal values for sex and body size*

Fontan & ventricular function

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- EAE & ASE recommendations on chamber quantification
  - *Standardized cross sections???*
    - 4Ch view
    - Parasternal long axis
  - ~~*Normal values for sex and body size*~~
- But embrace the idea behind it:
  - *Use reproducible cross section*
  - *Anatomic landmarks*
    - DILV ≠ HLFS ≠ unbalanced AVSD

Fontan &ventricular function

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Eyeballing

Quantify what you can quantify, in a reproducible way

Essential for longitudinal follow-up

- M-mode
- 2D
- 3D
- Doppler (incl TDI)

not for comparsion with normal values; patient as his/her own control

2D & color Doppler

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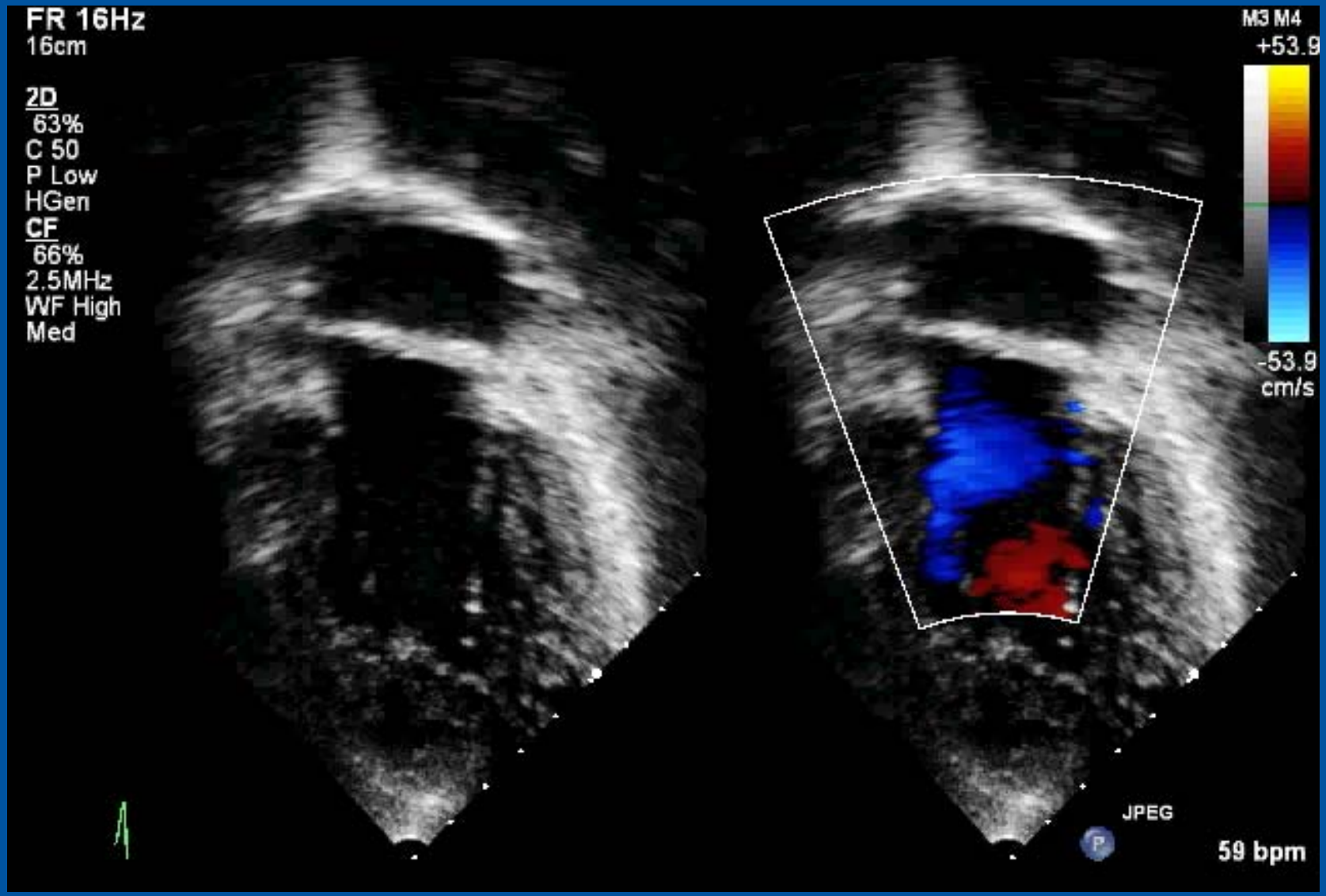

Inflow

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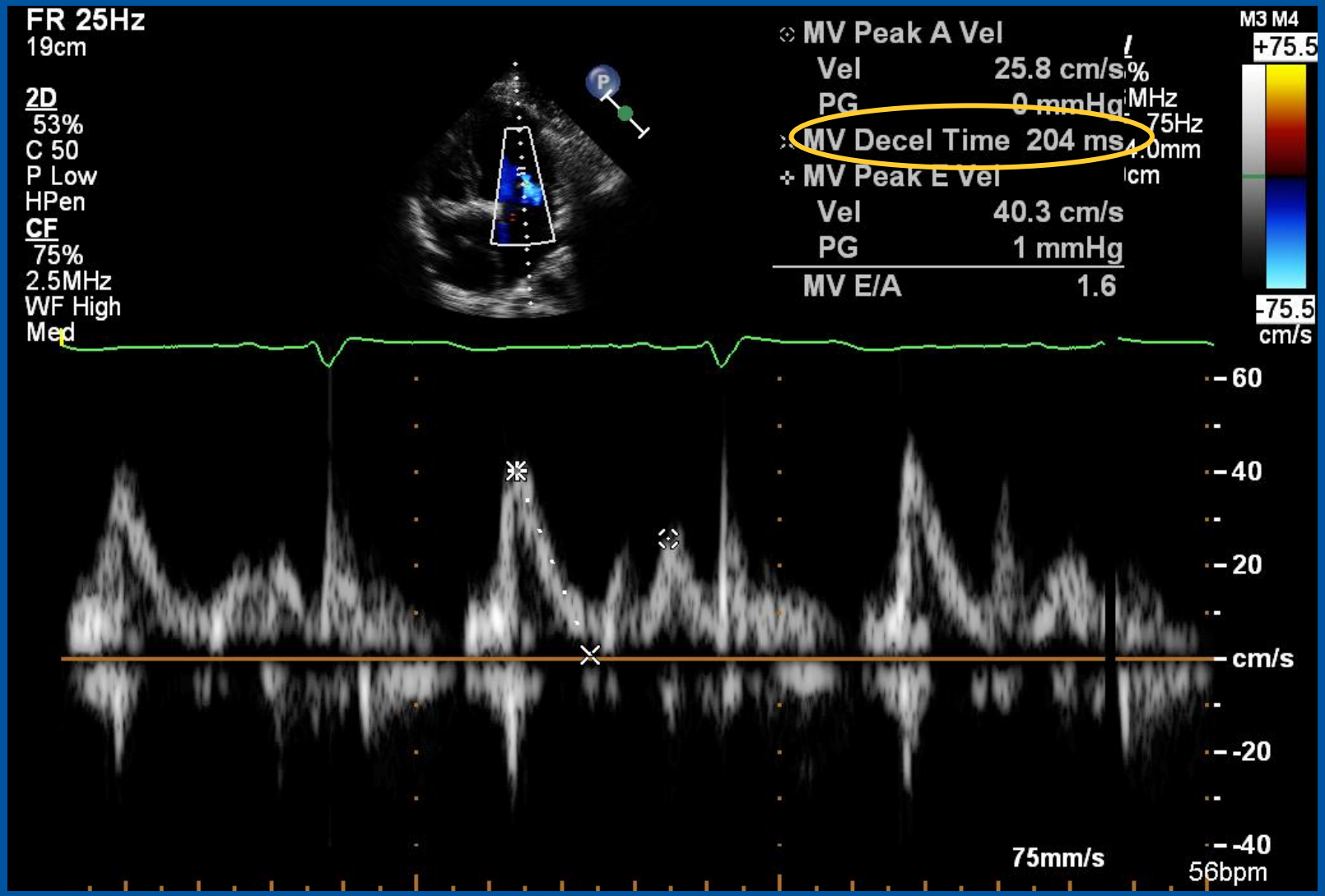

Tissue Doppler

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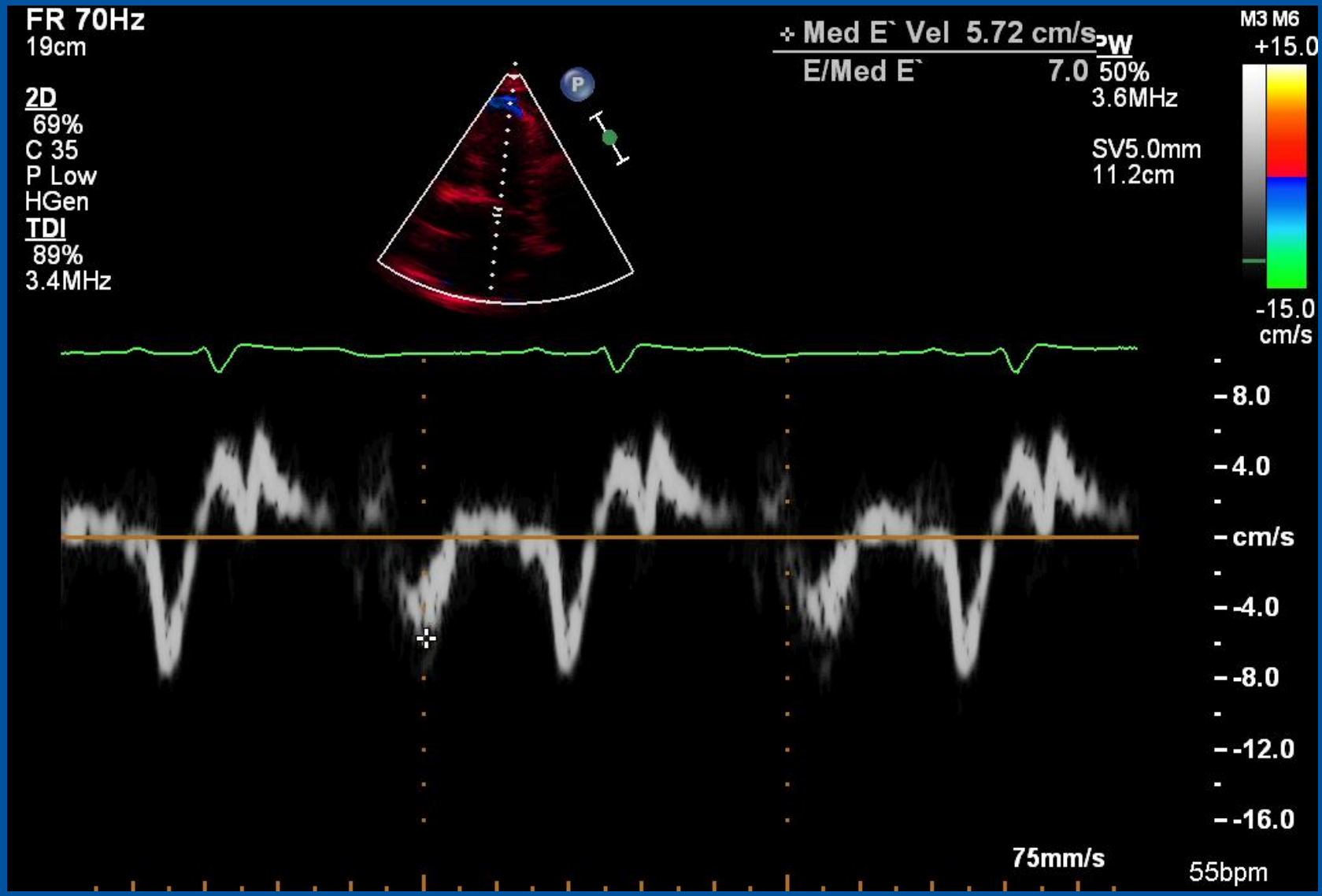

Ejection time, Tei-index

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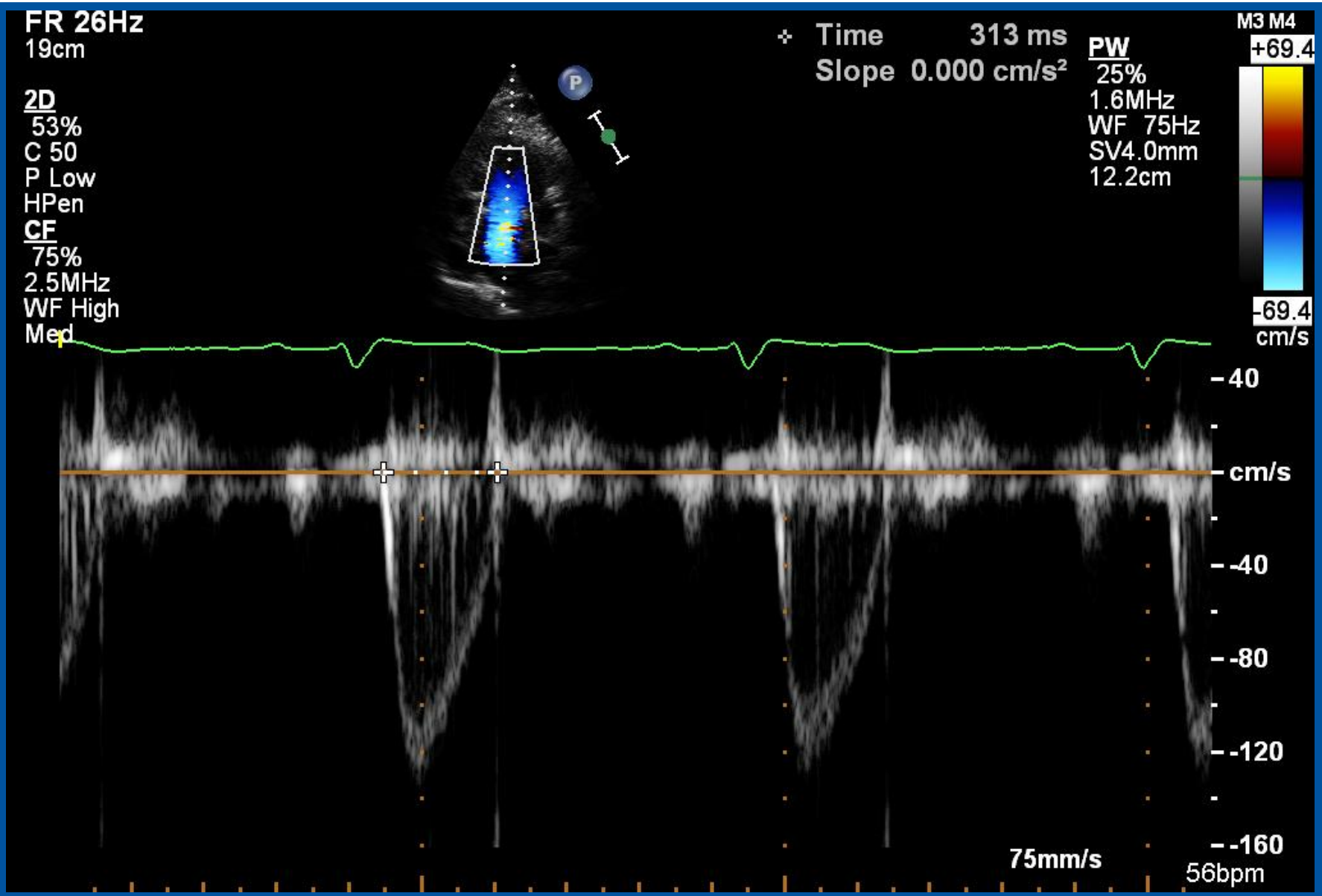

Relative duration systole in cardiac cycle (good functioning adult Fontan)

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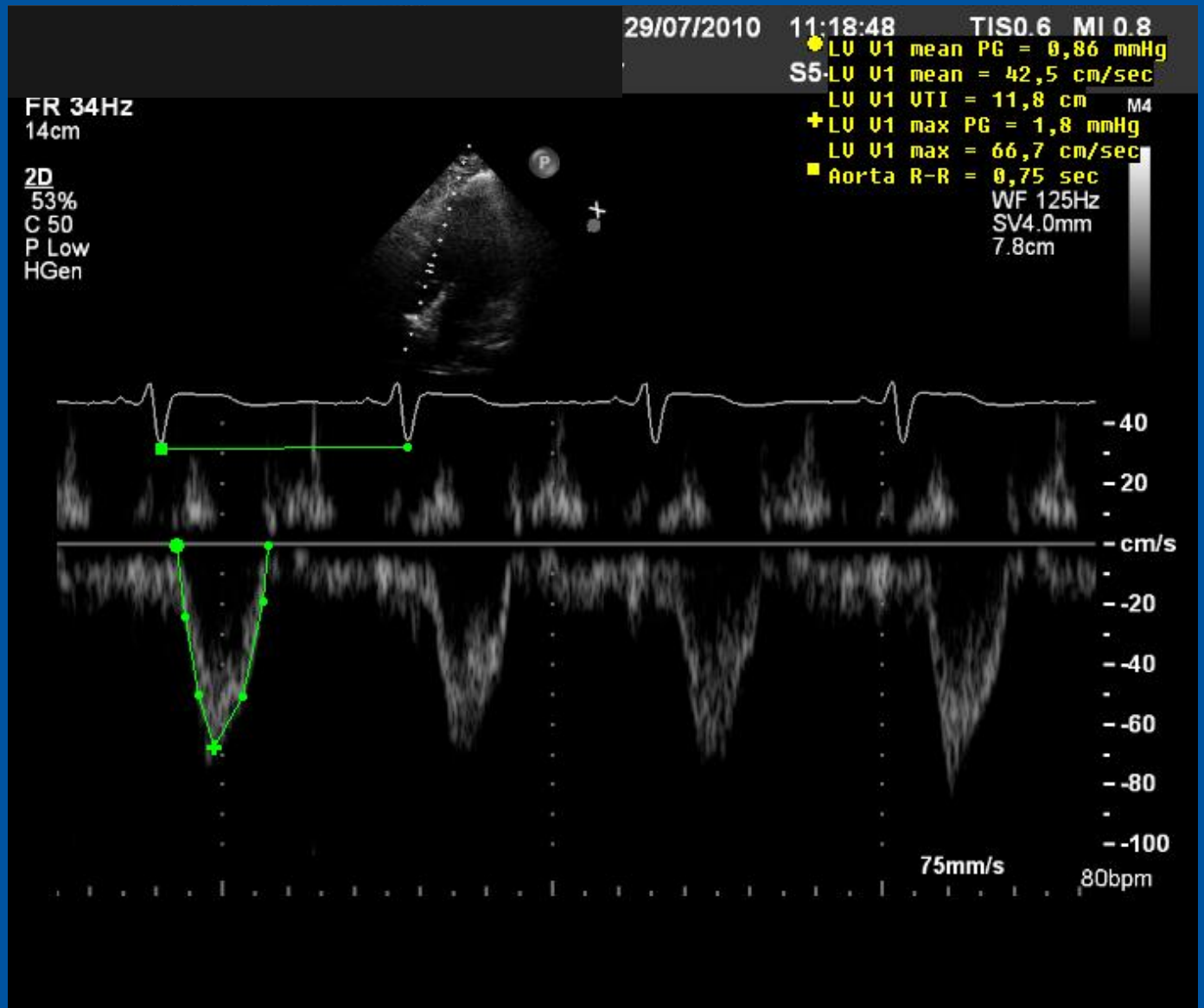

duration systole, incl IVCT & IVRT (poorly functioning adult Fontan)

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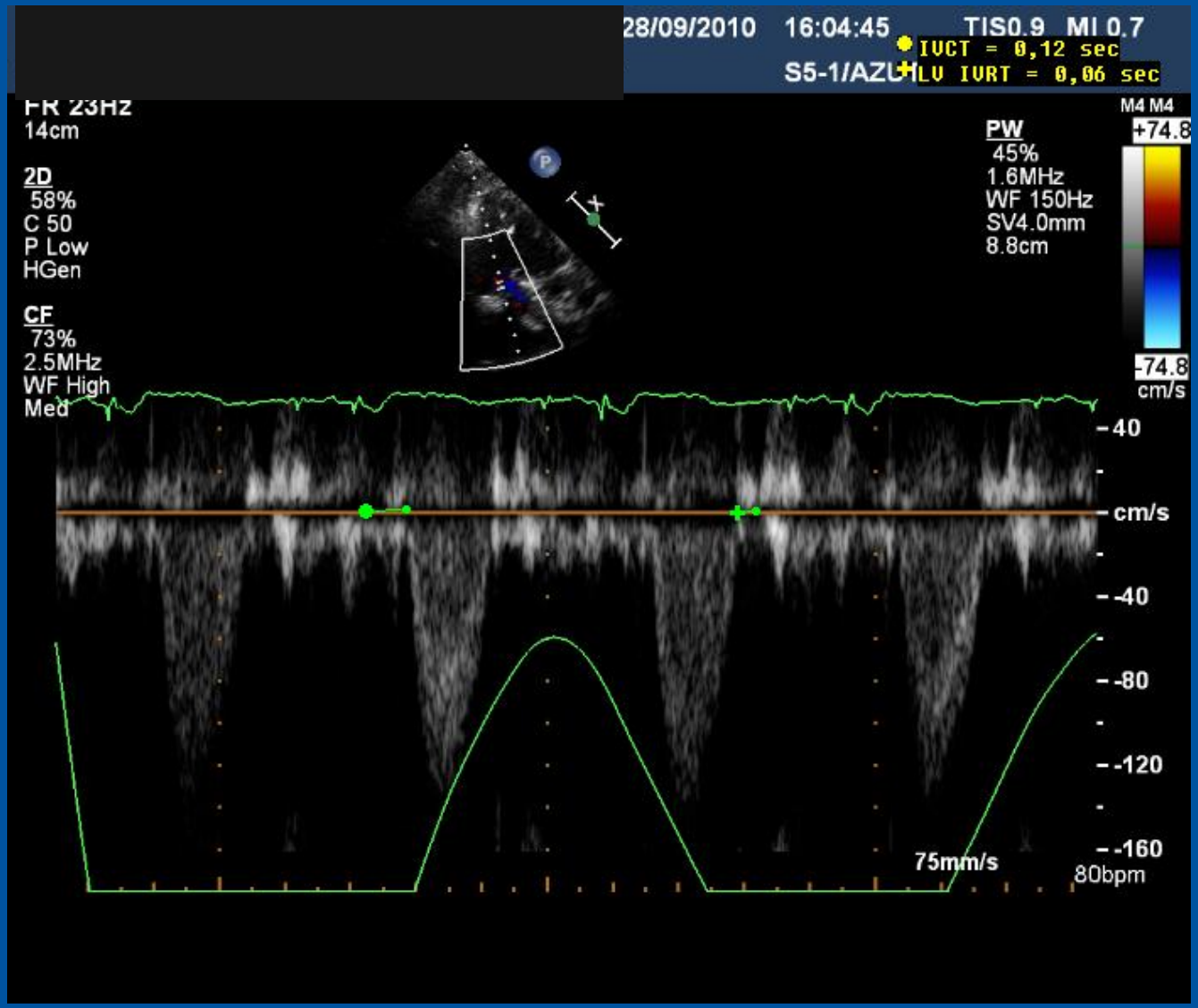

duration systole – very short diastole (3 yr old with HLHS)

