Pacing Therapy in Infants with Congenital Complete Heart Block in Hong Kong

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Background

- Data in pacing therapy in infants with congenital heart block are limited
- Aim to share our experience in the single tertiary centre in HK

Methodology

- All infants (aged <1) diagnosed with **congenital heart block**, with pacing therapy were recruited
- 2006-2018
- The following data were collected:
  - patient demographics
  - clinical progress and medication
  - pacing related parameters: pacing mode, pacing site, lead related complication
  - ECG parameters: escape rate, QRS duration
  - Echocardiographic data: LVIDD (z-score), LVFS (m-mode)

Results

- N= 8 (M:F = 5:3)
- FU duration: 77.3 months (IQR 35.8-136.2)
- Prenatal diagnosis: 6 (75%)
- Indication for pacing therapy:
  - evidence of low cardiac output (n=2)
  - asymptomatic extreme bradycardia (n=6)
- Mean age of first pacemaker implantation = 9.9 days (range 1-38)
- Mean escape ventricular rate = 53.8 +/- 6.2 bpm
- QRS duration 76.3 +/- 27.8 ms (wide escape = 1)

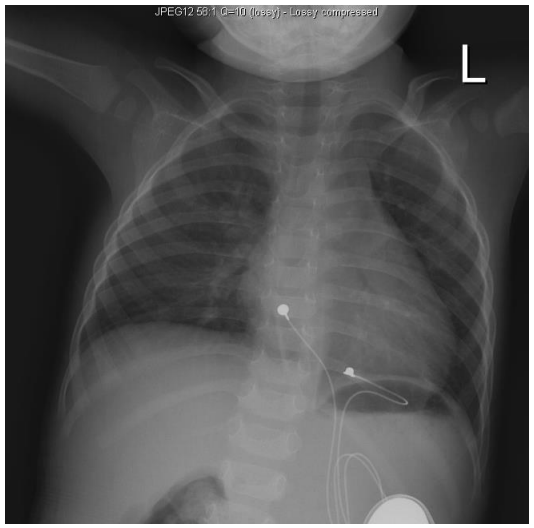
Pacing parameters

- All epicardial dual chamber system
- DDD 80-180 at initial programming
- 2006-2010 RV lead implantation; 2010-2018 LV lead implantation
- 3 patients had epicardial lead fracture, median age of 108 months (range 45-129)

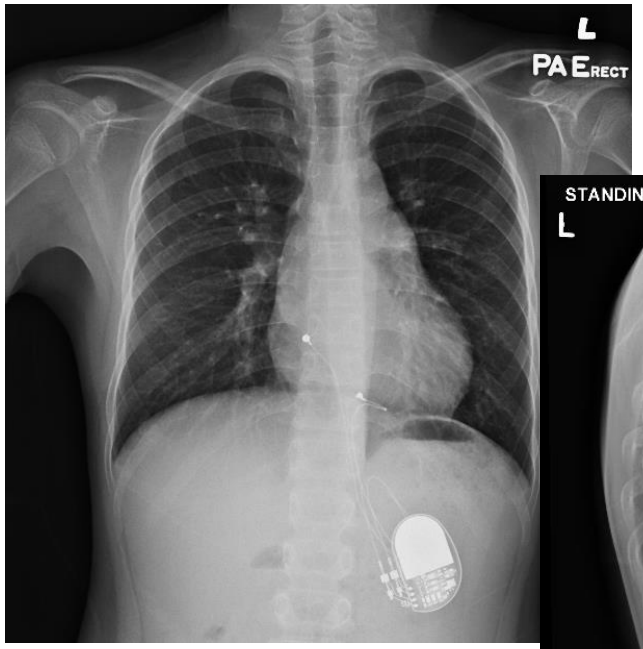
RV pacing

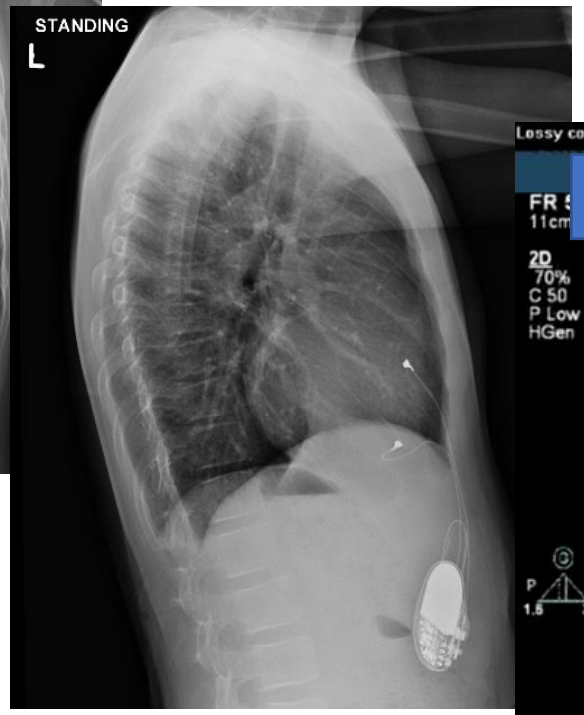
RV apical pacing

RV pacing 1 – no heart failure/ LV dysfunction since implant

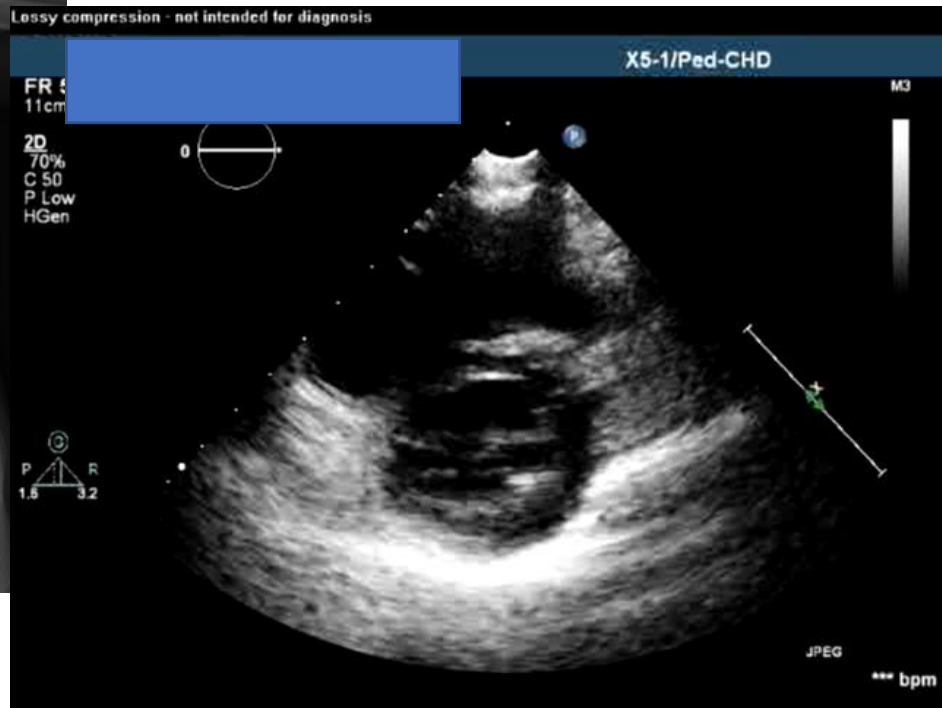


Initial - 2006





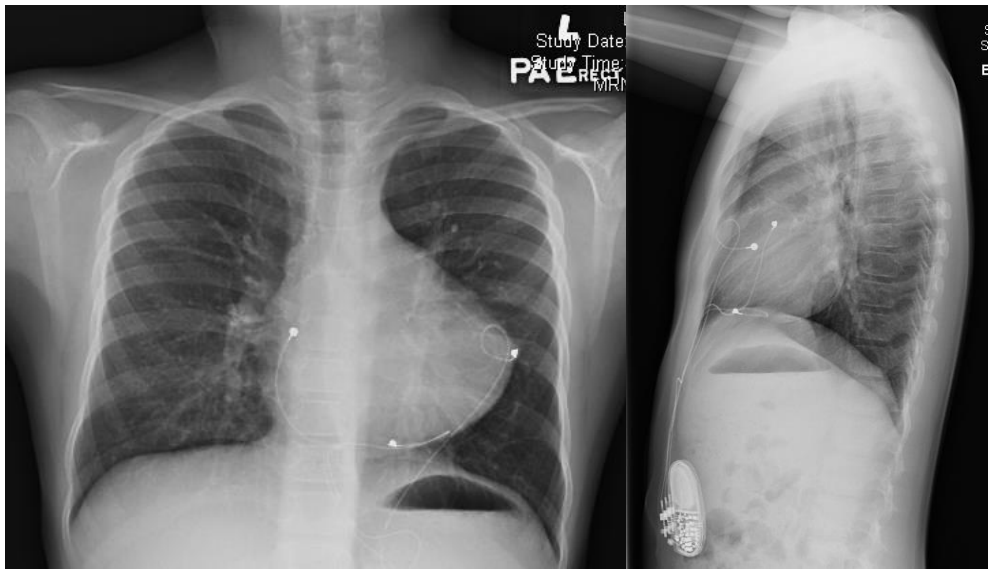
Latest film 2019

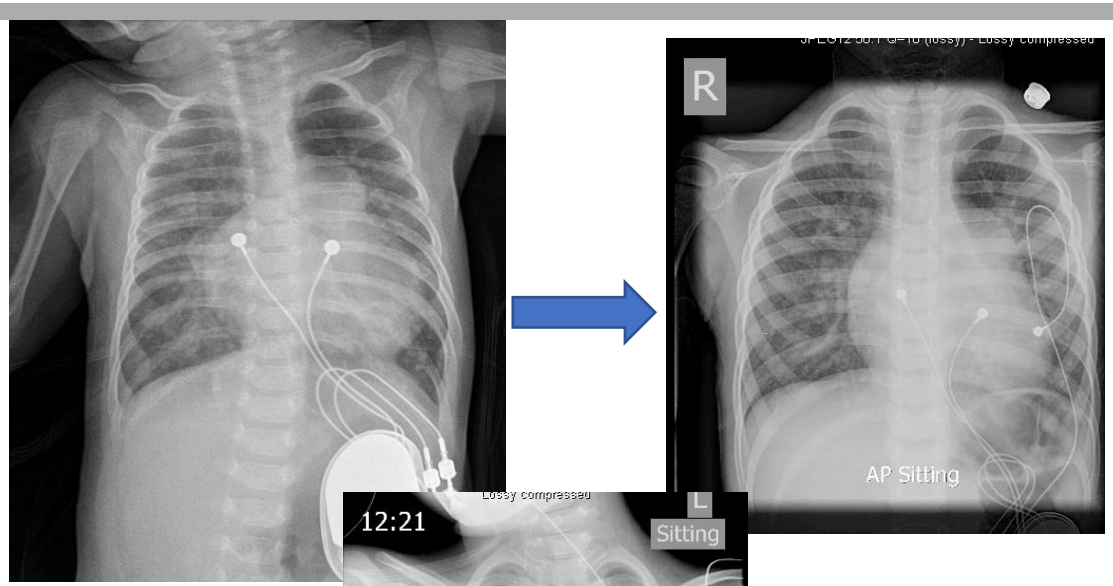


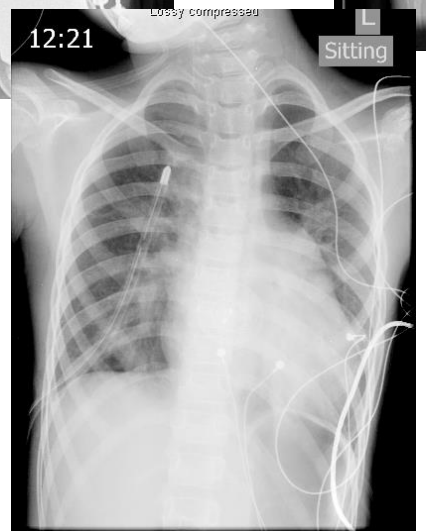
RV pacing 2 & 3 – develop heart failure @ 3-month → CRT

