

VT Case Studies

Moti Haim, MD

Rabin Medical Center

31M with WCT

December 19, 2006

History

- 31 yo male, no prior cardiac history
- 4 episodes of sustained palpitations associated with heavy exercise or stress (e.g. once after a near-miss auto accident)
- Documented WCT 200 bpm
- Terminated by IV procainamide on one occasion in the ED, and by IV lidocaine on another occasion

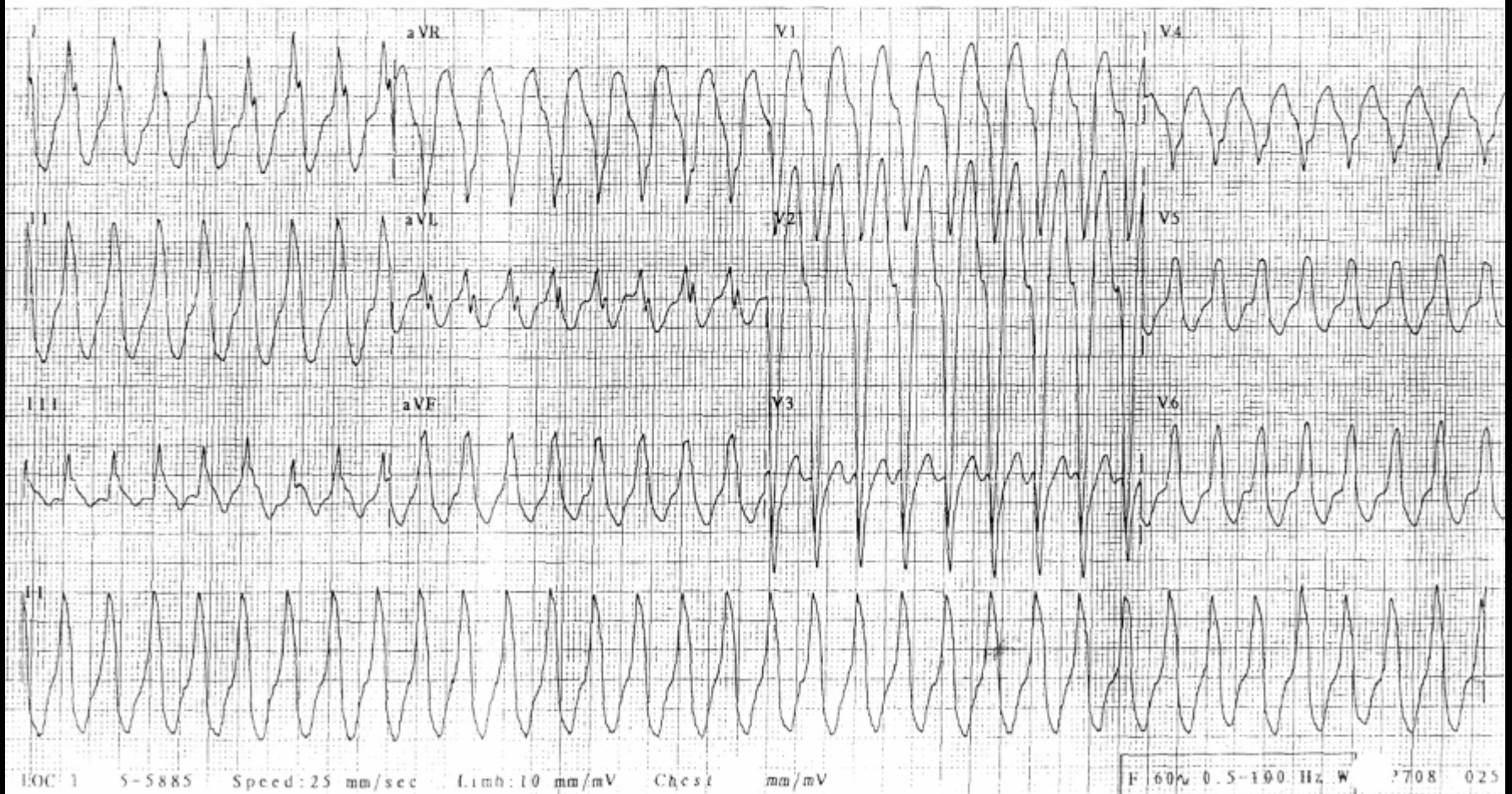
Clinical VT

Rate 201 Extreme tachycardia, rate 201 bpm, V rate > (220 age) of 150 SERIAL
PR 75 Left bundle branch block QRS 120 mS, terminal forces left
QRSD 136
QT 259
QTc 474

--AXIS--
P -63
QRS 51
T 225

- ABNORMAL ECG -

PRELIMINARY-MD MUST REVIEW



Clinical – 2nd event

31 years
Male Caucasian
Vent. rate 211 bpm
PR interval * ms
QRS duration 210 ms
QT/QTc 310/581 ms
P-R-T axes * 83 169

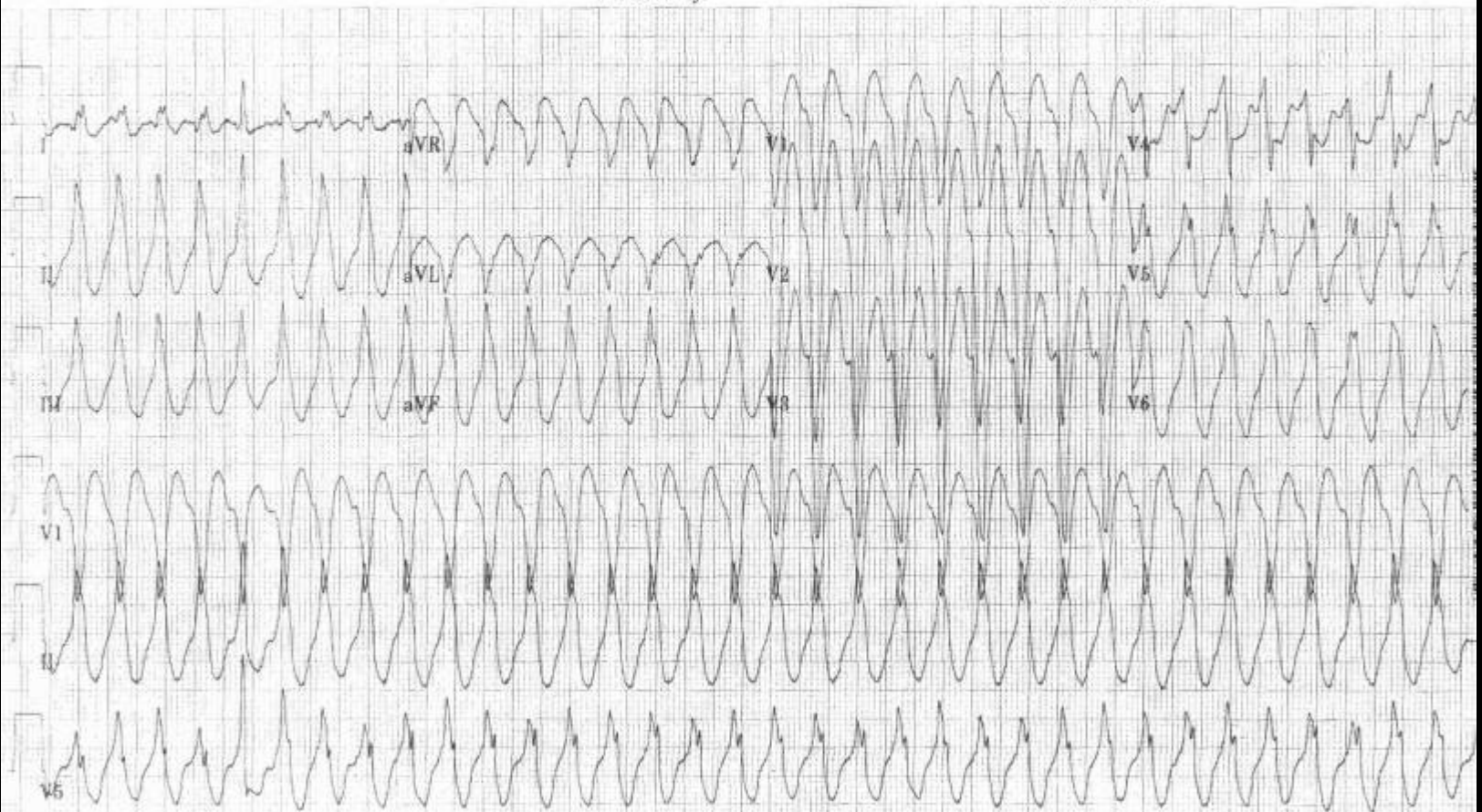
Wide QRS tachycardia with premature supraventricular complexes and fusion complexes
Left bundle branch block
Abnormal ECG

