Ebstein's and Tricuspid Valve Dysplasia

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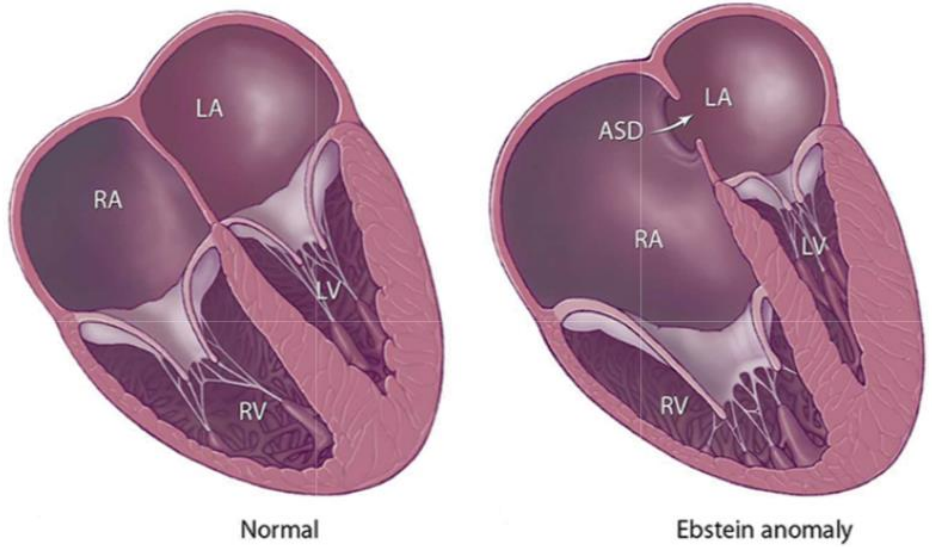
Disclosure

- I have no relevant disclosures

Children's Mercy

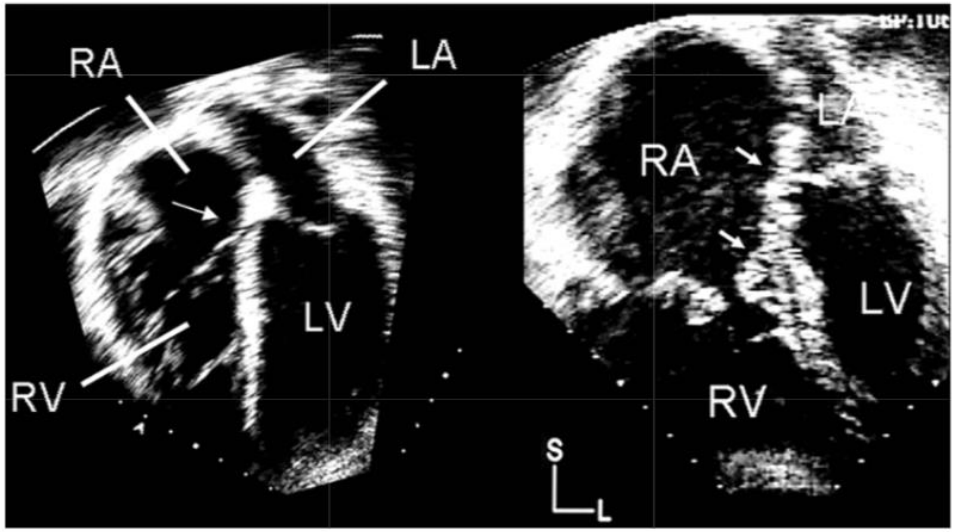
Objectives

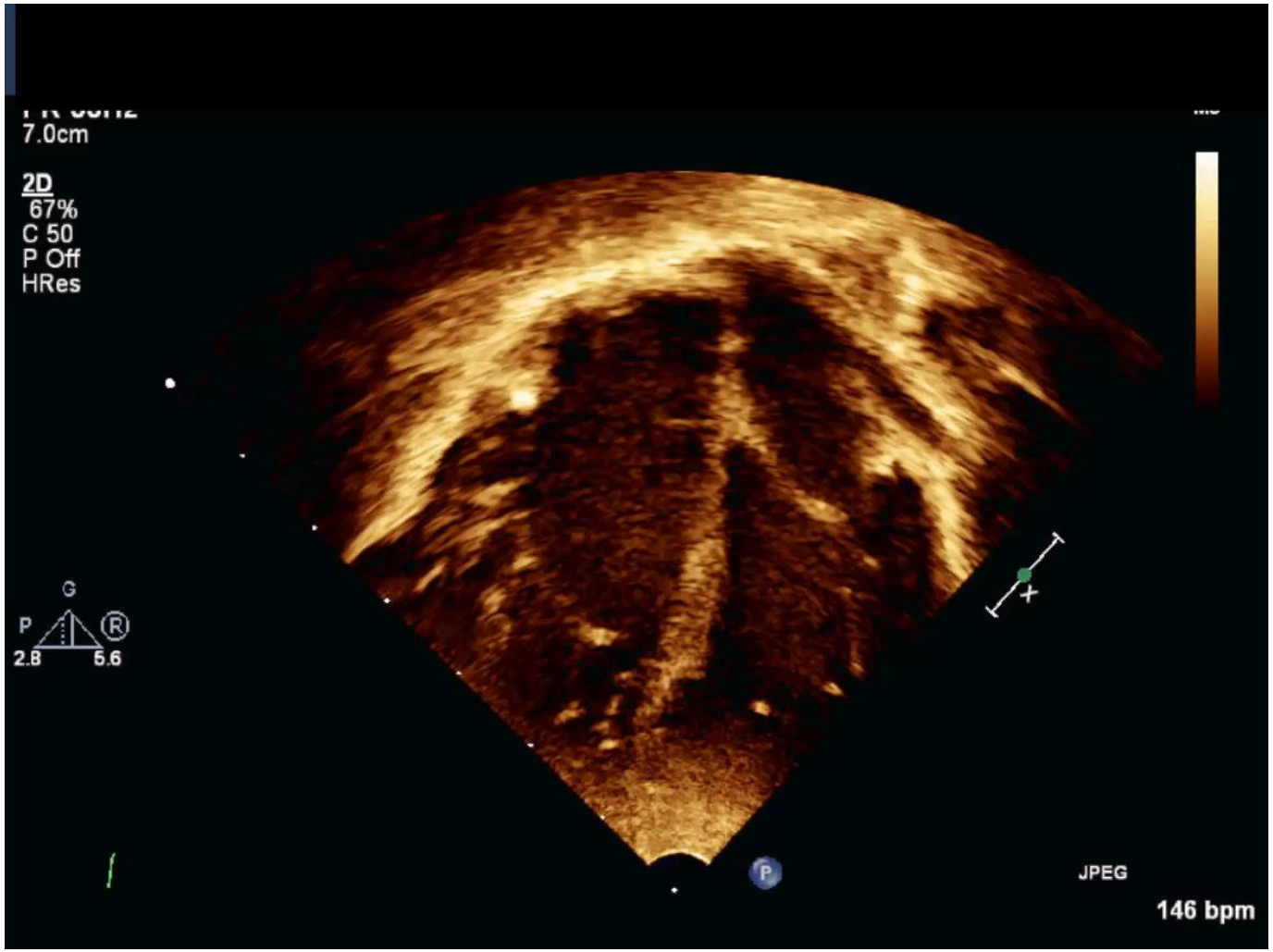
- Anatomy
- Physiology
- Imaging Considerations
- Management
  - Pre/Post natal
- Surgical Implications
- Outcomes



Normal

Ebstein anomaly





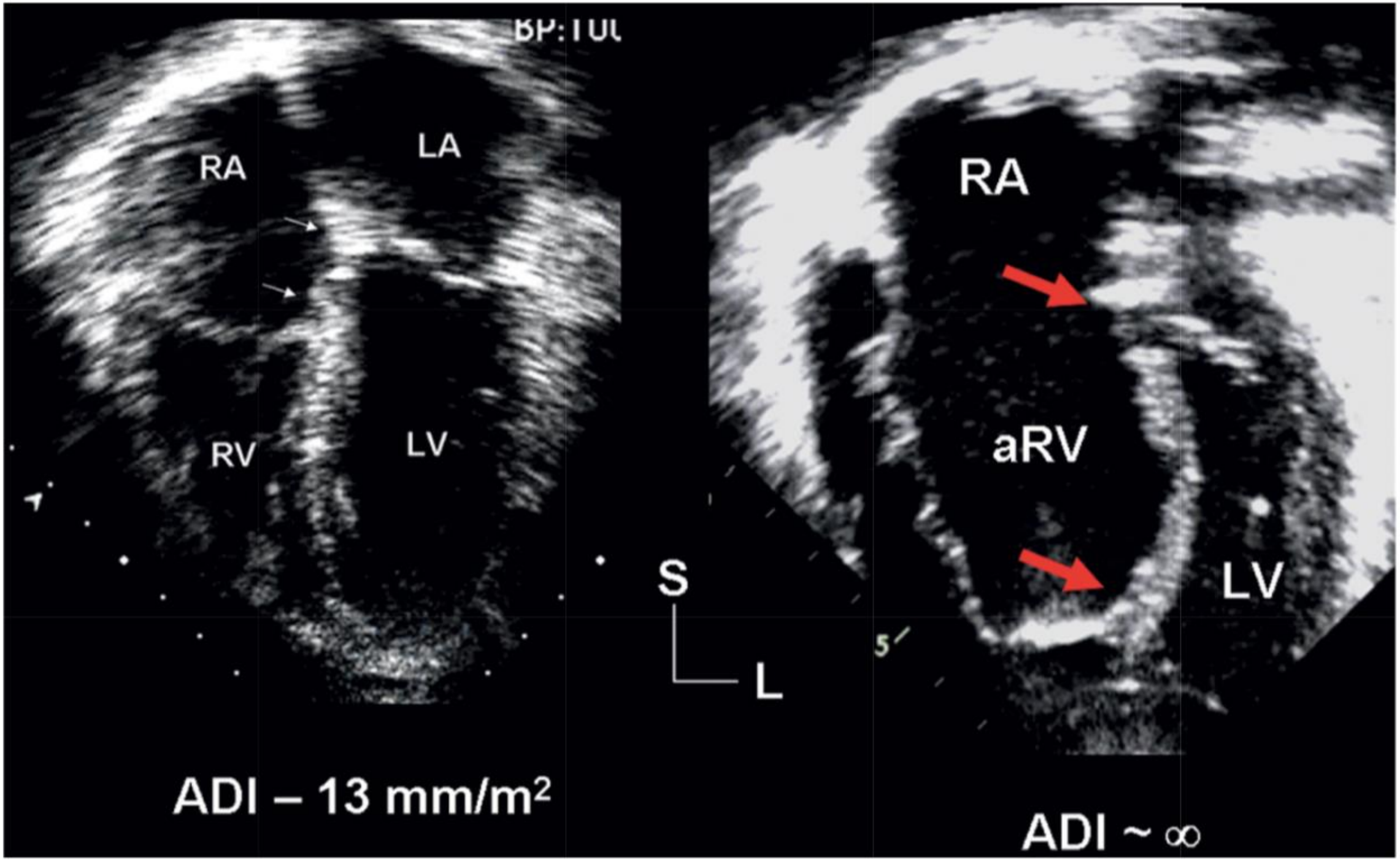
Ebstein's vs Tricuspid Valve Dysplasia

Ebstein's Anomaly

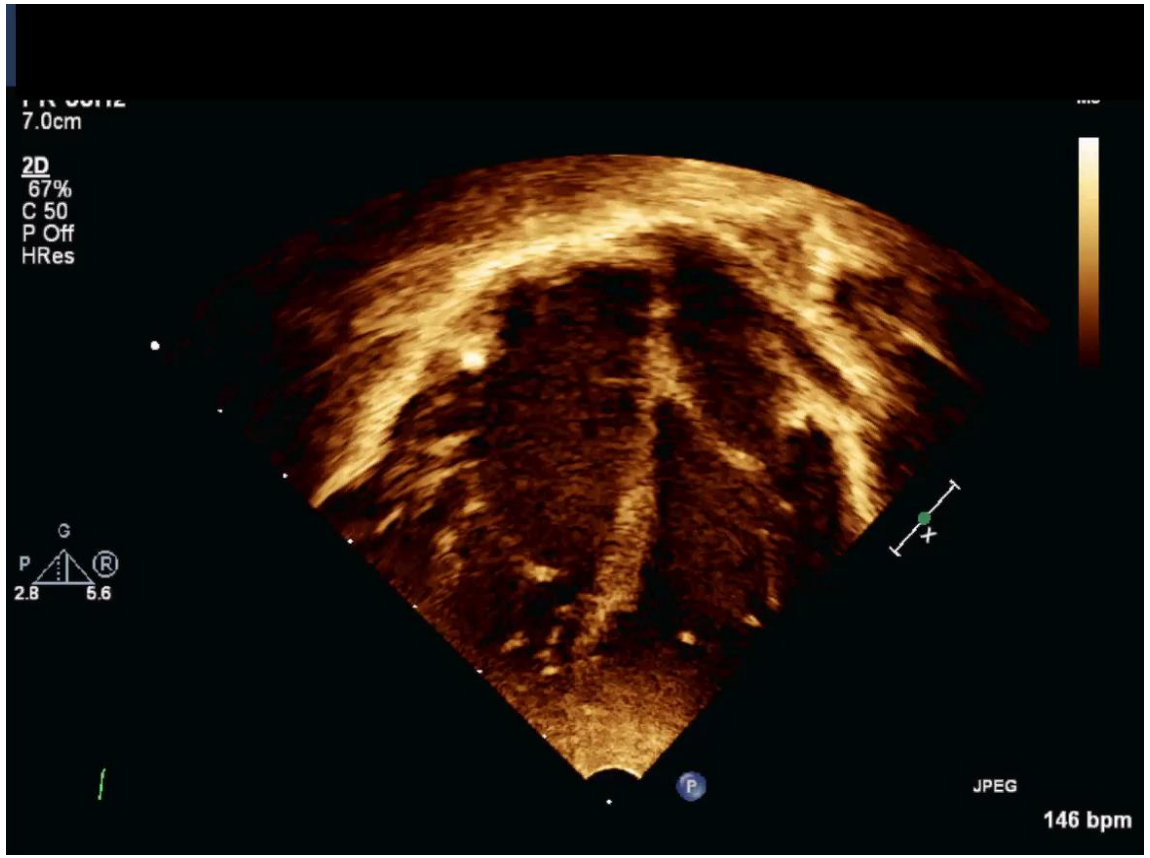
- Inferior/apical displacement of tricuspid valve septal leaflet
  - Incomplete delamination of tricuspid valve leaflets
  - > 8 mm/m^2
- Atrialization of the of the right ventricle
- Malformed leaflets

Tricuspid Valve Dysplasia

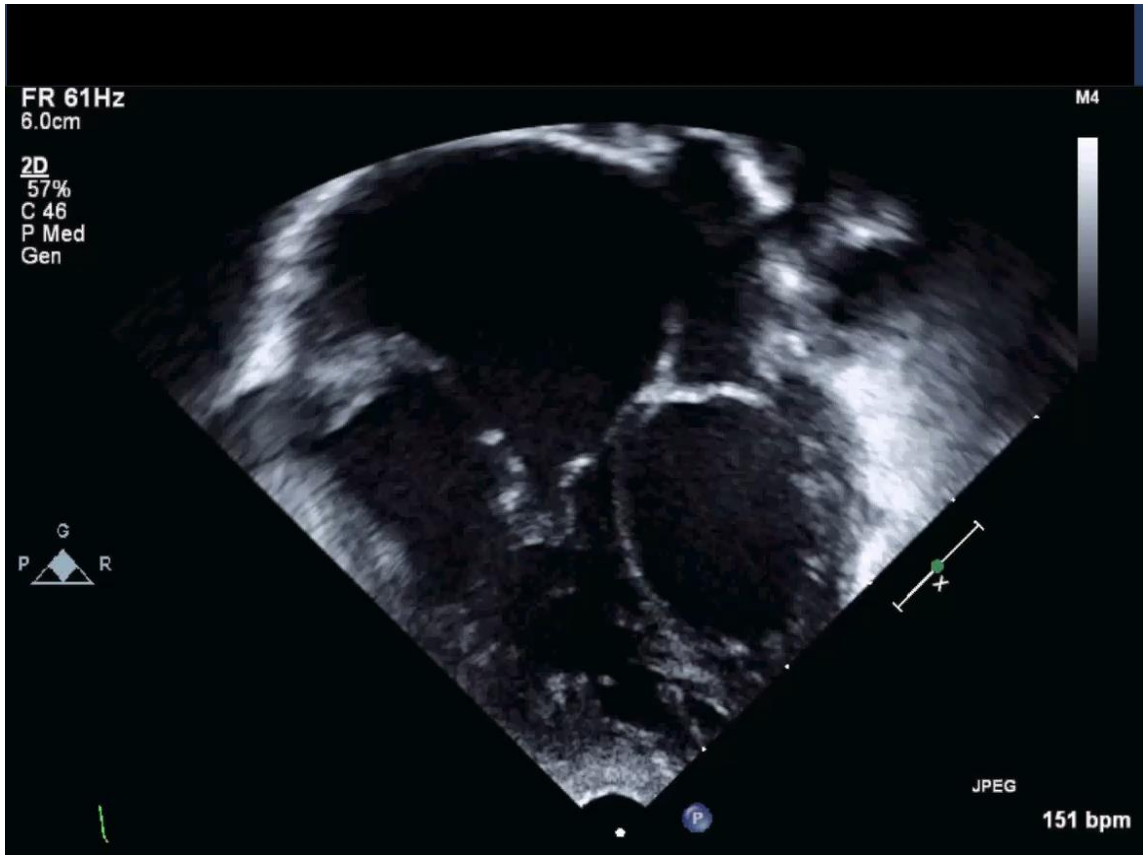
- Tricuspid valve attachments are not displaced but with thickened leaflets
- No atrialization
- Malformed leaflets