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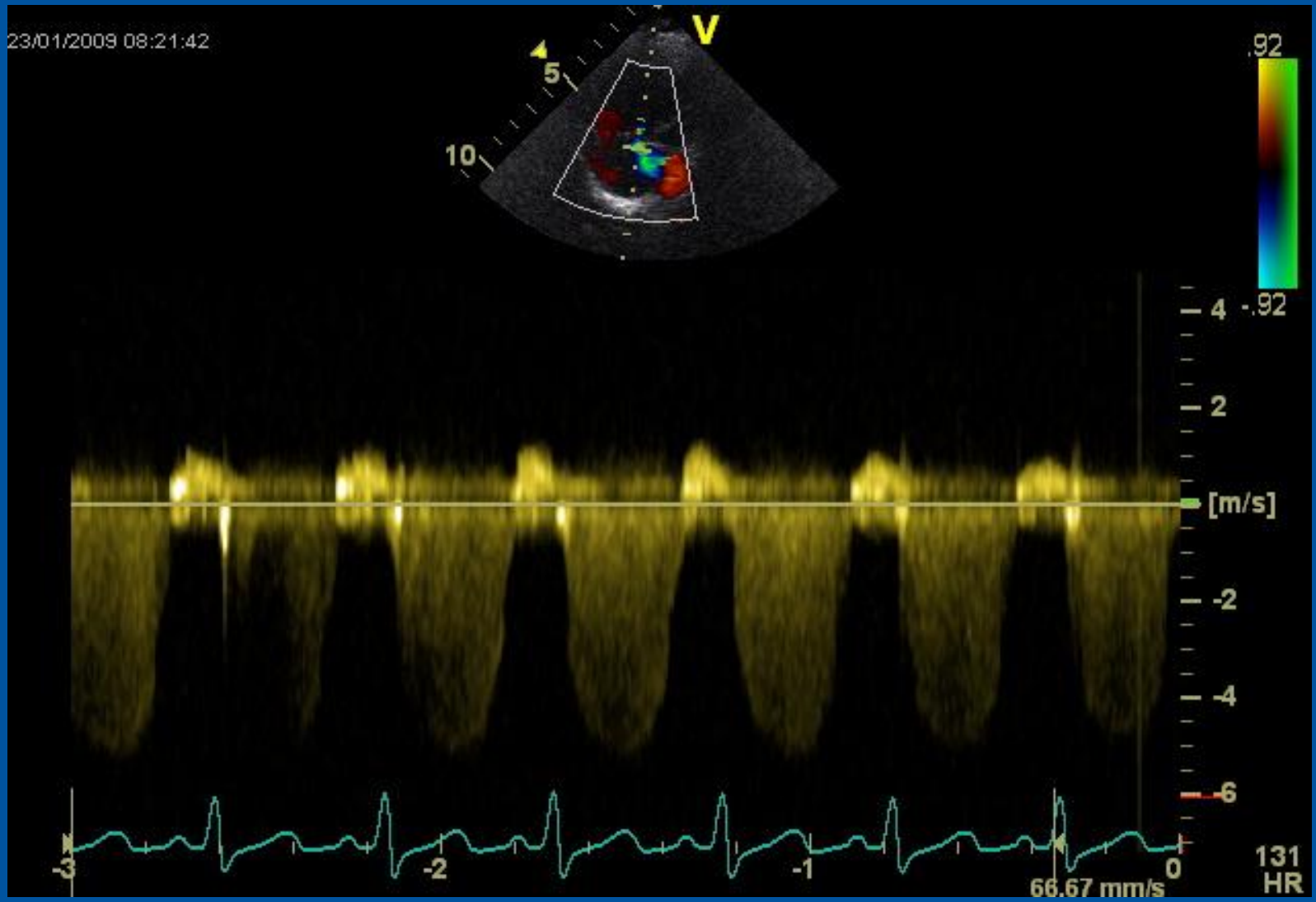

SD ratio in the RV

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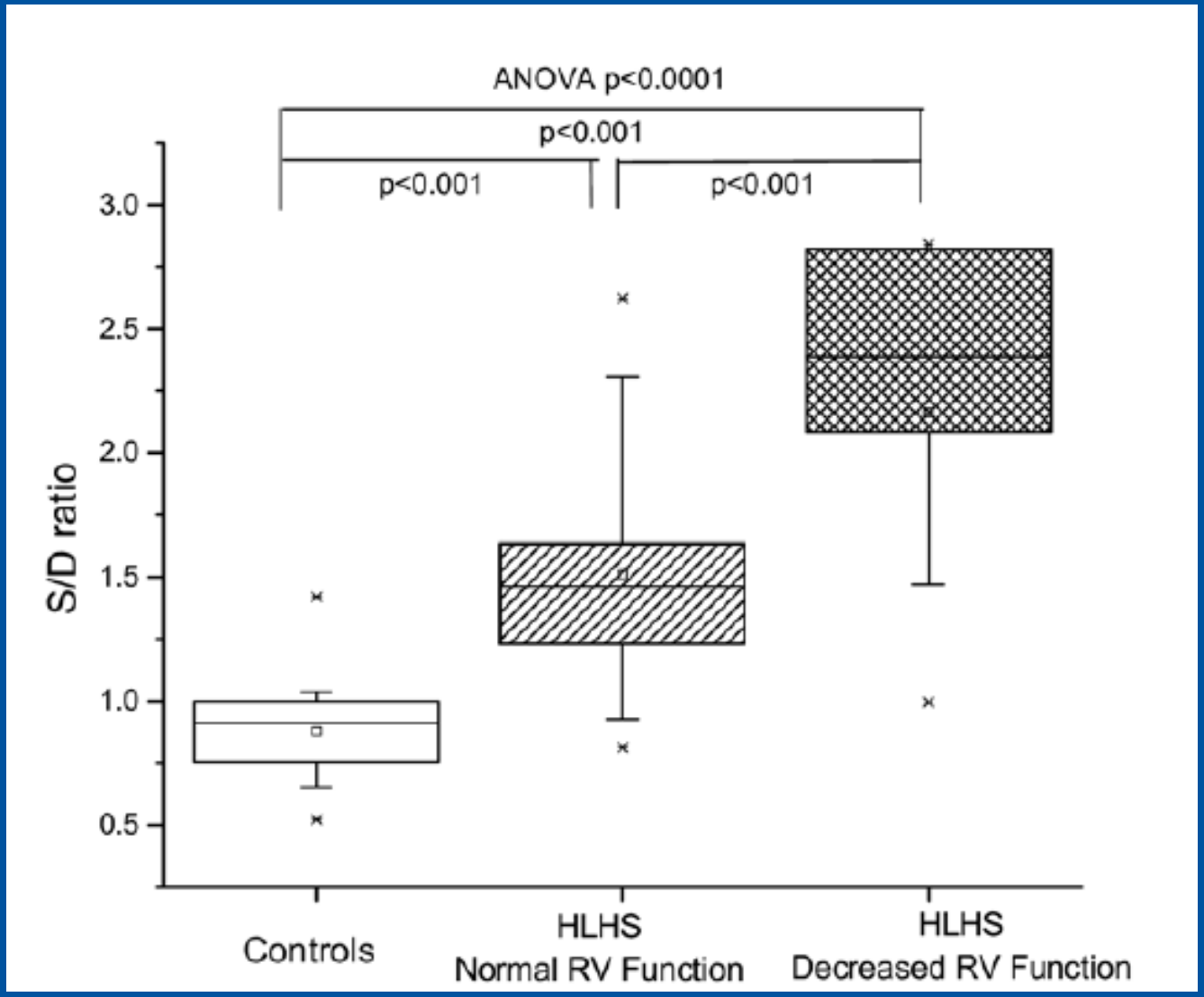

Friedberg, J Am Soc Echocardiogr 2007;20:749

duration systole – short diastole (21 yr old criss-cross, TCPC with PLE)

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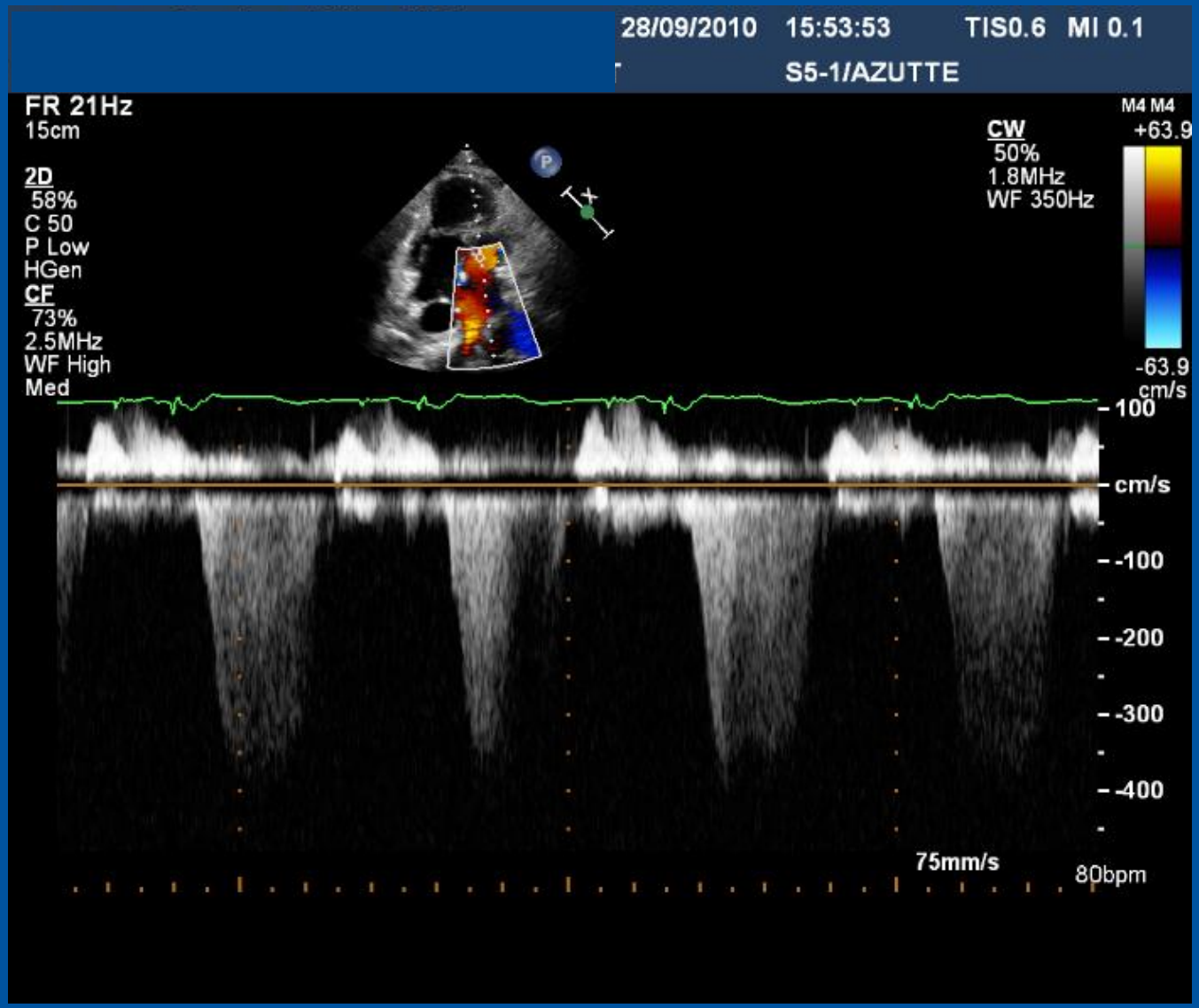

CW dP/dt

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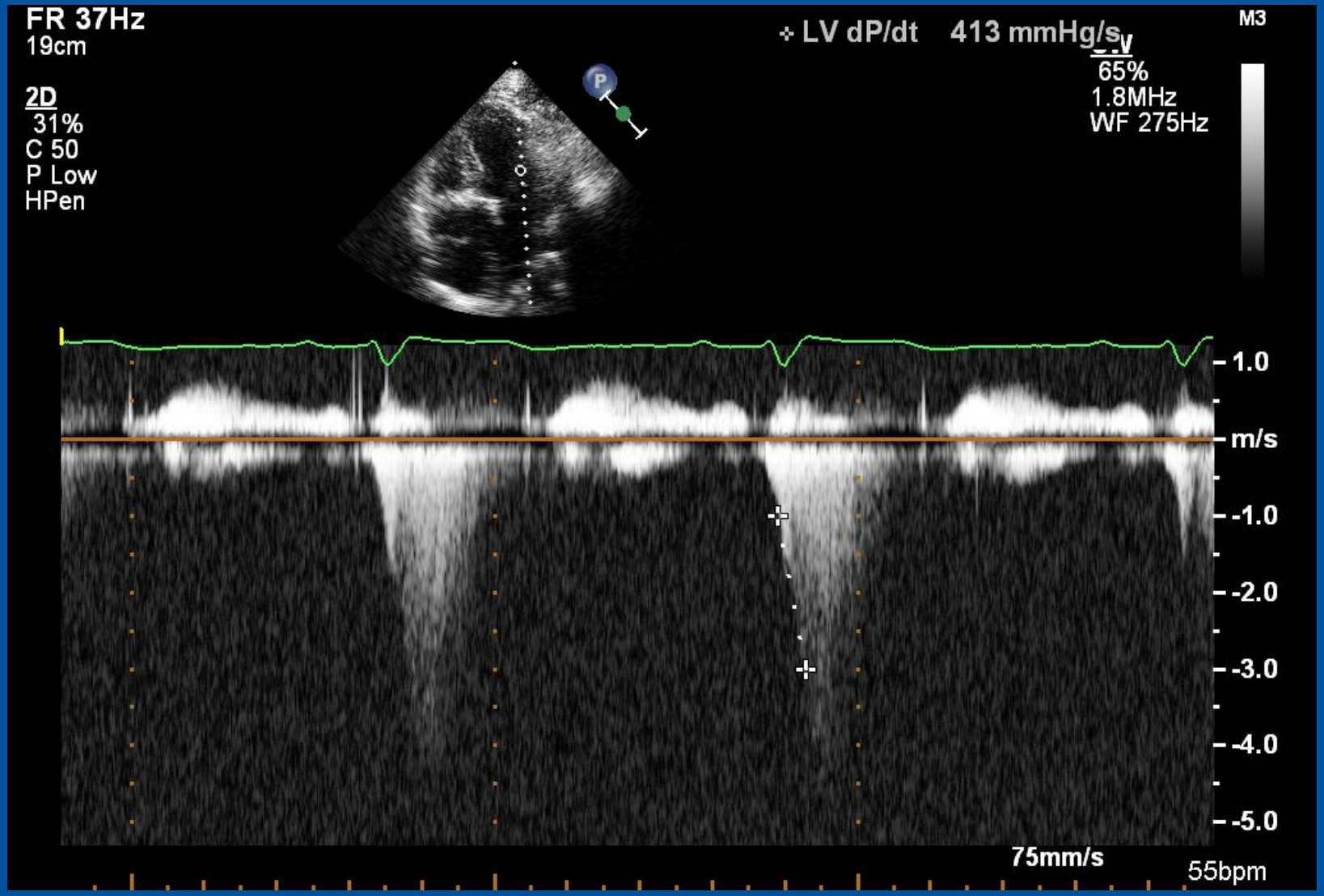

TAPSE

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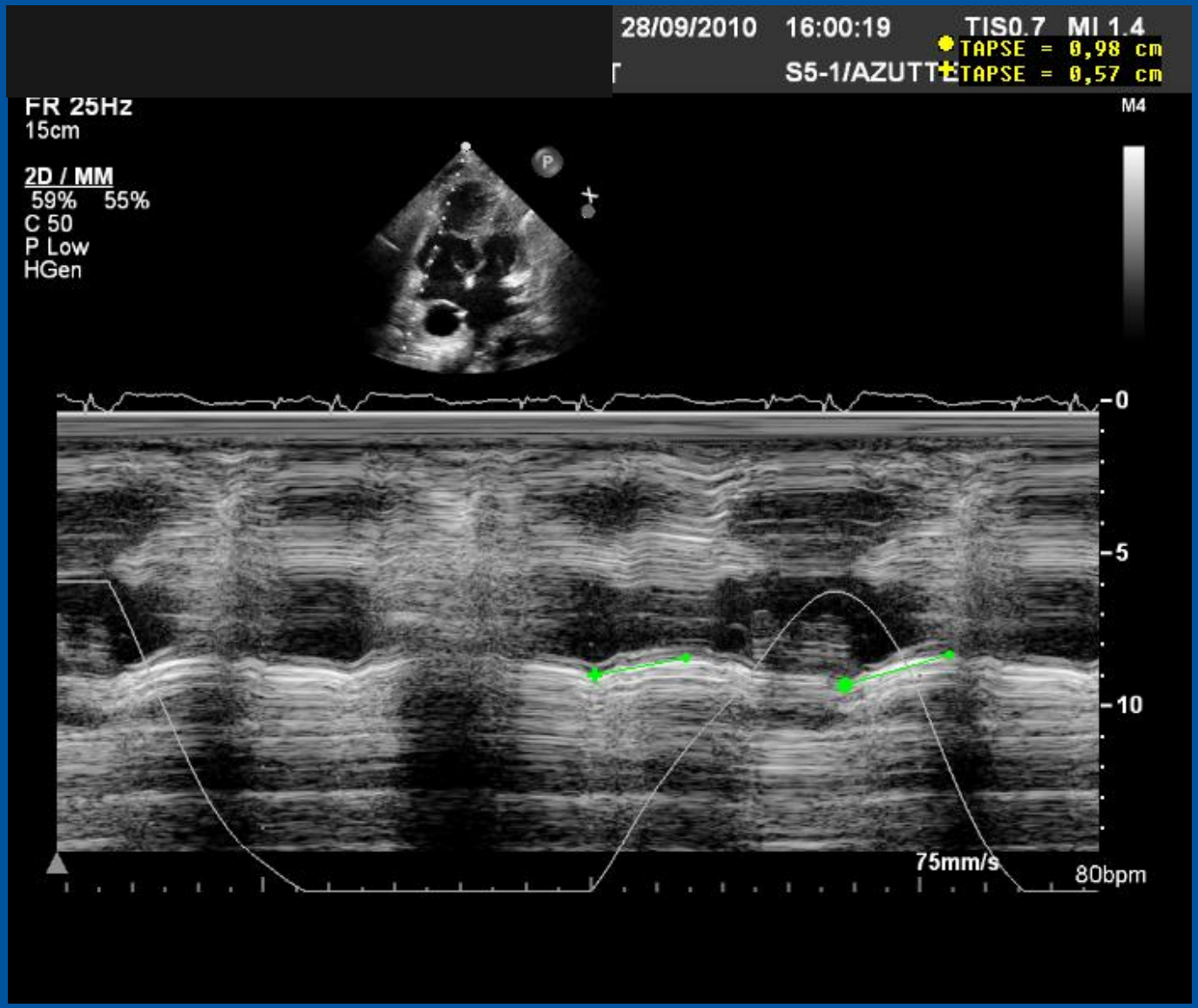

PV flow pattern: very low S in absence MR

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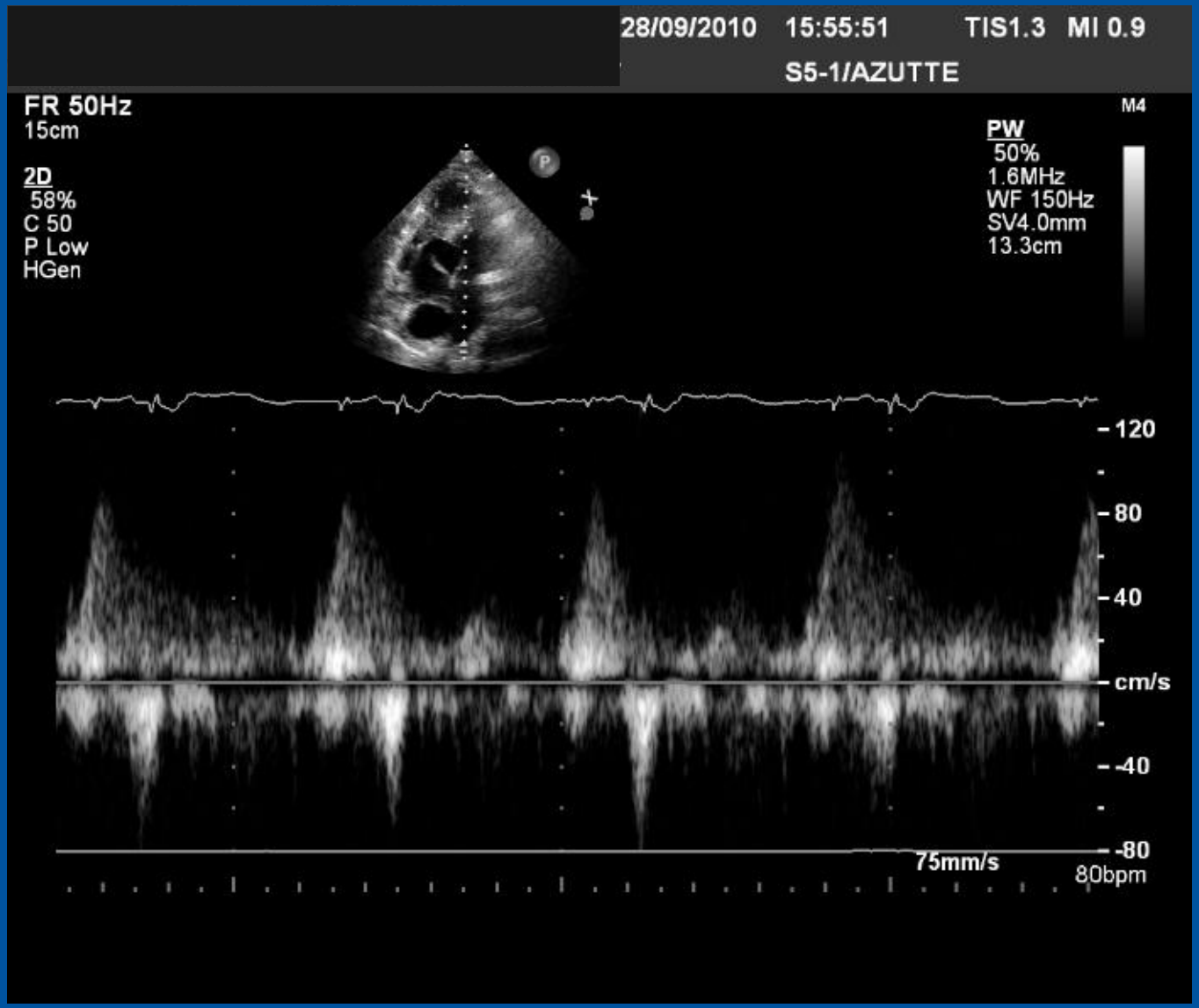