SA ✓ (2)

Technician:
Test ind:

Referred by:

Unconfirmed



LBBB inferior axis VT



Precordial transition

At or before V2/V3

V2-V4

V4 or later

LVOT VT

Coronary Cusp VT

RVOT VT

VT from above PV

➤ axis may be horizontal axis

Aortomitral continuity

➤ RBBB right inferior axis

Anterior

-R in lead I

Septal wall

➤ Transition V4 or earlier

➤ QRS < 140ms (close to AVCS)

➤ R amplitude (inferior leads) > free wall

Septal wall

➤ LBBB inferior axis

Lead I

Dominant R

Bifid R

rS

Posterior

+R in lead I

Free wall

➤ Transition V4 or later

➤ QRS > 140ms

➤ S amplitude (precordial) > septal wall

➤ S in V3 > 3mV

➤ Notched QRS (RR') (inferior leads)

RCC

NCC

LCC

Other methods to differentiate RVOT and LVOT VT

	RVOT VT	LVOT VT
R/S ratio in lead V3	> 1	< 1
QS amplitude	aVL > aVR	aVR > aVL
R/QRS ratio in lead V2	< 30%	

Other methods to differentiate Coronary Cusp VT from RVOT

	Coronary Cusp VT	RVOT VT
R-wave duration in leads V1 & V2	80-90ms	40-50ms
R/S ratio in lead V2	55%	10%

History (2)

- Cardiac MRI performed
 - Normal ejection fraction of LV and RV
 - Normal chamber sizes
 - No hyperenhancement or evidence of fatty infiltration in either ventricle
- Scheduled for SAECG and EPS

Baseline ECG



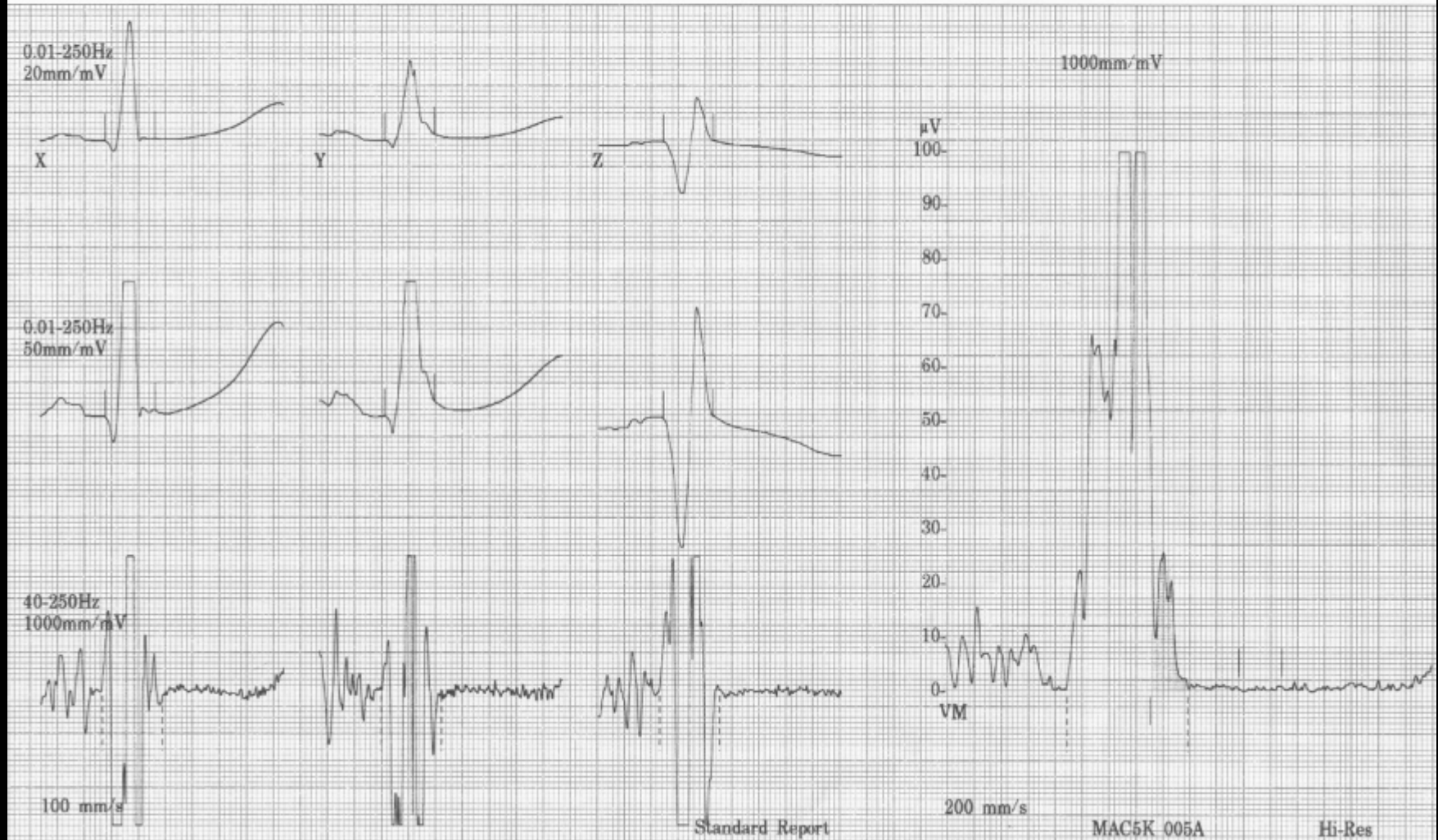
Analysis Filter : 40-250Hz
Std. QRS Duration (unfiltered) : 93 ms
Total QRS Duration (filtered) : 112 ms
Duration Of HFLA signals < 40 μ V : 35 ms
RMS Voltage in terminal 40 ms : 25 μ V
Mean Voltage in terminal 40 ms : 18 μ V

Number Of Beats Averaged: 41
Number Of Beats Detected: 45
Noise Level (Std. Devn.) : 0.30 μ V

