Bicuspid Aortic Valve: When Veggies are Bad for You

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April 27th, 2019

Objectives

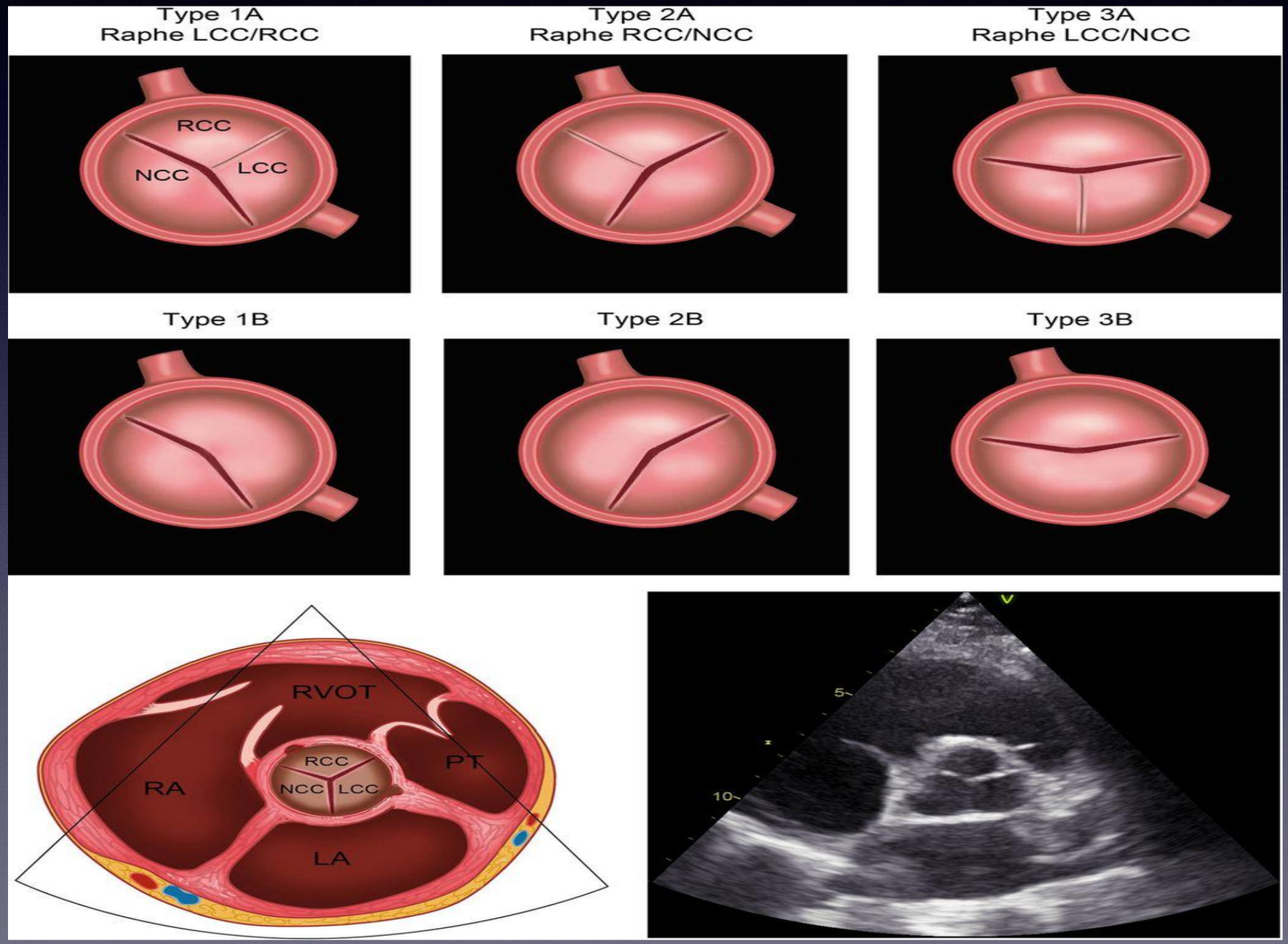
- Review basic congenital pathology
- Discuss typical complications of bicuspid aortic valve
- Discuss endocarditis and its prevalence in bicuspid aortic valves
- Case Report

Bicuspid Aortic Valve

- Bicuspid aortic valve (BAV) - Contains two commissures that present as an oval “football” shape opening on echo.

- Can also present as a functional BAV: where three cusps are present and there is fusion of two cusps.

Bicuspid Aortic Valve (cont.)



https://heart.bmj.com/content/102/12/943

Complications of Bicuspid Aortic Valves (BAV)

- Aortic stenosis and aortic insufficiency
- Coarctation of the aorta
- Aortic aneurysm
- Aortic dissection
- Infective endocarditis

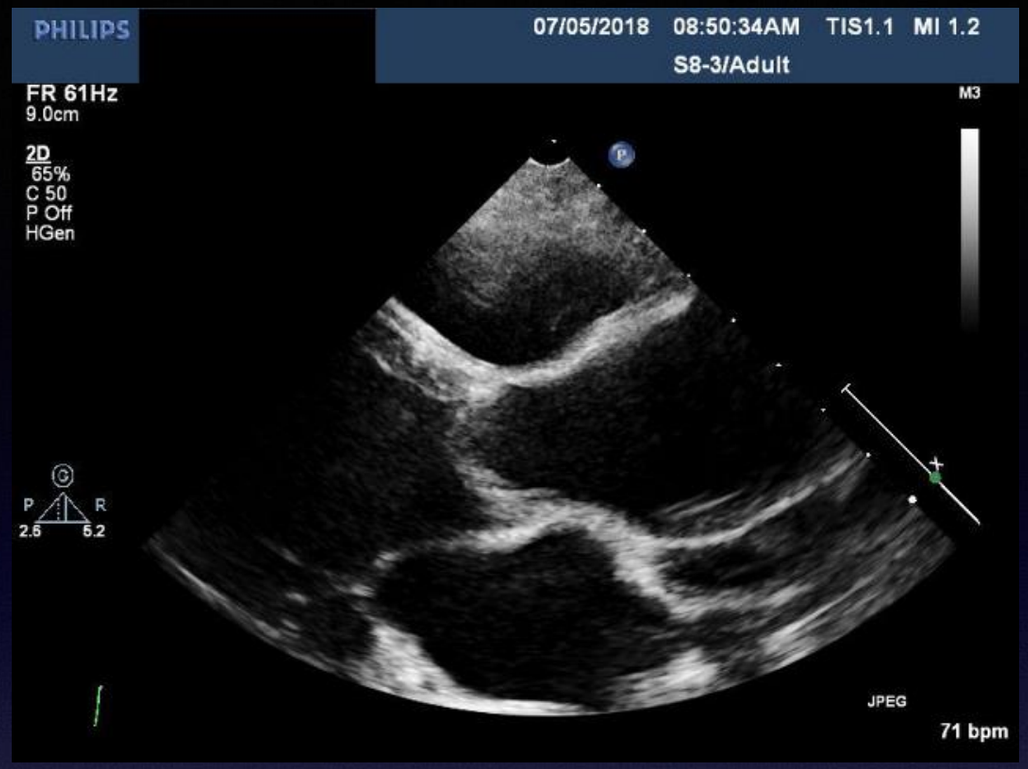
Endocarditis

- Patients with BAV are at a higher risk of developing endocarditis
- Most common causes: Staphlylococcus and Streptococci
- Most common symptoms: Flu-like symptoms (fever, fatigue, aching joints and muscles)
- Along with blood work, endocarditis can be confirmed by vegetation growth seen on echo

Case Study

- Patient is an adolescent male seen in the outpatient clinic for routine follow-up echo. Patient with known history of bicuspid aortic valve, aortic stenosis, aortic insufficiency and ascending aorta dilatation

