Difficult echocardiography in an adult patient with “repaired” congenital heart disease

Markus Schwerzmann, MD
Center for Congenital Heart Disease
University of Bern

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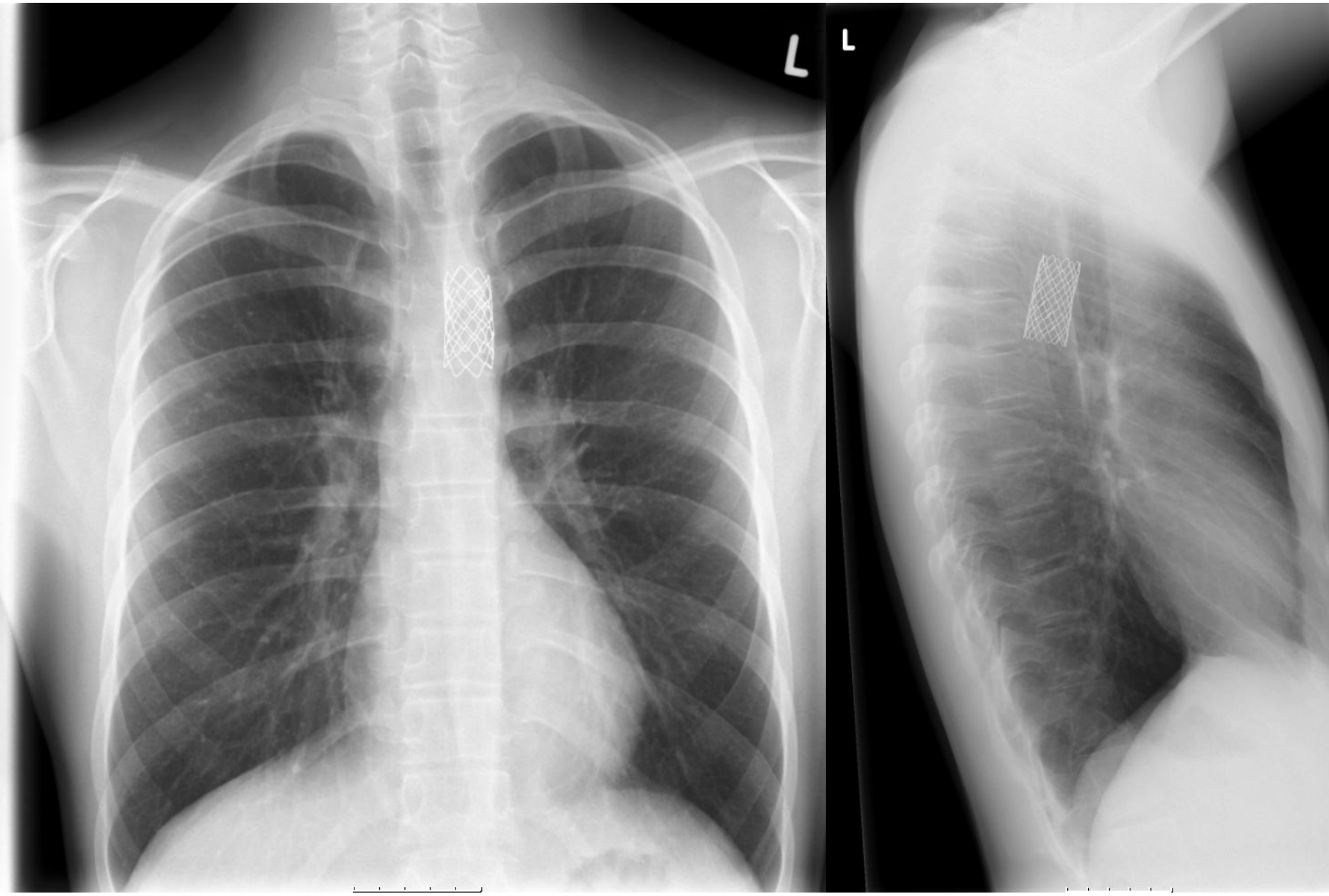
Clinical history

19 year old male, 168 cm, 54 kg, BMI 19 kg/m^2

Past medical history:

- Peter‘s anomaly (thinning and clouding of the cornea and attachment of the iris to the cornea, which causes blurred vision)
- At age 6: bicuspid aortic valve, mild coarctation
- At age 17: balloon dilatation of native coarctation /stent placement at the site of aortic coarctation; residual peak gradient across the stent: 2 mmHg

Regular follow-up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Clinical exam

Oxygen saturation 98% at room air

Heart rate 94 bmp and regular; normal S1, S2 with early systolic ejection click (tambour sound), 2/6 ejection murmer and 2/4 early diastolic murmer over the aorta, peripheral pulses easily palpable and not bounding, no radio-femoral delay.

