2ο ΠΑΝΕΛΛΗΝΙΟ ΣΥΝΕΔΡΙΟ ΠΝΕΥΜΟΝΙΚΗΣ ΥΠΕΡΤΑΣΗΣ

Πνευμονική Υπέρταση - Απεικόνιση ΙΙ: Περιστατικά πνευμονικής υπέρτασης με απεικονιστικό ενδιαφέρον

Σοφία Μ. Αράπη
Επιμελήτρια Α' Καρδιολογικής Κλινικής
ΓΝΑ 'Γ. Γεννηματάς'

Disclosures: None

Case presentation

- 39-year old female patient, with VSD- Eisenmenger syndrome
- She reports that VSD was diagnosed at birth. At 6months, considered inoperable due to severe PH
- 5- year old: diltiazem 60mg t.i.d. initiated
- 25-year old: illoprost added after RHC (5 inh/day), WHO FC II-III, improvement in WHO FC reported
- 27-year old: sildenafil 20 mg t.i.d. (stable WHO FC II, no RHC)

RHC

PASP: 132mmHg
Diast PAP: 64mmHg
Mean PAP: 94mmHg
RAP: 10mmHg
PAWP: 13mmHg

BNP = 111pg/ml

ECHOs: VSD described as either subaortic or perimembranous, dimensions: 1- 1,2cm, R-L shunting

RVSP: 70-80mmHg RV increased dimensions, normal function

6MWD: 517m, Borg scale dyspnea 0 to 0.5 / fatigue 1 to 2

Sat O2: 75% to 59%

During follow-up:

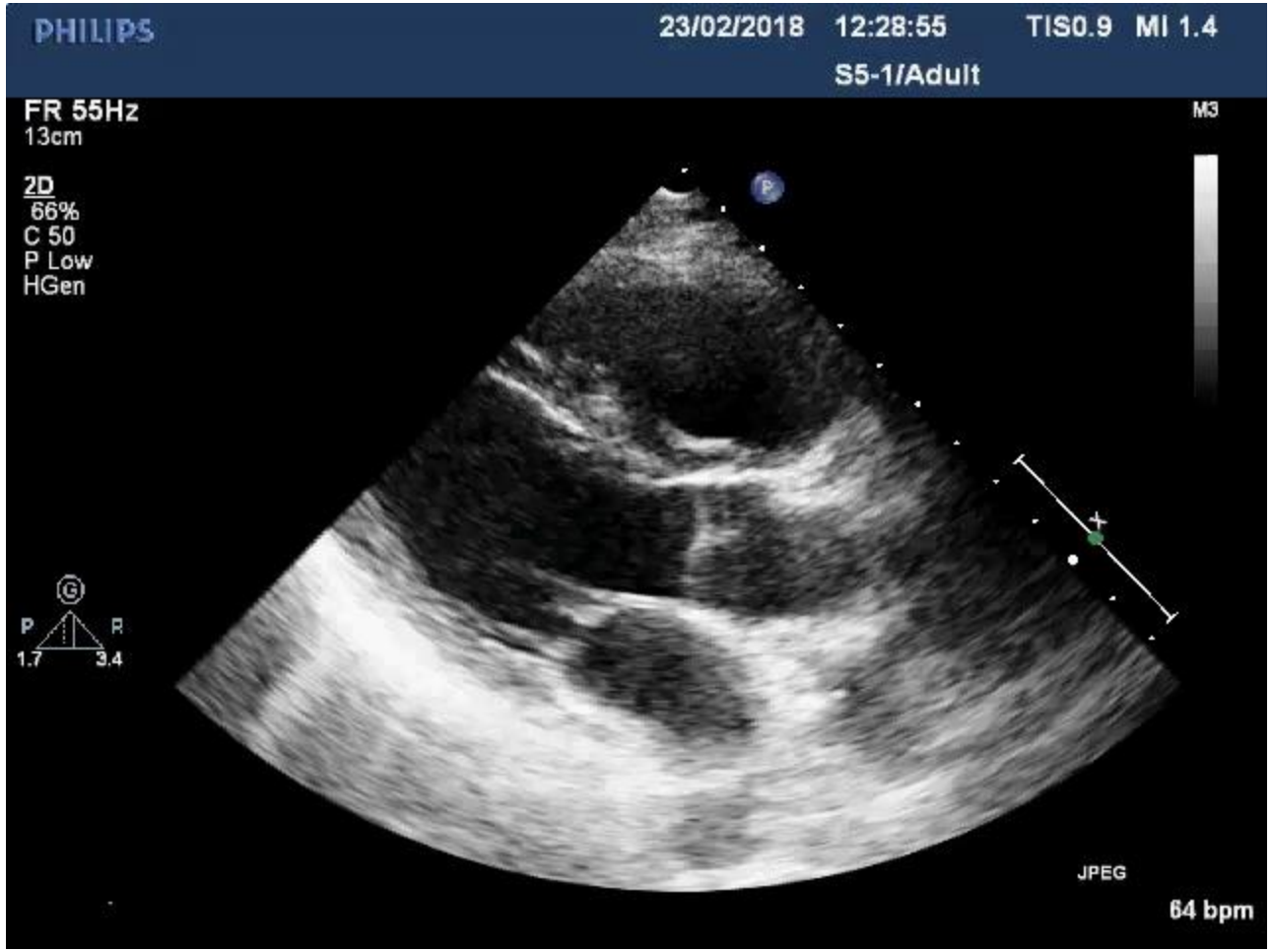
- Uptitration of sildenafil and illoprost inhalations
- Discontinuation of diltiazem
- ERA administered (initially bosentan - substantial increase in ALT, AST led to bosentan discontinuation, ambrisentan administration)

- Supplemental Oxygen therapy (nocturnal), periodically supplemental iron treatment due to low ferritin levels
- Patient required to gradually down-titrate inhalations in order to discontinue

- Currently receiving ambrisentan (10mg q.d.) and sildenafil (20mg t.i.d.)

During follow-up:

- **WHO FC II, no angina or syncope**
- **6MWD:** 630m, **Borg scale dyspnea 0 to 0 / fatigue 0 to 0.5**
  Sat O2: 81% to 74%
- **BNP =** 45 pg/ml
- HcT= 46.8%, MCV 94.7 fL, Fe= 86.9 µg/dl, ferritin 44.7ng/ml, UA: 3.7mg/dl, bil: 2,25mg/dl, Cr: 0.81mg/dL, Cr Cl: 81ml/min, GFR: 83.7 ml/min/1.73m²
- Has not accepted follow-up RHC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2D
66%
C 50
P Low
HGen
M3
P
G
P R
1.7 3.4
JPEG
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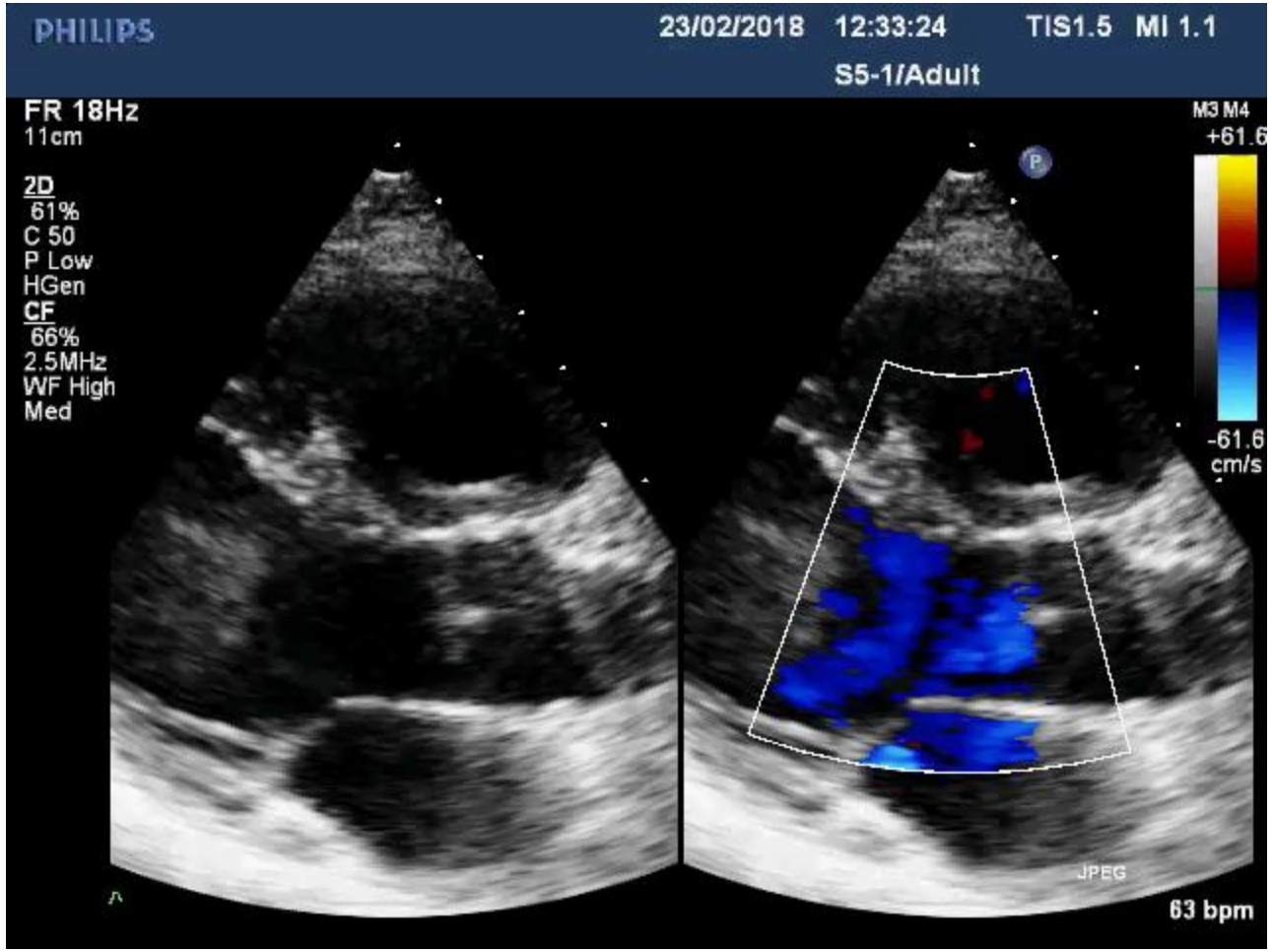
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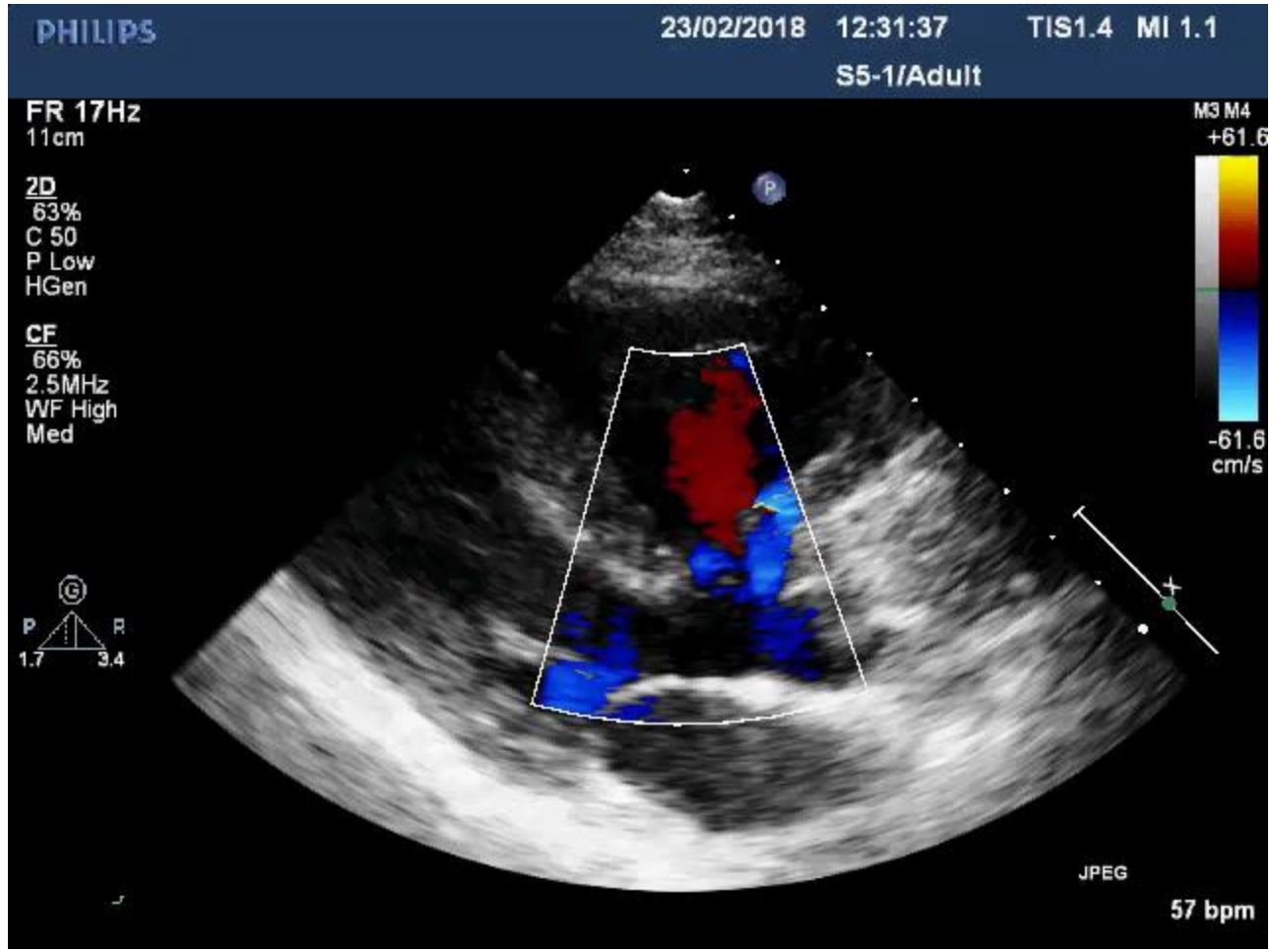
-61.6
cm/s
JPEG
63 bpm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HGen
CF
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cm/s
P
R
1.7
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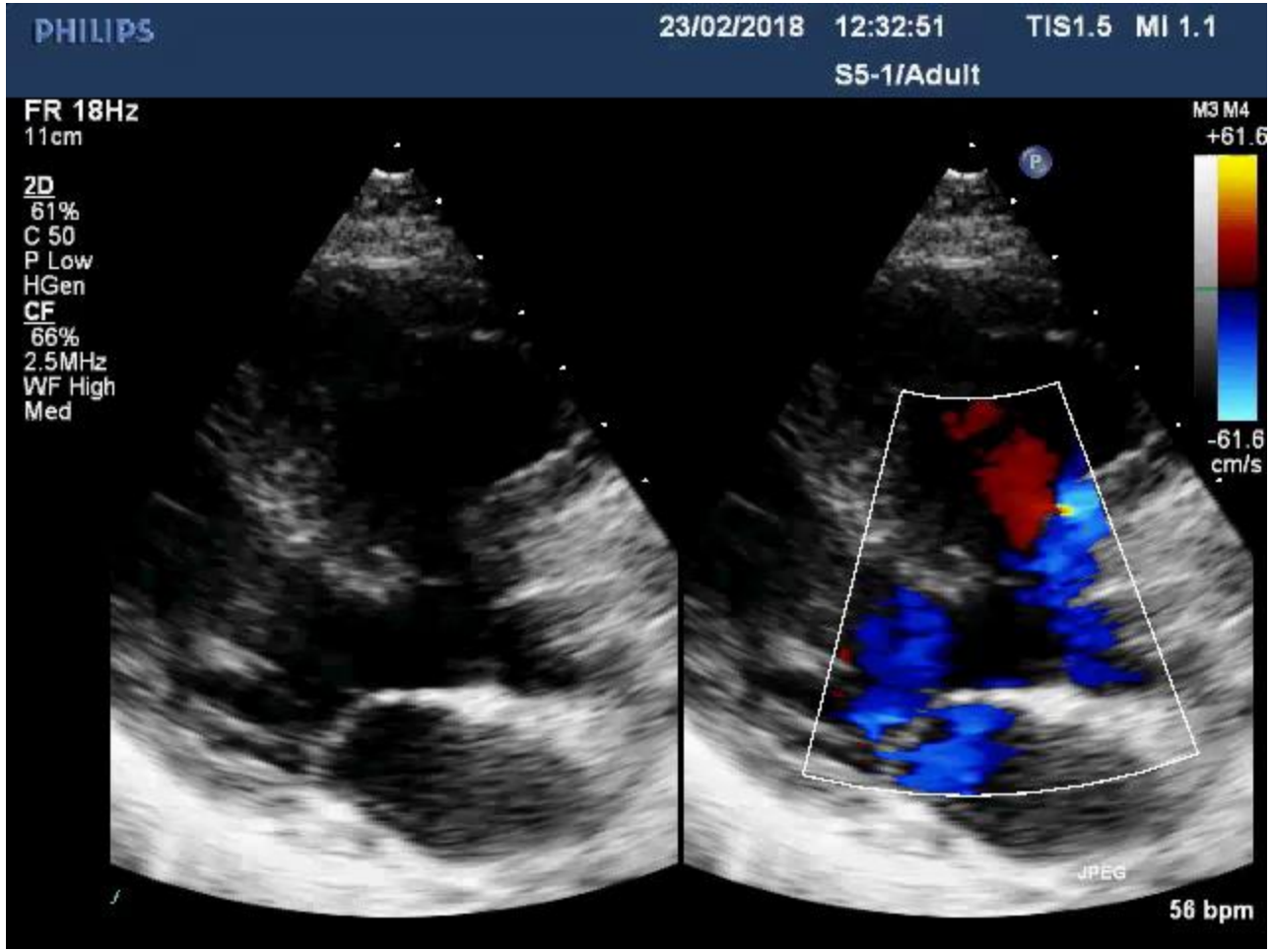
57 bpm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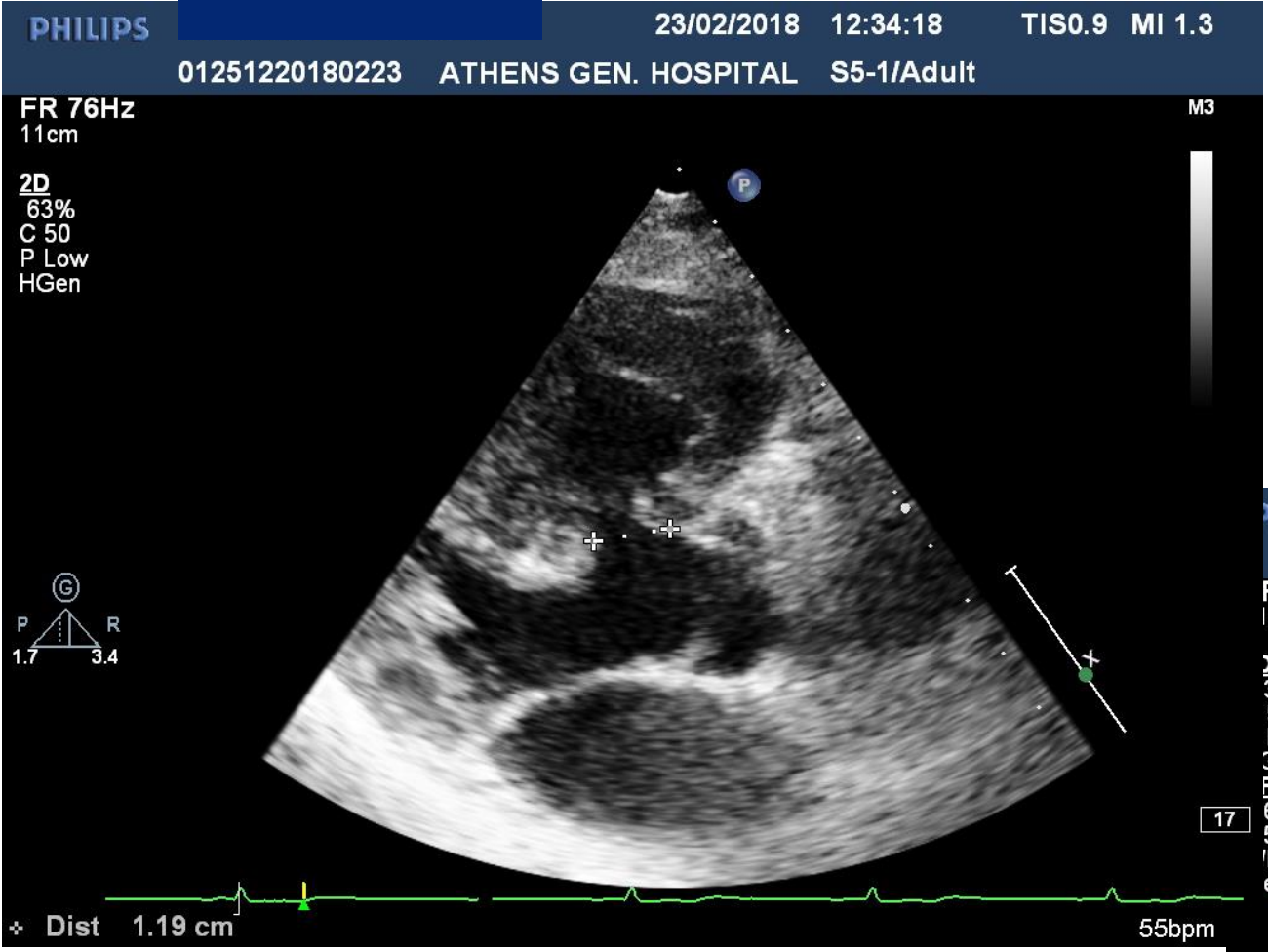
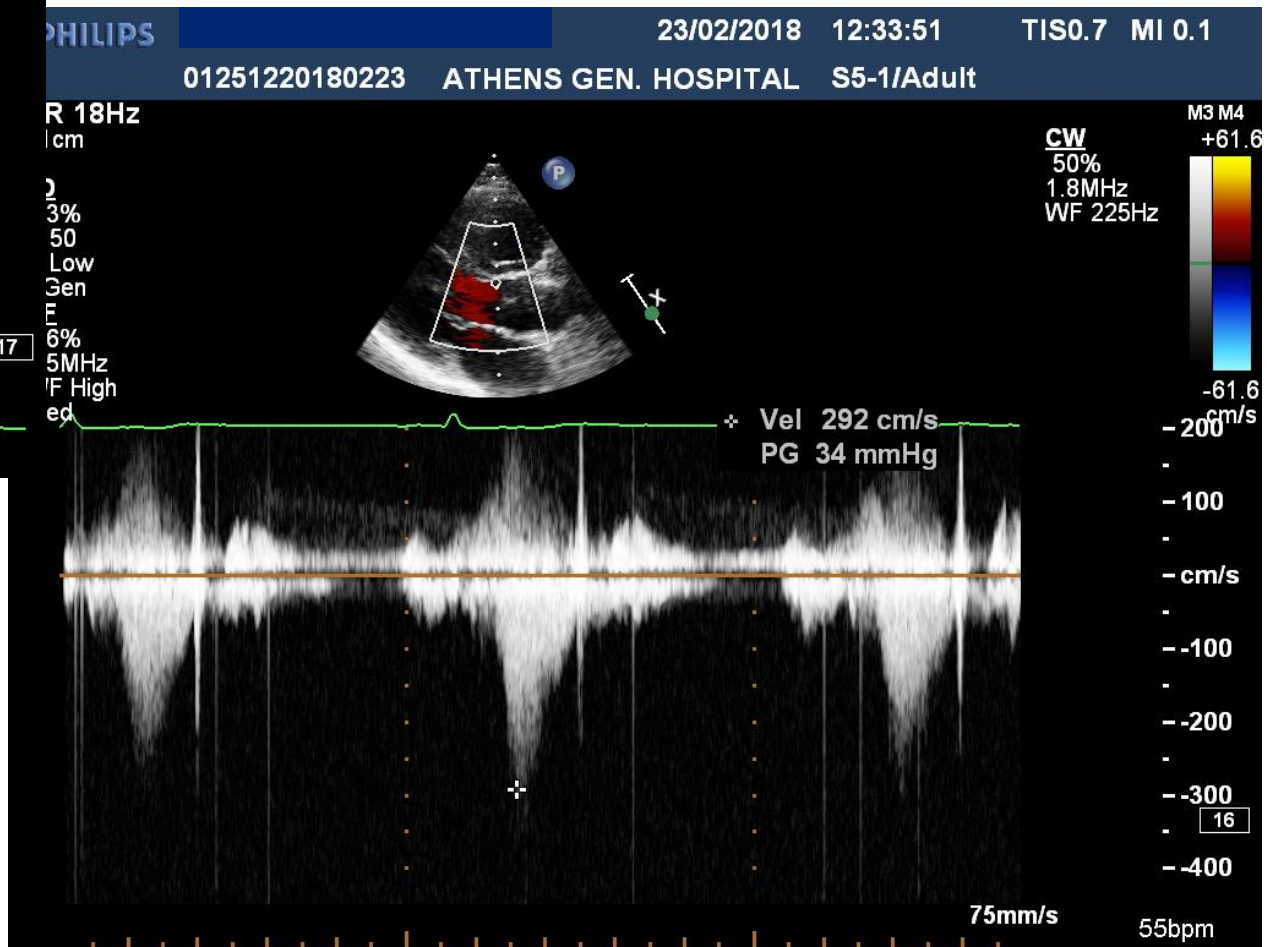
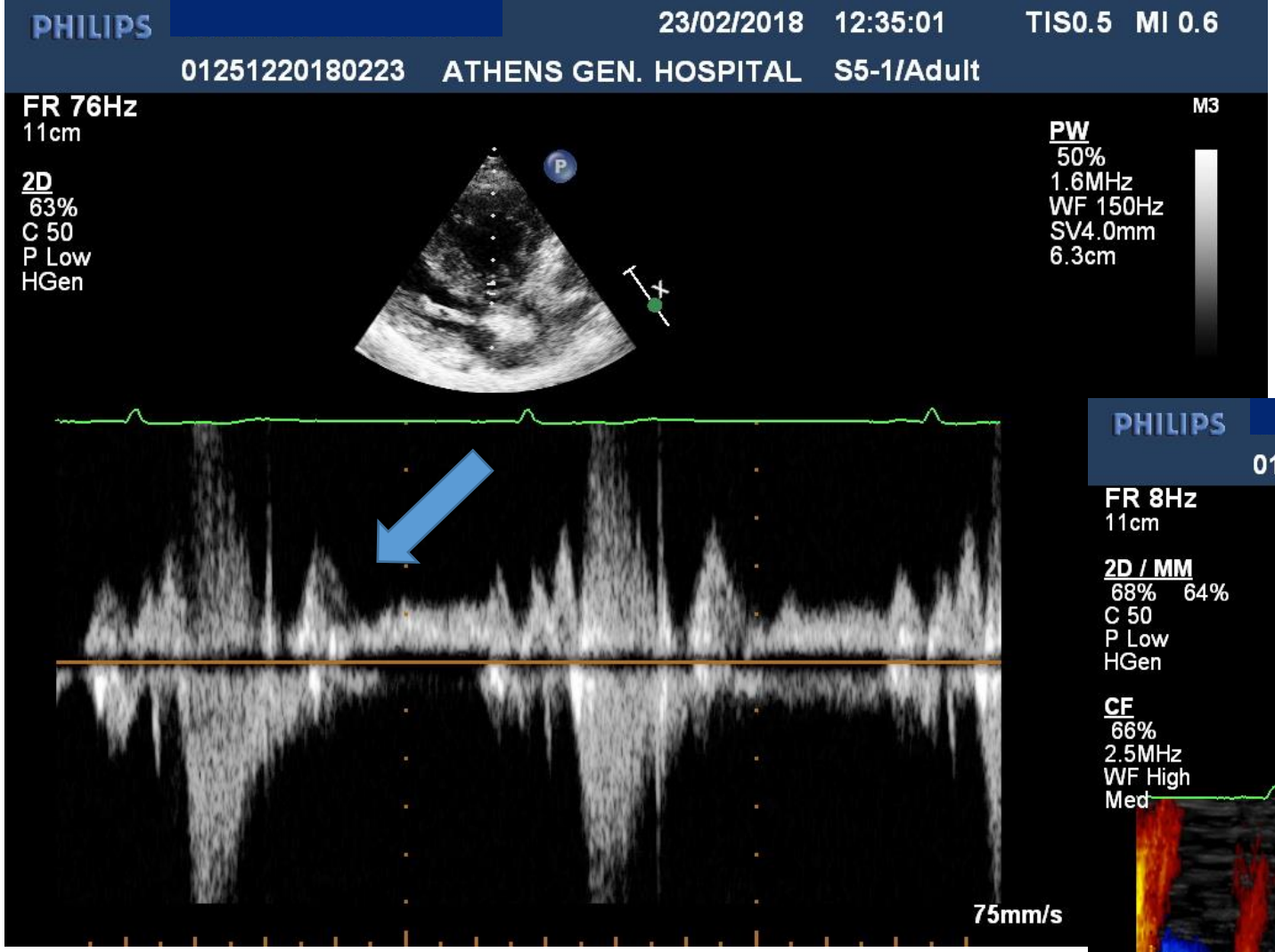
2D
61%
C 50
P Low
HGen
CF
66%
2.5MHz
WF High
Med
M3 M4
+61.6
-61.6
cm/s
JPEG
56 bpm

Perimembranous VSD *d.d. Outlet VSD*
= 1,19cm (PLAX) [1,39cm (mod PSAX)]

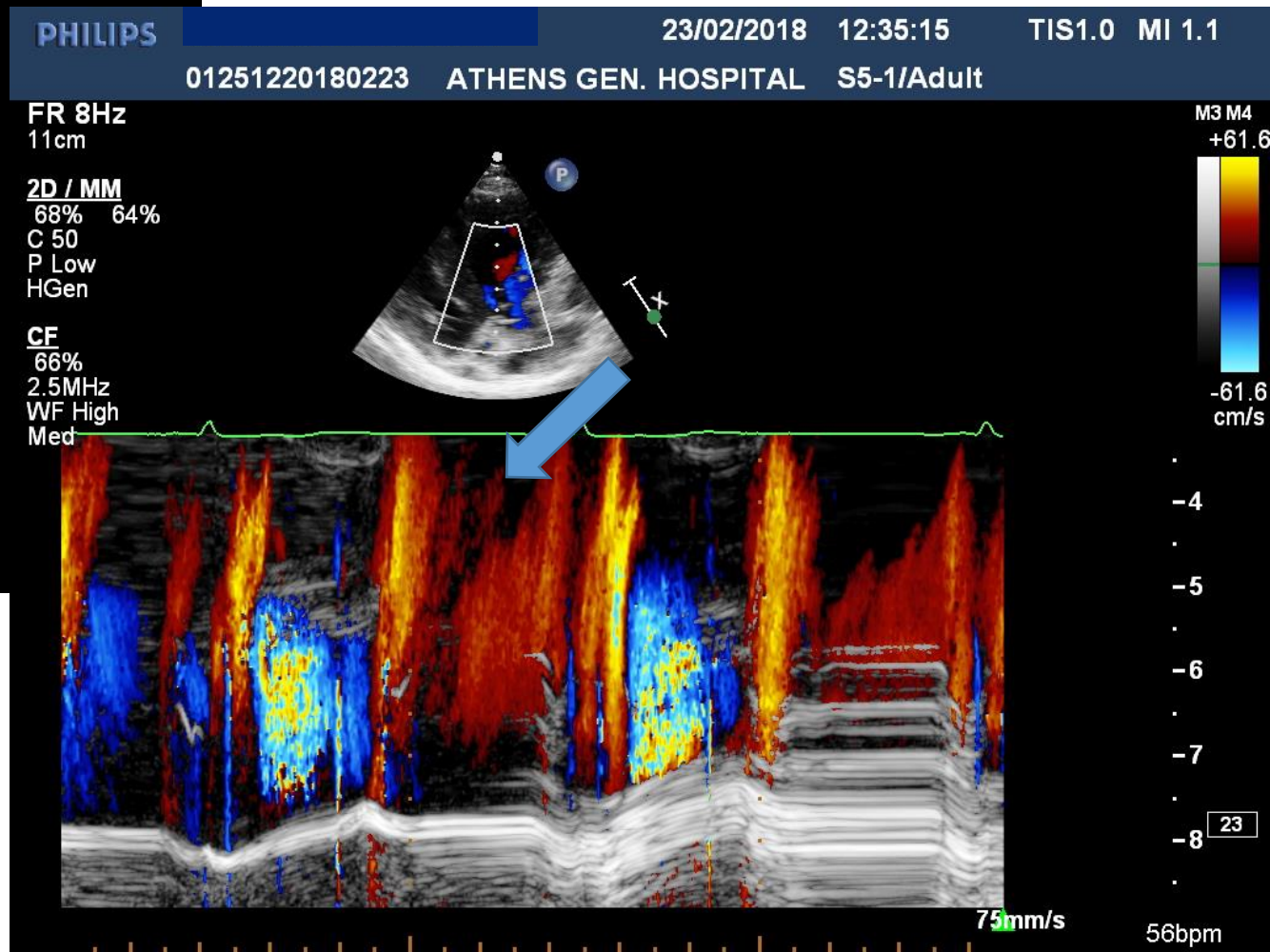
RIGHT- to- LEFT shunt (systole): max PG= 30mmHg
- estimated RVSP= (30+108)= 138mmHg