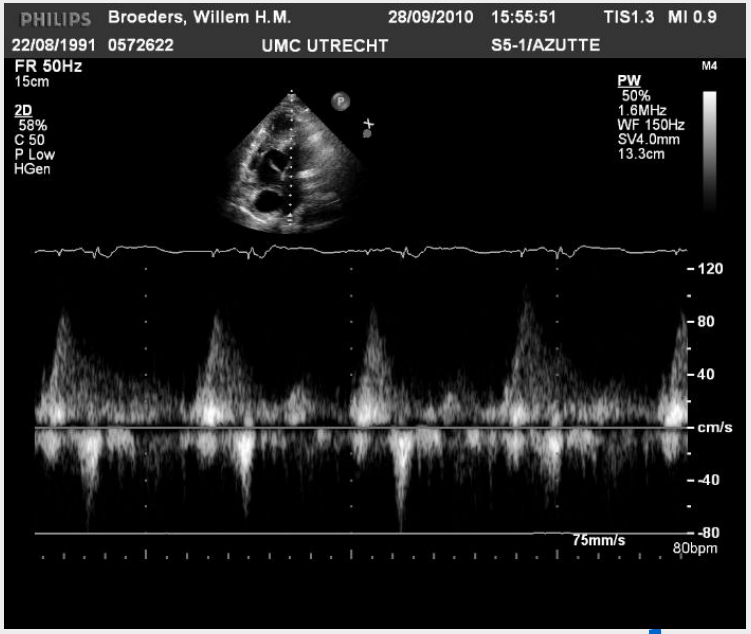

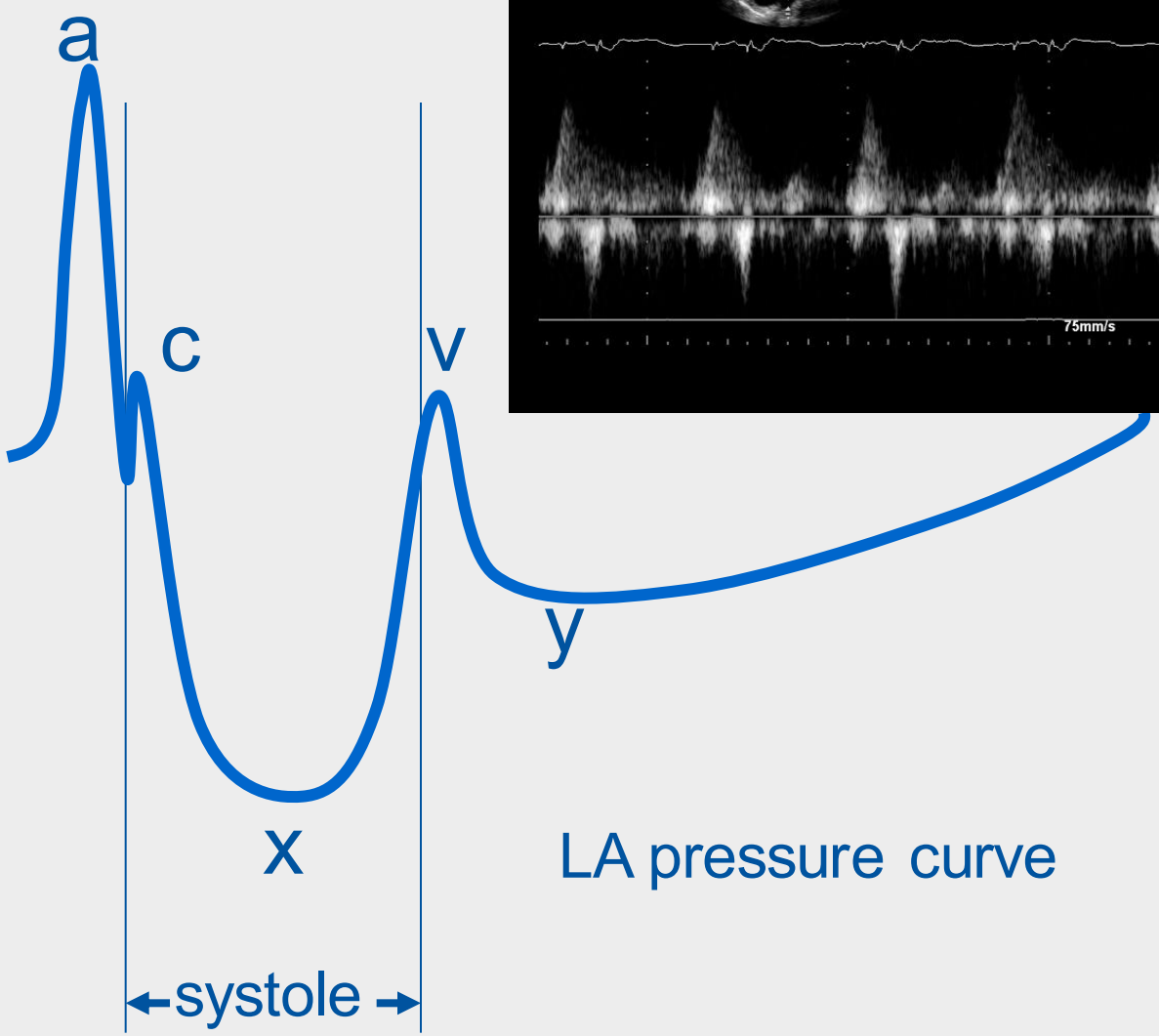

LA pressure curve

Echo & Doppler measurements

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- Absolute values difficult to interpret
- Change over time/ in different conditions may guide us:
  - *in clinical decision making*
  - *When to send a patient to the cathlab*

Echo & Doppler measurements

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- Absolute values difficult to interpret
- Change over time/ in different conditions may guide us:
  - *in clinical decision making*
  - *When to send a patient to the cathlab*
- The essence of the Fontan circulation: V = P/R we cannot measure this with echo

Visualisation of Fontan anatomy

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Visualisation of Fontan anatomy

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- Segmental approach
  - *Hepatic veins*
  - *VCI*
  - *Tunnel*
    - Intracardiac
    - Extracardiac
  - *RA*
  - *Connection with PA*
  - *VCS*
  - *Connection with PA*
  - *Pulmonary artery branches*
  - *Pulmonary veins*

Visualisation of Fontan anatomy & function

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- Segmental approach
  - *Hepatic veins*
  - *VCI*
  - *Tunnel*
    - Intracardiac
    - Extracardiac
  - *RA*
  - *Connection with PA*
  - *VCS*
  - *Connection with PA*
  - *Pulmonary artery branches*
  - *Pulmonary veins*

- Color Doppler patterns
- Pulsed Doppler flow patterns
- Shunts/leaks in the tunnel
- thrombi

VCI & hepatic veins

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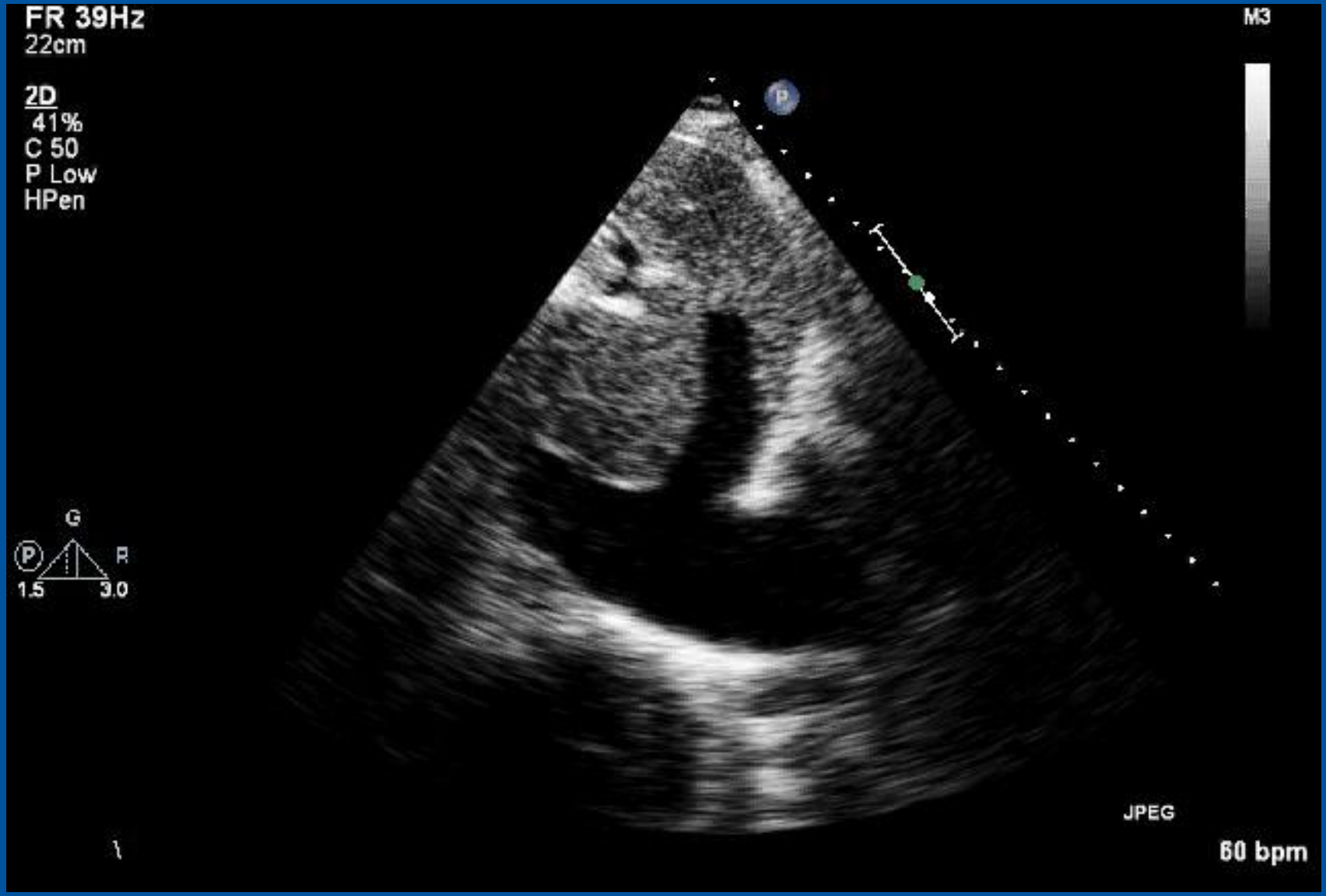

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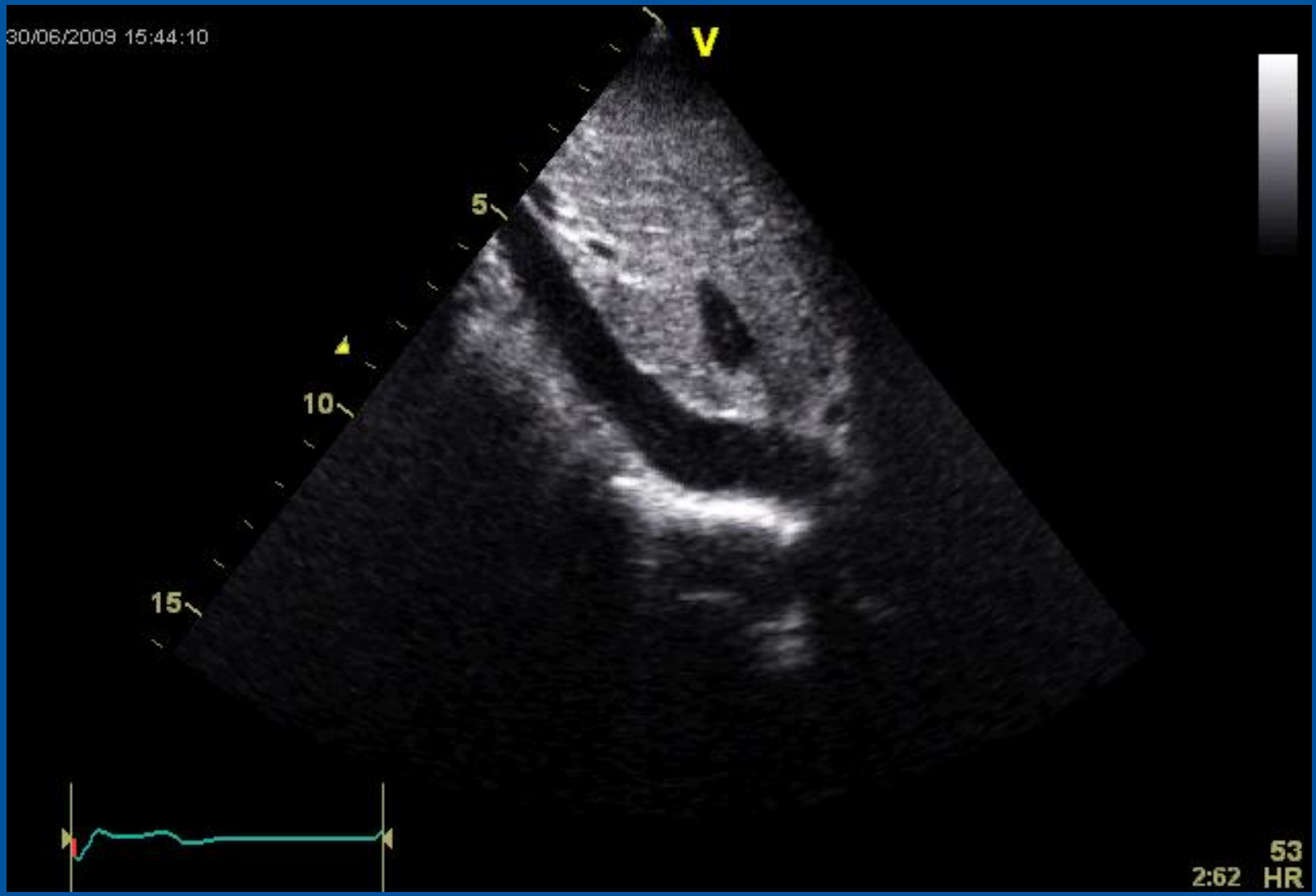

VCI pulsed Doppler:
Scale! Low velocity filter! Respiration!

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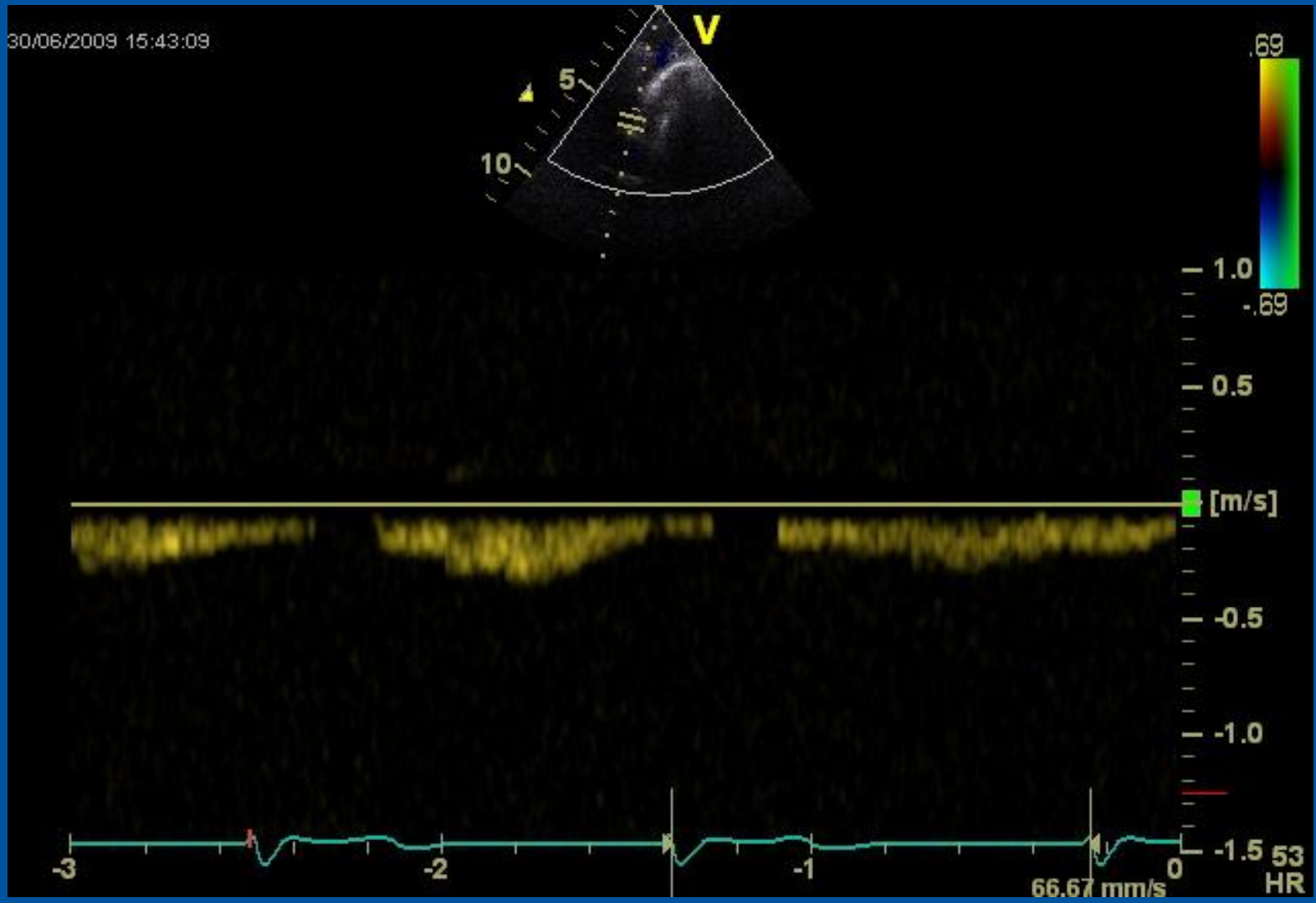

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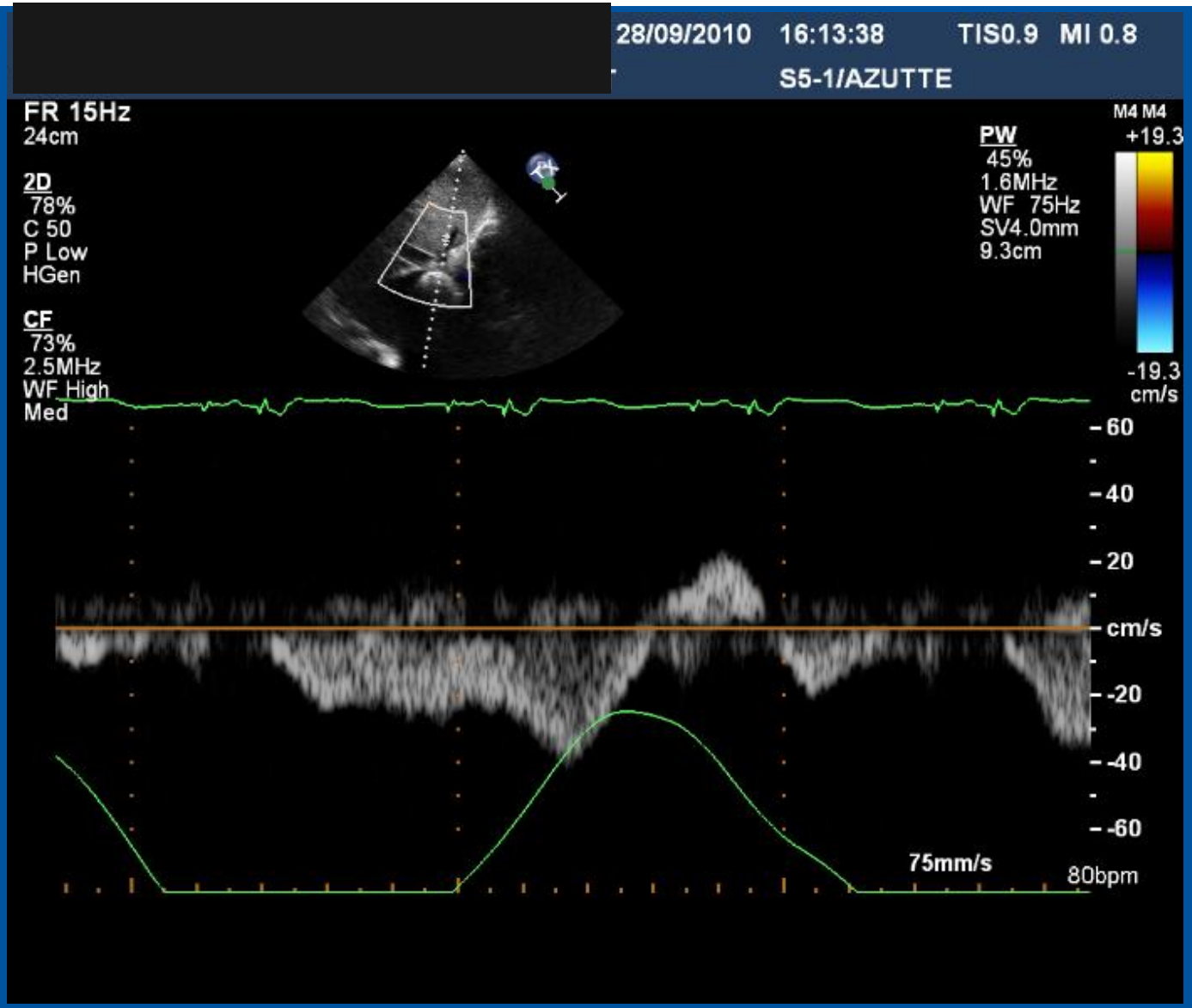