- History of Osteogenesis Imperfecta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S8-3/Adult
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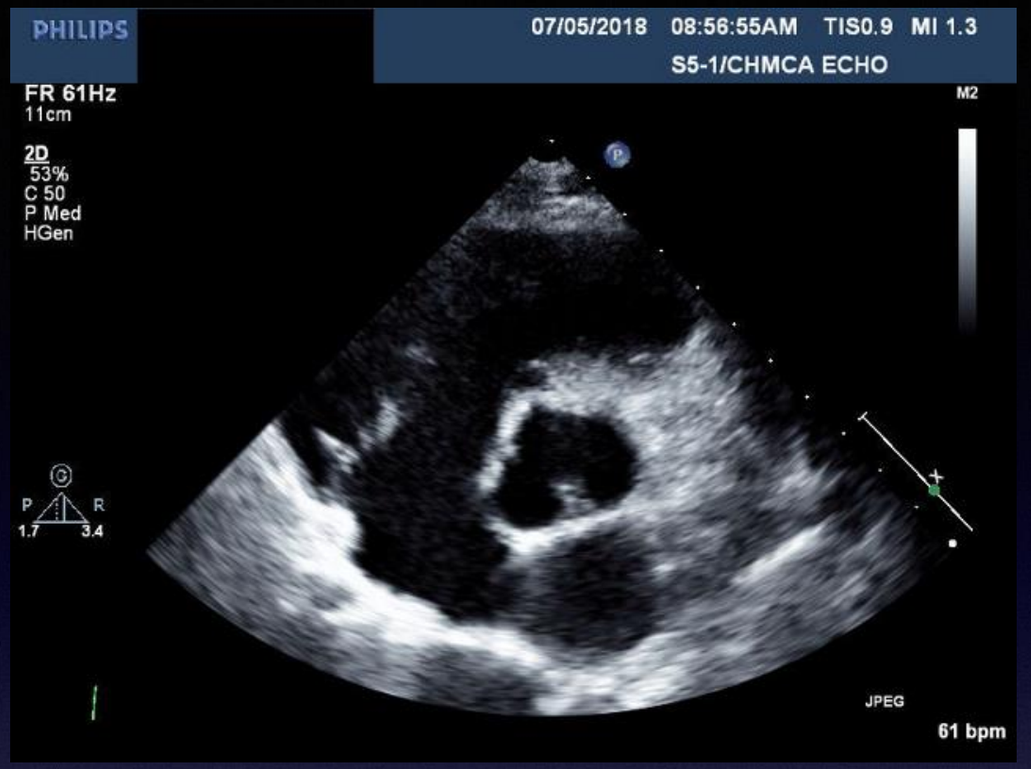
9.0cm
2D
65%
C 50
P Off
HGen
M3
P R
2.6 5.2
JPEG
71 bpm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S5-1/CHMCA ECHO
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2D
53%
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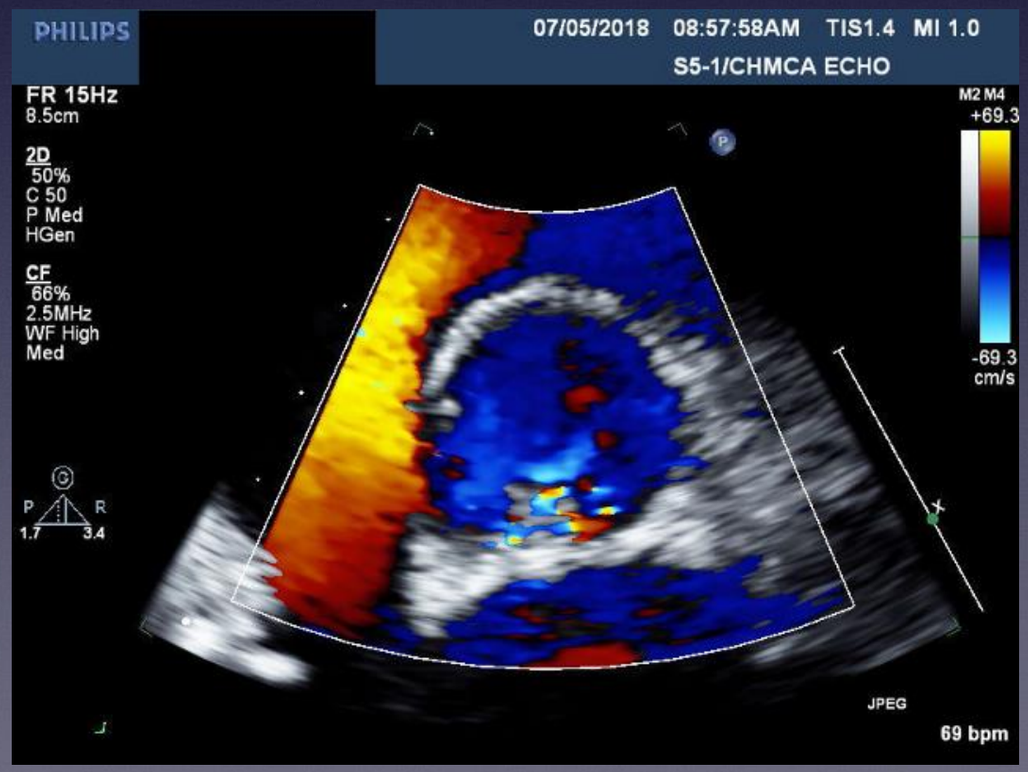
P Med
HGen
M2
P R
1.7 3.4
JPEG
61 bpm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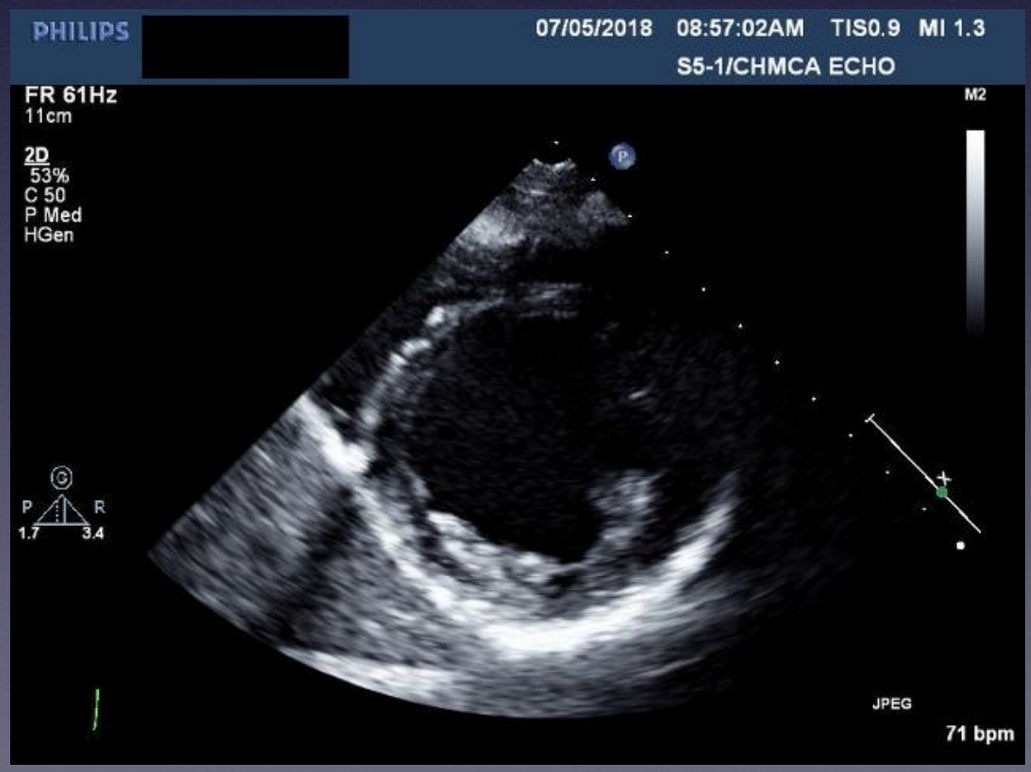
S5-1/CHMCA ECHO
FR 15Hz
8.5cm
2D
50%
C 50
P Med
HGen
CF
66%
2.5MHz
WF High
Med
M2 M4
+69.3
-69.3
cm/s
P R
1.7 3.4
JPEG
69 bpm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S5-1/CHMCA ECHO
FR 61Hz
11cm
2D
53%
C 50
P Med
HGen
M2
P R
1.7 3.4
JPEG
71 bpm


PHILIPS
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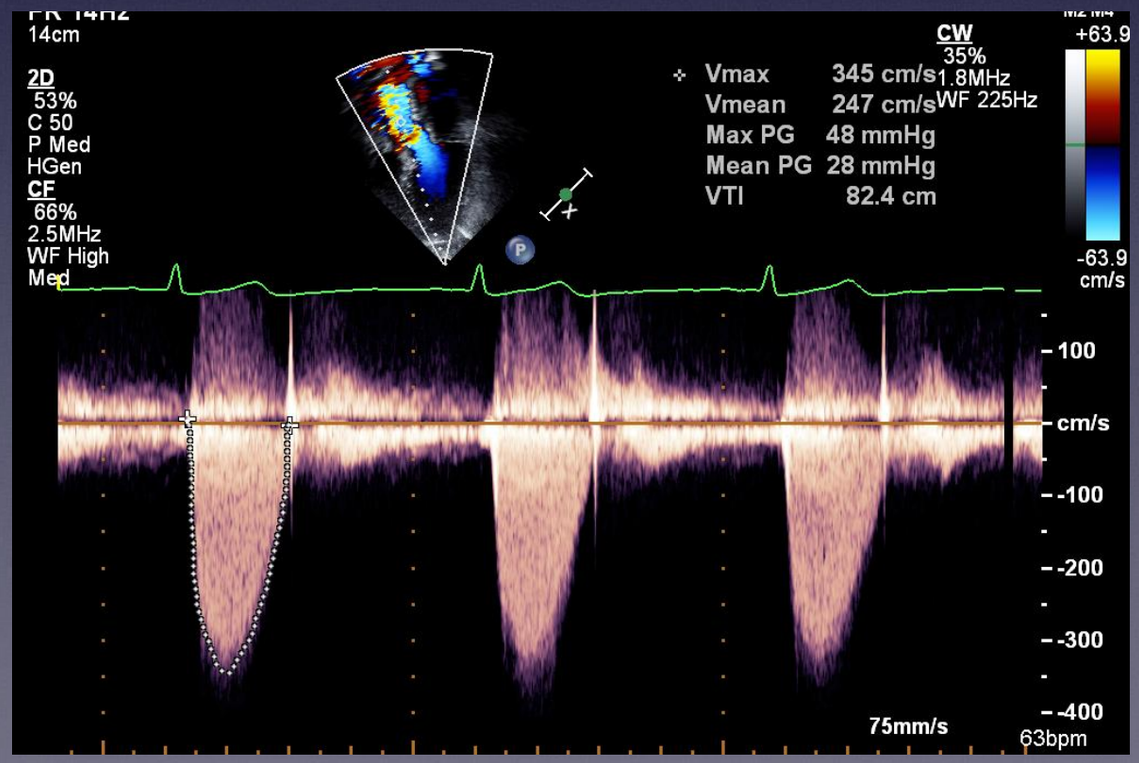
S5-1/CHMCA ECHO
FR 55Hz
13cm
2D
58%
C 48
P Med
HGen
M2
P R
1.7 3.4
JPEG
78 bpm


PHILIPS
06/20/2017 02:25:16PM TIS2.4 MI 1.2
S5-1/CHMCA ECHO
FR 14Hz
14cm
2D
50%
C 50
P Med
HGen
CF
66%
2.5MHz
WF High
Med
M2 M4
+63.9
-63.9
cm/s
P R
1.7 3.4
JPEG
66 bpm


14cm
2D
53%
C 50
P Med
HGen
CF
66%
2.5MHz
WF High
Med
CW
35%
1.8MHz
WF 225Hz
Vmax 345 cm/s
Vmean 247 cm/s
Max PG 48 mmHg
Mean PG 28 mmHg
VTI 82.4 cm
+63.9
-63.9
cm/s
100
cm/s
-100
-200
-300
-400
75mm/s
63bpm