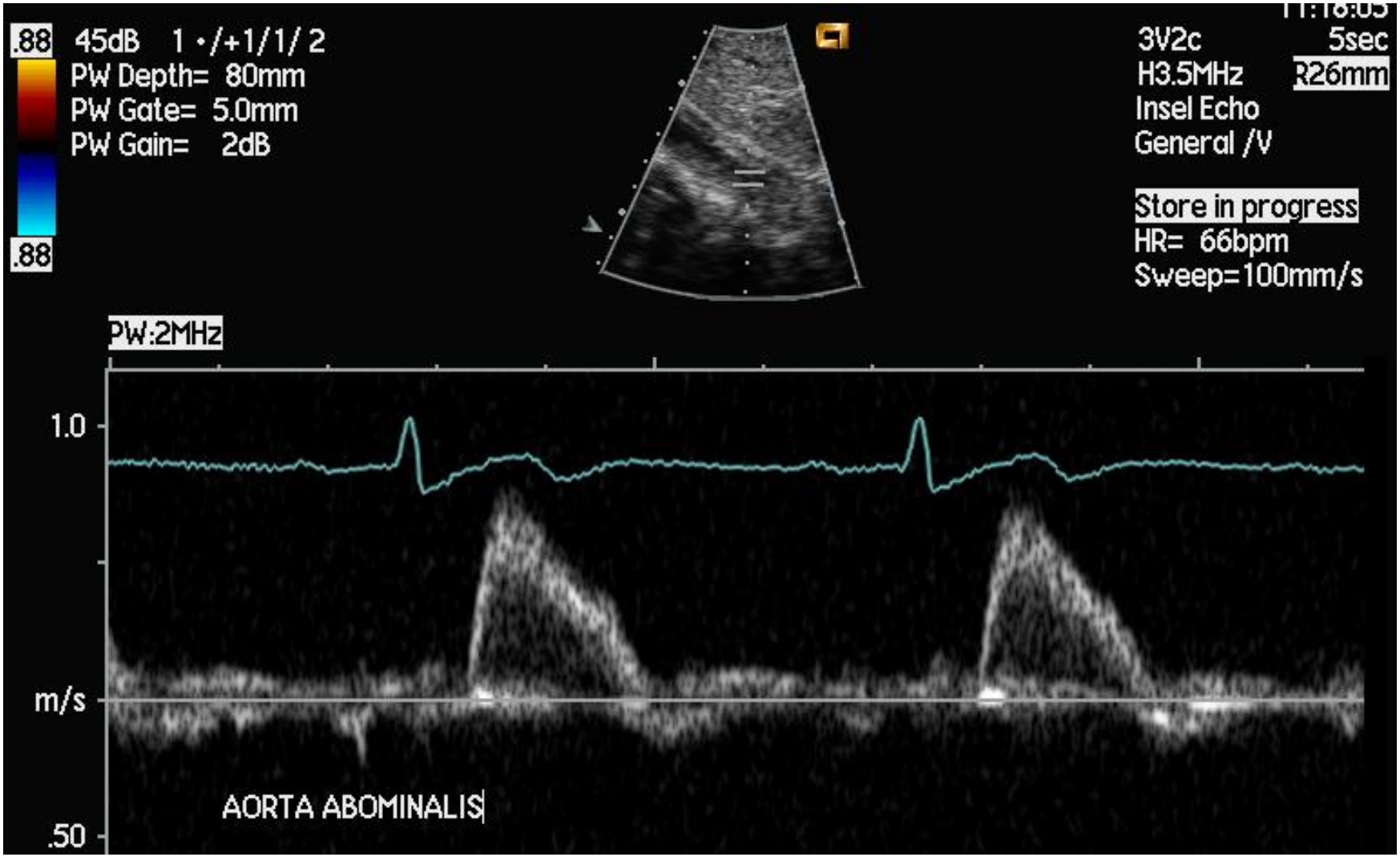
Blood pressure was 146/66 mmHg right arm, 136/64 mmHg left arm, 148 mmHg systolic at the right leg (dorsalis pedis).

Medication: perindopril 5 mg OD

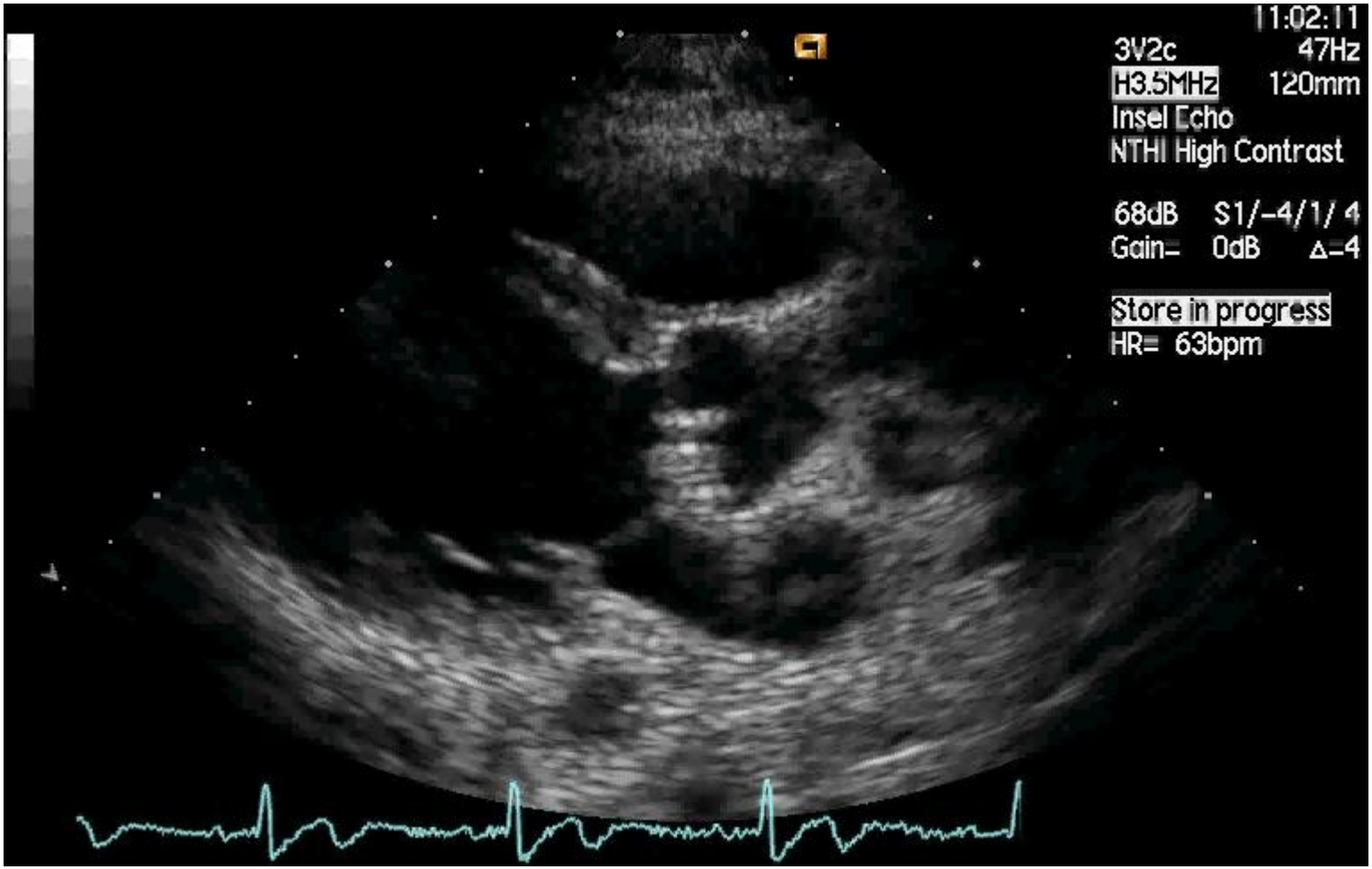
Question

- Based on the clinical findings, what would you expect to find at echocardiography