TCPC – driving force = inspiration
no influence of cardiac cycle

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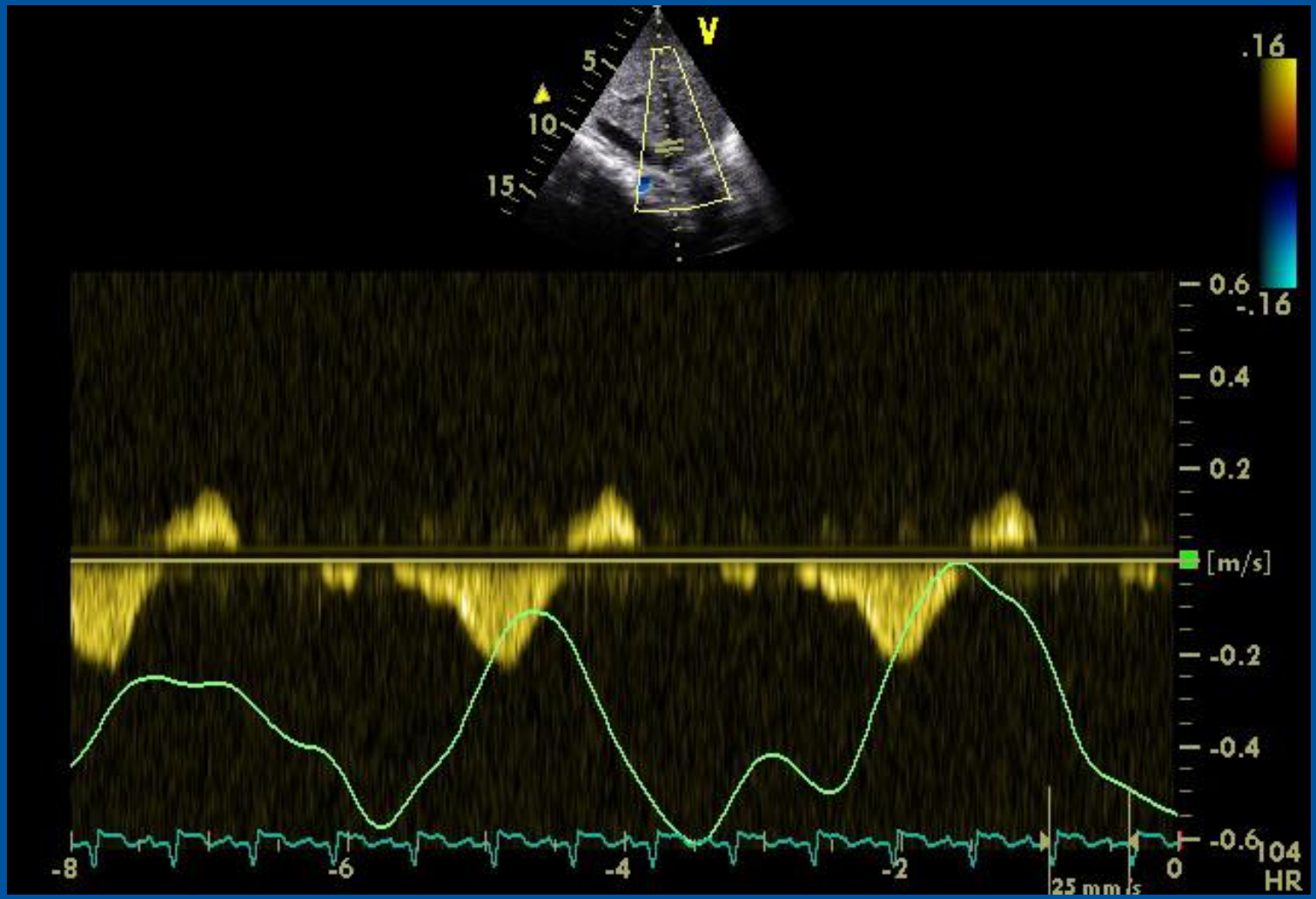

Connection VCI – extracardiac lateral tunnel

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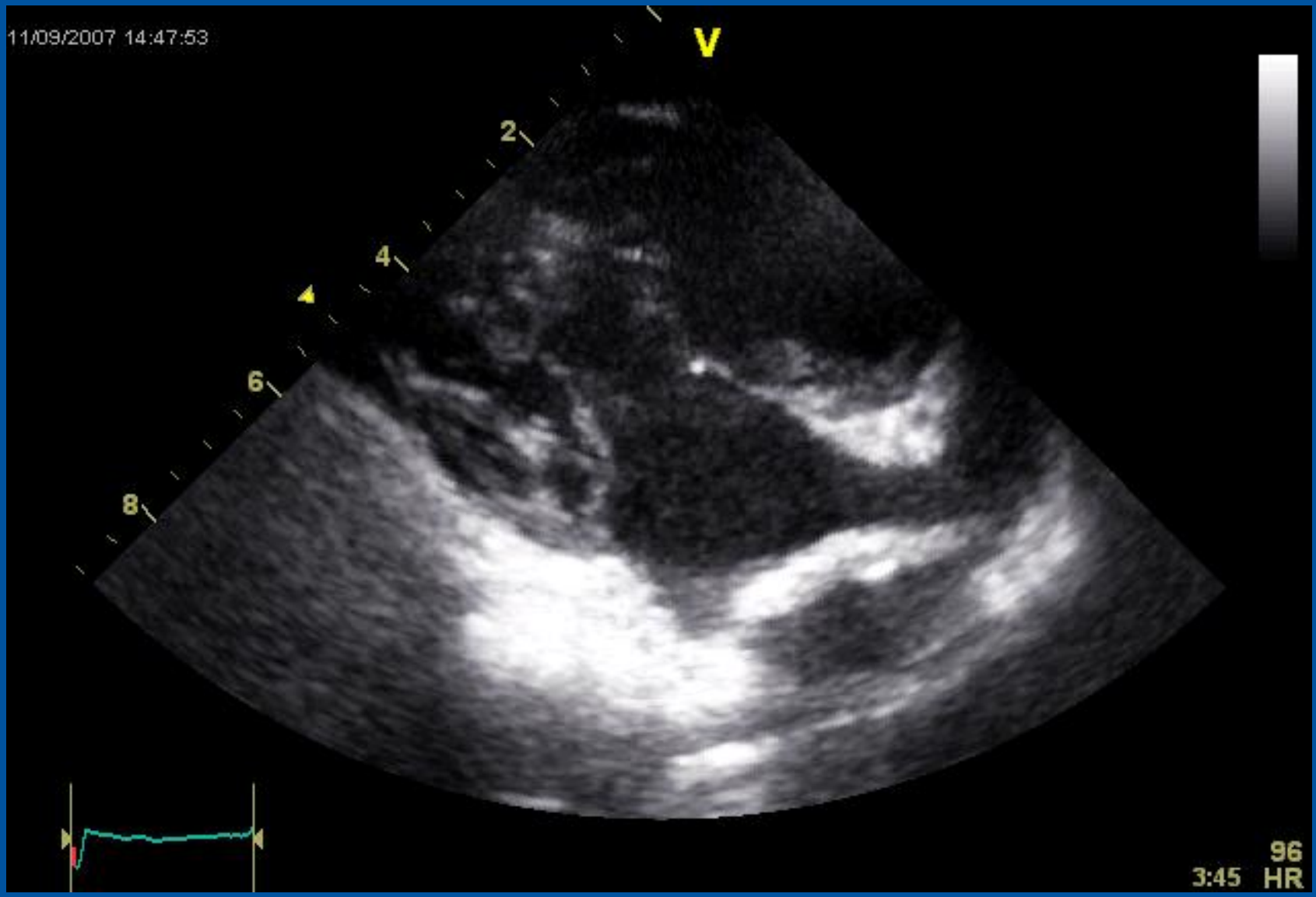

Lateral, extracardiac tunnel look for thrombi

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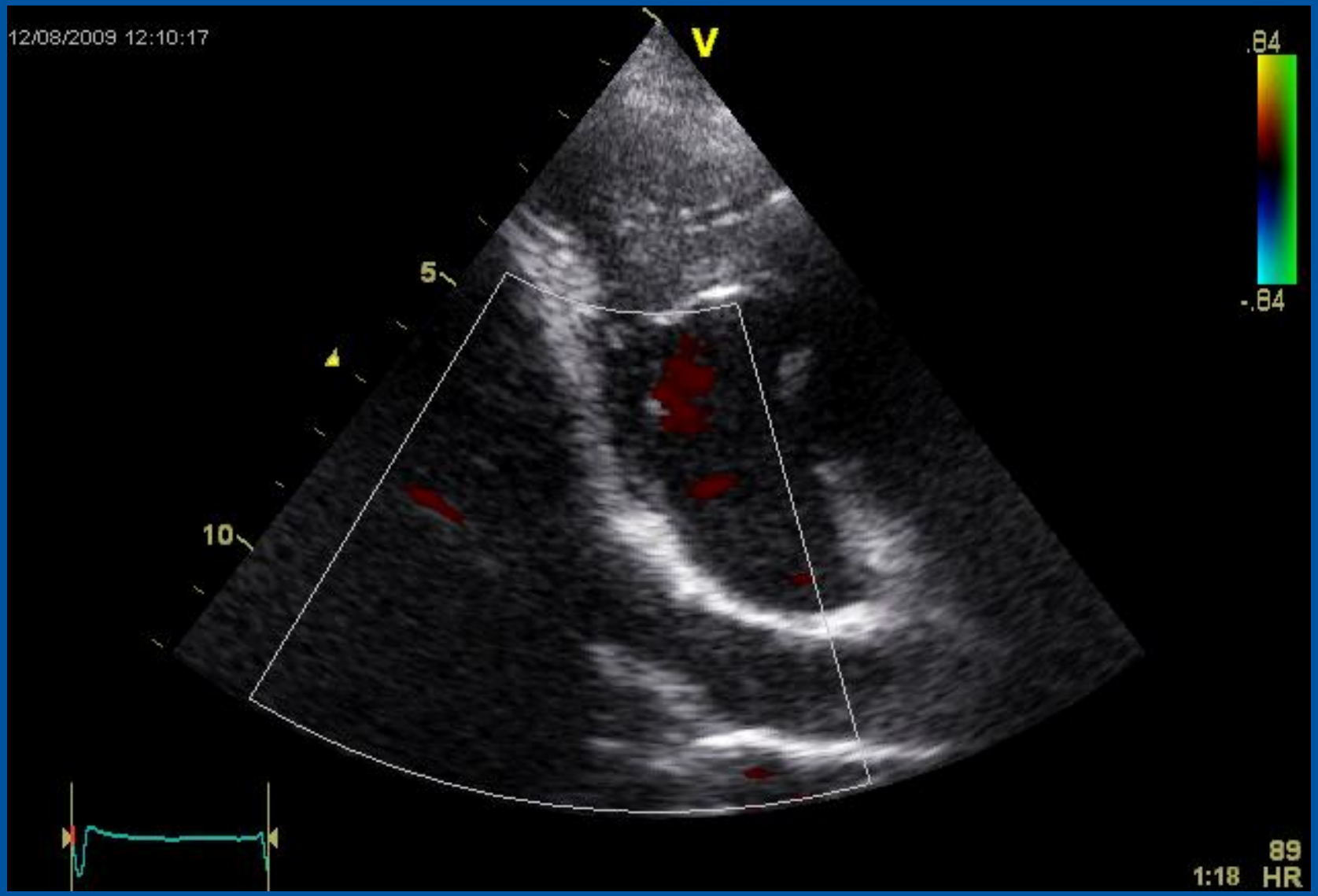

Apical 4 Ch view intracardiac tunnel

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Intra-atrial tunnel

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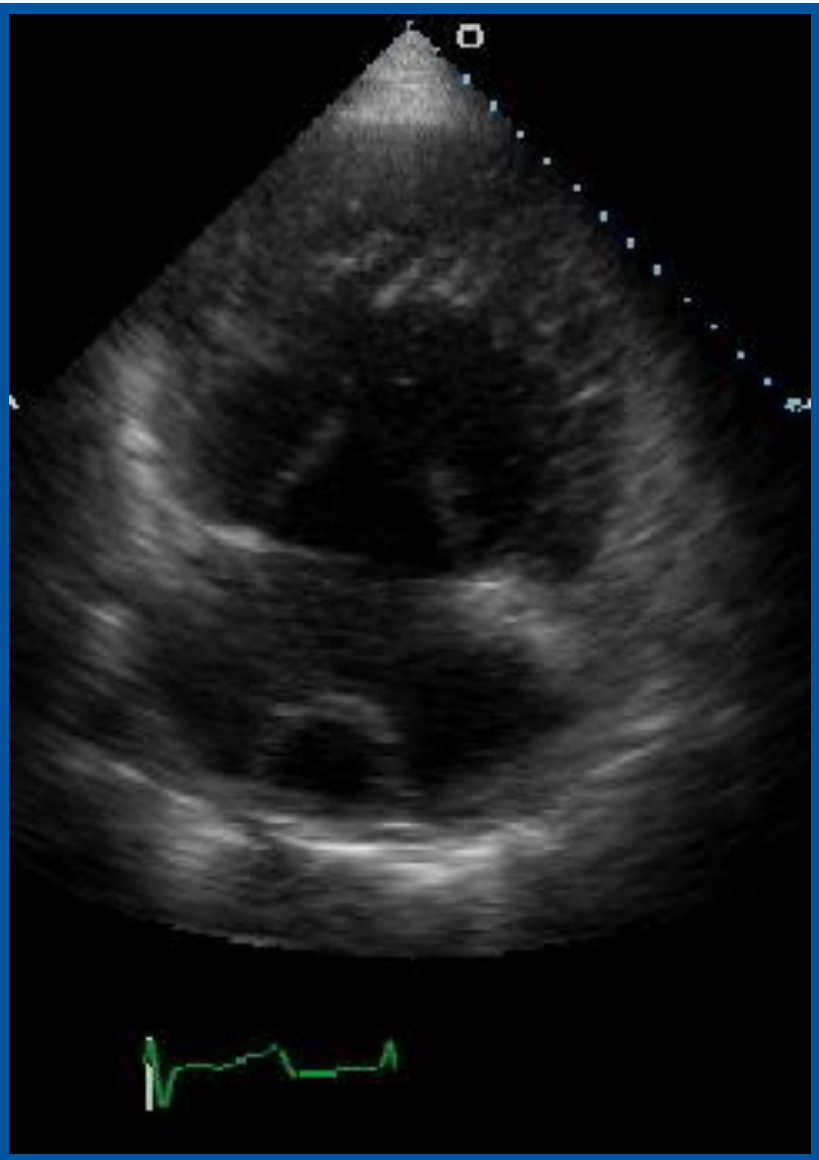

- Intra-atrial tunnel
- initially a small diameter
- No PV obstruction

- Apical scan; upside down

Intra-atrial tunnel; adult age

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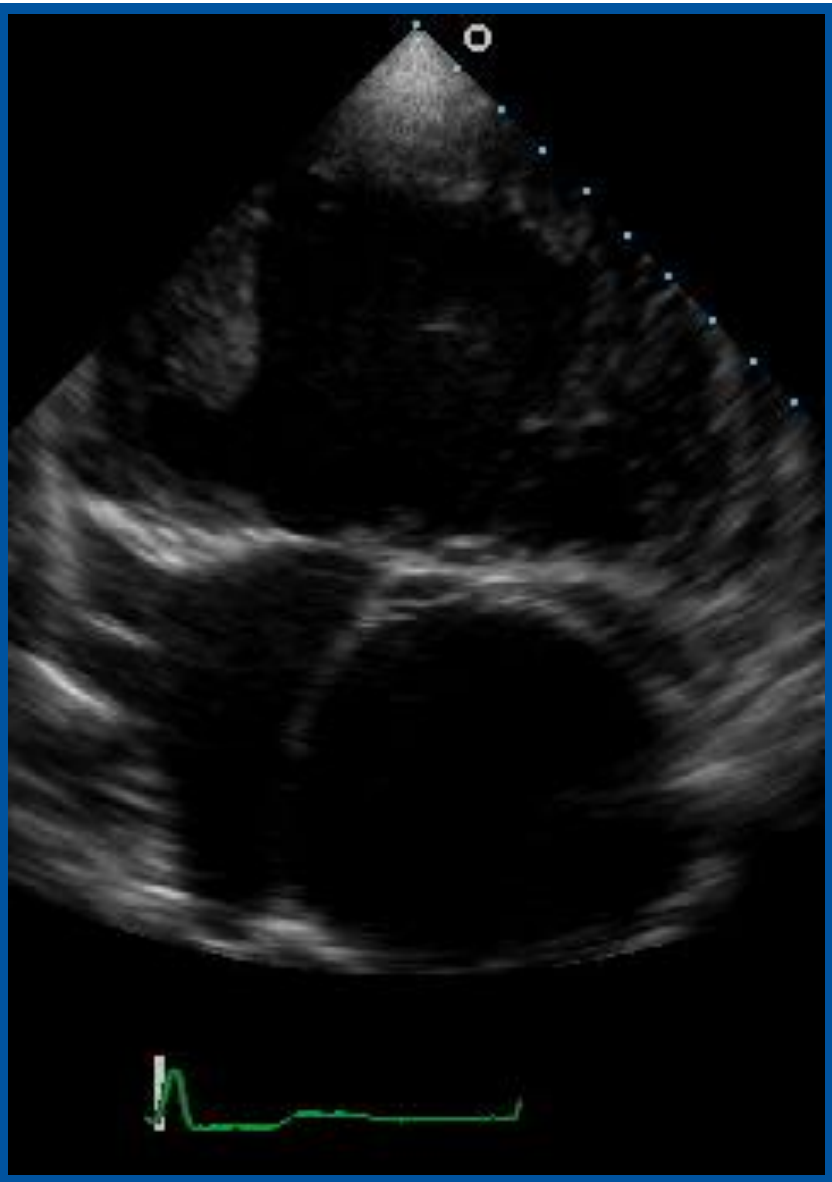

- can dilate in time,
- become very wide
- can give PV obstruction
- Looks like RA – PA
- (different flow patterns)

VCI Doppler in RA – PA conduit effect of atrial contraction

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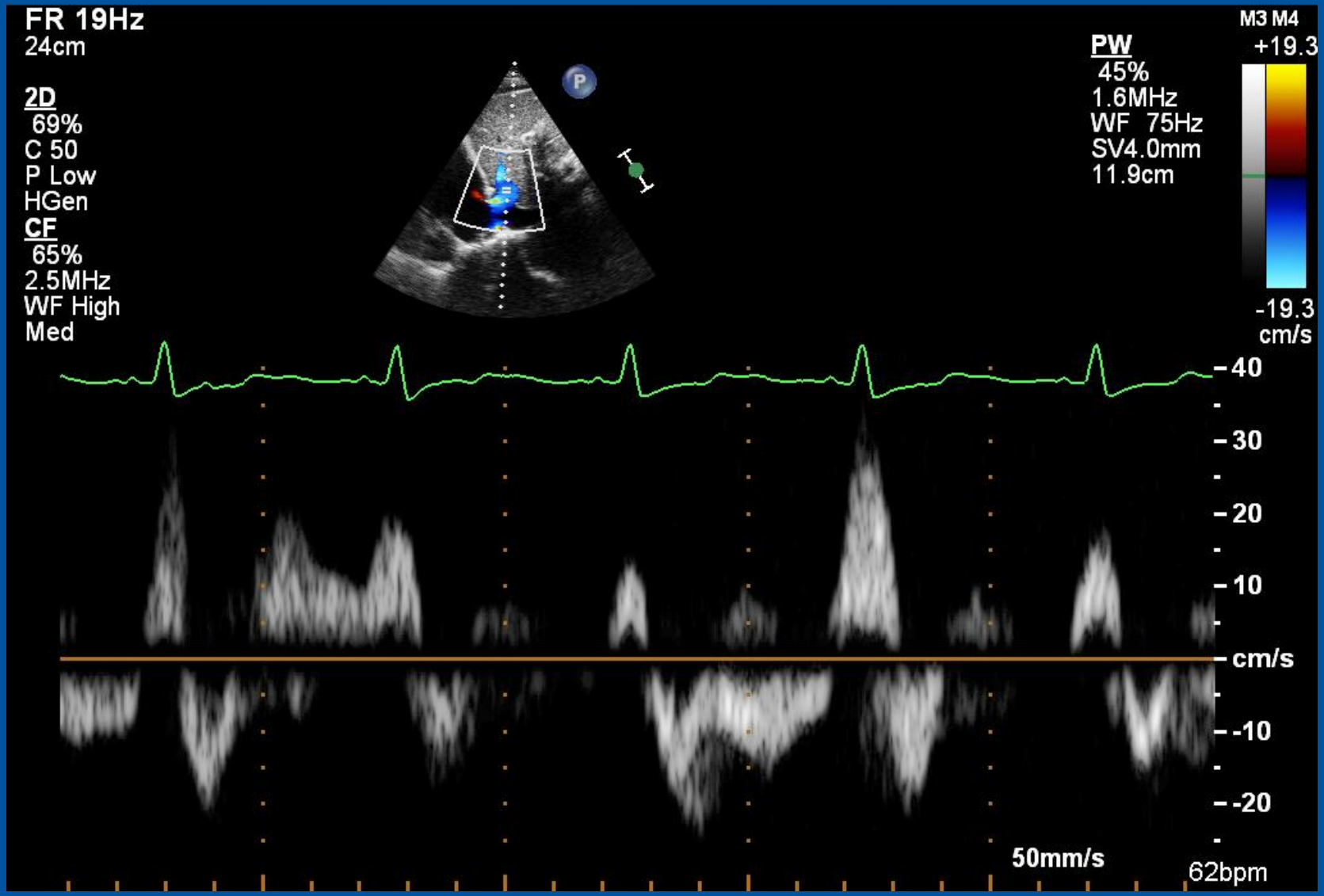