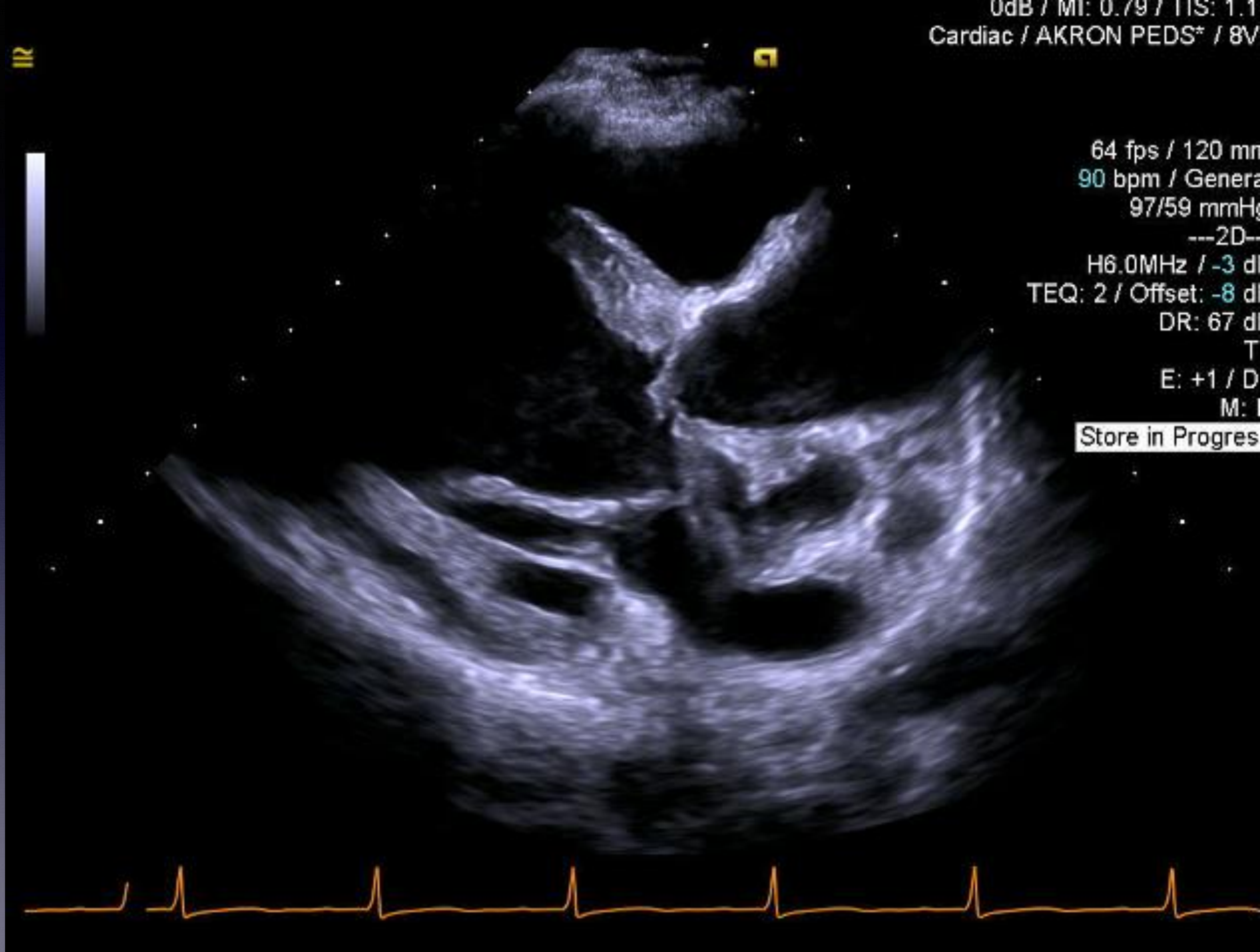
Case Study (cont.)

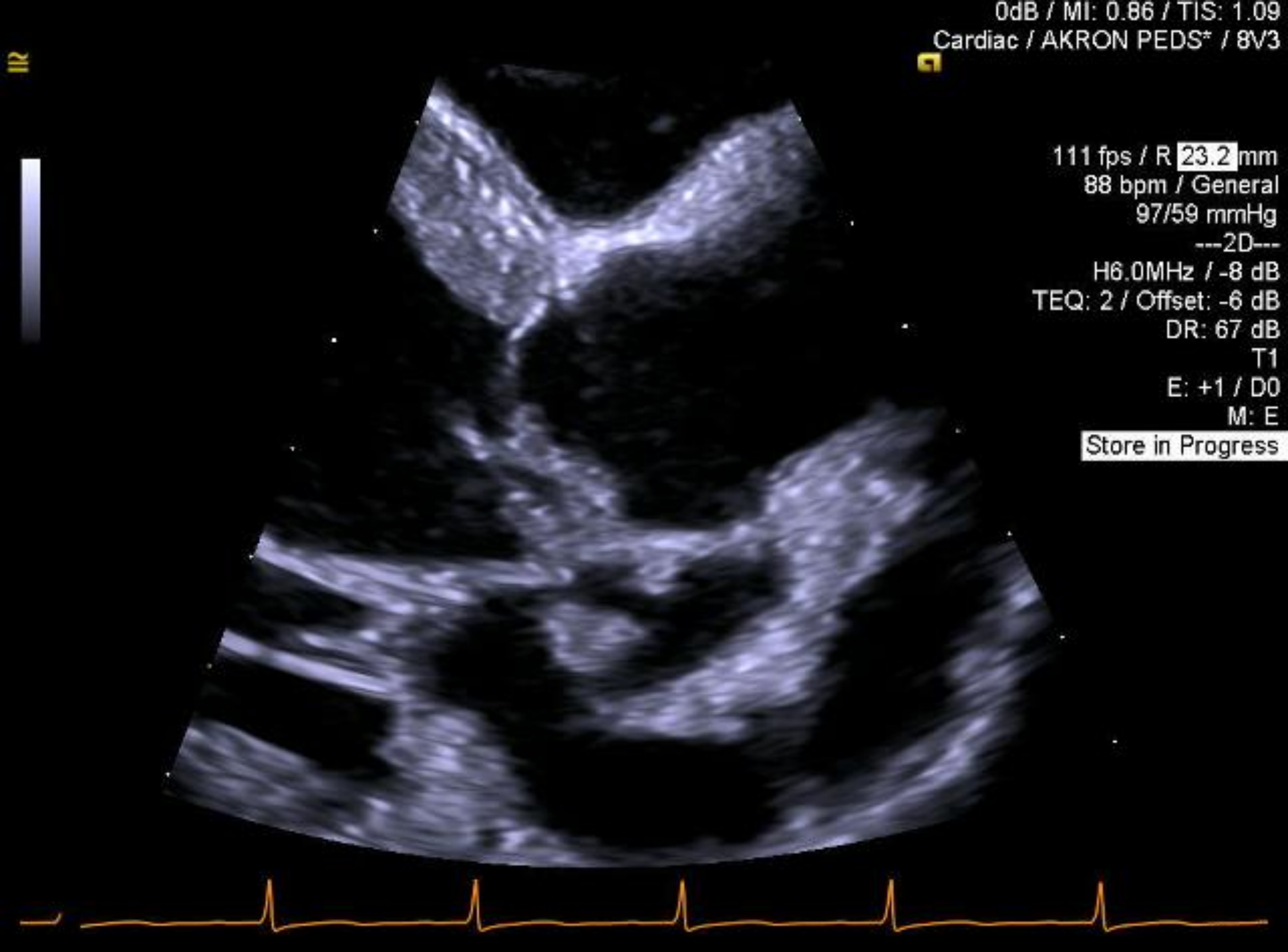
- The same adolescent male is now admitted to the hospital after being seen as an outpatient. Physician observed patient was pale and upon examination, evidence of an enlarged spleen and liver was noted. Physician wanted patient evaluated by Hematology/Oncology since he also presented with fatigue and increased sleep, fever, night sweats, significant weight loss, headaches, and anemia.

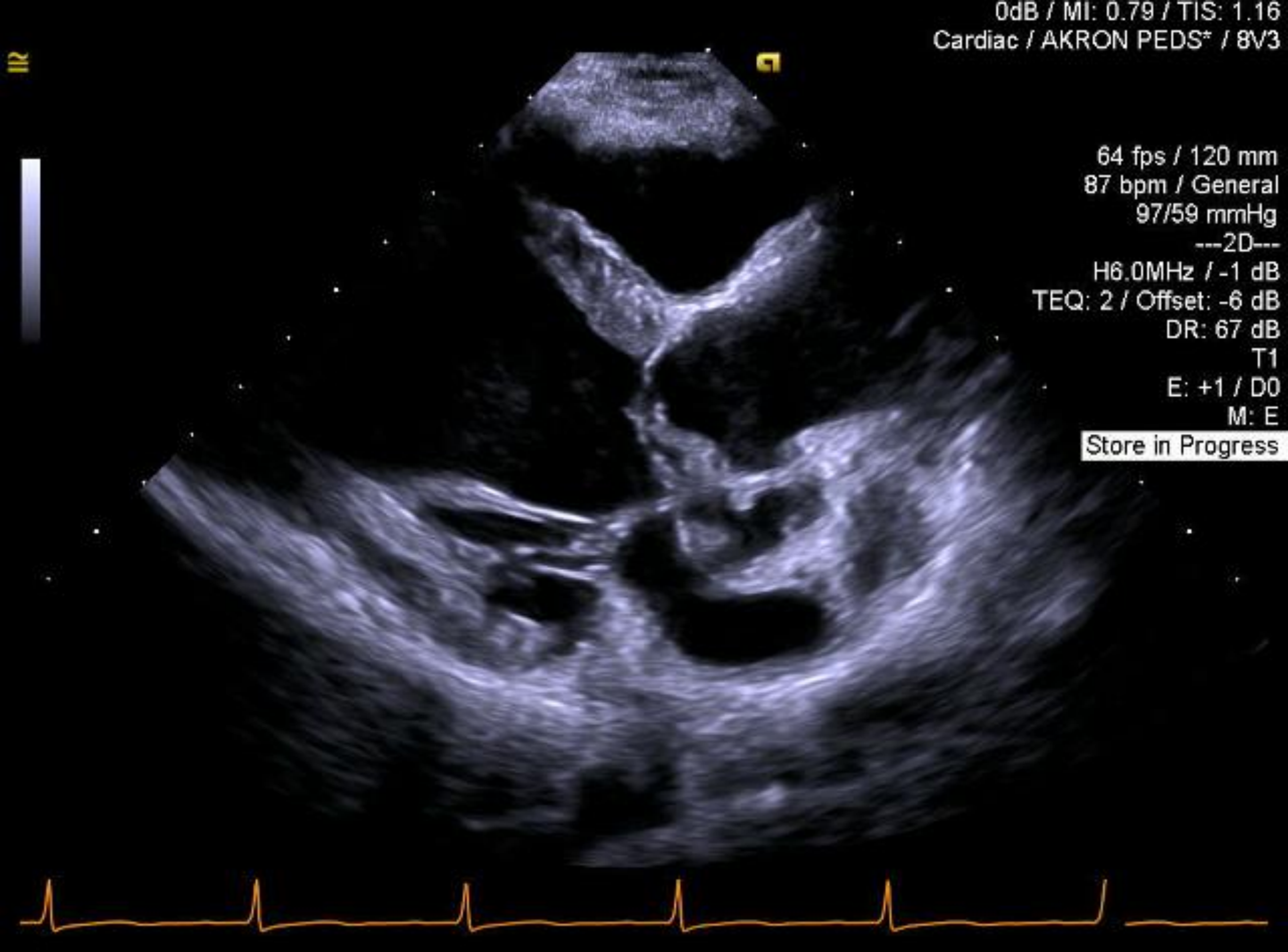
- Patient was admitted for work up of anemia and there were concerns for leukemia or lymphoma. A CT scan was ordered.