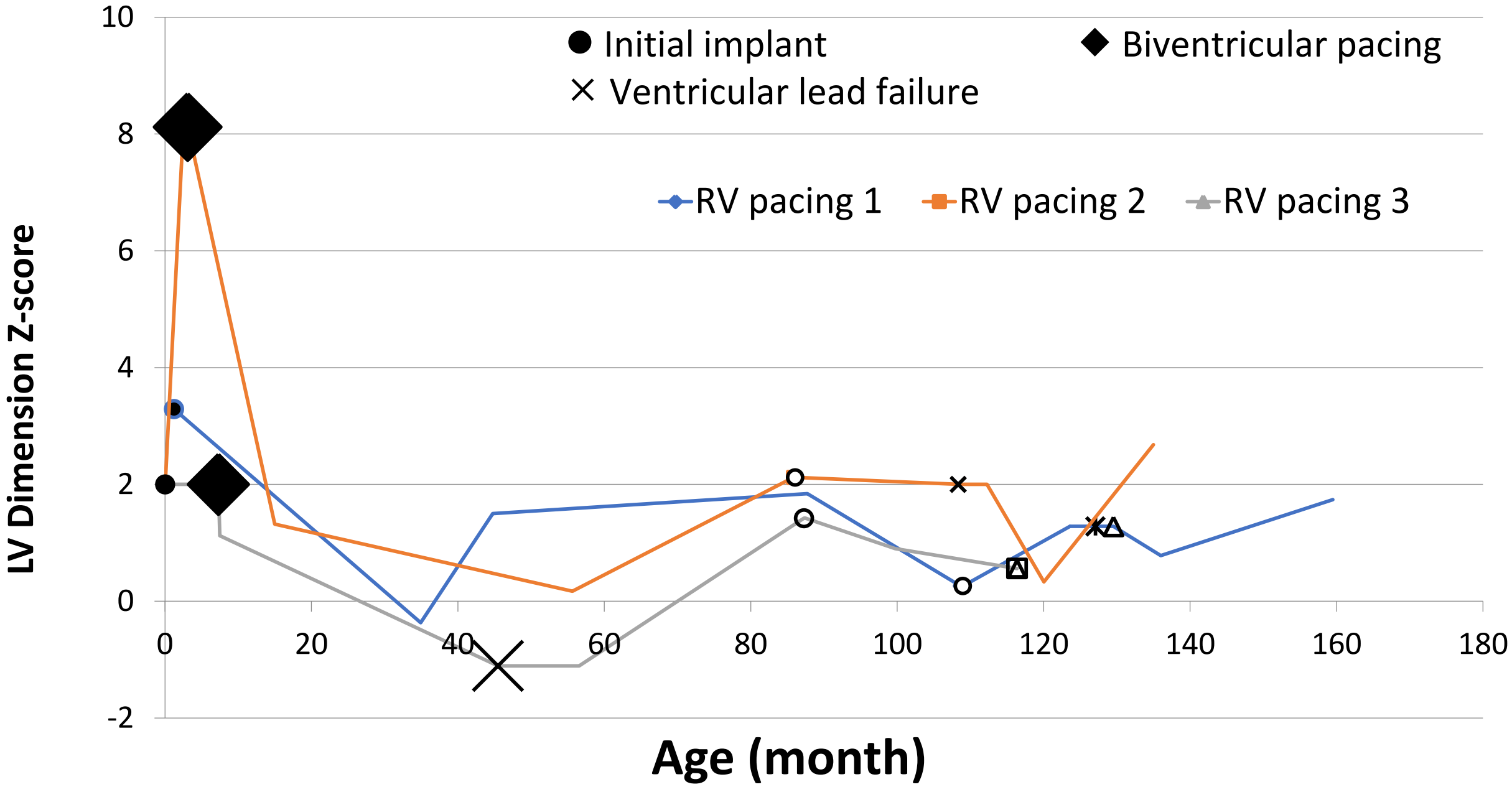
RV apical pacing

? RVOT pacing





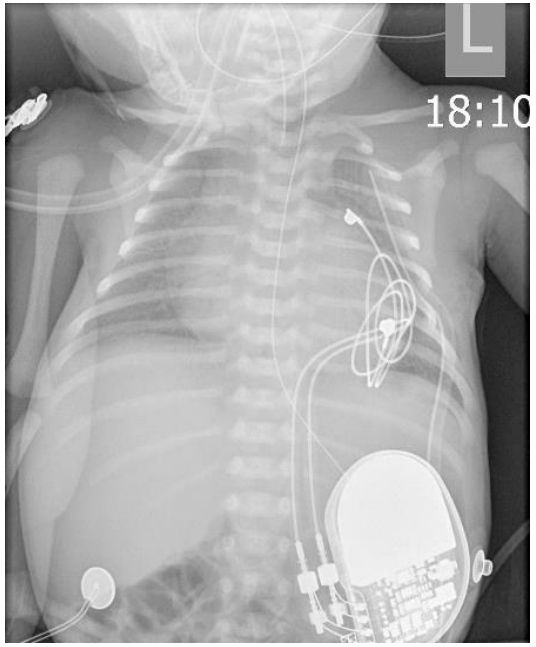


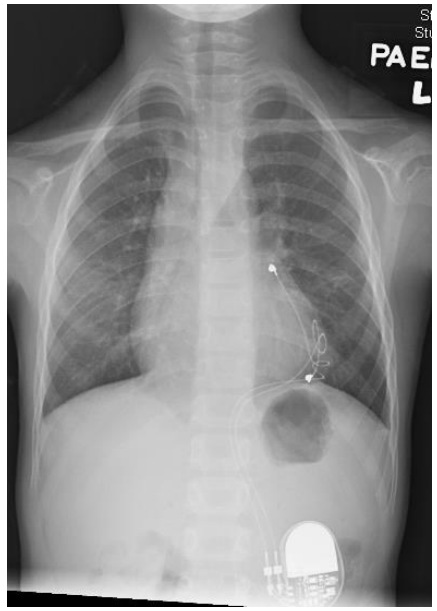

Initial implant
Biventricular pacing
Ventricular lead failure
RV pacing 1
RV pacing 2
RV pacing 3
LV Dimension Z-score
10
8
6
4
2
0
-2
0
20
40
60
80
100
120
140
160
180
Age (month)