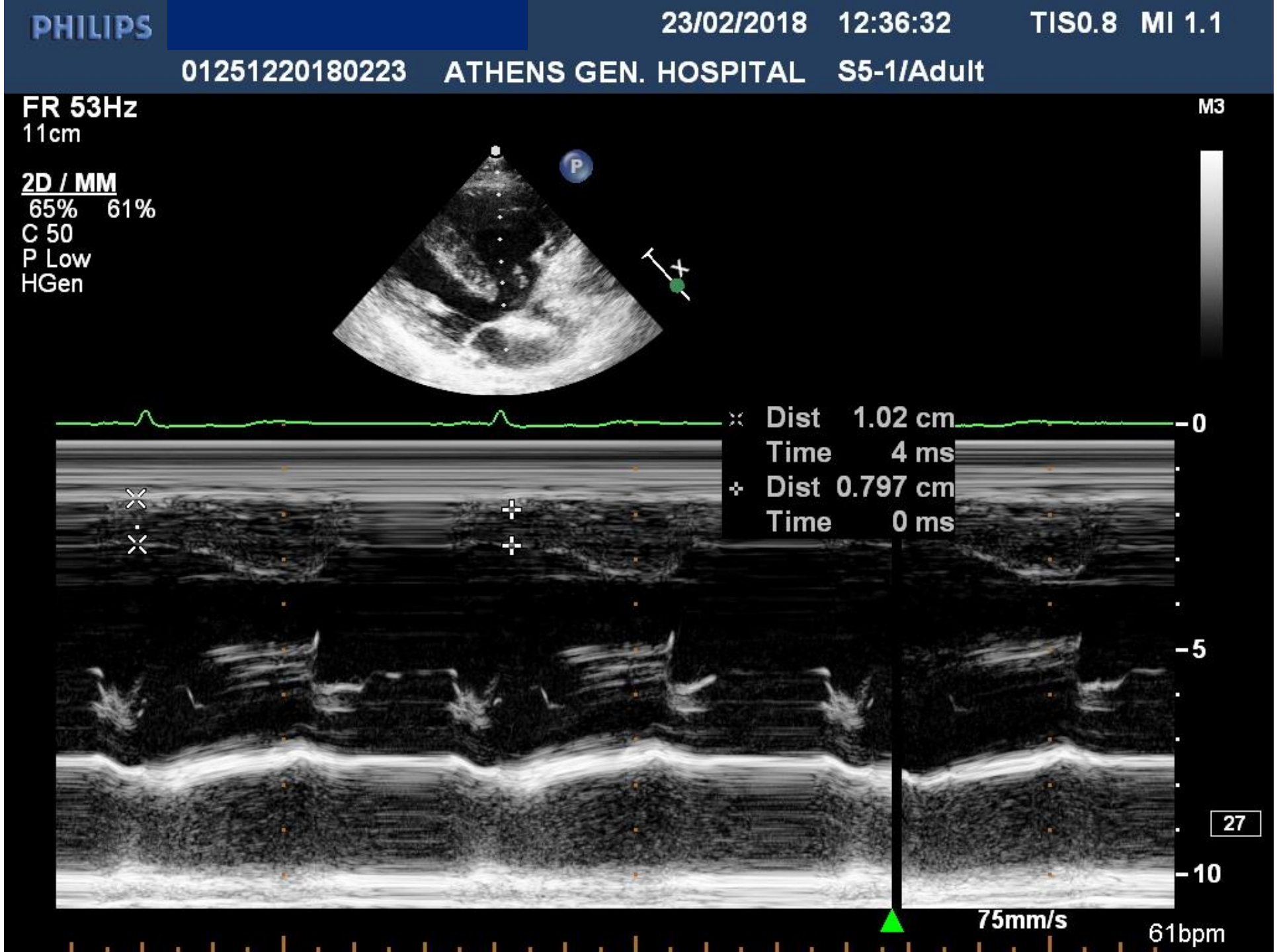






PW-Doppler and colour M-Mode:
Left- to- right shunt (diastole)

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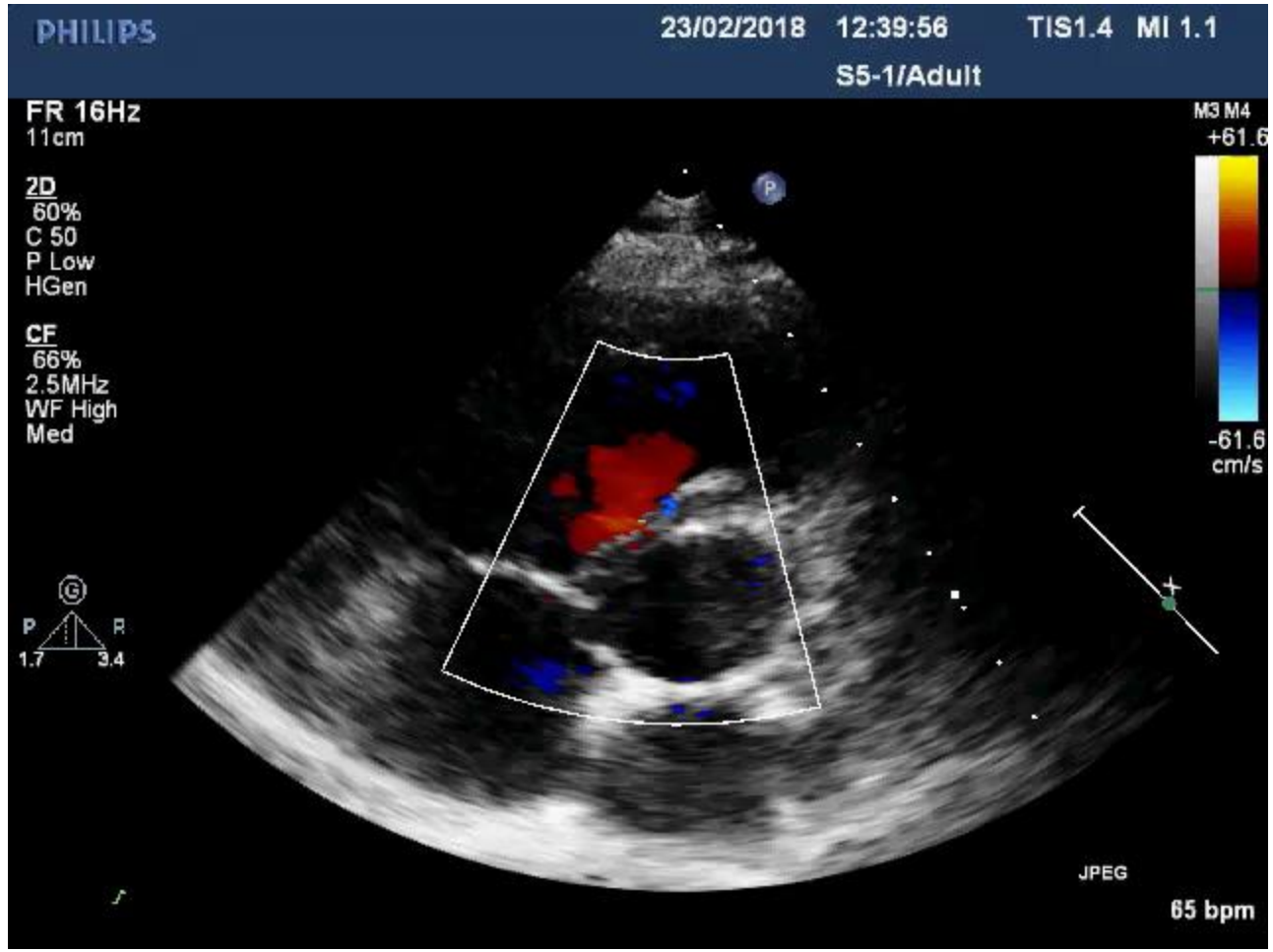
FR 53Hz
11cm
2D / MM
65% 61%
C 50
P Low
HGen
M3
Dist 1.02 cm
Time 4 ms
Dist 0.797 cm
Time 0 ms
0
5
10
27
75mm/s
61bpm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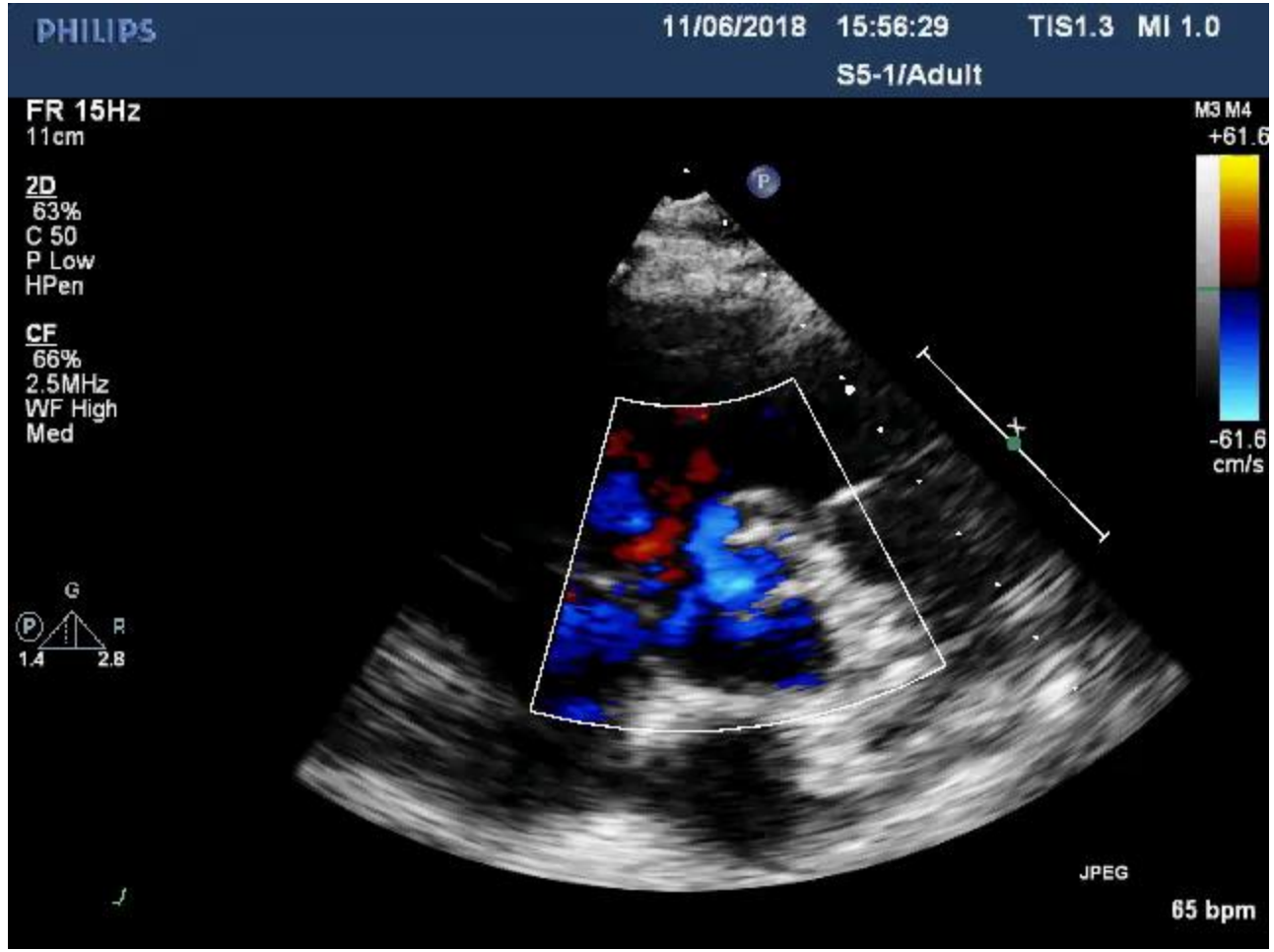
FR 16Hz
11cm
2D
60%
C 50
P Low
HGen
CF
66%
2.5MHz
WF High
Med
M3 M4
+61.6
-61.6
cm/s
P
R
1.7
3.4
JPEG
65 bpm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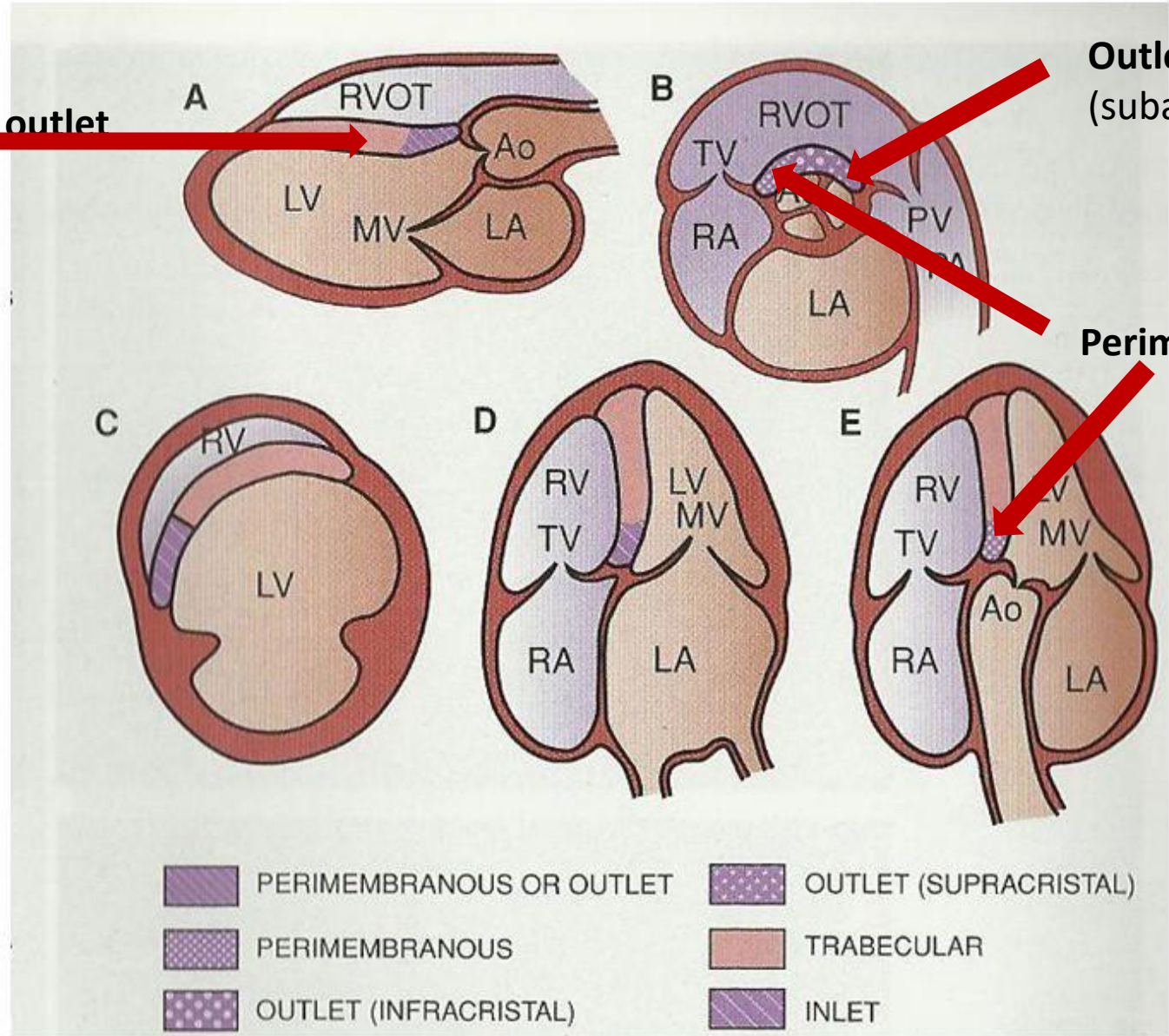
2D
63%
C 50
P Low
HPen
CF
66%
2.5MHz
WF High
Med
M3 M4
+61.6
-61.6
cm/s
G
P
R
1.4
2.8
JPEG
65 bpm

Common locations of vsd -2d echo

Perimembranous or outlet



Outlet (infra- , supracristal)
(subaortic, doubly-committed)

Perimembranous


PHILIPS
23/02/2018 12:38:13 TIS1.5 MI 1.1
S5-1/Adult
FR 22Hz
11cm
2D
61%
C 50
P Low
HGen
CF
66%
2.5MHz
WF High
Med
M3 M4
+61.6
-61.6
cm/s
P
R
1.7
3.4
JPEG
54 bpm


PHILIPS
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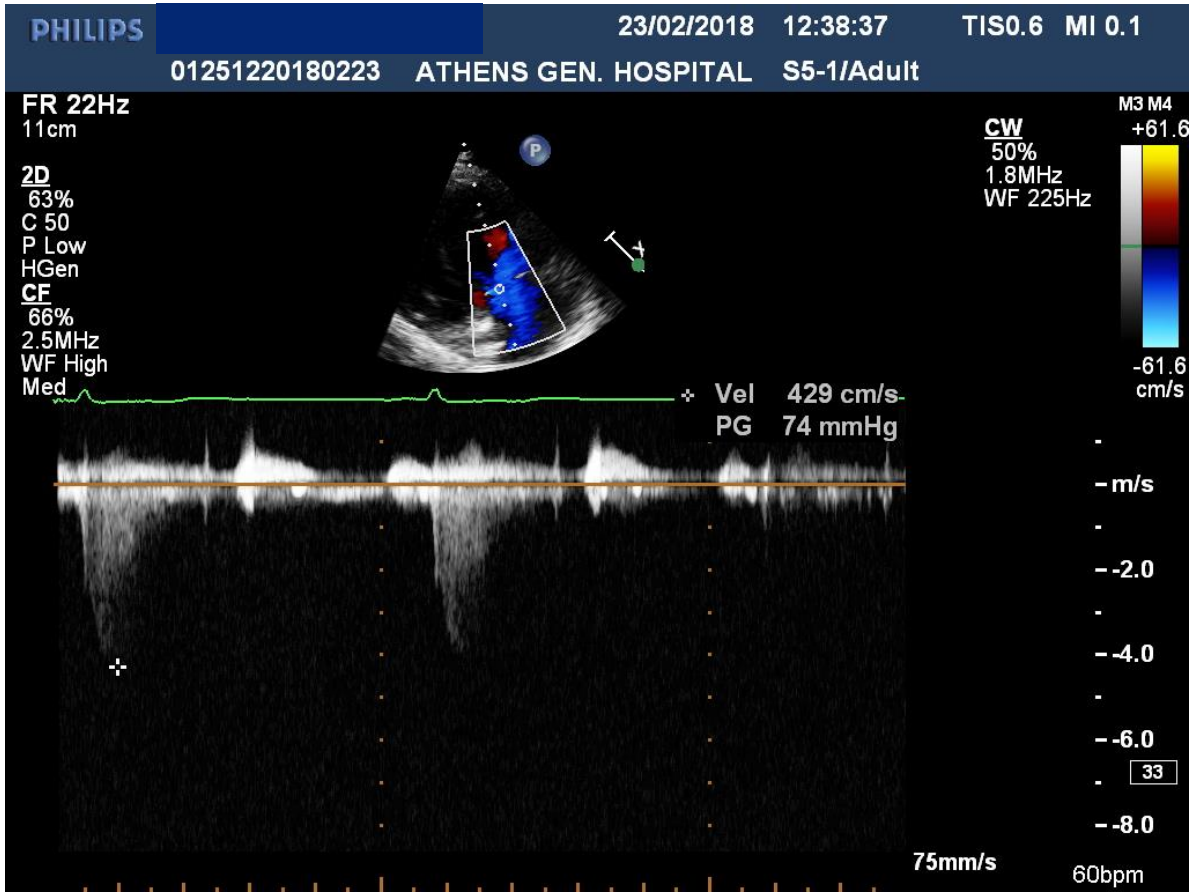
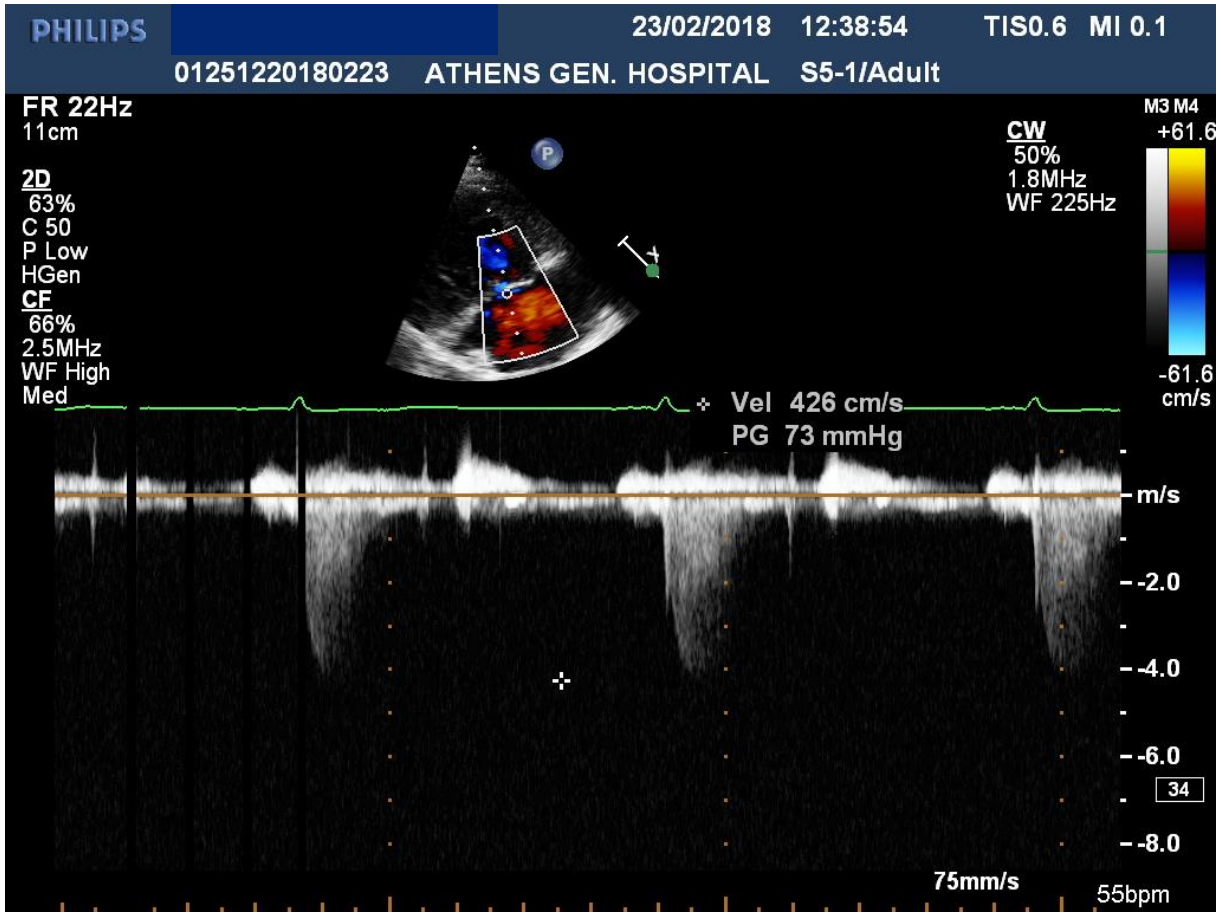
S5-1/Adult
FR 22Hz
11cm
2D
61%
C 50
P Low
HGen
CF
66%
2.5MHz
WF High
Med
M3 M4
+61.6
-61.6
cm/s
P
R
1.7
3.4
JPEG
55 bpm





- TR max Vel= 4,29cm/s (?)
- Estimated RVSP= 74mmHg (?)+ RAP

PASP, mean PAP evaluation (applying TR max Vel)

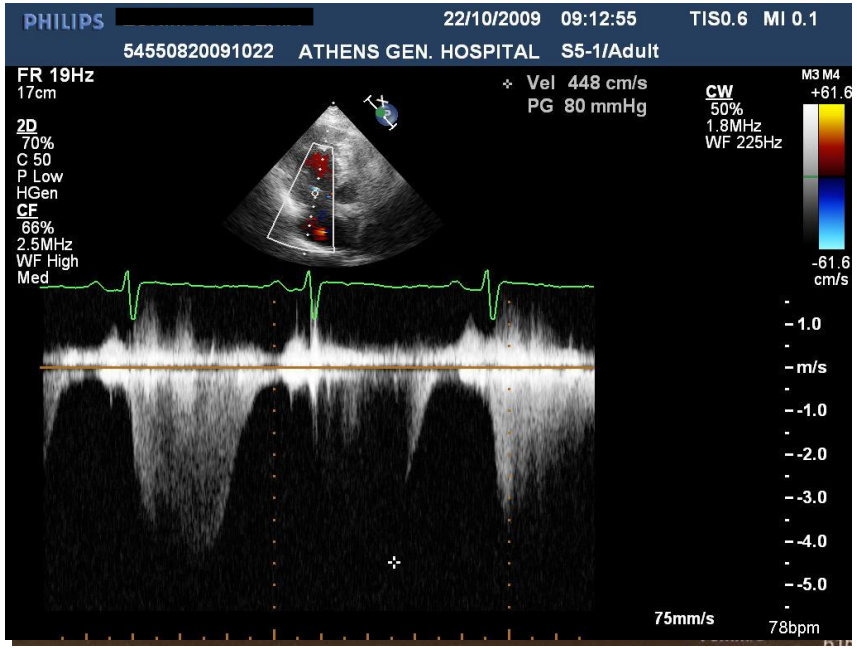
- **RVSP = $4V^2$+RAP**

RVSP = PASP (provided no RVOT obstruction)

- **Mean PAP= 0,61 X PASP +2mmHg**

- **MPAP = mean RV-RA syst gradient + RAP**

(Aduen)



- 10-25% unsatisfactory TR doppler ⟵ agitated saline contrast
- ↓TRV av ⇑ RAP (RV infarct, RV failure, severe TR)

- Severe TR ⟹ Underestimation of PASP with simplified Bernoulli equation
- Overestimations by >10mmHg for PASP are common


PHILIPS
23/02/2018 12:42:56 TIS1.4 MI 1.1
S5-1/Adult
FR 17Hz
12cm
2D
60%
C 50
P Low
HGen
CF
66%
2.5MHz
WF High
Med
M3 M4
+61.6
-61.6
cm/s
P
R
1.7
3.4
JPEG
55 bpm


PHILIPS
11/06/2018 15:54:14 TIS1.3 MI 0.8
S5-1/Adult
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11cm
2D
71%
C 50
P Low
HPen
CF
66%
2.5MHz
WF High
Med
M3 M4
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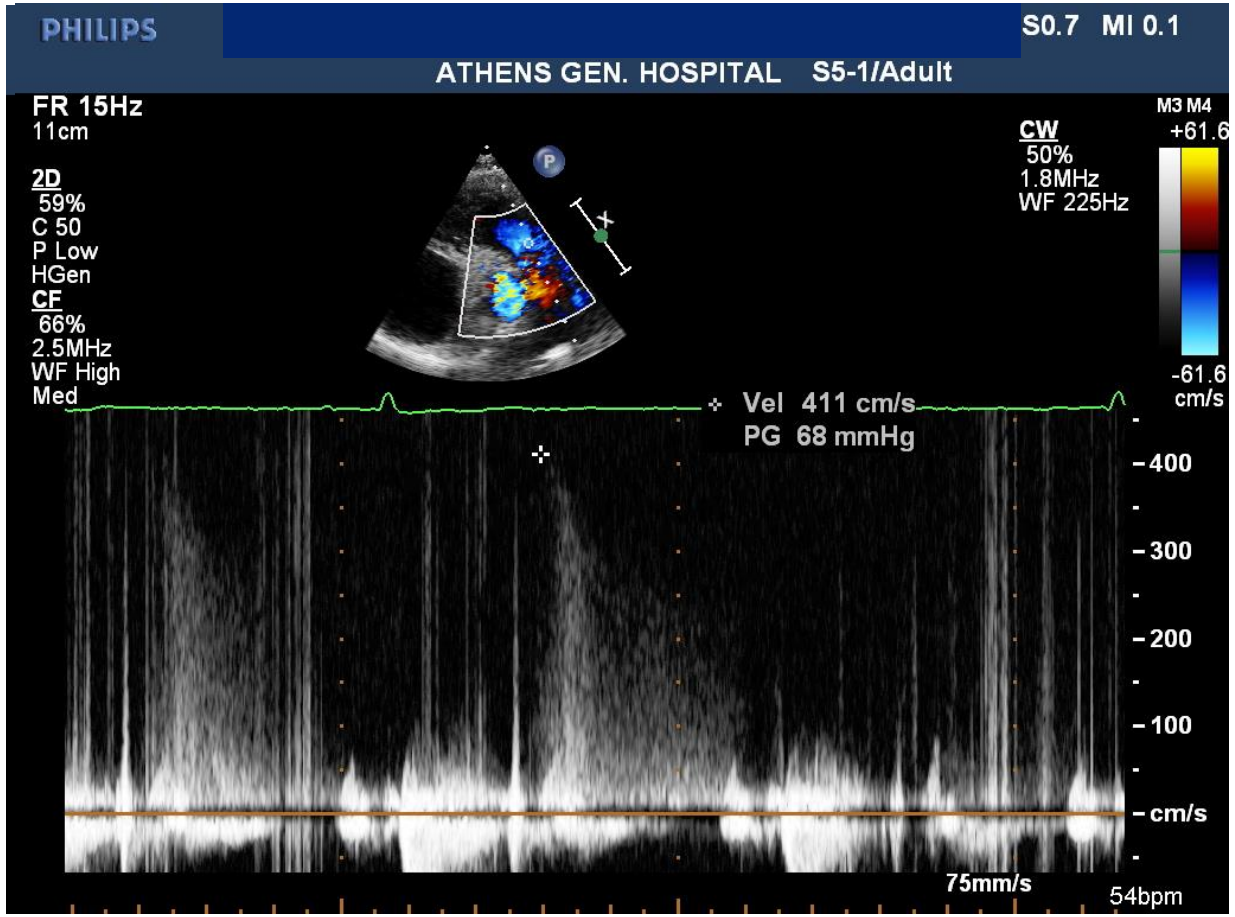
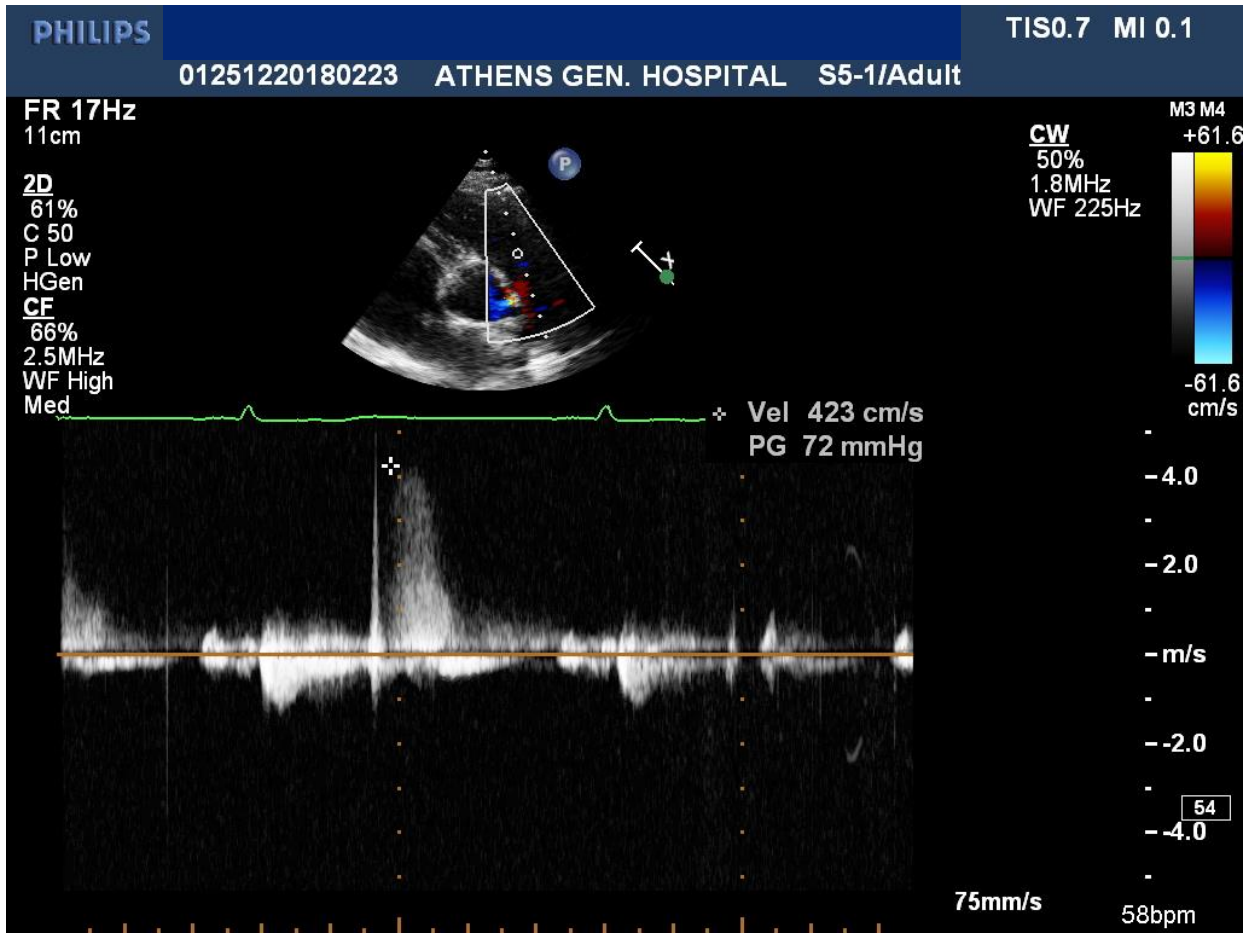
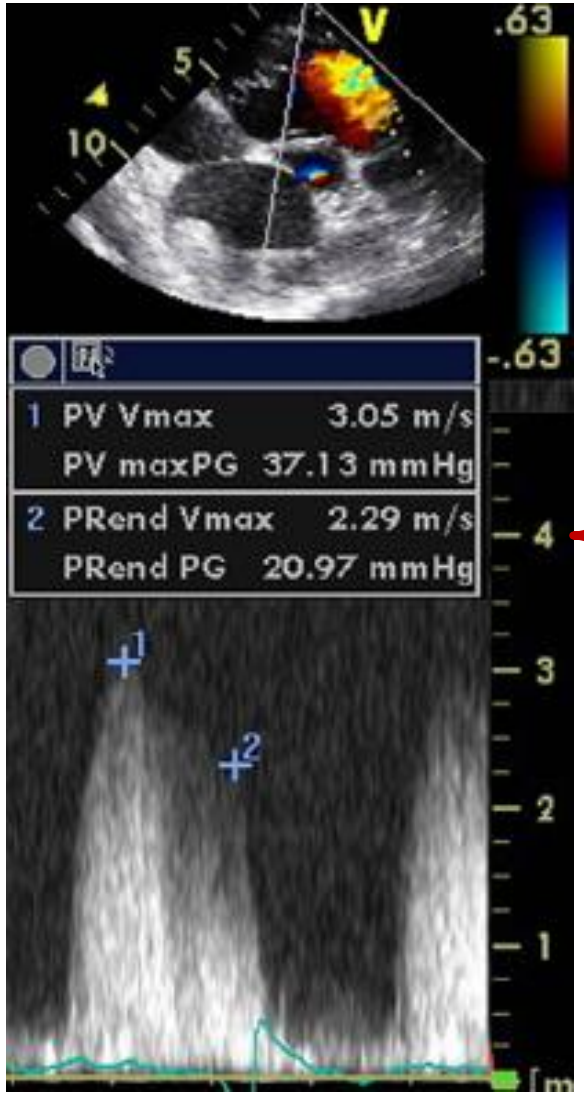
cm/s
G
P
R
1.4
2.8
JPEG
60 bpm