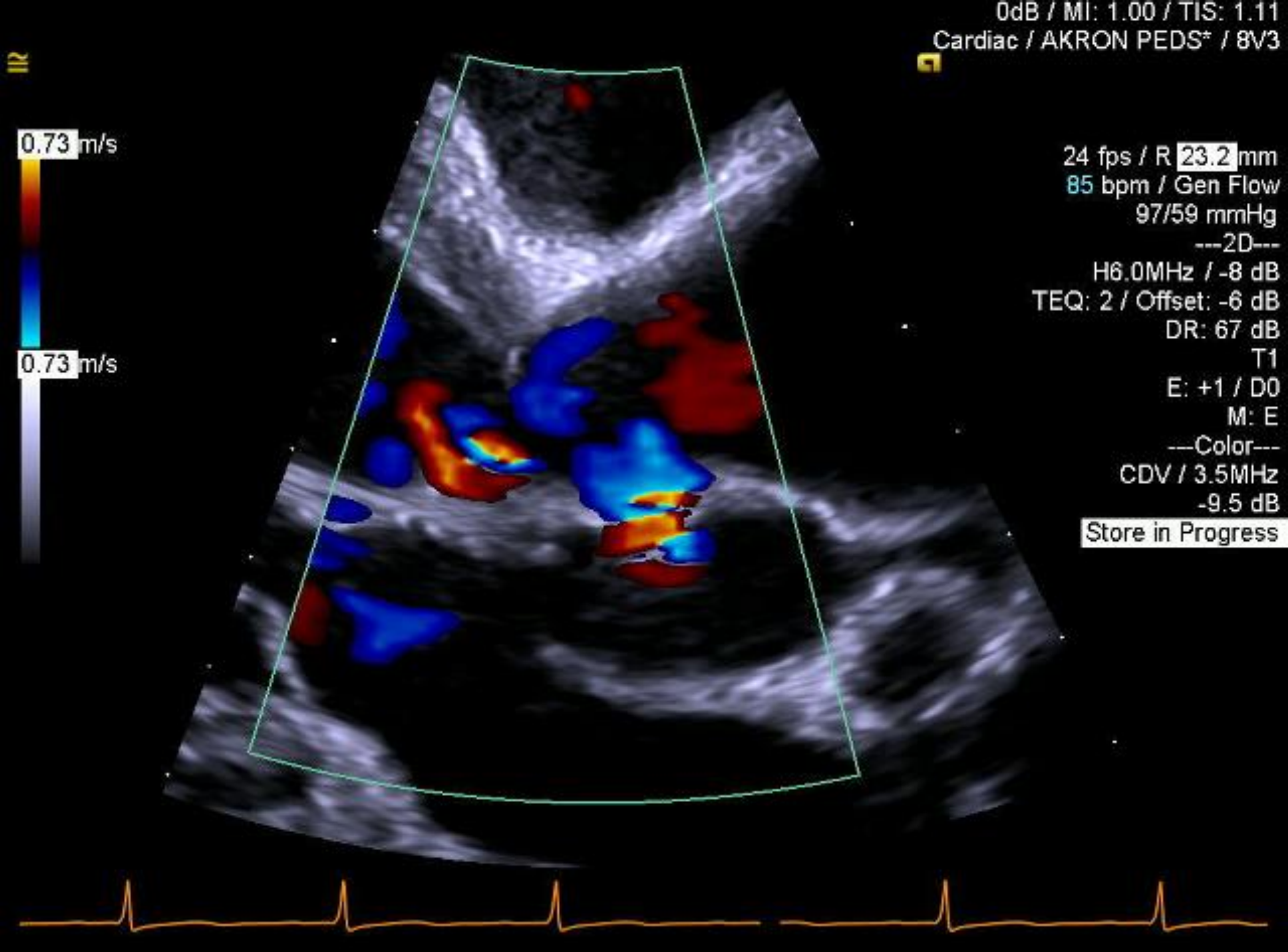
- CT scan showed abnormal aorta dilation. Also, dissection could not be ruled out due to artifact.

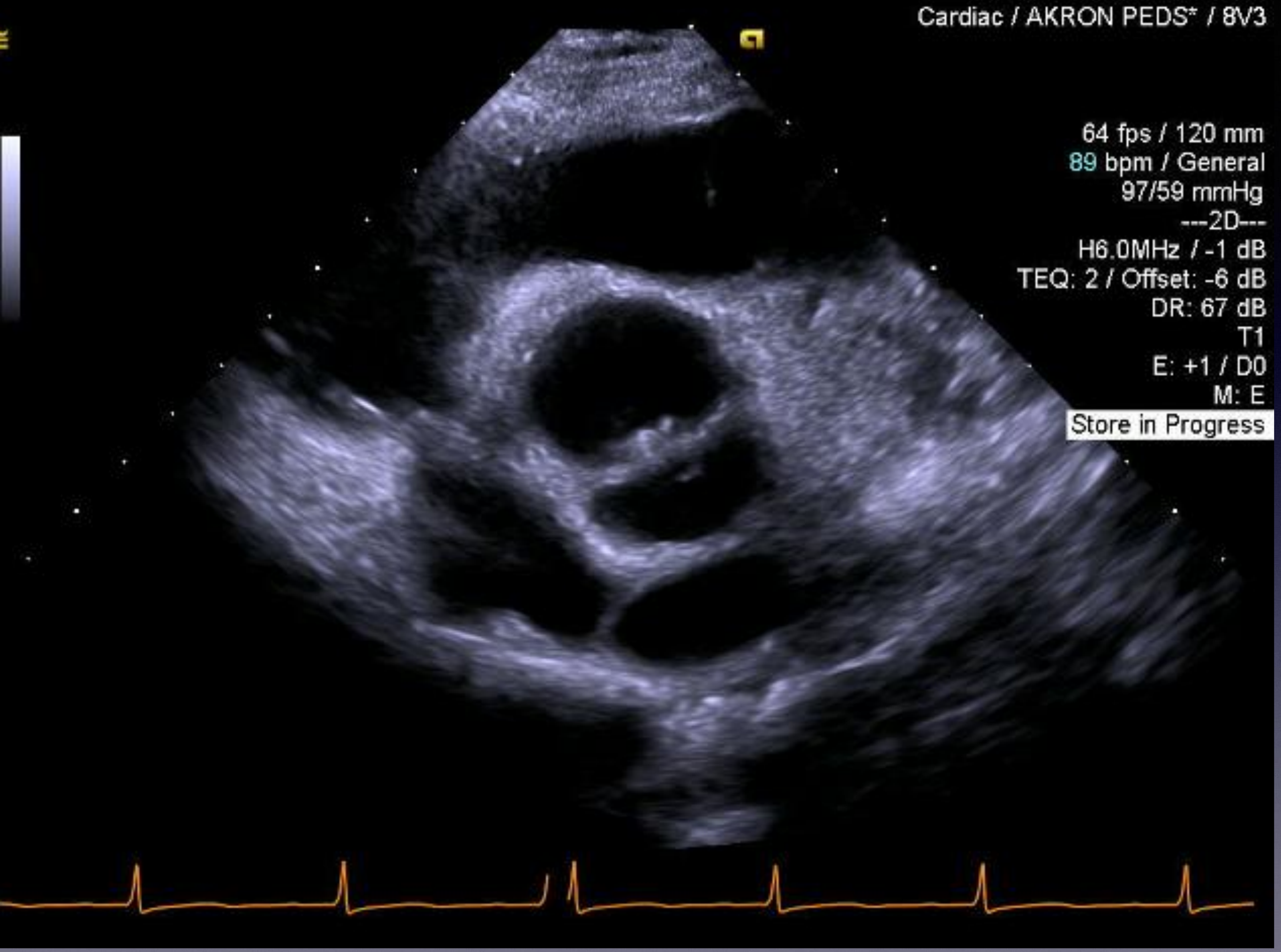
- Enter echo……..