Tricuspid atresia; RA – AP connectie

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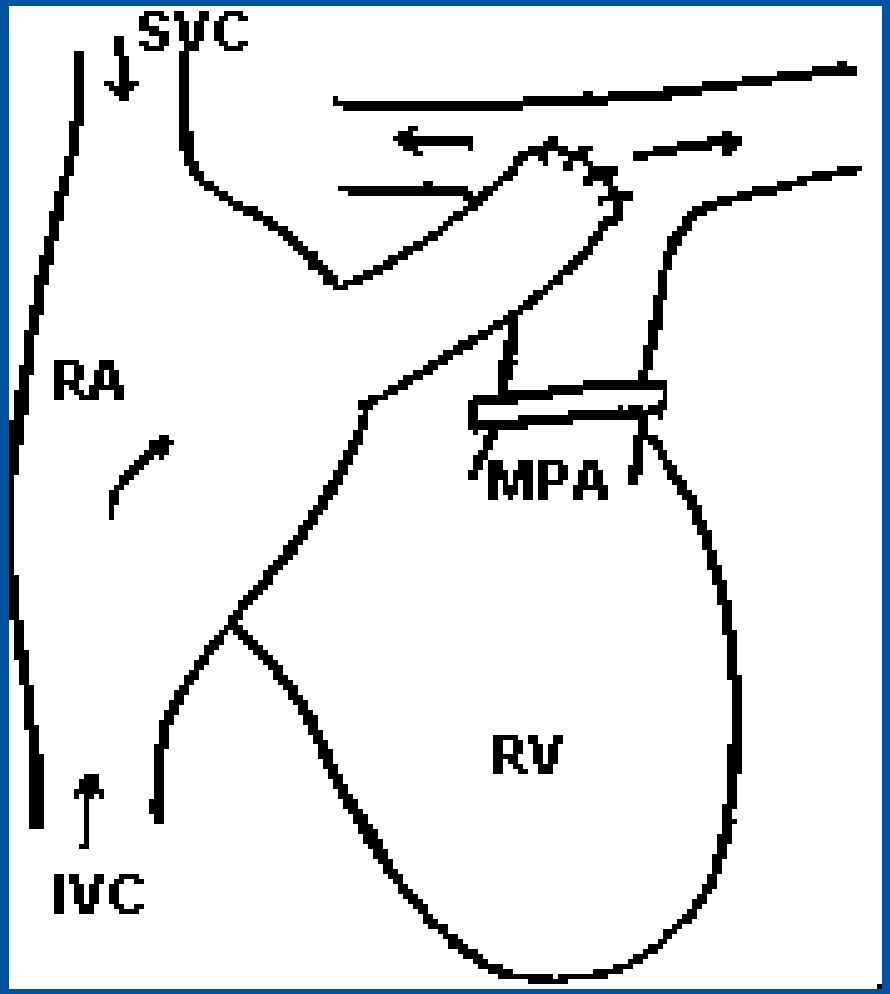

RA –PA conduit

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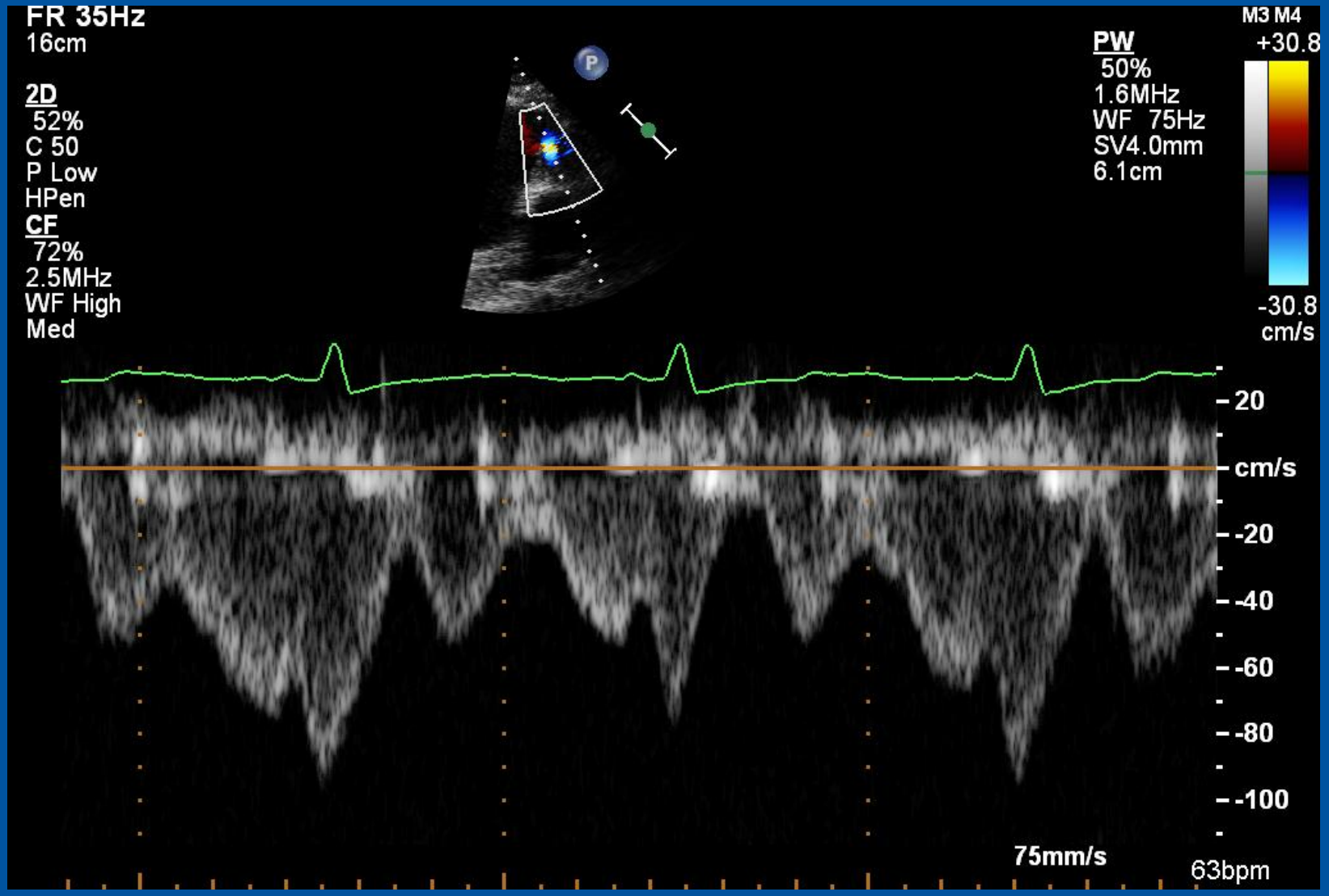

Atrio – ventricular connection

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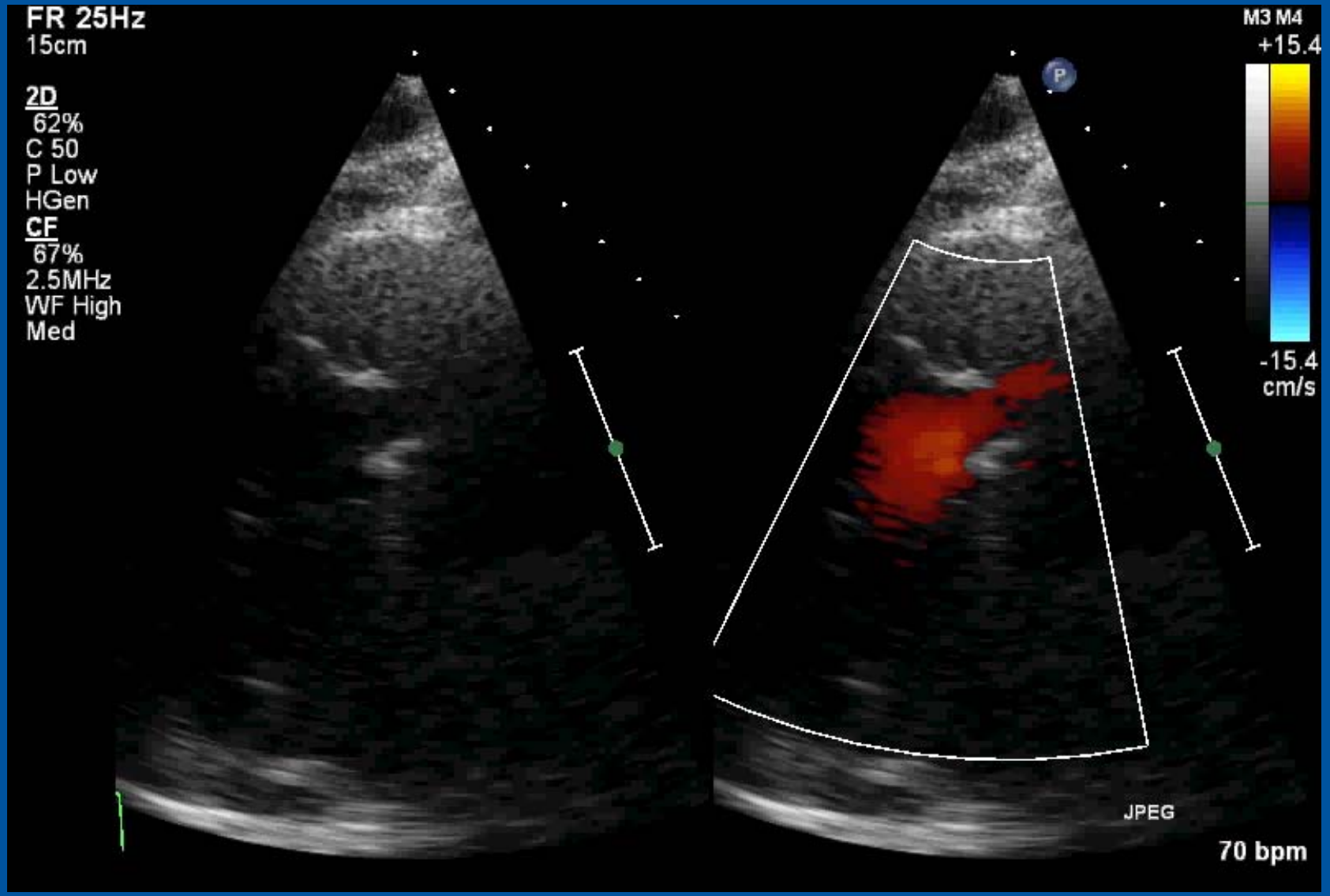

Tricuspid atresia & RA – RV connectie

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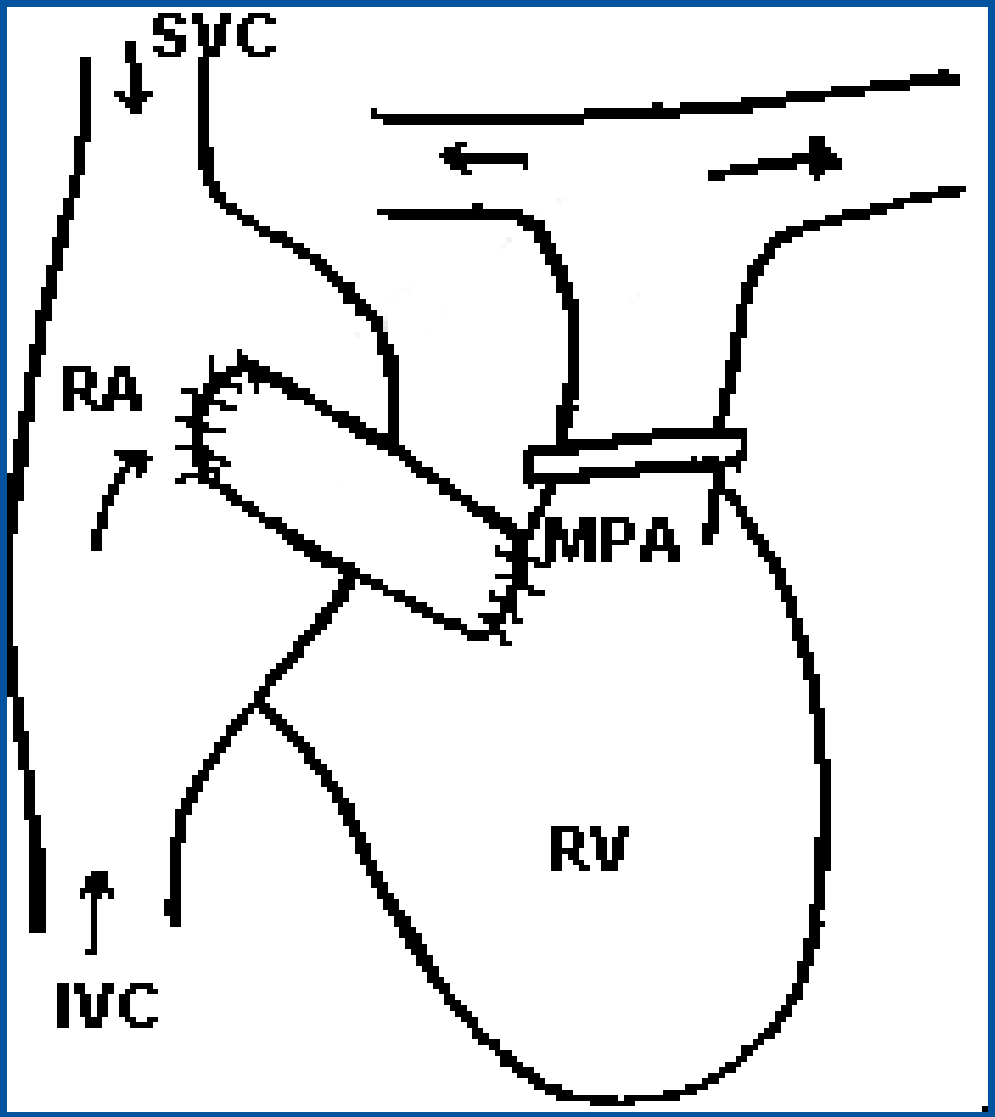

RA – RV conduit with stenosis

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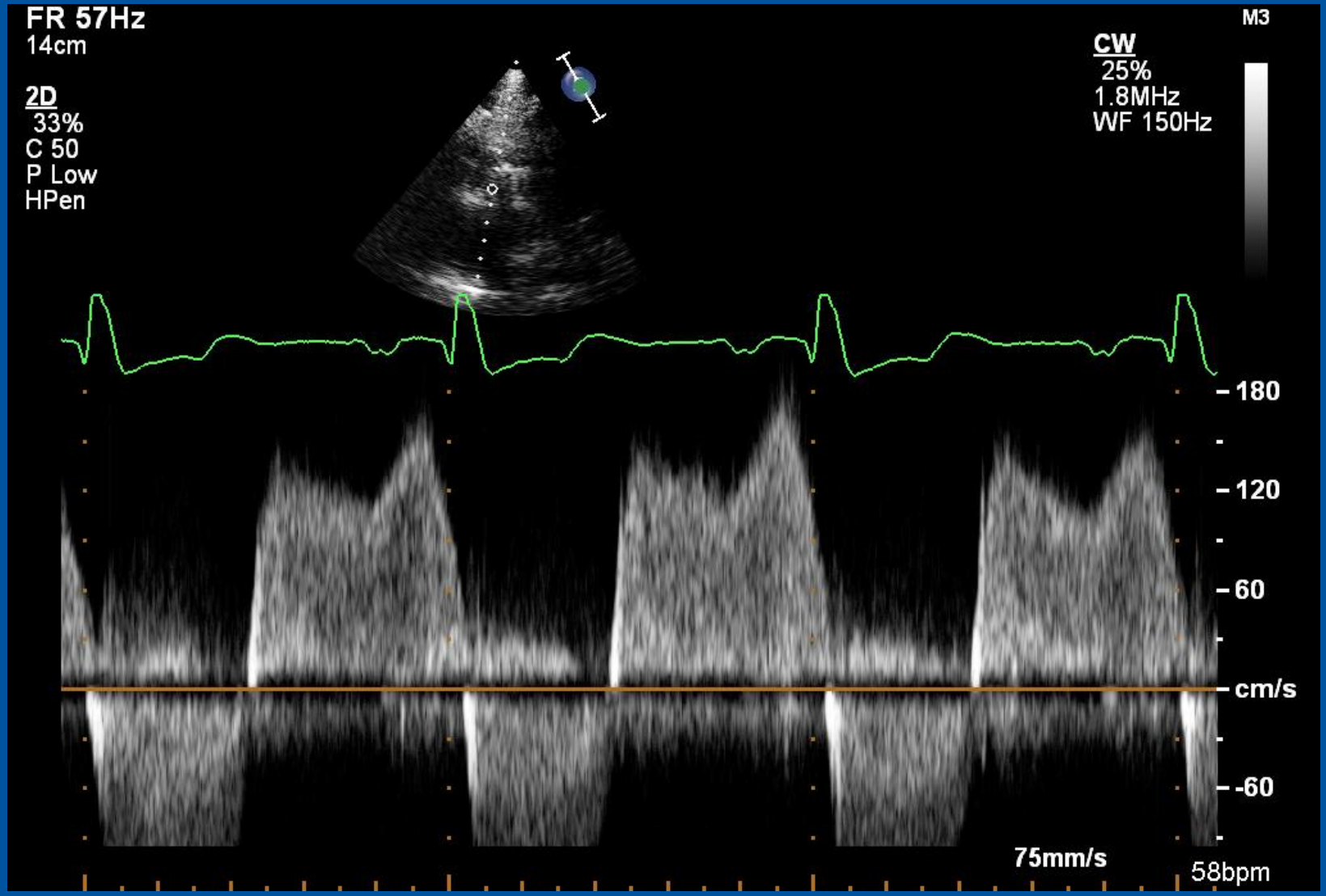

PA flow in RA – RV conduit:

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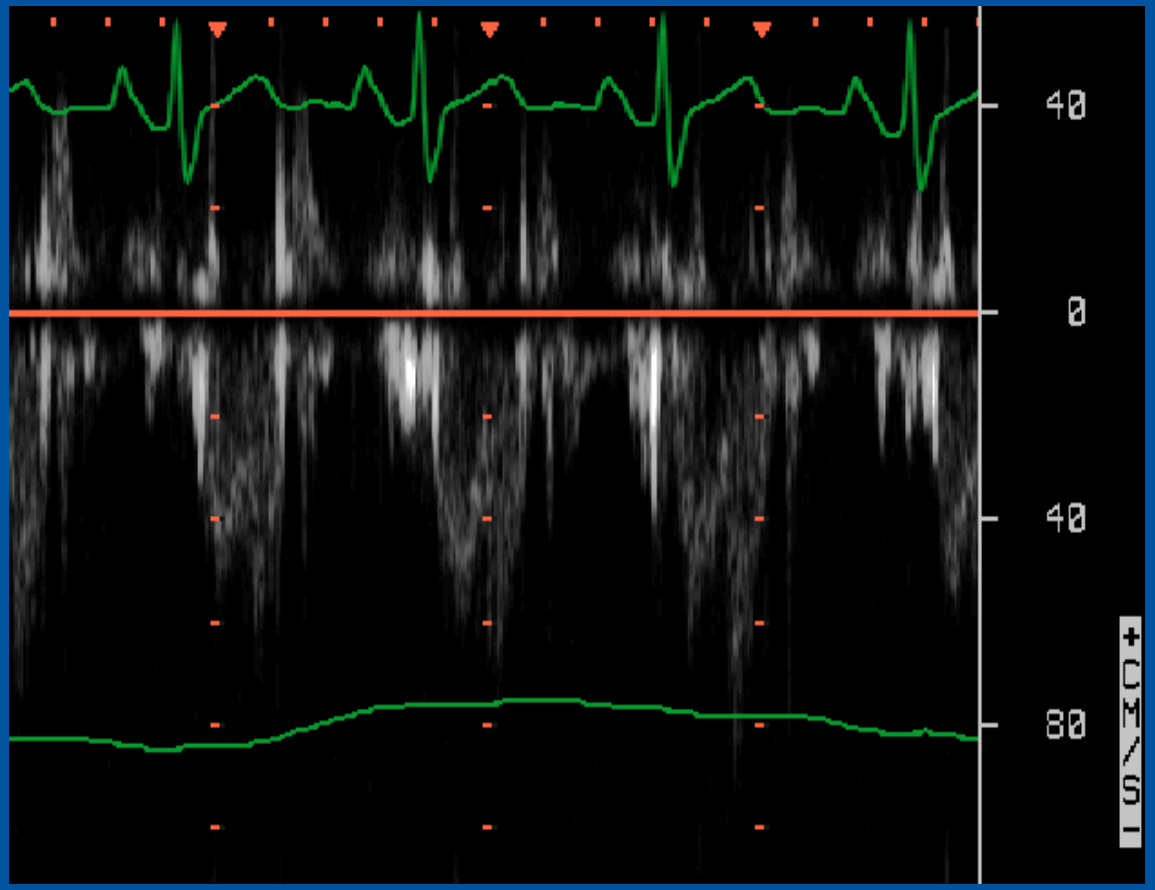

Pulsatile systolic flow in PA!