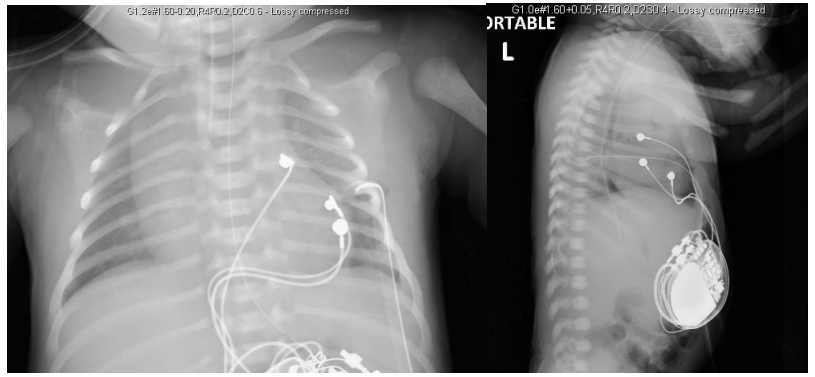
LV pacing

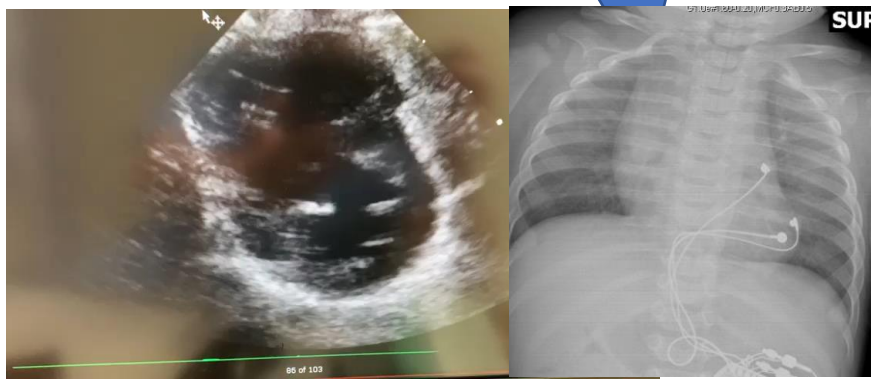
LV apical pacing

LV apical pacing – no heart failure/ LV dysfunction since implant

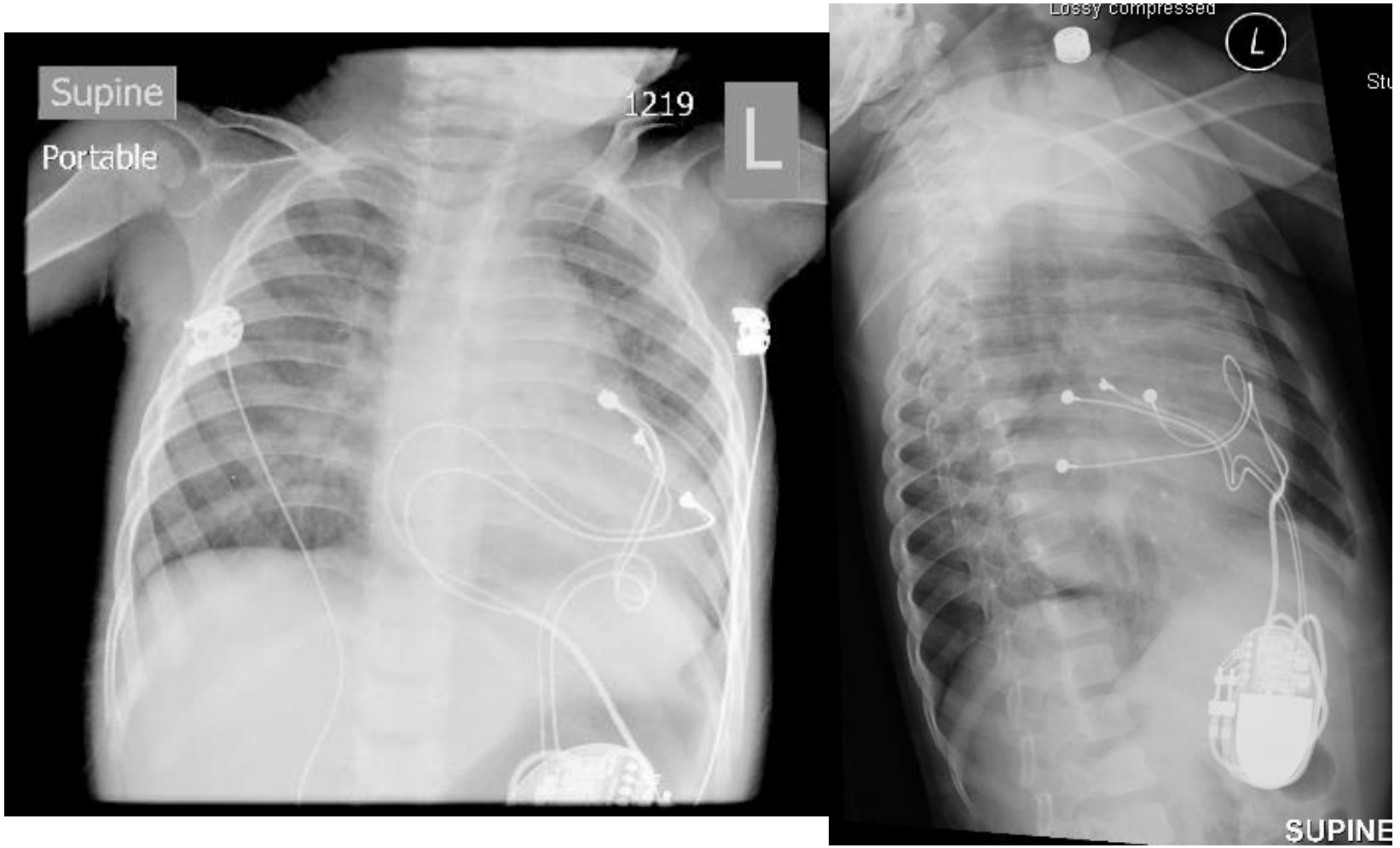




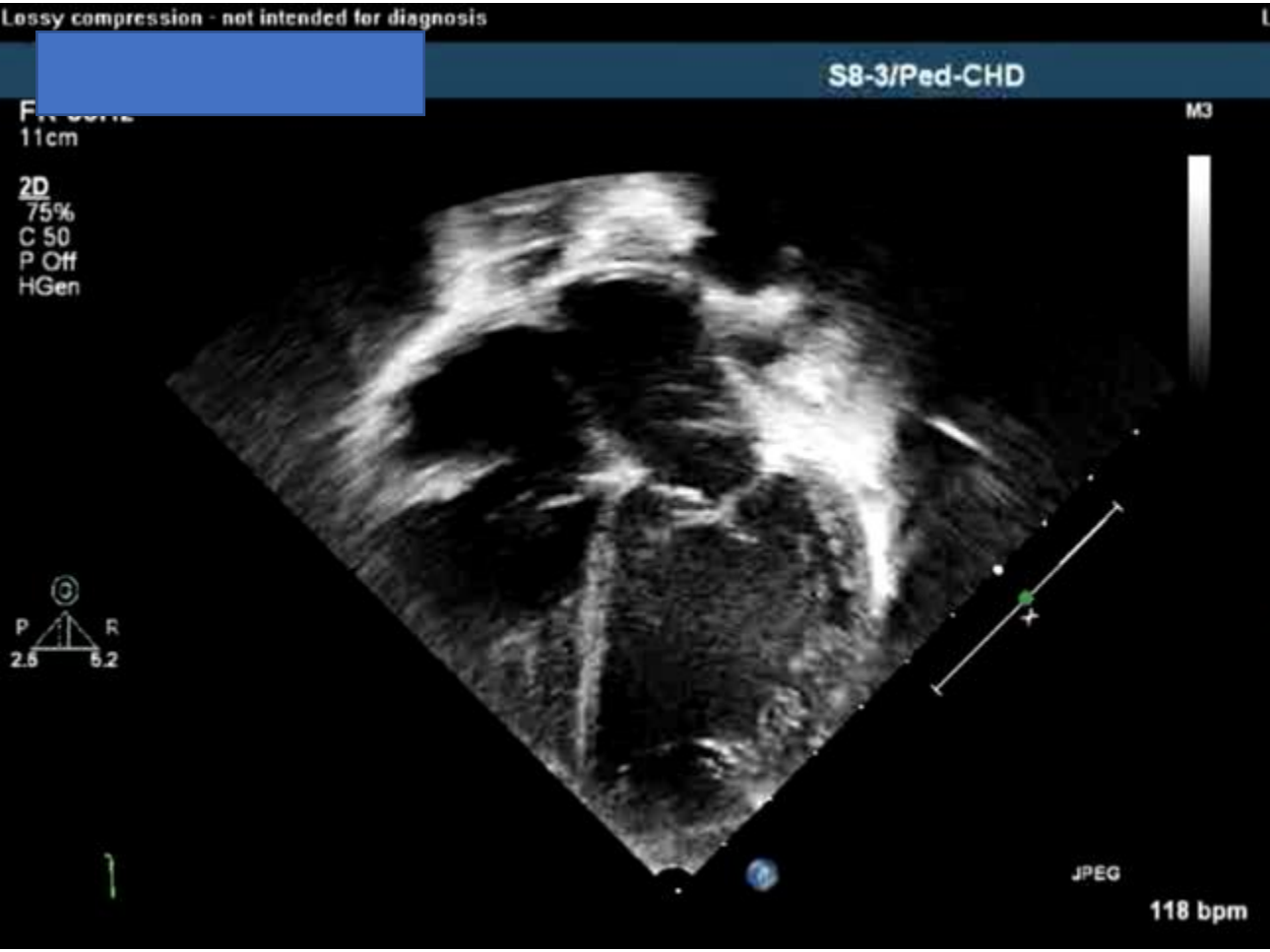


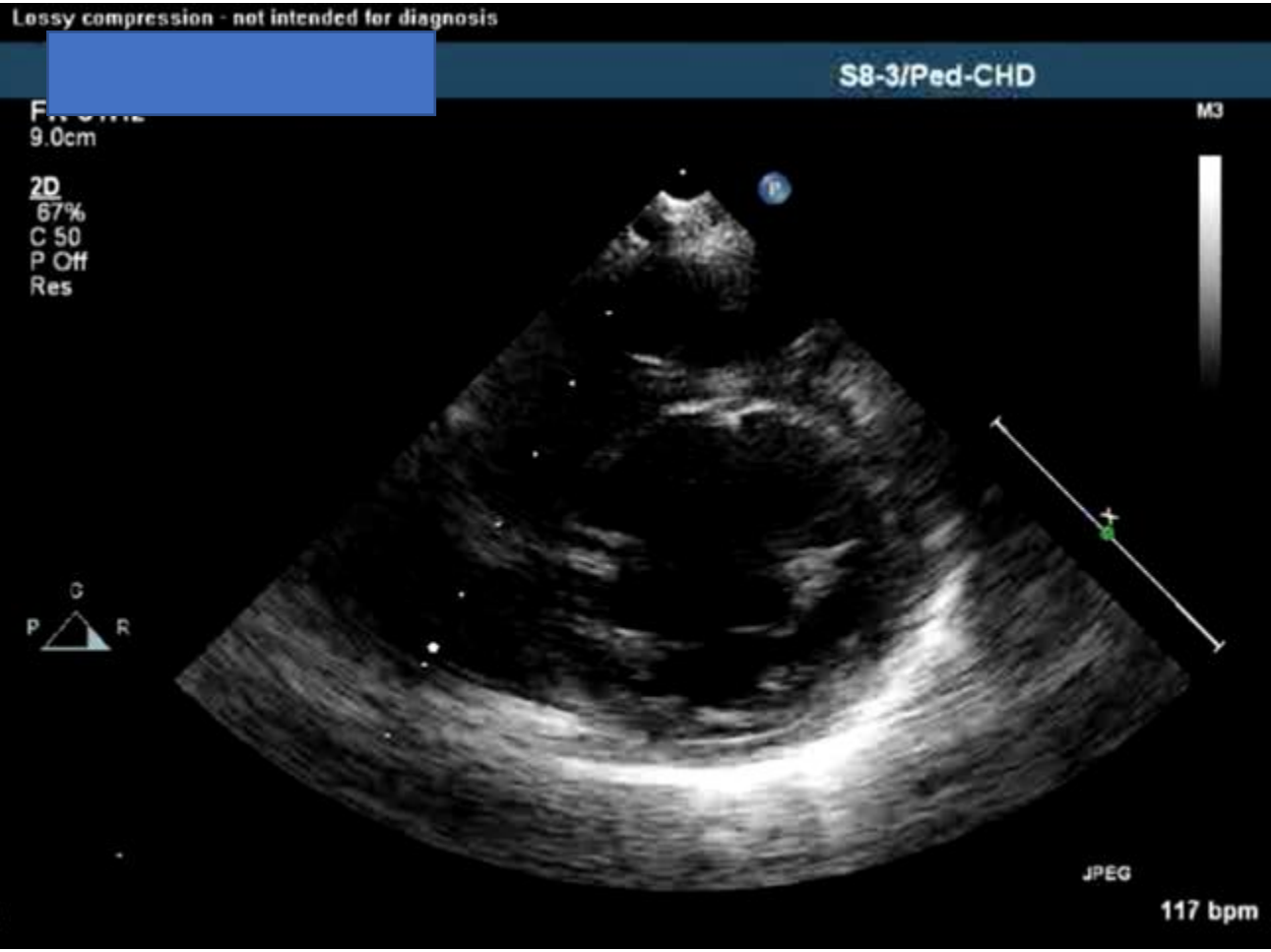


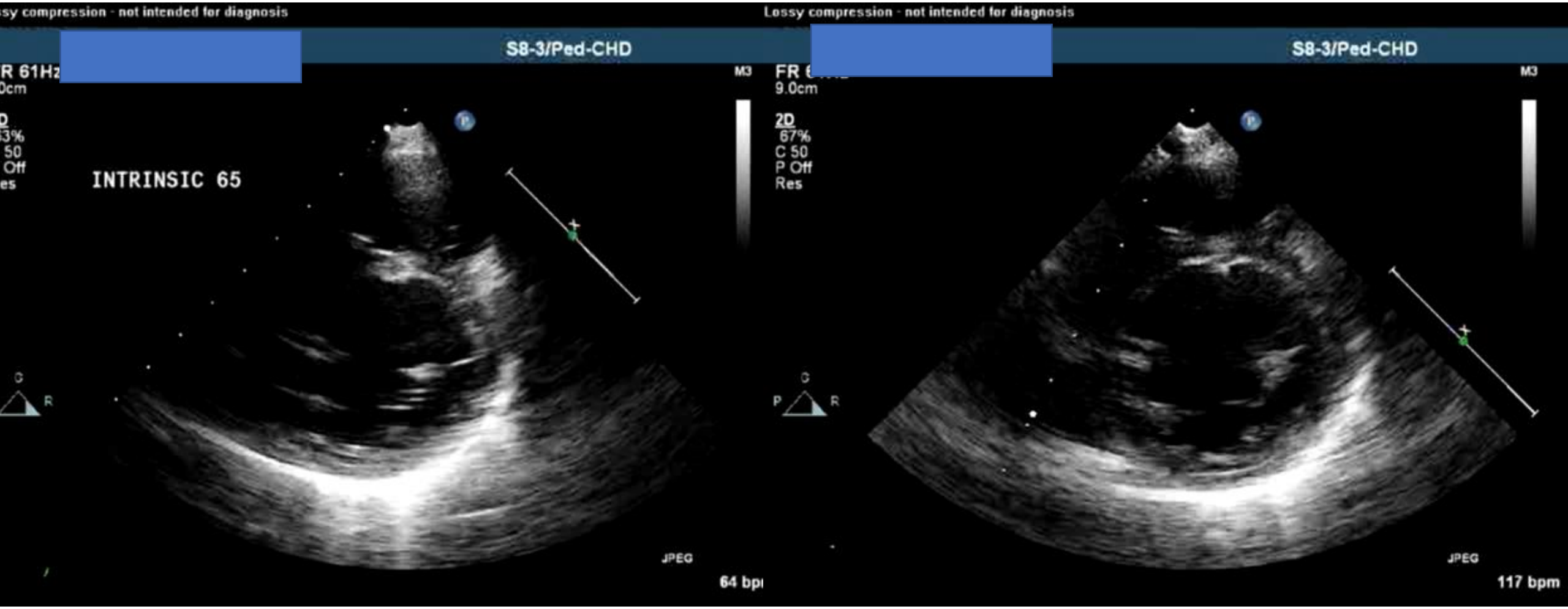
LV basal pacing 1
[one of unipolar lead at RVOT – because of difficult placement]



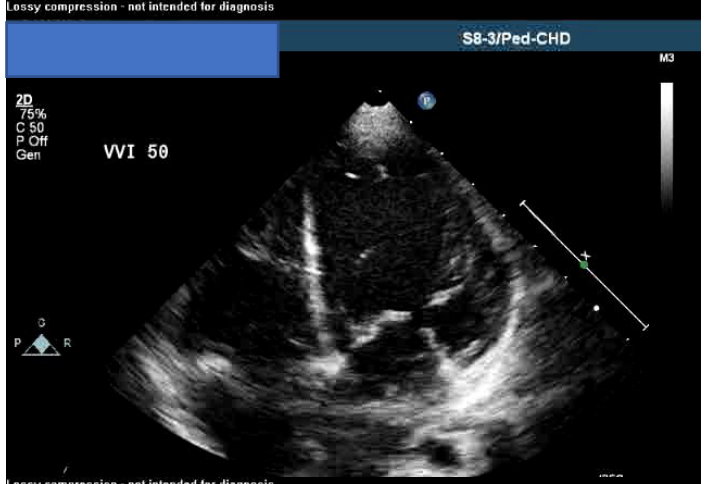
9 month
Holter:
average 145/min

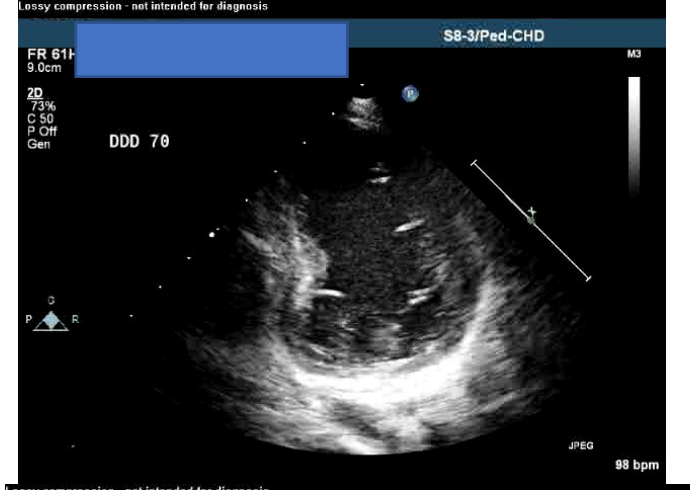

Lossy compression - not intended for diagnosis
S8-3/Ped-CHD
11cm
M3
2D
75%
C 50
P Off
HGen
P
R
2.5
5.2
JPEG
118 bpm

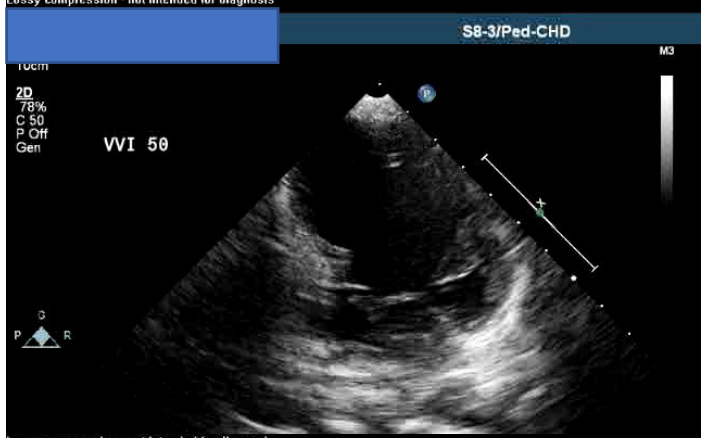

Lossy compression - not intended for diagnosis
S8-3/Ped-CHD
9.0cm
M3
2D
67%
C 50
P Off
Res
C
P
R
JPEG
117 bpm

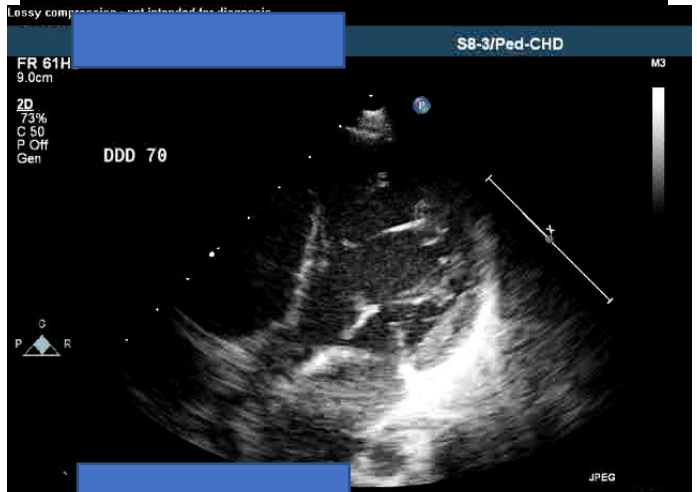


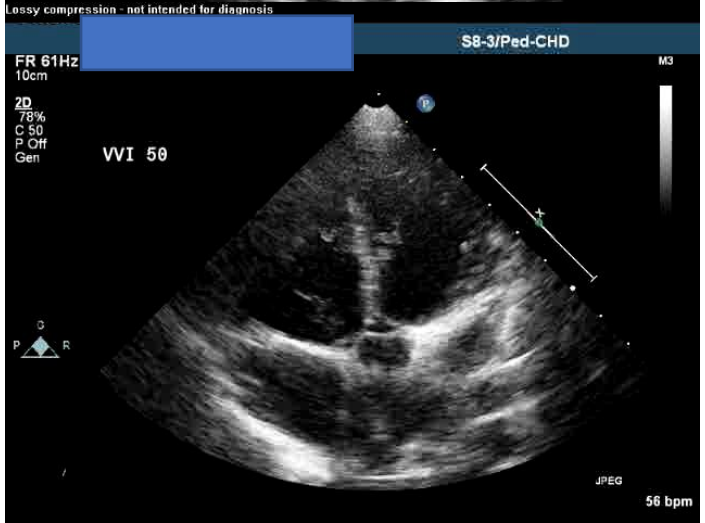
Off DDD to VVI backup pacing

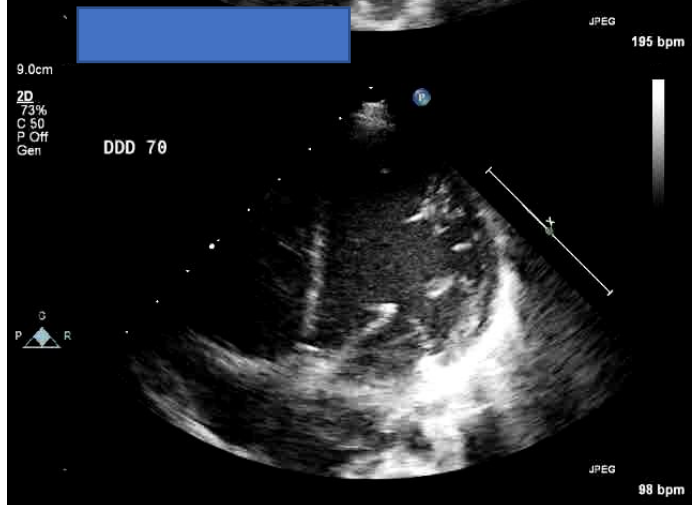

Lossy compression - not intended for diagnosis
S8-3/Ped-CHD
M3
2D
75%
C 50
P Off
Gen
VVI 50

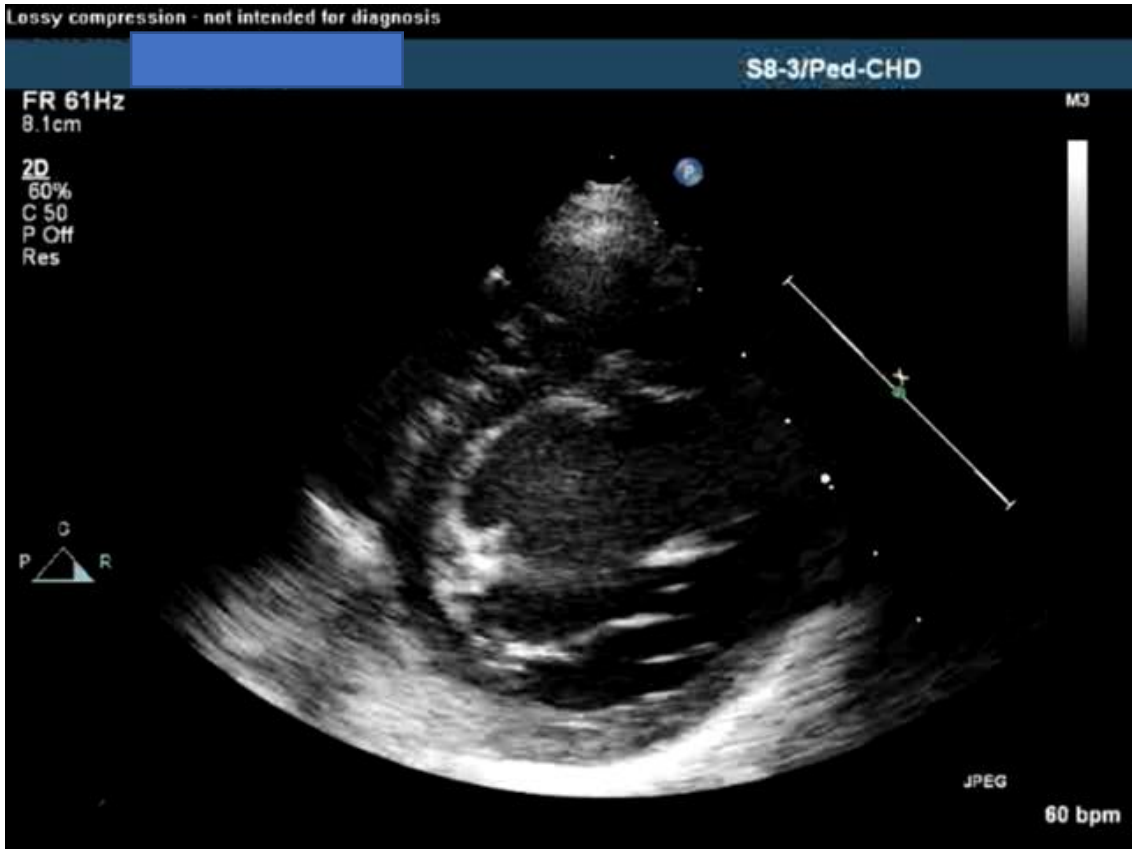

Lossy compression - not intended for diagnosis
S8-3/Ped-CHD
M3
9.0cm
2D
73%
C 50
P Off
Gen
DDD 70
JPEG
98 bpm