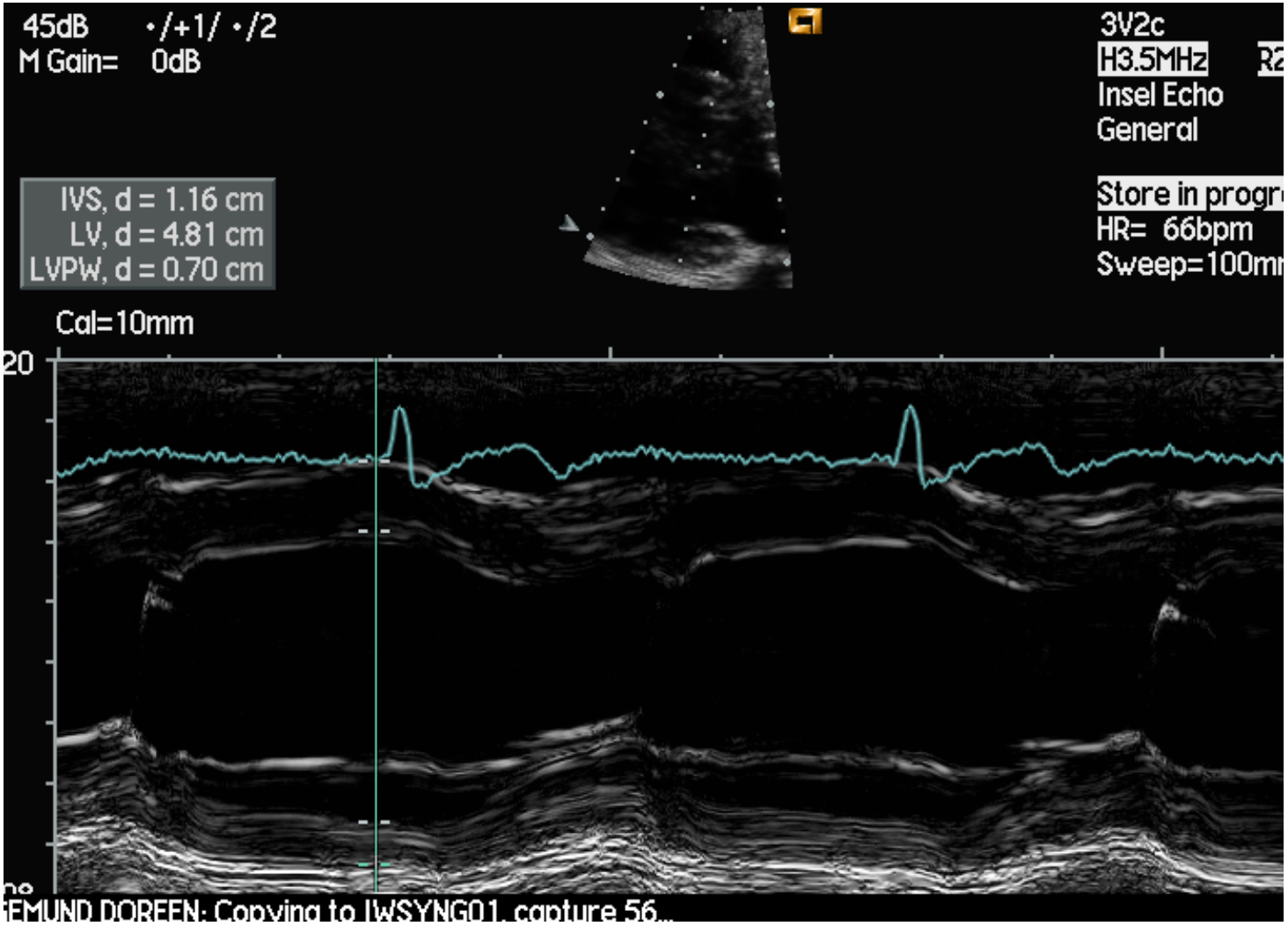
TTE


.88
45dB 1 •/+1/1/ 2
PW Depth= 80mm
PW Gate= 5.0mm
PW Gain= 2dB
3V2c
5sec
H3.5MHz
R26mm
Insel Echo
General /V
Store in progress
HR= 66bpm
Sweep=100mm/s
.88
PW:2MHz
1.0
m/s
.50
AORTA ABOMINALIS

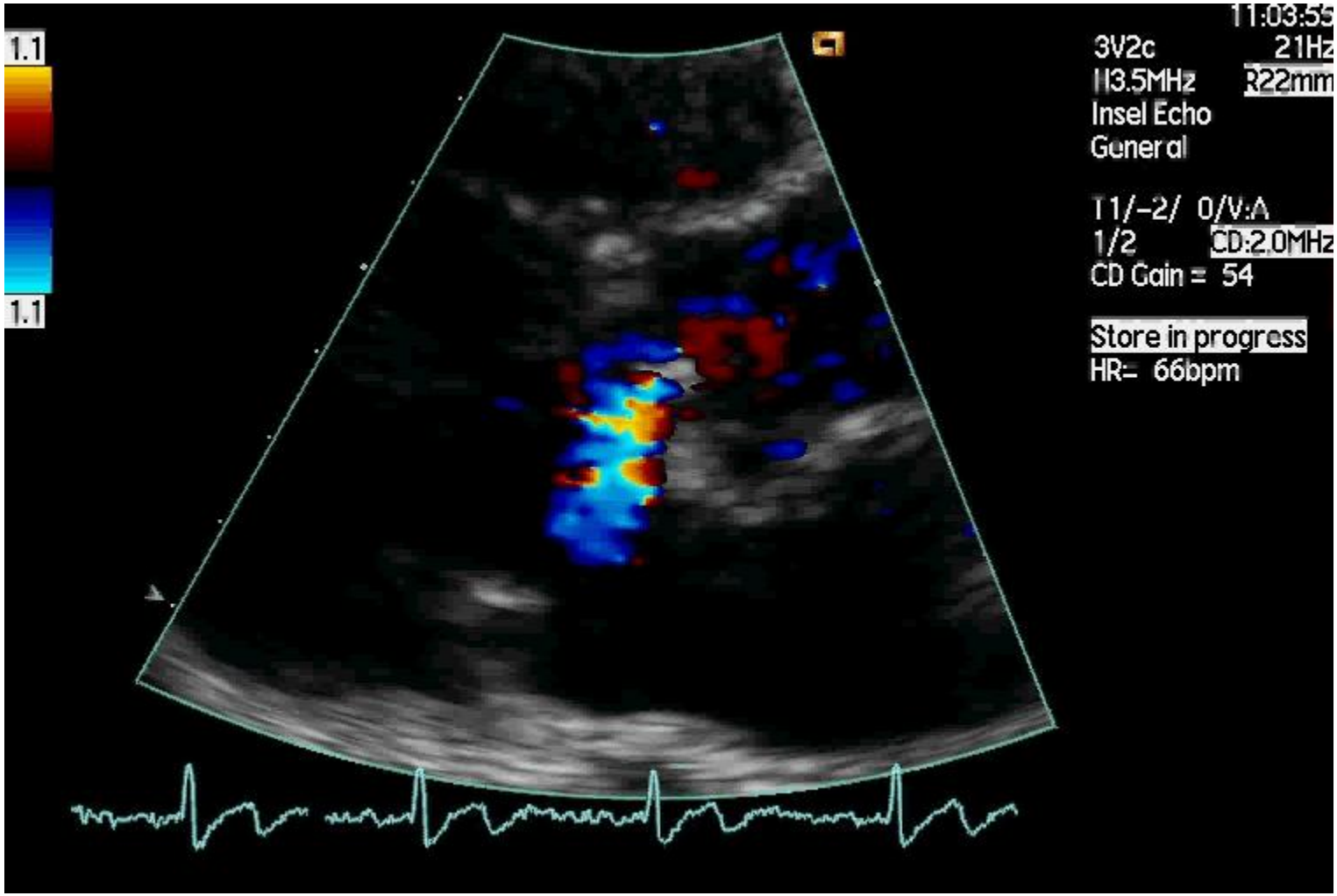
TTE


11:02:11
3V2c
47Hz
H3.5MHz
120mm
Insel Echo
NTHI High Contrast
68dB
S1/-4/1/ 4
Gain=
0dB
Δ=4
Store in progress
HR= 63bpm