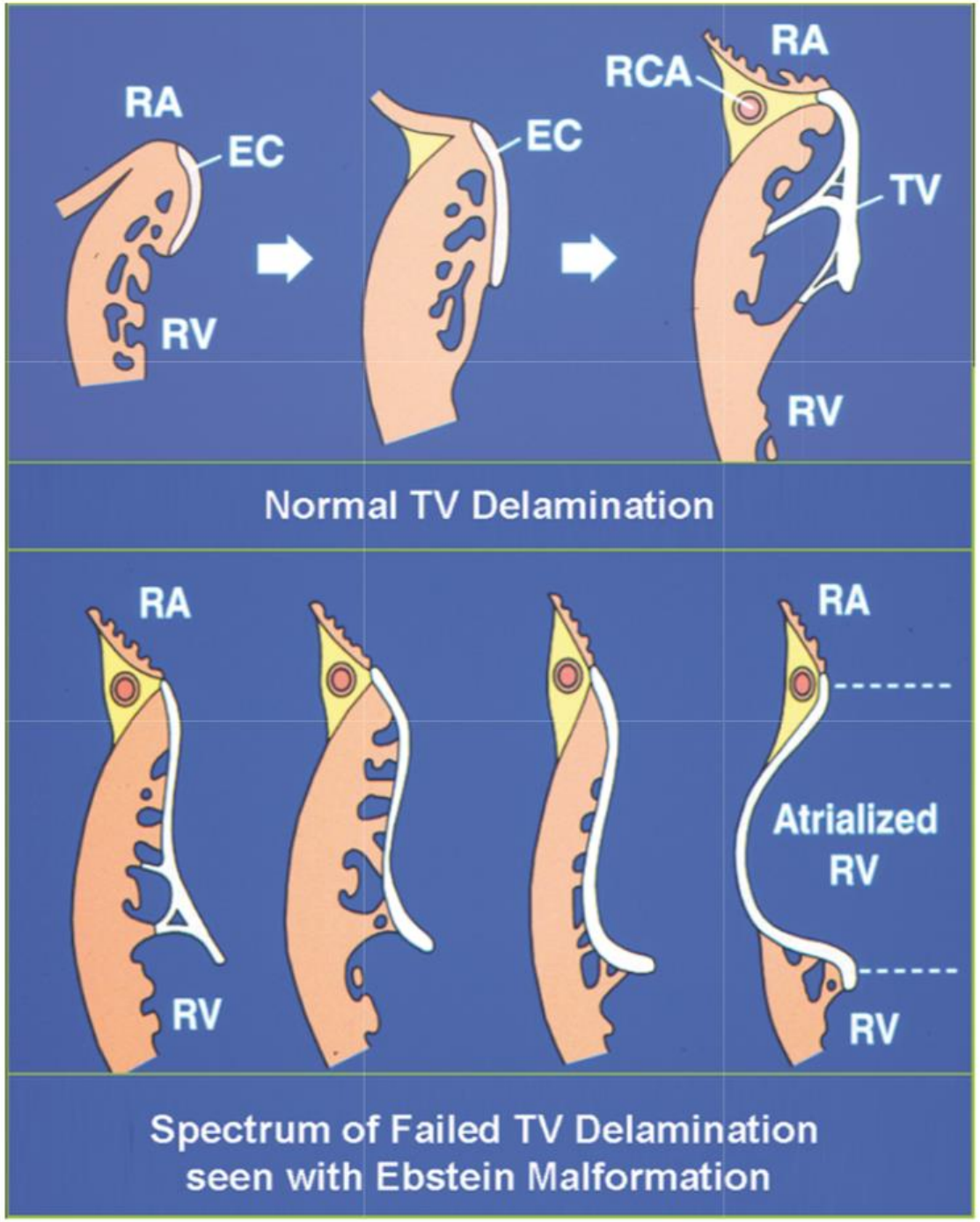

RA
LA
RV
LV
ADI – 13 mm/m²
S
L
RA
aRV
LV
ADI ~ ∞

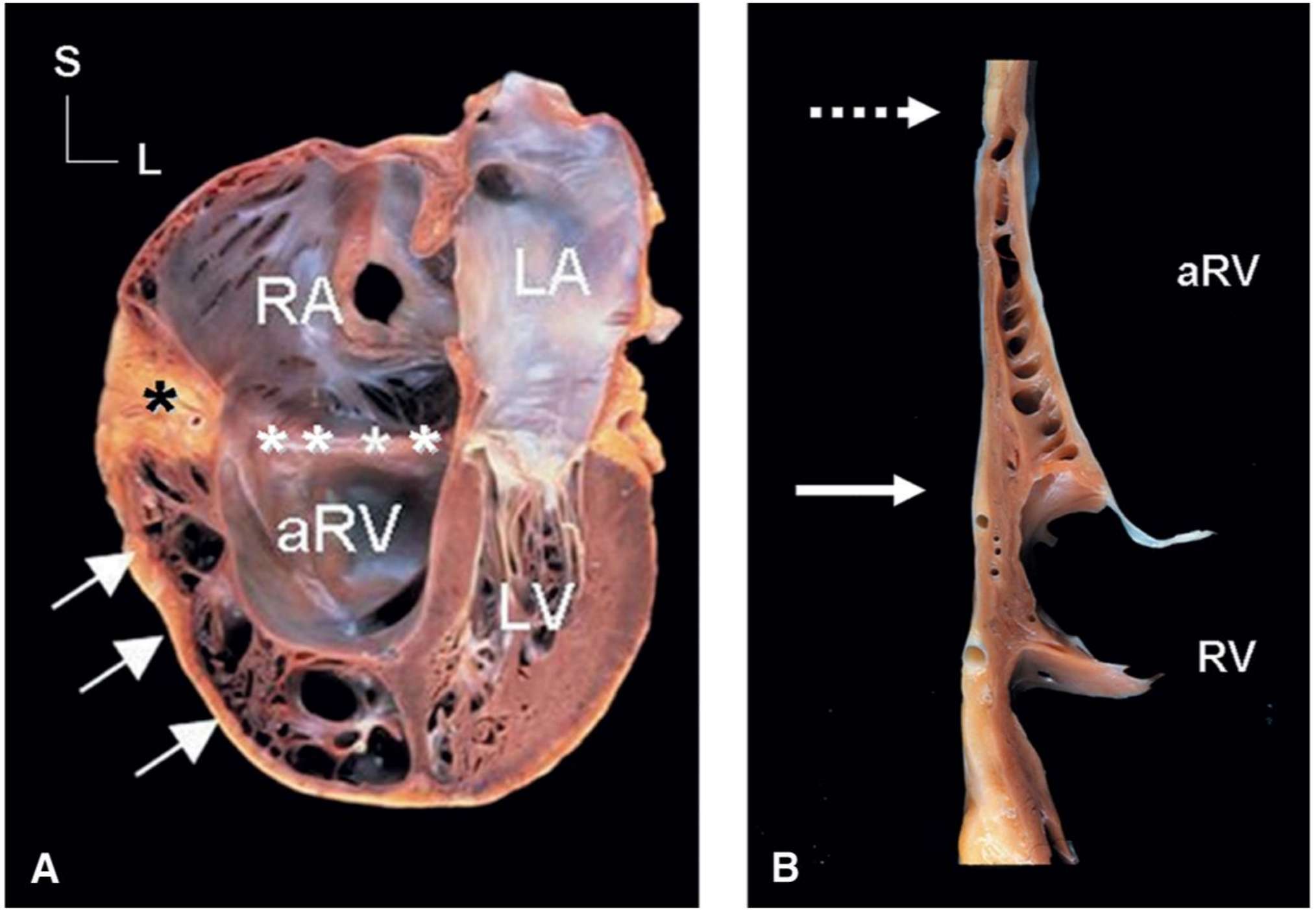
Ebstein's

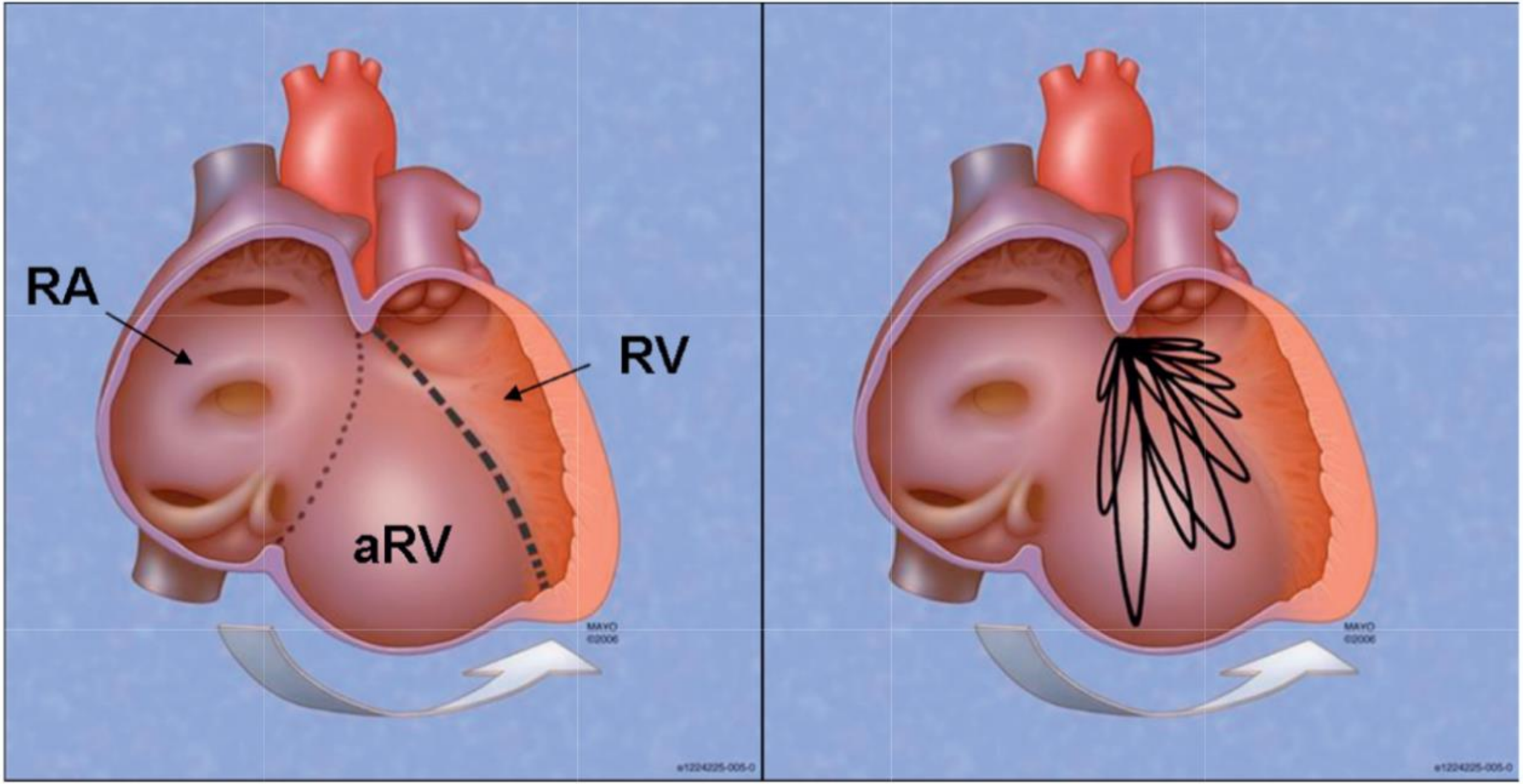


Tricuspid Valve Dysplasia



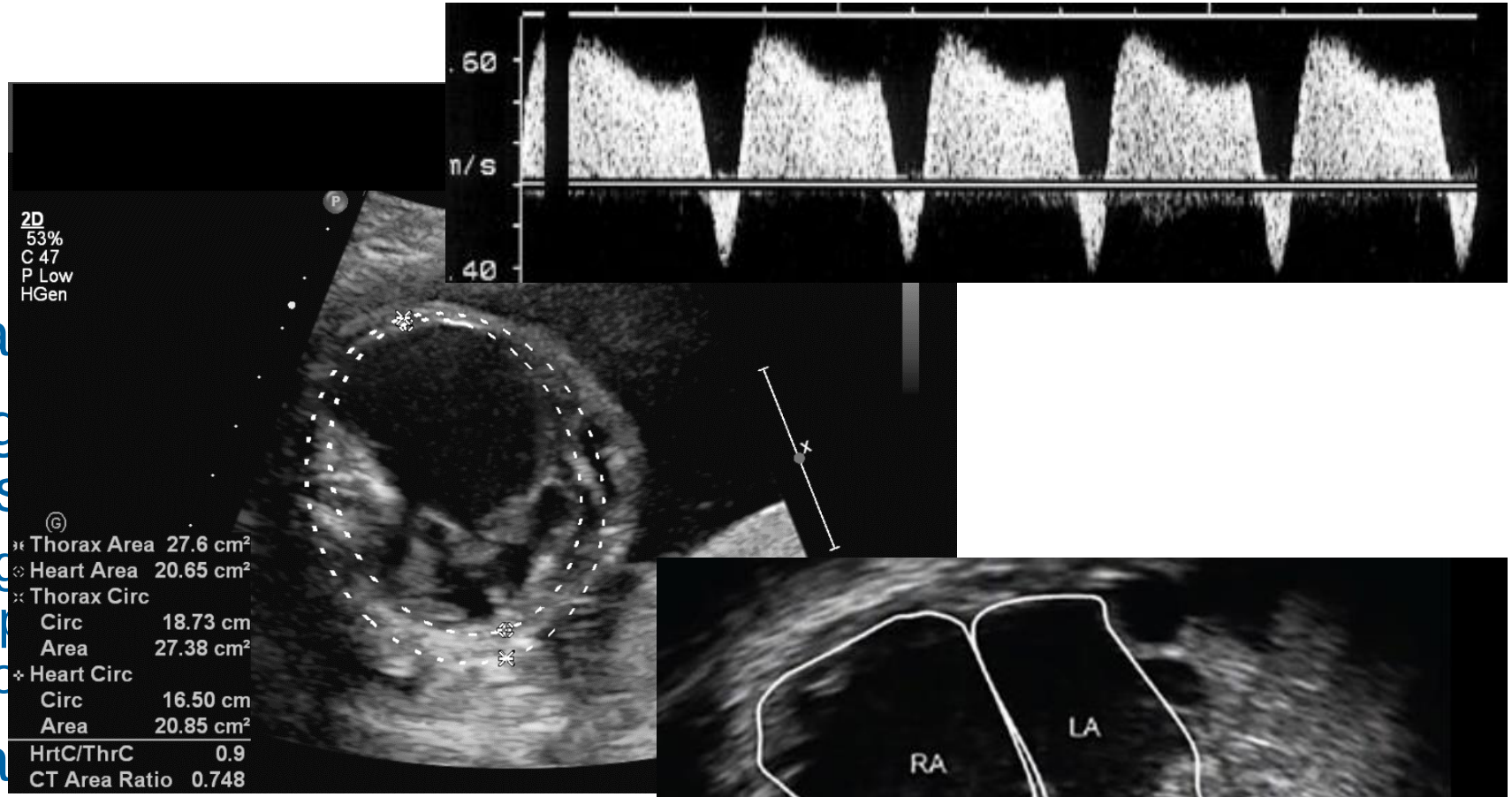

RA
EC
RV
EC
RCA
RA
TV
RV
Normal TV Delamination
RA
RV
RA
Atrialized
RV
RV
Spectrum of Failed TV Delamination
seen with Ebstein Malformation

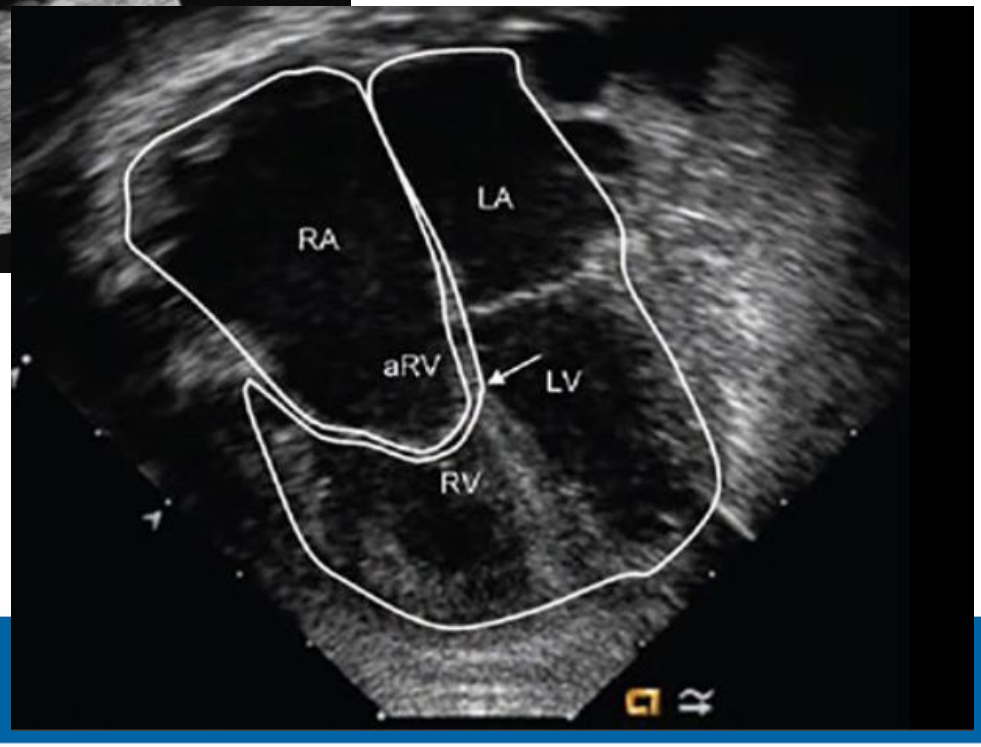

S
L
RA
LA
*
* * * *
aRV
LV
A
aRV
RV
B


RA
RV
aRV
MAYO
©2006
e1224225-005-0
MAYO
©2006
e1224225-005-0

Physiology

- Mild forms can be a
- Severe tricuspid reg
  hydrops, fetal demis
  - Severe tricuspid reg
    atresia and lung hyp
    particularly poor pro
- Cardiothoracic area
  absent pulmonary valve flow, retrograde PDA shunt, RV:LV >1.5 and Celermajer index >1.5 associated with poor prognosis





Imaging considerations

- Effusions/Hydrops?
- Degree/Severity of tricuspid regurgitation?
- Right atrial and ventricular size and function?
- Pulmonary valve patency?
  - Atresia, insufficiency, antegrade flow
- Ductal shunt
- Pulmonary arteries
- LVOT obstruction
- LV function

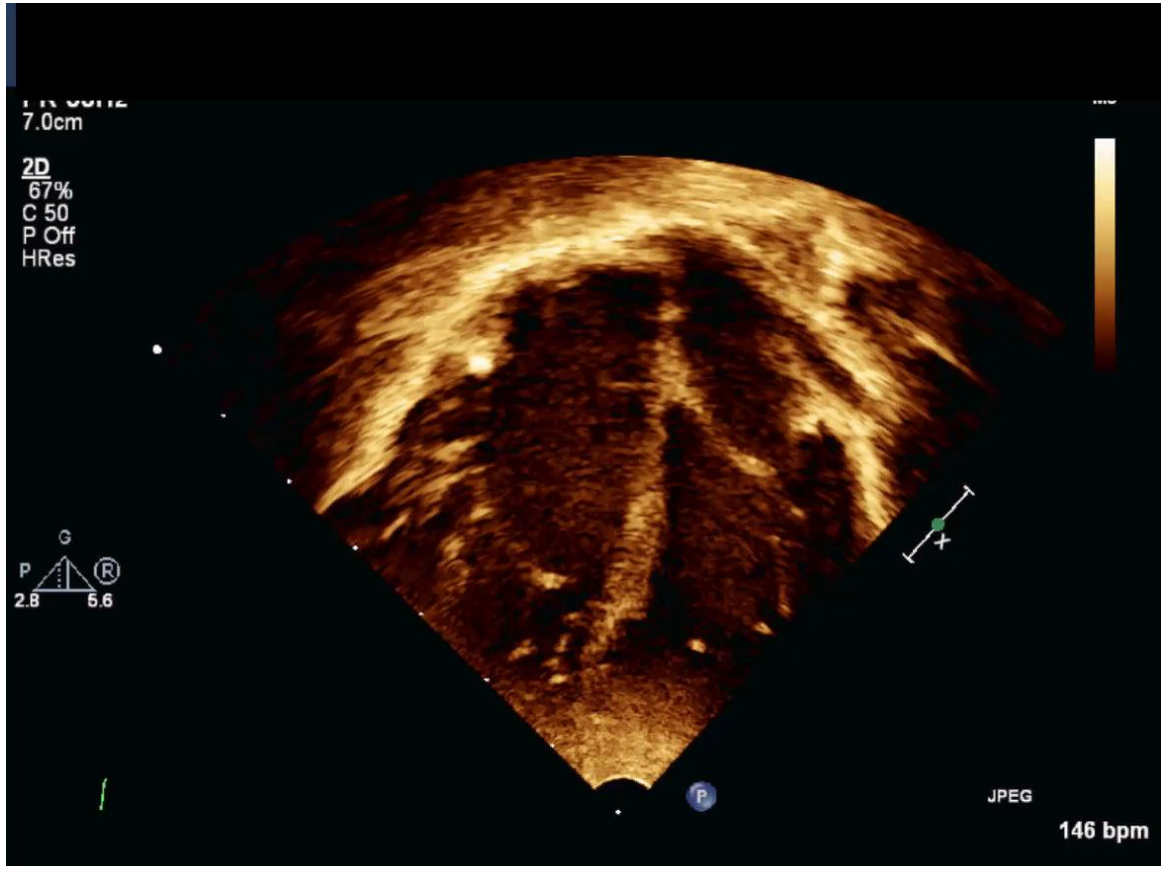


Table 2
Prognostic (SAS) score

	Weighting		
Variable	0	1	2
Cardiothoracic ratio	<0.65	0.65–0.75	>0.75
Celermajer index	<1.0	1.0–1.5	>1.5
Pulmonary valve flow	Normal	Reduced	Absent
Duct flow	Anterograde	Both	Retrograde
Right–left ventricular Ratio	<1.5	1.5–2.0	>2.0

Pulmonary–aortic valve ratio	1.047 ± 0.192	1.074 ± 0.300	0.81
Patent foramen ovale–			
Atrial septal length ratio	0.604 ± 0.141	0.581 ± 0.172	0.74
Celermajer index	1.689 ± 0.513	0.776 ± 0.324	<0.001
Functional tricuspid valve opening–			
Annulus ratio	0.665 ± 0.113	0.607 ± 0.189	0.06
Left ventricluar output (z score)	−0.611 ± 1.908	0.438 ± 1.457	0.15
Tricuspid regurgitant jet (severe)	13	2	<0.001
Ductal flow (retrograde)	10	1	0.003
Pulmonary flow (reduced or absent)	12	4	0.02

Comparison of continuous variables was via the *t* test with unequal variances assumed, categorical variables via Fisher's exact test (with continuity correction). Numbers represent mean ± SD for continuous variables.

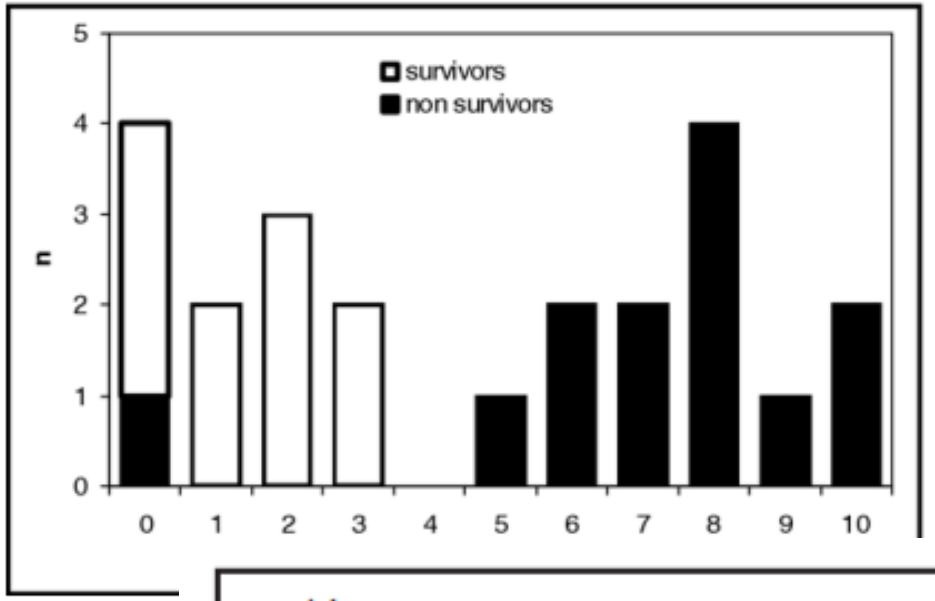


Figure 2. Relati
outcome.