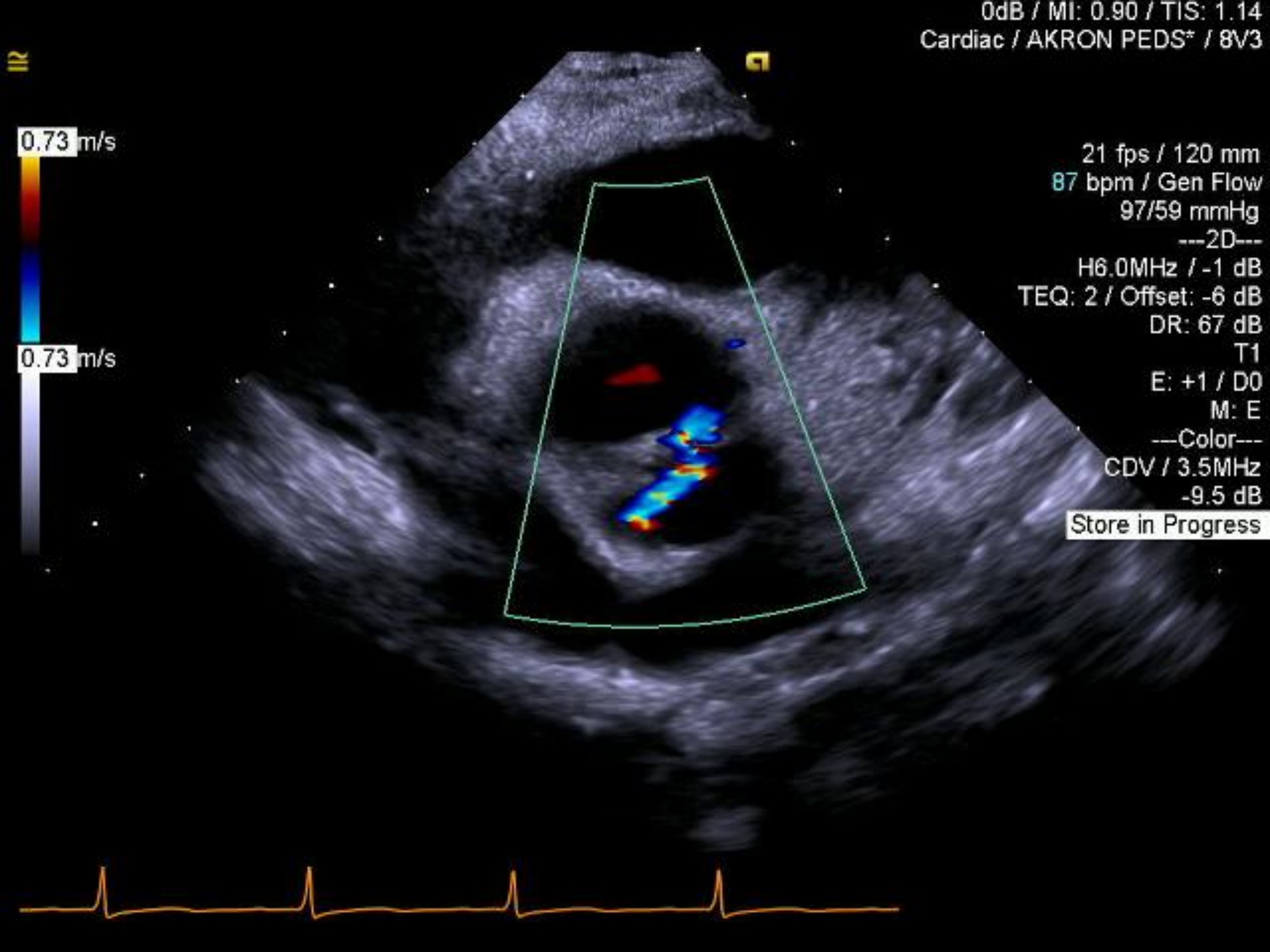

0dB / MI: 0.79 / TIS: 1.1
Cardiac / AKRON PEDS* / 8V
64 fps / 120 mm
90 bpm / Genera
97/59 mmHg
---2D--
H6.0MHz / -3 dB
TEQ: 2 / Offset: -8 dB
DR: 67 dB
T
E: +1 / D
M:
Store in Progres

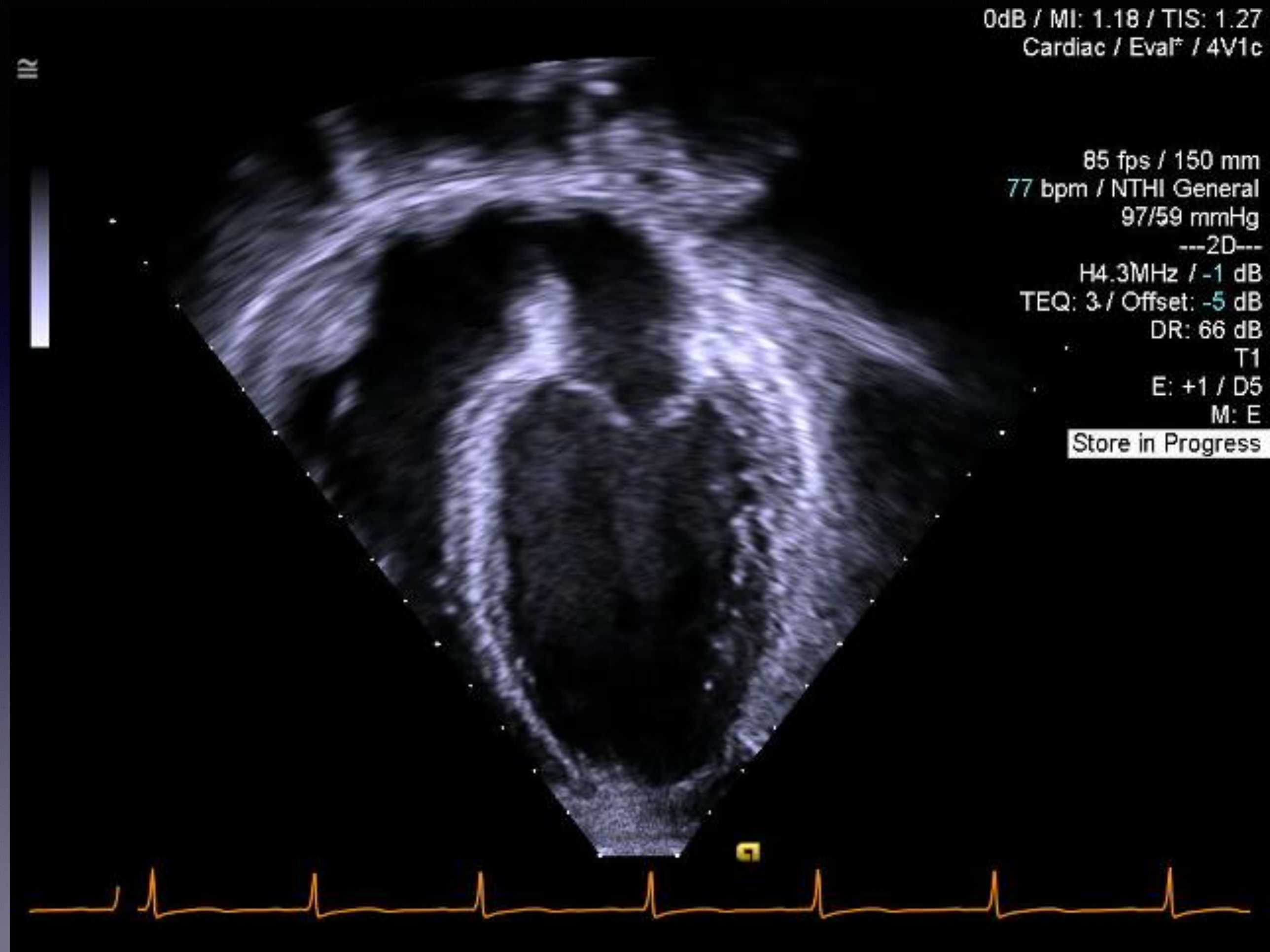

0dB / MI: 0.86 / TIS: 1.09
Cardiac / AKRON PEDS* / 8V3
111 fps / R 23.2 mm
88 bpm / General
97/59 mmHg
---2D---
H6.0MHz / -8 dB
TEQ: 2 / Offset: -6 dB
DR: 67 dB
T1
E: +1 / D0
M: E
Store in Progress