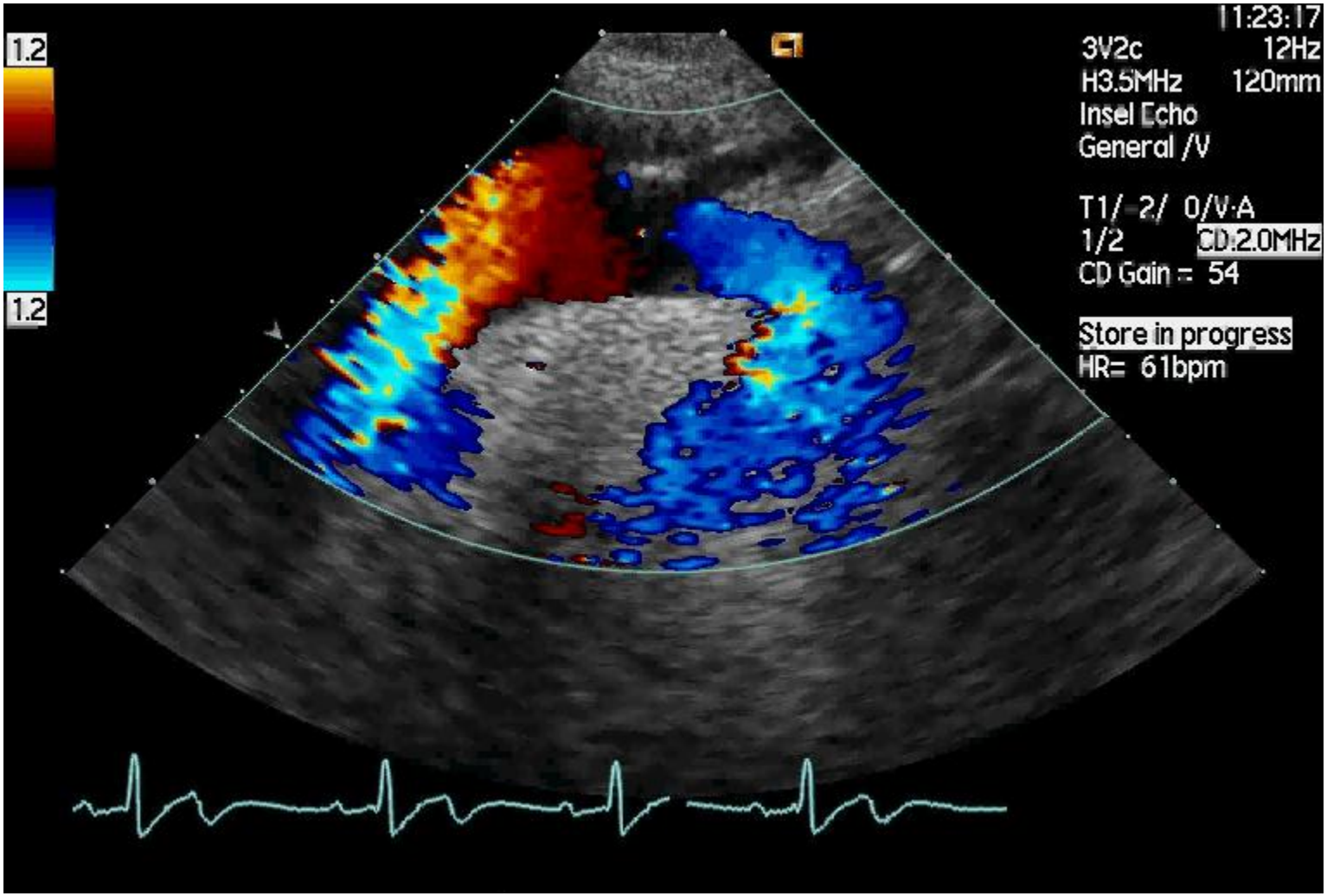
TTE


45dB •/+1/ •/2
M Gain= 0dB
3V2c
H3.5MHz
Insel Echo
General
Store in progr
HR= 66bpm
Sweep=100m
IVS, d = 1.16 cm
LV, d = 4.81 cm
LVPW, d = 0.70 cm
Cal=10mm
20
EMUND DOREEN: Copying to IWSYNG01, capture 56...