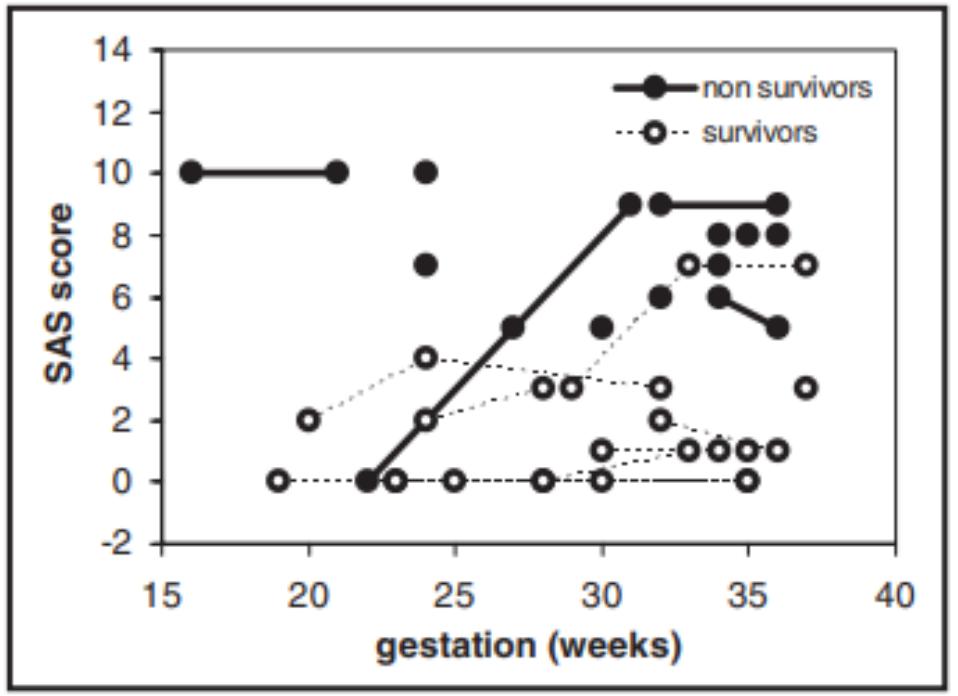


Figure 3. Evolution of the SAS score with gestation.

Improving Outcomes in Fetuses and Neonates With Co
(Ebstein's Malformation) or Dysplasia of the T

Table 1
Predictors of survival and live birth among 33 fetuses with Ebstein's malformation or TV dysplasia

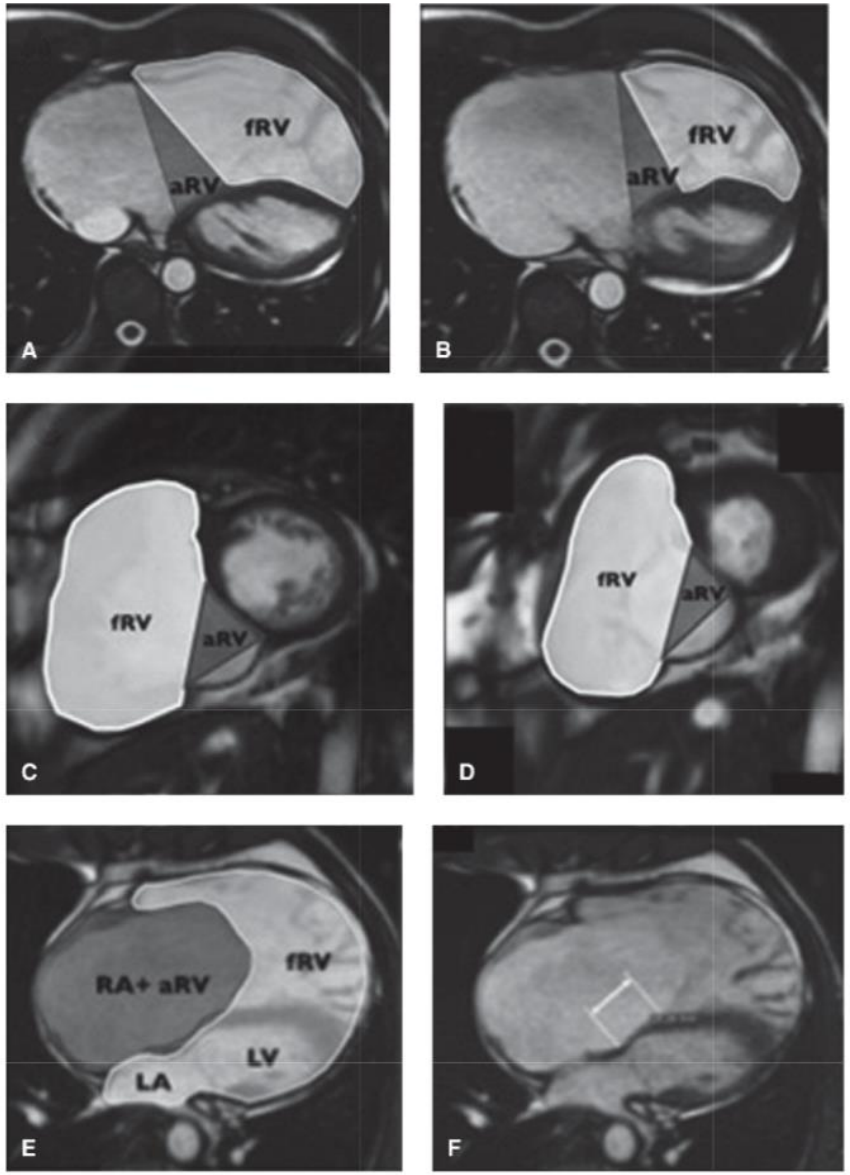
Variable	Deaths (n = 26)*	Survivors (n = 7)	p Value	Not Born Live (n = 17)	Born Live (n = 16)	p Value
Demographic variables						
Gestational age at diagnosis (wks)	24 ± 7	25 ± 7	0.67	20 ± 4	29 ± 6	<0.001
Maternal age at diagnosis (yrs)	31 ± 5	35 ± 7	0.07	31 ± 5	33 ± 6	0.45
Yr of diagnosis						
1984–1996	14 (54%)	1 (14%)	0.09	8 (47%)	7 (44%)	0.85
1997–2004	12 (46%)	6 (86%)		9 (53%)	9 (56%)	
Anatomic and physiologic variables†						
Cardiothoracic area ratio	0.51 ± 0.09	0.35 ± 0.12	0.002	0.52 ± 0.10	0.50 ± 0.14	0.55
RA area index	1.27 ± 0.47	1.02 ± 0.59	0.27	1.13 ± 0.49	1.06 ± 0.64	0.75
RA area index						
>1	21 (81%)	2 (29%)	0.01	12 (74%)	9 (56%)	0.48
<1	5 (19%)	5 (71%)		5 (29%)	7 (44%)	
RA area index						
>0.75	24 (92%)	2 (29%)	0.002	14 (82%)	9 (56%)	0.14
<0.75	2 (8%)	5 (71%)		3 (18%)	7 (44%)	
Severe TR (grade 4)						
Present	26 (100%)	3 (43%)	0.001	17 (100%)	12 (75%)	0.04
Absent	0 (0%)	4 (57%)		0 (0%)	4 (25%)	
TV Z score	7.5 ± 3.3	3.3 ± 3.0	0.01	8.1 ± 2.4	6.4 ± 3.7	0.19
TV Z score						
>3	25 (96%)	2 (29%)	0.001	17 (100%)	13 (81%)	0.10
<3	1 (4%)	5 (71%)		0 (0%)	3 (19%)	
Anterograde flow across the pulmonary valve						
Present	5 (19%)	5 (71%)	0.01	2 (12%)	5 (31%)	0.23
Absent	21 (81%)	2 (29%)		15 (88%)	11 (69%)	
Pulmonary regurgitation						
Present	6 (23%)	2 (29%)	0.76	6 (35%)	4 (25%)	0.71
Absent	20 (77%)	5 (71%)		11 (65%)	12 (75%)	
Fetal hydrops						
Present	10 (38%)	0 (0%)	0.06	7 (41%)	3 (19%)	0.17
Absent	16 (62%)	7 (100%)		10 (59%)	13 (81%)	

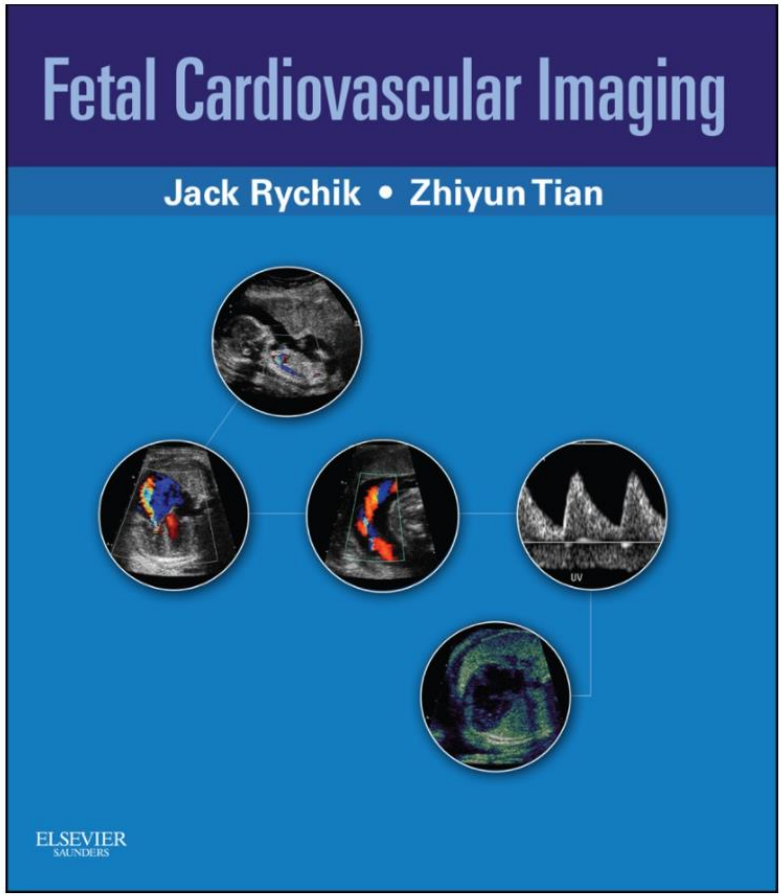
* Elective terminations of pregnancies are included among fetal deaths.
† Comparison between deaths and survivors is based on the most recent fetal echocardiogram, and comparison between live-born and non–live-born patients is based on the first fetal echocardiogram.

Table 2
Predictors of neonatal outcome among 49 live-born patients with Ebstein's malformation or TV dysplasia

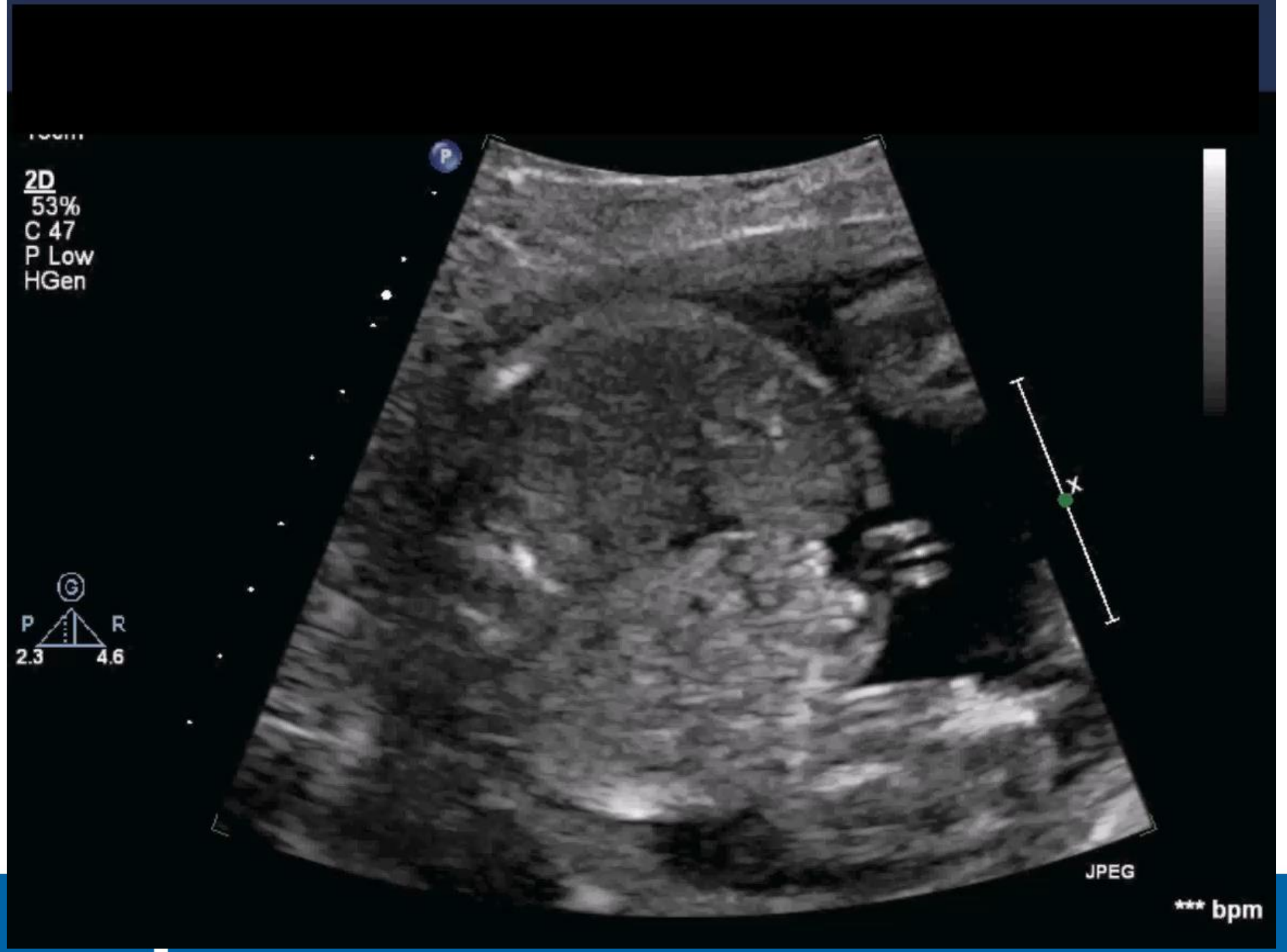
Variable	Deaths (n = 14)	Survivors (n = 35)	p Value
Demographic variables			
Diagnosis			
Prenatal	9 (64%)	7 (20%)	0.003
Postnatal	5 (36%)	28 (80%)	
Yr of diagnosis			
1984–1996	10 (71%)	19 (54%)	0.27
1997–2004	4 (29%)	16 (46%)	
Weight at first neonatal echocardiogram (kg)	2.7 ± 0.4	3.3 ± 0.6	0.006
Anatomic/physiologic variables			
RA area index	1.34 ± 0.43	0.74 ± 0.24	<0.001
RA area index			
>1	12 (86%)	3 (9%)	<0.001
<1	2 (14%)	32 (91%)	
RA area index			
>0.75	14 (100%)	15 (43%)	<0.001
<0.75	0 (0%)	20 (57%)	
Severe TR (grade 4)			
Present	13 (93%)	8 (23%)	<0.001
Absent	1 (7%)	27 (77%)	
TV Z score	3.7 ± 1.9	2.3 ± 1.6	0.04
TV Z score			
>3	11 (79%)	8 (23%)	<0.001
<3	3 (21%)	27 (77%)	
Anterograde flow across the pulmonary valve			
Present	1 (7%)	27 (77%)	<0.001
Absent	13 (93%)	8 (23%)	
Pulmonary regurgitation			
Present	3 (21%)	10 (28%)	0.45
Absent	11 (79%)	25 (62%)	
Neonatal supraventricular/ventricular tachycardia			
Present	1 (7%)	5 (14%)	0.66
Absent	13 (93%)	30 (86%)	

LOVE WILL.