- PR peak Vel= 4,2m/s
- Mean PAP= (70mmHg+ RAP)



Hemodynamic assessment of pulmonary circulation



PAEDP = 4x PREDVel² + RAP

(EDPG> 5mmHg = syst or diast RV dysfunction,

↑ BNP,↓ FC)

Ristow B JASE 2005

MPAP = 4 X peak PRVel² *(Masuyama)*

MPAP = 4 X peak PRVel²+ RAP *(Abbas)*

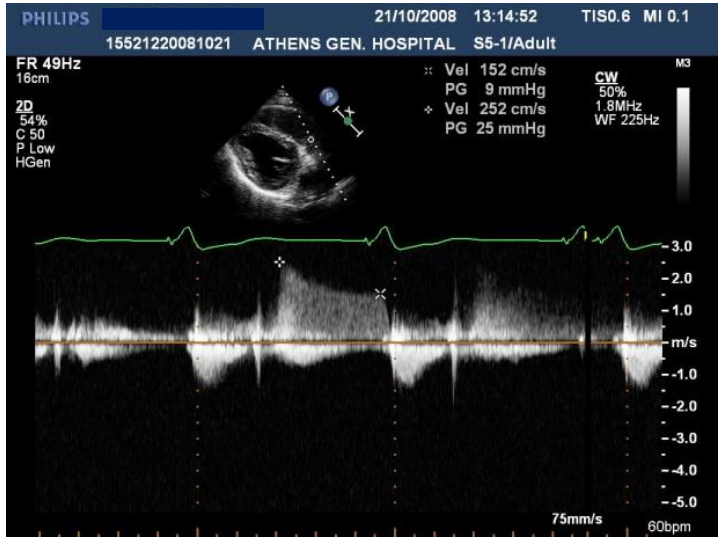
(MPAP = PAEDP + 1/3 (PASP-PAEDP))

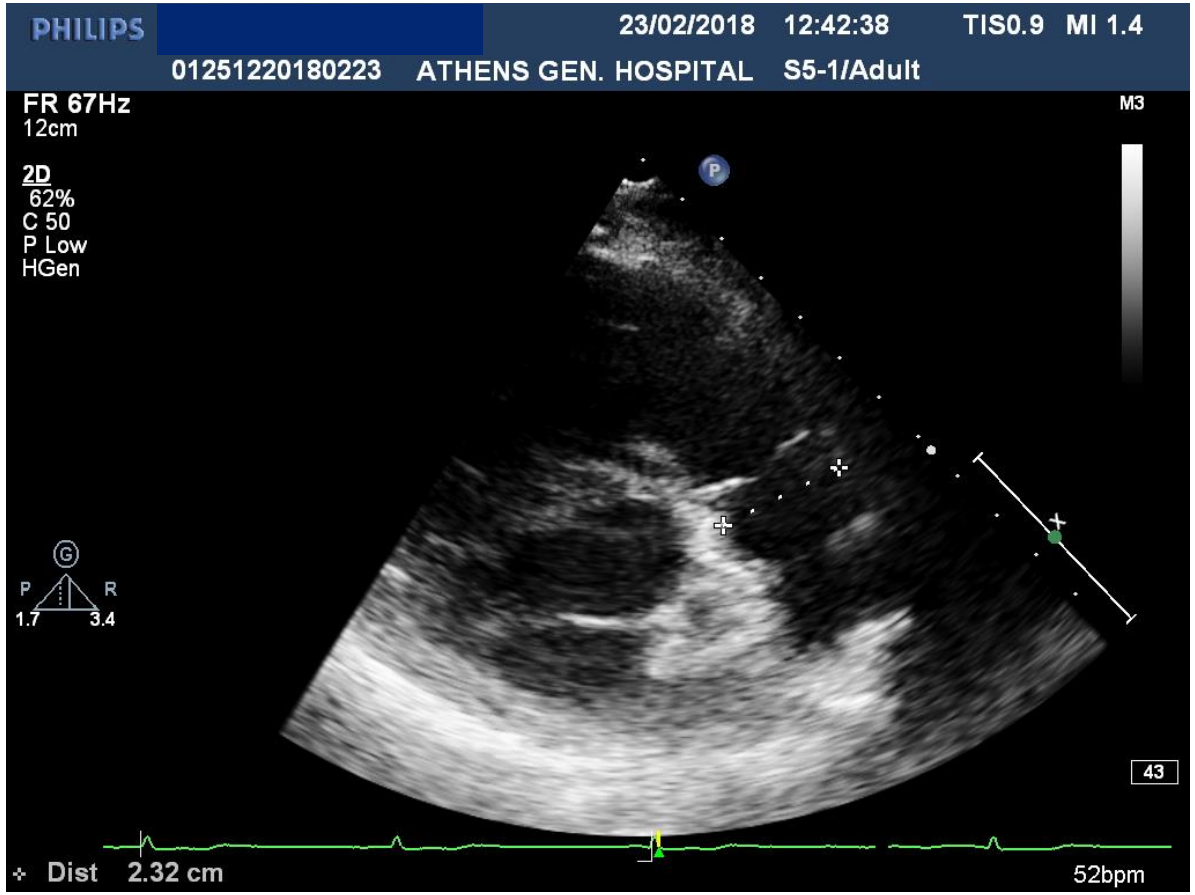
Masuyama T Circulation 1986

Abbas AE Am J Cardiol 2003

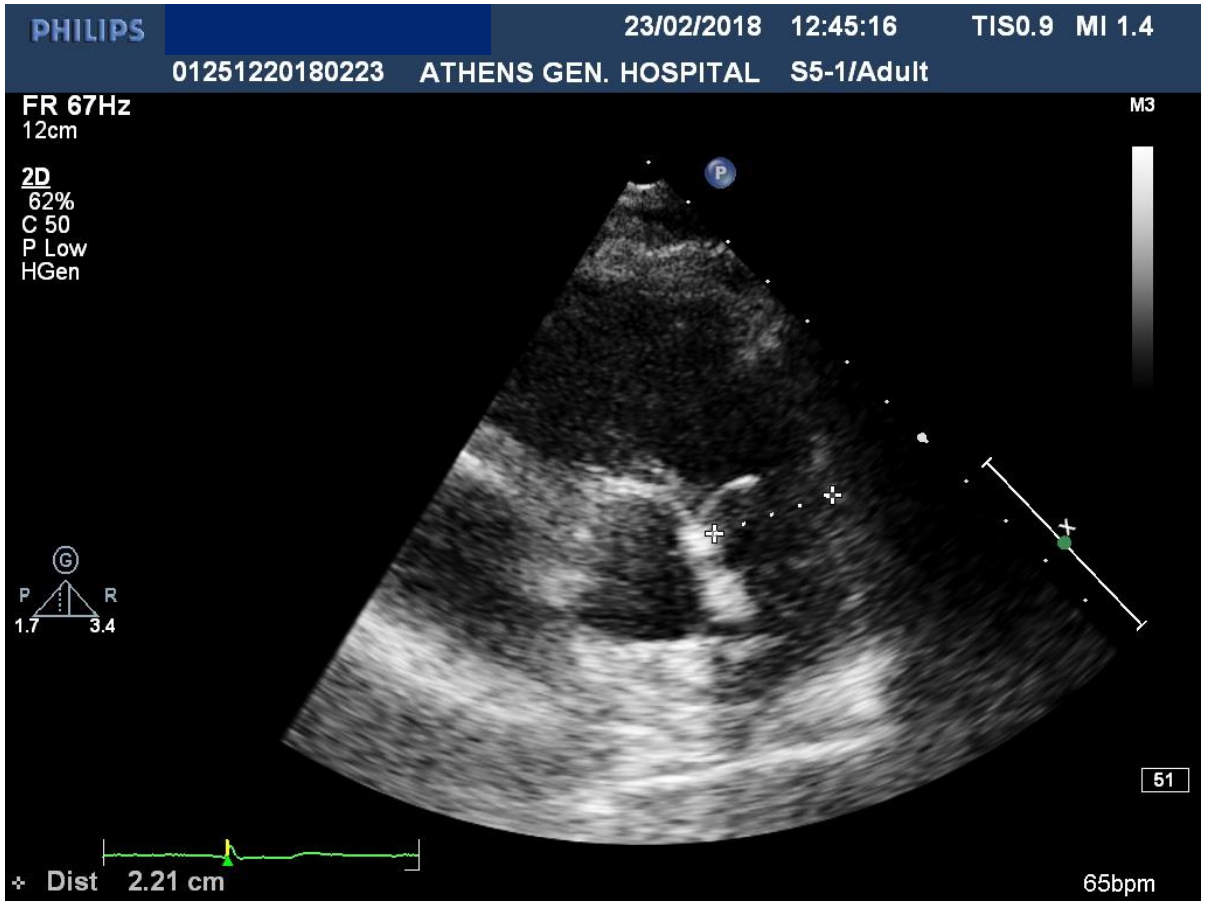
Aduen JF JASE 2009

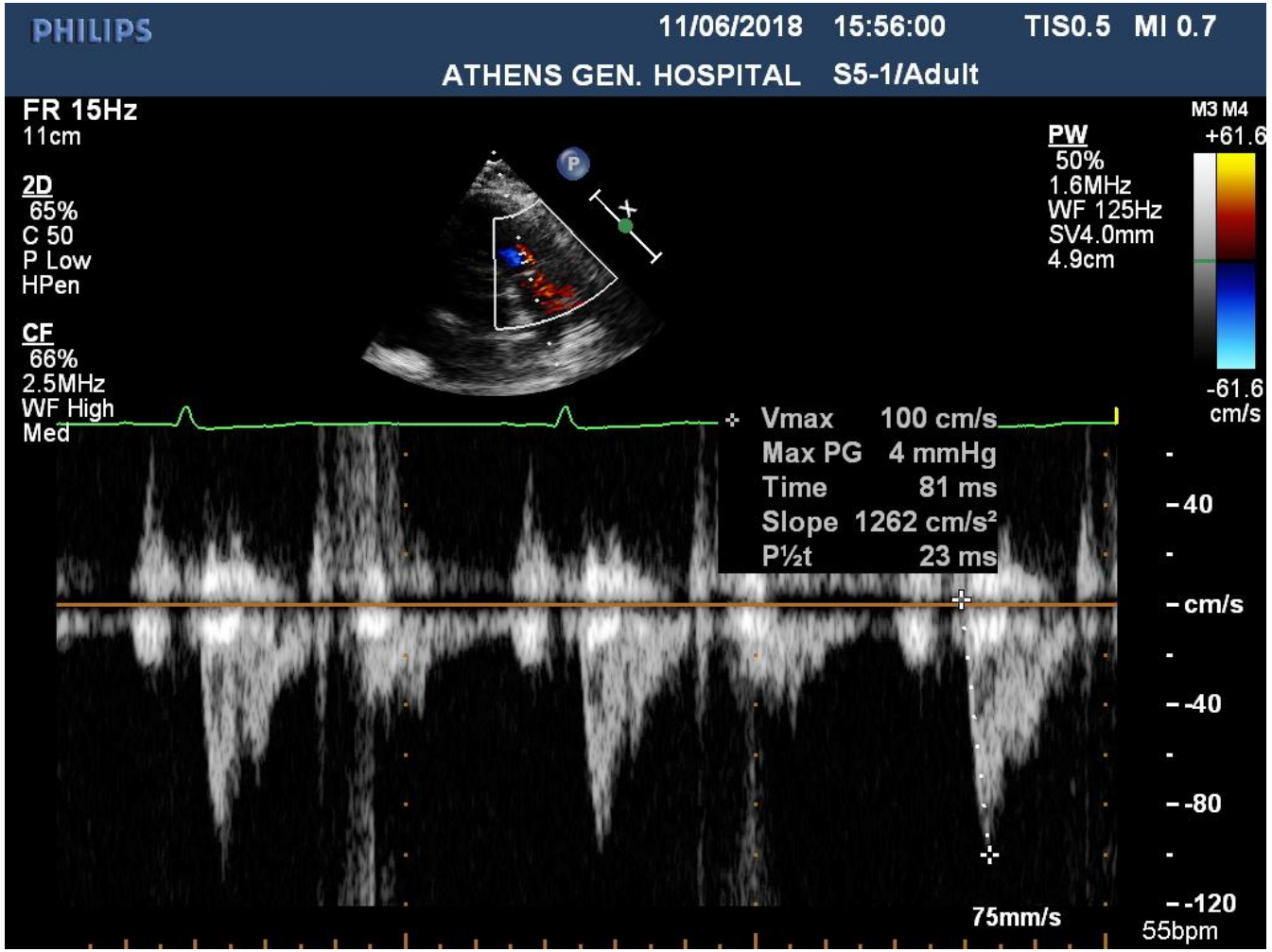
Rudski LG JASE 2010





PA diameter: 23 mm



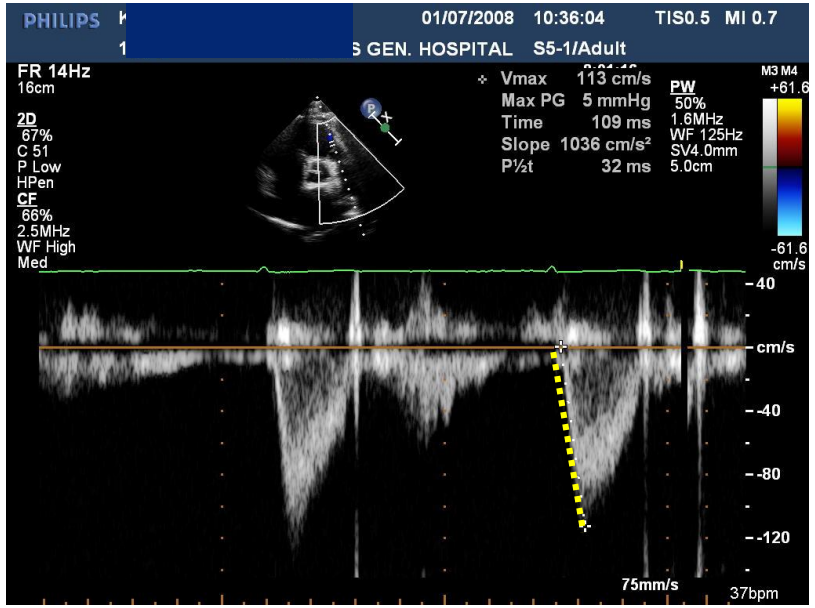


RVOT PW-Doppler:

AcT= 81ms

Midsystolic notch

Hemodynamic assessment of pulmonary circulation



RVOT PW Doppler - AcT

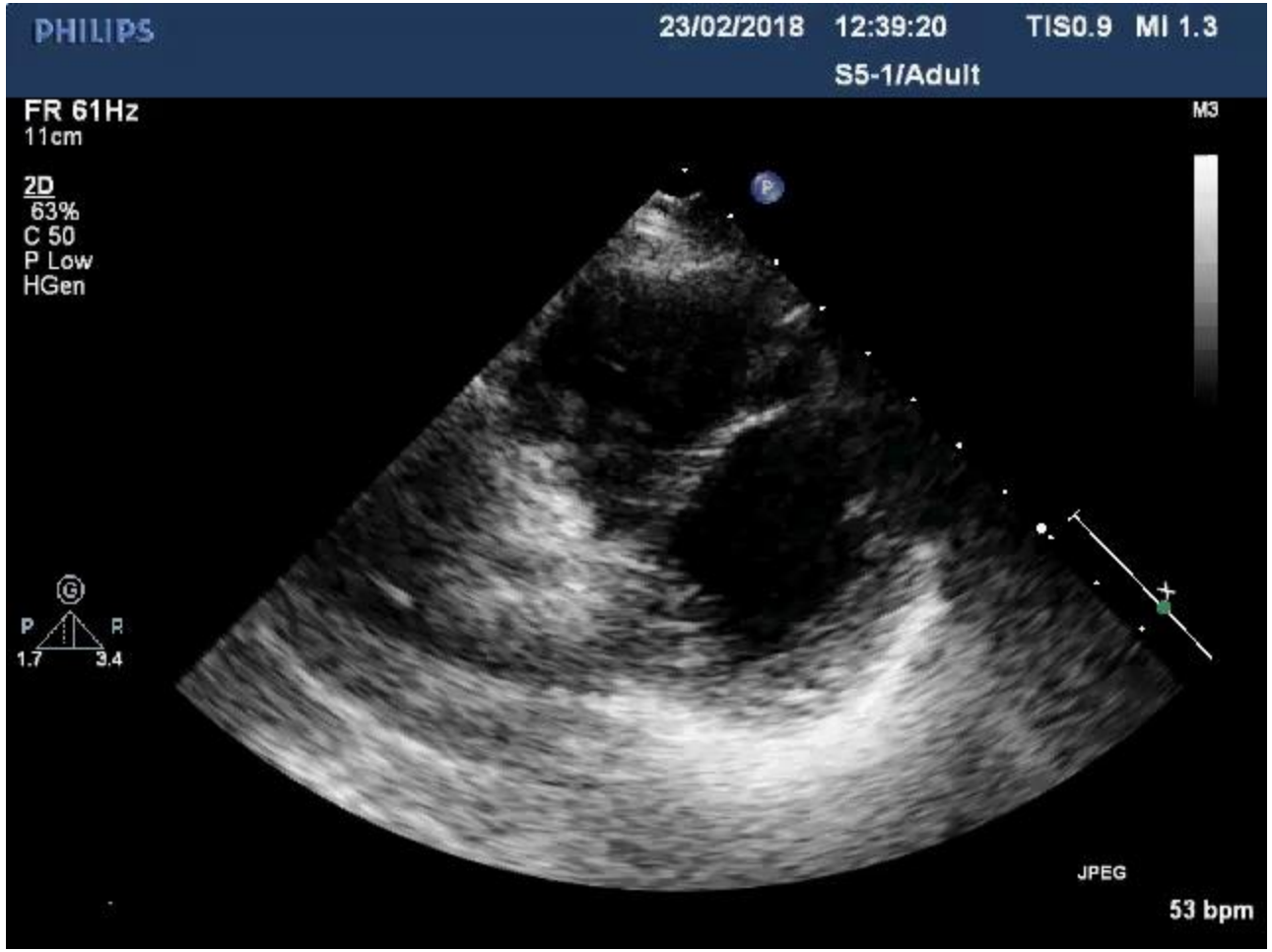
MPAP = 79 – (0.45 X AcT) *(Mahan's equation)*

- *AcT depends CO, HR (< 60bpm ή > 100bpm)*

- ***MPAP = 90 – (0.62 X AcT) when AcT <120ms***

Mahan G Circulation 1983

Dabestani A Am J Cardiol 1987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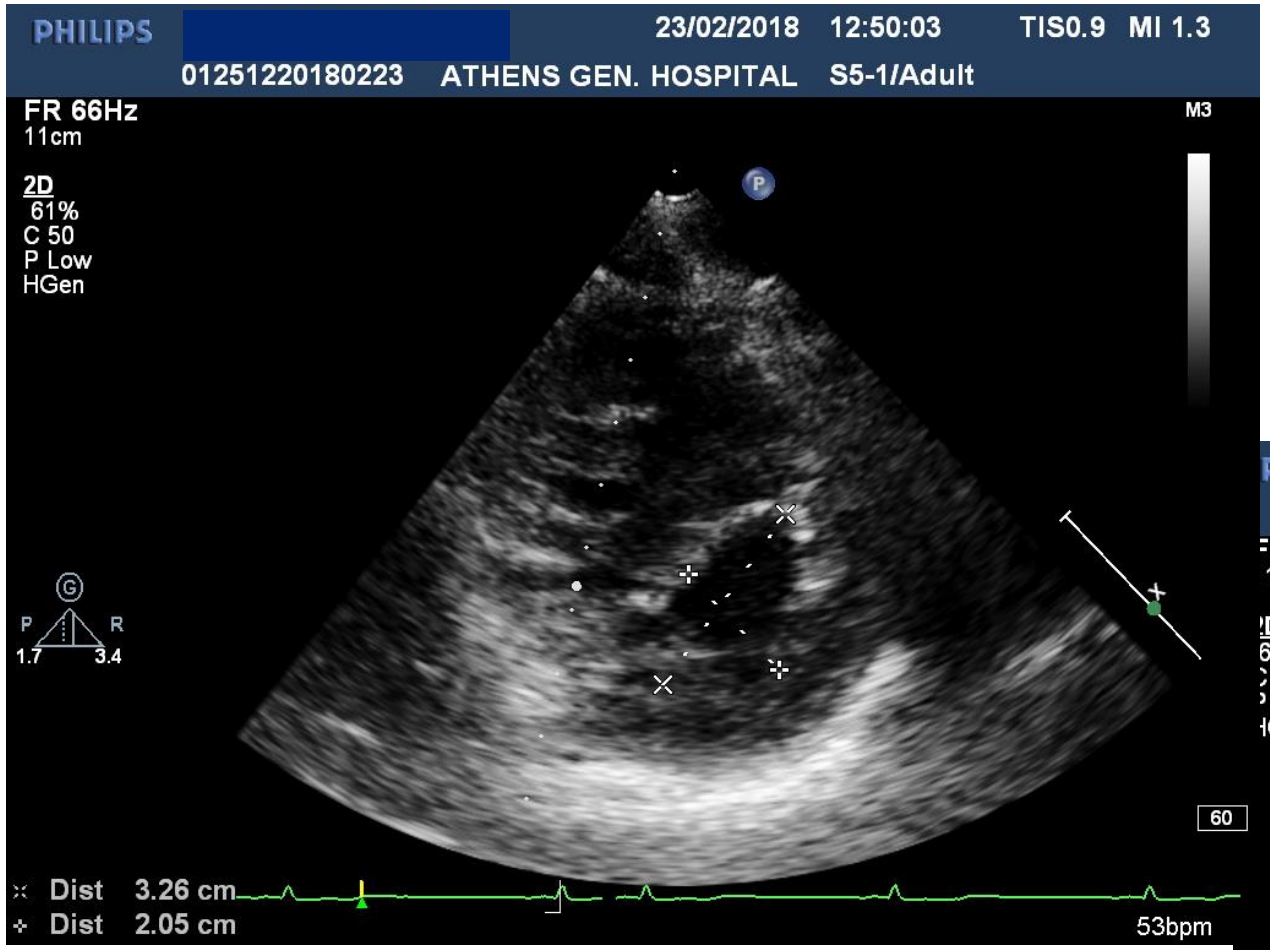
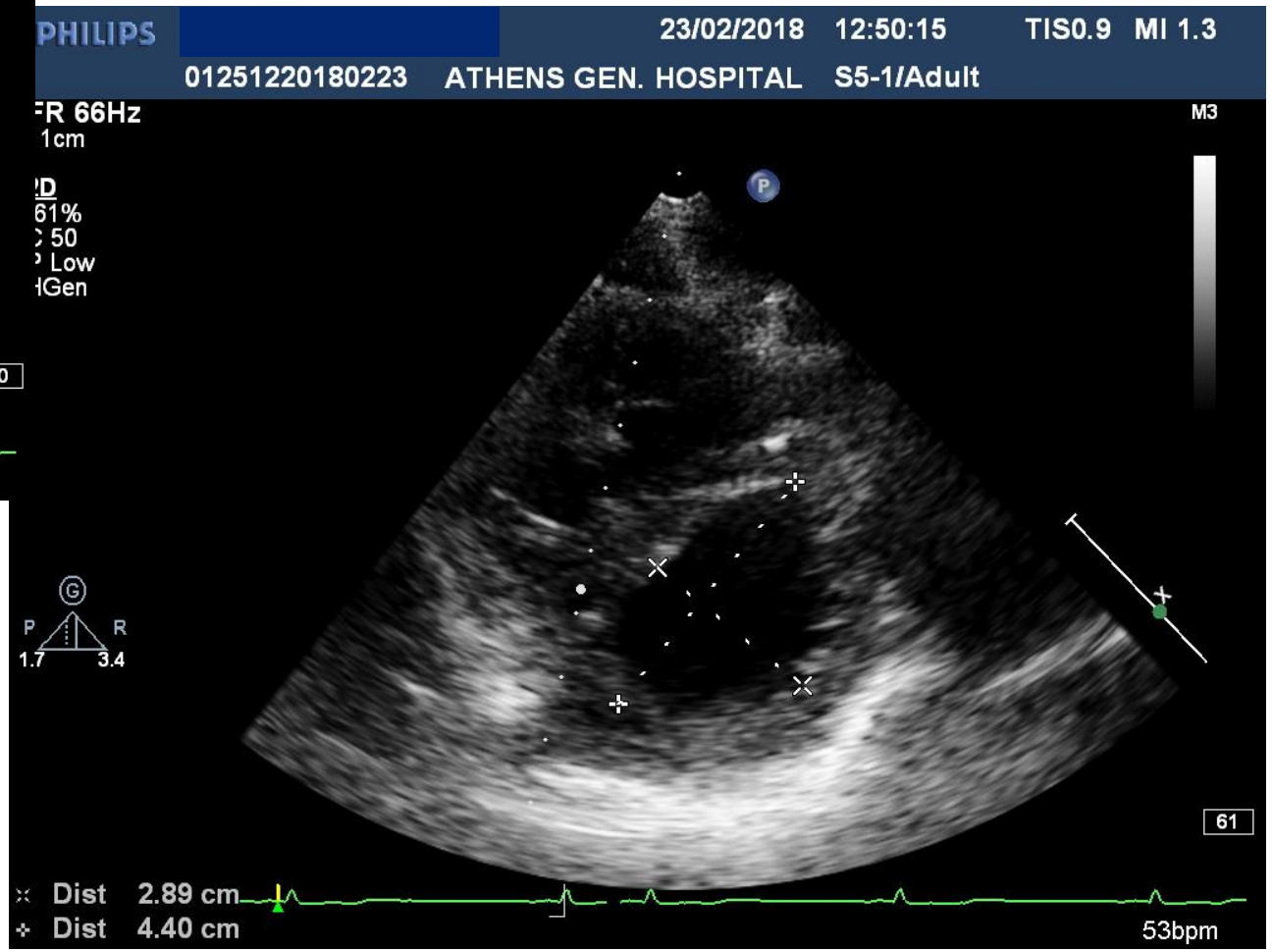
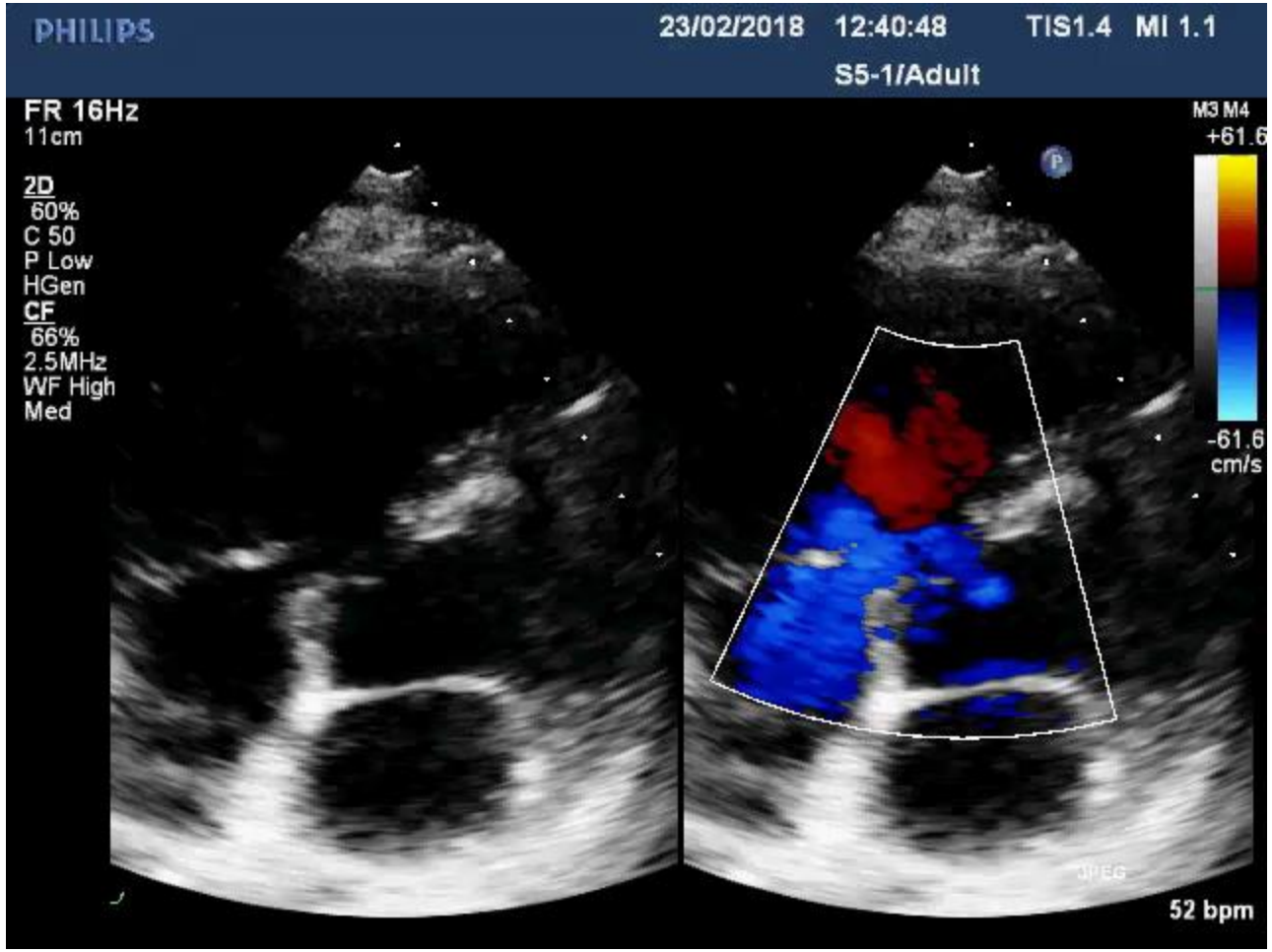
2D
63%
C 50
P Low
HGen
M3
P
R
1.7
3.4
JPEG
53 bpm