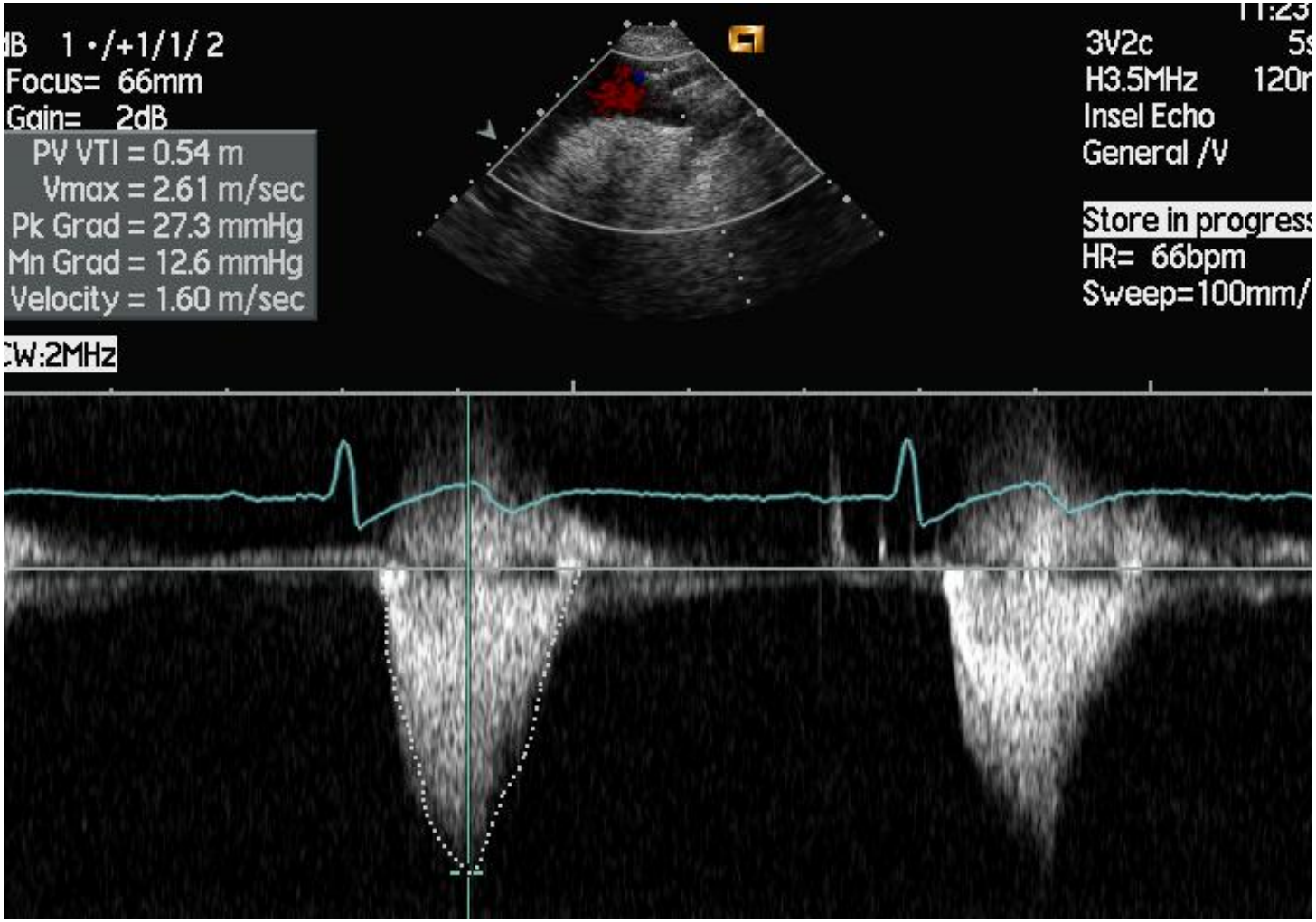
TTE


1.1
1.1
3V2c
21Hz
3.5MHz
R22mm
Insel Echo
General
T1/-2/ 0/V:A
1/2
CD:2.0MHz
CD Gain = 54
Store in progress
HR= 66bpm

TTE


1.2
1.2
11:23:17
3V2c
12Hz
H3.5MHz
120mm
Insel Echo
General /V
T1/ 2/ 0/V·A
1/2
CD:2.0MHz
CD Gain = 54
Store in progress
HR= 61bpm

TTE


B 1 •/+1/1/ 2
Focus= 66mm
Gain= 2dB
PV VTI = 0.54 m
Vmax = 2.61 m/sec
Pk Grad = 27.3 mmHg
Mn Grad = 12.6 mmHg
Velocity = 1.60 m/sec
CW:2MHz
3V2c
H3.5MHz
Insel Echo
General /V
Store in progress
HR= 66bpm
Sweep=100mm/

TTE

Report:
- Bicuspid aortic valve (fused right and left-coronary cusp) with mild to moderate regurgitation, no aortic stenosis
- ascending aorta not dilated
- Peak/mean gradient at the isthmic region: 15/30 mmHg

What do you think?

- I‘m confindent with the clinical exam indicating no important blood pressure differences between upper and lower extremity
  ⇨ the peak gradient of 30 mmHg measured at echocardiography is not correct

- I‘m confindent with the echo exam indicating a > 20 mmHg blood pressure gradient across the stent at the coarctation site
  ⇨ the clinical assessment is not reliable (size BP cuff, no Doppler used for peripheral pulses, collateral vessels…)

- We need further investigations

Physics

Relationship between pressure and flow

- Conservation of energy:

  pressure = potential energy

  flow = kinetic energy

- Bernoulli equitation (fluid, steady laminar flow):

  energy density = $P + \frac{1}{2} \rho v^2 + \rho g h = \text{constant}$

  $P_1 + \frac{1}{2} \rho v_1^2 + \rho g h_1 = P_2 + \frac{1}{2} \rho v_2^2 + \rho g h_2$

  $P_1 - P_2 = \frac{1}{2} \rho (v_2^2 - v_1^2)$

- Bernoulli equitation complete (fluid, pulsatile)

  ΔP = convective + inertial + shear stress component

This means

• the simplified Bernoulli formula cannot be used to quantify pressure gradients in coarctation

• even the more complex Bernoulli equitation $P_1 – P_2 = ½ \rho (v_2^2 - v_1^2)$ does not take into account the viscous friction component, particularly in stented vessels

⇨ Peak echo gradients do not reflect real pressure gradients in the setting of aortic coarctation/stenting