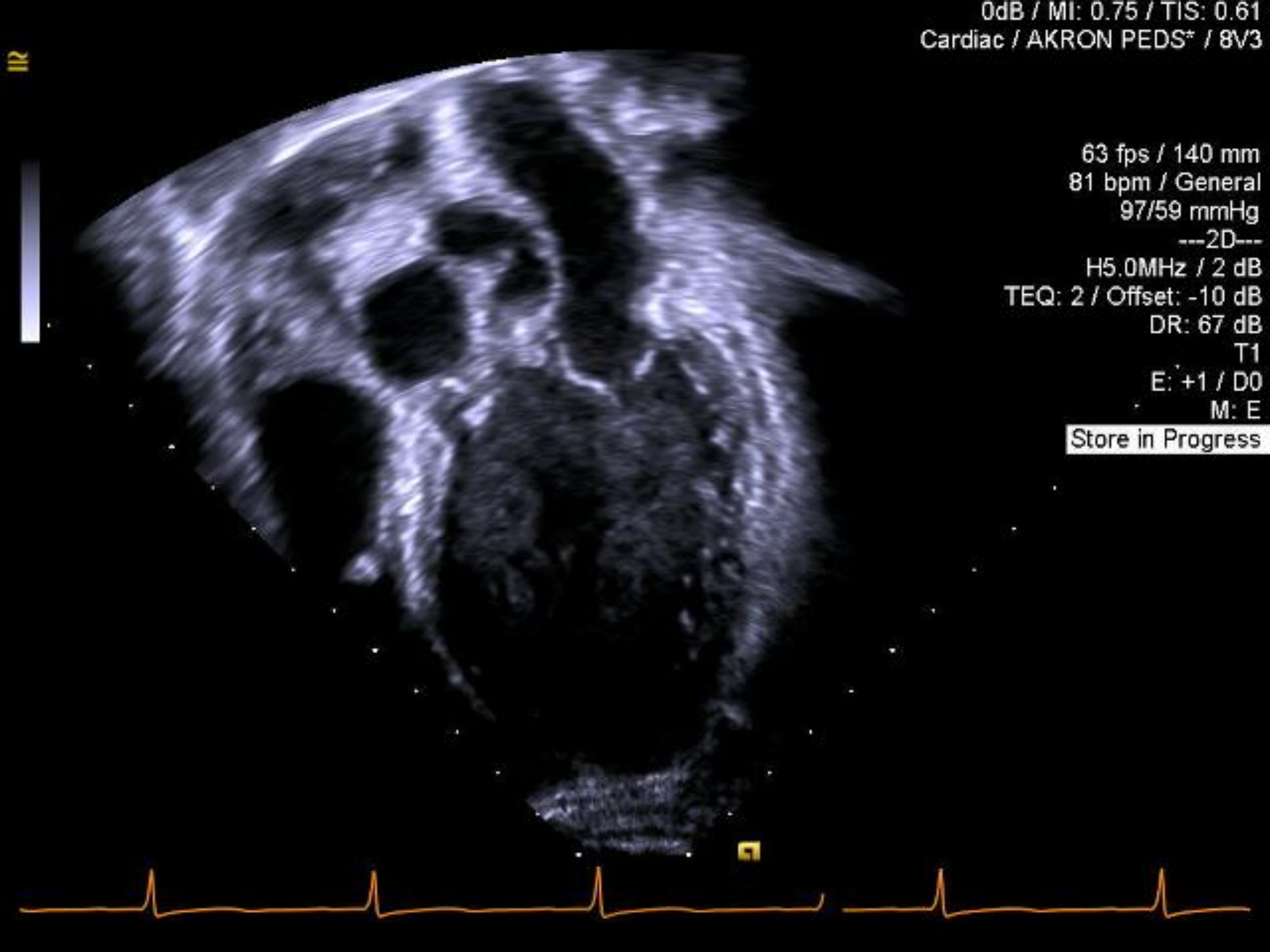

0dB / MI: 0.79 / TIS: 1.16
Cardiac / AKRON PEDS* / 8V3
64 fps / 120 mm
87 bpm / General
97/59 mmHg
---2D---
H6.0MHz / -1 dB
TEQ: 2 / Offset: -6 dB
DR: 67 dB
T1
E: +1 / D0
M: E
Store in Progress


0dB / MI: 1.00 / TIS: 1.11
Cardiac / AKRON PEDS* / 8V3
0.73 m/s
0.73 m/s
24 fps / R 23.2 mm
85 bpm / Gen Flow
97/59 mmHg
---2D---
H6.0MHz / -8 dB
TEQ: 2 / Offset: -6 dB
DR: 67 dB
T1
E: +1 / D0
M: E
---Color---
CDV / 3.5MHz
-9.5 dB
Store in Progress


Cardiac / AKRON PEDS* / 8V3
64 fps / 120 mm
89 bpm / General
97/59 mmHg
---2D---
H6.0MHz / -1 dB
TEQ: 2 / Offset: -6 dB
DR: 67 dB
T1
E: +1 / D0
M: E
Store in Progress


0dB / MI: 0.90 / TIS: 1.14
Cardiac / AKRON PEDS* / 8V3
0.73 m/s
0.73 m/s
21 fps / 120 mm
87 bpm / Gen Flow
97/59 mmHg
---2D---
H6.0MHz / -1 dB
TEQ: 2 / Offset: -6 dB
DR: 67 dB
T1
E: +1 / D0
M: E
---Color---
CDV / 3.5MHz
-9.5 dB
Store in Progress


0dB / MI: 1.18 / TIS: 1.27
Cardiac / Eval* / 4V1c
85 fps / 150 mm
77 bpm / NTHI General
97/59 mmHg
---2D---
H4.3MHz / -1 dB
TEQ: 3 / Offset: -5 dB
DR: 66 dB
T1
E: +1 / D5
M: E
Store in Progress


0dB / MI: 0.75 / TIS: 0.61
Cardiac / AKRON PEDS* / 8V3
63 fps / 140 mm
81 bpm / General
97/59 mmHg
---2D---
H5.0MHz / 2 dB
TEQ: 2 / Offset: -10 dB
DR: 67 dB
T1
E: +1 / D0
M: E
Store in Progress

0dB / MI: 1.18 / TIS: 1.07
Cardiac / Eval* / 4V1c

0.84 m/s

0.84 m/s

27 fps / 150 mm
81 bpm / Gen Flow
97/59 mmHg
---2D---
H4.3MHz / -1 dB
TEQ: 3 / Offset: -5 dB
DR: 66 dB
T1
E: +1 / D5
M: E
---Color---
CDV / 2.0MHz
-5.5 dB
Store in Progress

THE HEART CENTER-A SAINI
1/31/2019 4:02:56 PM

0dB / MI: 0.10 / TIS: 1.00
Cardiac / Eval* / 4V1c

1 AoV VTI = 0.810 m
AoV Vmax = 3.57 m/s
AoV Vmean = 2.64 m/s
AoV Peak Grad = 50.9 mmHg
AoV Mean Grad = 30.8 mmHg
AoV AT = 119 msec
AoV ET = 307 msec

0.84
97 mm
0°
0.84 m/s

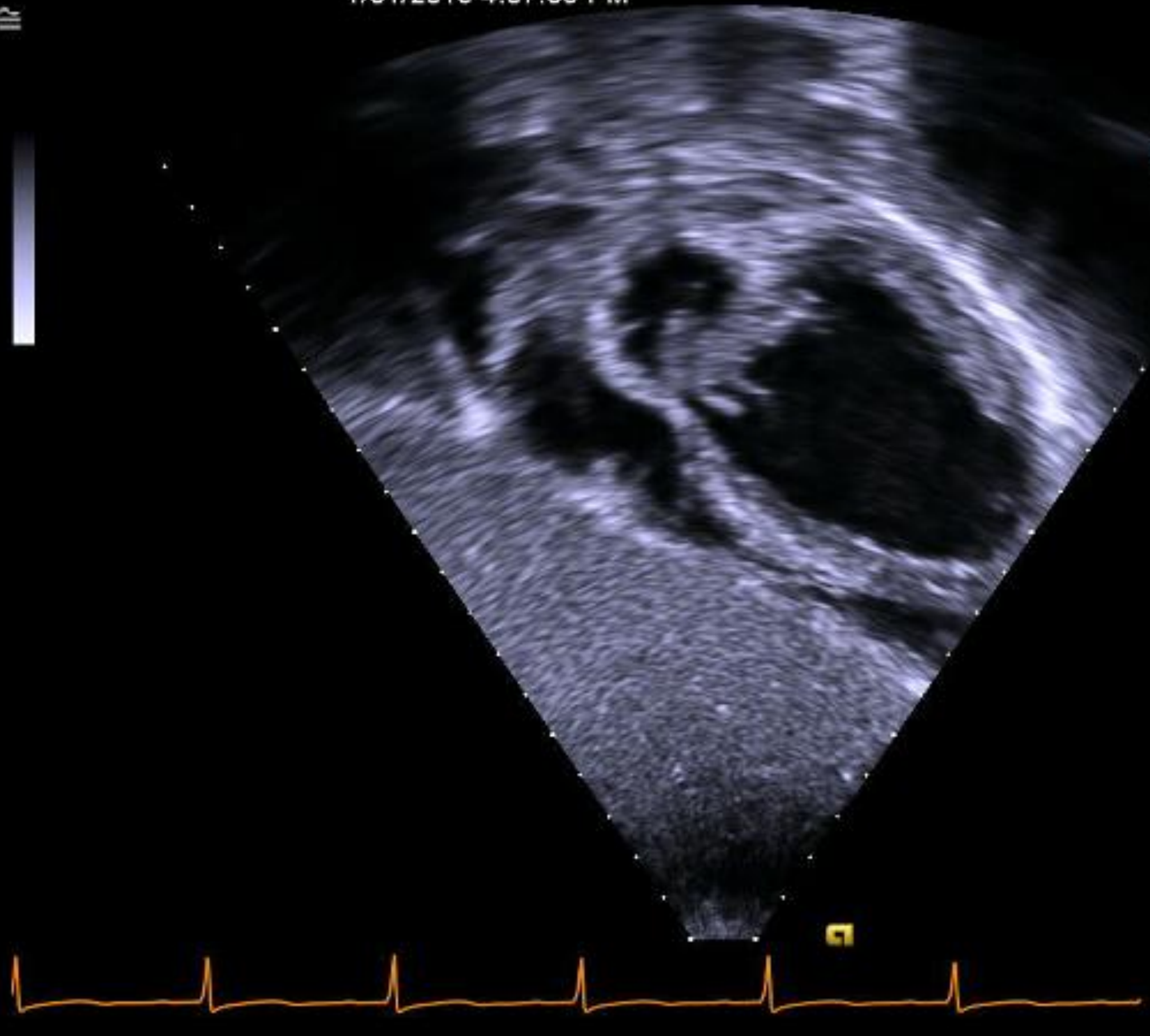
27 fps / 150 mm
78 bpm / General
97/59 mmHg
---2D---
H4.3MHz / -5 dB
TEQ: 3 / Offset: -9 dB
DR: 66 dB
T1
E: +1 / D5
M: E
---Color---
CDV / 2.0MHz
-5.5 dB
---CW---
1.75MHz
-4 dB / DR: 50
Sweep: 100
2

m/s
1.00
2.00
3.00
4.00

THE HEART CENTER-A SAINI
1/31/2019 4:07:39 PM

0dB / MI: 1.13 / TIS: 1.23
Cardiac / Eval* / 4V1c

79 fps / 190 mm
84 bpm / NTHI General
97/59 mmHg
---2D---
`H4.3MHz / 13 dB
TEQ: 3 / Offset: 9 dB
DR: 66 dB
T1
E: +1 / D5
M: E
Store in Progress




THE HEART CENTER-A SAINI
1/31/2019 4:10:53 PM
0dB / MI: 1.18 / TIS: 1.25
Cardiac / Eval* / 4V1c
59 fps / 210 mm
76 bpm / NTHI General
97/59 mmHg
---2D---
H4.3MHz / 17 dB
TEQ: 3 / Offset: 13 dB
DR: 66 dB
T1
E: +1 / D5
M: E
Store in Progress

Diagnosis…

(Drumroll please….)