Systolic eccentricity index= 1,59

Diastolic eccentricity index= 1,59

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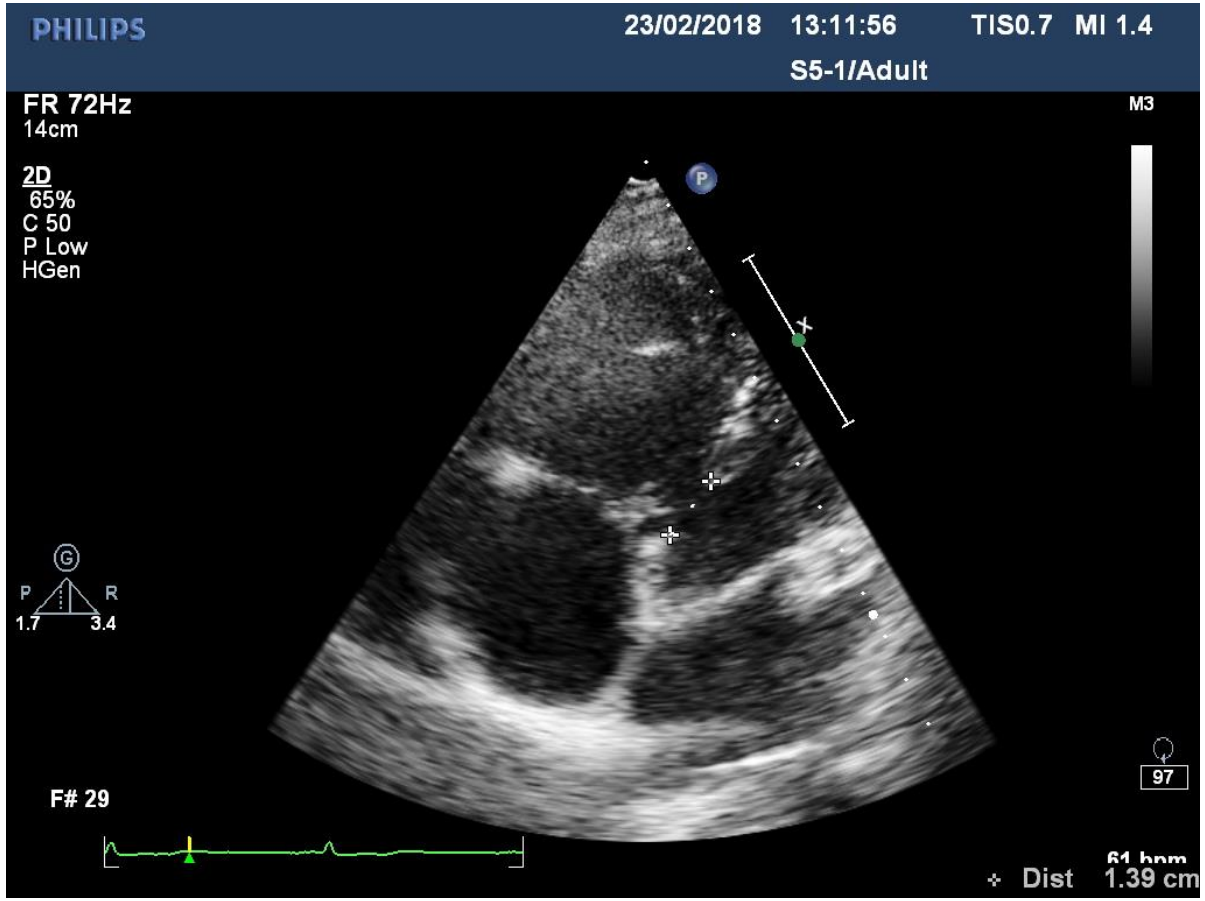
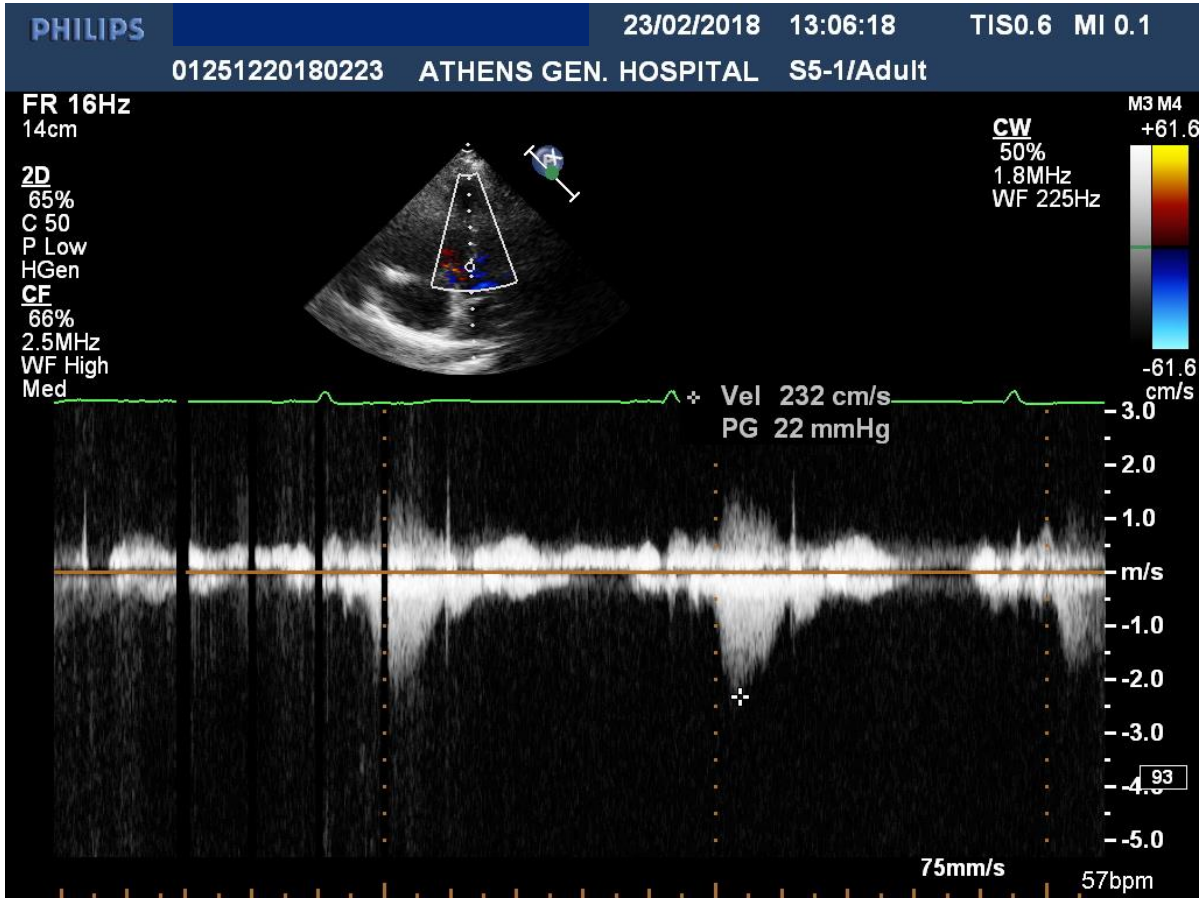
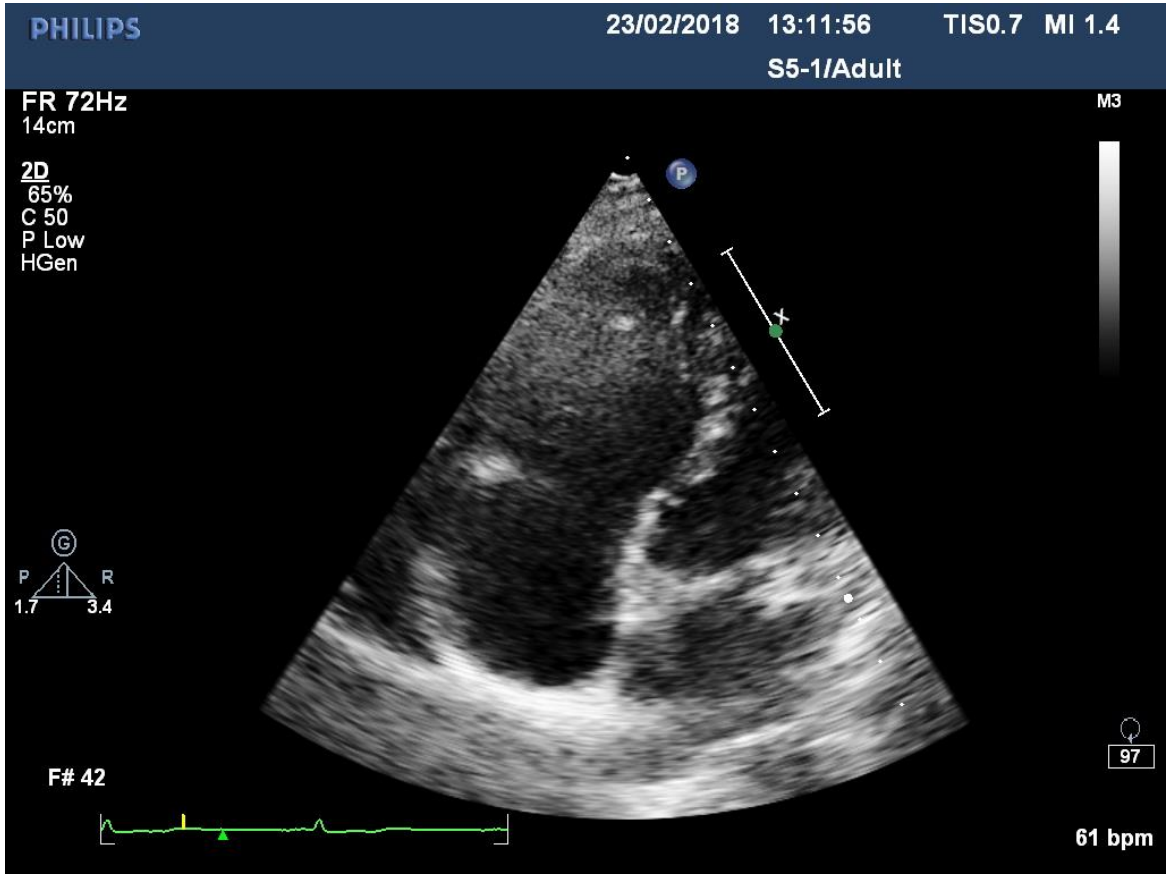
2D
60%
C 50
P Low
HGen
CF
66%
2.5MHz
WF High
Med
M3 M4
+61.6
-61.6
cm/s
52 bpm

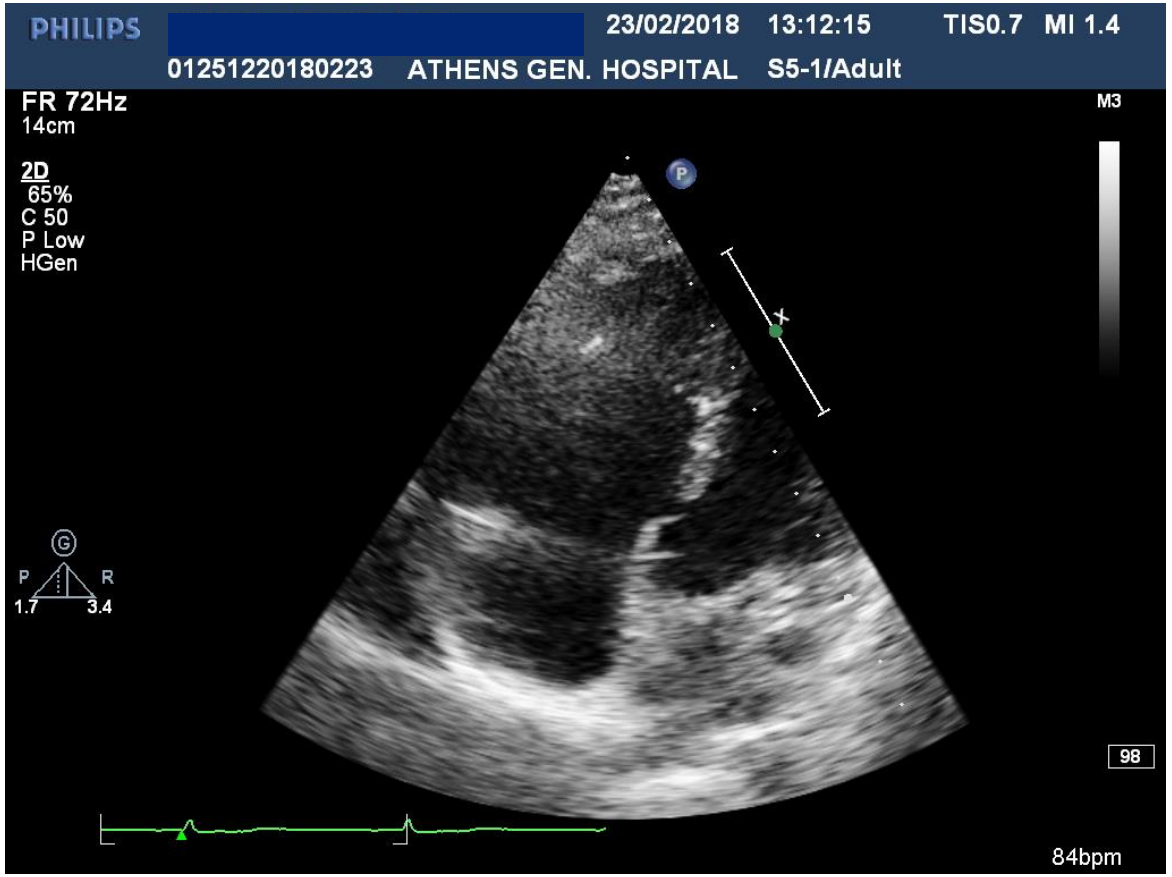


Perimembranous VSD = 1.39cm (mod view)

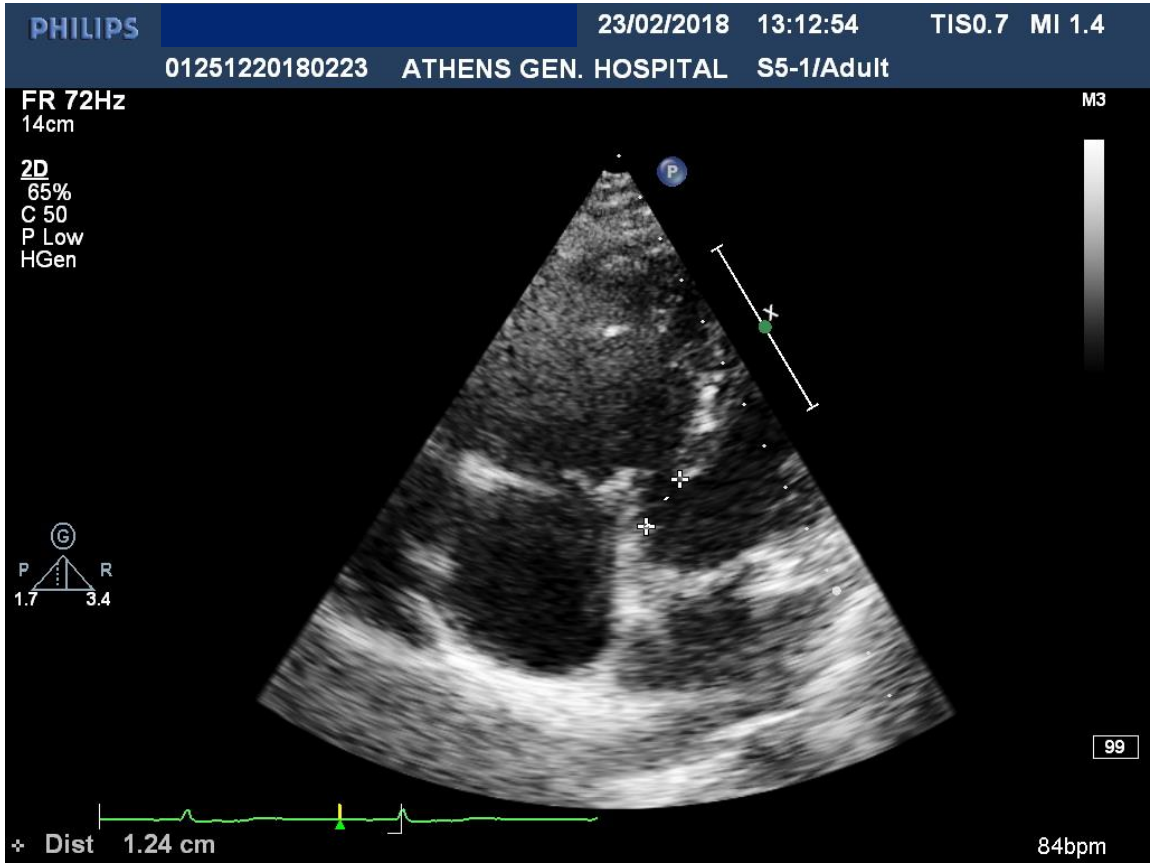


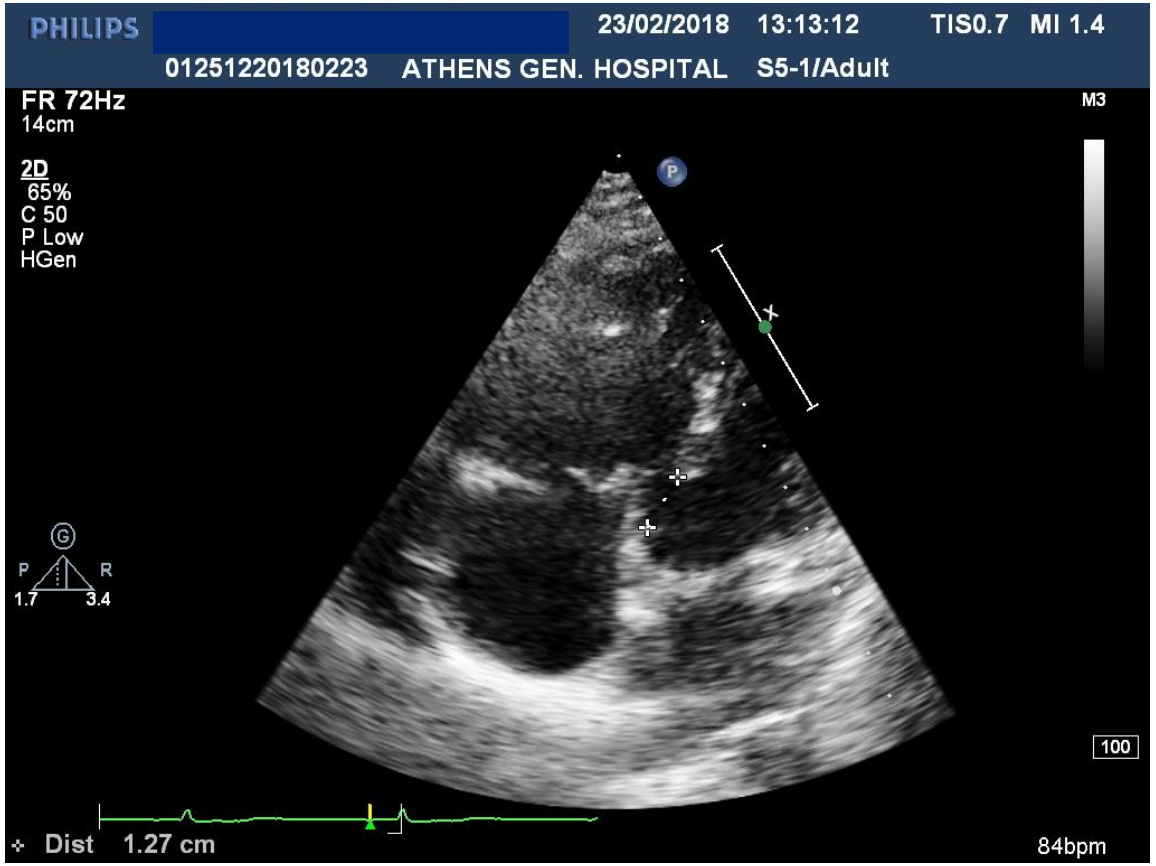
RIGHT- to- LEFT shunt (systole): max PG= 22mmHg
- estimated RVSP= (22+108)= 130mmHg



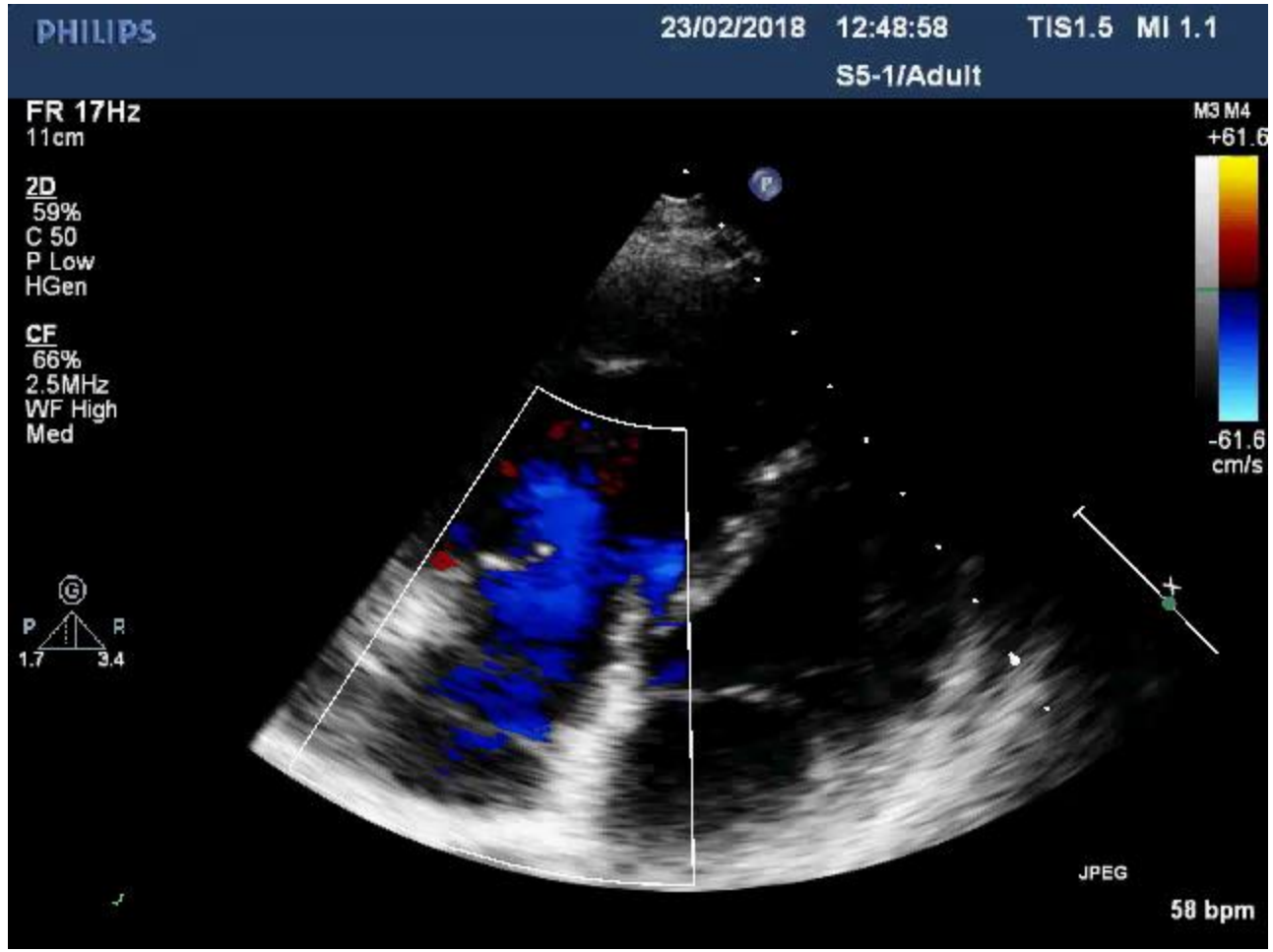


Tricuspid septal leaflet trying to ‘close’ VSD





Diastole: 1.24-1.27 cm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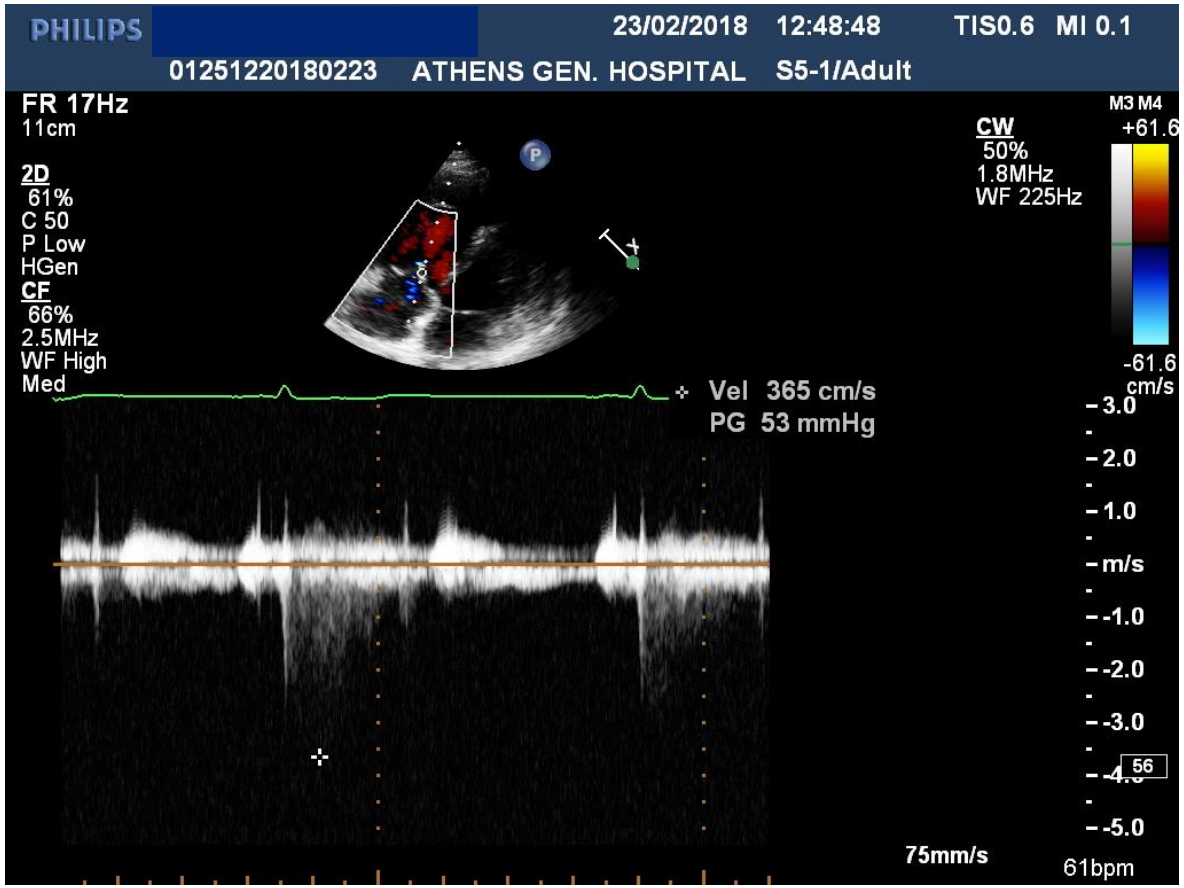
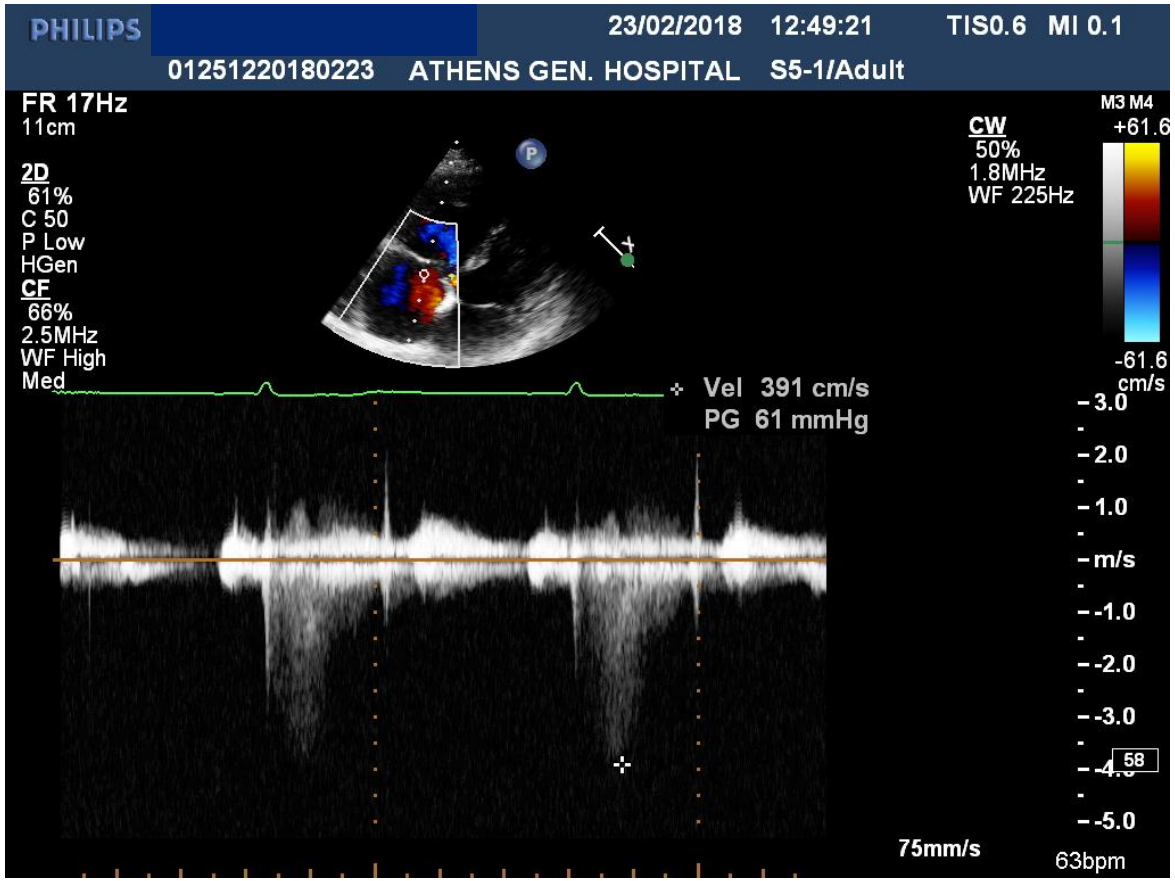
S5-1/Adult
FR 17Hz
11cm
2D
59%
C 50
P Low
HGen
CF
66%
2.5MHz
WF High
Med
M3 M4
+61.6
-61.6
cm/s
P
R
1.7
3.4
JPEG
58 bpm



- TR max Vel= 3.91cm/s (?)
- Estimated RVSP= 61mmHg (?)+ RAP




PHILIPS
23/02/2018 12:50:44 TIS0.9 MI 1.4
S5-1/Adult
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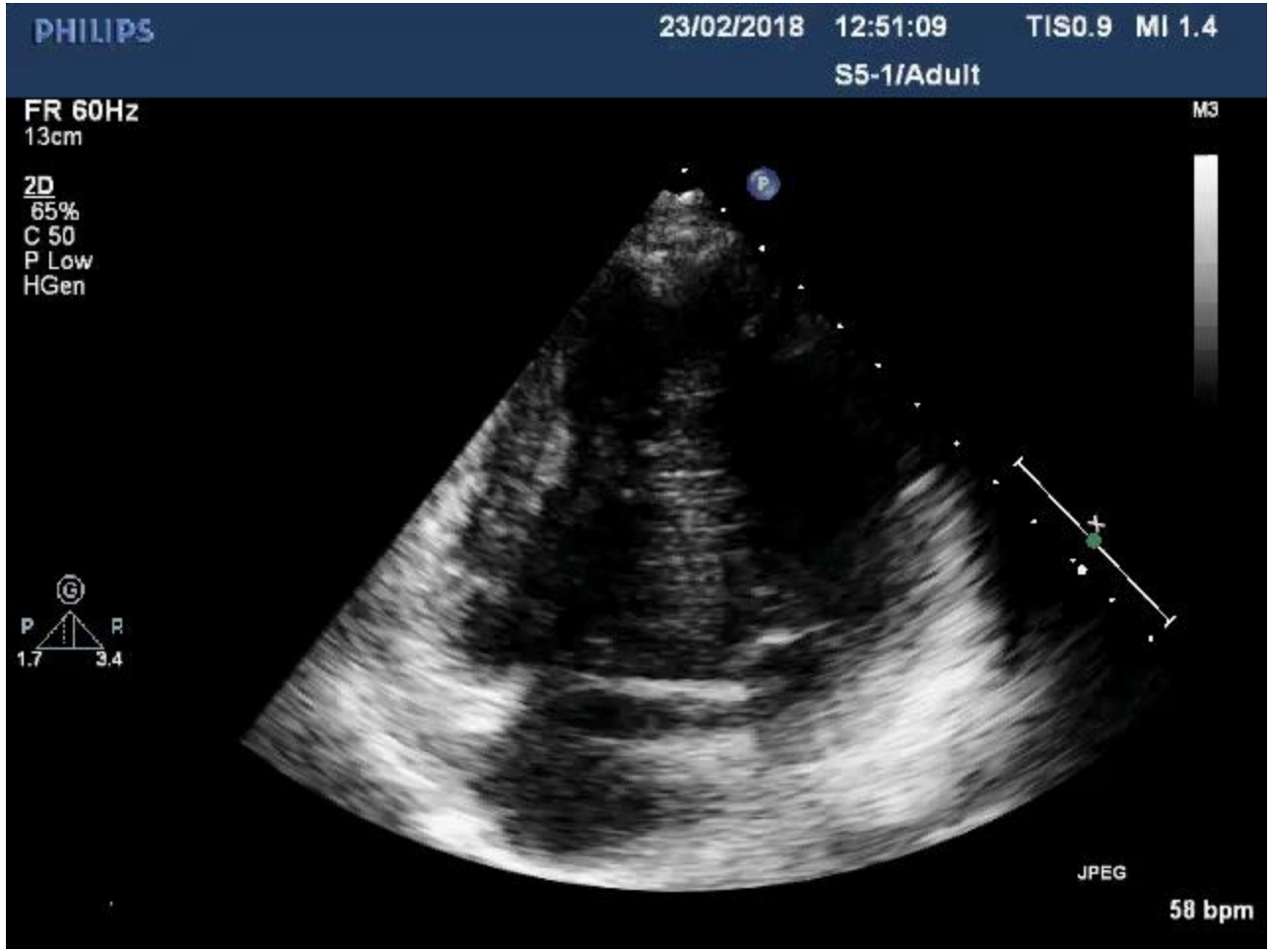
13cm
2D
65%
C 50
P Low
HGen
M3
P
G
P R
1.7 3.4
JPEG
55 bpm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S5-1/Adult
FR 60Hz
13cm
2D
65%
C 50
P Low
HGen
M3
P
G
P R
1.7 3.4
JPEG
58 bpm