fRV
aRV
A
fRV
aRV
B
fRV
aRV
C
fRV
aRV
D
RA+ aRV
fRV
LV
LA
E
F

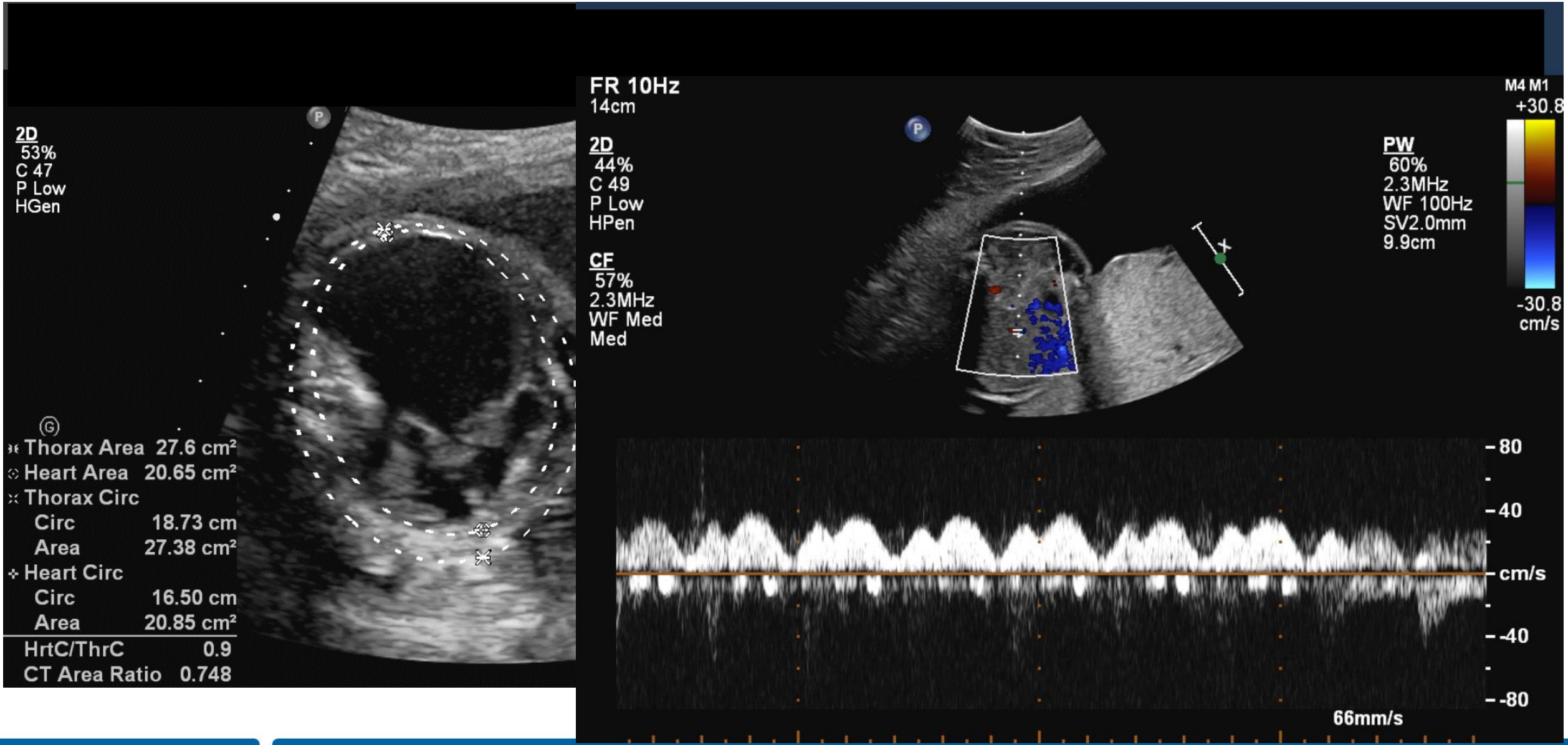

Fetal Cardiovascular Imaging
Jack Rychik • Zhiyun Tian
UV
ELSEVIER
SAUNDERS

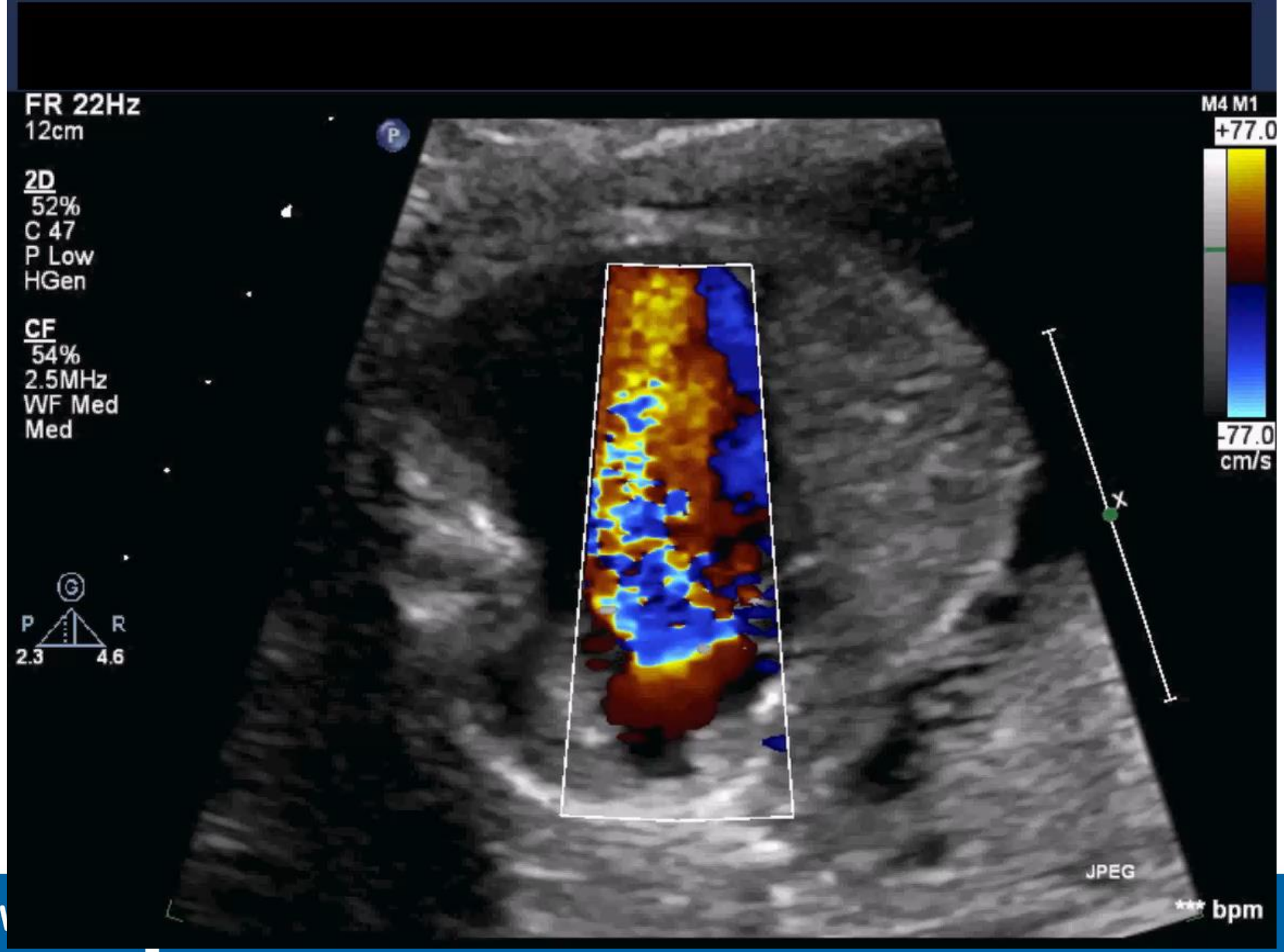

FR 28Hz
16cm
2D
49%
C 47
P Low
HGen
SWEEP
M4
P
G
P
R
2.3
4.6
JPEG
*** bpm

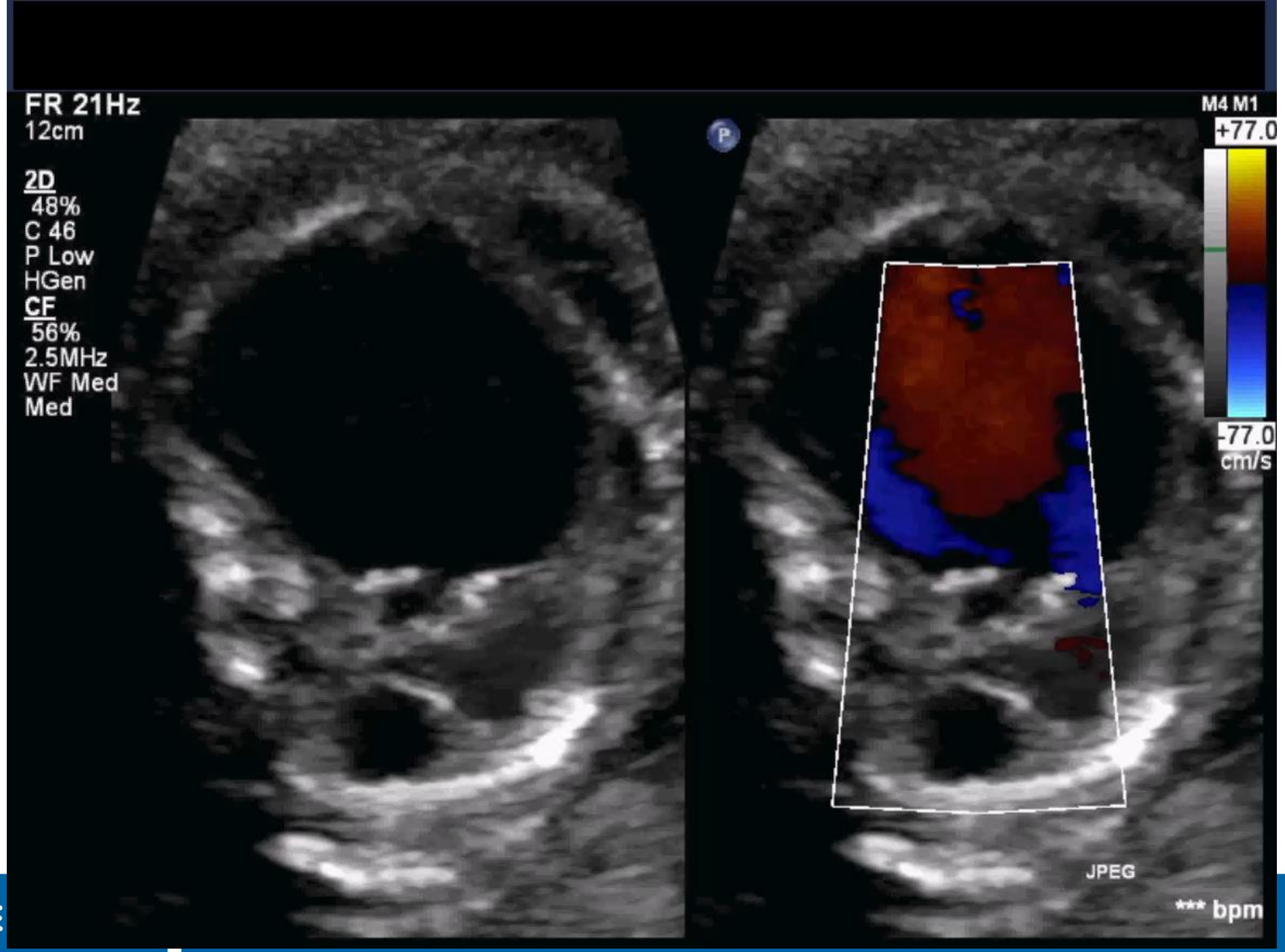

2D
53%
C 47
P Low
HGen
P
R
2.3
4.6
JPEG
*** bpm

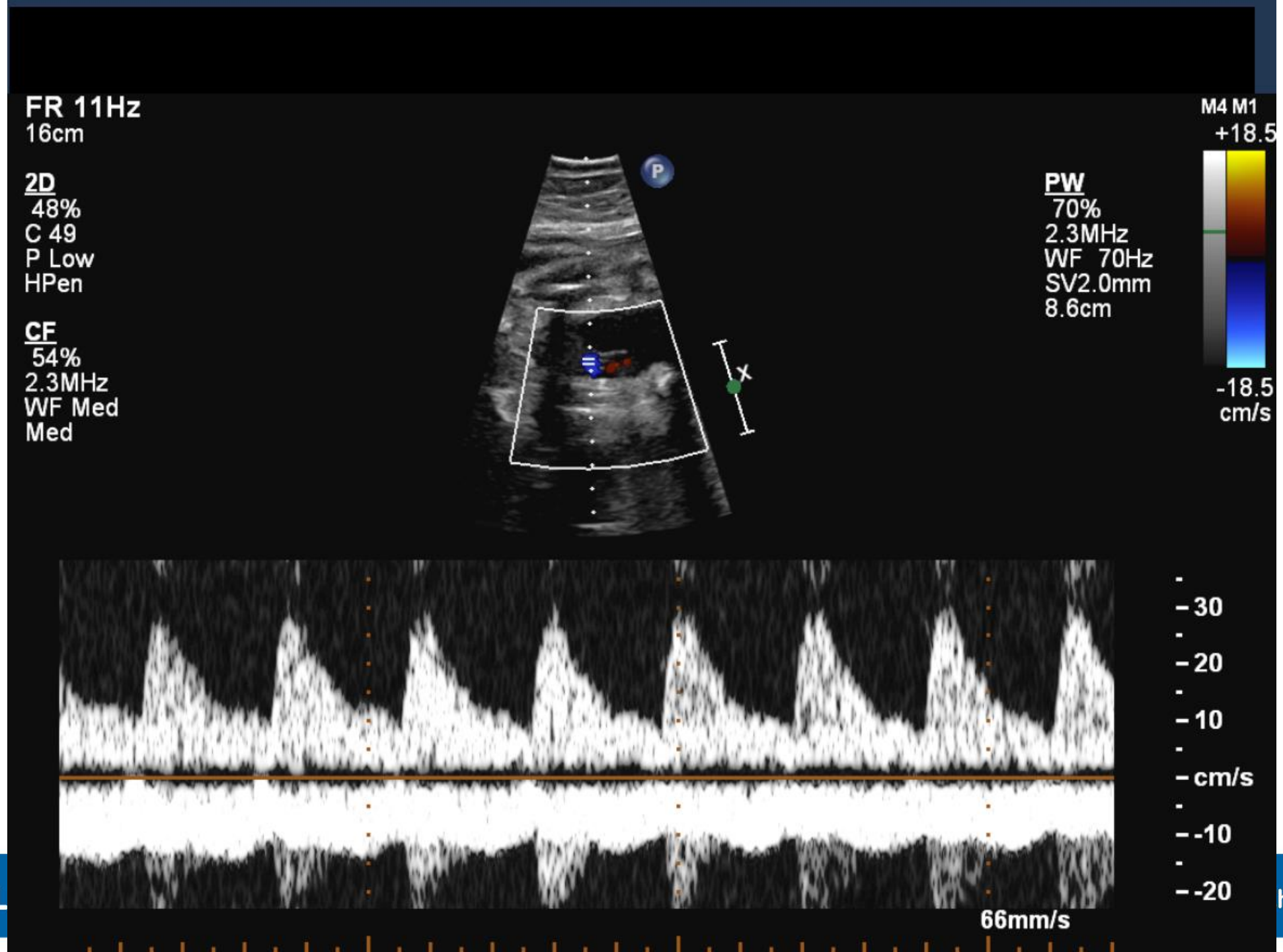

FR 40Hz
15cm
2D
49%
C 50
P Low
HPen
M4
G
P
R
1.8
3.6
JPEG
*** bpm


FR 47Hz
12cm
2D
51%
C 47
P Low
HGen
M4
P
X
G
P
R
2.3
4.6
JPEG
*** bpm

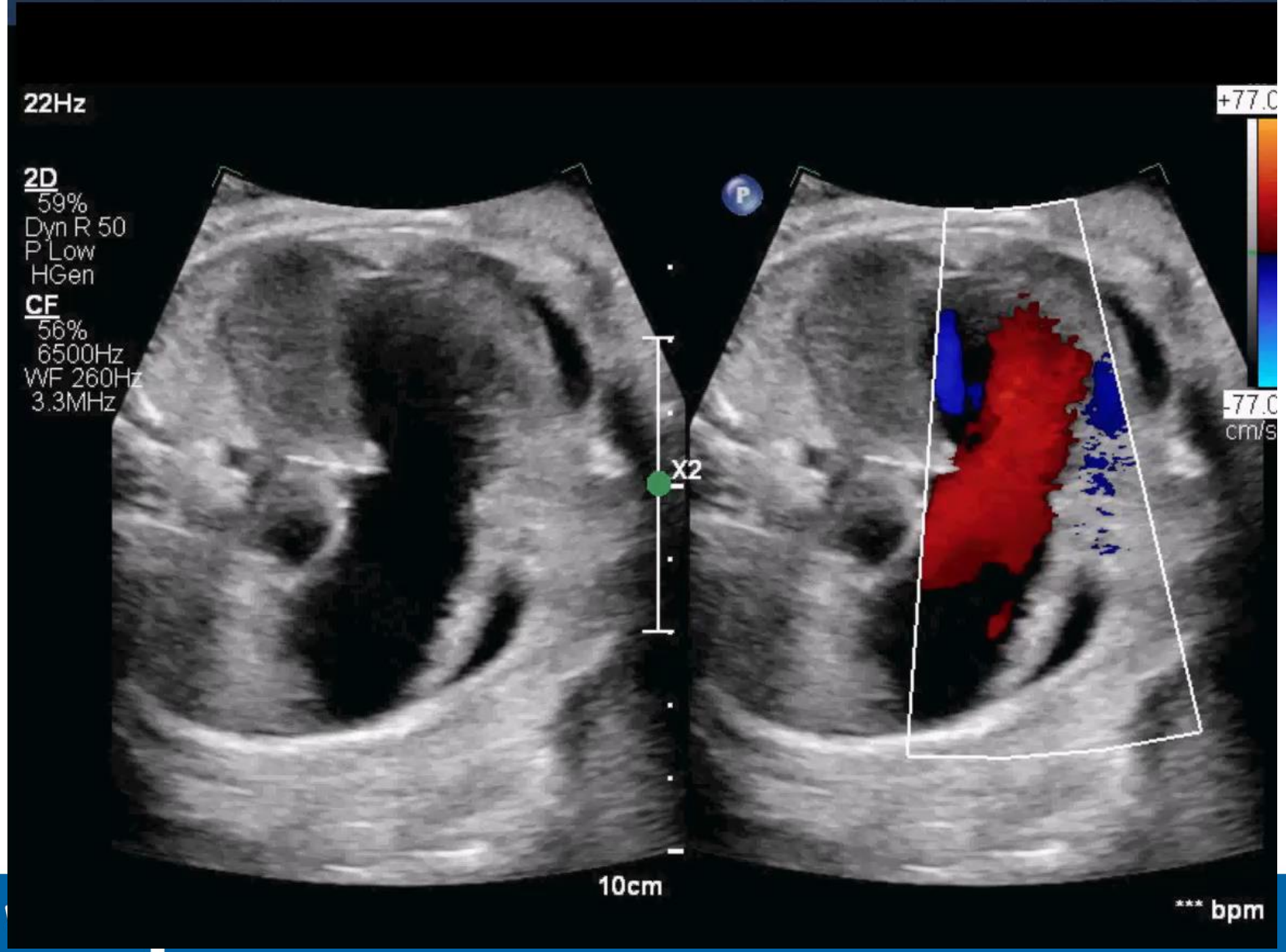

2D
53%
C 47
P Low
HGen
Thorax Area 27.6 cm²
Heart Area 20.65 cm²
Thorax Circ
Circ 18.73 cm
Area 27.38 cm²
Heart Circ
Circ 16.50 cm
Area 20.85 cm²
HrtC/ThrC 0.9
CT Area Ratio 0.748
FR 10Hz
14cm
2D
44%
C 49
P Low
HPen
CF
57%
2.3MHz
WF Med
Med
PW
60%
2.3MHz
WF 100Hz
SV2.0mm
9.9cm
M4 M1
+30.8
-30.8
cm/s
80
40
cm/s
-40
-80
66mm/s

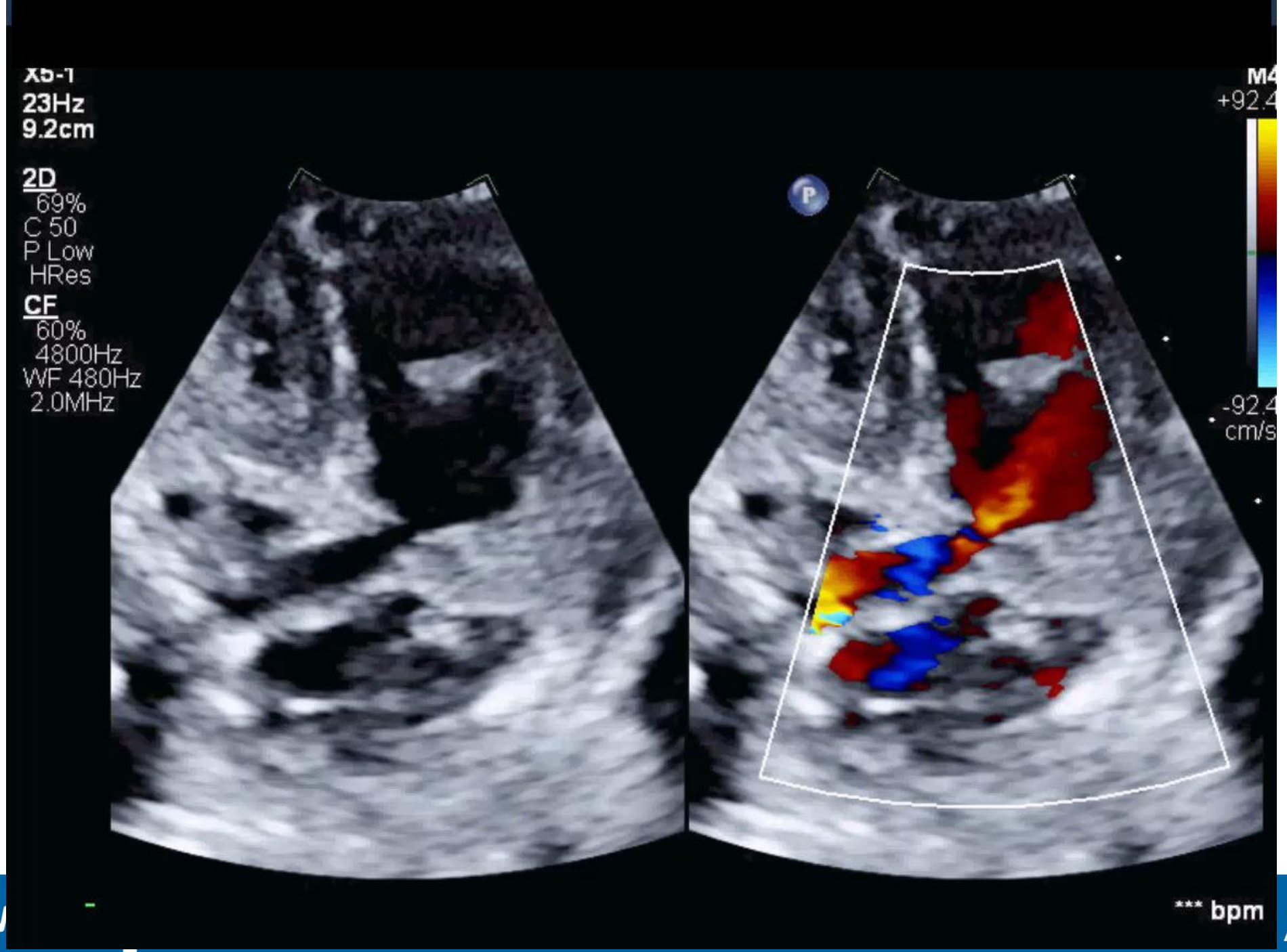

FR 22Hz
12cm
2D
52%
C 47
P Low
HGen
CF
54%
2.5MHz
WF Med
Med
M4 M1
+77.0
-77.0
cm/s
P
R
2.3
4.6
JPEG
*** bpm


FR 21Hz
12cm
2D
48%
C 46
P Low
HGen
CF
56%
2.5MHz
WF Med
Med
M4 M1
+77.0
-77.0
cm/s
JPEG
*** bpm

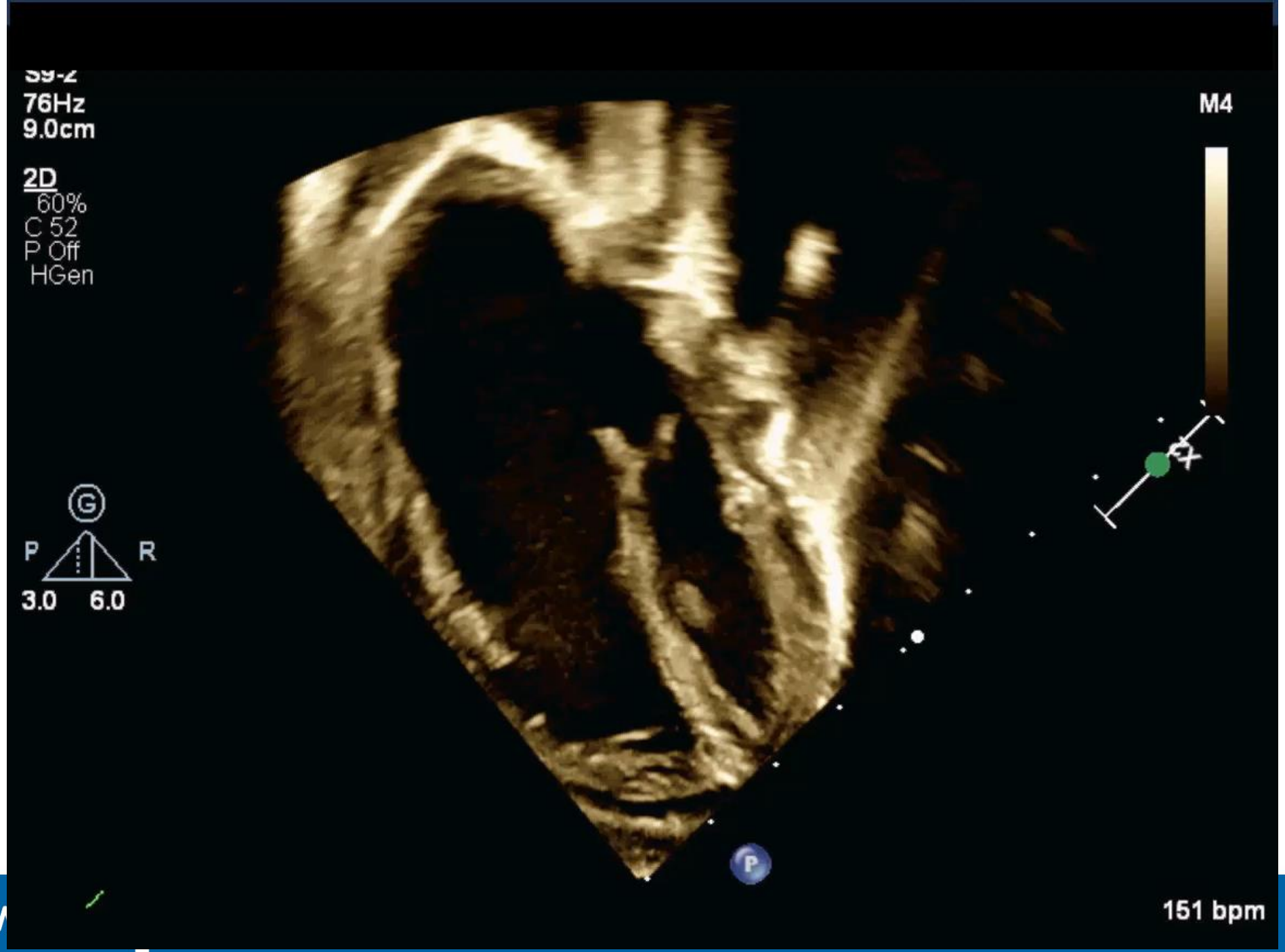

FR 11Hz
16cm
2D
48%
C 49
P Low
HPen
CF
54%
2.3MHz
WF Med
Med
PW
70%
2.3MHz
WF 70Hz
SV2.0mm
8.6cm
M4 M1
+18.5
-18.5
cm/s
30
20
10
cm/s
-10
-20
66mm/s