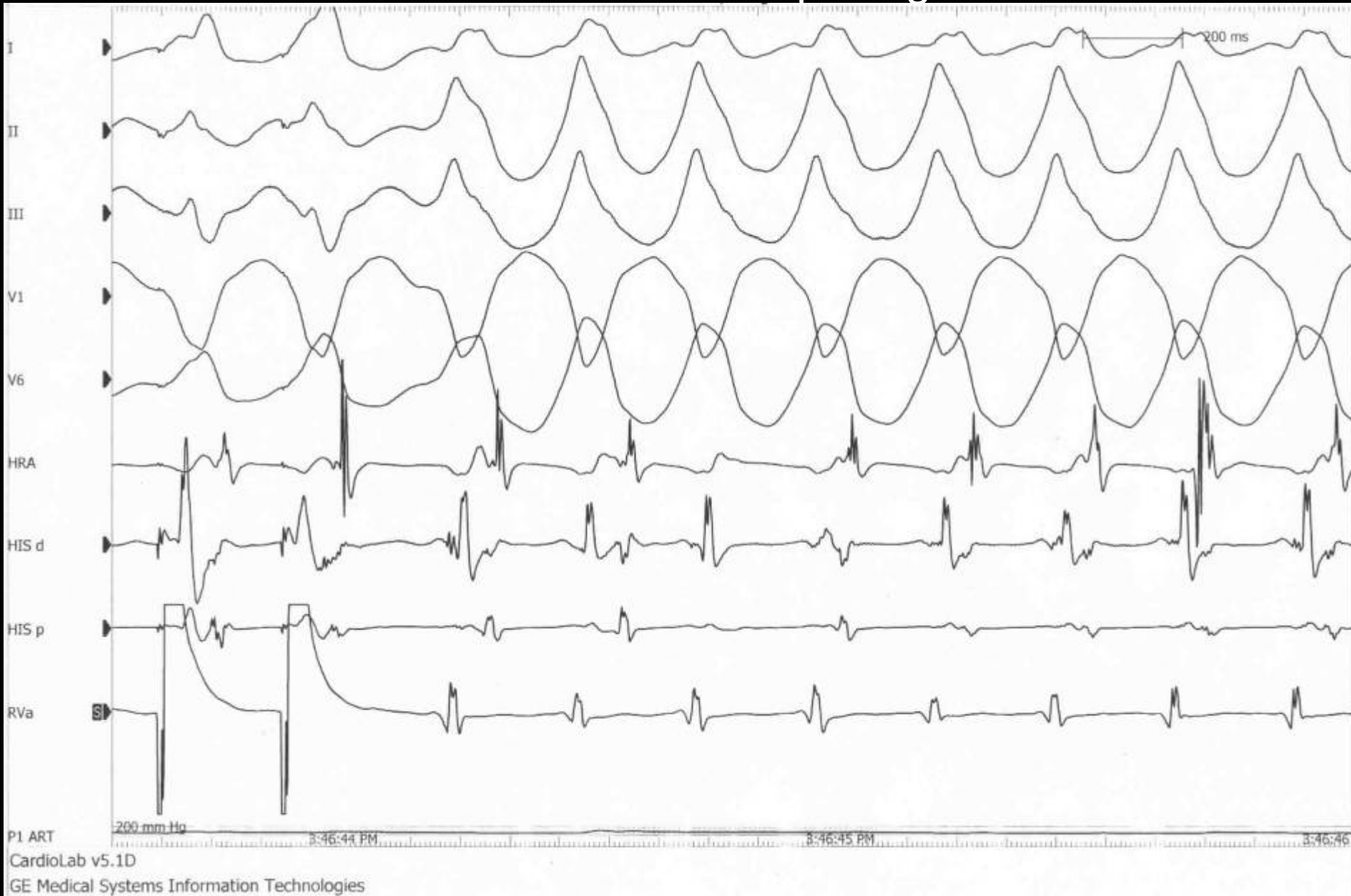
Technician:

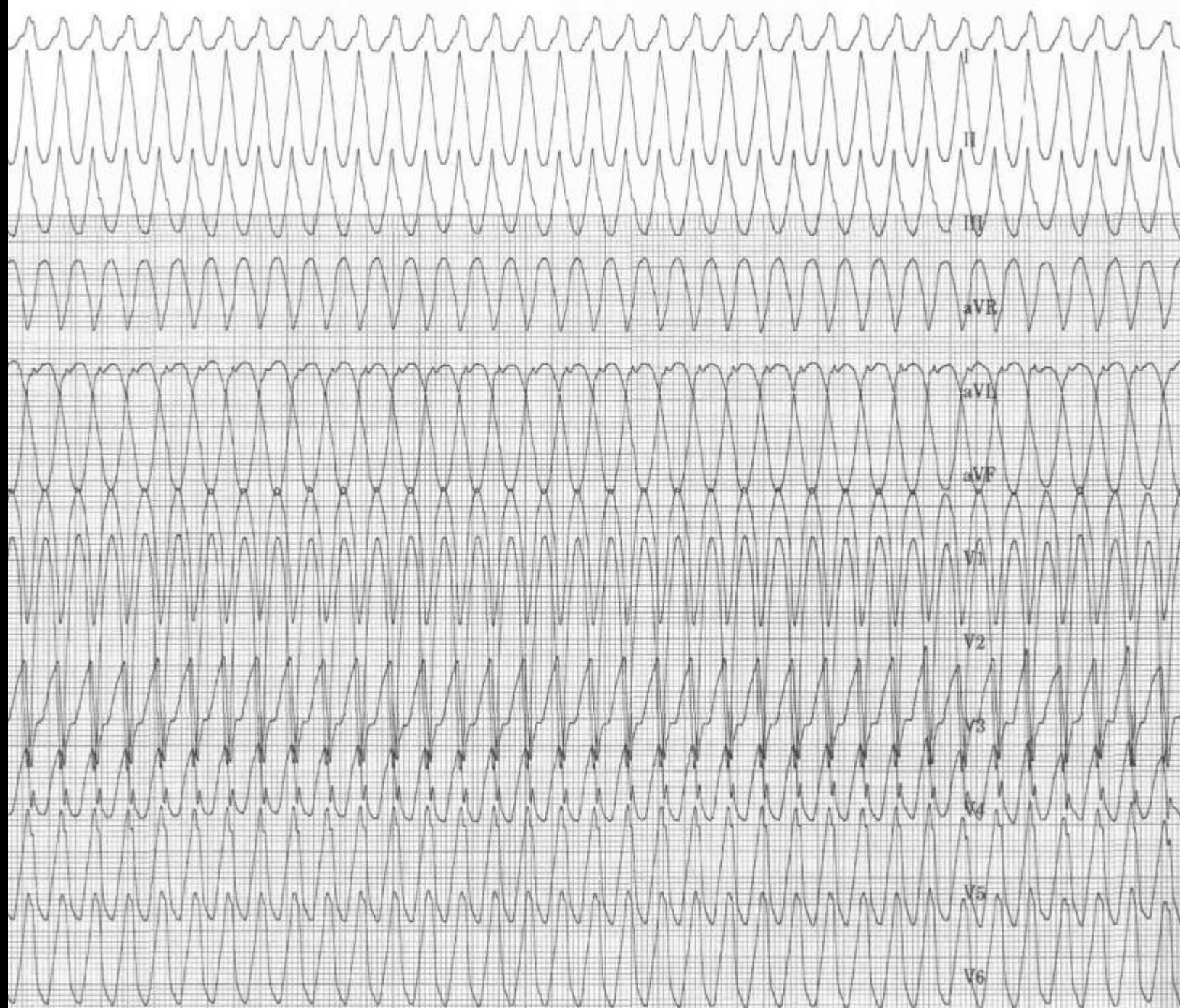
Referred by:

Unconfirmed

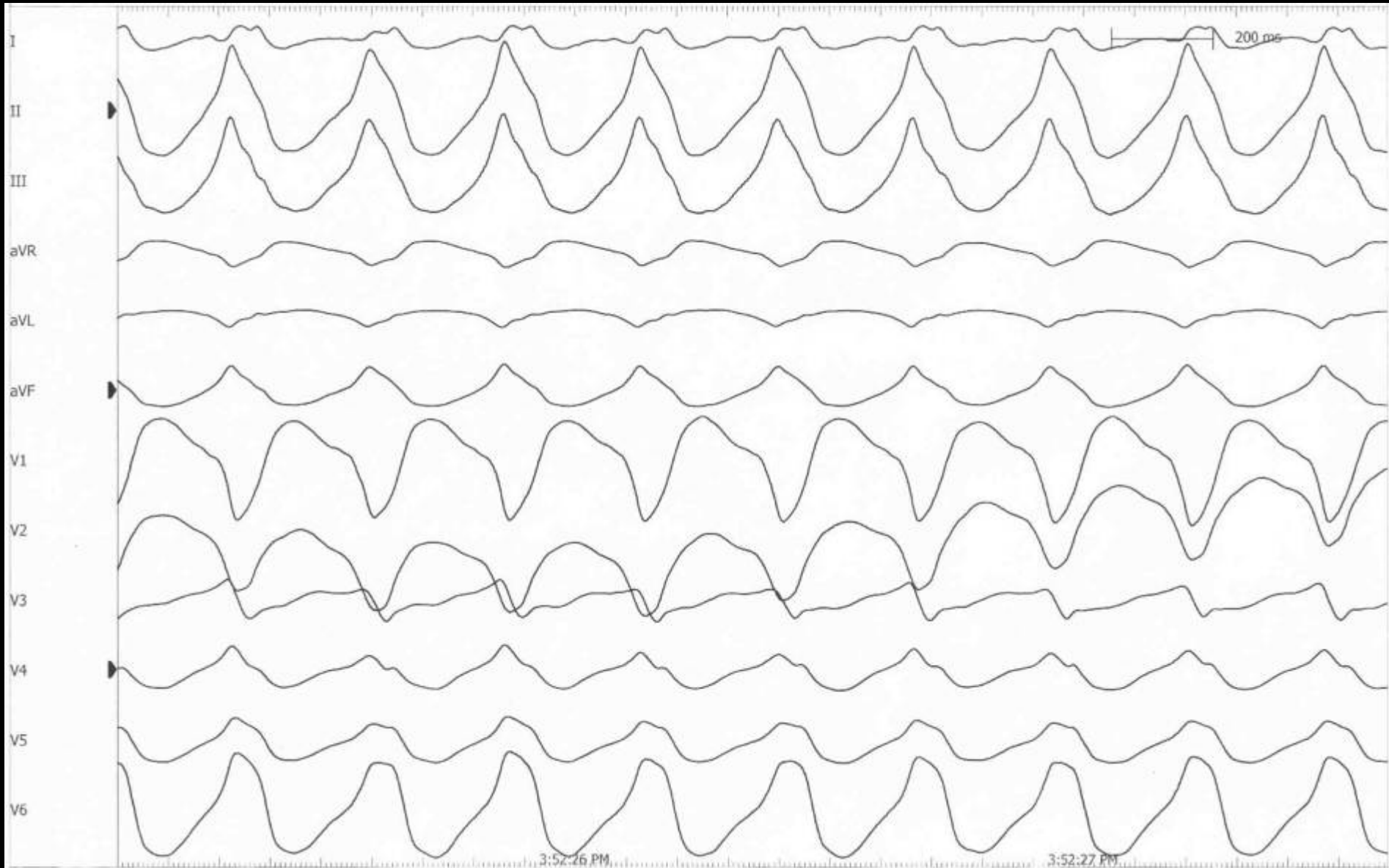


Induction with RV-pacing

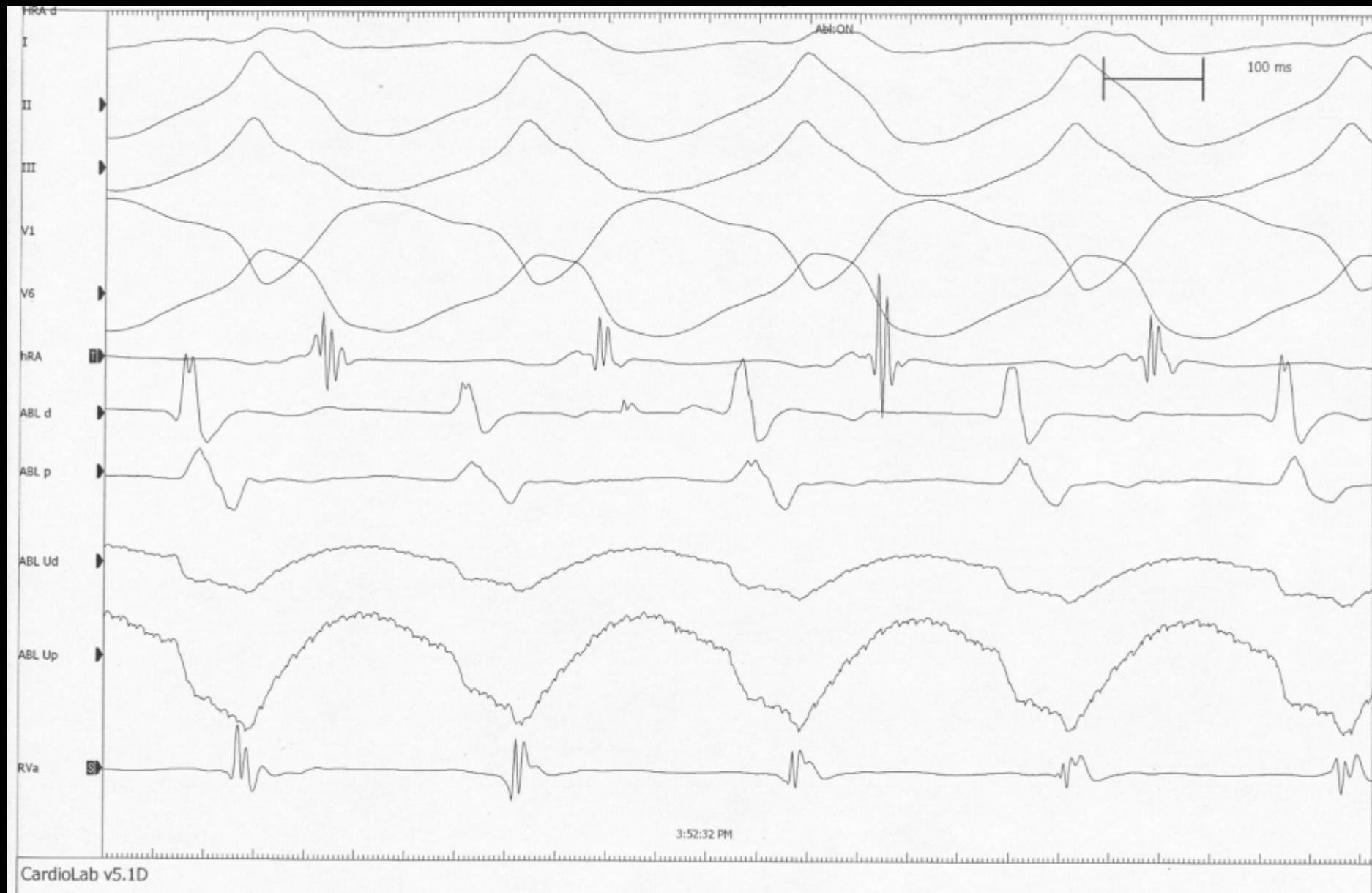




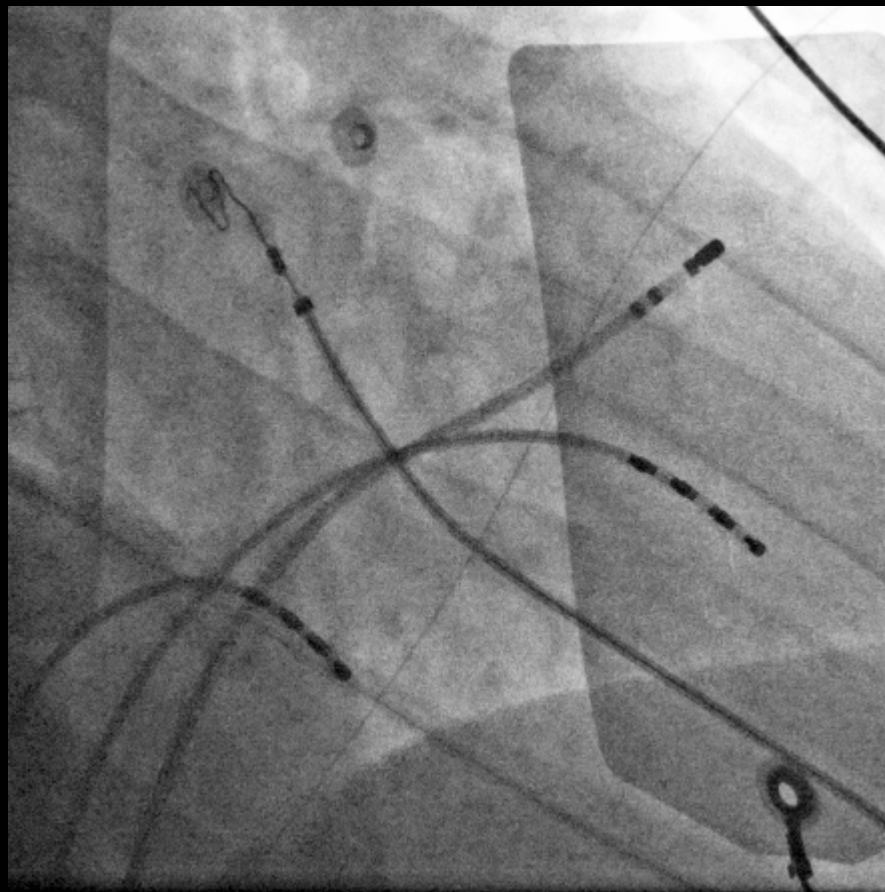
VT



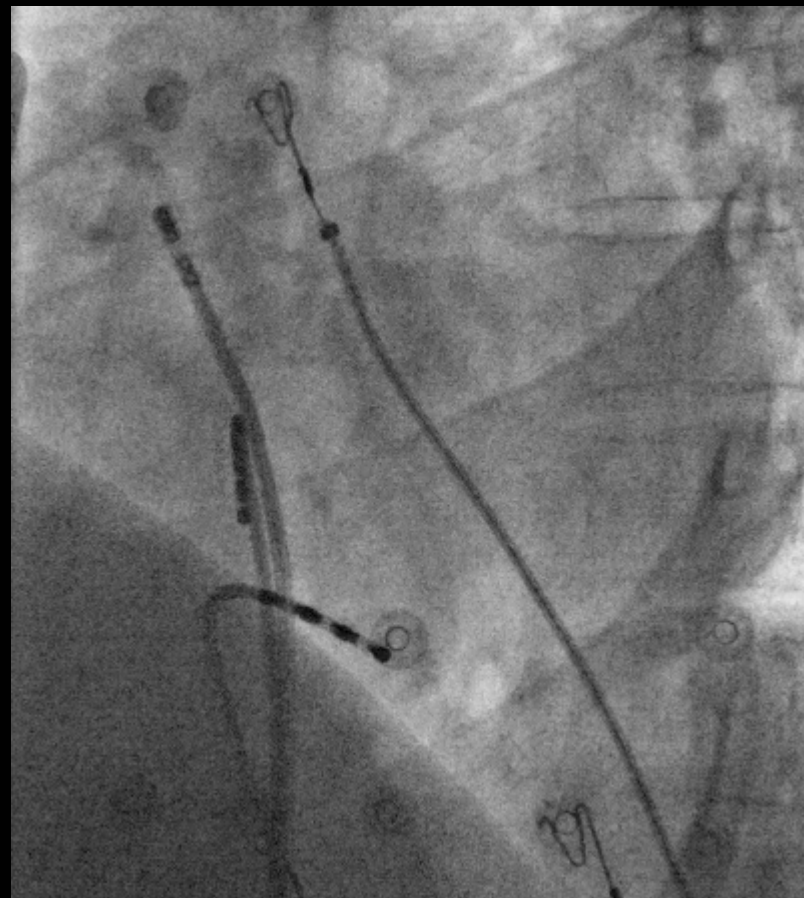
Successful ablation site



Successful ablation site