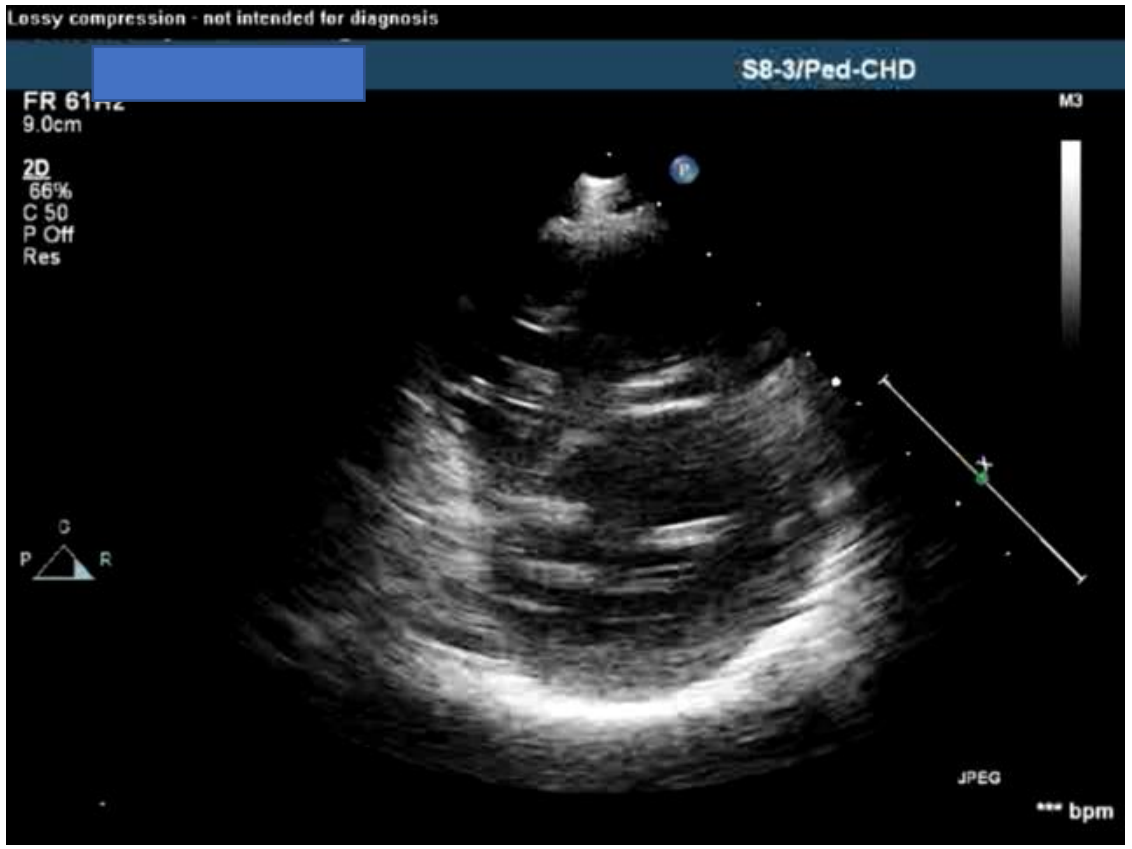

Lossy compression - not intended for diagnosis
S8-3/Ped-CHD
M3
2D
78%
C 50
P Off
Gen
VVI 50


Lossy compression - not intended for diagnosis
S8-3/Ped-CHD
M3
9.0cm
2D
73%
C 50
P Off
Gen
DDD 70
JPEG
195 bpm


Lossy compression - not intended for diagnosis
S8-3/Ped-CHD
M3
FR 61Hz
10cm
2D
78%
C 50
P Off
Gen
VVI 50
JPEG
56 bpm