Doppler patterns & type of Fontan

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- Doppler patterns in different types of connection are different
  - *Doppler patterns in different patients with the same type of connections are different*
- registration of ECG – p-waves – and respiration curve
- Off-line studying on tracings:
  - *What explains flow patterns?*
  - *What explains phasic changes?*

intracardiac tunnel VCI - PA

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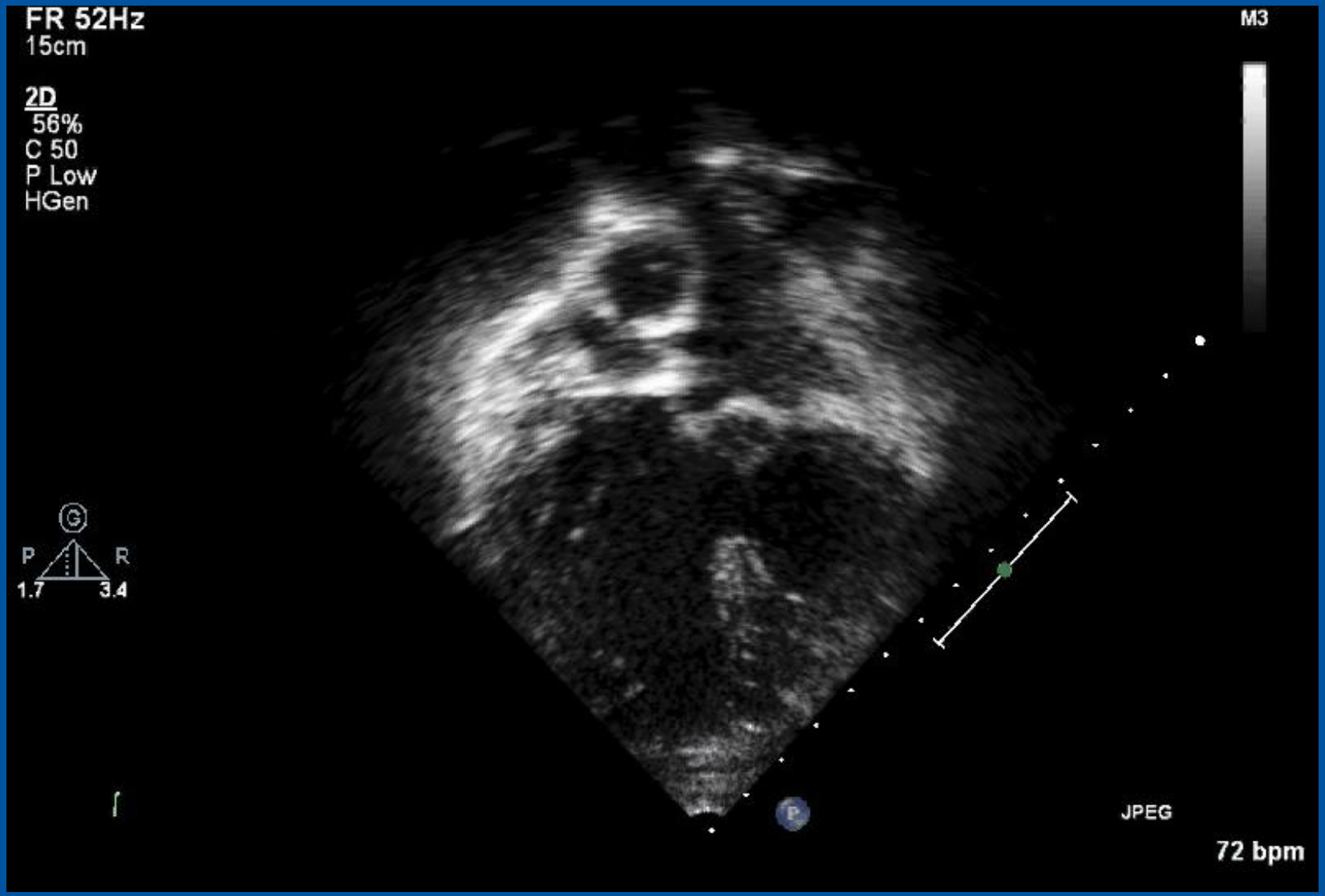

Look for shunts/abnormal flows

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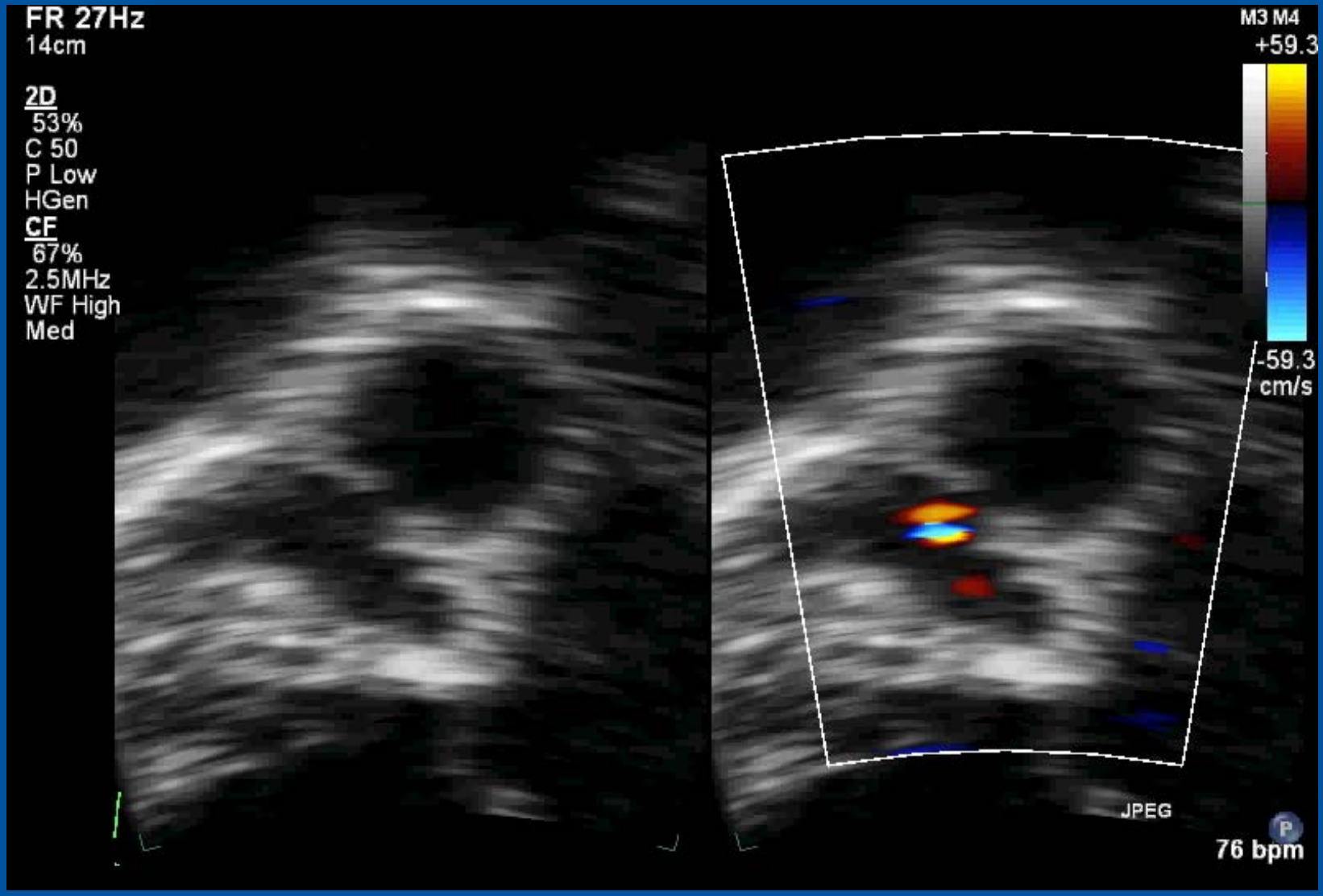

Baffle leakage with R – L shunt

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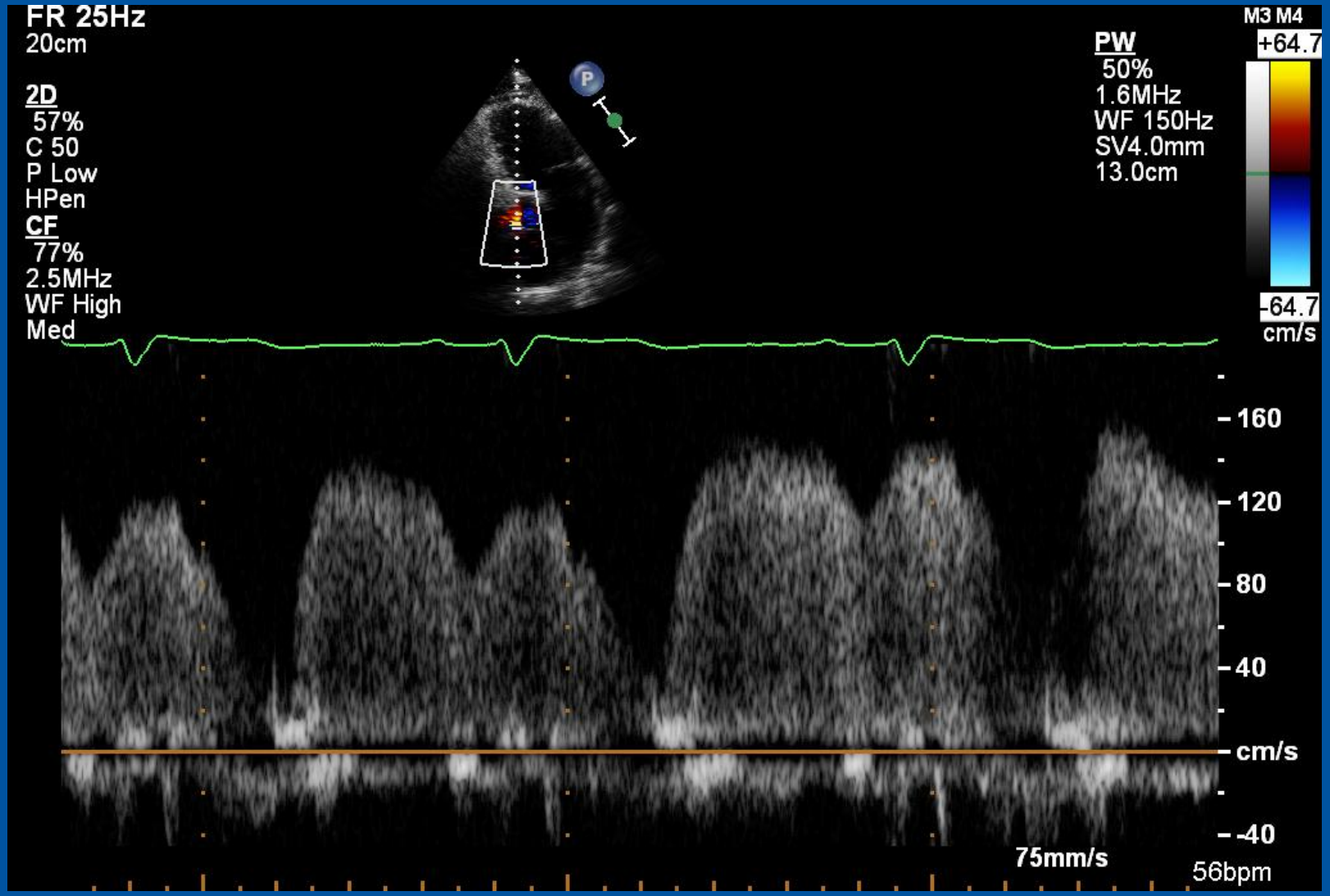

Baffle leakage with R – L shunt

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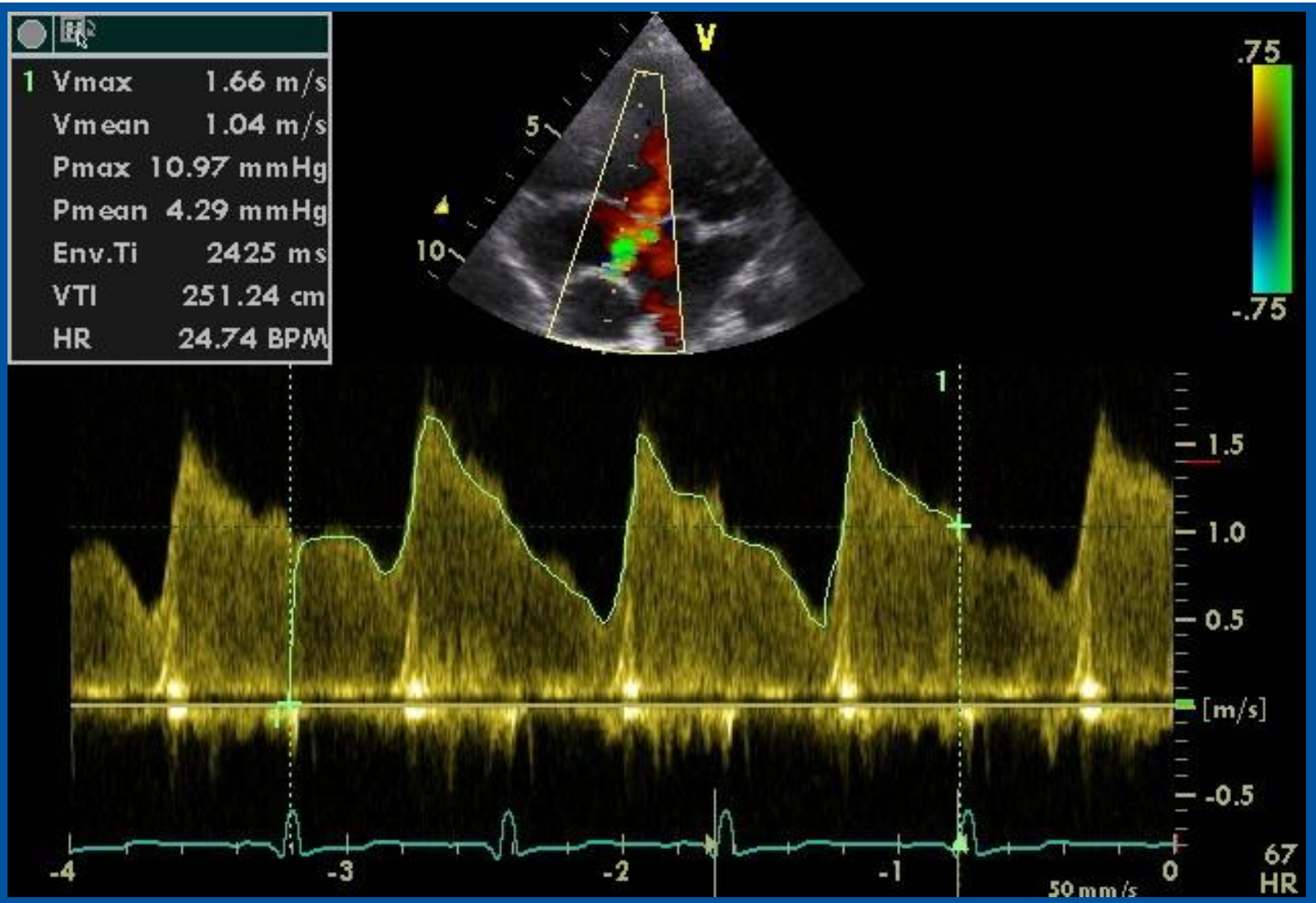

Courtesy Jan Marek

What to do with a baffle leak?

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- Beyond the scope of this talk

Bidirectional Glenn anastomosis

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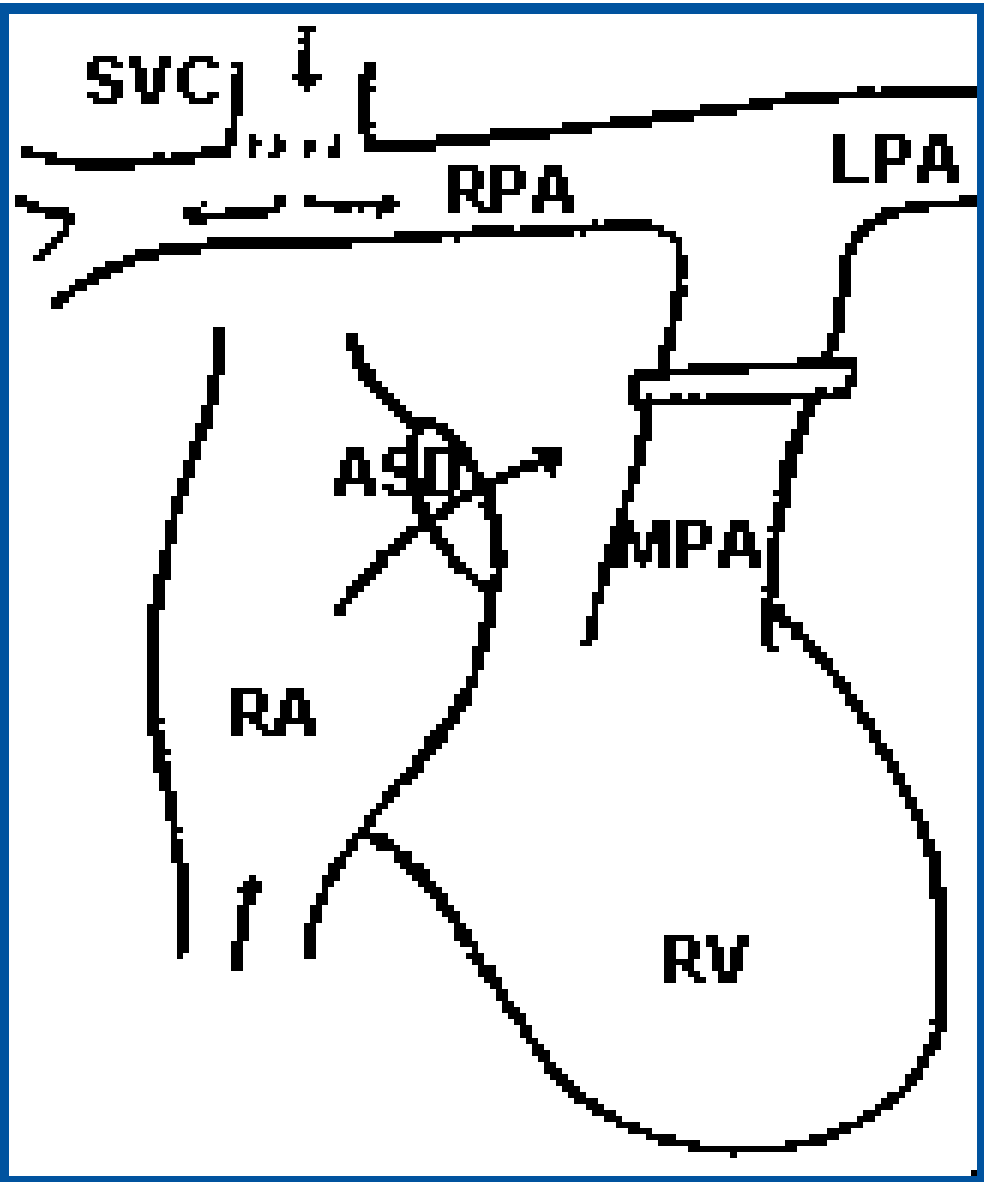

VCS – Glenn from R supraclavicular

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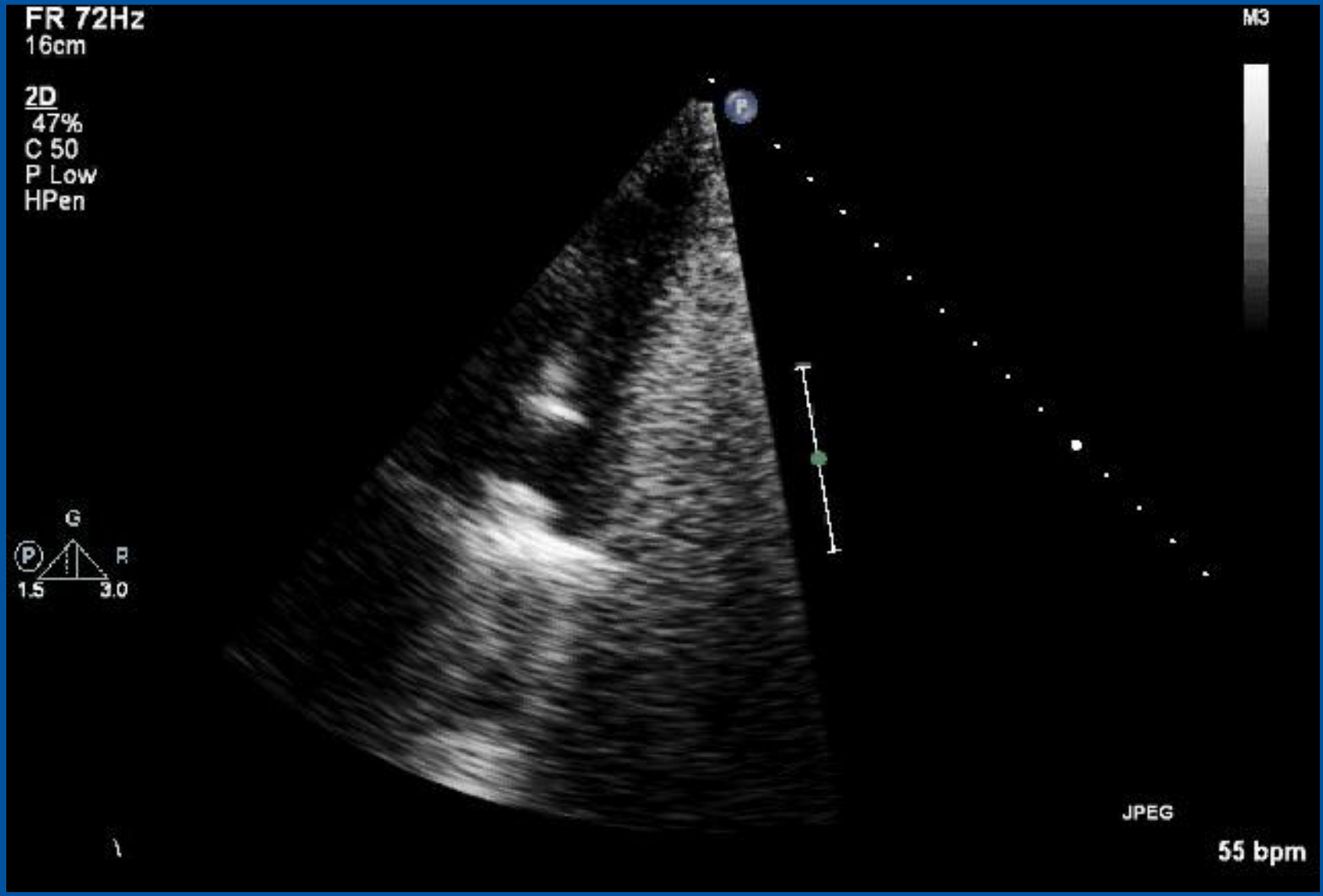