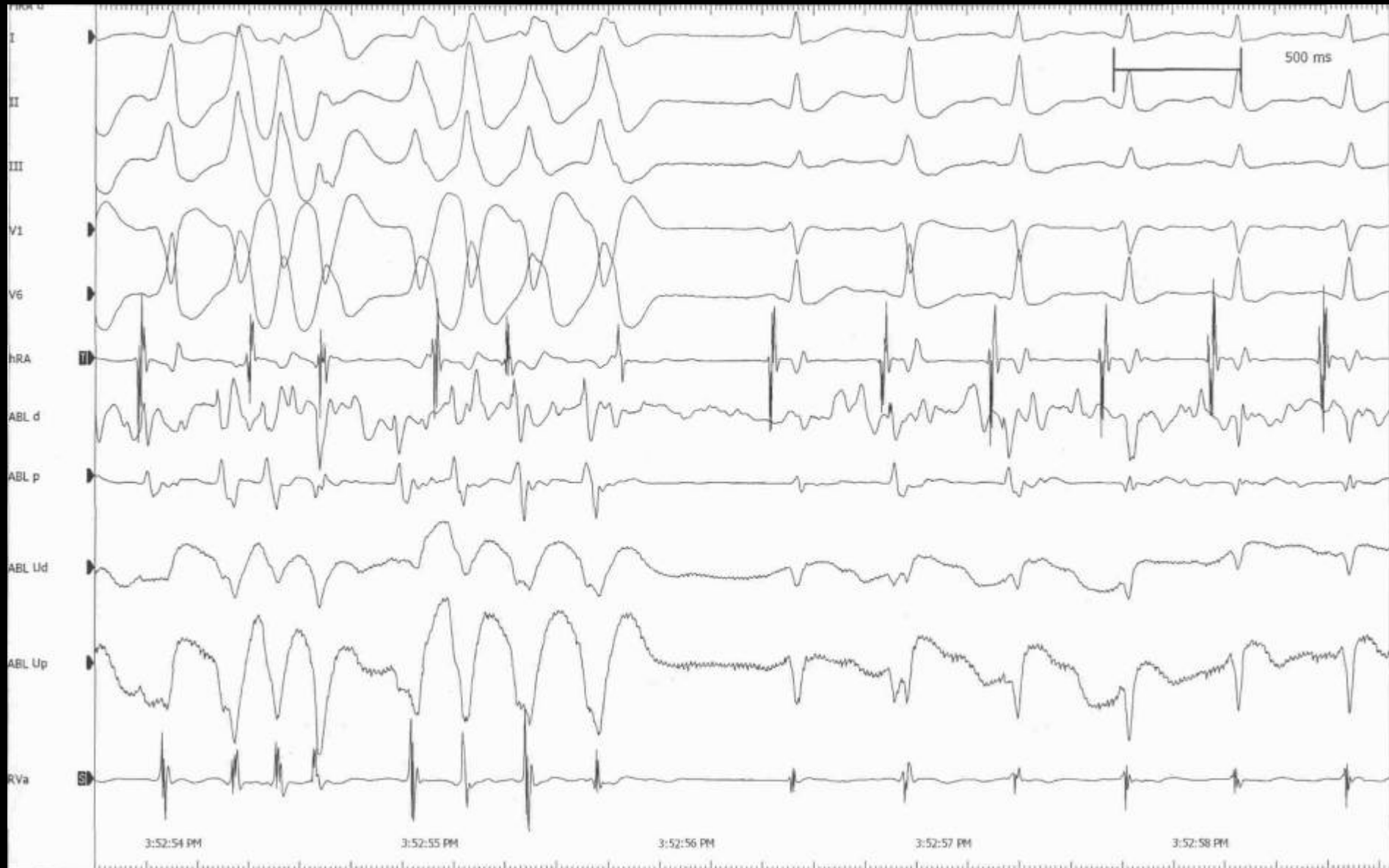


RAO

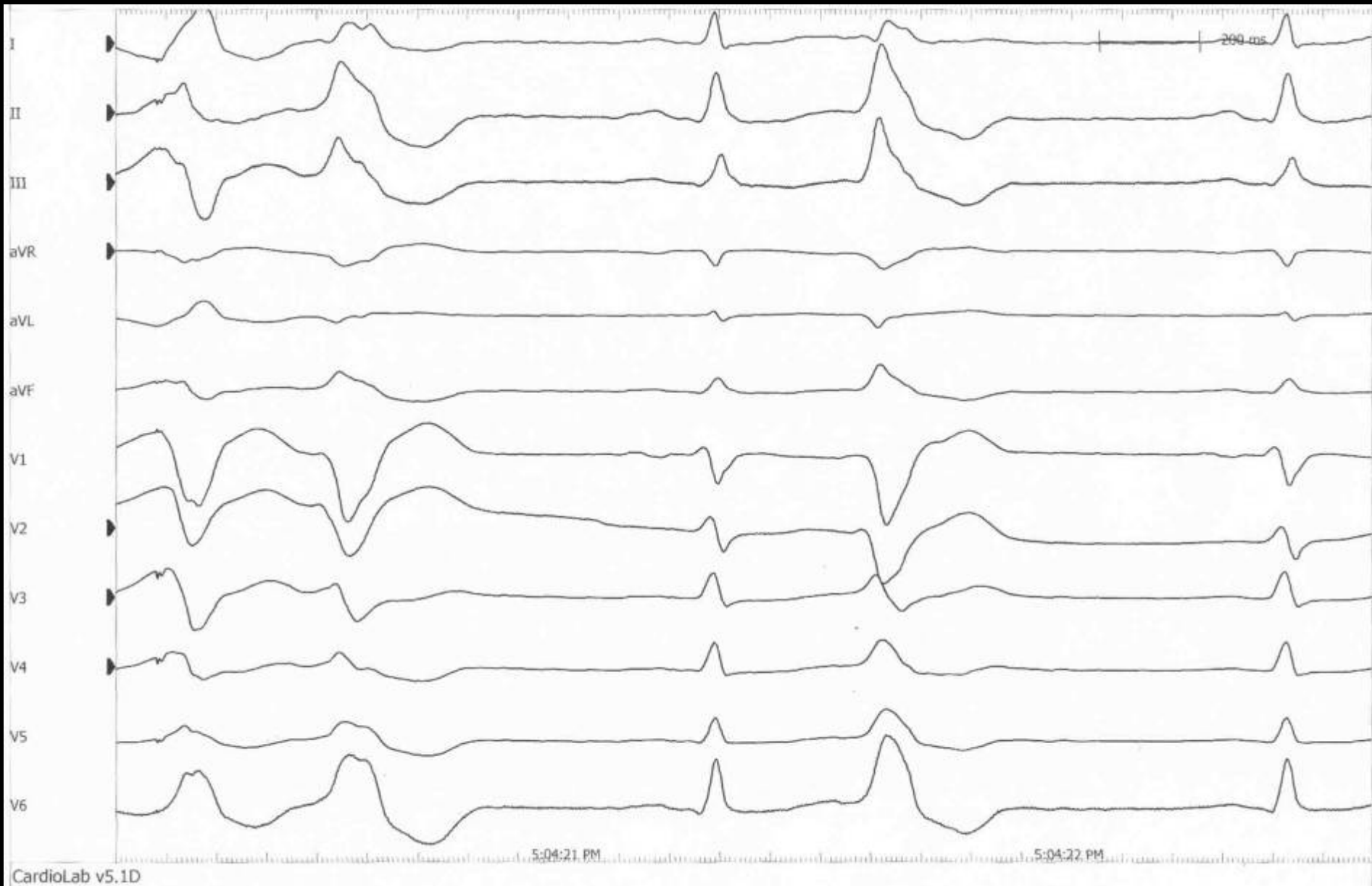


LAO

Termination during first ablation



Persistent VPDs after ablation



VPD ablation site



Conclusion

- After total of 3 ablations, no inducible VT with up to quadruple extrastimuli or burst pacing on up to 4mcg/min Isuprel, 1mg atropine, and up to 2mg/min epinephrine
- Occasional single VPDs with similar morphology induced with burst pacing on epinephrine
- Discharged in good condition, off of beta blocker
- Given short acting metoprolol to take in case of recurrent palpitations