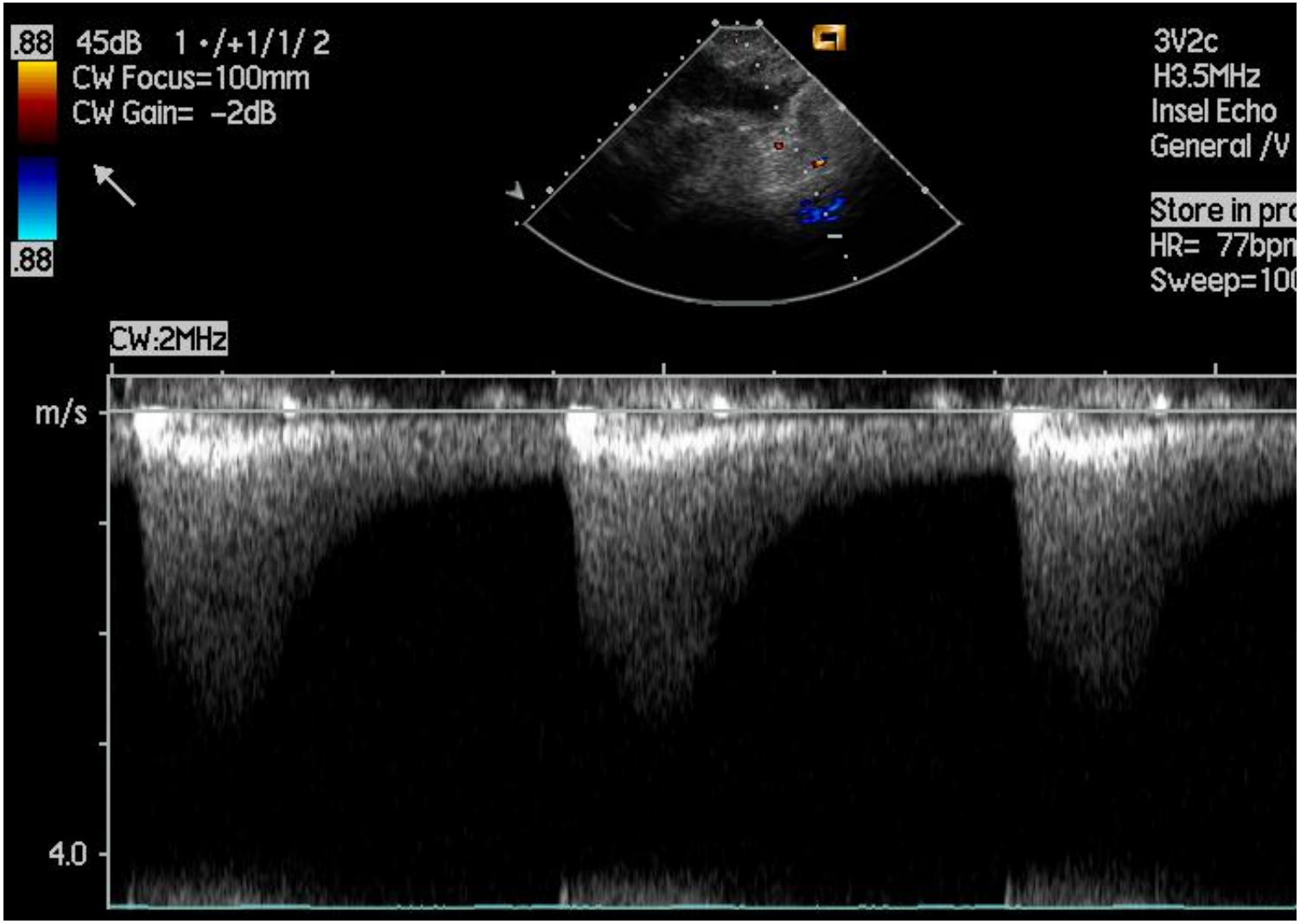
Echo signs of significant coarctation in the adult

- Doppler flow signal isthmus - serrated pattern
  - rapid acceleration
  - early high-velocity
  - gradual deceleration throughout diastole (diastolic spill over)

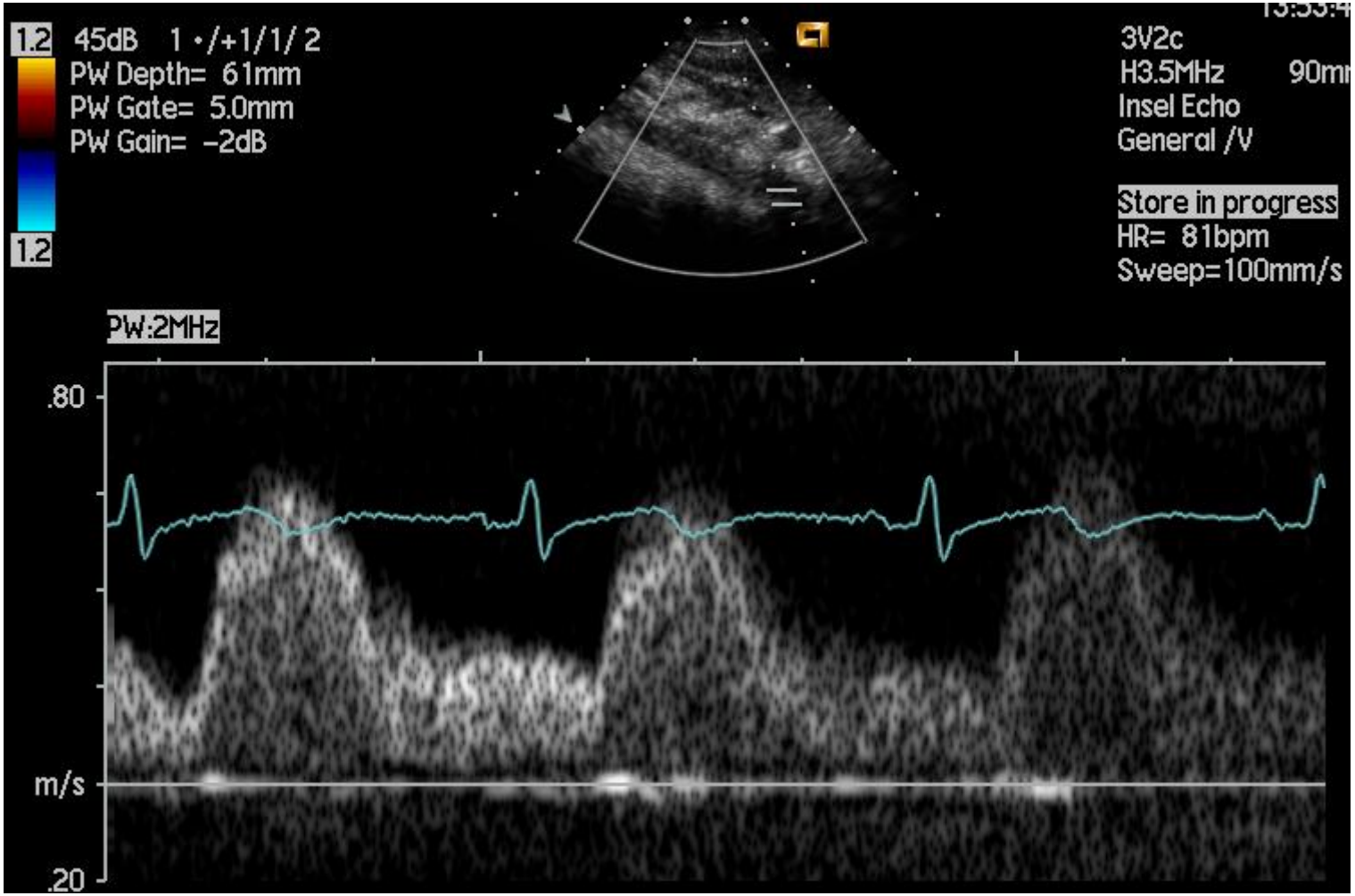
- Doppler flow signal abdominal aorta
  - low-velocity systolic-diastolic flow

- Signs of left ventricular pressure overload:
  - LV hypertrophy
  - LV diastolic dysfunction
  - Left atrial enlargement

Echo signs of significant coarctation in the adult



Echo signs of significant coarctation in the adult



6 months later…

• presents with exertional chest pain 8/10, responding to nitroglycerin, no symptoms at rest

• physicial exam: unchanged; BP right arm supine 152/78 mmHg, systolic pressure right leg 148 mmHg

• ECG unchanged;
• TTE unchanged

• Exercise ECG: left-sided stabbing chest pain beginning at mild/moderate exercise, increasing in intensity during exercise; BP peaks at 210/92 mmHg; no ECG changes