VCS – Glenn from R supraclavicular

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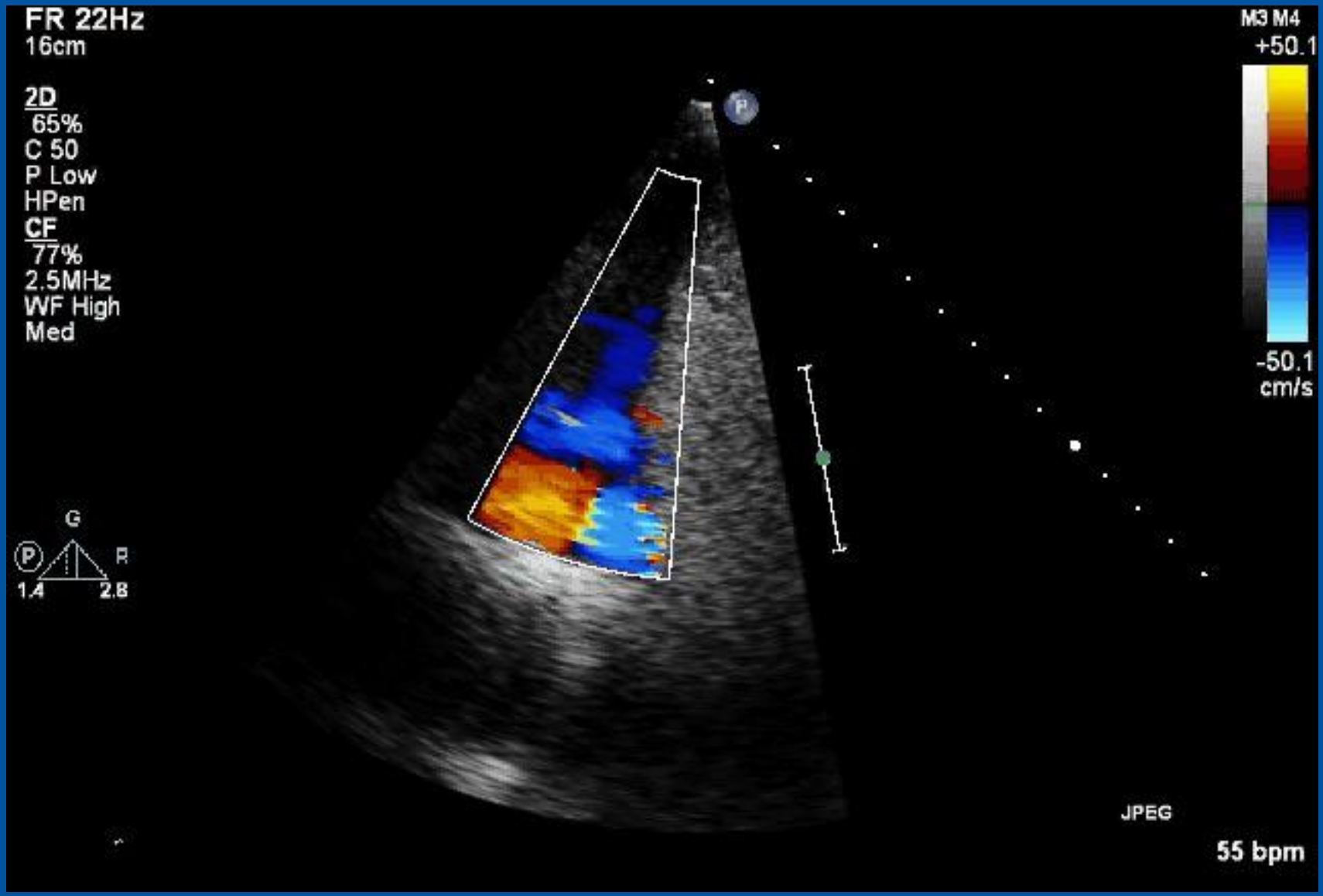

Glenn anastomose

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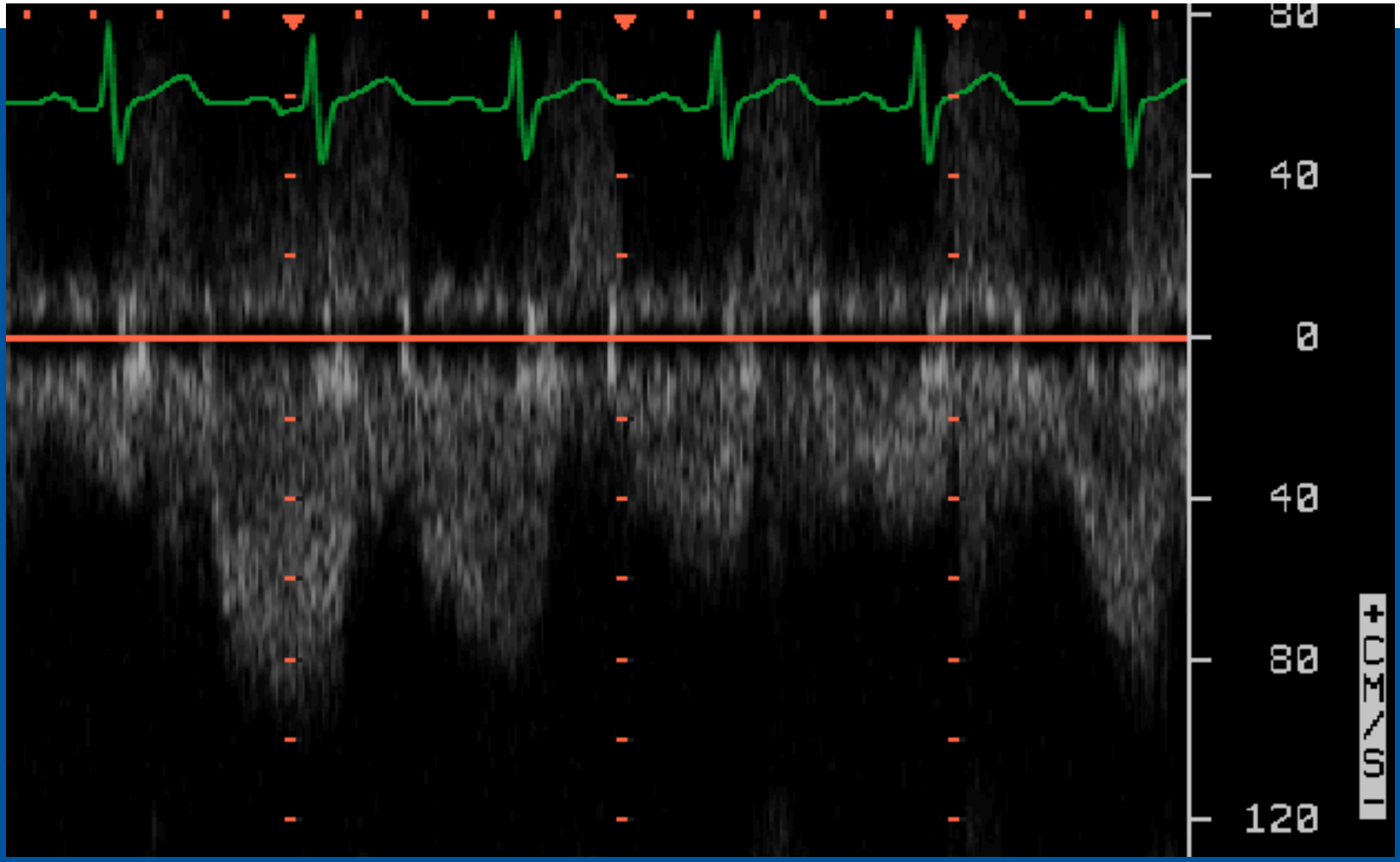

- VCS

APL from SSN; almost always obtainable

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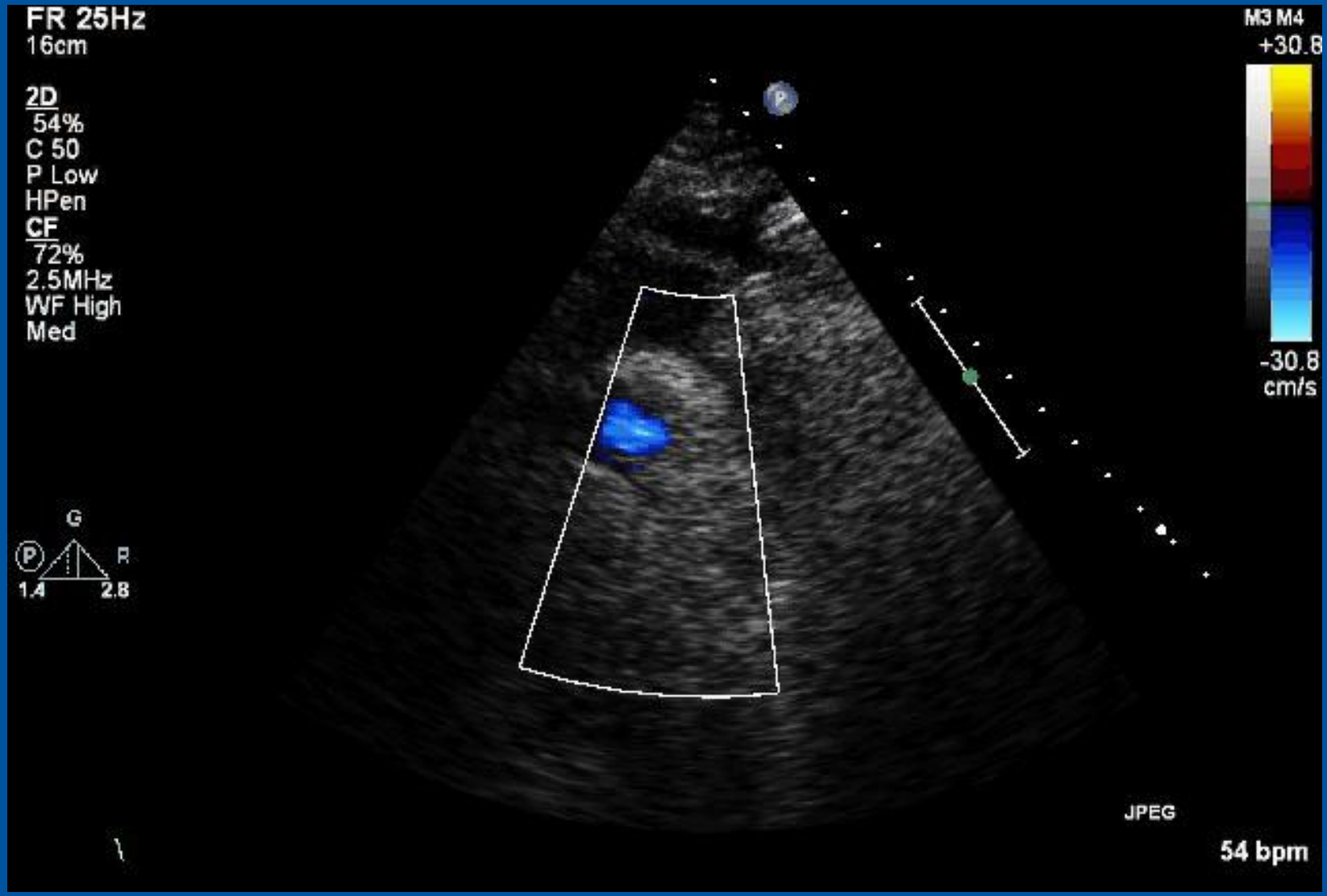

APL bij TCPC

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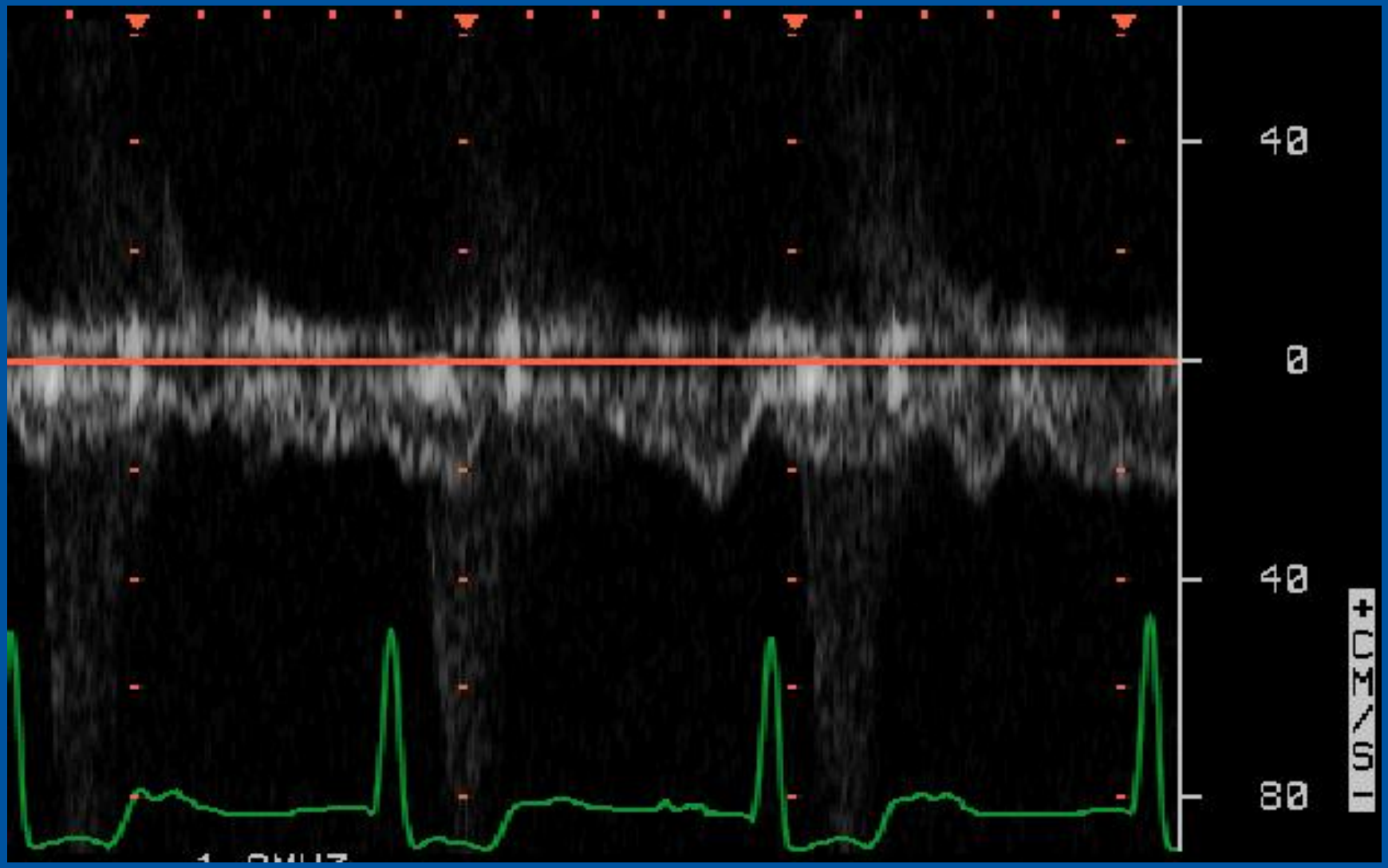

From SSN: LPA

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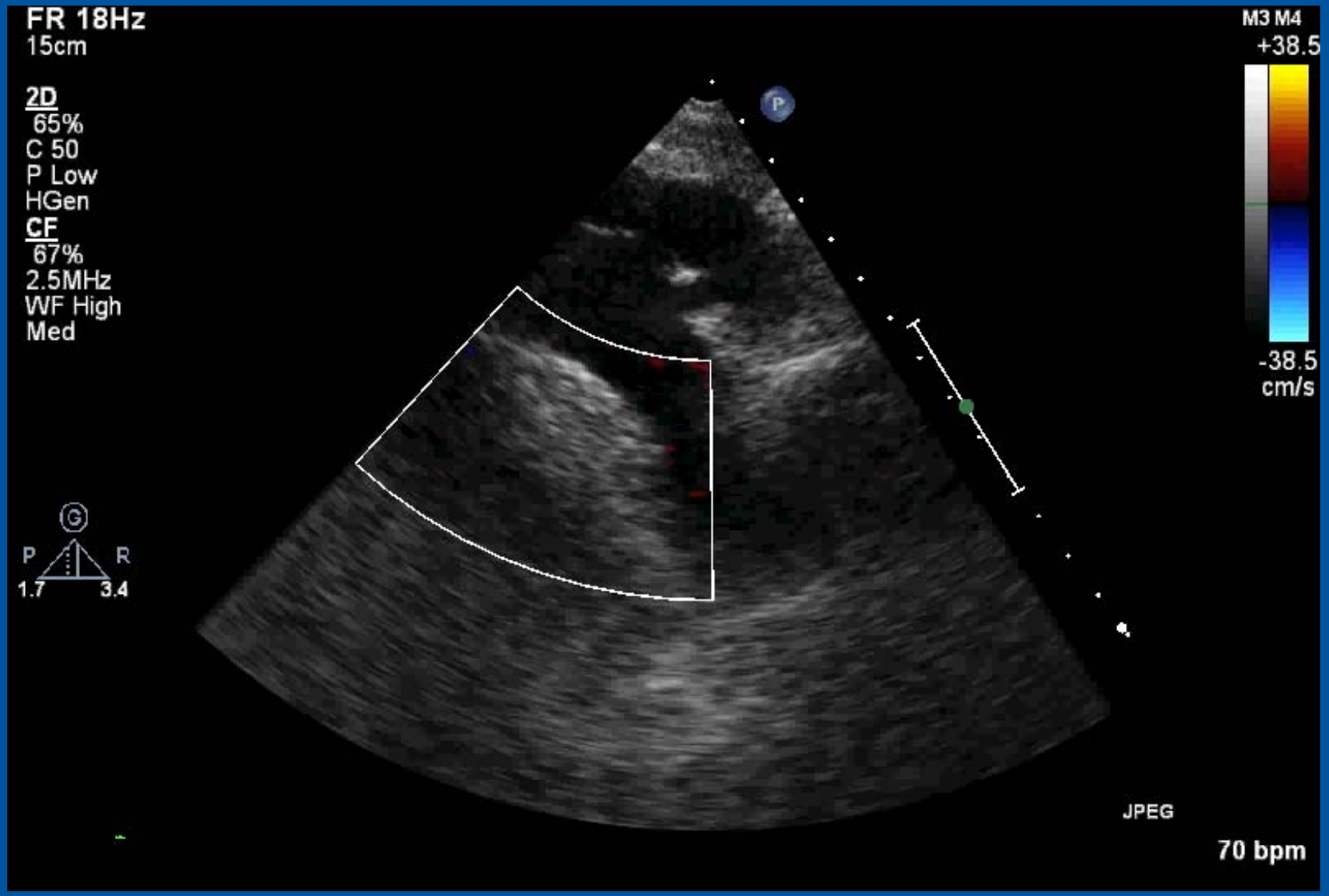

From SSN: LPA

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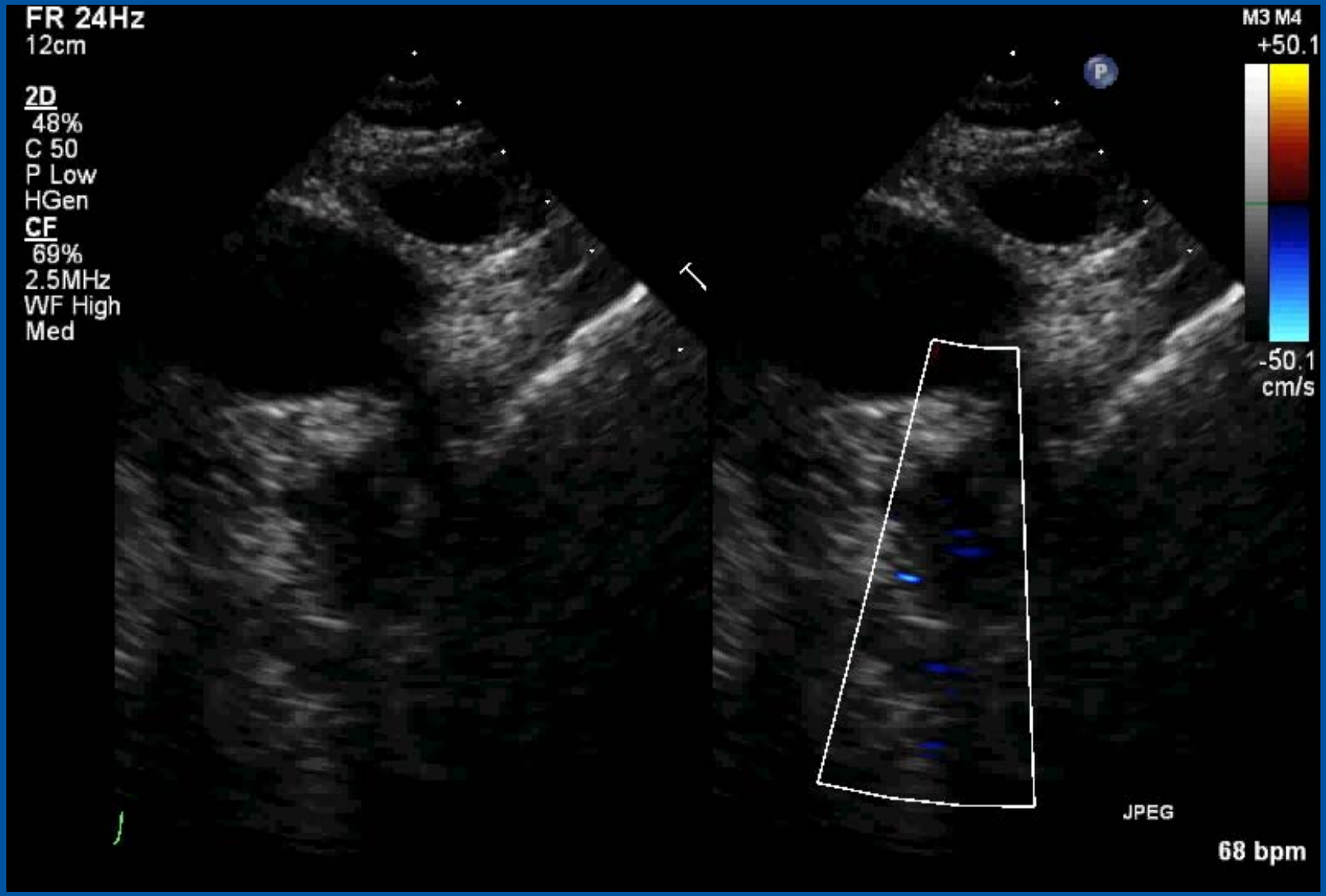