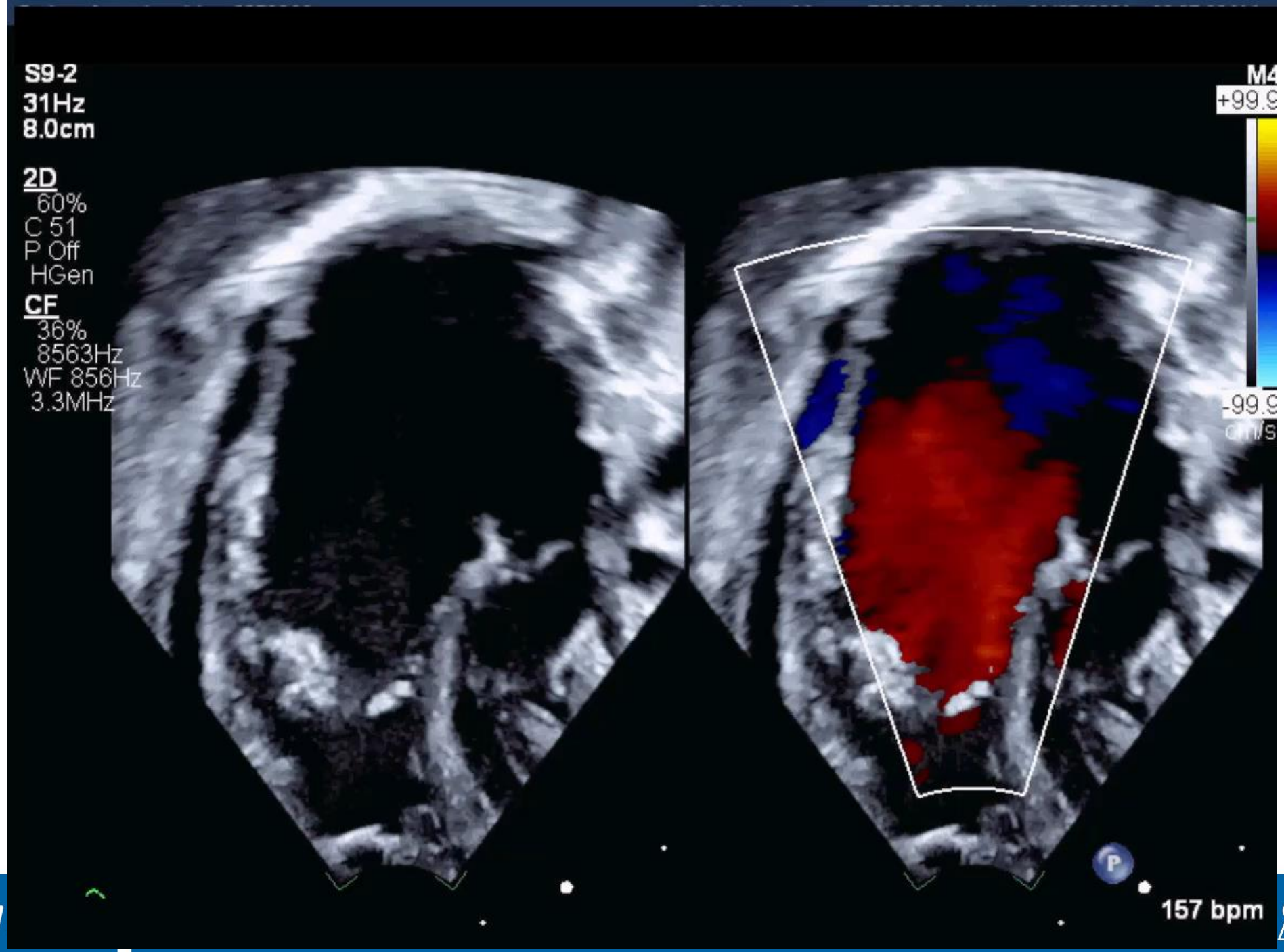

55Hz
RS
2D
47%
Dyn R 50
P Low
HGen
P
R
2.0
4.0
M2
X2
12cm
*** bpm

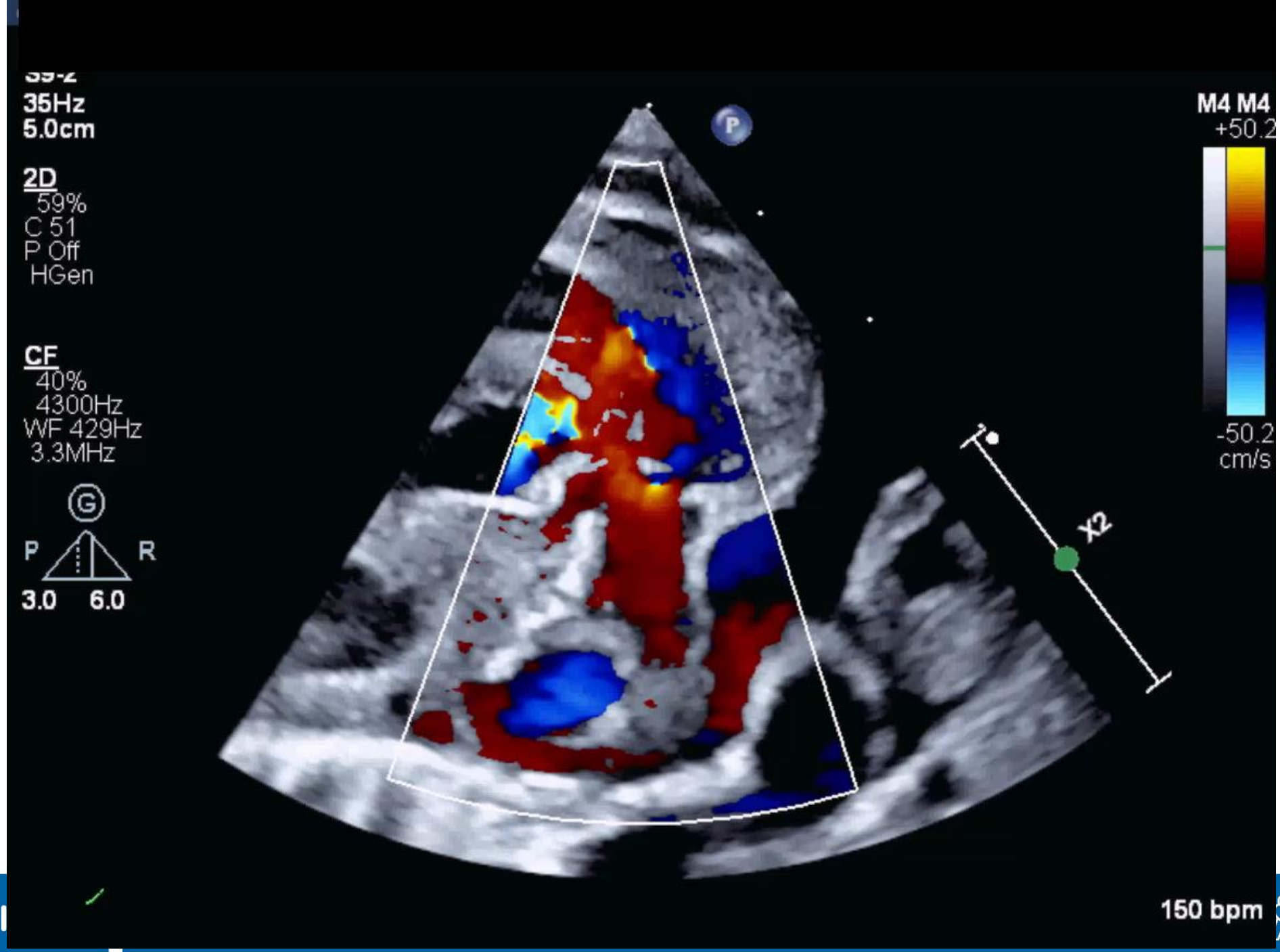

22Hz
2D
59%
Dyn R 50
P Low
HGen
CF
56%
6500Hz
WF 260Hz
3.3MHz
+77.0
-77.0
cm/s
X2
10cm
*** bpm

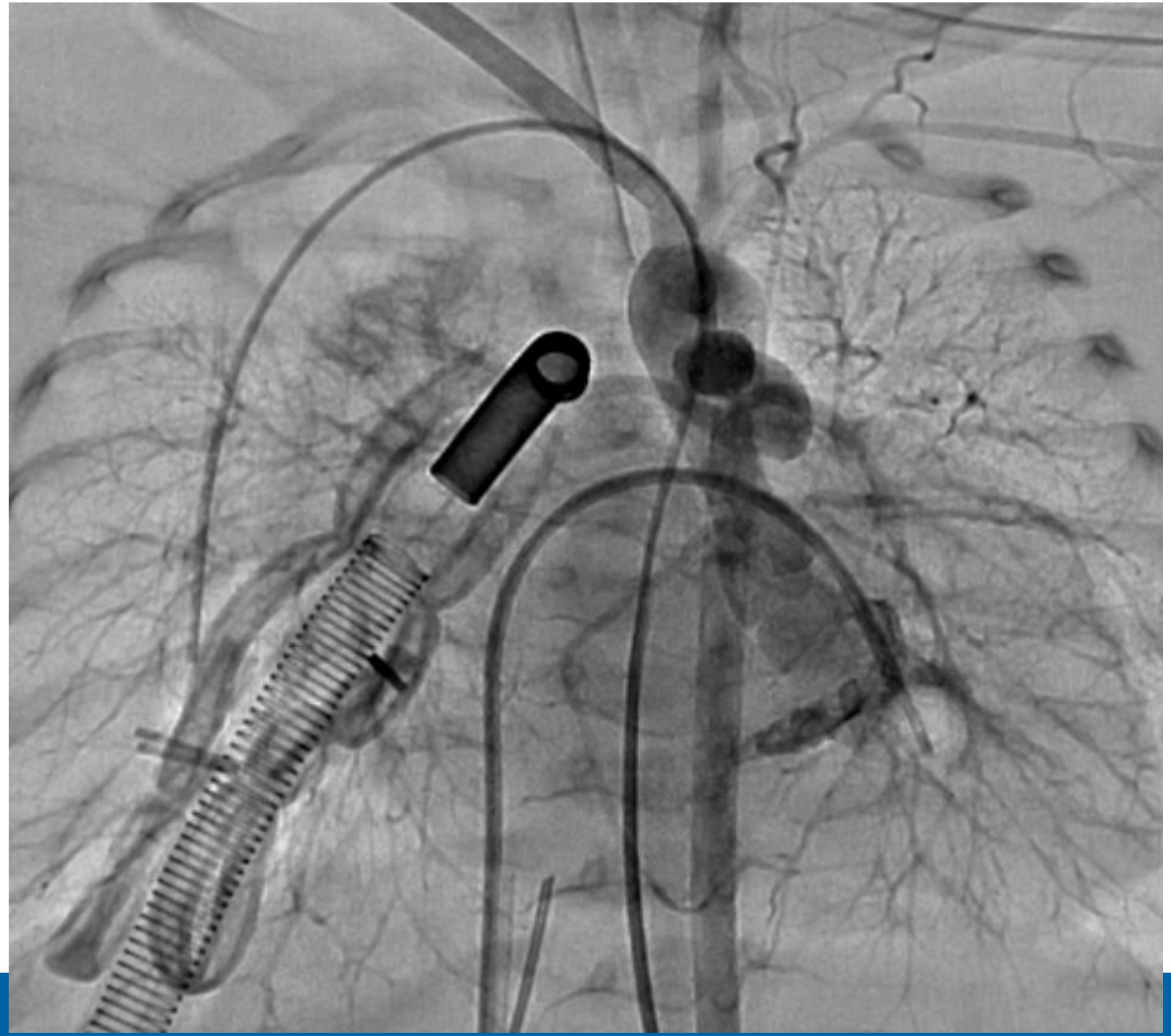

X5-1
23Hz
9.2cm
2D
69%
C 50
P Low
HRes
CF
60%
4800Hz
WF 480Hz
2.0MHz
+92.4
-92.4
cm/s
*** bpm

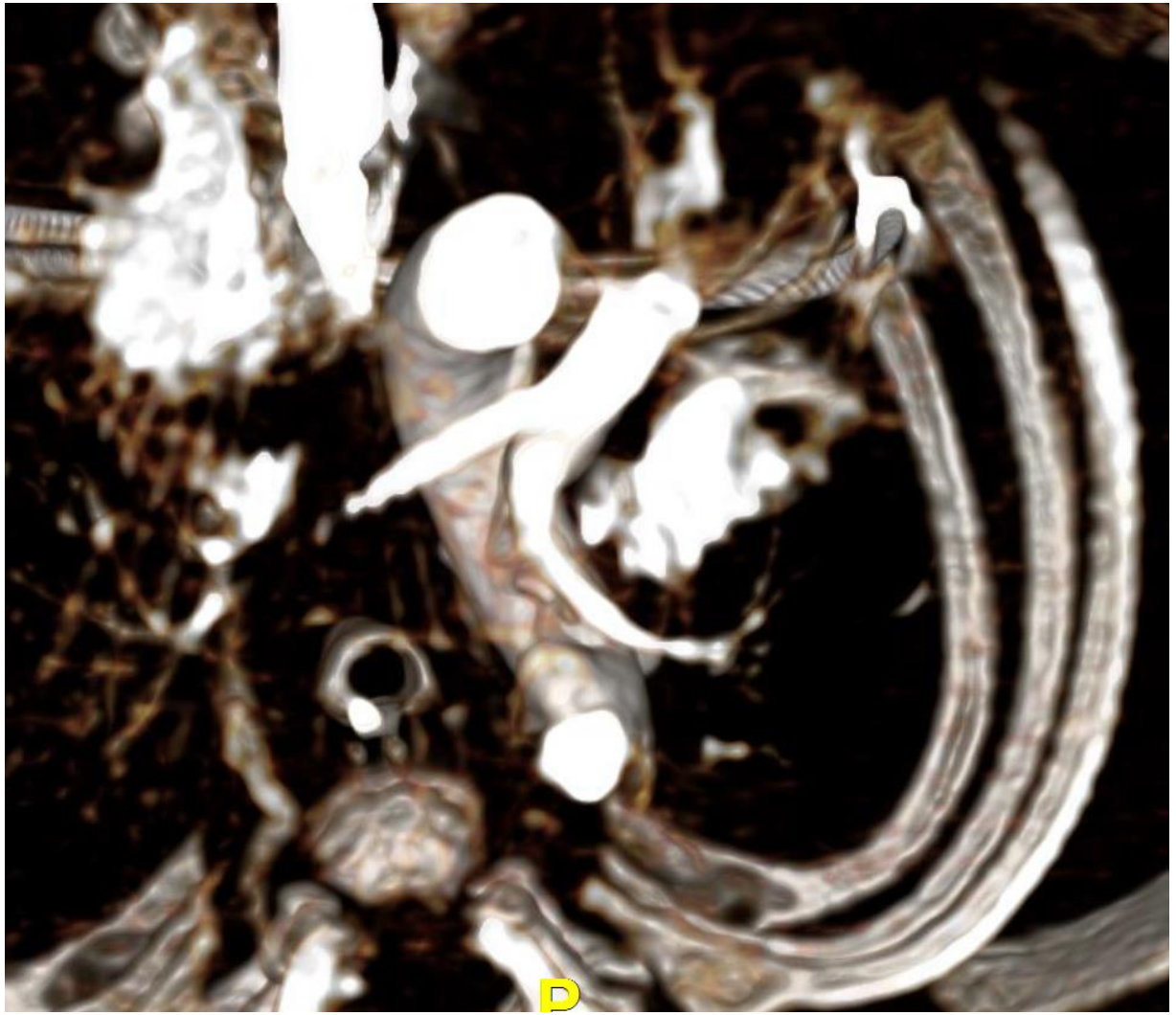

S9-2
92Hz
6.0cm
2D
61%
C 49
P Off
HGen
M4
P
X2
G
P
R
3.0
6.0
147 bpm

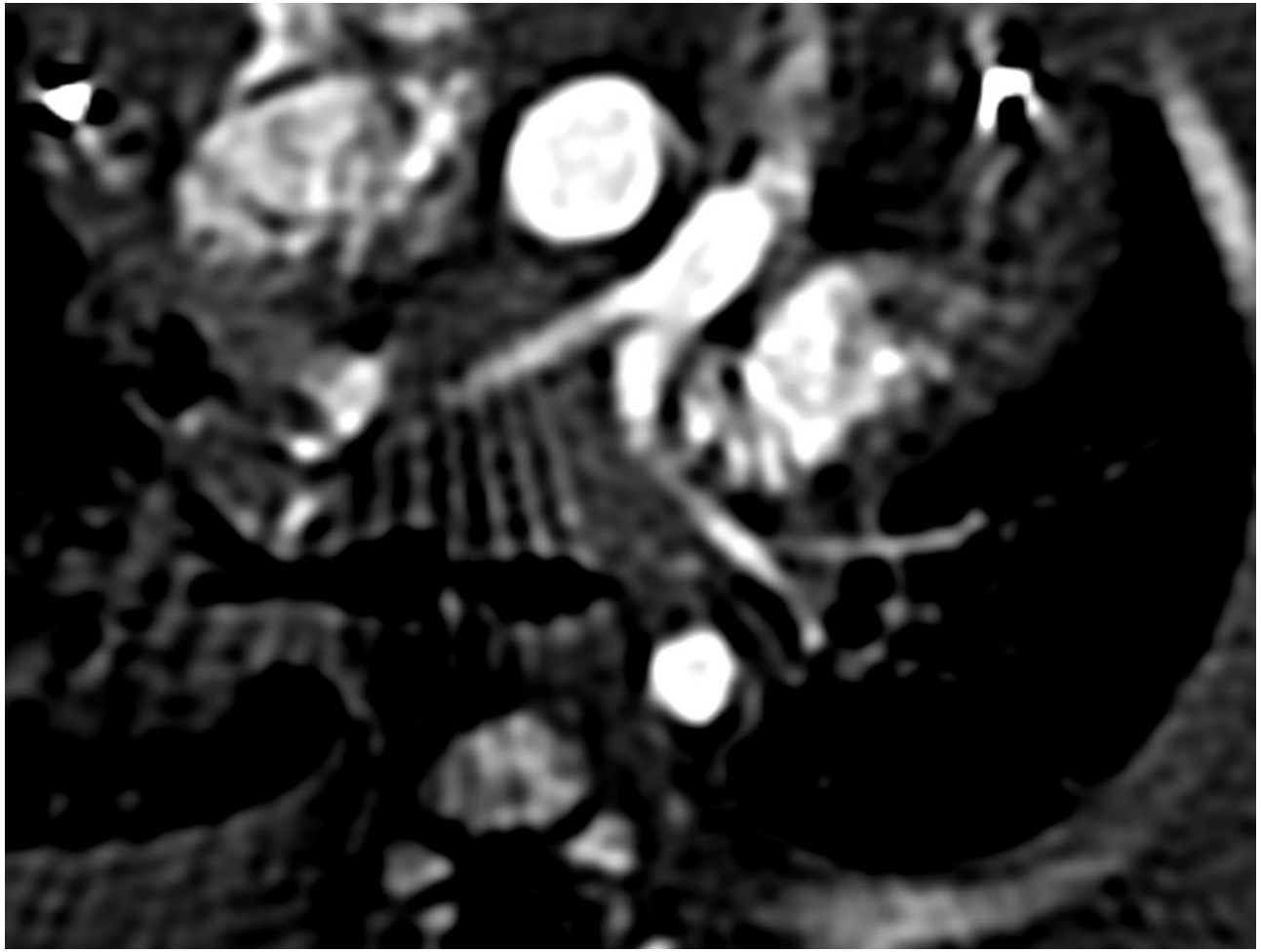

76Hz
9.0cm
2D
60%
C 52
P Off
HGen
M4
G
P
R
3.0
6.0
151 bpm


S9-2
31Hz
8.0cm
2D
60%
C 51
P Off
HGen
CF
36%
8563Hz
WF 856Hz
3.3MHz
M4
+99.9
-99.9
cm/s
157 bpm


35Hz
5.0cm
2D
59%
C 51
P Off
HGen
CF
40%
4300Hz
WF 429Hz
3.3MHz
G
P
R
3.0
6.0
M4 M4
+50.2
-50.2
cm/s
X2
150 bpm