2. **What additional information, concerning the RV, can you derive from this A4C view?**

a) there is a hypertrophied moderator band
b) enhanced RV trabeculations
c) possible double-chambered RV
d) no extra findings from this echo view

2. **What additional information, concerning the RV, can you derive from this A4C view?**

a) there is a hypertrophied moderator band
b) enhanced RV trabeculations
c) possible double-chambered RV
d) no extra findings from this echo view


PHILIPS
11/06/2018 16:04:58 TIS0.7 MI 1.3
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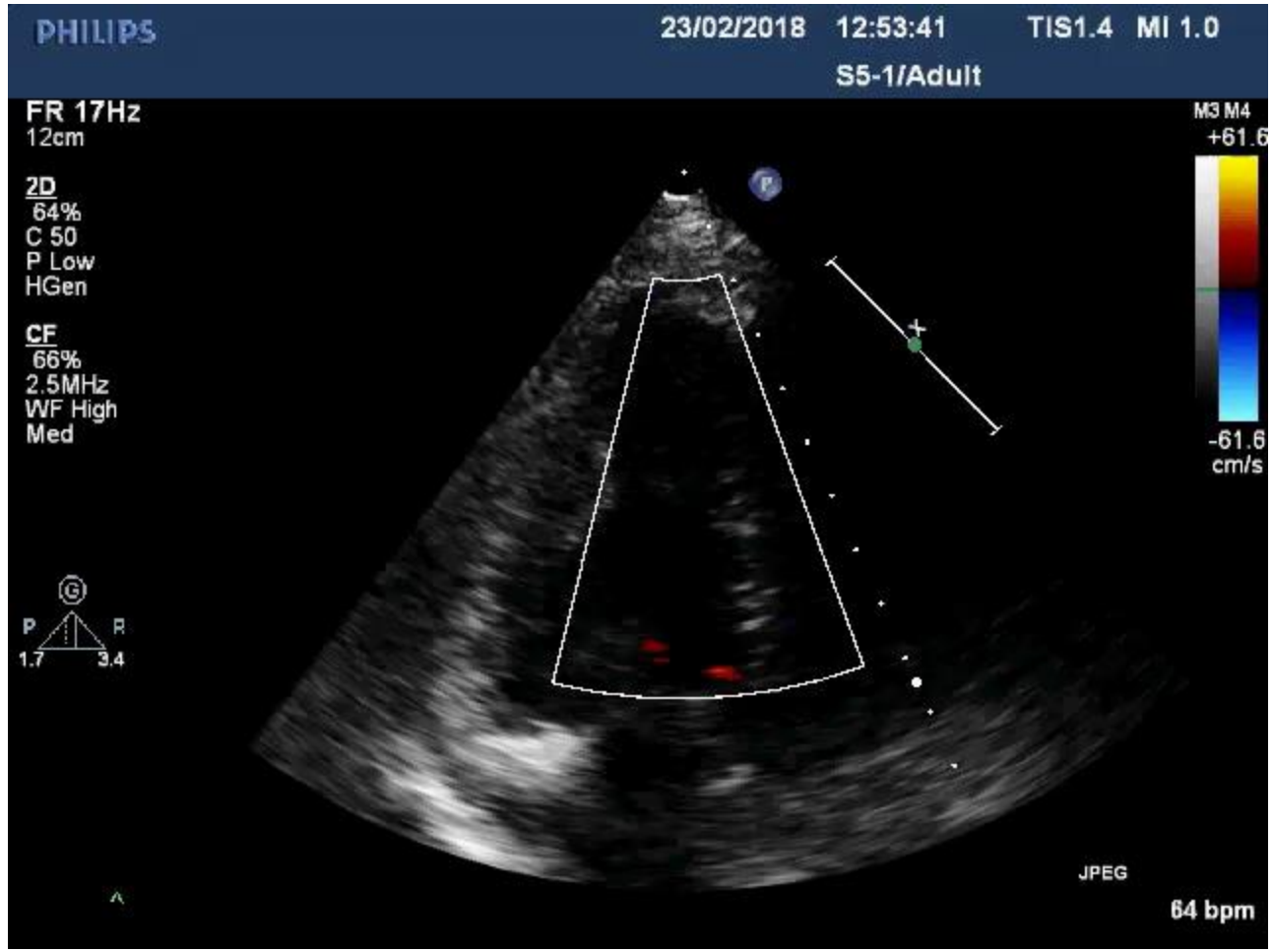
P
G
P R
1.7 3.4
JPEG
56 bpm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2D
64%
C 50
P Low
HGen
CF
66%
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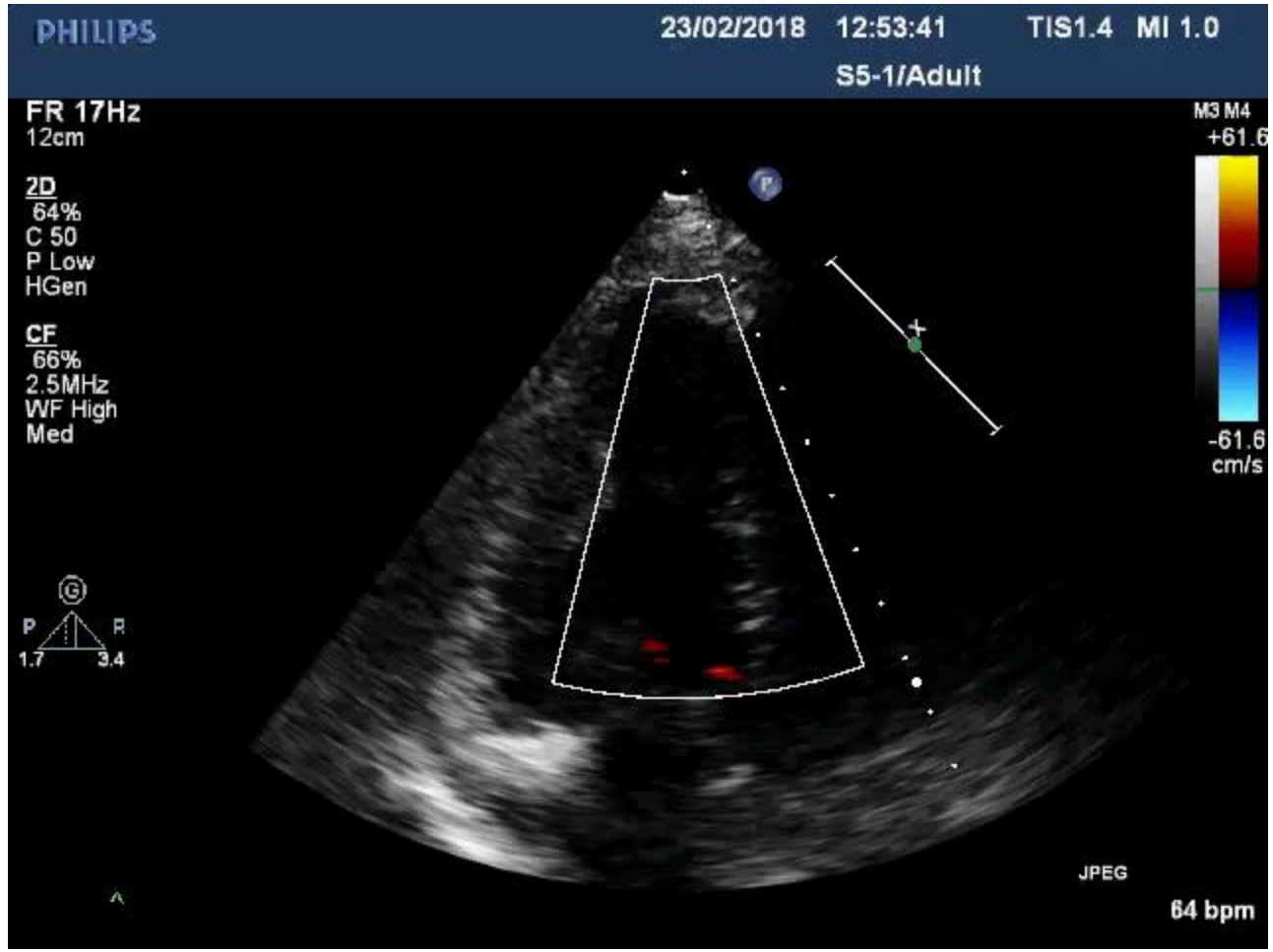
WF High
Med
M3 M4
+61.6
-61.6
cm/s
P
R
1.7
3.4
JPEG
64 bpm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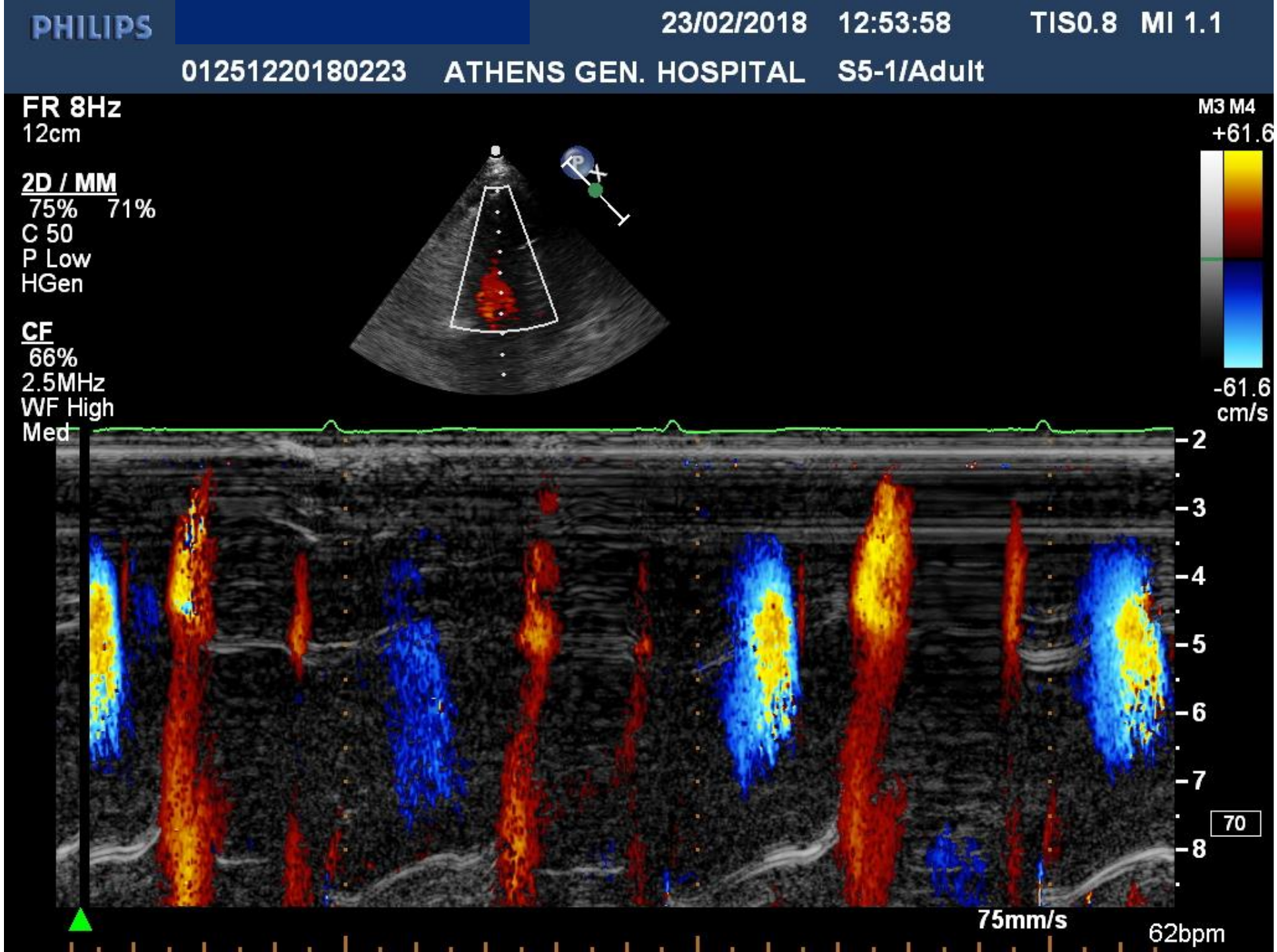
S5-1/Adult
FR 17Hz
12cm
2D
64%
C 50
P Low
HGen
CF
66%
2.5MHz
WF High
Med
M3 M4
+61.6
-61.6
cm/s
P
R
1.7
3.4
JPEG
64 bpm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23/02/2018 12:53:58 TIS0.8 MI 1.1
01251220180223 ATHENS GEN. HOSPITAL S5-1/Adult
FR 8Hz
12cm
2D / MM
75% 71%
C 50
P Low
HGen
CF
66%
2.5MHz
WF High
Med
M3 M4
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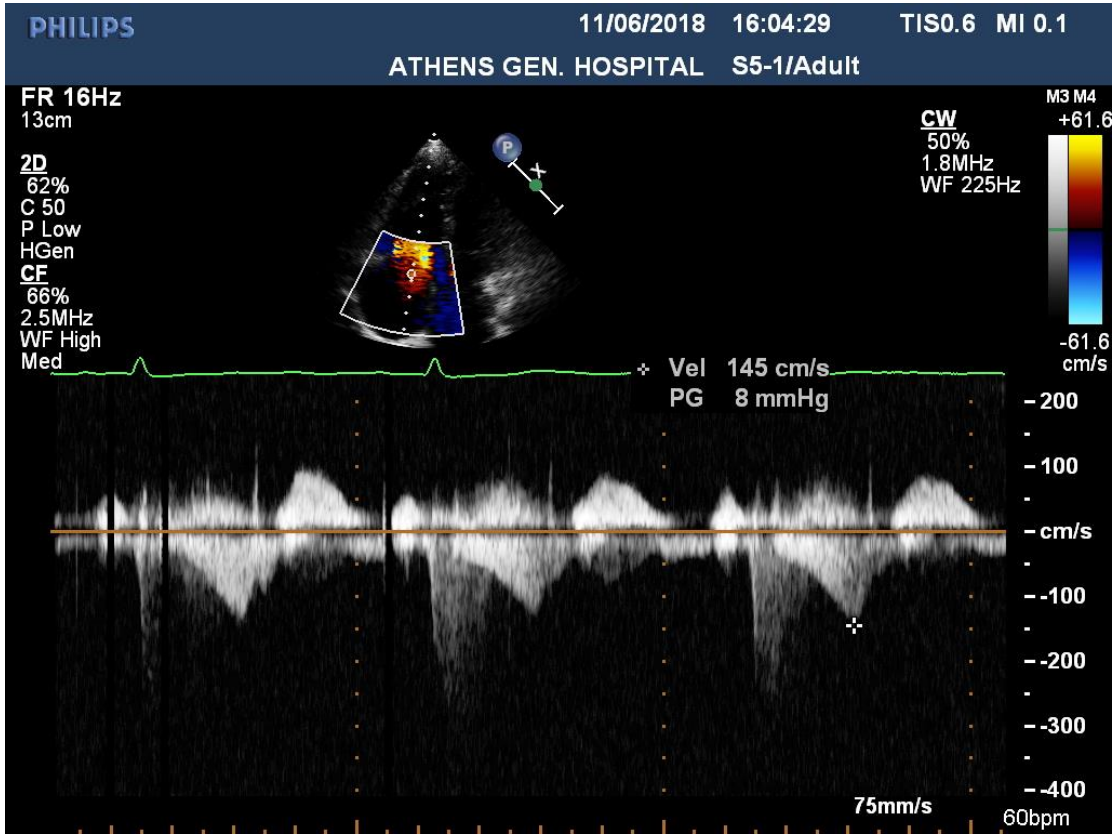
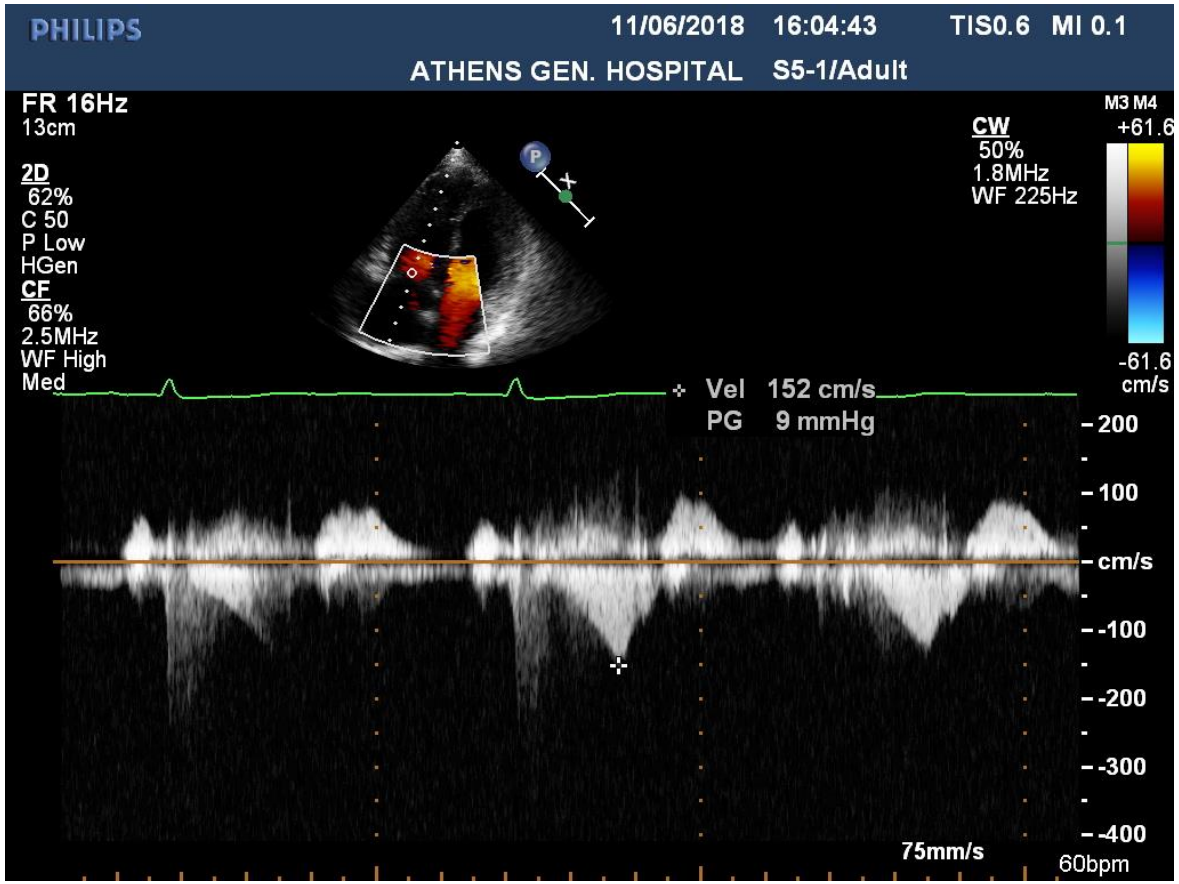
-61.6
cm/s
2
3
4
5
6
7
8
70
75mm/s
62bpm



Max PG estimated to be 9 mmHg

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10cm
2D
62%
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P Low
HPen
M3
G
P
R
1.4
2.8
JPEG
62 bpm


PHILIPS
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66%
C 50
P Low
HPen
CF
66%
2.5MHz
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G
P
R
1.4
2.8
JPEG
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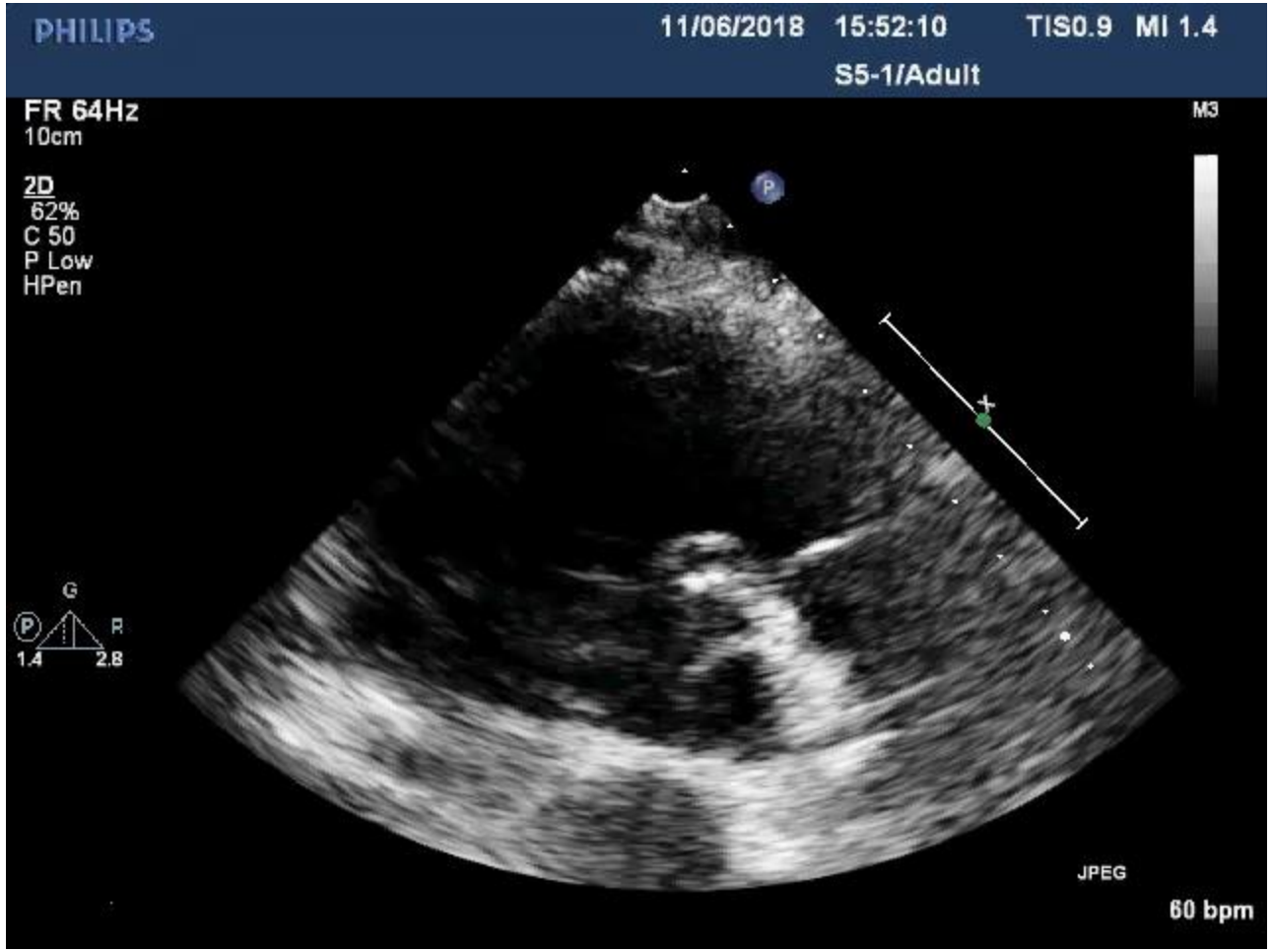
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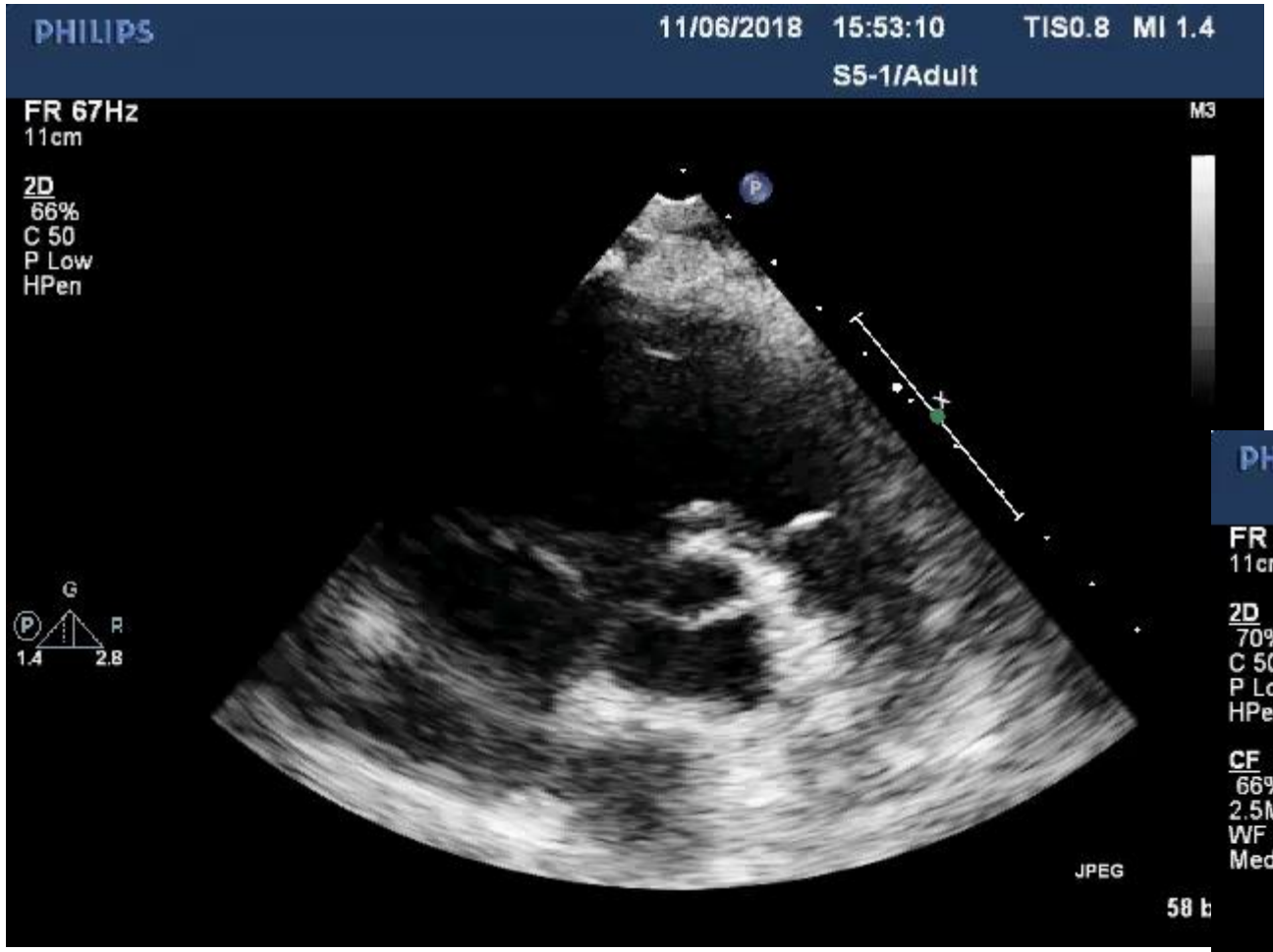
P
R
1.4
2.8
JPEG
60 bpm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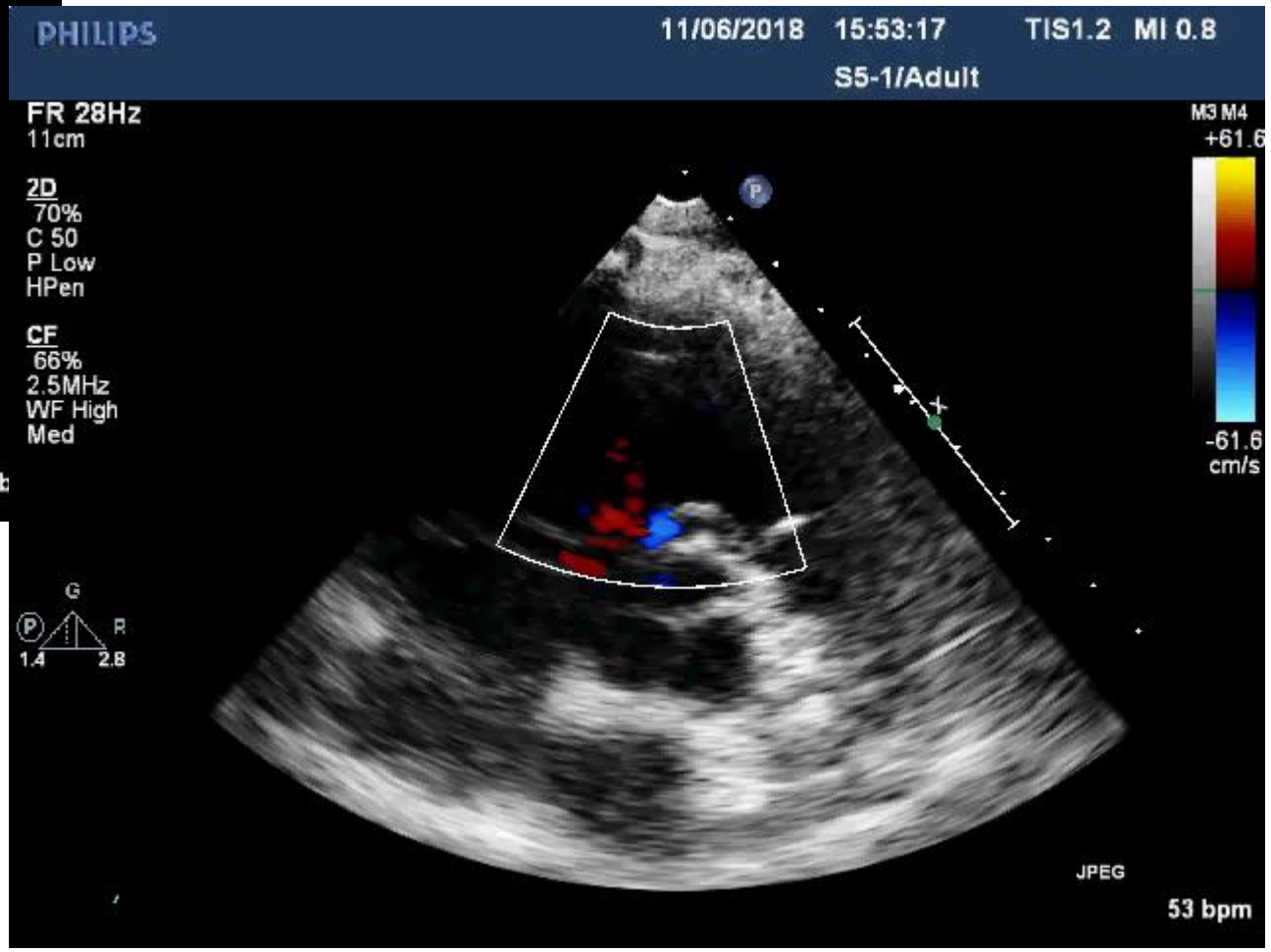
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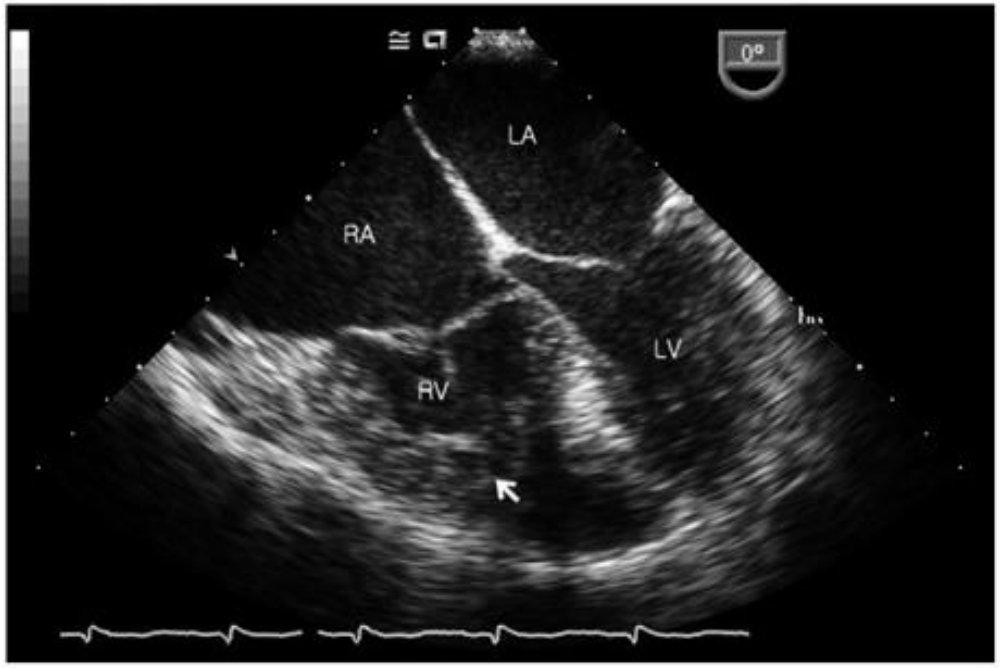
2D
70%
C 50
P Low
HPen
CF
66%
2.5MHz
WF High
Med
M3 M4
+61.6
-61.6
cm/s
G
P
R
1.4
2.8
JPEG
53 bpm