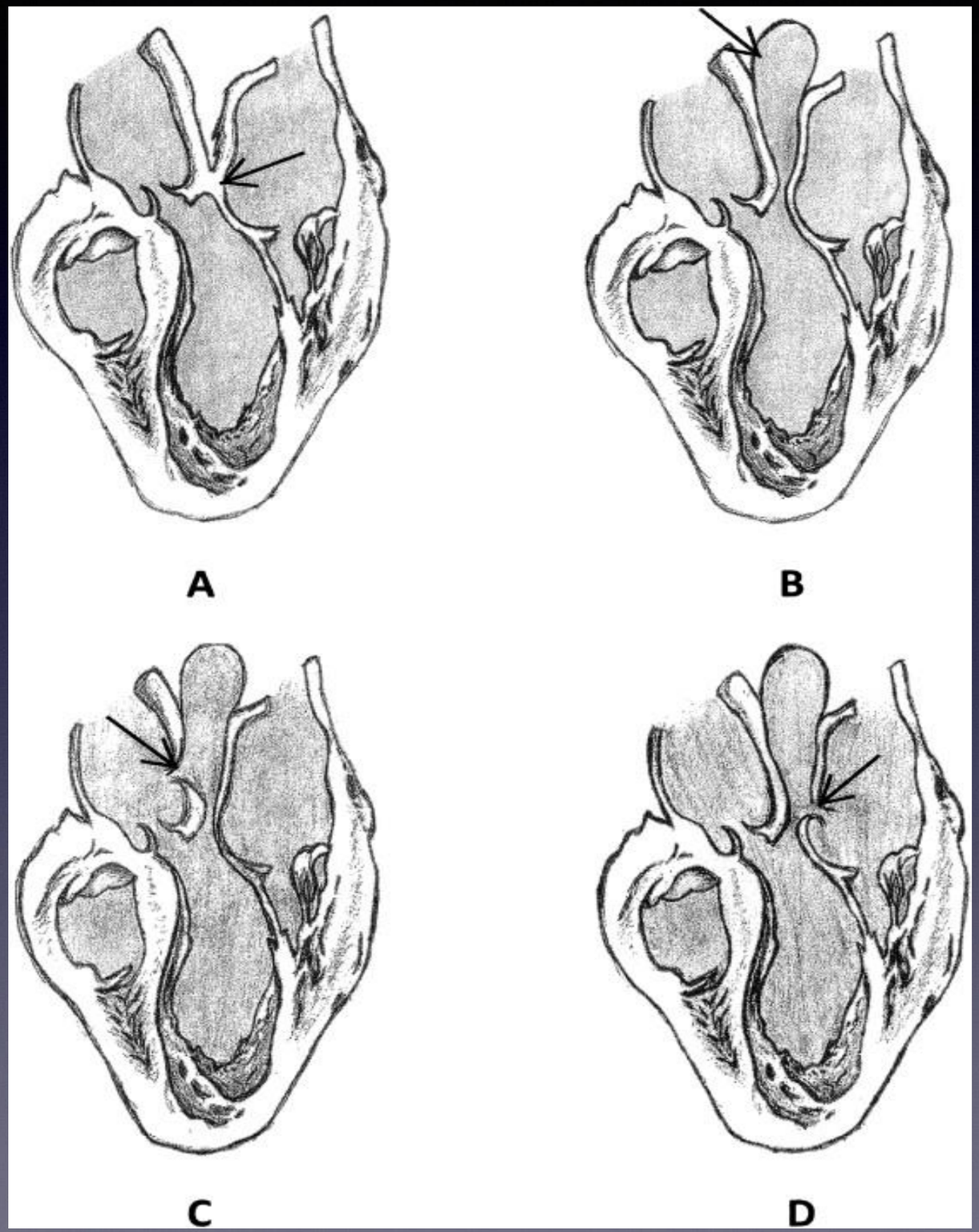
Pseudoaneurysm of Mitral-Aortic Intervalvular Fibrosa

Definition:

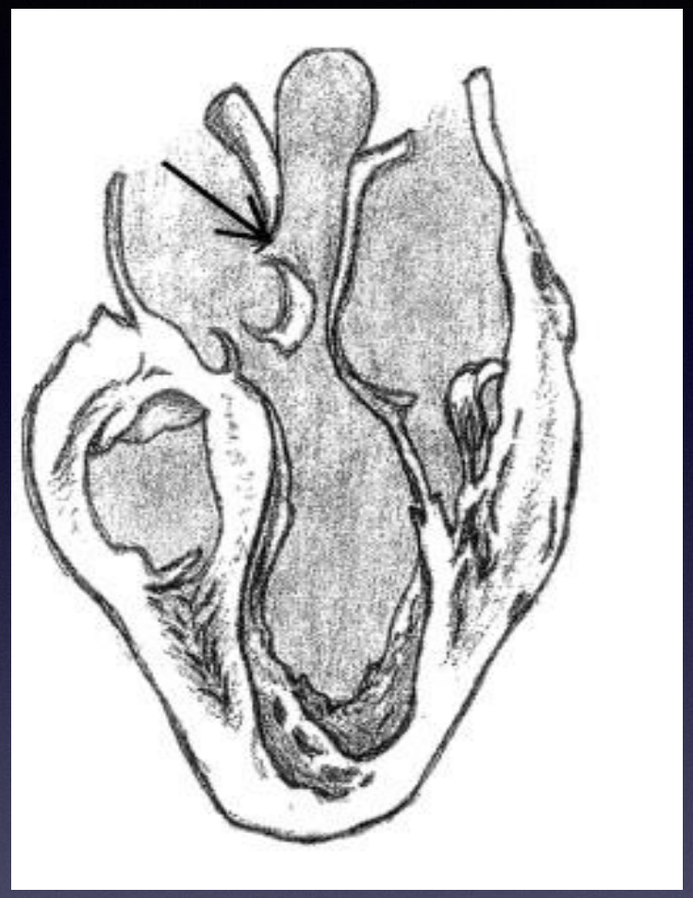
"Pseudoaneurysm at the interannular zone between the mitral and aortic valves and its communication with the left ventricular outflow tract between the left coronary or noncoronary aortic cusp and the anterior leaflet of the mitral valve."

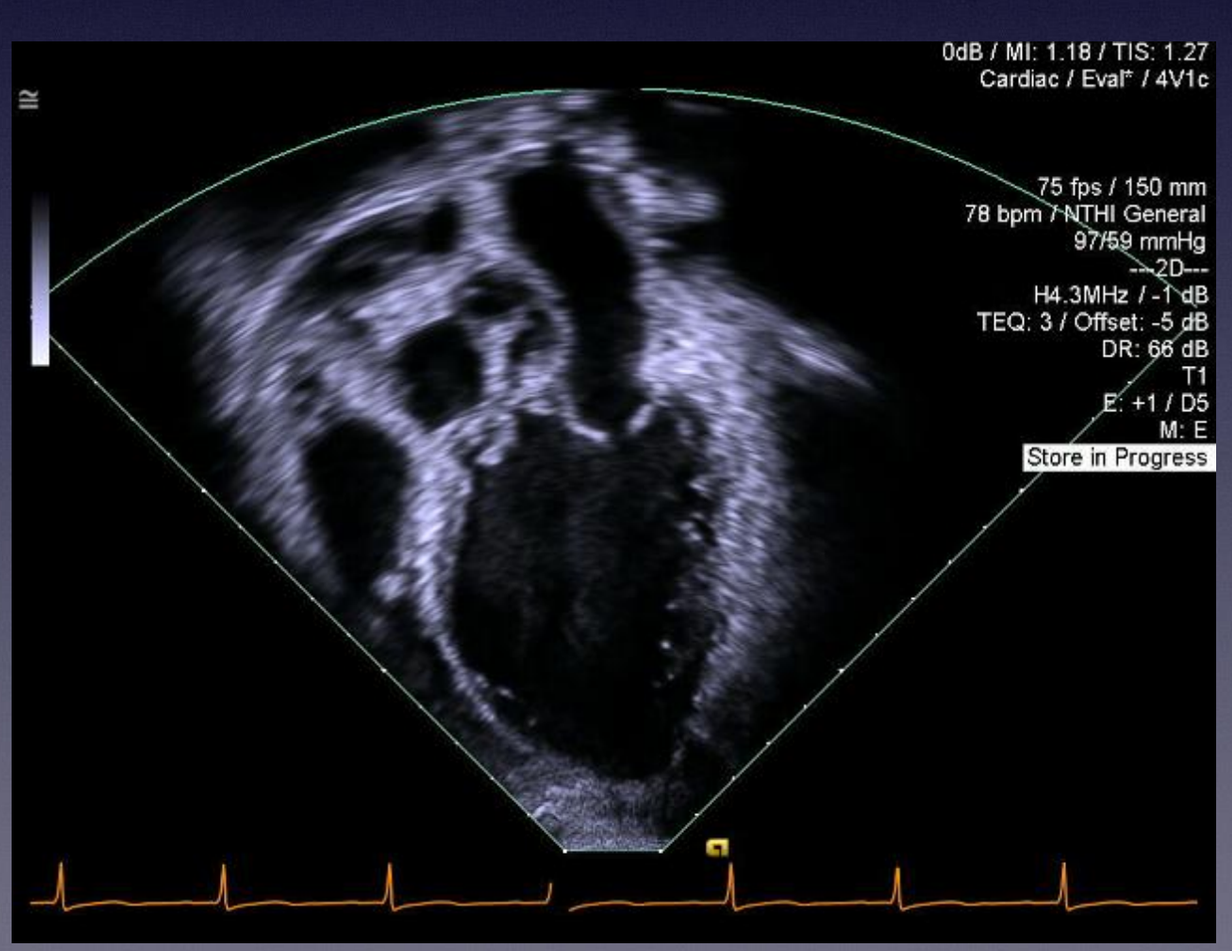
http://www.clinmedres.org/content/1/1/49/F2.expansion