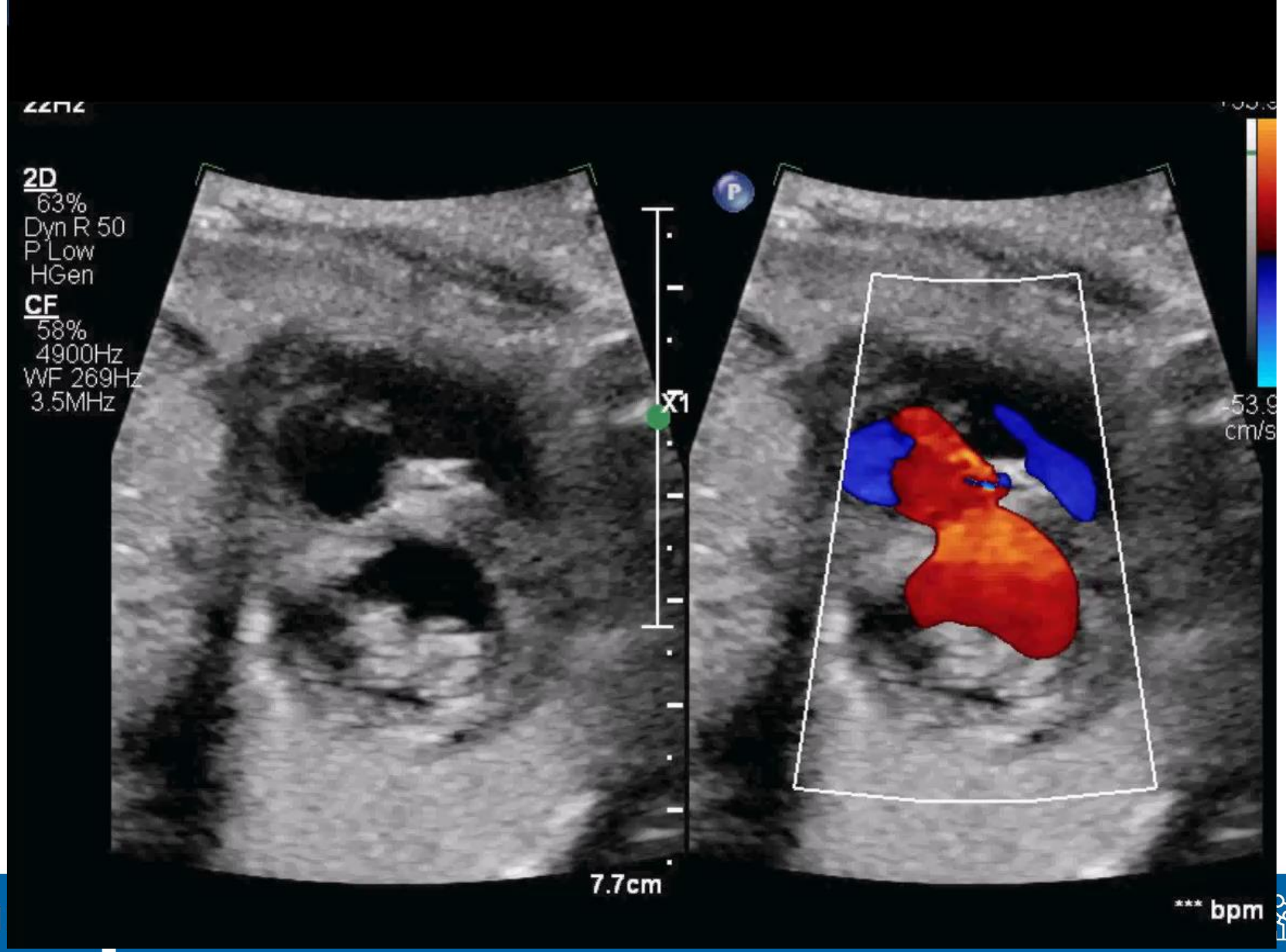



P

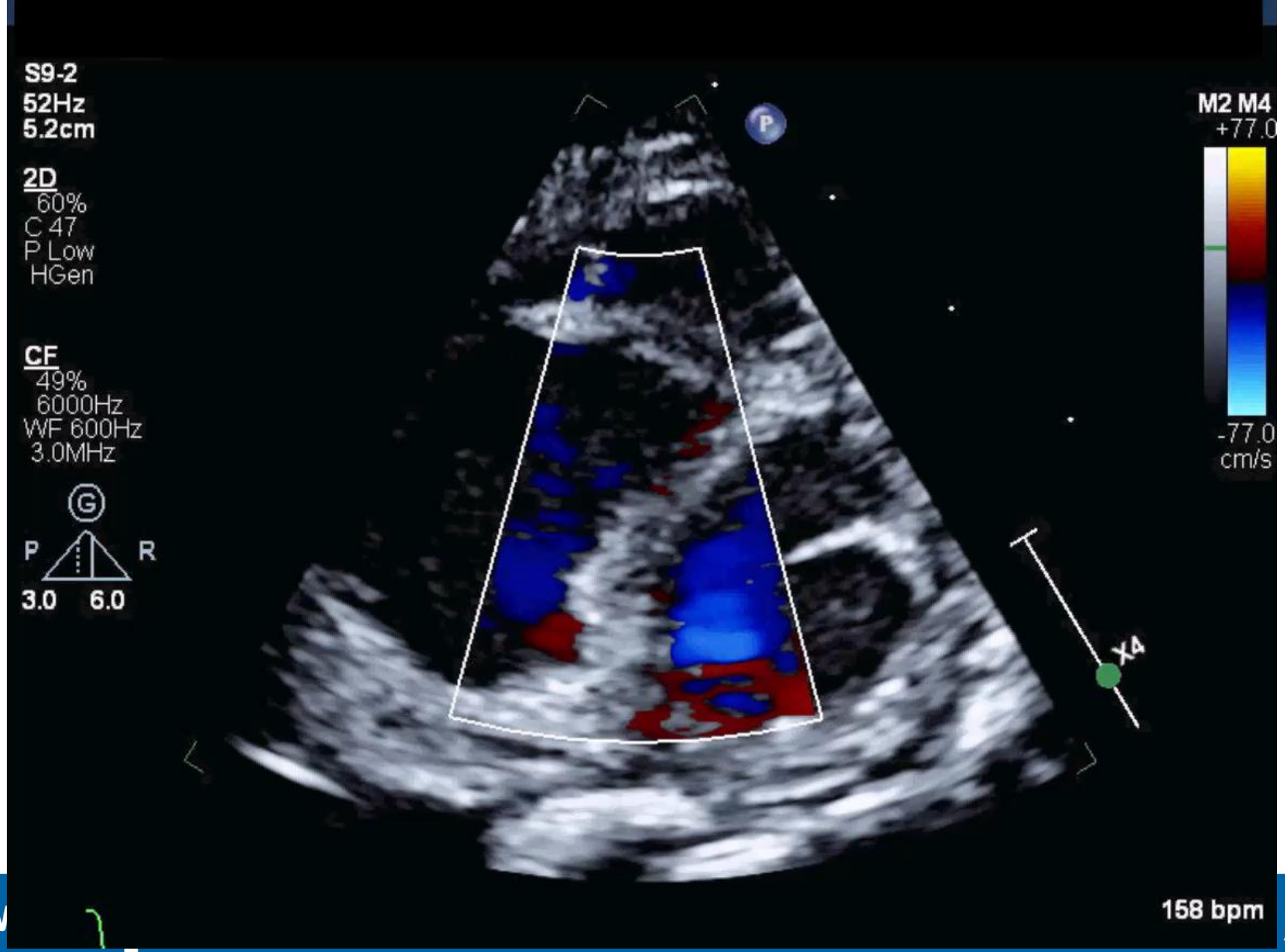



71Hz
RS
2D
37%
Dyn R 48
P Low
HGen
M2
G
P
R
2.0
4.0
X1
8.3cm
*** bpm

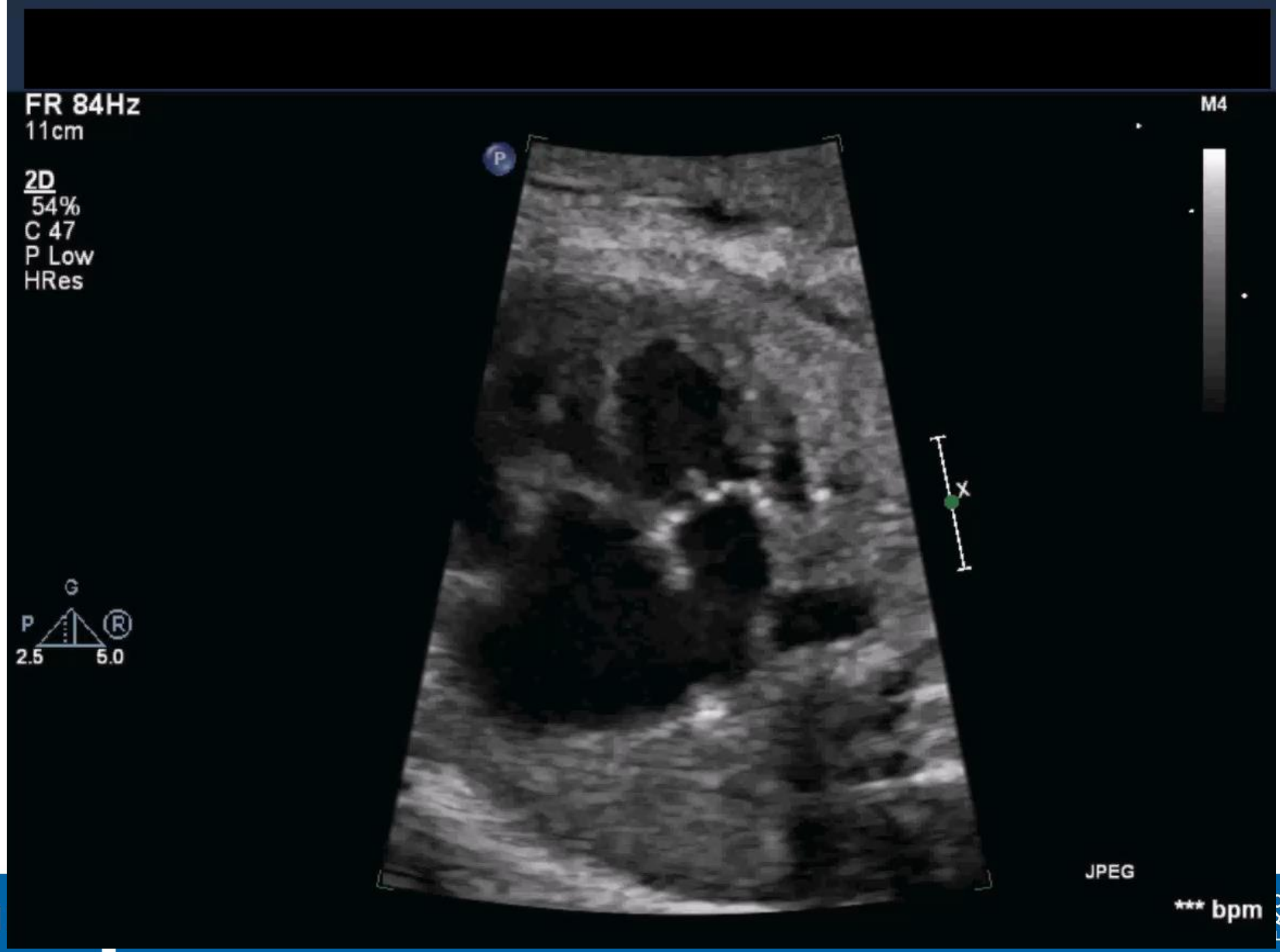

101Hz
RS
2D
36%
Dyn R 46
P Low
HGen
G
P
R
2.0
4.0
M2
X1
7.8cm
*** bpm

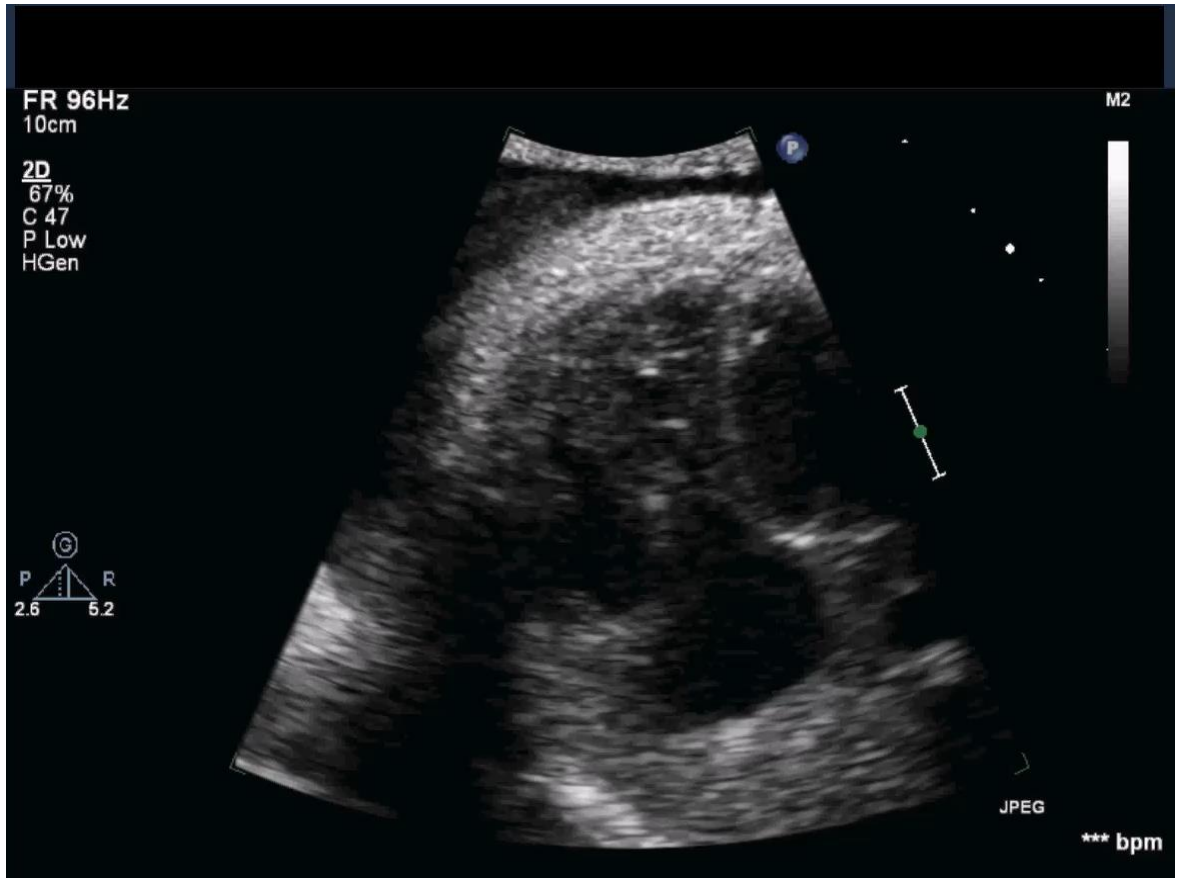

2D
63%
Dyn R 50
P Low
HGen
CF
58%
4900Hz
WF 269Hz
3.5MHz
X1
-53.9
cm/s
7.7cm
*** bpm

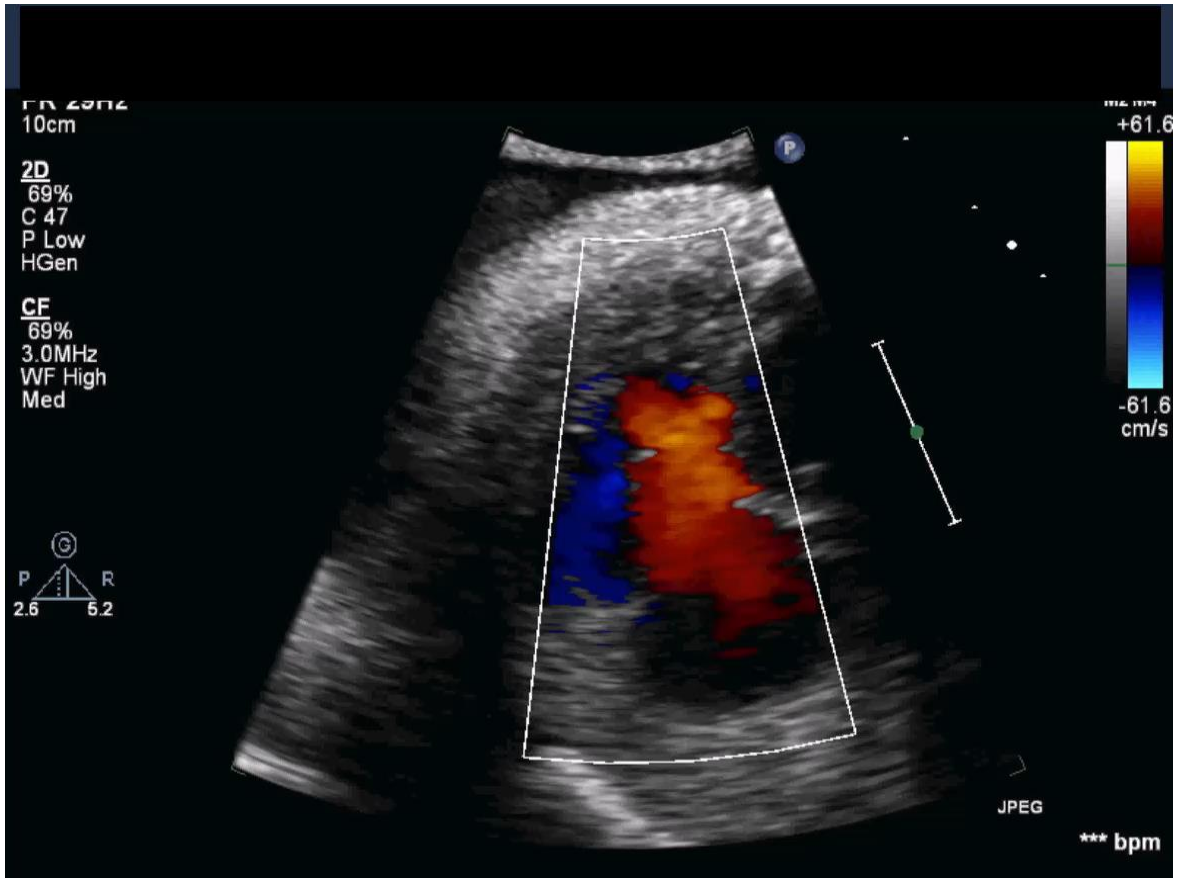

S9-2
89Hz
7.0cm
2D
58%
C 50
P Low
HGen
M2
G
P
R
3.0
6.0
148 bpm

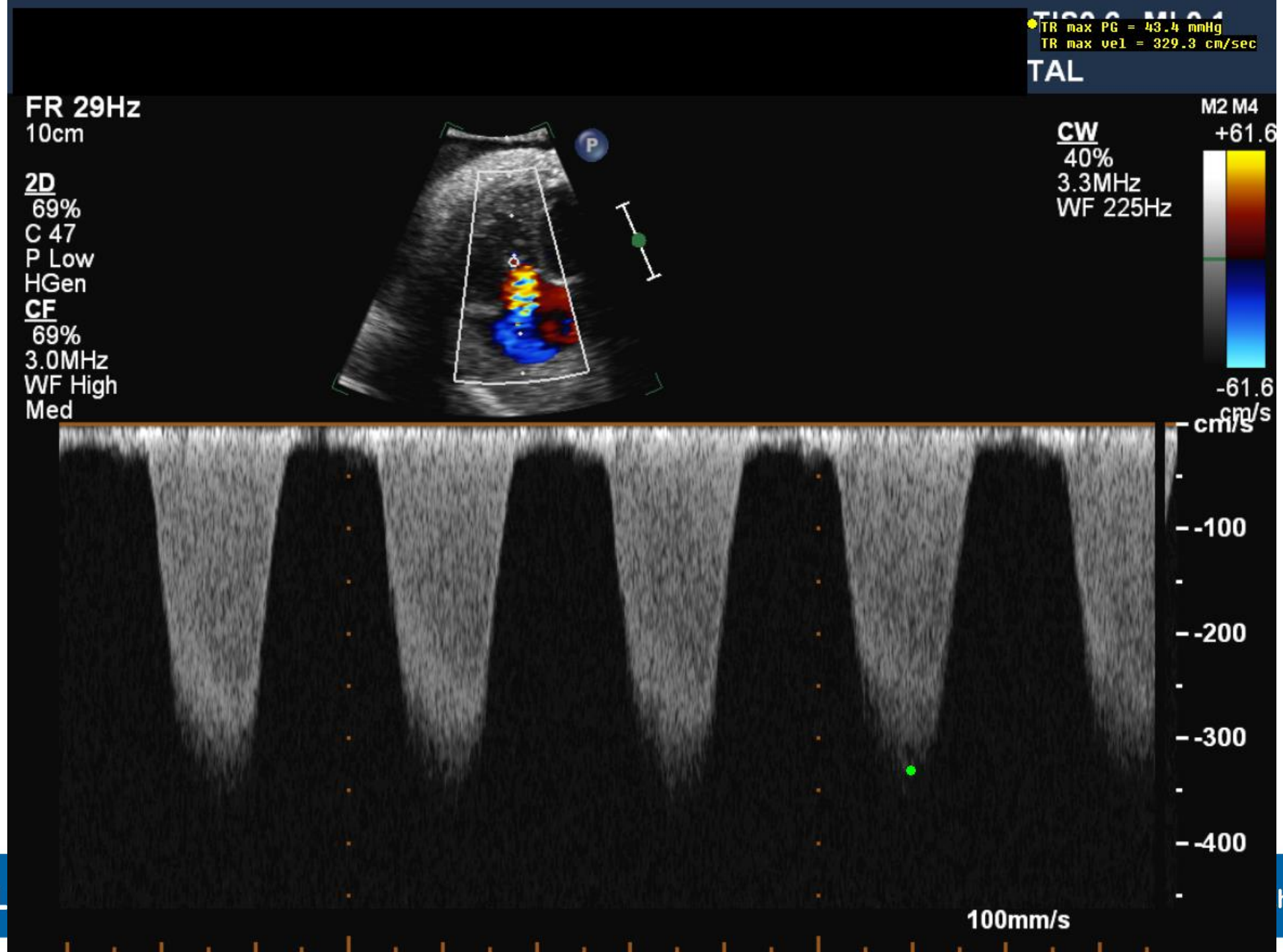

S9-2
52Hz
5.2cm
2D
60%
C 47
P Low
HGen
CF
49%
6000Hz
WF 600Hz
3.0MHz
G
P
R
3.0
6.0
M2 M4
+77.0
-77.0
cm/s
X4
158 bpm