Double-chambered RV (DCRV)

- ✓ A form of septated RV, caused by the presence of abnormally located or hypertrophied muscular bands, that devide the RV cavity into a proximal and a distal chamber (Pressure in distal chamber equals PASP, provided there is no PVS)
- ✓ No uniformity is observed in the position of the anomalous muscle bundles or in the manner in which the RV is divided



Sanatani Shubhayan. The Heart org. Medscape Jan 07 2016
Hoffman P et al. Heart 2004 Jul; 90 (7):789-793
Restio A et al. Ped Cardiol 1984; 5: 197-204

Double-chambered RV (DCRV)

High horizontal obstruction

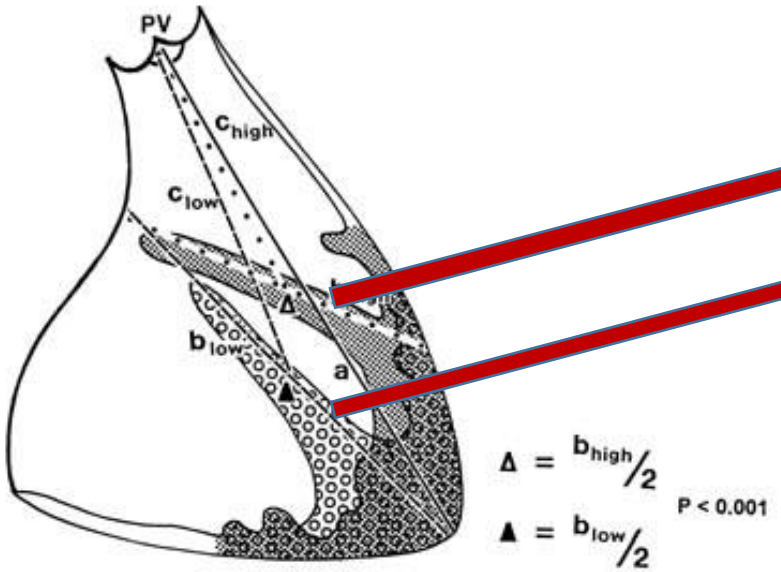
Patient	PV - Δ (C high)	PV-Apex (a)	Ratio
# 4	46	149	0.3
# 8	39	146	0.26
# 10	40	133	0.3
# 13	40	150	0.26
# 14	41	143	0.28
# 17	37	140	0.26
# 19	42	143	0.29
# 21	39	144	0.27
# 22	40	143	0.27
# 24	37	142	0.26
# 25	36	143	0.25
Mean	**39.7**	**143.3**	**0.27**
SD	**2.76**	**4.52**	**0.02**

Low oblique obstruction

Patient	PV - ▲ (C low)	PV - Apex (a)	Ratio
# 2	57	140	0.40
# 3	59	151	0.39
# 5	56	144	0.38
# 7	51	148	0.34
# 9	55	146	0.37
# 11	59	148	0.39
# 12	57	146	0.39
# 15	52	142	0.36
# 23	58	140	0.41
Mean	**56**	**145**	**0.38**
SD	**2.87**	**3.81**	**0.02**

Measurements in mm, SD = Standard deviation
PV = pulmonary valve

Angiocardiographic measurements in patients with double chambered right ventricle



Two positions of muscle bands:

high (horizontal)- adjacent to the PV, or

low (oblique) position- close to the apex

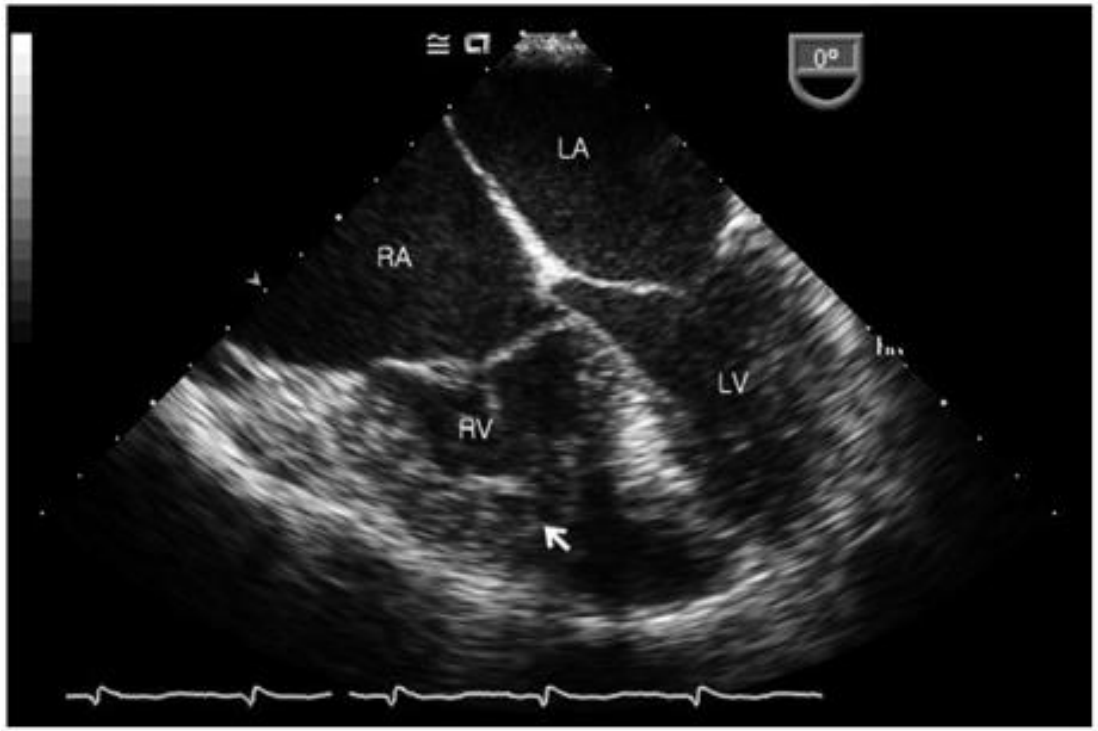
Either position of the **muscular shelf divides the apical trabeculated RV in 2.**

Sanatani Shubhayan. The Heart org. Medscape Jan 07 2016
Hoffman P et al. Heart 2004 Jul; 90 (7):789-793
Restivo A et al. Ped Cardiol 1984; 5: 197-204

Double-chambered RV (DCRV)

- ✓ Frequent associated lesions (80-90%) include a **VSD (that involves the membranous septum- the most common defect described),** pulmonary valve stenosis and discrete subaortic stenosis

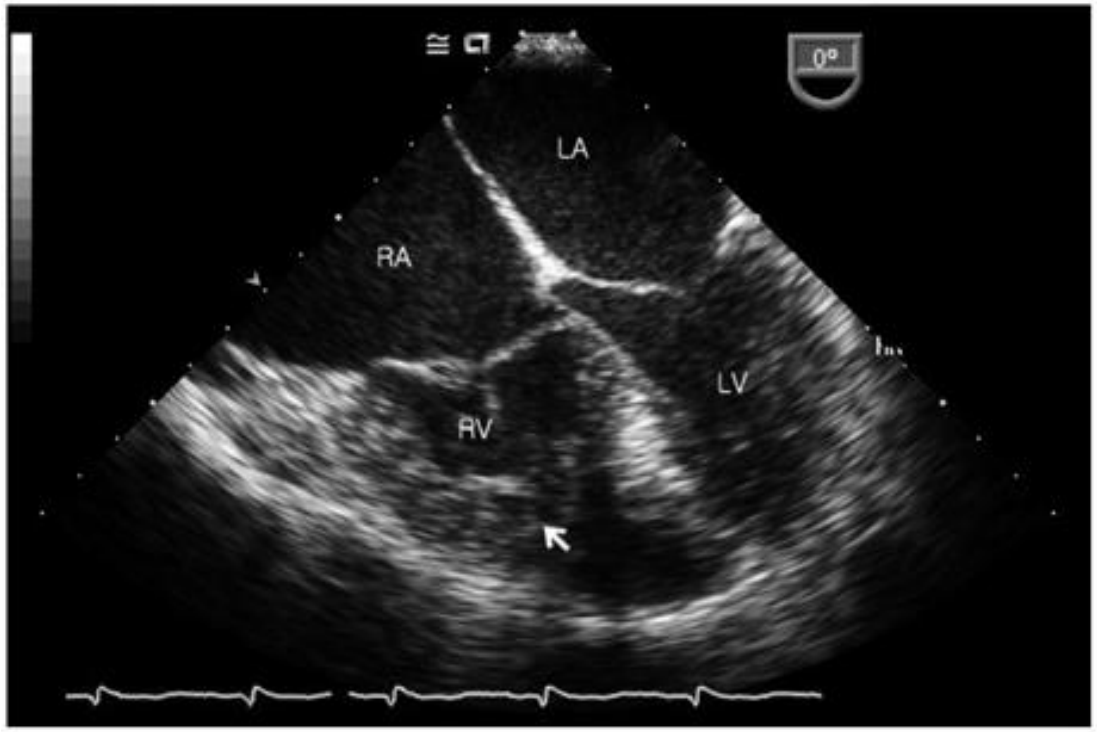
- ✓ It has been suggested that the increased blood flow within the RVOT may act as a stimulus for hypertrophy of the crista supraventricularis in pts with VSD



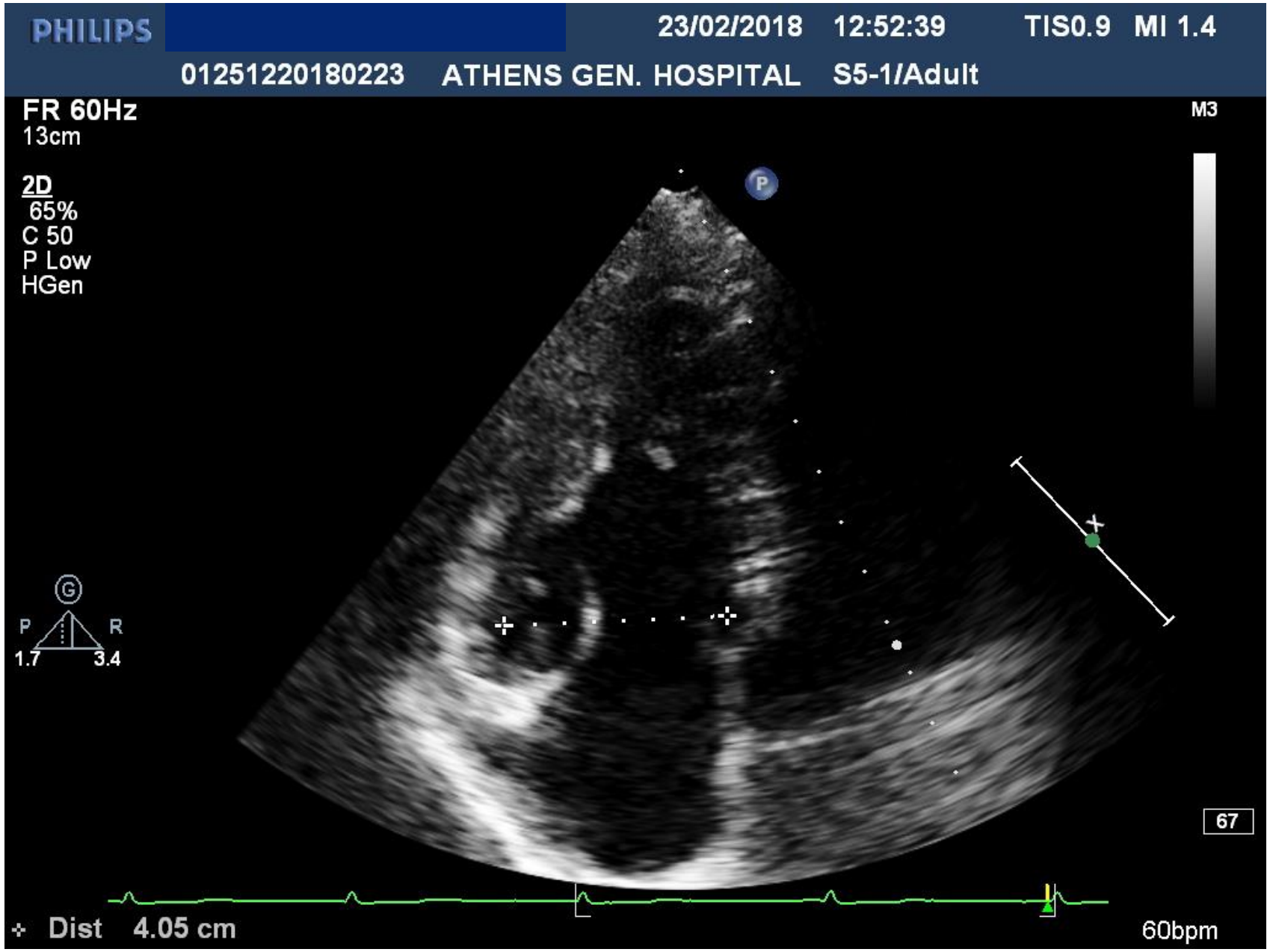
Sanatani Shubhayan. The Heart org. Medscape Jan 07 2016
Hoffman P et al. Heart 2004 Jul; 90 (7):789-793
Restio A et al. Ped Cardiol 1984; 5: 197-204

Double-chambered RV (DCRV)

Group 1: Pulmonary stenosis with intact ventricular septum
Group 2: Tetralogy of Fallot
Group 3: Large VSD
Group 4: Hemodynamically insignificant DCRV as an associated anomaly

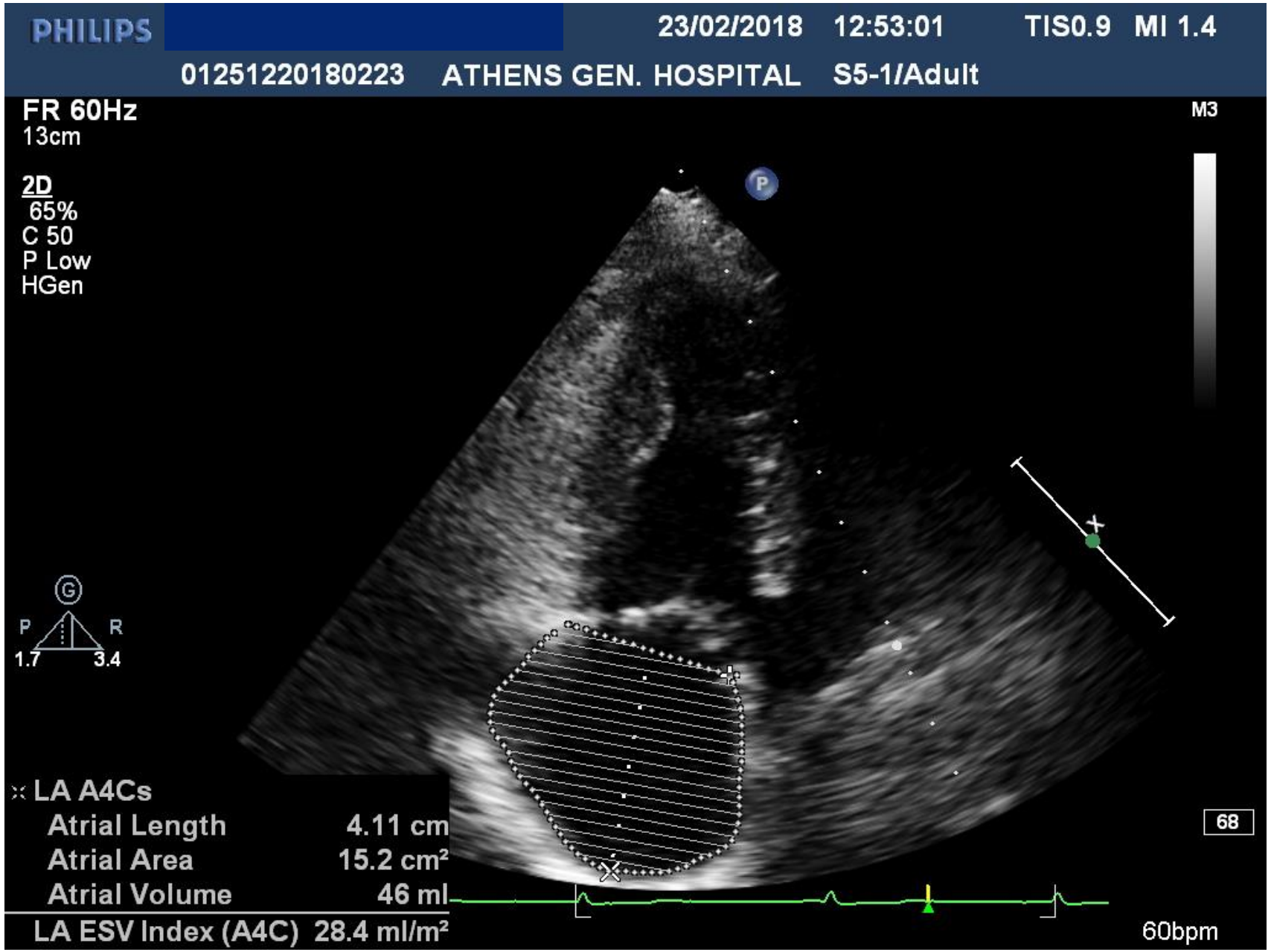


Rowland TW et al. Am Heart Journal 1975; 89: 455-62

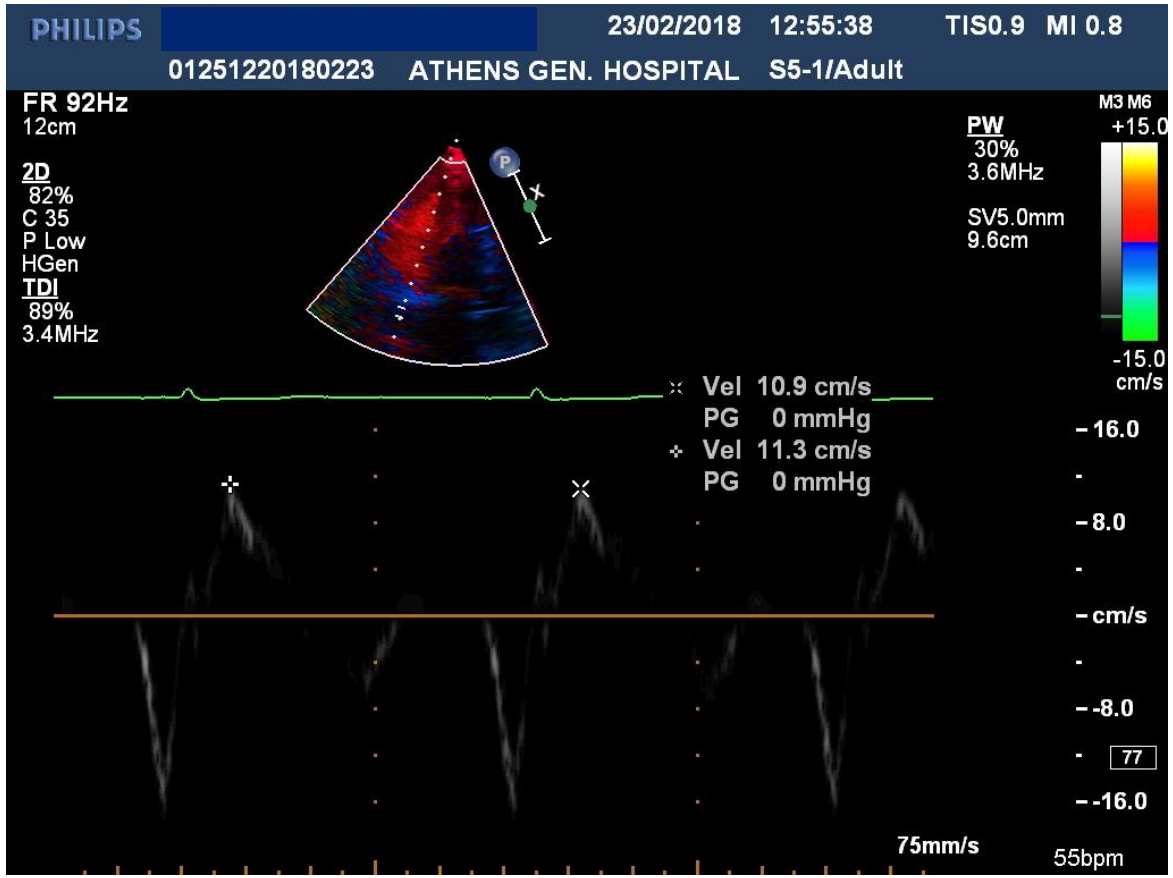


Patient BSA: 1.62m²

- RVD1= 40.5mm
- RV/LV basal diameter ratio>1



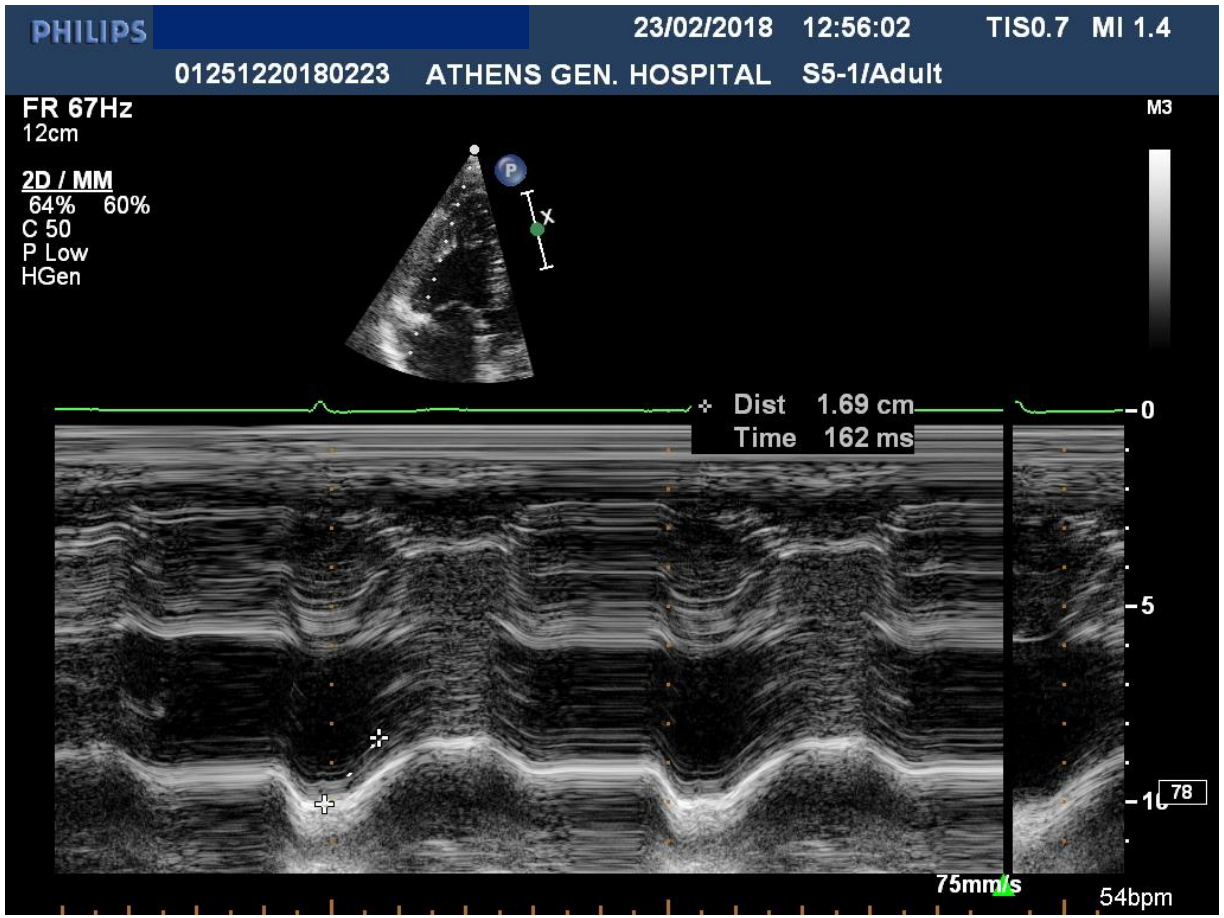
- RA area: 15.2cm² (RA /LA: 1.03)
- RAVI: 28.4ml/m²