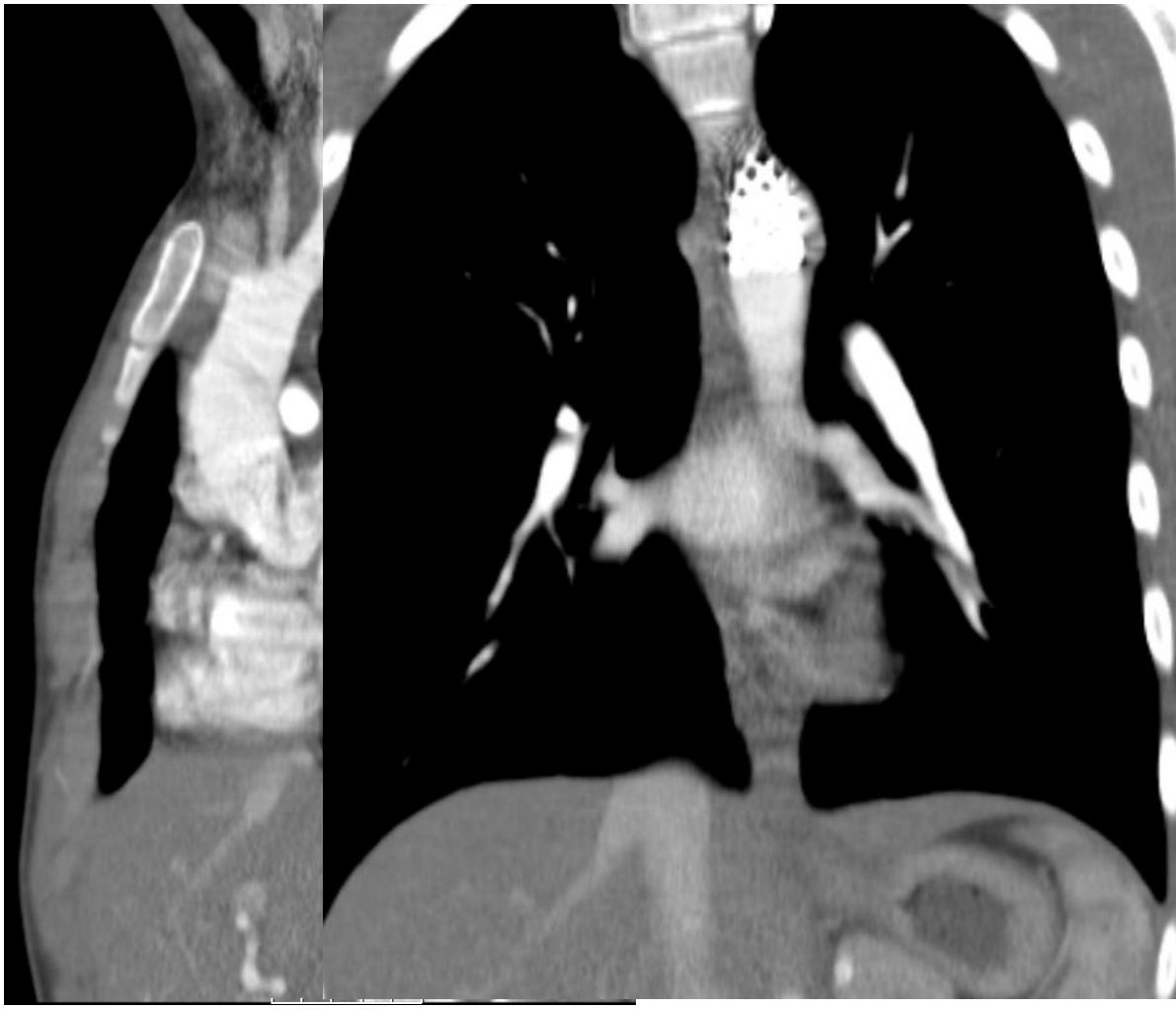
Questions – What is your next step

- increase perindopril to 10 mg OD; pain killers (paracetamol) as needed

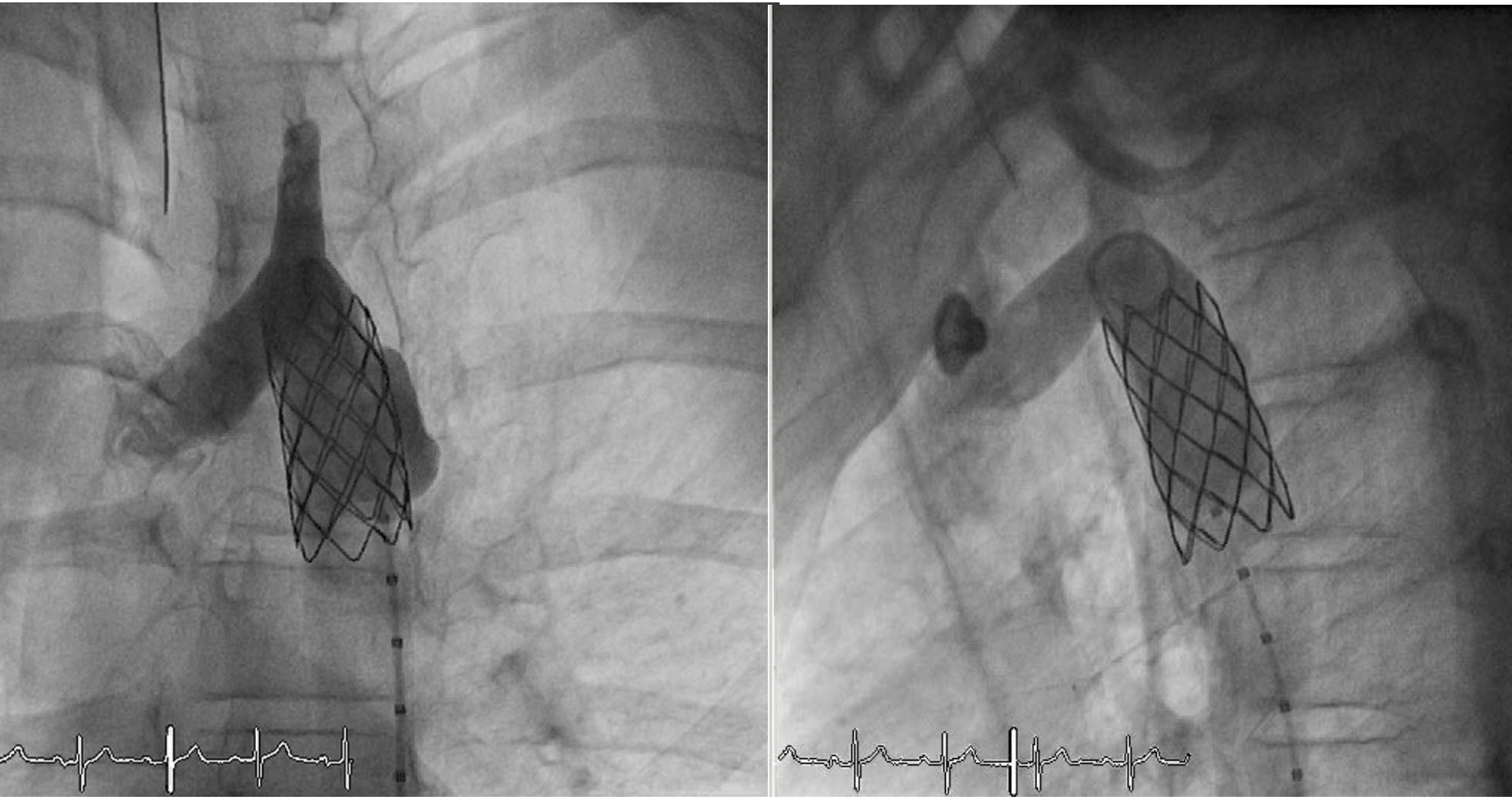
- further investigations are needed

- no additional steps are needed – close f/u in 2 months

CT-scan



Cath



Cath

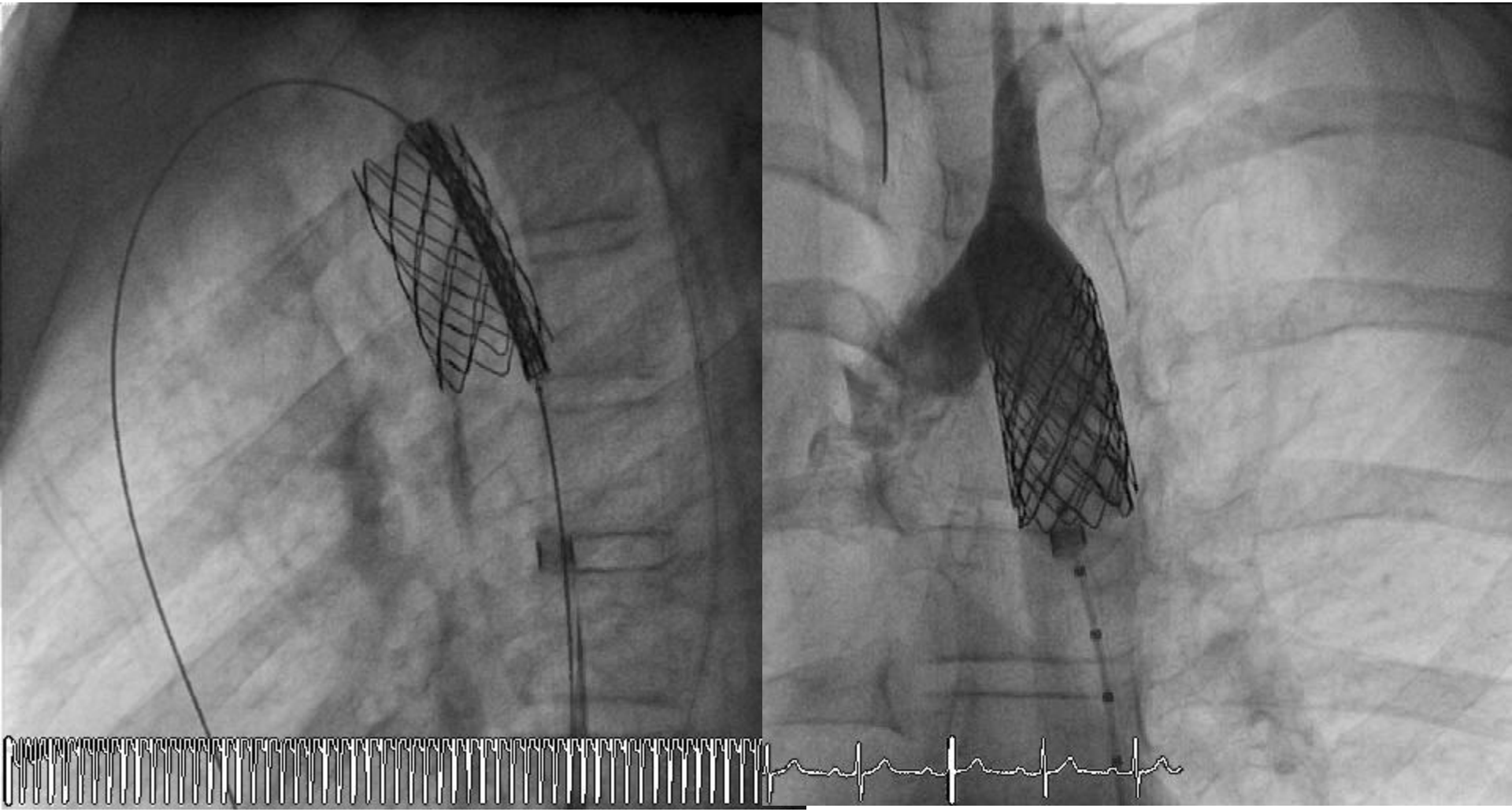


Table 7 Intermediate Follow-up Outcomes by Integrated Imaging

	Surgery (n = 16)	Balloon (n = 16)	Stent (n = 56)	p Value (2-Sided)
Any complications*	25.0%	43.8%	12.5%	0.020‡
Aortic wall injury	12.5%	43.8%	7.1%	0.003‡
Dissection/intimal tear	0.0%	6.3%	1.8%	0.598
Aneurysm	12.5%	43.8%	5.4%	<0.001
Coarct:Dao ratio, mean	0.98	0.79	0.80	0.011‡
Coarct:Dao ratio ≥0.6	88%	93%	89%	1.000
Any reobstruction	18.8%	18.8%	14.3%	0.923
Mild†	6.3%	18.8%	12.5%	
Moderate	6.3%	0%	1.8%	
Severe	6.3%	0%	0%	

*Defined as any moderate to severe reobstruction, aortic wall injury (aneurysm, dissection, intimal tear) or stent fracture. †Mild reobstruction was not considered as a complication in our analysis. ‡$p < 0.05$.

Coarct:Dao = narrowest coarctation dimension (mm)/the dimension of the descending aorta at the level of the diaphragm (mm).

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Discussion – Echo in coarctation

• remember its physical limitations, particularly in patients with a stent

• Severity of coarctation:
- flow pattern in descending/abdominal aorta

• Also assess
- aortic valve and ascending aorta
- associated lesions

• Not very sensitive for the detection of late complications after coartation repair
- additional imaging methods are needed