FR 50Hz
12cm
2D
56%
C 41
P Low
HRes
M4
P
G
P
R
2.5
5.0
JPEG
*** bpm

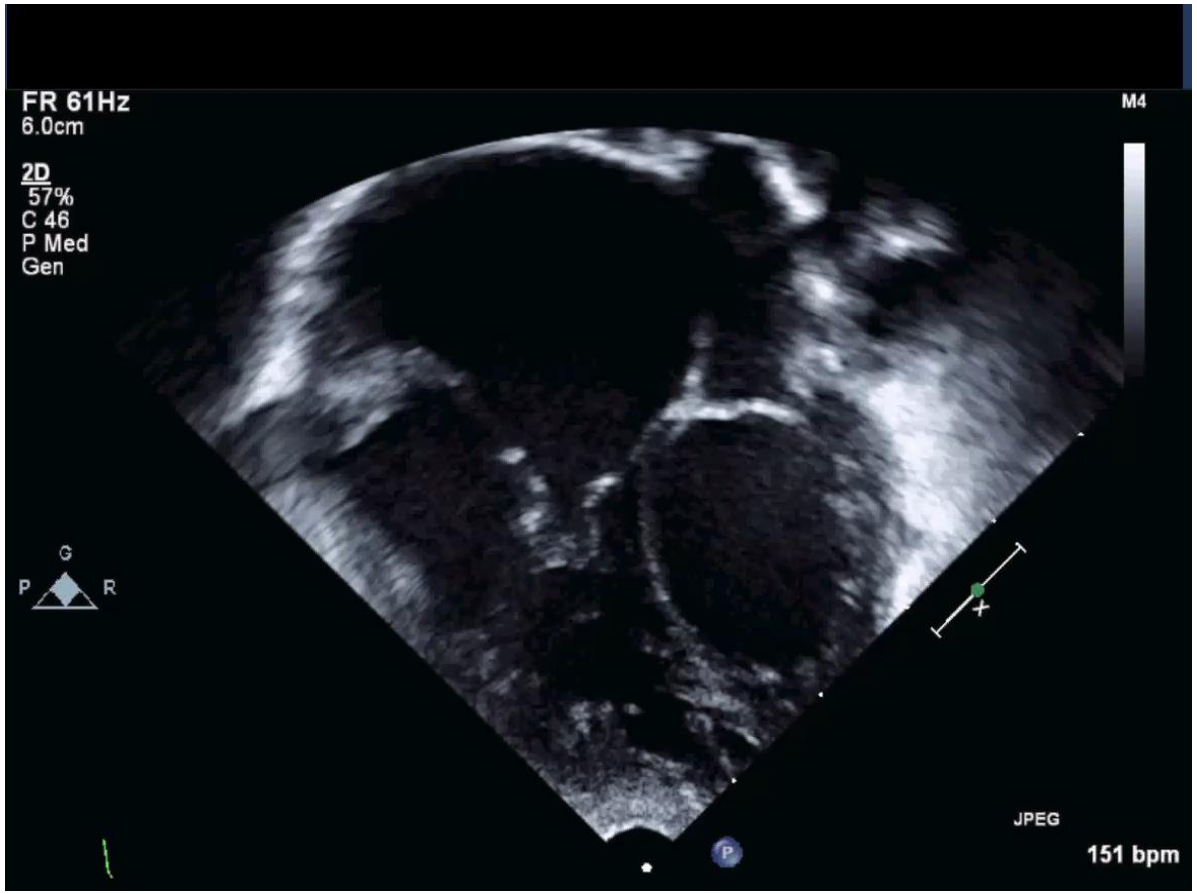

FR 84Hz
11cm
2D
54%
C 47
P Low
HRes
M4
P
X
G
P
R
2.5
5.0
JPEG
*** bpm

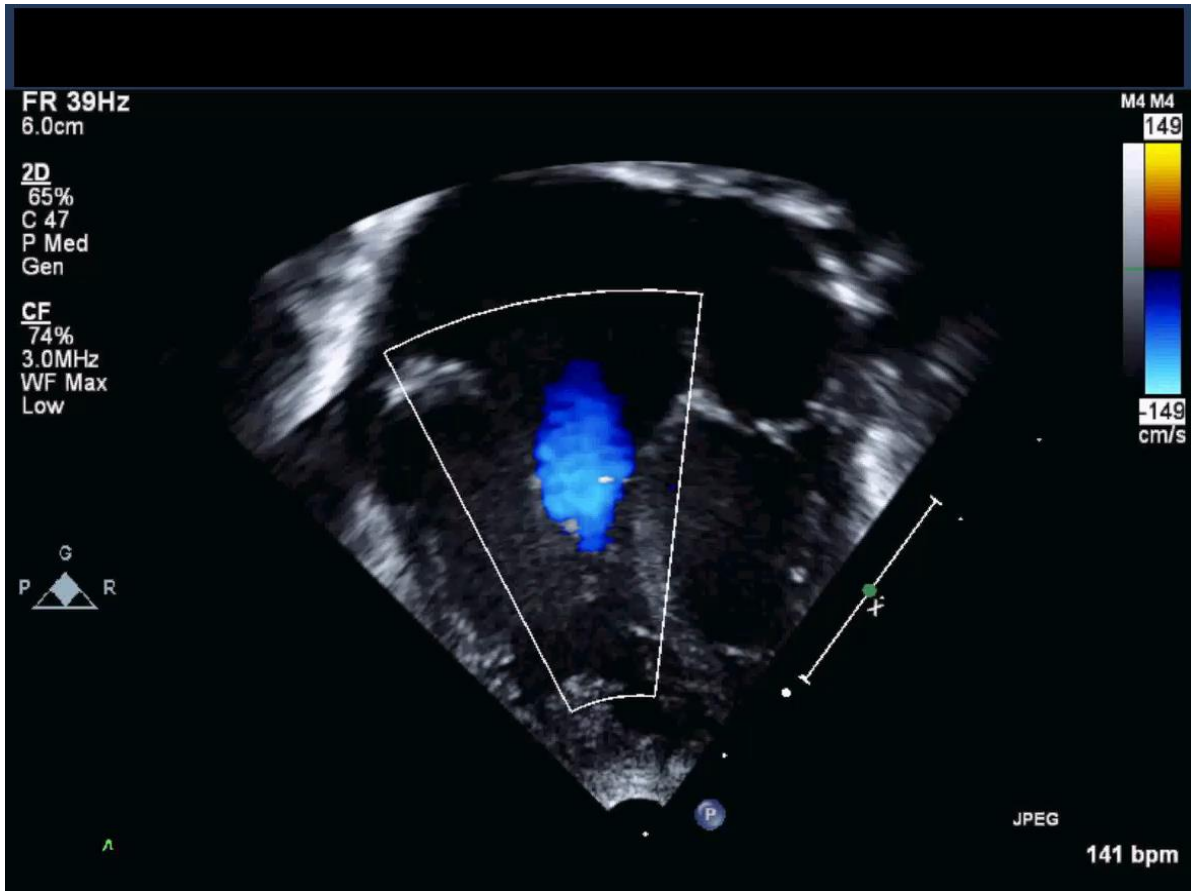

FR 96Hz
10cm
2D
67%
C 47
P Low
HGen
M2
P
G
P
R
2.6
5.2
JPEG
*** bpm


10cm
2D
69%
C 47
P Low
HGen
CF
69%
3.0MHz
WF High
Med
+61.6
-61.6
cm/s
P
G
P
R
2.6
5.2
JPEG
*** bpm


TR max PG = 43.4 mmHg
TR max vel = 329.3 cm/sec
TAL
FR 29Hz
10cm
2D
69%
C 47
P Low
HGen
CF
69%
3.0MHz
WF High
Med
CW
40%
3.3MHz
WF 225Hz
M2 M4
+61.6
-61.6
cm/s
cm/s
-100
-200
-300
-400
100mm/s


FR 64Hz
9.4cm
2D
56%
C 41
P Low
HRes
M4
P
G
P
R
2.5
5.0
X
JPEG
*** bpm


FR 61Hz
6.0cm
2D
57%
C 46
P Med
Gen
M4
G
P
R
JPEG
151 bpm


FR 39Hz
6.0cm
2D
65%
C 47
P Med
Gen
CF
74%
3.0MHz
WF Max
Low
M4 M4
149
-149
cm/s
G
P
R
JPEG
141 bpm

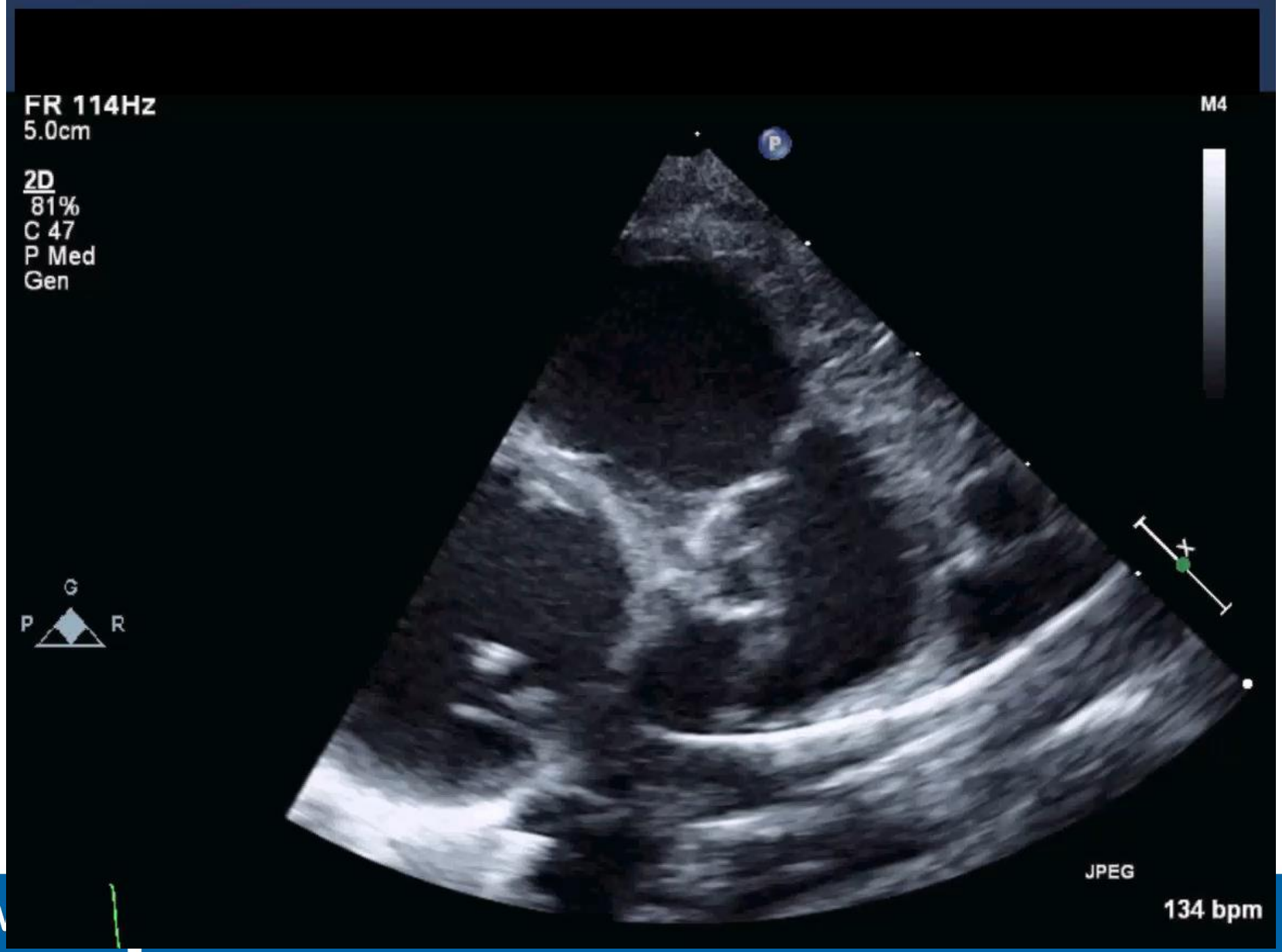
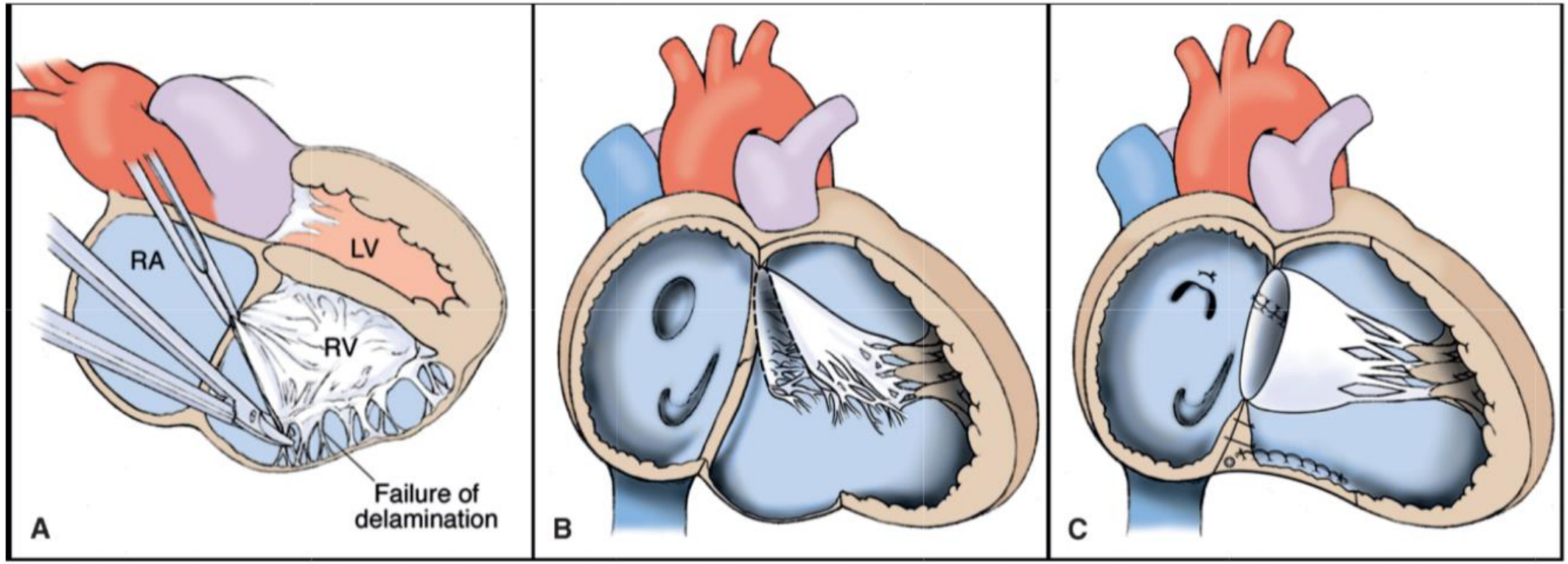
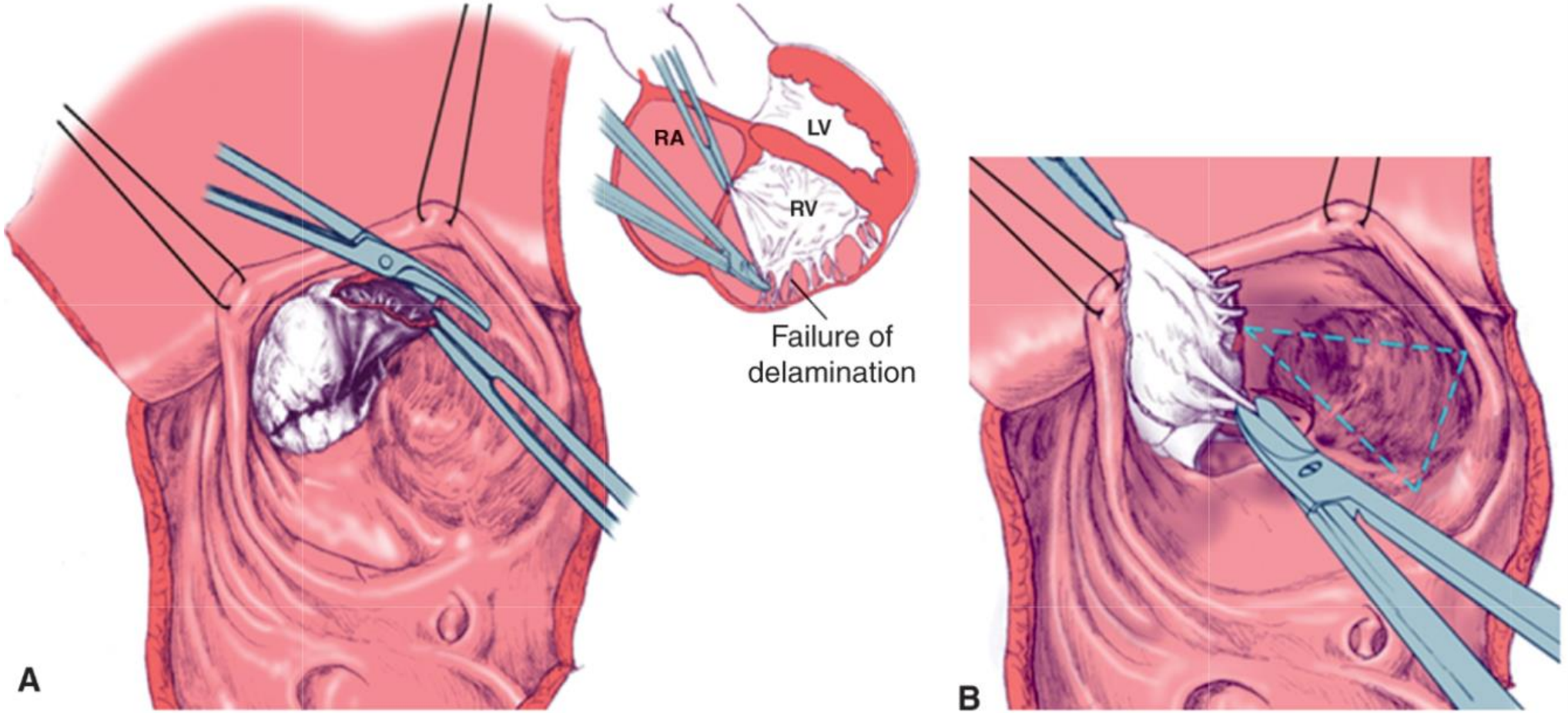
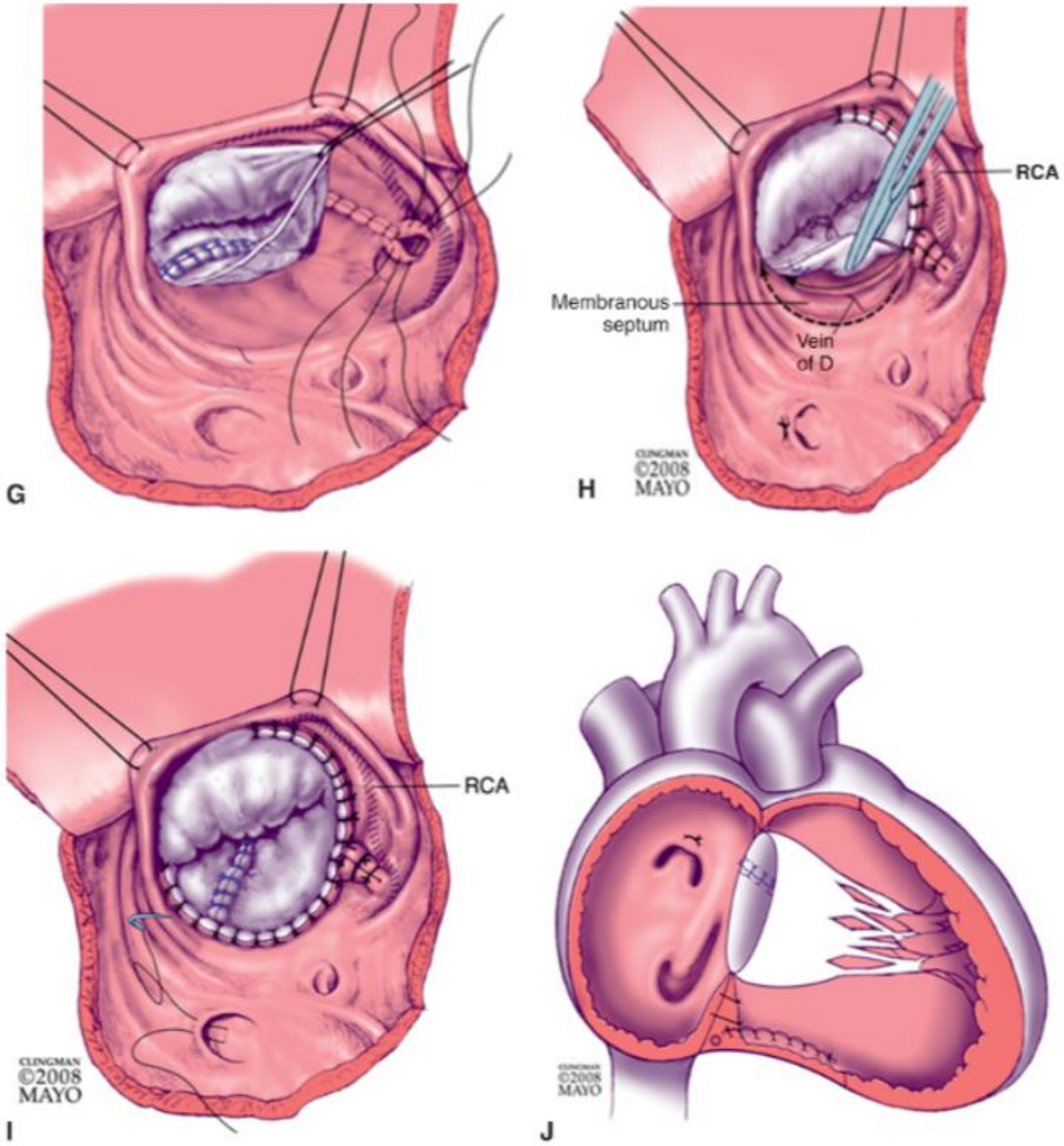