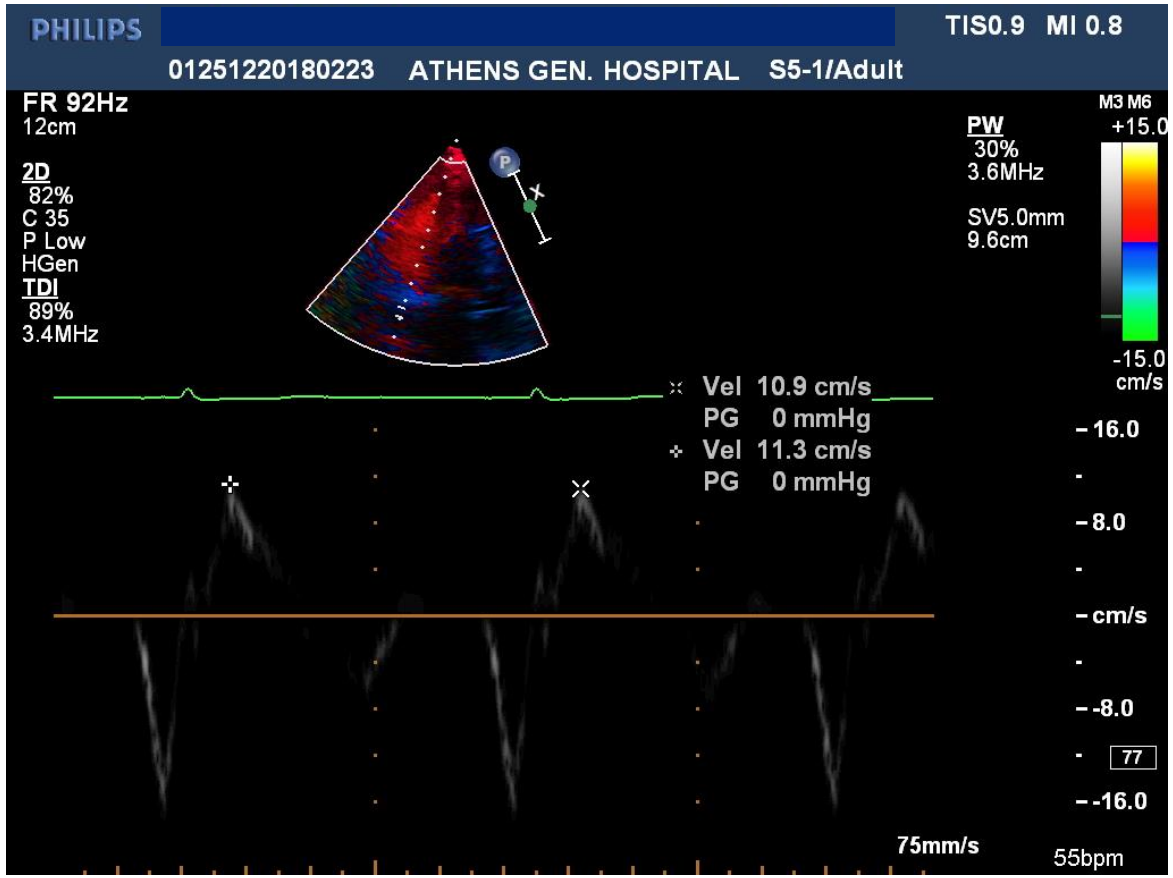


RV Longitudinal systolic function

TAPSE= 17 mm

S-TDI= 10.9 cm/s

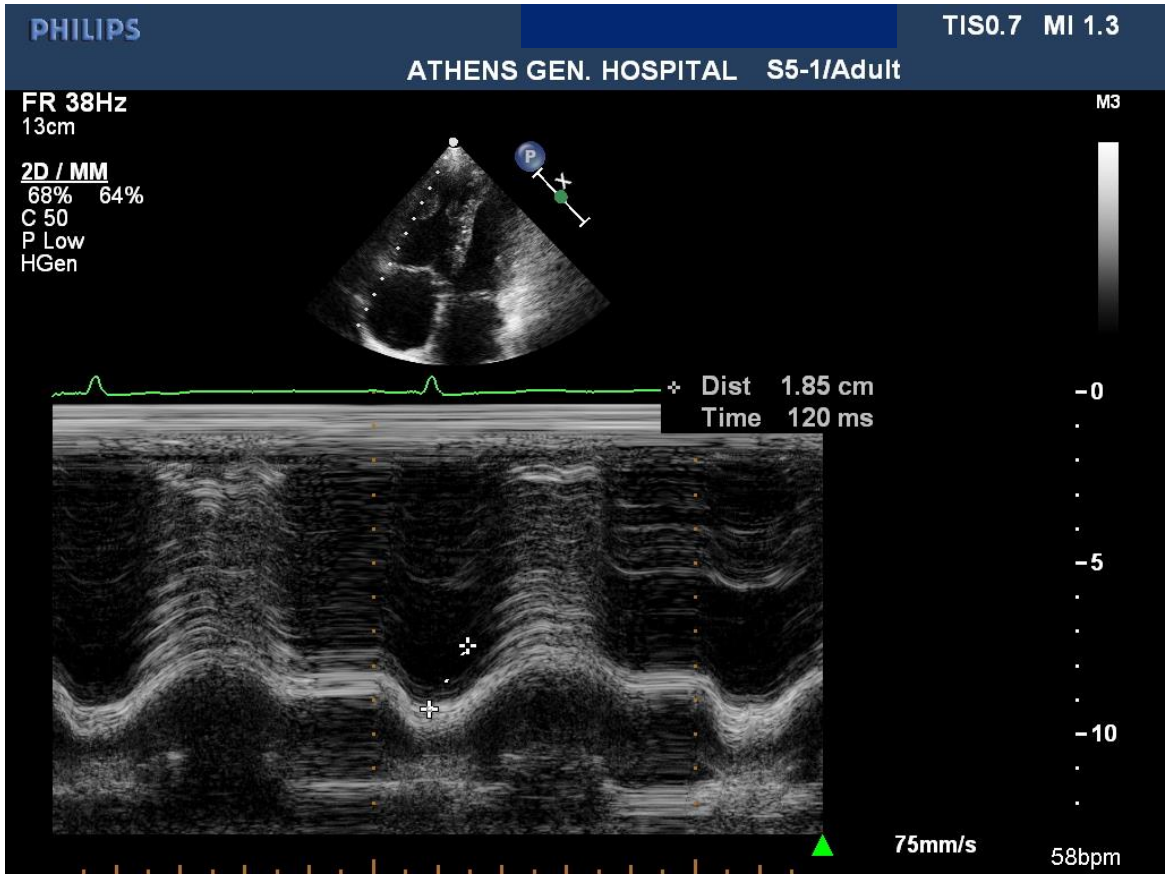


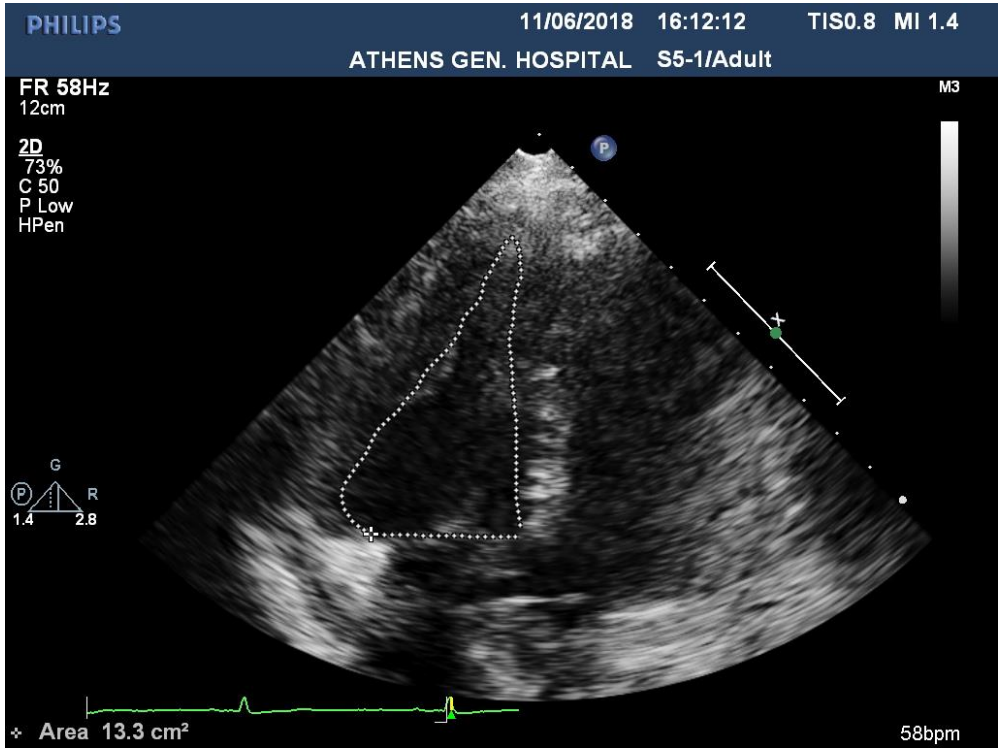


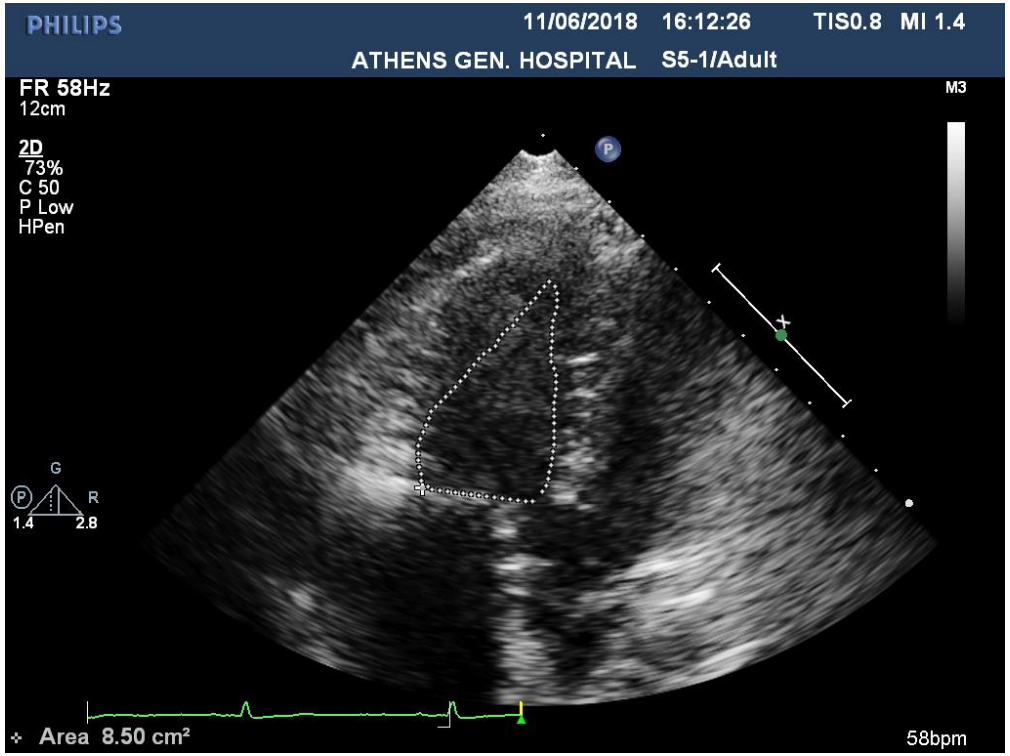
RV Longitudinal systolic function

TAPSE= 18 mm

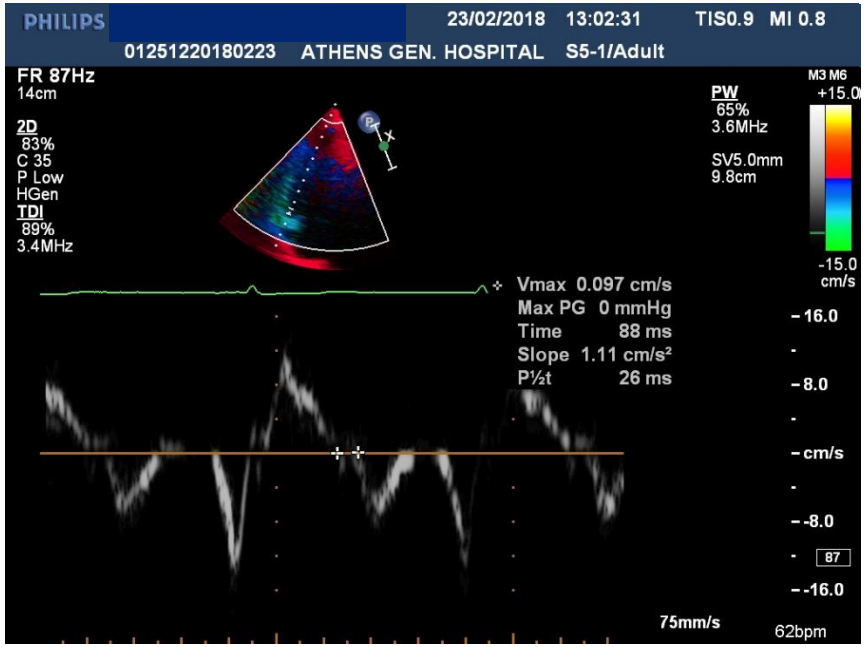
S-TDI= 10.9 cm/s







Fractional area change= 36%



RV MPI (Tei) index:

$$\frac{\text{IVCT + IVRT}}{\text{ET}}$$

= 0.52

3. **Which of the following statements is true?**

a) RV MPI (Myocardial Performance Index) is an index of RV longitudinal systolic function
b) The patient's RIMP value implies normal RV function
c) The patient's RIMP value implies RV dysfunction
d) RAP has to be estimated as normal, in order to conclude that the patient's RIMP value is not falsely low.

3. **Which of the following statements is true?**

a) RV MPI (Myocardial Performance Index) is an index of RV longitudinal systolic function
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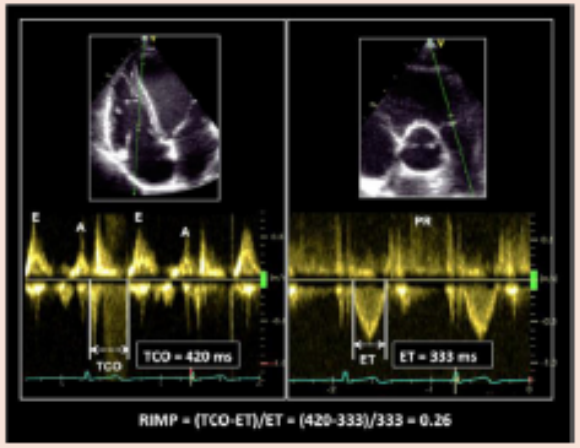
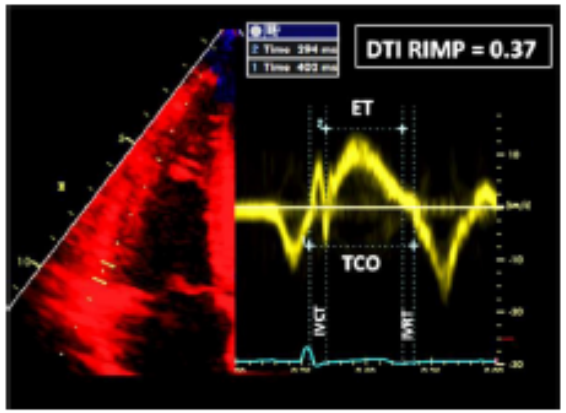
c) The patient's RIMP value implies RV dysfunction
d) RAP has to be estimated as normal, in order to conclude that the patient's RIMP value is not falsely low.

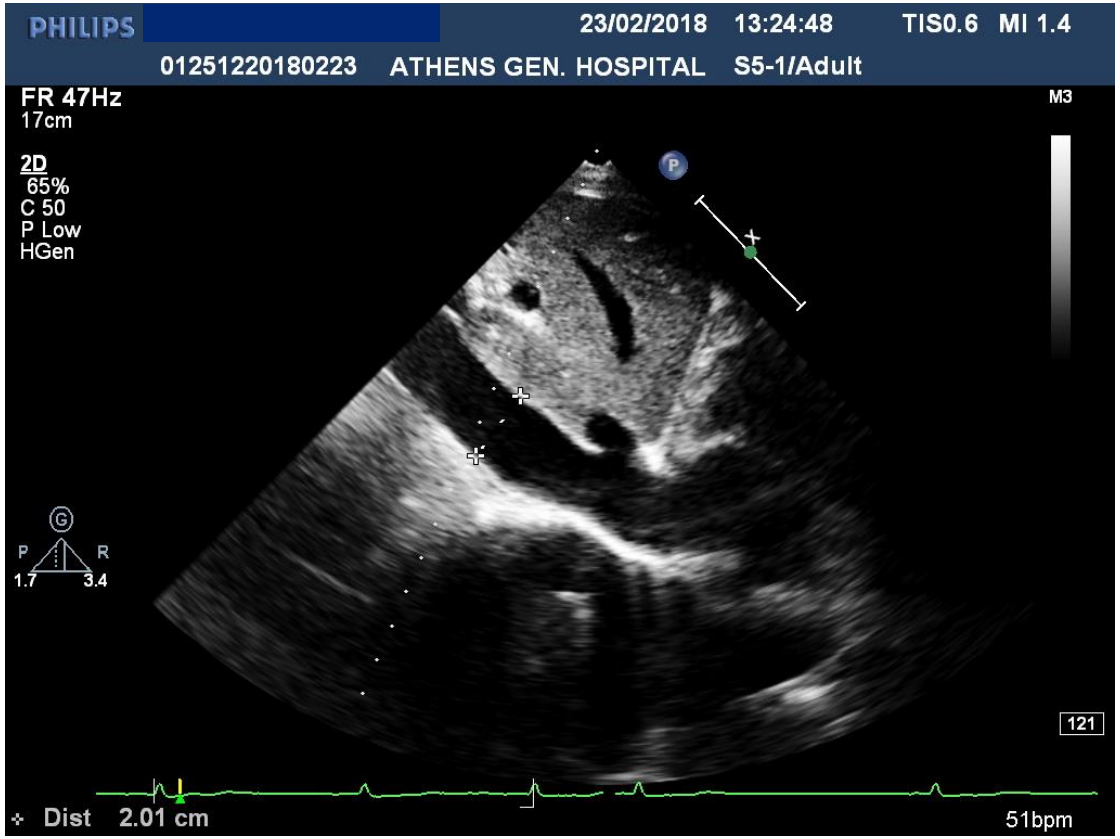
Table 10 Normal values for parameters of RV function

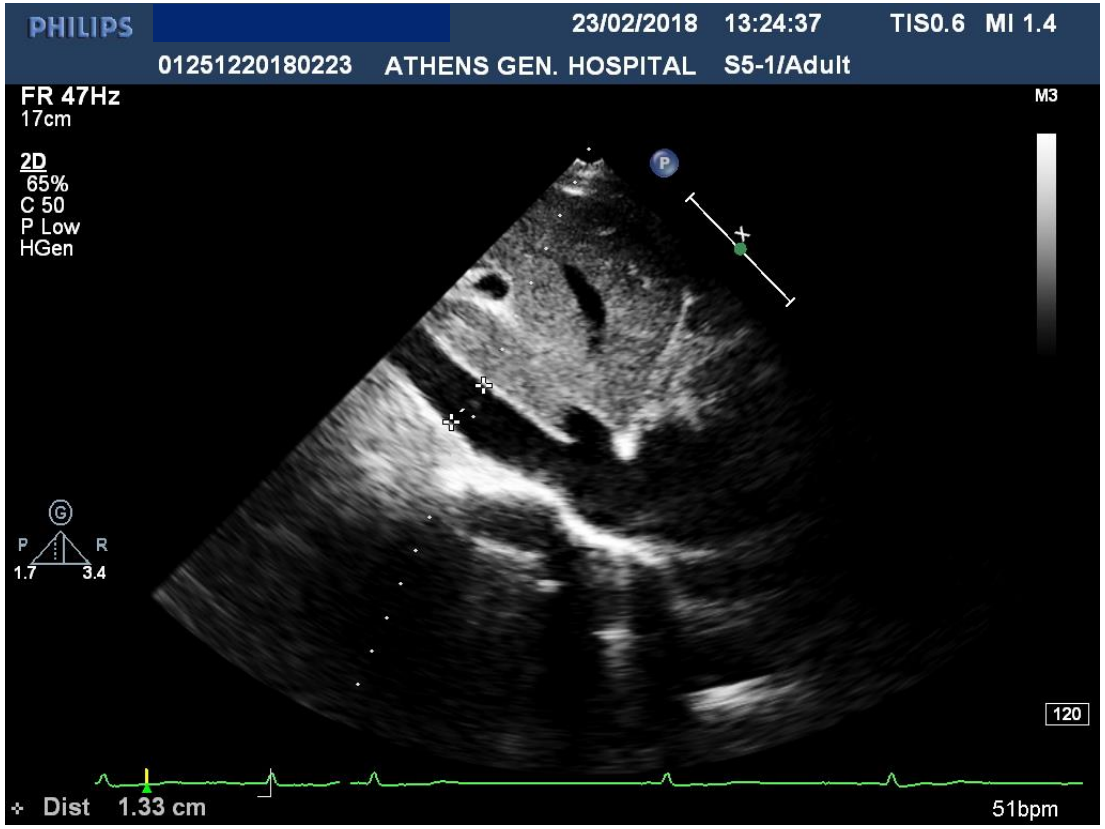
Parameter	Mean ± SD	Abnormality threshold
TAPSE (mm)	24 ± 3.5	<17
Pulsed Doppler S wave (cm/sec)	14.1 ± 2.3	<9.5
Color Doppler S wave (cm/sec)	9.7 ± 1.85	<6.0
RV fractional area change (%)	49 ± 7	<35
RV free wall 2D strain* (%)	−29 ± 4.5	>−20 (<20 in magnitude with the negative sign)
RV 3D EF (%)	58 ± 6.5	<45
Pulsed Doppler MPI	0.26 ± 0.085	>0.43
Tissue Doppler MPI	0.38 ± 0.08	>0.54
E wave deceleration time (msec)	180 ± 31	<119 or >242
E/A	1.4 ± 0.3	<0.8 or >2.0
e′/a′	1.18 ± 0.33	<0.52
e′	14.0 ± 3.1	<7.8
E/e′	4.0 ± 1.0	>6.0

Lang RM et al. JASE 2015

RV global function Pulsed Doppler RIMP 	RIMP (Tei index) by pulsed Doppler: RIMP = (TCO – ET)/ET	• Prognostic value • Less affected by heart rate	• Requires matching for R-R intervals when measurements are performed on separate recordings • Unreliable when RA pressure is elevated
Tissue Doppler RIMP 	RIMP by tissue Doppler: RIMP = (IVRT + IVCT)/ET = (TCO – ET)/ET	• Less affected by heart rate • Single-beat recording with no need for R-R interval matching	• Unreliable when RA pressure is elevated

Lang RM et al. JASE 2015





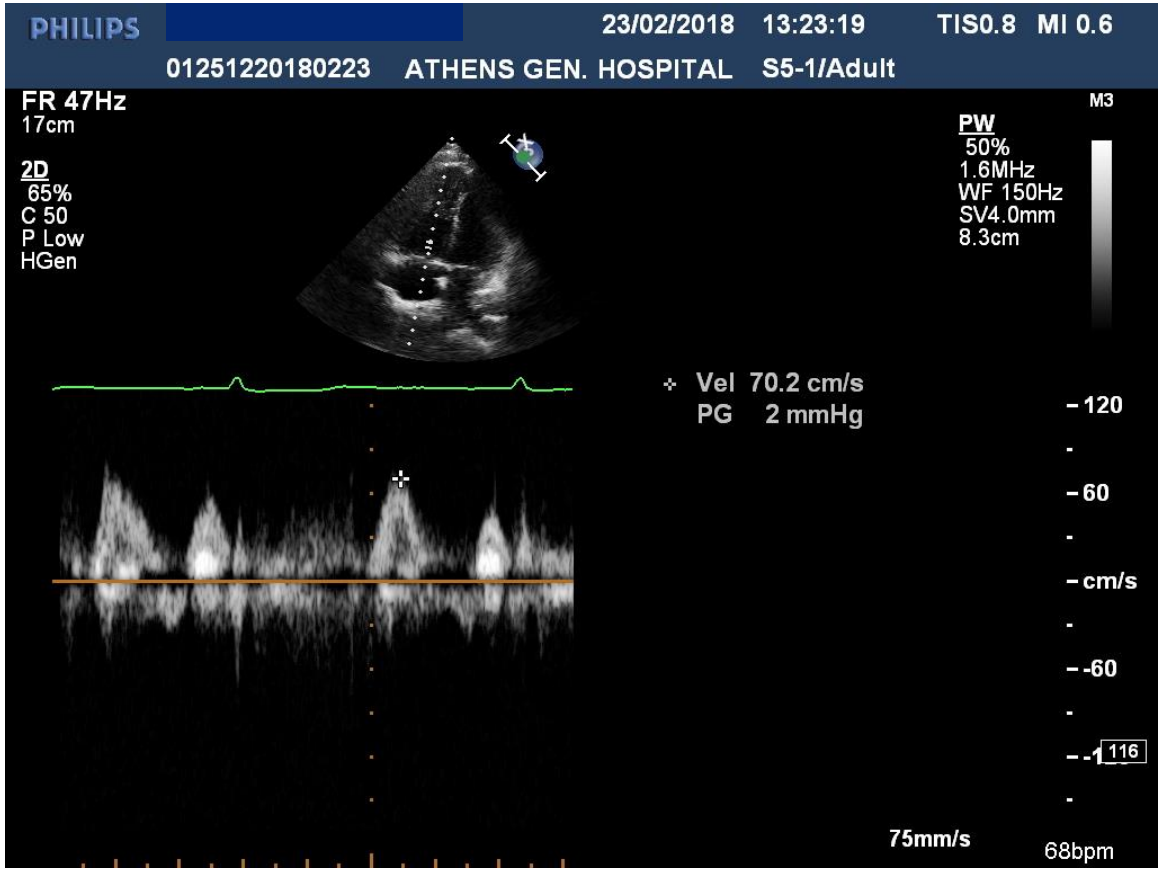
RAP evaluation

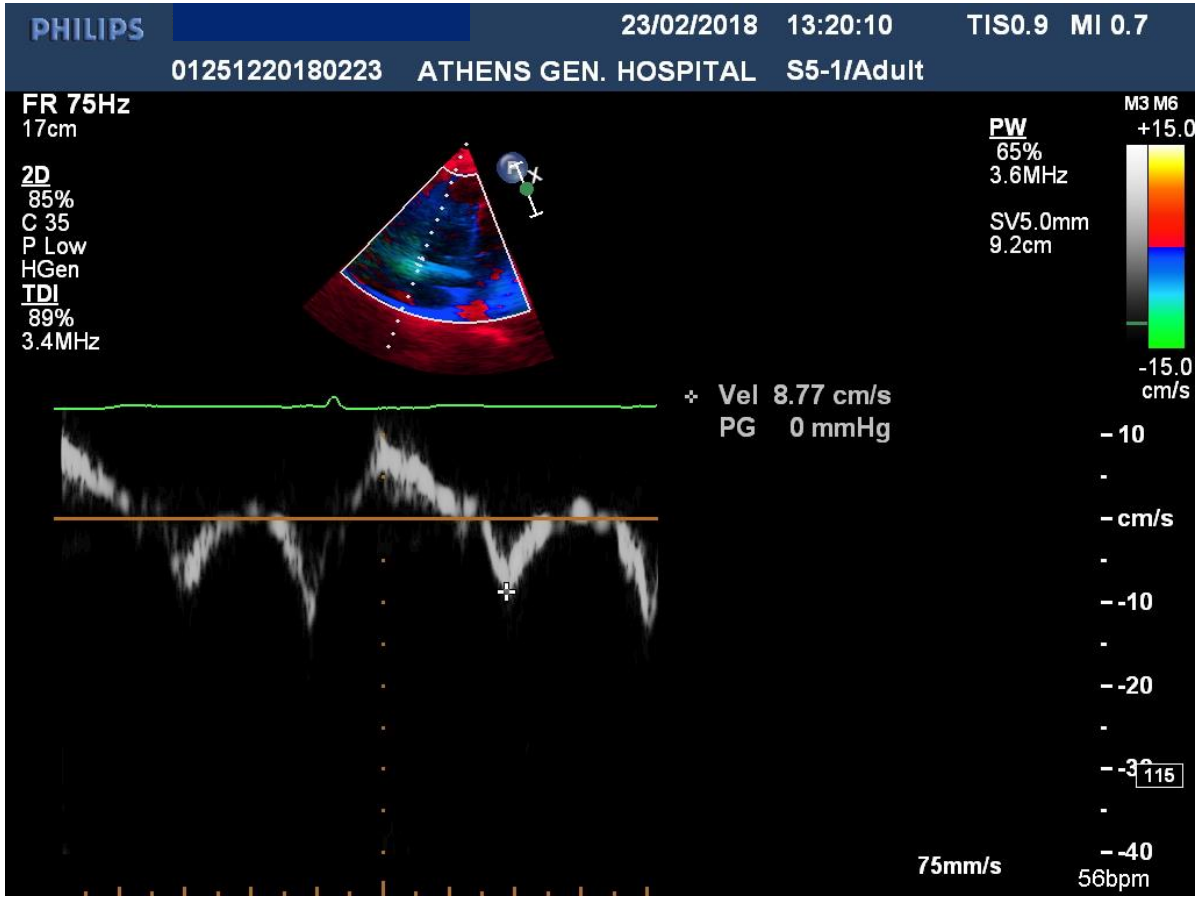
IVC <2.1mm but with decreased inspiratory collapse: <50% with a sniff

RAP conventionally accepted to be 8mmHg (range: 5-10mmHg)

or additional parameters evaluated to better identify elevated RAP

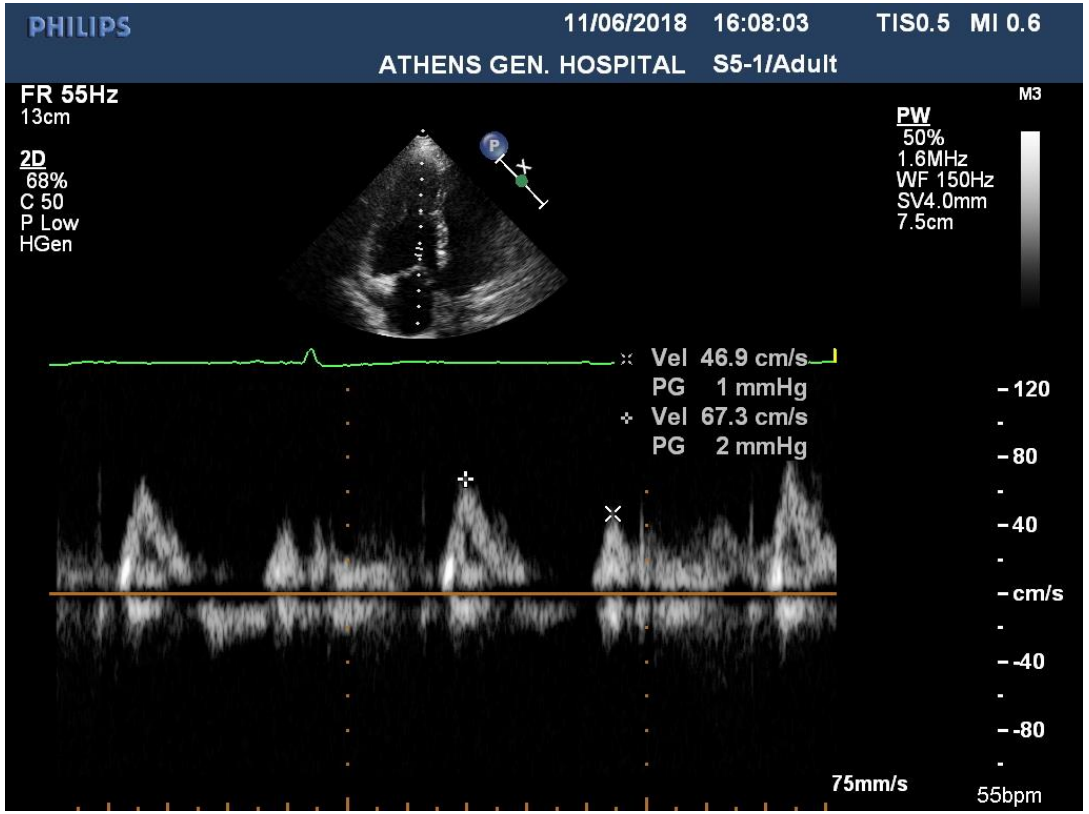
i.e. estimate RV diastolic pressure applying E/e' (or HV systolic fraction)

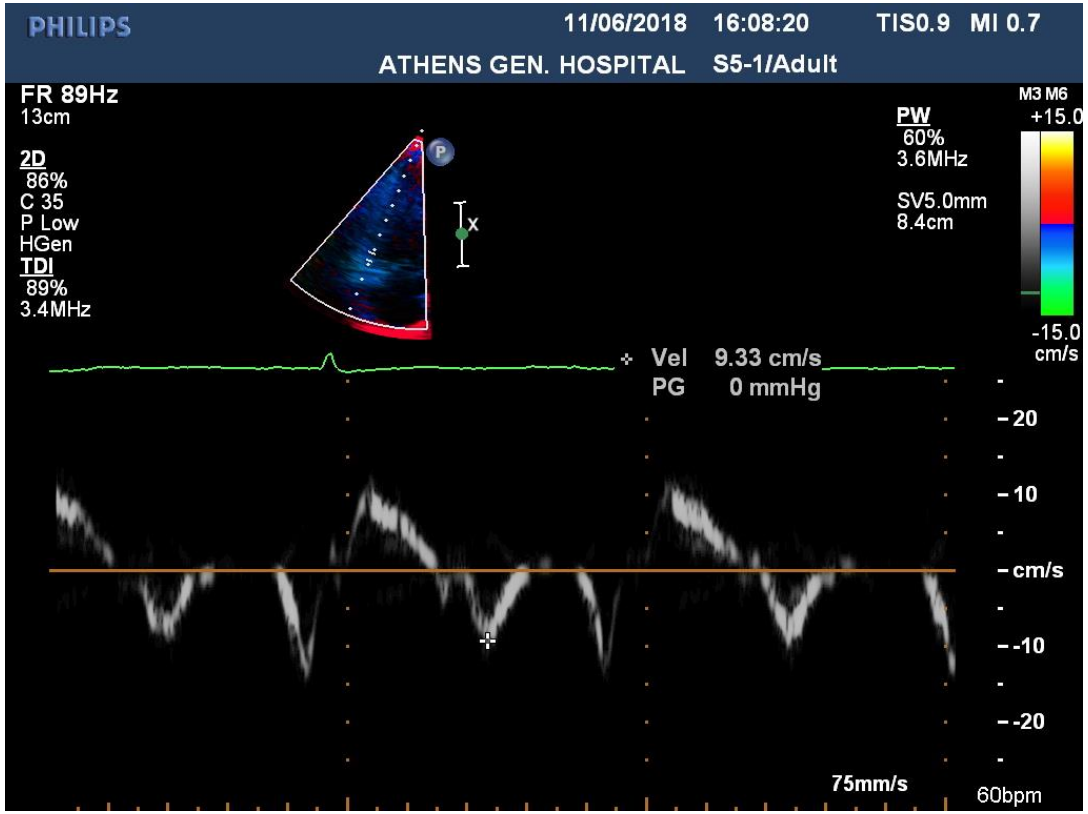




E/e′= 8 (> 6 identifies increased RVEDP) – RAP= 15mmHg

- RV MPI may not be reliable





E/e΄= 7,2 (> 6 identifies increased RVEDP) – RAP= 15mmHg

- RV MPI may not be reliable

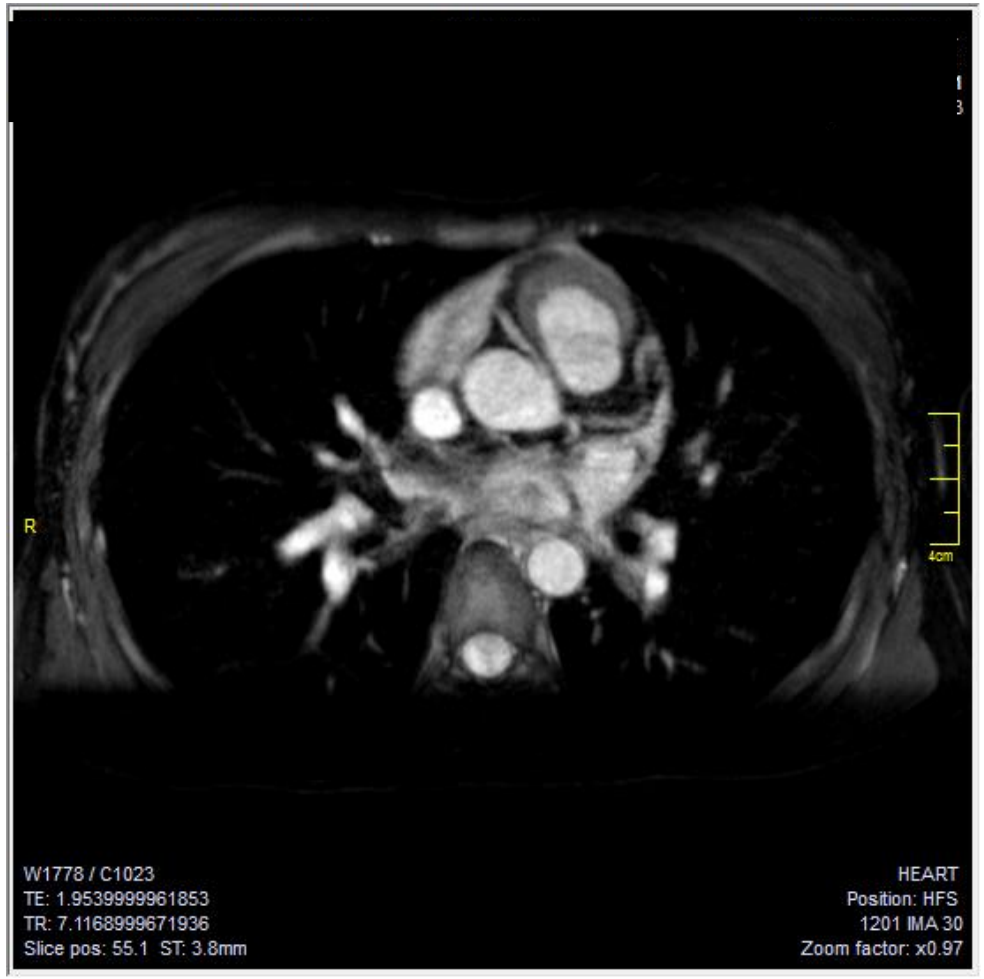
CMR

- Μεσοκοιλιακή επικοινωνία στο βασικό τμήμα του ΜΚΔ, με απεικονιστικές ενδείξεις δεξιά-αριστερά επικοινωνίας (Qp/Qs: 0,7) και ενδείξεις σημαντικής πνευμονικής υπέρτασης (ευρήματα συμβατά με σύνδρομο Eisenmenger). Όγκος επικοινωνίας (shunt volume): 14ml/καρδιακό κύκλο.