APL in LA – AP connection

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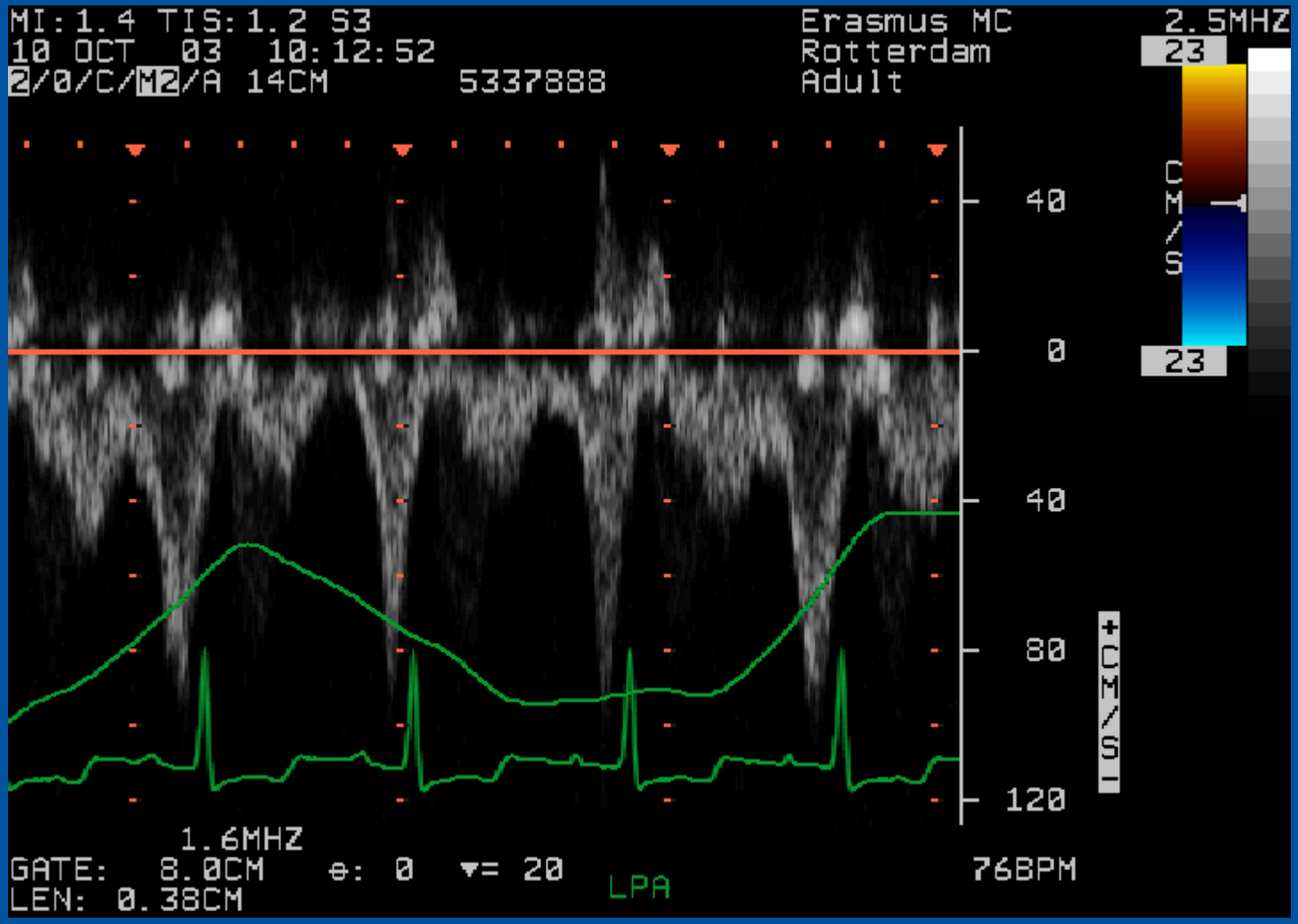

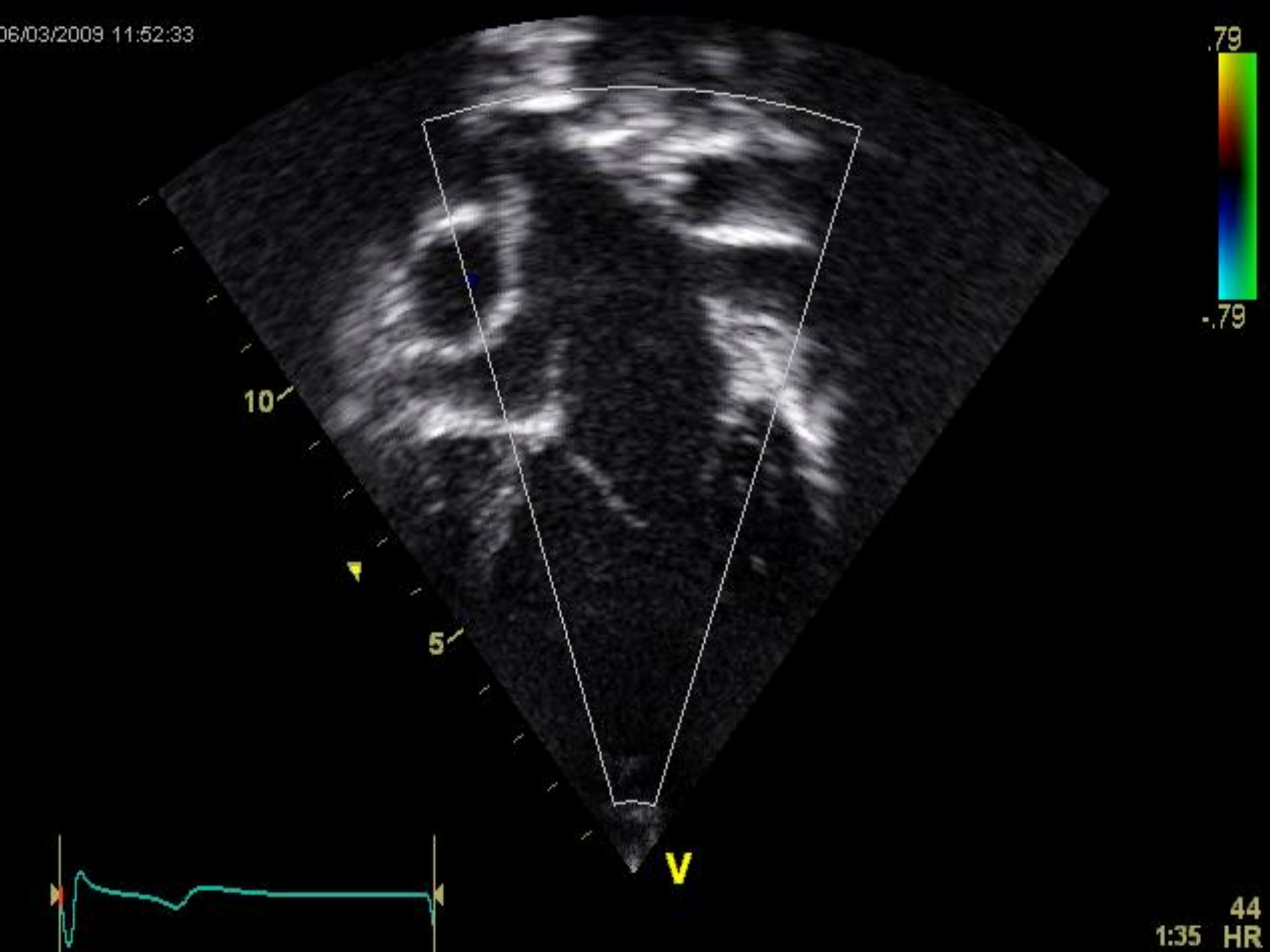
06/03/2009 11:52:33
.79
-.79
10
5
V
44
1:35 HR

R upper pulmonary vein

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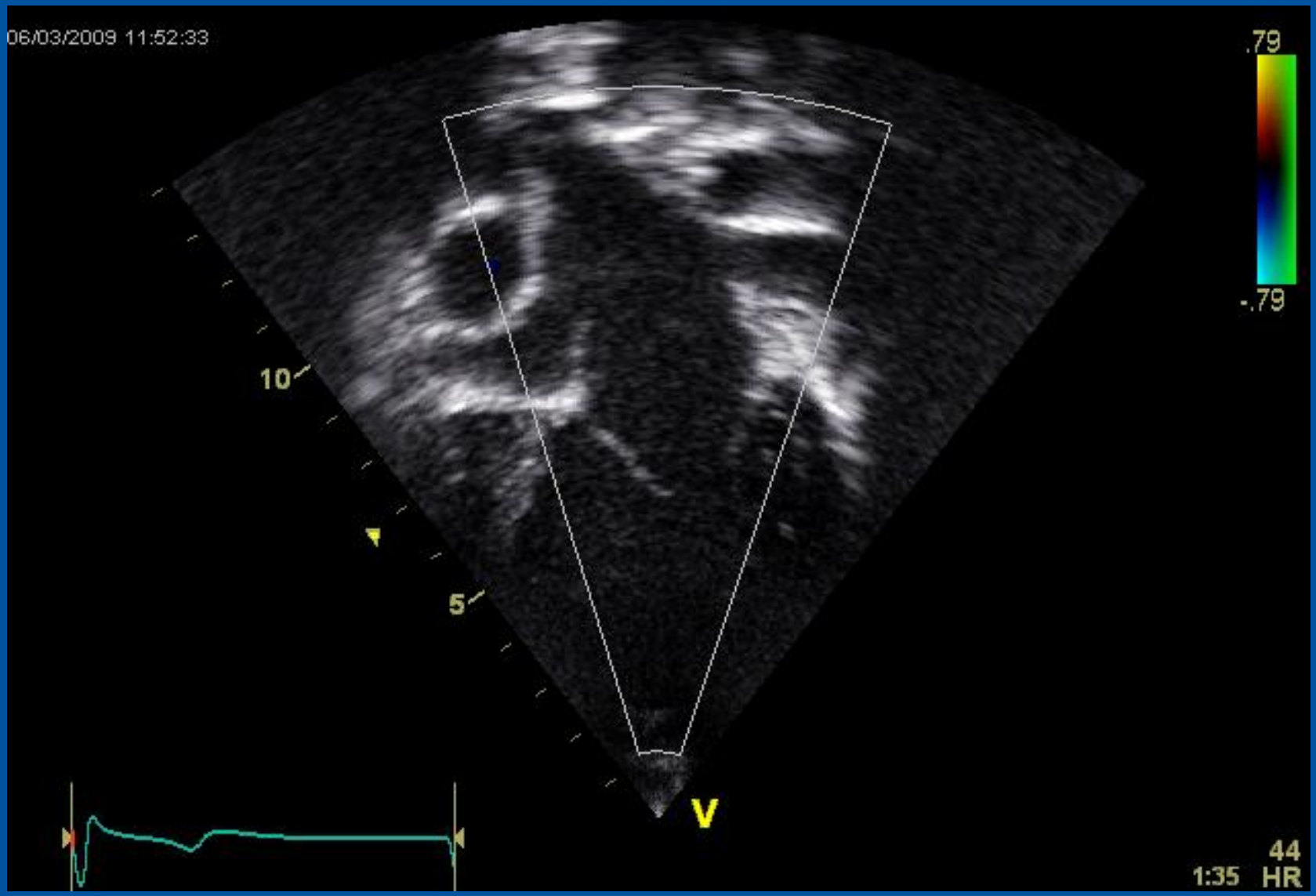

PV Doppler

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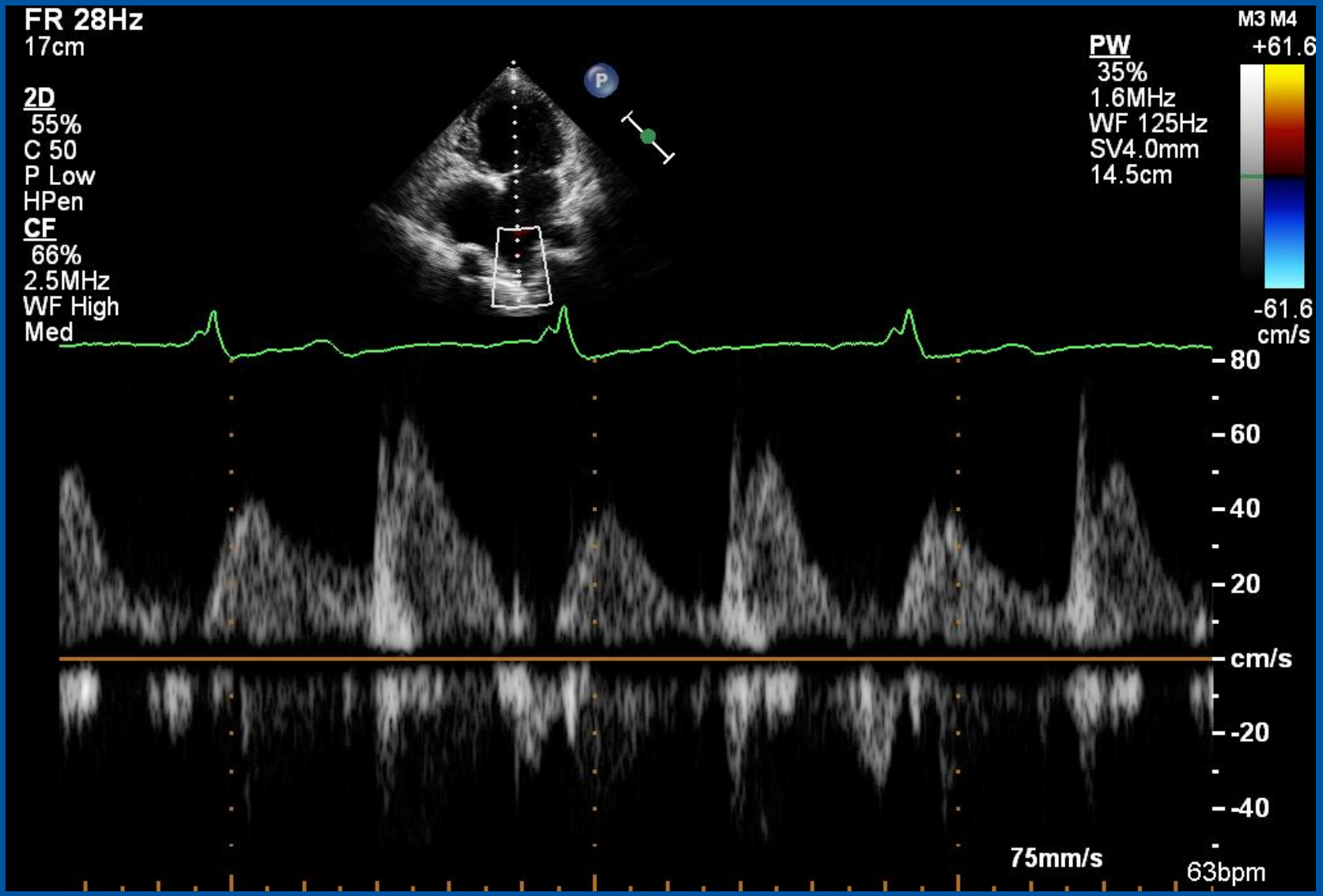

RUPV obstruction (RA – RV connection)

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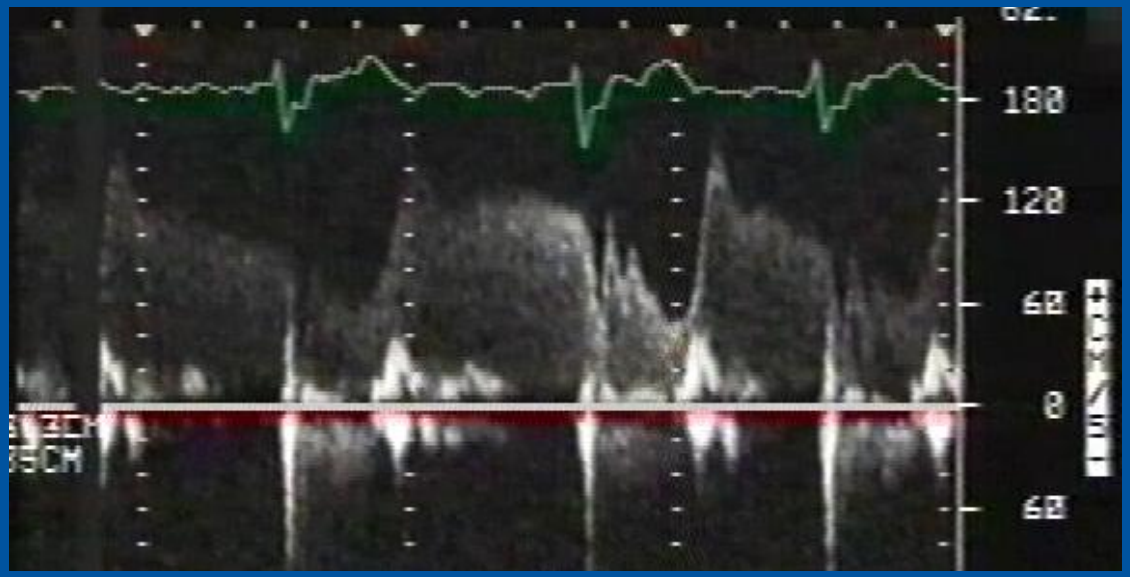
•AP4K



•PW

•Always look for the R pulmonary veins, especially if the IAS is bulging

Dilated RA in RA-AP connection beware of R PV obstruction

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RUPV obstruction

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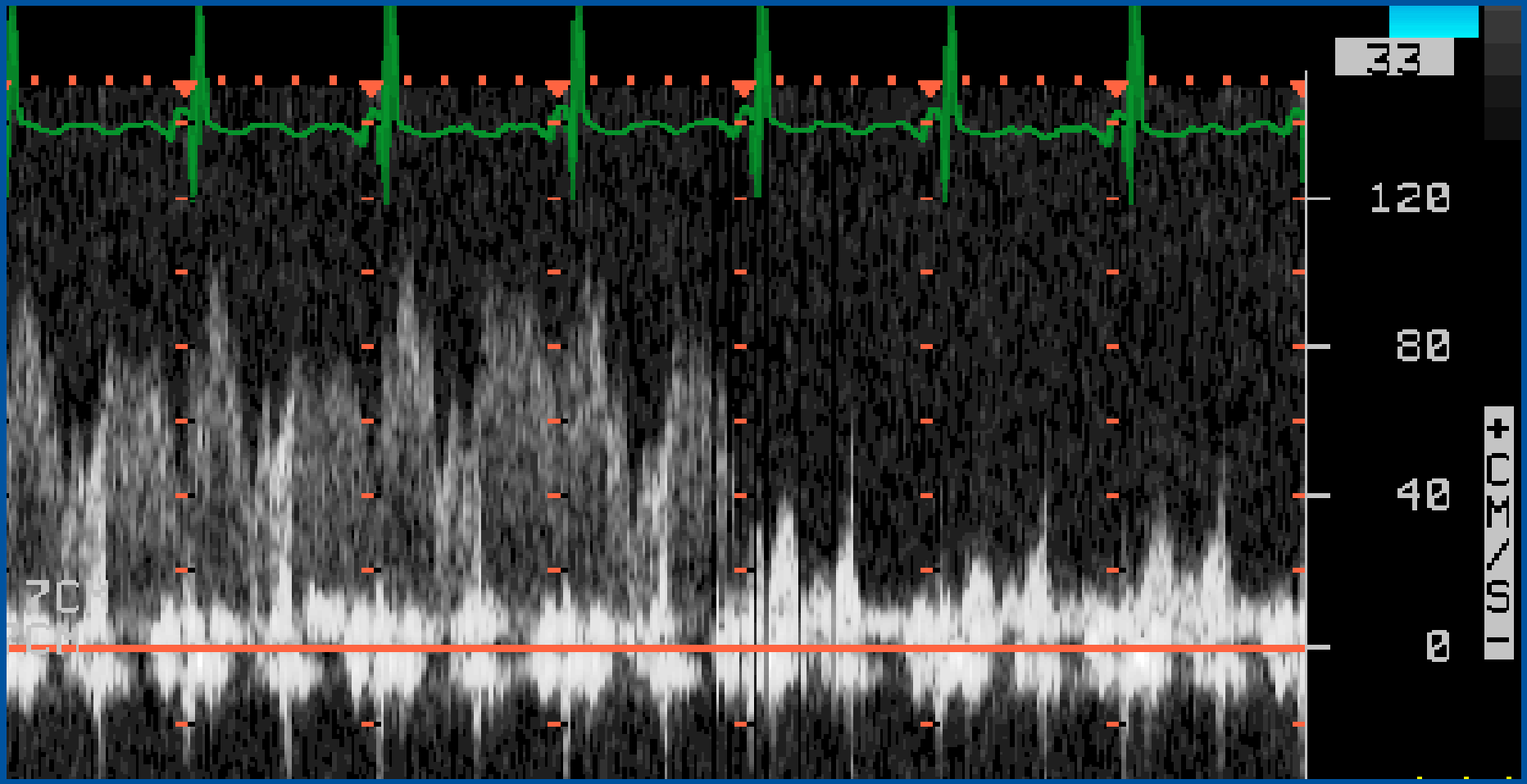

Pulsed wave Doppler (“pull-back”)

Conclusion

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Conclusion

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Why a Fontan-circulation

Indications

How does it work

Types of Fontan circulation

Historical overview → essential for the understanding of echo& Doppler images

Role of echocardiography in the follow-up

*Measuring **function** in a reproducible way for longitudinal follow up*

Conclusion

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Fontan & echo = difficult & fascinating

You need to understand Fontan physiology, especially when you consider intervention
(elective redo of "old" RA- PA to TCPC)

Echo is very helpful, but do not hesitate to send a patient to the cathlab when in doubt

$V = P/R$