9.0cm
2D
73%
C 50
P Off
Gen
DDD 70
JPEG
98 bpm





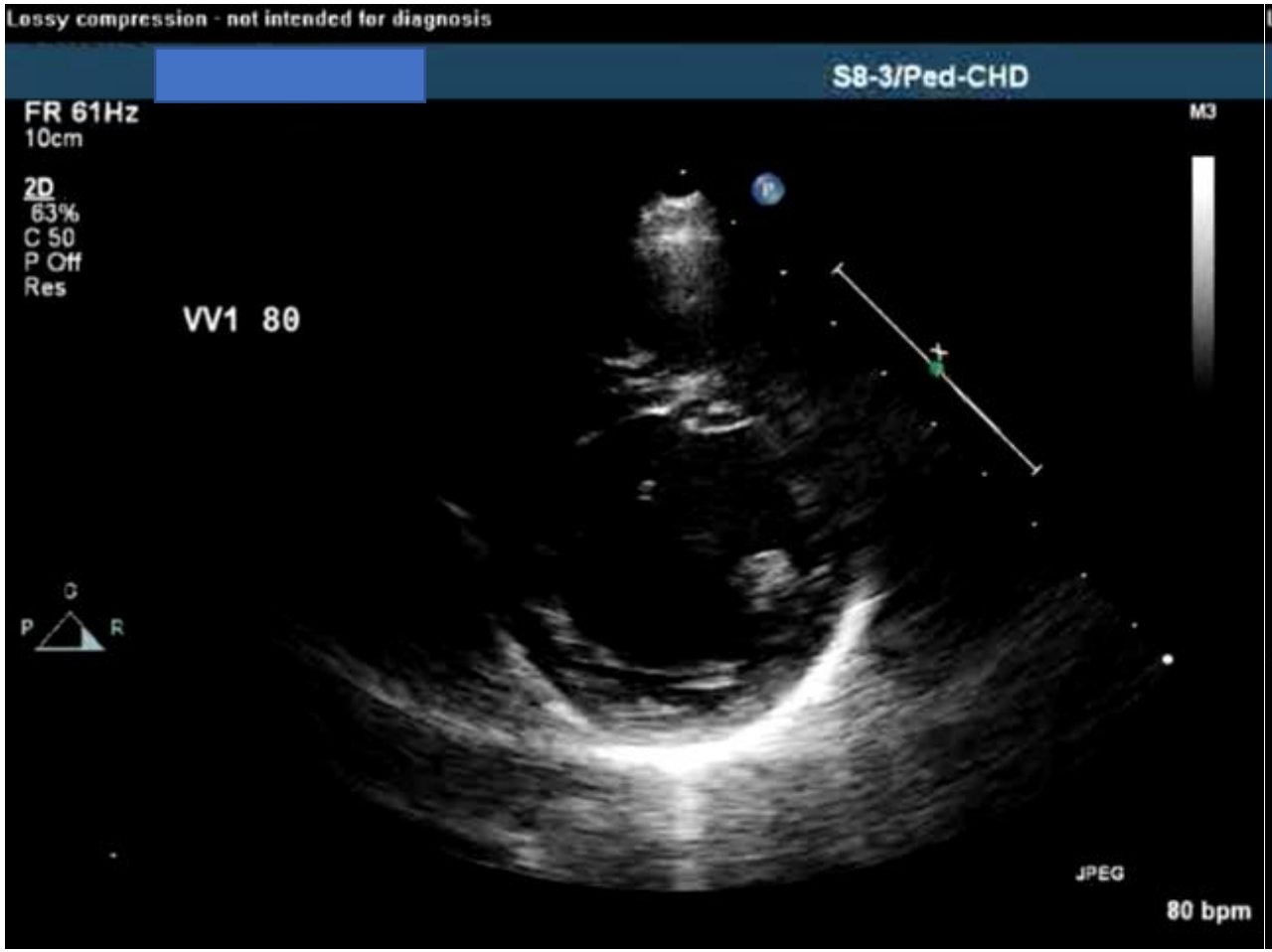
1 month later

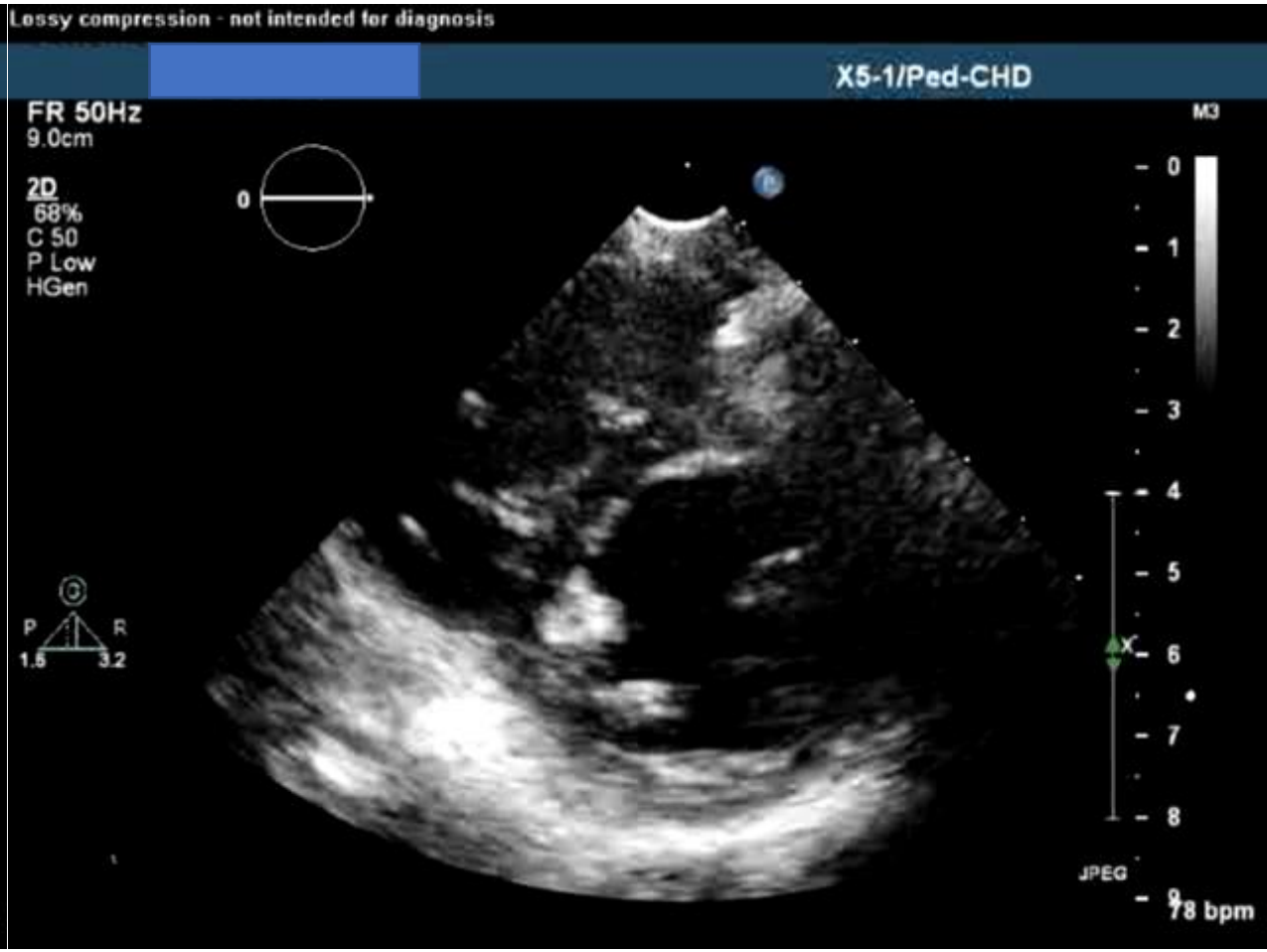
3 month later

VP ~ 20-30 %

? Pacing induced CM

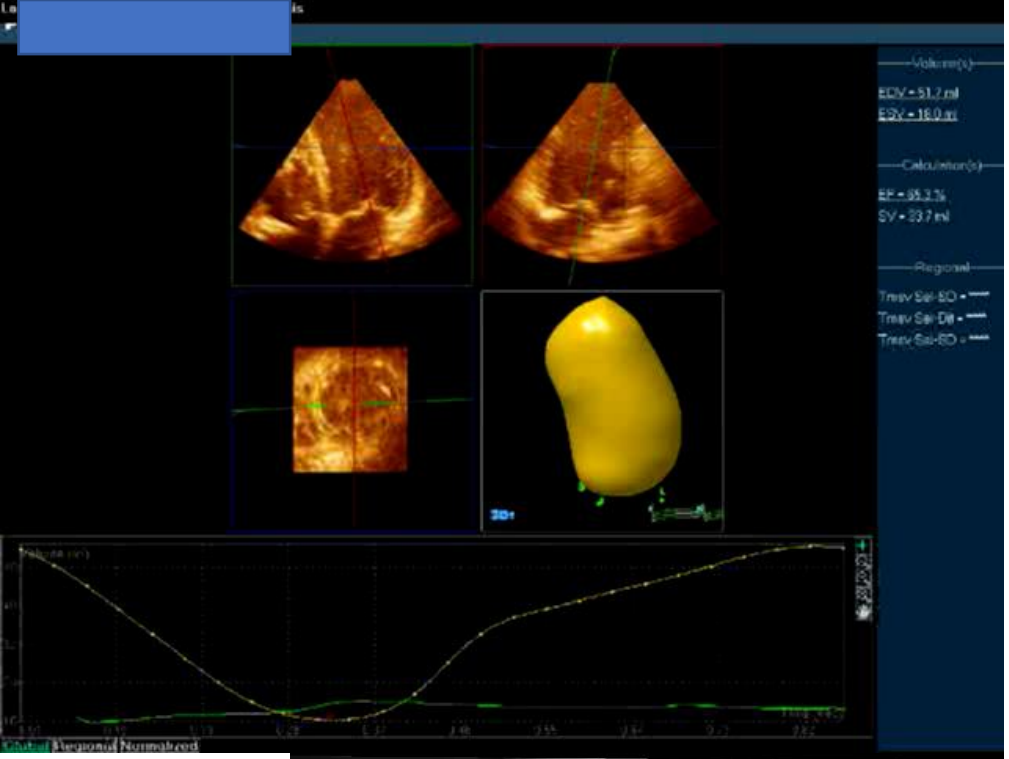
Re-challenge on DDD

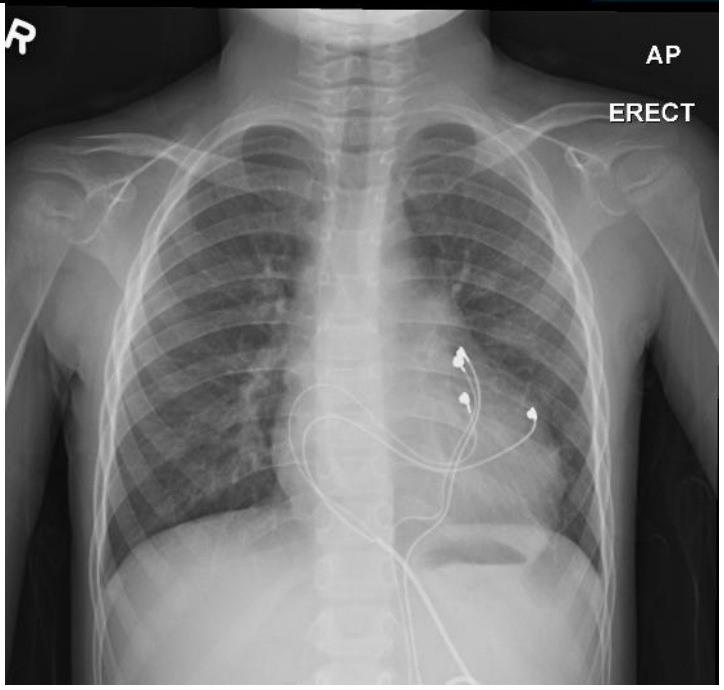


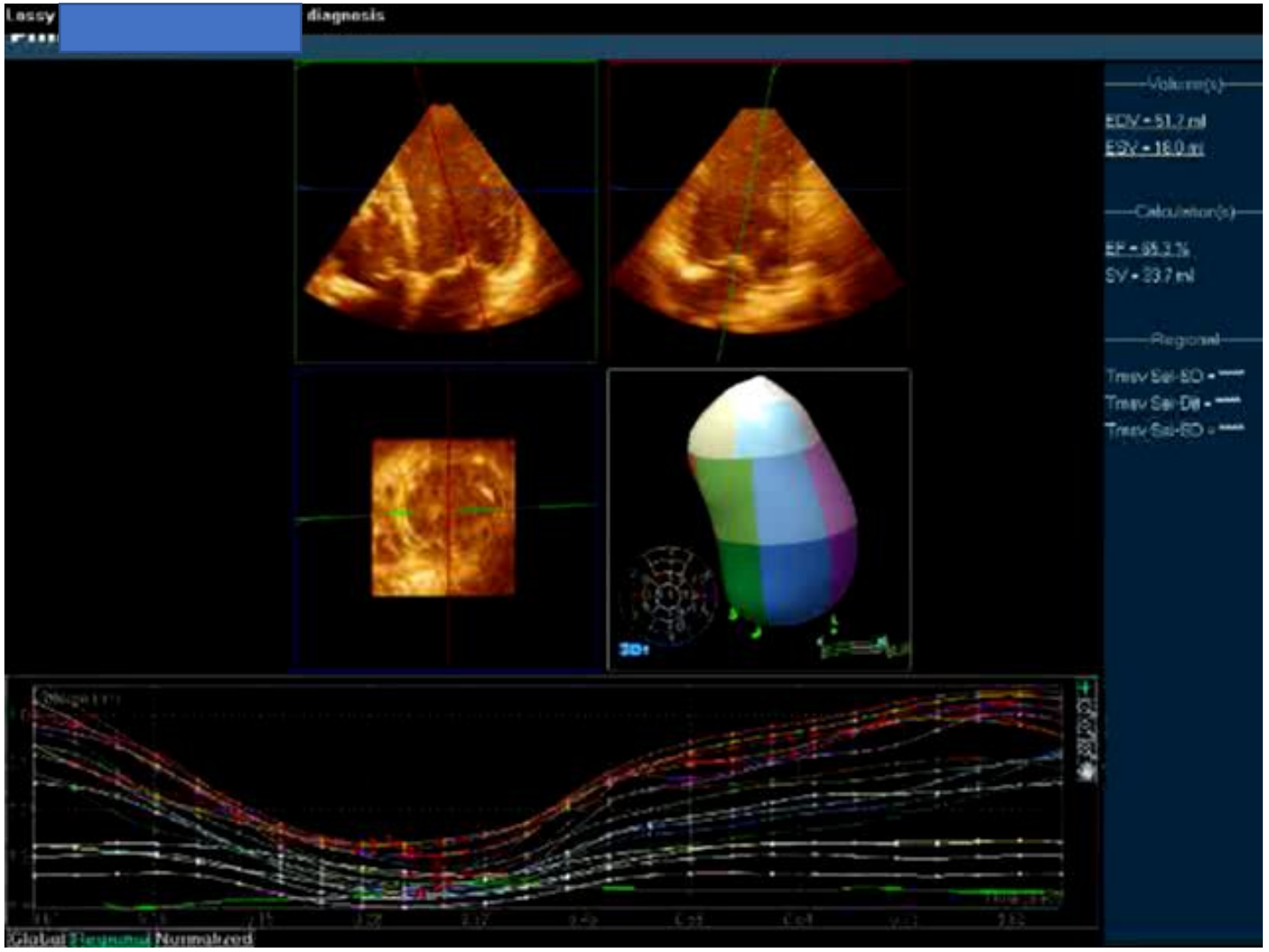


100% VP

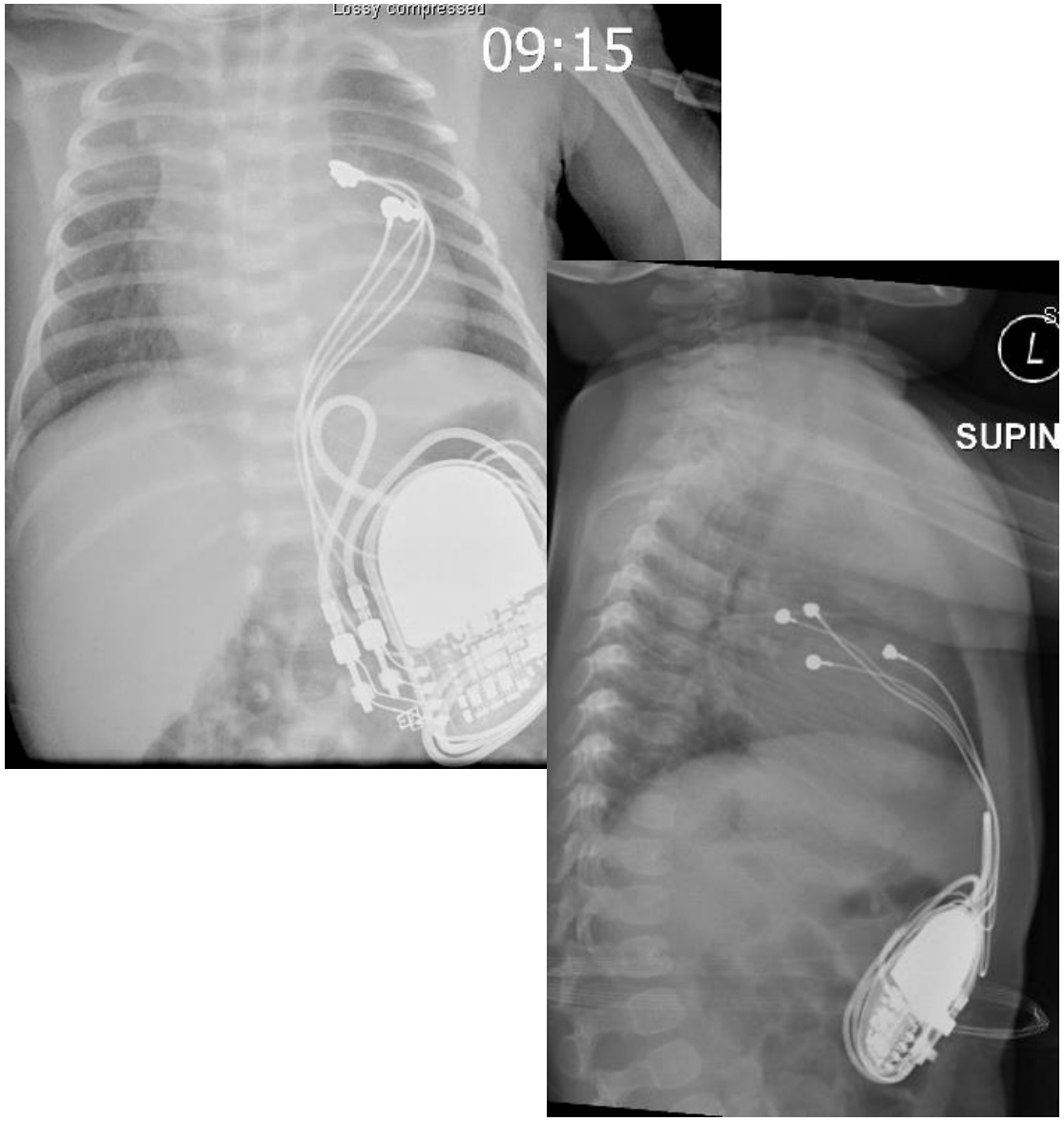
Resume DDD


EDV = 51.2 ml
ESV = 18.0 ml
EF = 65.2 %
SV = 33.7 ml
Global Regional Normalized

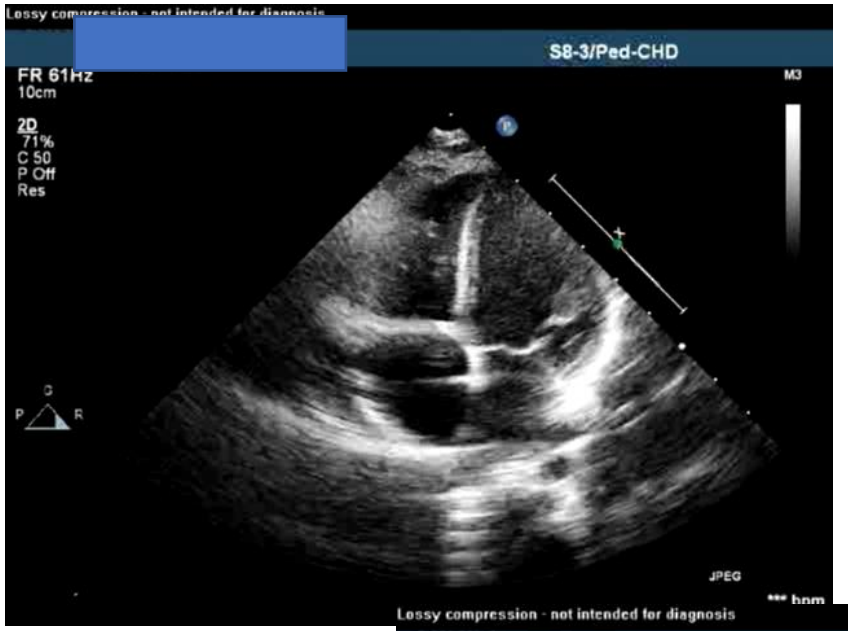

R
AP
ERECT

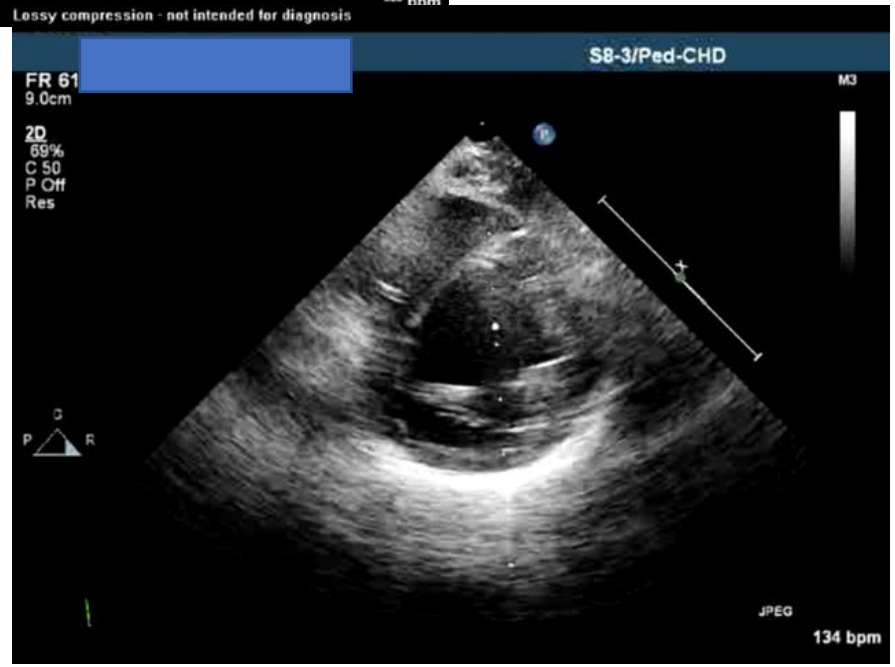

diagnosis
EDV = 51.2 ml
ESV = 18.0 ml
EF = 65.2 %
SV = 33.7 ml
Global Regional Normalized