51 yo man with WCT

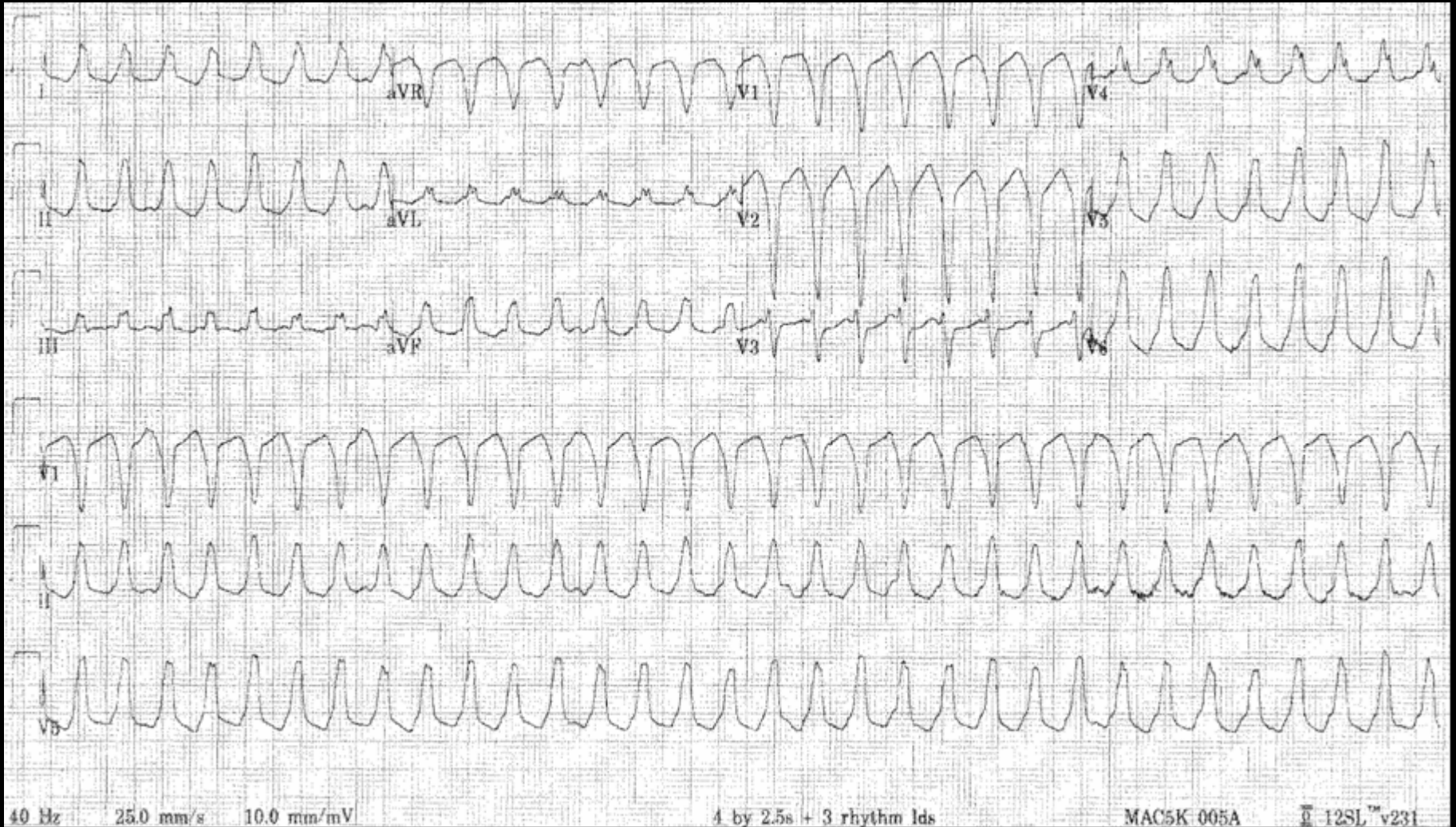
History

- 51 yo man with h/o HTN, no cardiac hx
- Several days of fatigue, malaise
- Worked up with stress test and echo, both unremarkable
- Developed sudden onset of fatigue, weakness, and presented to a local ED

Ancillary History

- Brother died suddenly at age 47, unknown cause
- Father developed heart failure age 53, died of a “heart attack”
- Social History
 - Rare alcohol consumption
 - No tobacco or drug use
 - Physically active

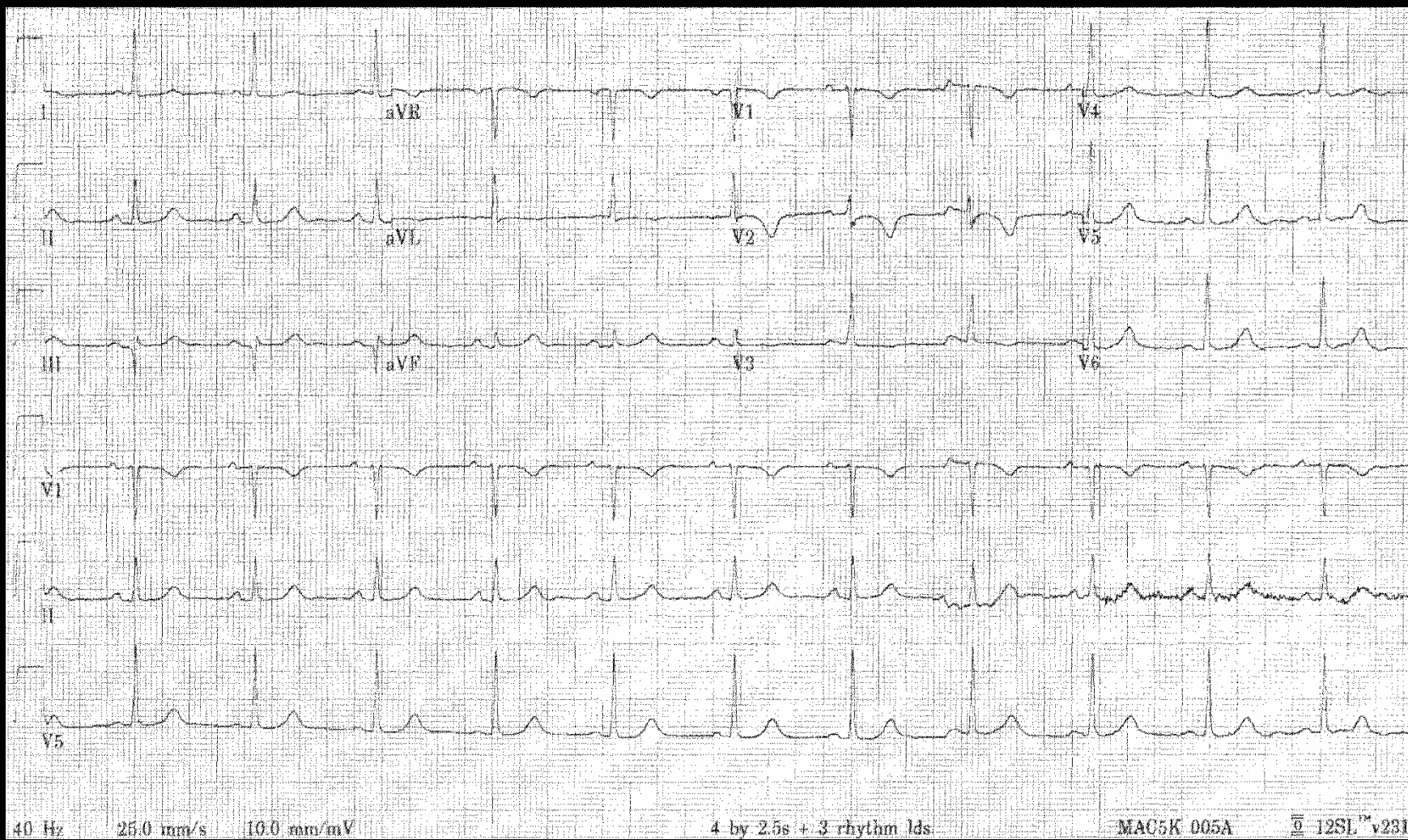
ECG on presentation



Course

- DC CV at 100J failed to convert from WCT
- Adenosine IV administered with no effect
- DCCV at 100J converts to SR

Baseline ECG



Course

- Transferred to BIDMC for further evaluation and EPS
- SAECG done prior to EPS

Analysis Filter : 40-250Hz
Std. QRS Duration (unfiltered) : 98 ms
Total QRS Duration (filtered) : 127 ms
Duration Of HFLA signals < 40μV : 51 ms
RMS Voltage in terminal 40 ms : 7 μV
Mean Voltage in terminal 40 ms : 5 μV