FR 114Hz
5.0cm
2D
81%
C 47
P Med
Gen
M4
P
G
P
R
JPEG
134 bpm

TABLE 38.5 Indications of Surgical Intervention in Ebstein Anomaly

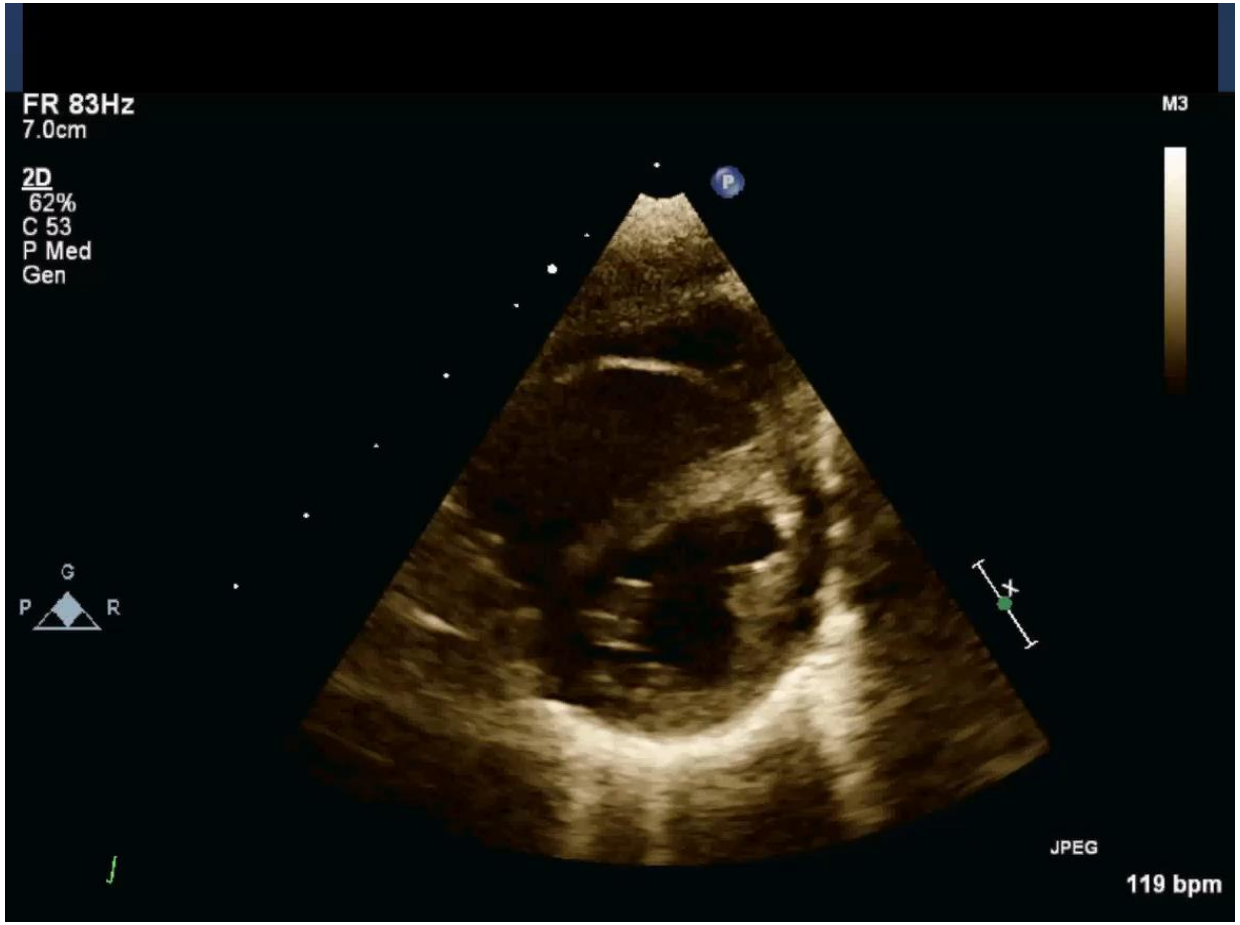
Decreased exercise tolerance
Cyanosis
Progressive right ventricular dilatation (Cardiothoracic ratio >60%)
Prior to significant right ventricular dysfunction
Onset or progression of atrial arrhythmias
Prior to left ventricular dysfunction



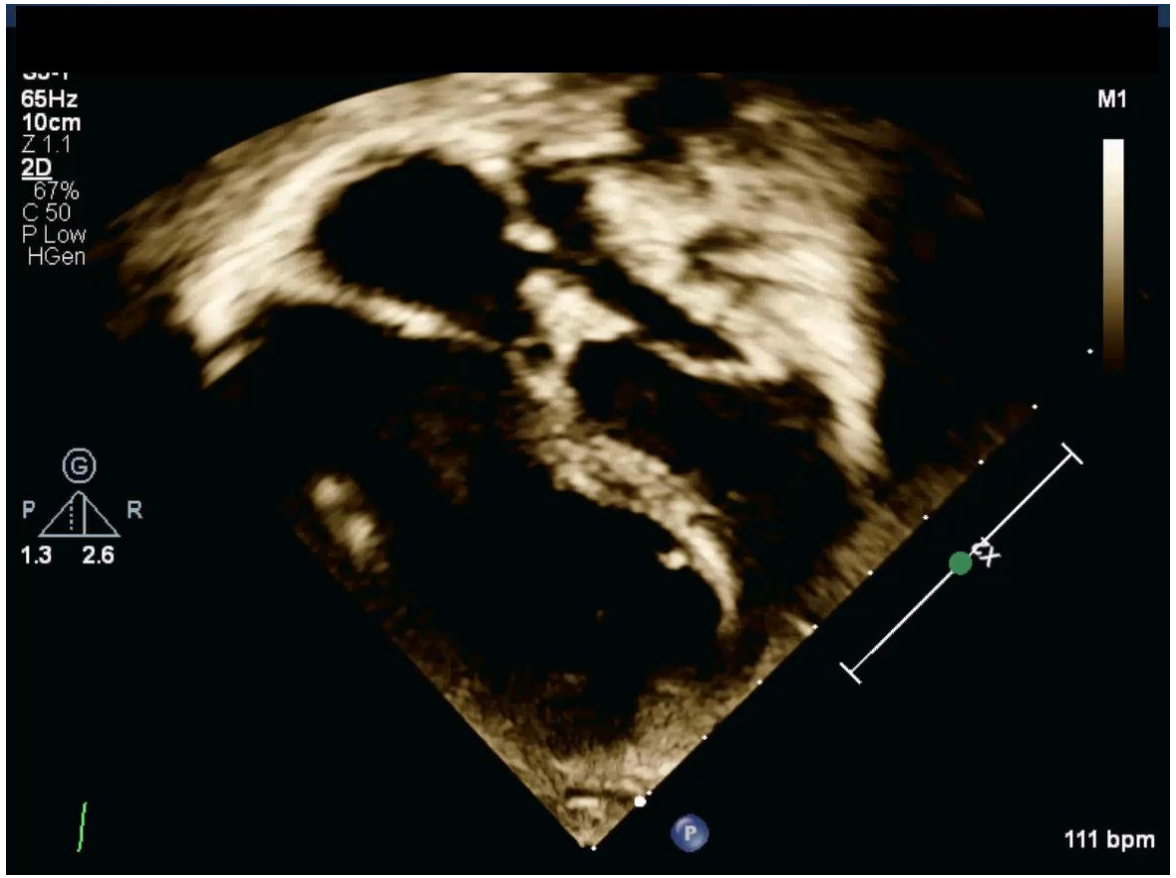

RA
LV
RV
Failure of
delamination
A
B

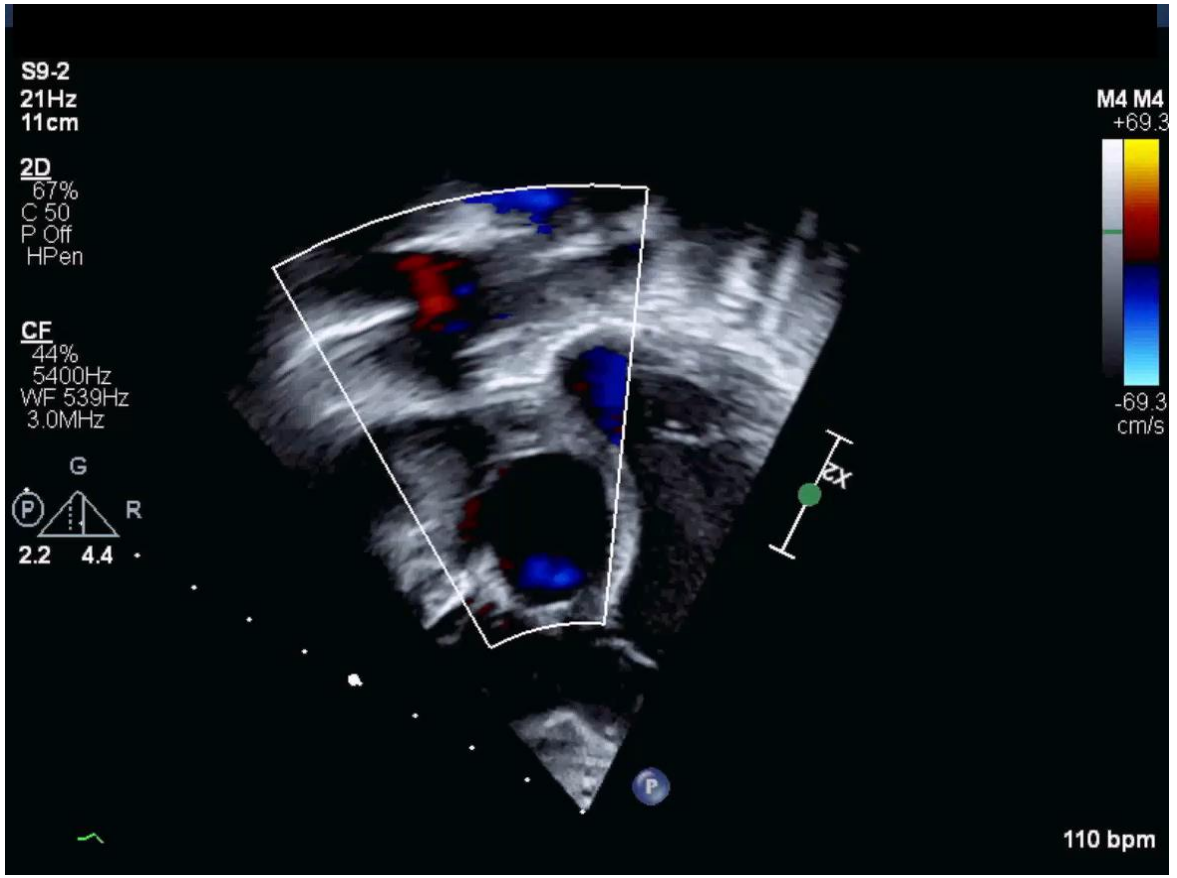


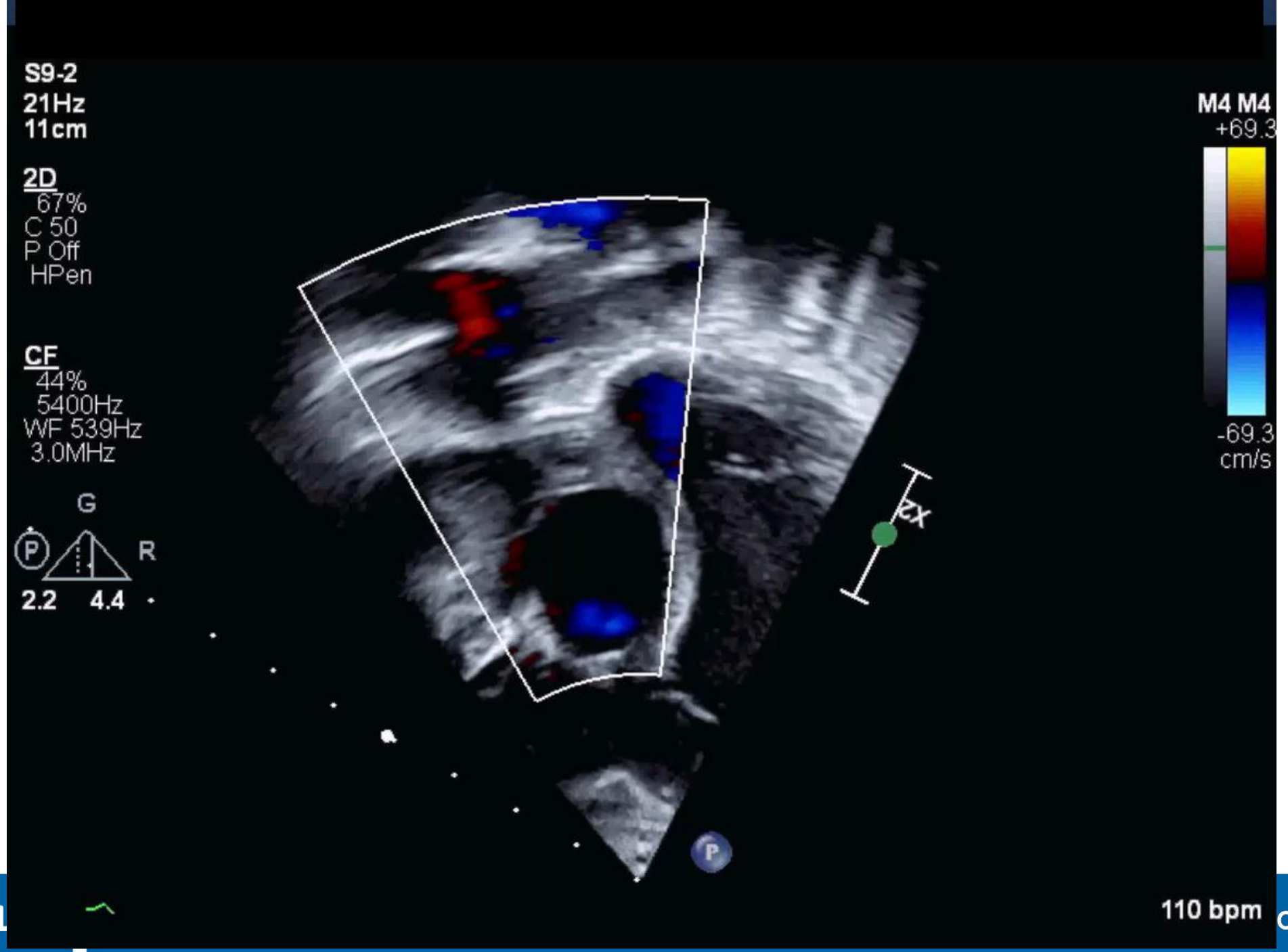

FR 63Hz
7.0cm
2D
56%
C 53
P Med
Gen
M3
G
P
R
JPEG
118 bpm


FR 83Hz
7.0cm
2D
62%
C 53
P Med
Gen
M3
G
P
R
JPEG
119 bpm


55Hz
12cm
Z 1.1
2D
67%
C 50
P Off
HPen
M4
G
P
R
2.2
4.4
118 bpm


65Hz
10cm
Z 1.1
2D
67%
C 50
P Low
HGen
M1
G
P
R
1.3
2.6
111 bpm


S9-2
21Hz
11cm
2D
67%
C 50
P Off
HPen
CF
44%
5400Hz
WF 539Hz
3.0MHz
G
P
R
2.2
4.4
M4 M4
+69.3
-69.3
cm/s
ex
110 bpm


S9-2
21Hz
11cm
2D
67%
C 50
P Off
HPen
CF
44%
5400Hz
WF 539Hz
3.0MHz
G
R
2.2
4.4
M4 M4
+69.3
-69.3
cm/s
ex
110 bpm

The outcomes of operations for 539 patient

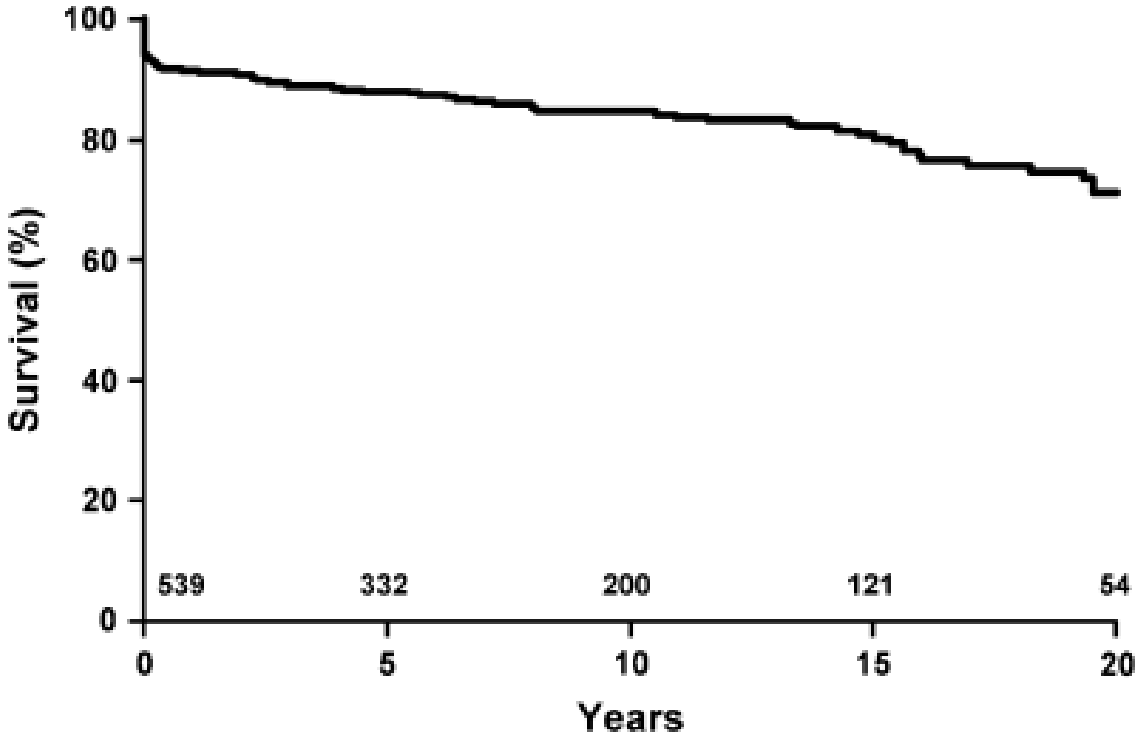


Figure 2. Long-term survival for all 539 patients. Time 0 is the time of the first cardiac operation at the Mayo Clinic.

…n K. Danielson, MD,[a] Frank Cetta, M…
MS,[d] and David J. Driscoll, MD,[b] for …

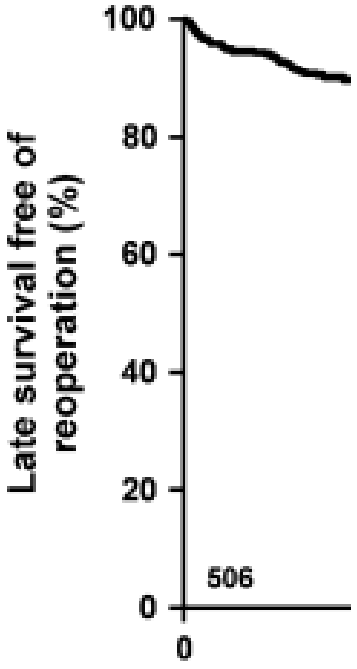


Figure 3. Survival … valve or other) for a… the Mayo Clinic.

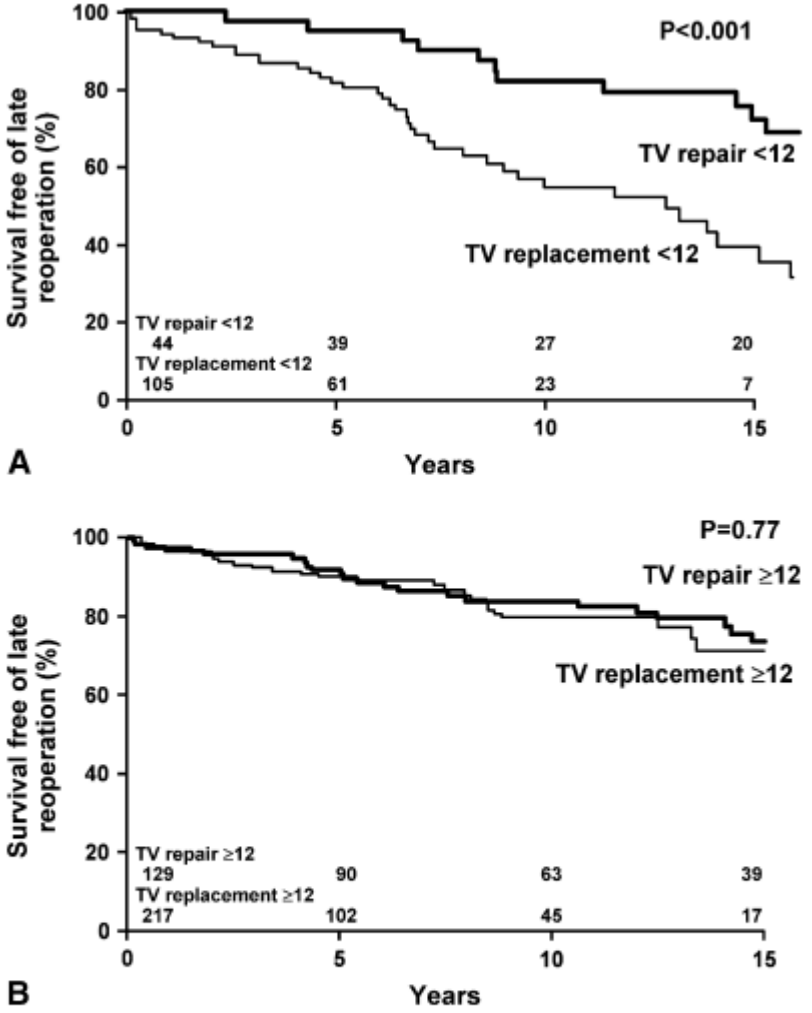


Figure 4. A and B, Survival free from late reoperation on the tricuspid valve *(TV)* for patients less than 12 or 12 or more years of age. The 35 patients who had TV replacement and had prior TV repair followed by TV replacement were removed from the TV replacement group.

References

- Jack Rychik, Zhiyun Tian. *Fetal Cardiovascular Imaging.* (2012).
- Hugh Allen, Robert Shaddy, Daniel Penny, Timothy Feltes, and Frank Cetta. (2016). *Moss and Adams' Heart Disease in Infants, Children, and Adolescents: Including the fetus and young adult* (9th ed). Wolters Kluwer.
- Gil Wernovsky, Robert Anderson, Krishna Kumar, Kathleen Mussatto, Andrew Redington, James Tweddell. (2020). *Anderson's Pediatric Cardiology* (4th ed). Elsevier.

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  - Rita France RDCS, RDMS, RT
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Questions?

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