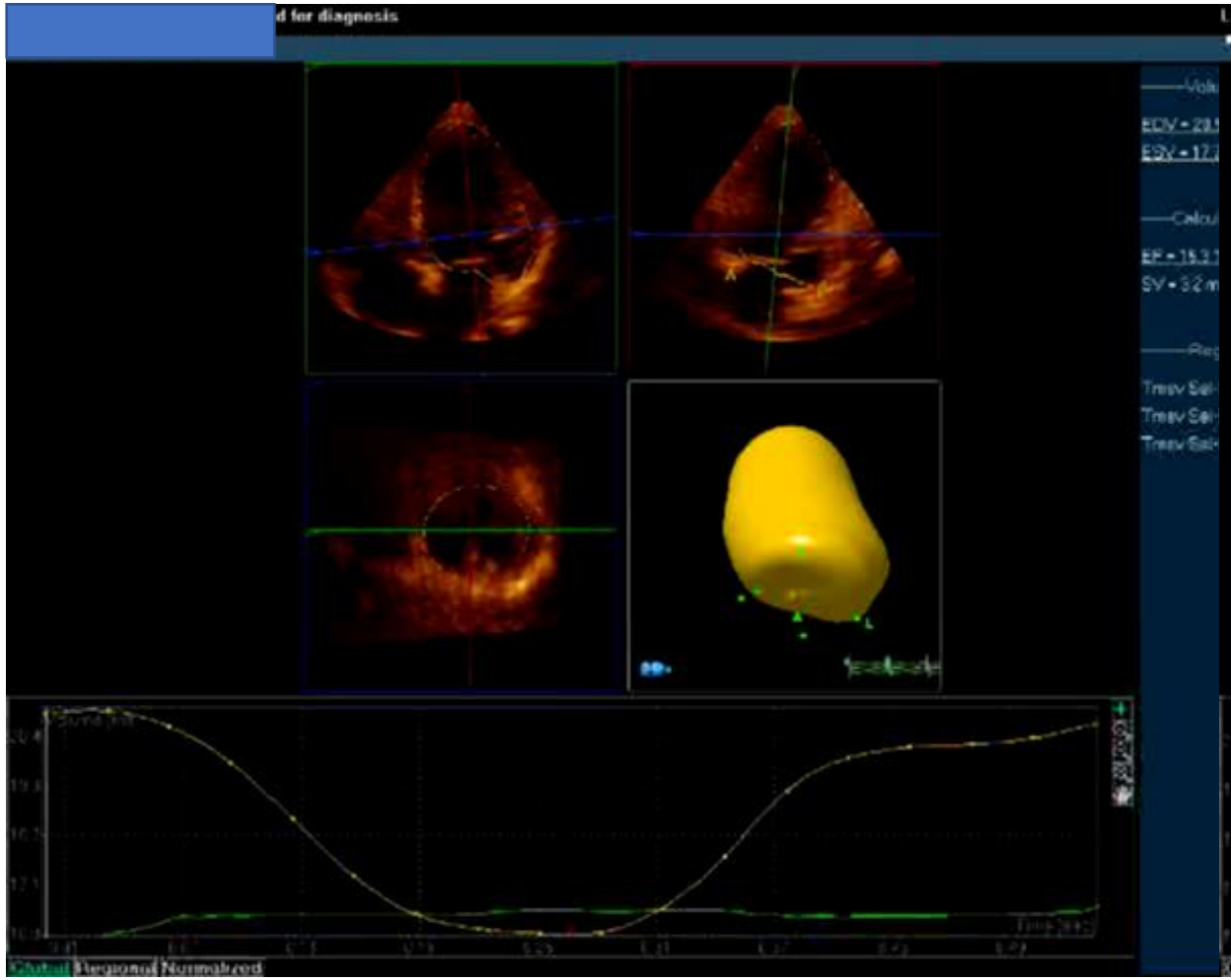
LV basal pacing 2

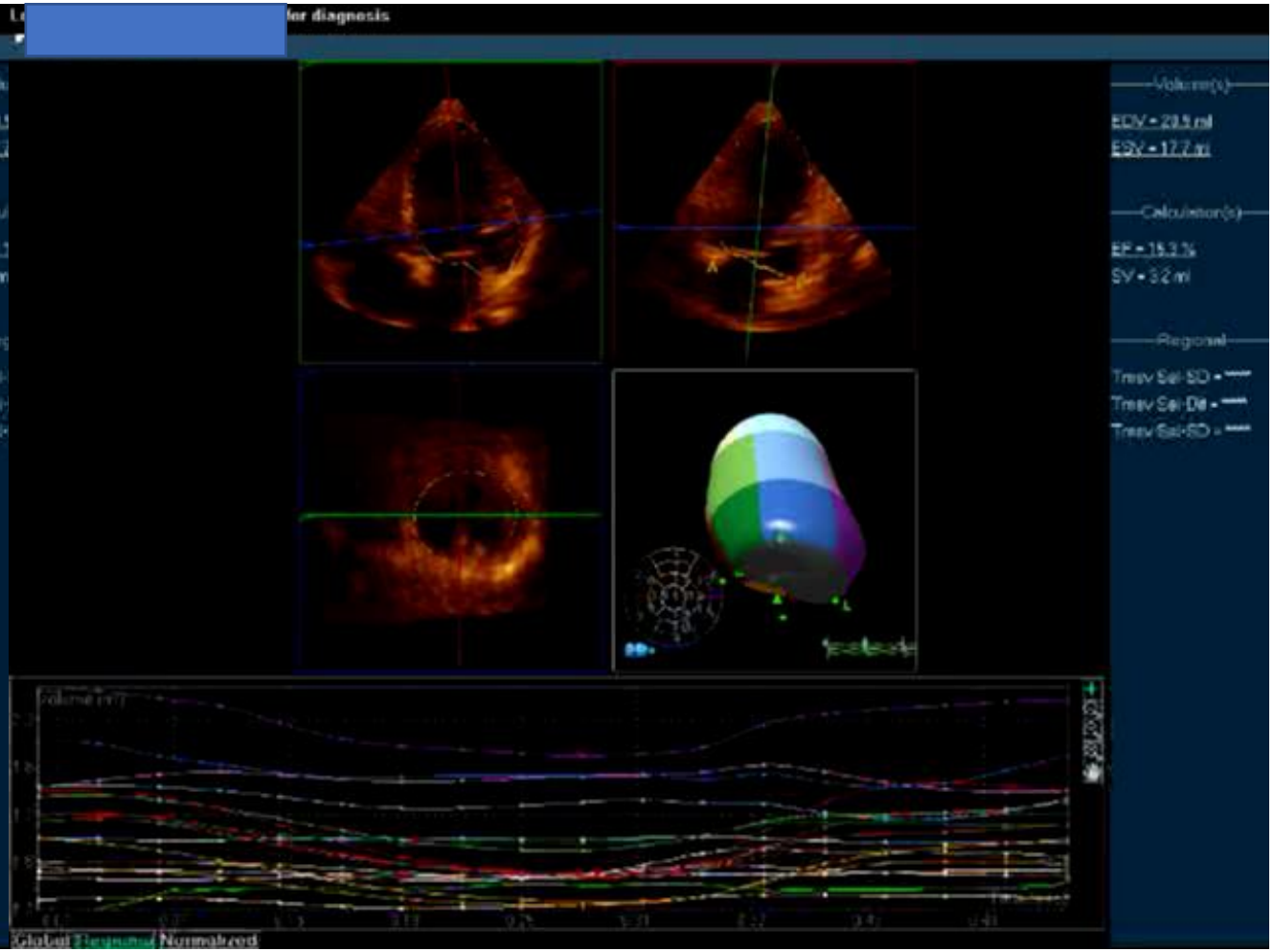


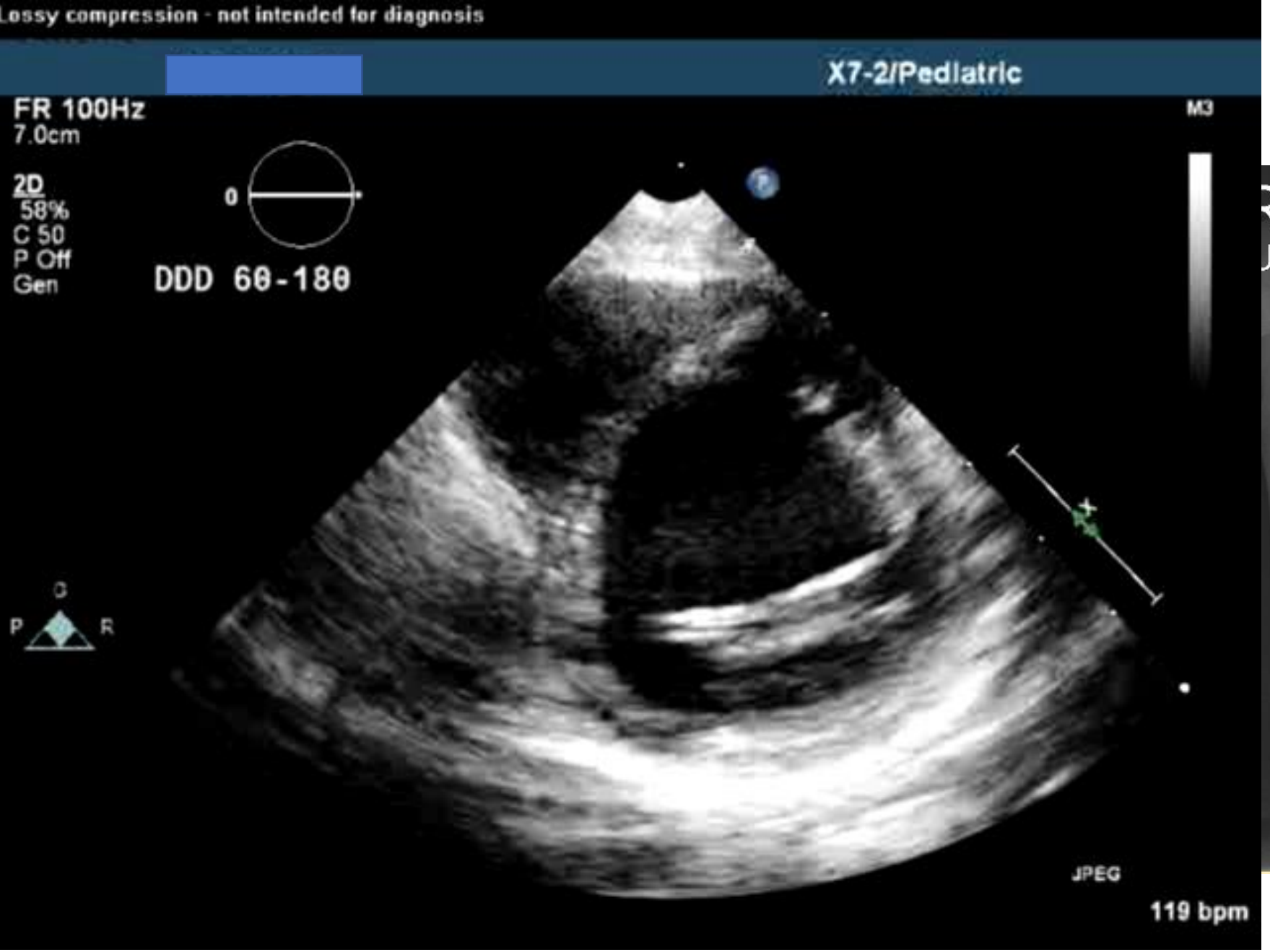
LV dysfunction at 6 month

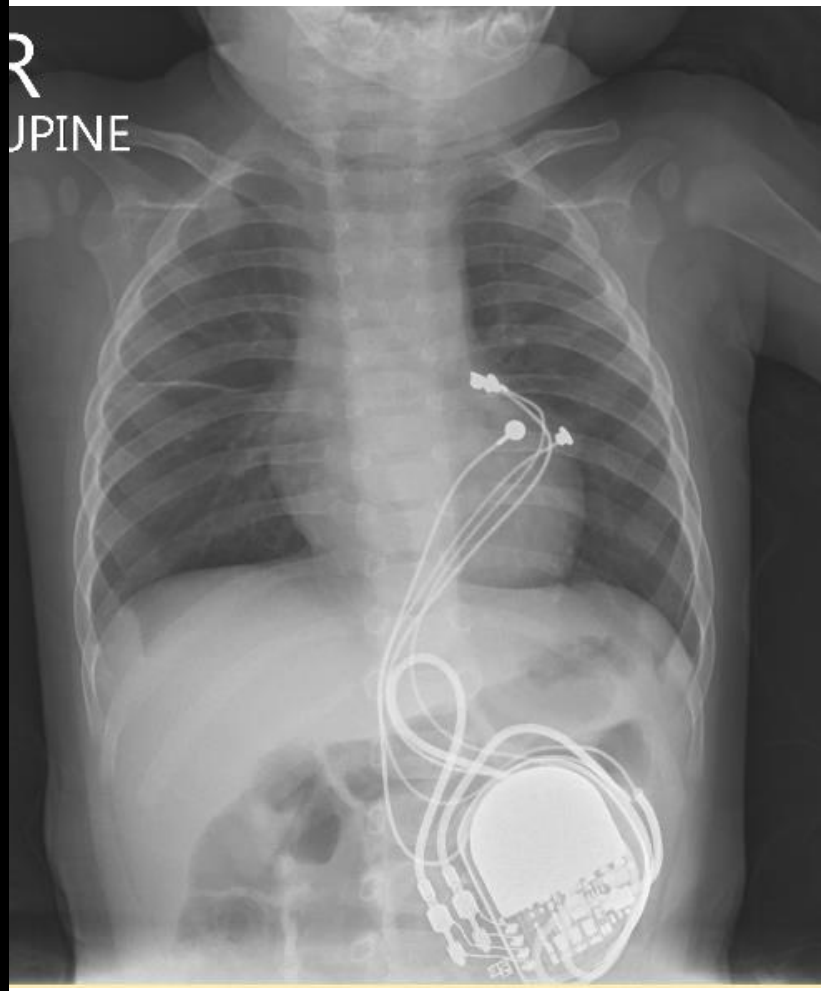





d for diagnosis
Global Regional Normalized

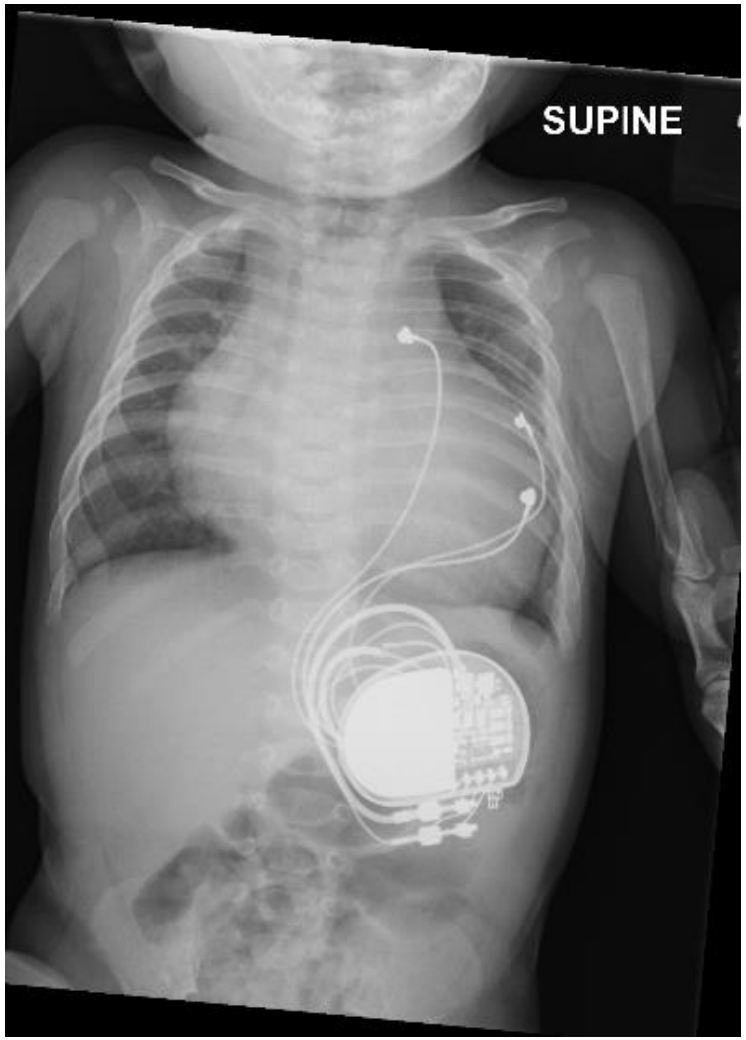

for diagnosis
Global Regional Normalized

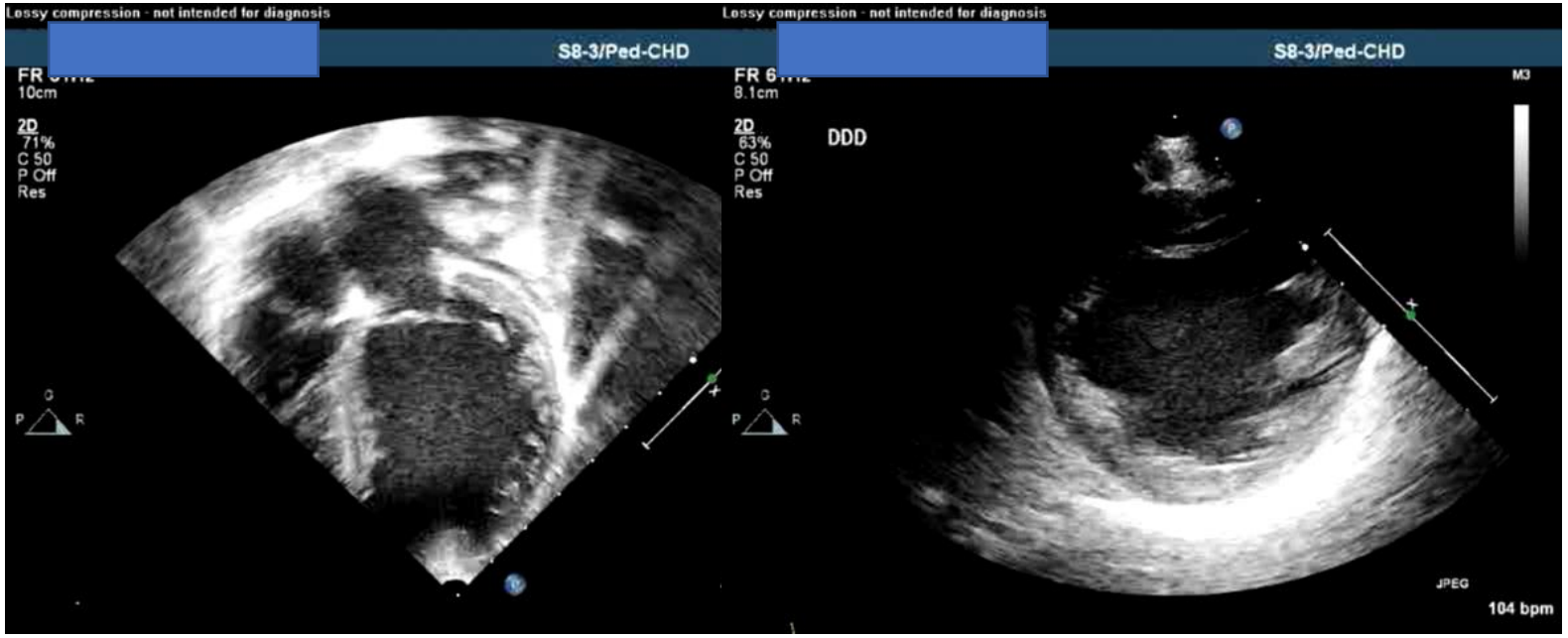

Lossy compression - not intended for diagnosis
X7-2/Pediatric
FR 100Hz
7.0cm
2D
58%
C 50
P Off
Gen
DDD 60-180
M3
JPEG
119 bpm