- Φυσιολογικό μέγεθος της αριστερής κοιλίας, με φυσιολογική συστολική λειτουργικότητα

- Διάταση και υπερτροφία της δεξιάς κοιλίας, με φυσιολογική συστολική λειτουργικότητα (EF= 61%)

- **Ανώμαλη έκφυση της δεξιάς στεφανιαίας αρτηρίας από τον αριστερό κόλπο του Valsalva, με οξεία γωνίωση προς τα δεξιά και πορεία αυτής μεταξύ της αορτής και του στελέχους της πνευμονικής αρτηρίας (διαμέτρου 31mm), μέχρις ότου εισέλθει στη δεξιά κολποκοιλιακή αύλακα.**

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2D
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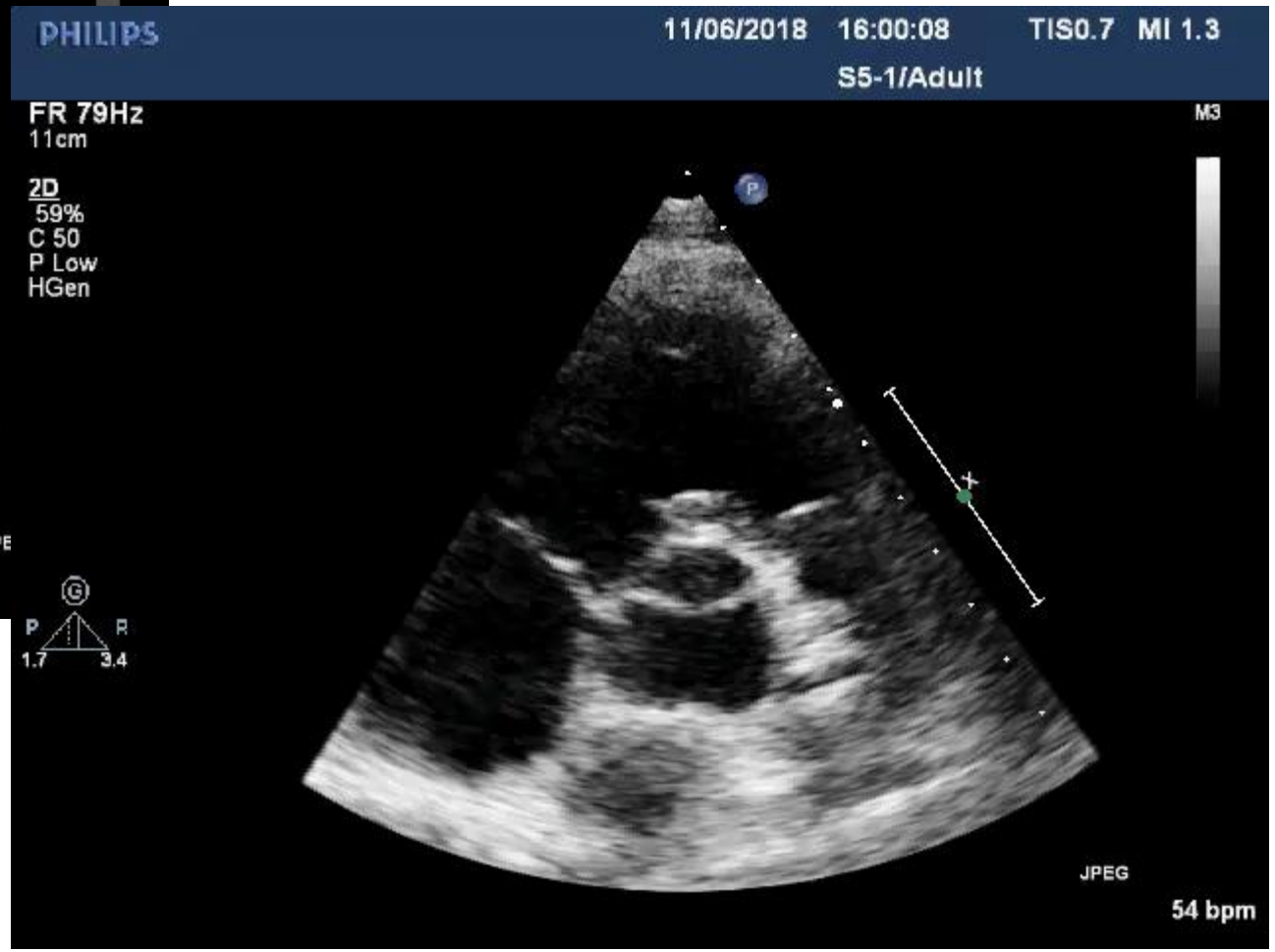
C 50
P Low
HGen
M3
P R
1.7 3.4
JPE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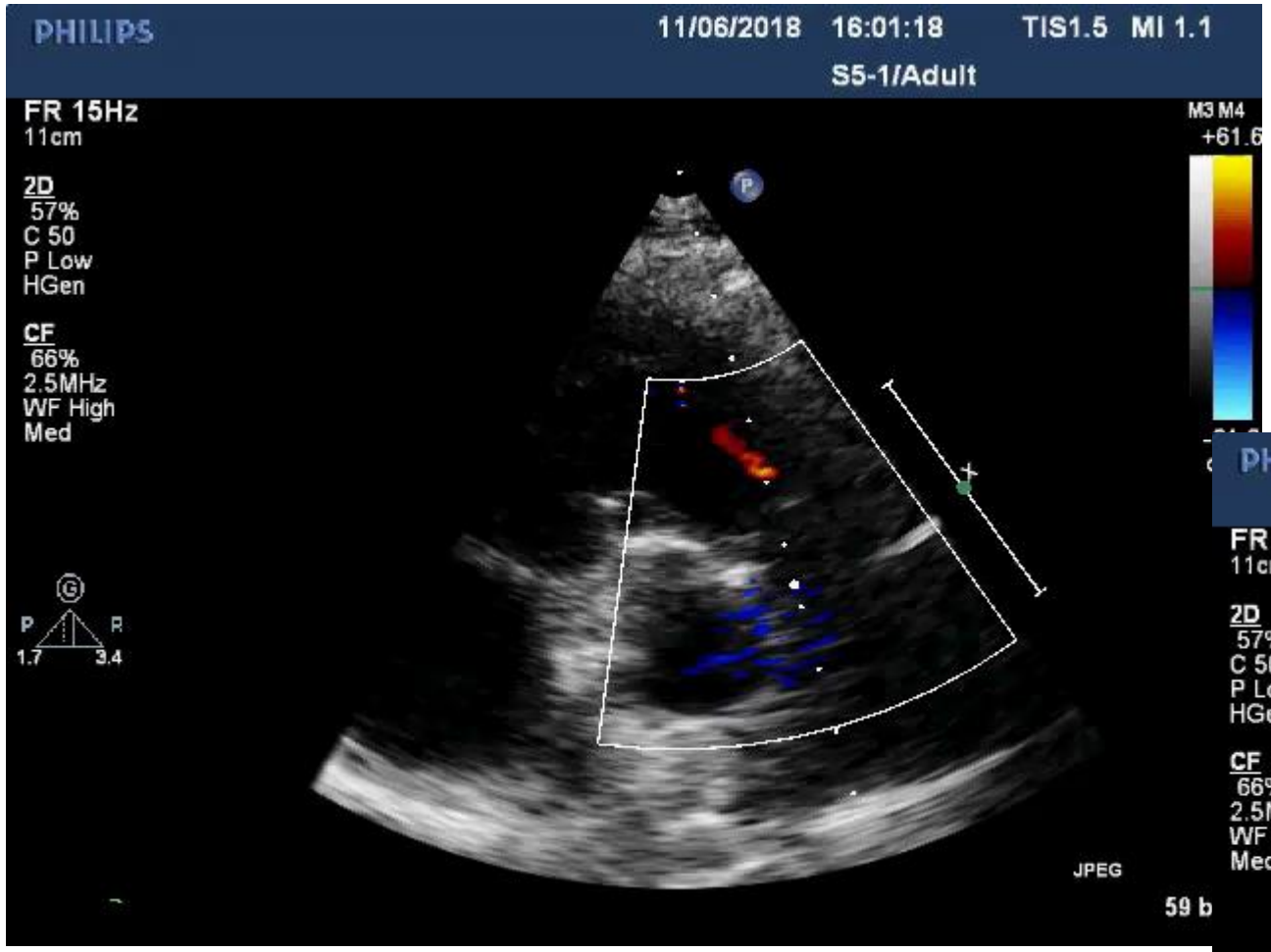
P R
1.7 3.4
JPEG
54 bpm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CF
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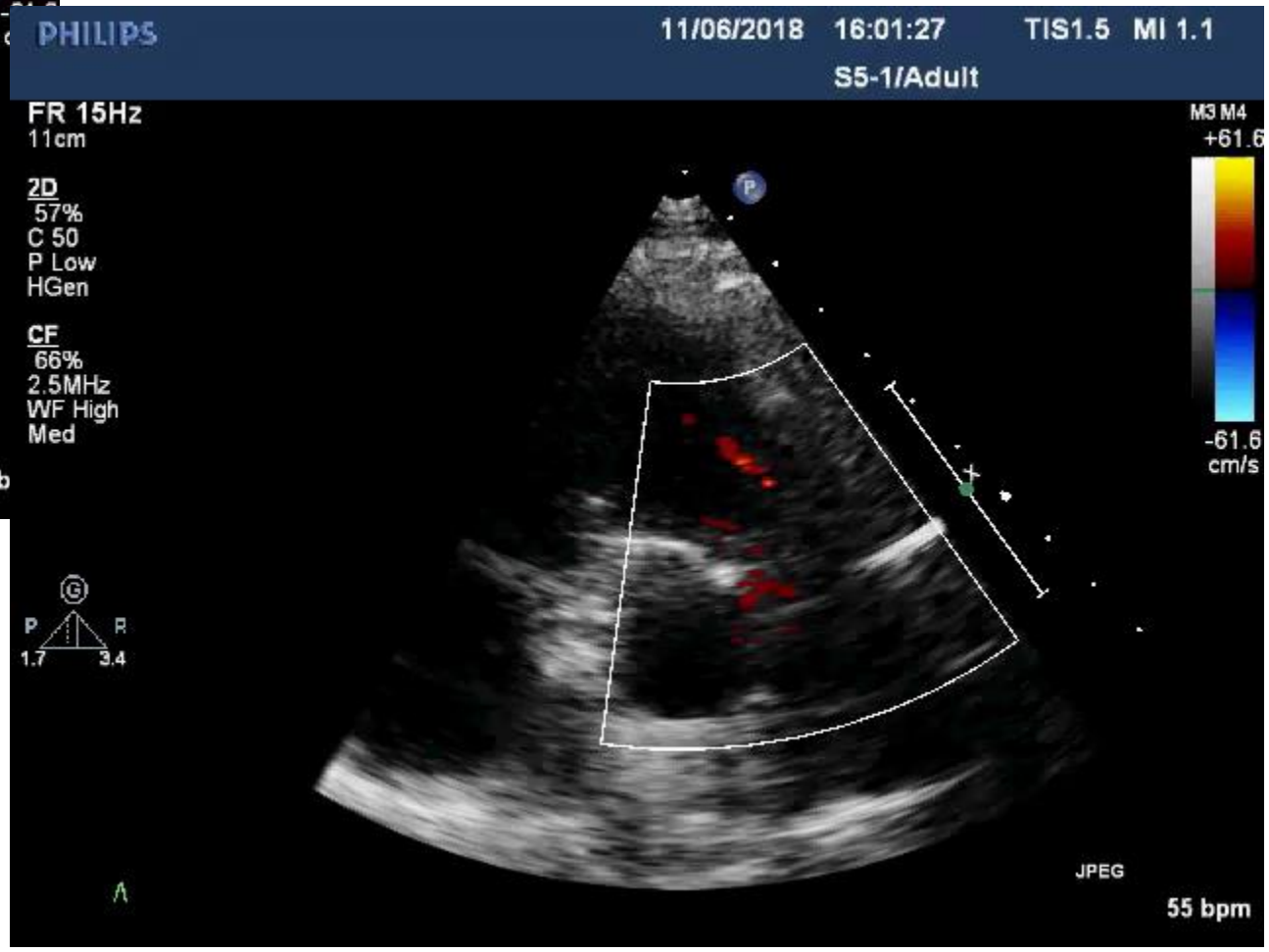
2.5MHz
WF High
Med
M3 M4
+61.6
P
R
1.7
3.4
JPEG
59 b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2D
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C 50
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CF
66%
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M3 M4
+61.6
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cm/s
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1.7
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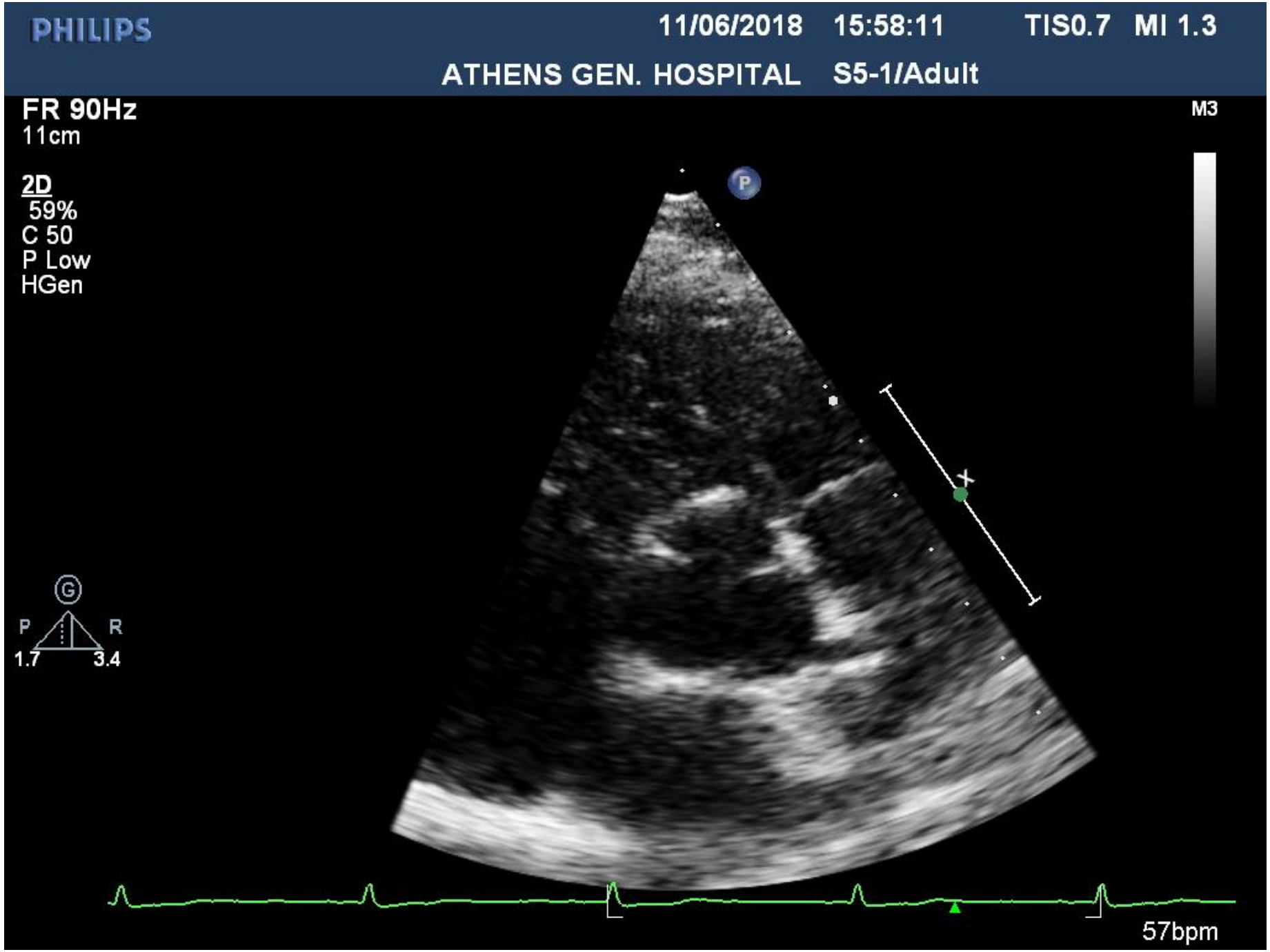
JPEG
55 bpm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P
G
P
R
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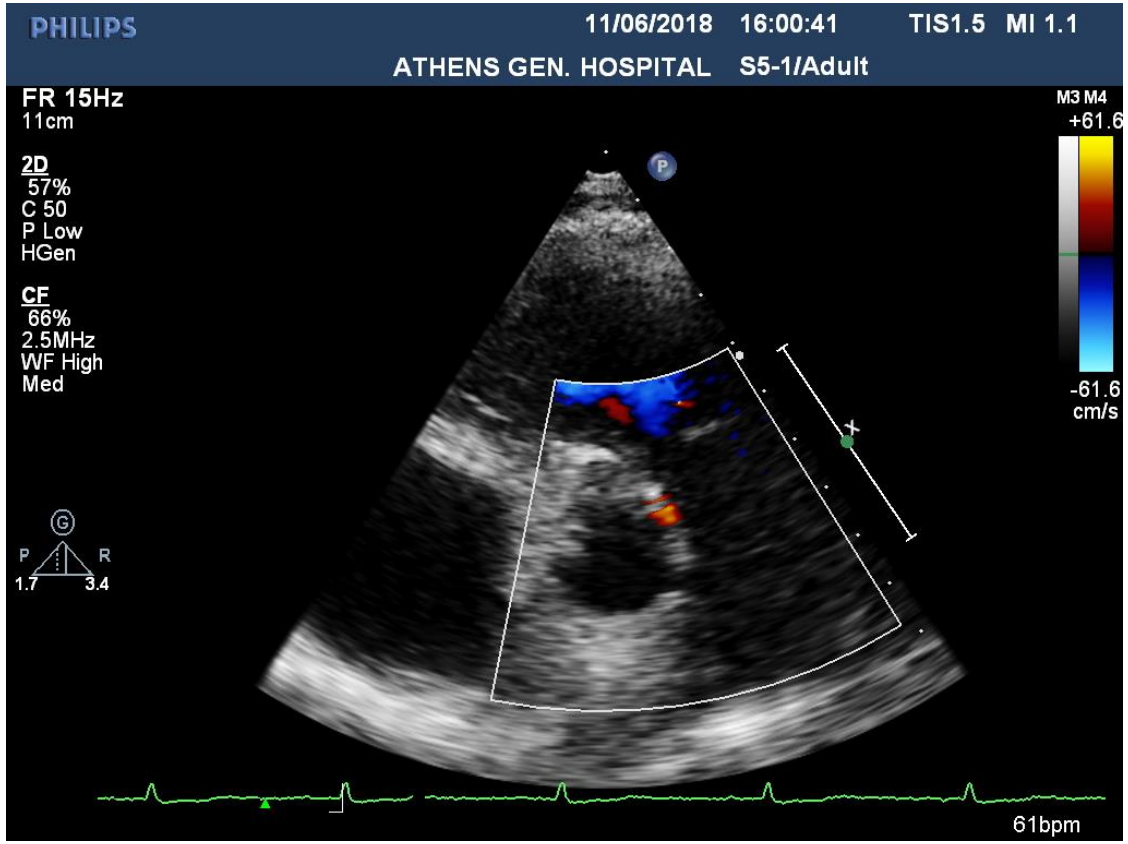
3.4
57bpm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P Low
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CF
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WF High
Med
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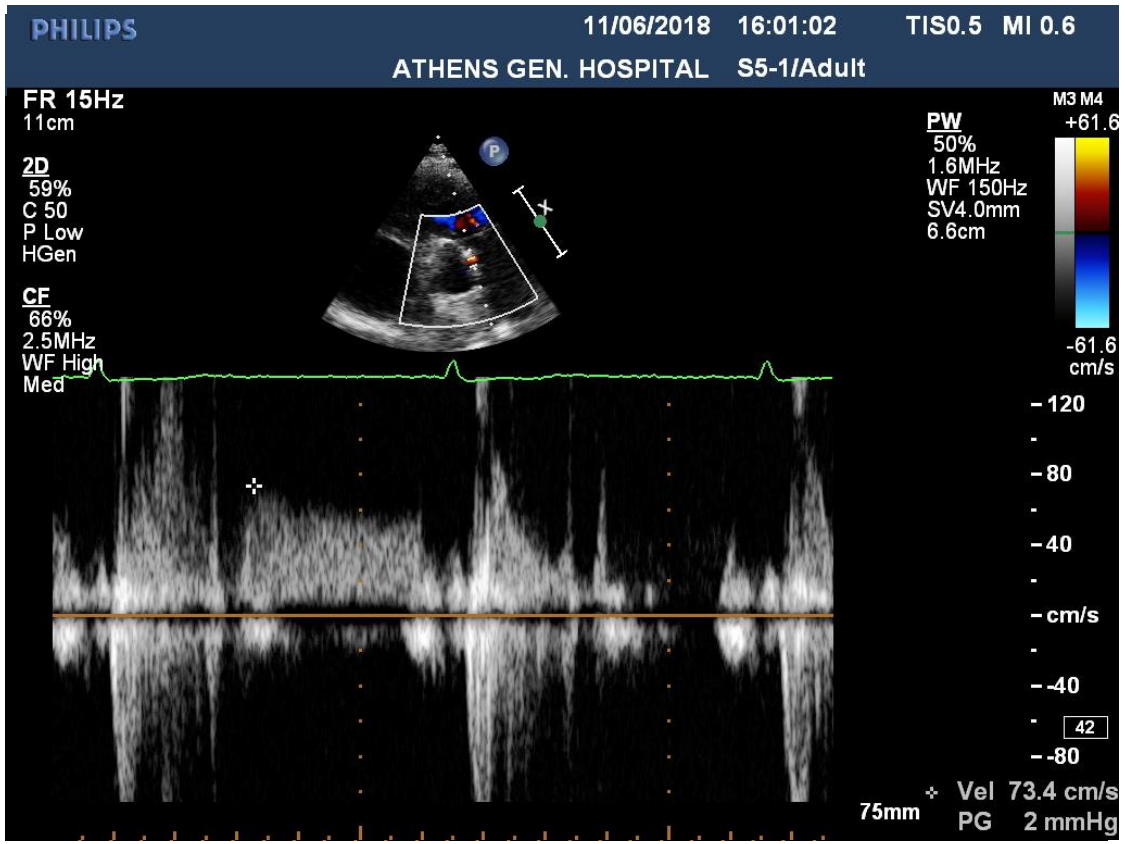
+61.6
-61.6
cm/s
P
R
1.7
3.4
61bpm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2D
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C 50
P Low
HGen
CF
66%
2.5MHz
WF High
Med
PW
50%
1.6MHz
WF 150Hz
SV4.0mm
6.6cm
M3 M4
+61.6
-61.6
cm/s
120
80
40
cm/s
-40
42
-80
75mm
Vel 73.4 cm/s
PG 2 mmHg

Extrinsic compression of the LM stem in PH pts

- Expected likelihood **19% -**

  increasing with **PA diam> 40mm** & **PA/Ao ratio> 1.21**

  and when the angle of the LMA with the left sinus of Valsalva is <30°

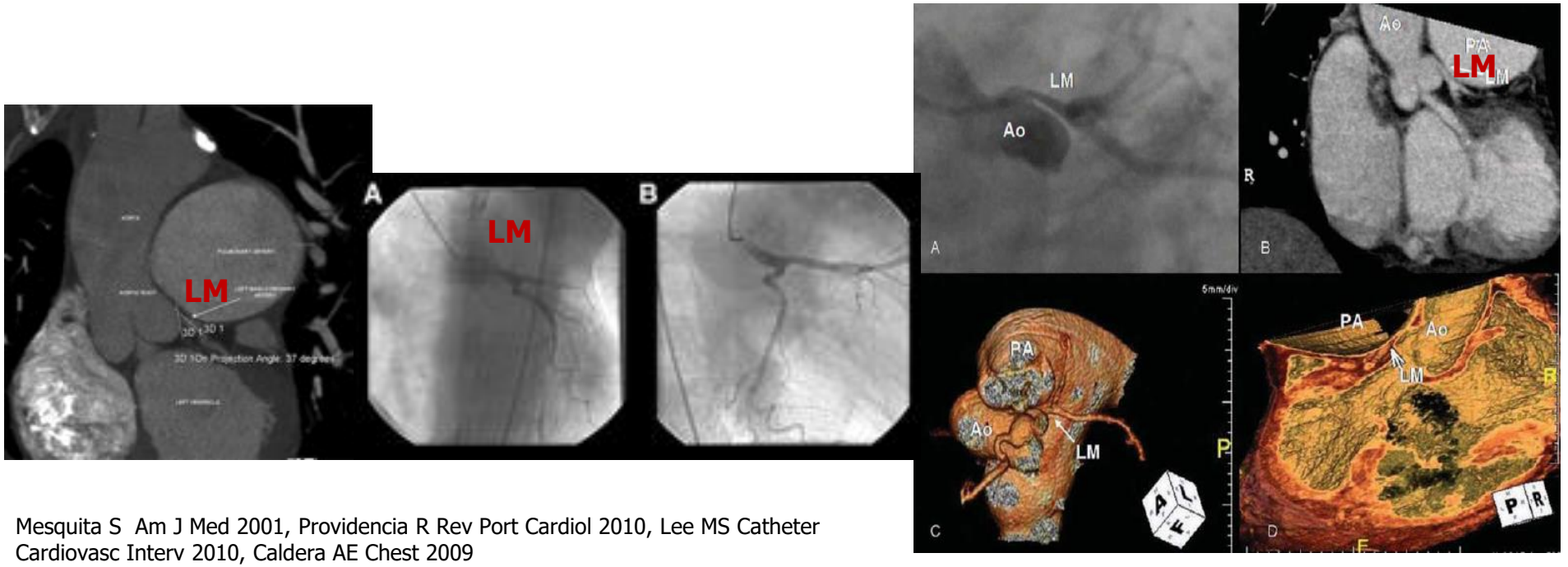
- **50% angina : Coronary Angiography** and/or

  MDCT angiography (or MRI) :

  in asymptomatic pts with high risk anatomy i.e. marked PA dilatation

- **PCI:** feasible, safe & effective treatment option

  (given the high risk of post-op RV failure and mortality with surgical revascularization)



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LEFT MAIN COMPRESSION IN EISENMENGER SYNDROME

Search results: Items: 9

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Ikegami R, Ozaki K, Ozawa T, Hirono S, Ito M, Minamino T.
Intern Med. 2018 May 15;57(10):1421-1424. doi: 10.2169/internalmedicine.9534-17. Epub 2018 Jan 11.
Select item 271435522.

ST-Segment-Elevation Myocardial Infarction Attributable to **Left Main** Coronary Artery **Compression**.
Plácido R, Martins SR, Canas da Silva P, Infante de Oliveira E, Campos P, Almeida AG, Pinto FJ.
Circulation. 2016 May 3;133(18):1828-9. doi: 10.1161/CIRCULATIONAHA.115.021102. No abstract available.

Two rare conditions in an **Eisenmenger** patient: **left main** coronary artery **compression** and Ortner's **syndrome** due to pulmonary artery dilatation.
Andjelkovic K, Kalimanovska-Ostric D, Djukic M, Vukcevic V, Menkovic N, Mehmedbegovic Z, Topalovic M, Tesic M.
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Left main coronary artery **compression** in a young woman with **Eisenmenger syndrome**.
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Extrinsic **compression** of the **left** coronary ostium by the pulmonary trunk: management in a case of **Eisenmenger syndrome**.
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Lee J, Kwon HM, Hong BK, Kim HK, Kwon KW, Kim JY, Lee KJ, Kang TS, Kim DS, Shin YH, Leem JS, Kim HS.
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Korean J Intern Med. 2001 Dec;16(4):265-9.

Total occlusion of left main coronary artery by dilated main pulmonary artery in a patient with severe pulmonary hypertension.

Lee J[1], Kwon HM, Hong BK, Kim HK, Kwon KW, Kim JY, Lee KJ, Kang TS, Kim DS, Shin YH, Leem JS, Kim HS.

Abstract

A 34-year-old woman was admitted to the hospital because of recently aggravated right heart failure without angina for 5 months. When she was 25 years old, patch repair with Polytetrafluoroethylene (PTFE) was performed for the secondum type of atrial septal defect (ASD) with moderate pulmonary hypertension. The chest PA, echocardiography and cardiac catheterization at current admission revealed Eisenmenger syndrome without intracardiac shunt. Chest CT scan with contrast revealed markedly dilated pulmonary trunk, both pulmonary arteries and concave disfigurement of the left side of the ascending aorta suggesting extrinsic compression, as well as total occlusion of the ostium of the left main coronary artery that was retrogradly filled with collateral circulation from the right coronary artery. The coronary angiography showed

normal right coronary artery and the collaterals that come out from the conus branch to the mid-left anterior descending artery (LAD) and that from distal right coronary artery to the left circumflex artery (LCX) and to the distal LAD, respectively.

On aortography, the left main coronary artery was not visualized with no stump, suggestive of total occlusion of the ostium of the left main coronary artery. From our experience, it is possible to say that the occlusion of the ostium of the left main coronary can be induced by the dilated pulmonary artery trunk due to ASD with pulmonary hypertension and that, if the ASD closure was too late, the narrowing or obstruction of the left coronary artery could not be resolved even after operation owing to irreversible pulmonary hypertension

Anomalous origin of the right coronary artery from the left sinus of Valsalva

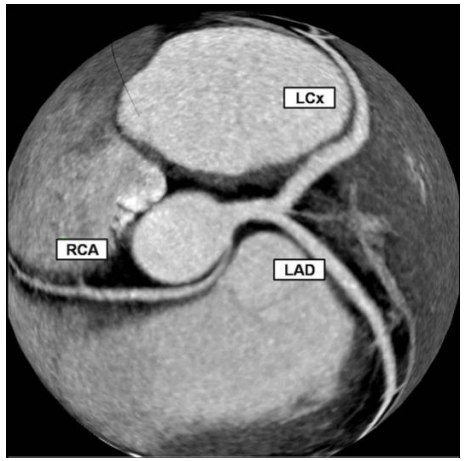
Right ACAOS (anomalous coronary artery from the opposite sinus) is more common and more benign than left ACAOS

The incidence of right ACAOS from coronary angiography is between 0.12% and 0.92%
The prevalence is estimated at 0.1% to 0.3% of the general population

Myocardial iscemia and **sudden death**, however, can be associated with both types of anomalies

Right ACAOS with interarterial course is a group of ACAOS with high risk for developing myocardial ischemia and sudden death (**mechanical compression between PA and Ao**)

Most individuals with ACAOS remain asymptomatic, but when symptoms occur they may include sudden cardiac death, dyspnea, angina pectoris, dizziness, palpitations, and syncope



Setianto BY et al. Case Reports in Cardiology 2016
Lilly SM et al. Proc (Bayl Univ Med Cent) 2011
Berbarie RF et al. Am J Cardiol 2006
Erez E et al. Ann Thorac Surg 2006

Anomalous origin of the right coronary artery from the left sinus of Valsalva

• **Imaging, usually with CT (MDCT) or MRI, is helpful in defining high-risk features:**

- shape and size of the orifice (usually slit-like) (flap-like closure)
- presence of an intramural segment within the aortic wall (mechanical compression)
- acute angle of take-off and kinking of the RCA as it exits the LMCA or while running off from the opposite sinus
- size of myocardium supplied by the anomalous artery

• compression of narrowed segment of coronary artery by aorta or pulmonary artery particularly during strenuous activity
• Possibility of spasm of the anomalous RCA as a result of endothelial injury

• **CTCA used to define the characteristics of coronary anomalies**

• **The presence of ischemic symptoms** may be helpful in defining a high-risk population. Such symptoms are uncommon, particularly in young patients who may present with a single episode of syncope or sudden cardiac death.

• **Stress testing** can also be useful to objectively demonstrate myocardial ischemia if the management strategy remains unclear. However, previous studies have demonstrated that exercise stress testing is unlikely to provide clinical evidence of myocardial ischemia

Lilly SM et al. Proc (Bayl Univ Med Cent) 2011
Yusuke J et al. Tex Heart Inst J 2011Angellini P. Tex Heart Inst J 2002

Anomalous origin of the right coronary artery from the left sinus of Valsalva

- **Treatment options** include: observation, percutaneous intervention (stenting), or surgery:

➢ **Observational approach** would include restriction from further competitive sports.

➢ **Stenting** can be technically difficult, with unclear long-term success rates, and currently is done in relatively few centers.

➢ **Cardiac surgery:**

A) coronary bypass (in older patients or in those whose anomalies are found incidentally, it is more likely that bypass would be considered if obstructive CAD were present; otherwise an observational approach may be advised).

B) corrective procedures have become a preferred strategy, particularly in young patients without obstructive CAD, given the concern of graft longevity over decades and competitive flow in the native coronary arteries that may increase the rate of graft failure .

➢ **Corrective surgical procedures include**:

a) direct implantation of the anomalous artery,

b) unroofing the intramural segment of the vessel within the aortic wall, and/or osteoplasty, which aims to create a new ostium at the end of the intramural segment as the artery branches away from the aorta.

Setianto BY et al. Case Reports in Cardiology 2016
Lilly SM et al. Proc (Bayl Univ Med Cent) 2011

4. **How to proceed next, in view of the anomalous RCA origin and dilated PA?**

a) Immediate complete work-up: MDCT - Coronary CT Angiography, CAA, CPET, 24-hour Holter ECG Monitoring

b) After excluding ischemia and arrhythmias, observation and follow-up CMR- MRA (or MDCT) to check on PA dilatation progression. Re-assess patient accordingly

c) Stenting of RCA. ‘Watch and wait’ for LM compression in the future

d) Corrective surgery

4. **How to proceed next, in view of the anomalous RCA origin and dilated PA?**

a) Immediate complete work-up: MDCT - Coronary CT Angiography, CAA, CPET, 24-hour Holter ECG Monitoring

b) After excluding ischemia and arrhythmias, observation and follow-up CMR- MRA (or MDCT) to check on PA dilatation progression. Re-assess patient accordingly

c) Stenting of RCA. 'Watch and wait' for LM compression in the future

d) Corrective surgery

During follow-up:

- **WHO FC II, no angina or syncope**

- **6MWD:** 630m, **Borg scale dyspnea 0 to 0 / fatigue 0 to 0.5**
  Sat O2: 81% to 74%

- **BNP =** 45 pg/ml

- HcT= 46.8%, MCV 94.7 fL, Fe= 86.9 µg/dl, ferritin 44.7ng/ml, UA: 3.7mg/dl, bil: 2,25mg/dl, Cr: 0.81mg/dL, Cr Cl: 81ml/min, GFR: 83.7 ml/min/1.73m²

- Ambulatory ECG - Holter monitoring: no significant arrhythmias detected
- Has not accepted follow-up RHC
- New CMR- MRA, CPET are due

Ευχαριστώ πολύ για την προσοχή σας!!!