Types of Pseudoaneurysm Mitral-Aortic Intervalvular Fibrosa

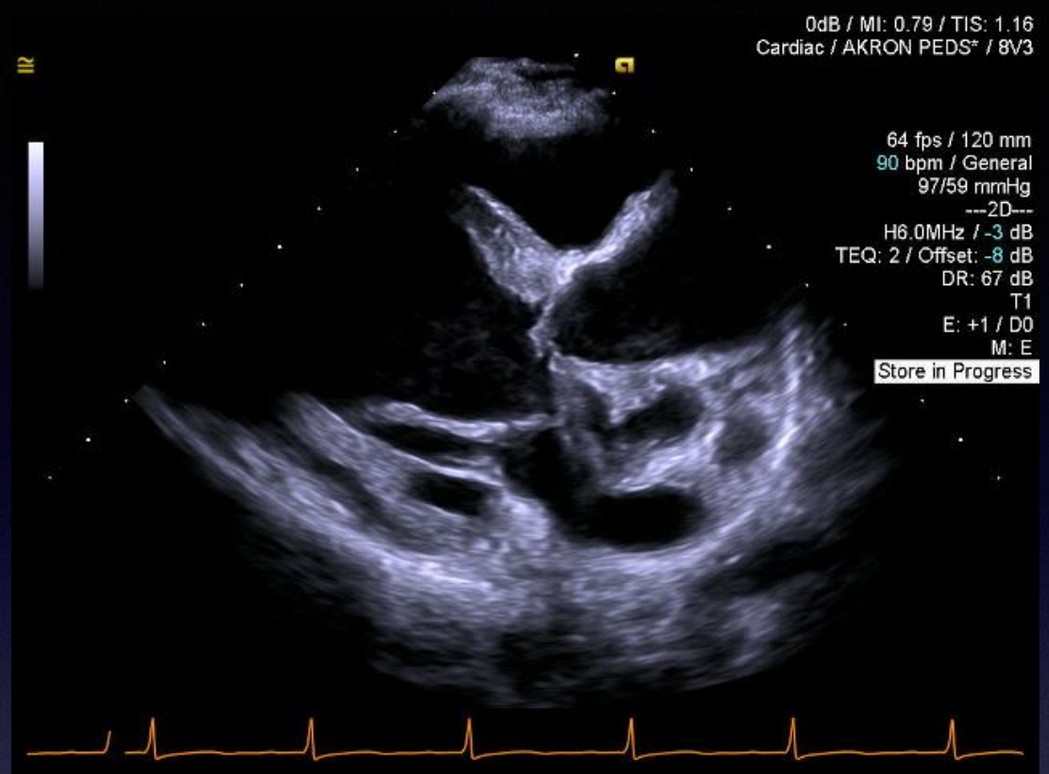


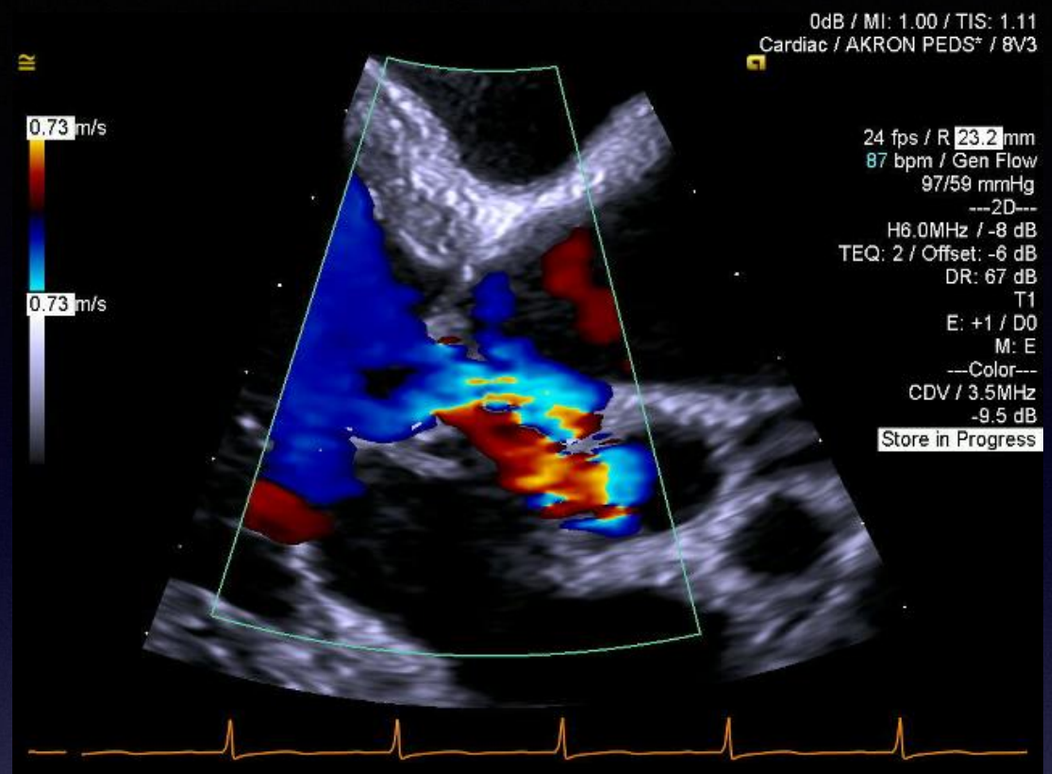
Pseudoaneurysm of the Mitral-Aortic Intervalvular Fibrosa with fistula to aorta

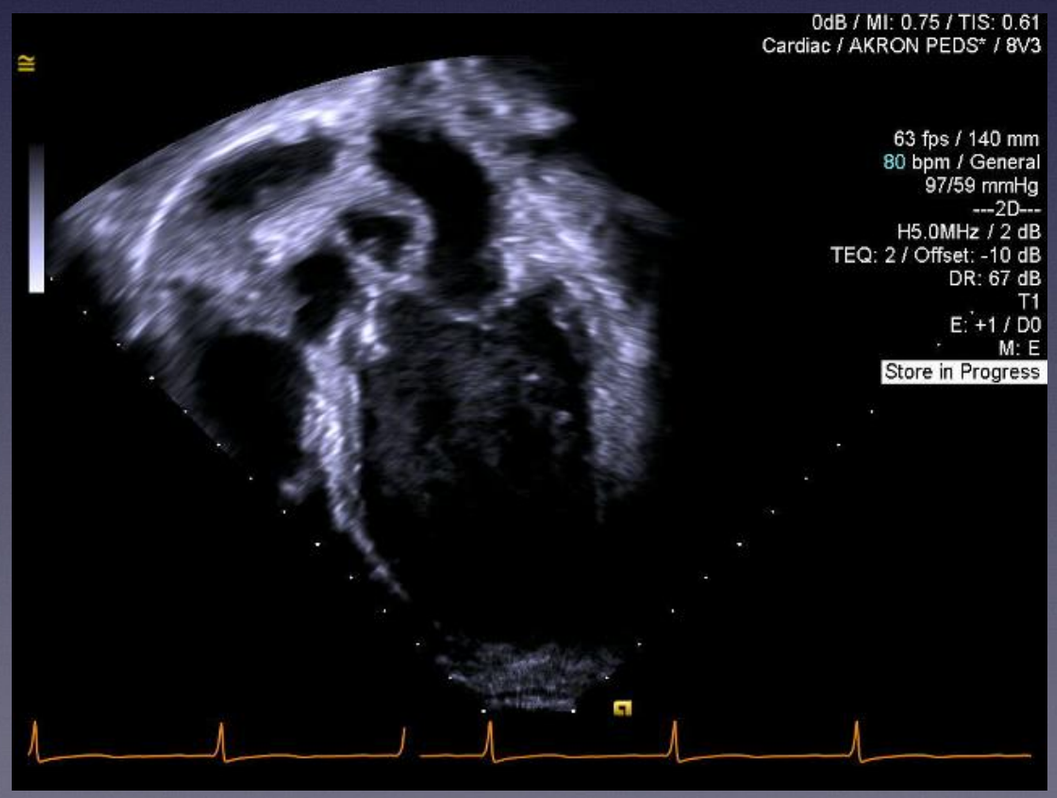




In review:


0dB / MI: 0.79 / TIS: 1.16
Cardiac / AKRON PEDS* / 8V3
64 fps / 120 mm
90 bpm / General
97/59 mmHg
---2D---
H6.0MHz / -3 dB
TEQ: 2 / Offset: -8 dB
DR: 67 dB
T1
E: +1 / D0
M: E
Store in Progress


0dB / MI: 1.00 / TIS: 1.11
Cardiac / AKRON PEDS* / 8V3
0.73 m/s
0.73 m/s
24 fps / R 23.2 mm
87 bpm / Gen Flow
97/59 mmHg
---2D---
H6.0MHz / -8 dB
TEQ: 2 / Offset: -6 dB
DR: 67 dB
T1
E: +1 / D0
M: E
---Color---
CDV / 3.5MHz
-9.5 dB
Store in Progress


0dB / MI: 0.75 / TIS: 0.61
Cardiac / AKRON PEDS* / 8V3
63 fps / 140 mm
80 bpm / General
97/59 mmHg
---2D---
H5.0MHz / 2 dB
TEQ: 2 / Offset: -10 dB
DR: 67 dB
T1
E: +1 / D0
M: E
Store in Progress


0dB / MI: 1.18 / TIS: 1.07
Cardiac / Eval* / 4V1c
0.84 m/s
0.84 m/s
27 fps / 150 mm
81 bpm / Gen Flow
97/59 mmHg
---2D---
H4.3MHz / -1 dB
TEQ: 3 / Offset: -5 dB
DR: 66 dB
T1
E: +1 / D5
M: E
---Color---
CDV / 2.0MHz
-5.5 dB
Store in Progress

- In a literature review of relevant articles published from 1966 to 2009, Sudhakar et al, identified 89 patients with pseudoaneurysm of the mitral-aortic intervalvular fibrosa.

- Endocarditis and aortic valve surgery were the two most frequently associated causative factors.

- Aortic regurgitation has been suggested as a contributing factor for the development of pseudoaneurysm of the mitral-aortic intervalvular fibrosa.

- Complications of Pseudoaneurysm of mitral-aortic intervalvular fibrosa:
  - Compression; coronary artery, pulmonary artery, or mitral valve
  - Fistula formation; between aorta or left atrium
  - Rupture; pericardial tamponade, death
  - Thrombosis; TIA or CVA
  - Infection; endocarditis, fistula formation
  - Heart failure

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