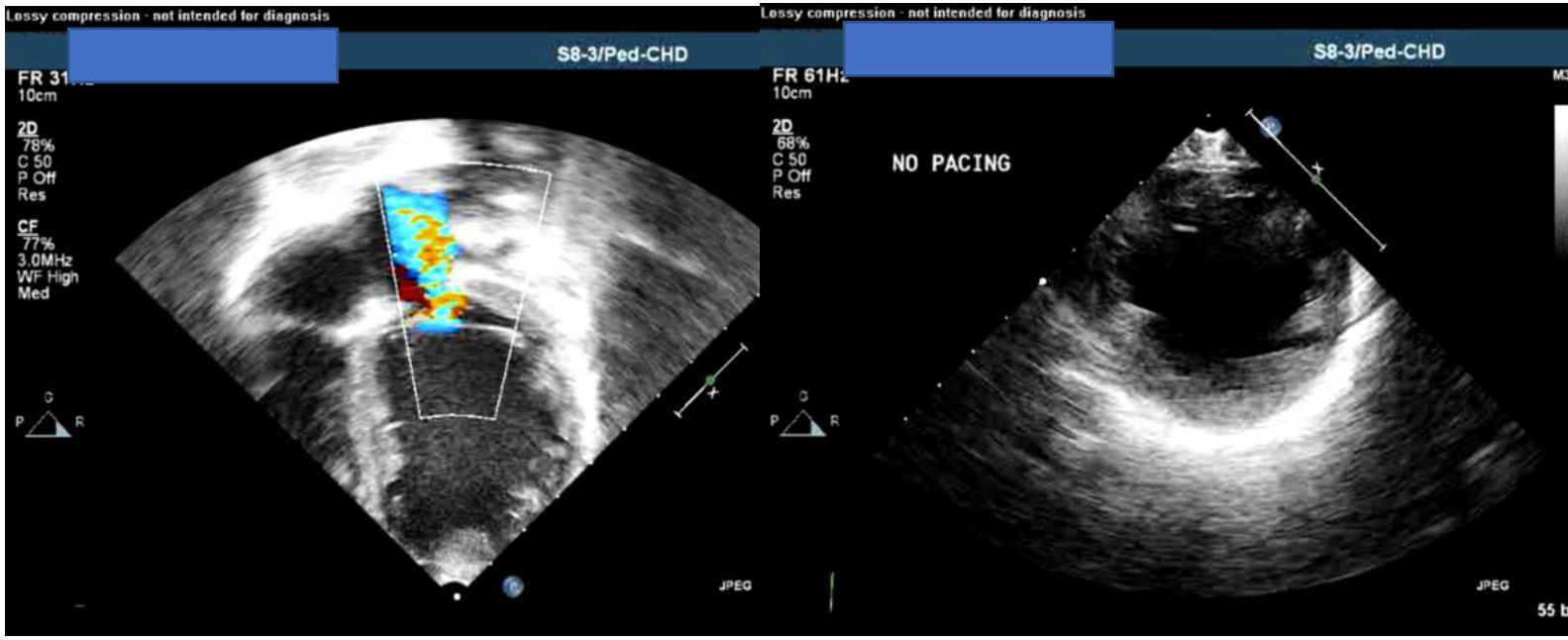

R
UPINE

LV basal?/ apical? pacing

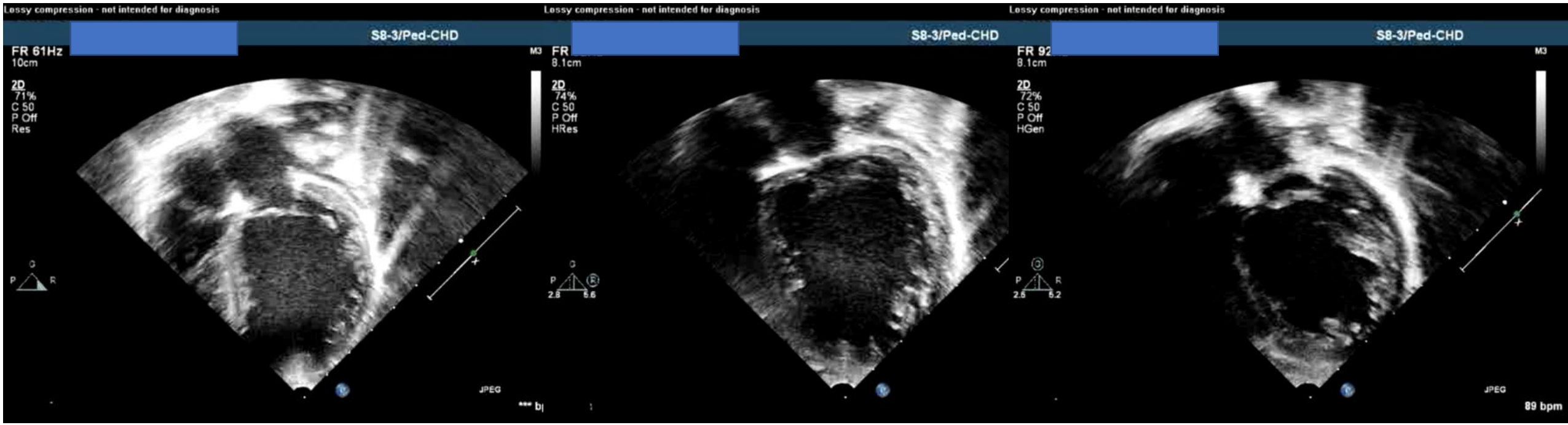
4 month







Decreasing upper tracking rate 120 bpm - no escape rhythm on VVI 60 bpm

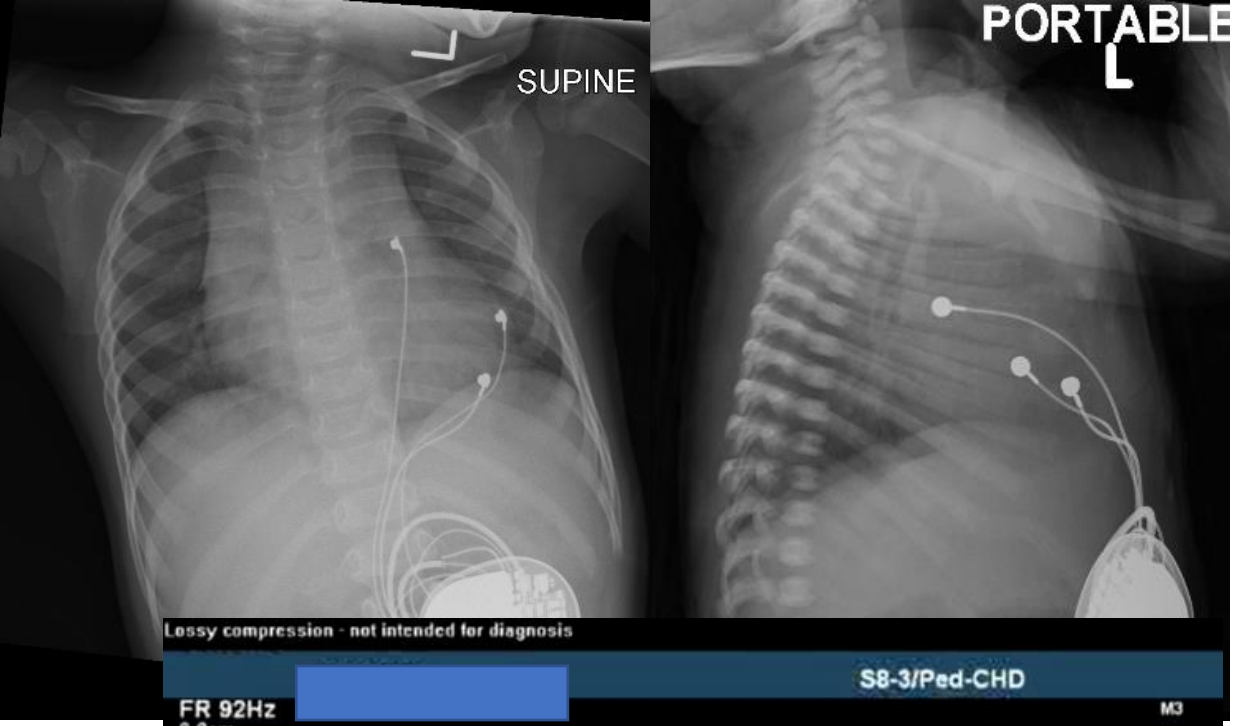


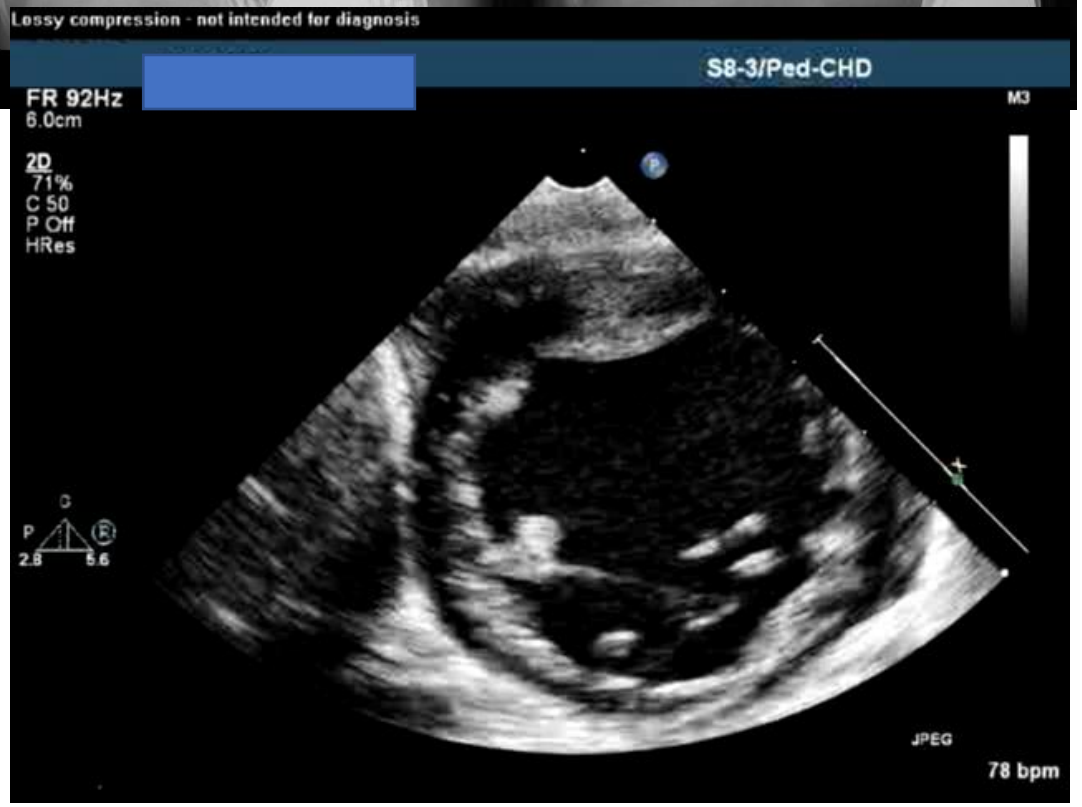
At presentation

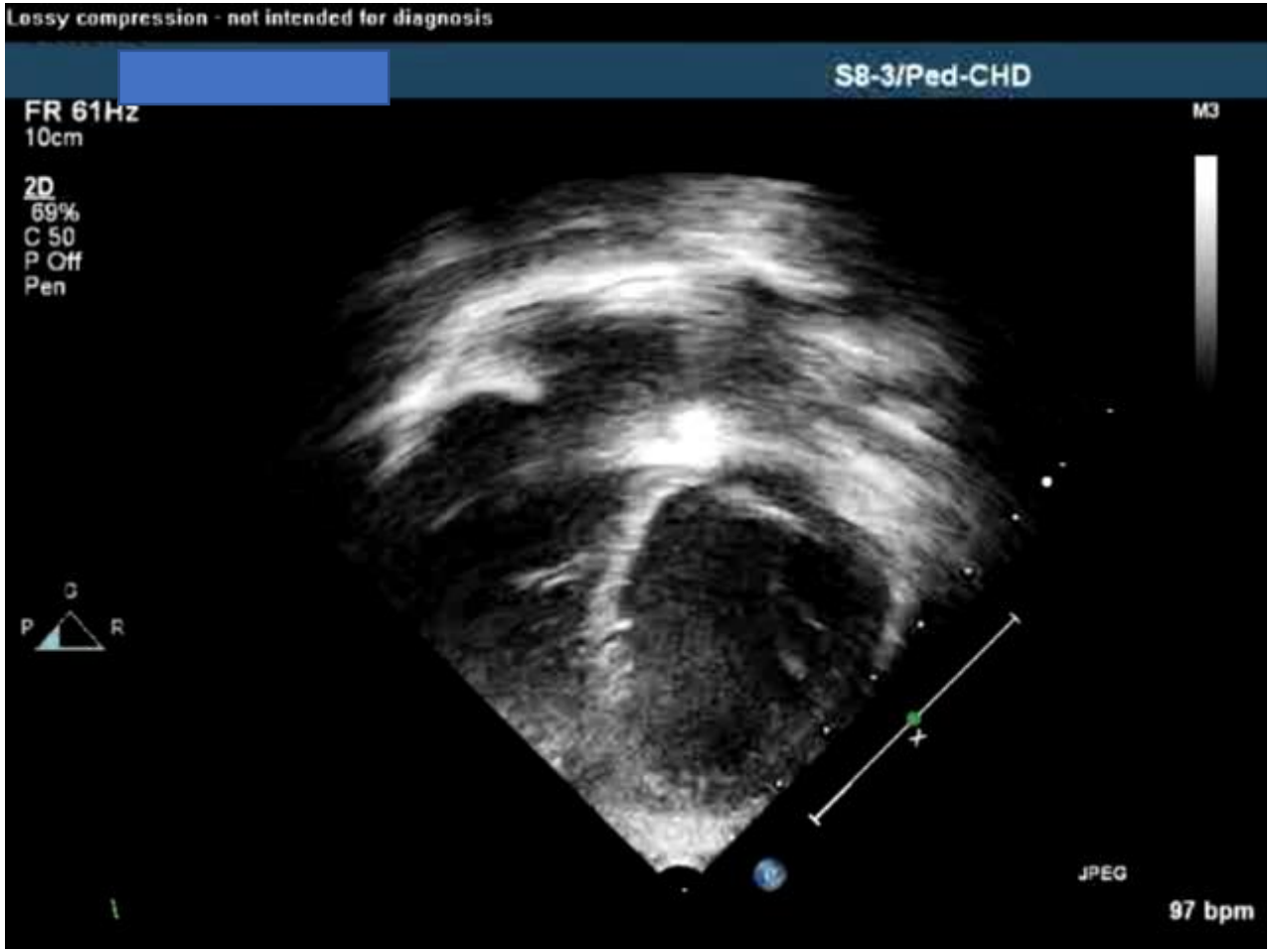
1 week

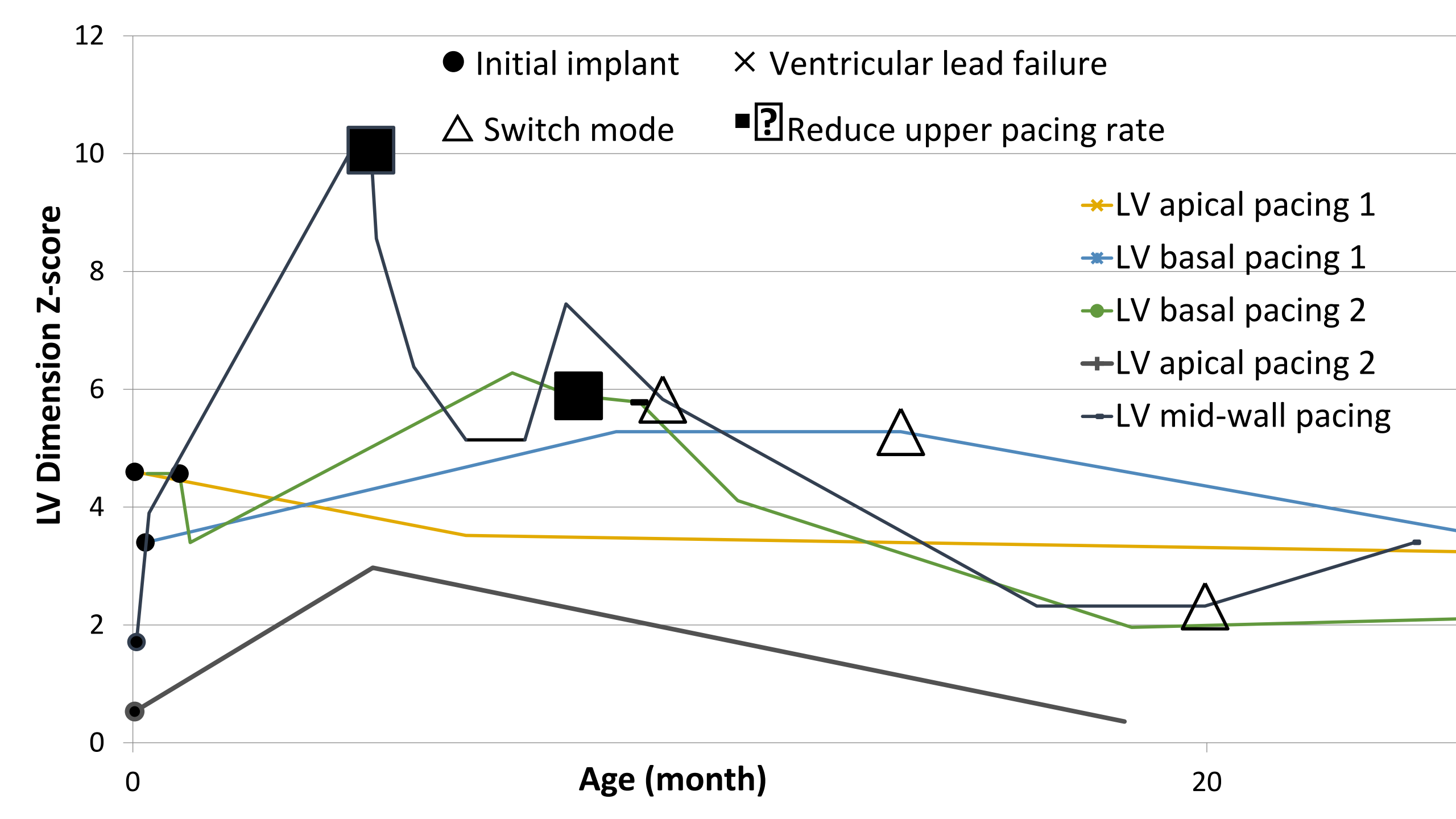
3 month

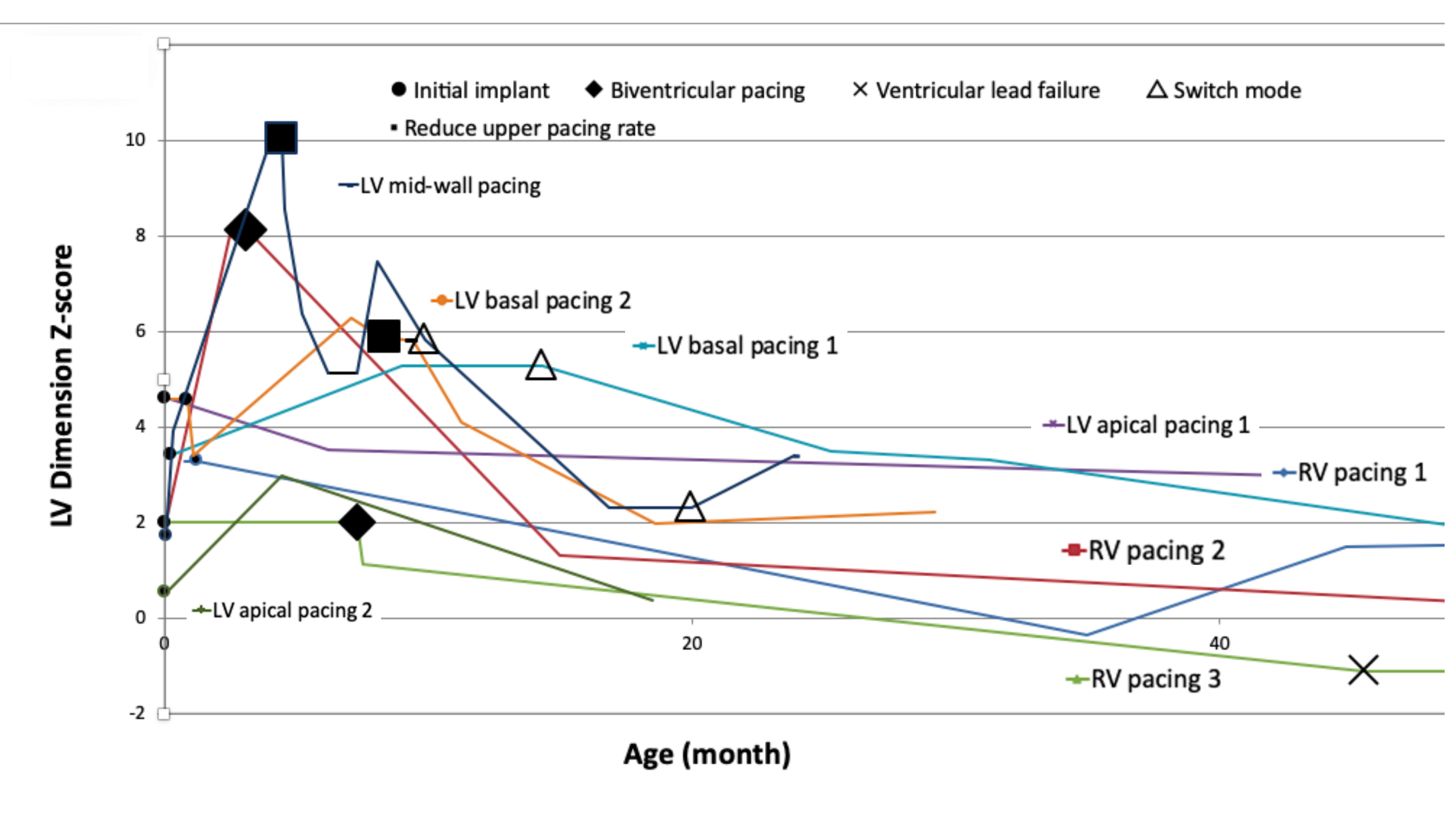
DDD pacing


SUPINE
PORTABLE
L


Lossy compression - not intended for diagnosis
S8-3/Ped-CHD
FR 92Hz
6.0cm
2D
71%
C 50
P Off
HRes
M3
JPEG
78 bpm


Lossy compression - not intended for diagnosis
S8-3/Ped-CHD
FR 61Hz
10cm
2D
69%
C 50
P Off
Pen
M3
JPEG
97 bpm


12
10
8
6
4
2
0
LV Dimension Z-score
0
20
Age (month)
Initial implant
Ventricular lead failure
Switch mode
Reduce upper pacing rate
LV apical pacing 1
LV basal pacing 1
LV basal pacing 2
LV apical pacing 2
LV mid-wall pacing


Initial implant
Biventricular pacing
Ventricular lead failure
Switch mode
Reduce upper pacing rate
LV mid-wall pacing
LV basal pacing 2
LV basal pacing 1
LV apical pacing 1
RV pacing 1
RV pacing 2
RV pacing 3
LV apical pacing 2
LV Dimension Z-score
Age (month)
10
8
6
4
2
0
-2
0
20
40

Learning point

- Pacing site of infant congenital heart block
- Management of cardiac dysfunction –
  Promoting intrinsic rhythm / reduce pacing rate Vs CRT
- Monitoring of cardiac function for congenital heart block post pacing therapy is crucial

Limitation

- Retrospective
- Limited Echo parameters
- Cardiomyopathy
- Medical therapy

Pediatr Cardiol (2017) 38:1309–1310
DOI 10.1007/s00246-017-1629-2

CASE REPORT

Pacing Induced Ventricular Dysfunction in a Child: Improvement with Reduction in Paced Rate

Shilpi Garg[1] · Seshadri Balaji[1]

31 weeker
1.669 gram
VVI, RV lead
110 → 80 bpm

Europace (2010) 12, 1316–1321
doi:10.1093/europace/euq258

CLINICAL RESEARCH
Cardiomyopathies

Determinants of early dilated cardiomyopathy in neonates with congenital complete atrioventricular block

Massimo Stefano Silvetti[1]*, Fabrizio Drago[1], and Lucilla Ravà[2]

Methods and results Retrospective analysis of a single-centre experience. Since 1992, 25 patients, aged 25 (1–355) days [median (range)], with normal ejection fraction (EF), underwent PM implantation (13 DDD, 12 VVIR) with an RV-pacing site. Follow-up was 4 (0.3–16) years. DCM occurred after 4 (3–23) months in eight patients (32%). Univariate analysis identified the following risk factors: younger age at implantation [5 (1–85) days vs. 90 (1–355) $P = 0.007$], a broad QRS (50 vs. 18% $P = 0.03$), prolonged QTc at implantation (63 vs. 0%, $P = 0.001$), and greater duration of heart rate >160 bpm during the first month after implantation (18 vs. 2%, $P = 0.03$). By multivariate analysis prolonged QTc was the only significant risk factor for DCM (hazard ratio: 23, $P < 0.001$, 95% confidence interval: 4–128). One patient died of heart failure, one was lost to follow up, and two were compensated on anticongestive therapy. EF normalized in four patients after resynchronization therapy (two patients), normalization of AV conduction or changing pacing mode to allow predominant narrow QRS junctional rhythm (one patient each).

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Orhan Uzun, Cardiff UK

- In fetuses with CHB due to maternal Lupus, the heart rates over 55 are well tolerated and if the fetal HR is over 65-70bpm then the outcome is excellent. Infants do not tolerate sinus tachycardia tracked by high ventricular pacing rates very well. Immature myocardium is incapable of coping with DDD pacing (tracking sinus tachycardia with abnormal ventricular depolarisation) or high rate ventricular pacing above rates 100-120 ppm. Atrial kick is also less relevant in these ages therefore DDD pacing may not be the best choice. In fact children up to age of 6-8 tolerate VVI pacing remarkably well. Once they are very active then pacing mode can be switched to DDD.
- We had 3 cases of DDD pacing induced cardiomyopathy due to tracking of fast sinus rates in immature infants. Using beta blocker or mixed one like carvedilol also may prevent sinus tachycardia being tracked in such cases with DDD pacing, if one has to do DDD pacing but in my experience is not necessary. Reducing pacemaker rate to 90 and changing pacing mode from DDD to VVI 90bpm and Captopril support LV function recovered in my cases. In one older child biventricular pacing was required and the mechanism was again tracking of atrial high rates by ventricular pacing in DDD mode.
- Histogram in this case is obvious that >85% of the time the baby had a pacing heart rates of 110ppm and >60% of the time over 140. This is not desirable. We routinely choose VVI pacing at rate of 90-95 in infants with no problem whatsoever. Problem occurs when a pacemaker is implanted elsewhere and programmed as HR of 130-140 even with VVI mode or DDD mode with tracking rates of 180 and higher basal pacing rates of over 100bpm. We instantly switch it to VVI and rate of 90ppm. None of these patients had CMP.
- In summary, (1) it would be helpful to check lupus antibodies in the infant. Maternal lupus induced cardiomyopathy in infants may occur but also lupus itself may be present too. A few months would be enough for CMP to appear or sometime becomes evident at birth. (2) Mode switch to VVI and fixed HR of 90ppm. (3) Support myocardial function with Captopril or carvedilol as appropriate. (4) Bi-ventricular pacing if necessary.

Ferran Rosés, Spain

- I agree with Shu's, Fabrizio's and Nico's views that in some cases the most probable mechanism is a combination of rapid pacing plus mild dyssynchrony and immaturity, and it is really difficult to know which is more relevant as it seems difficult to predict in which patients this phenomenon will occur.

- Specifically in this case, I believe that the rapid progression is due to the fact that the epicardial LV leads are placed towards the base of the LV wall and therefore they cause a reverse LV dyssynchrony pattern. We have had 3 cases like this one and the 3 we decided to upgrade to a CRTP with a new lead placed in the RV Apex, and the 3 of them recovered completely. I think when leads are implanted in very small babies it is difficult for the surgeons to reach the true LV Apex, and they have to be sure they place the anode more distally because if they get it to too basal, this reverse LV dyssynchrony patterns seems to cause a very rapid LV dysfunction.

- Changing to VVI would be a good option in some other cases, but when there is such a rapid progression, I would be inclined to upgrade to a CRTP.
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Shu

- This is a case of significant interest to me, and this type of case inspired the recent study proposed by Drs Shah and Tan that was circulated to this group just a few days ago. I have unfortunately a few such cases. Those, combined with a few cases of infant post-op AV block, lead me to suspect that rapid V pacing in the immature myocardium promotes dysfunction in a very rapid manner. I have changed my practice over the years to leave patients with upper rates around 100, perhaps 120, for a few years even. While it is only anecdote, this approach has not resulted in the phenomenon described below. I think it is a function of rate, immature myocardium, and likely abnormal activation. I am not sure that we will be able to easily separate these, unless we do animal studies.
- Since it has occurred in post-op, non-lupus cases, I do not think this is lupus mediated.
- Once it occurs, I have reduced the rate and in some cases added resynchronization and heart function treatment as well. The best response has been with resynchronization.
- I am looking forward to learning from this group, as it would be very good to be able to study a few such cases, and potentially understand the histology, etc.

Alberto Sciegata, Argentina

- 3 years ago we had a similar case which was resolved downgrading the PM to VVI 60 bpm with hysteresis at 40 bpm and the patient recovered his LV function very soon. I agree with Dr. Sanatani about the phisiopatology of such cases including high rate DDD, immature myocardium and abnormal activation with a wide QRS. Our usual practice in programming PM for CHB with very low heart rate is DDD 60/180 bpm if the QRS is narrow (less than 120 ms) and there is not evidence of dysinchrony by Echo. When this is not possible we prefer VVI with hysteresis at low rates trying to mantain the own rythm . We think that epicardial stimulation is better from the LV apex of course.

Fabrizio Drago

- This is our experience..
- Sometimes “physiological” pacing in DDD and mild dissyncrony can cause severe LV dysfunction in neonates and infants.
- In very particular cases , when possible, in order to recover the function, we have , carefully and successfully , even switched off DDD pacing and come back to spontaneous rhythm with narrow QRS, even if at low rate, with VVI pacing at the lowest rate, (just in case) .
- Then we have started again ventricular pacing with just a little increase of HR with the time.