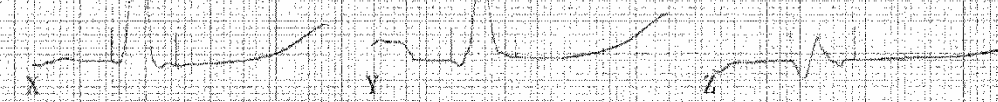
Number Of Beats Averaged: 94
Number Of Beats Detected: 97
Noise Level (Std. Devn.) : 0.30 μV

Abnormal Filtered QRS > 114ms

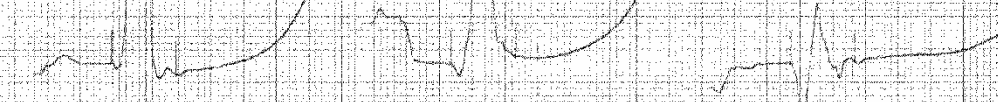
Abnormal duration < 40: 38ms

Abnormal RMS voltage < 20uV

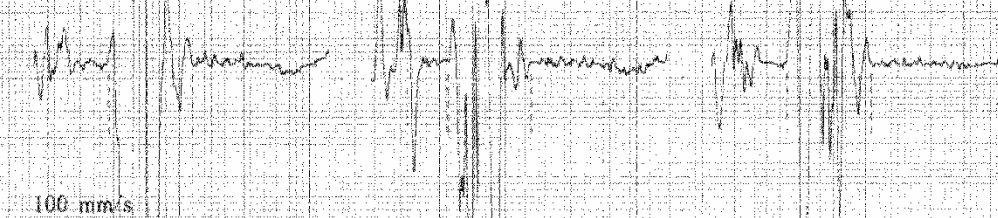
0.01-250Hz
20mm/mV



0.01-250Hz
50mm/mV



40-250Hz
1000mm/mV



1000mm/mV

μV

100

90

80

70

60

50

40

20

20

10

0

VM

200 mm/s

MAC5K 005A

Hi-Res

Standard Report

100 mm/s

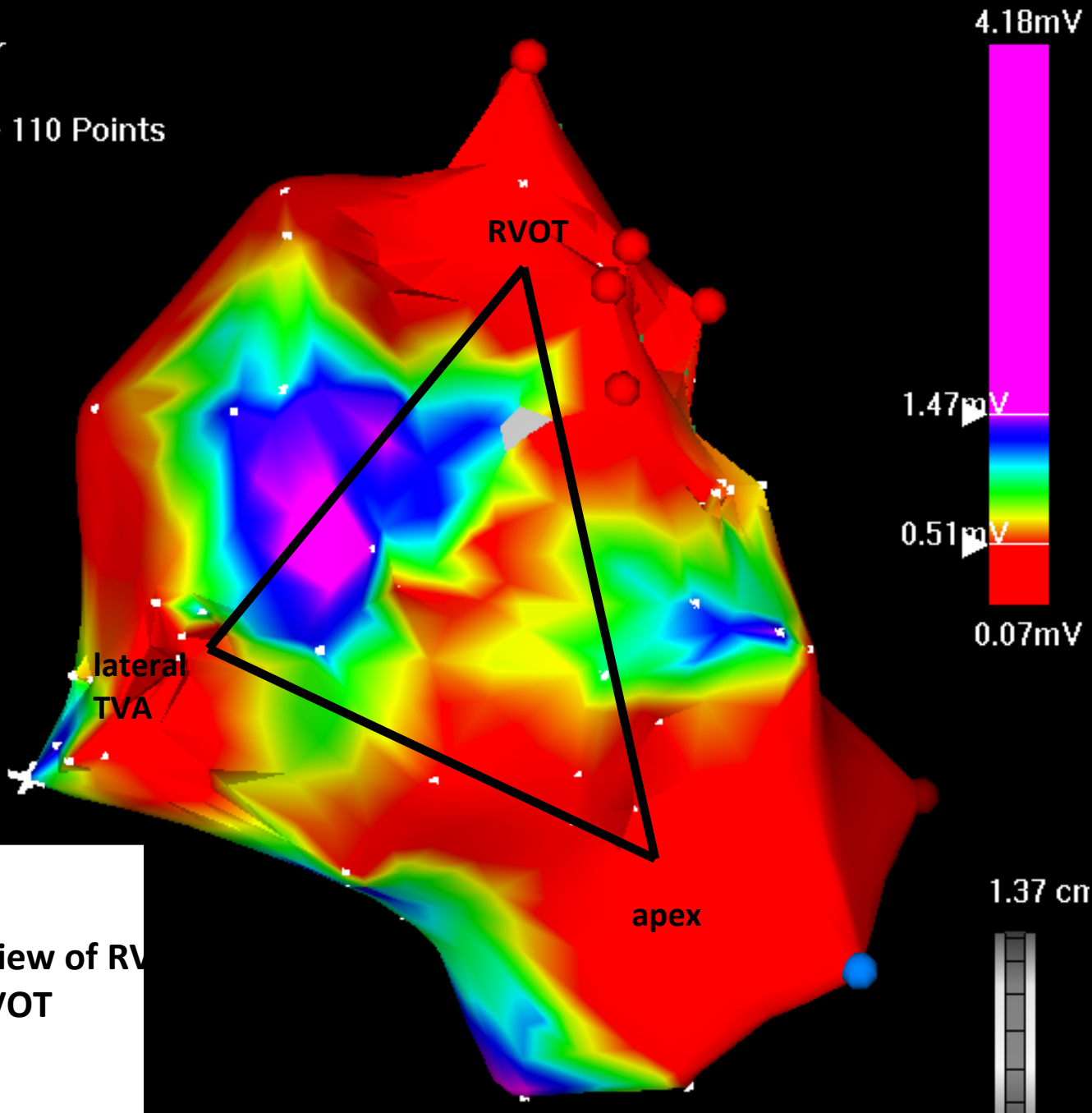
SAECG

- SAECG detects slow conduction or delayed depolarization potentials (as ventricular late potentials) through myocardium that are disrupted by scar, fibrosis using computerized averaging of multiple ECG complexes
 - Slowed propagation in the myocardium → delayed ventricular activation → substrate for reentrant arrhythmias
 - Used to risk stratify patients with ventricular arrhythmias and risk of sudden death
- Markers of high risk for sudden death (in pts with CAD) include (Gomes, circulation 2001. "MUSTT" study)
 - Filtered QRS duration (fQRS) >114ms (sens 91%, spec 90%)
 - Low Amplitude Signal duration (LAS40) ≥38ms
 - High frequency root mean square (RMS40) <20μV

Analysis Filter : 40-250Hz		Number Of Beats Averaged: 555
Std. QRS Duration (unfiltered) : 111 ms		Number Of Beats Detected: 642
Total QRS Duration (filtered) : 139 ms		Noise Level (Std. Devn.) : 0.46 μV
Duration Of HFLA signals < 40μV : 73 ms		
RMS Voltage in terminal 40 ms : 4 μV		
Mean Voltage in terminal 40 ms : 3 μV		

Bipolar

1-1-ReMap > 110 Points

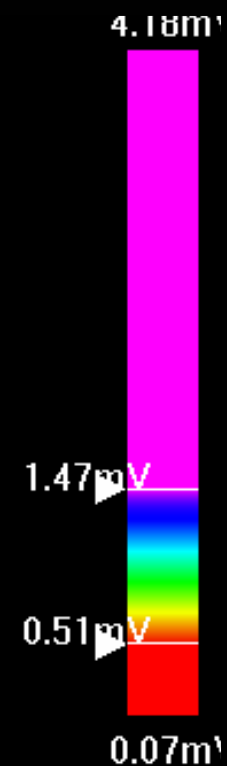
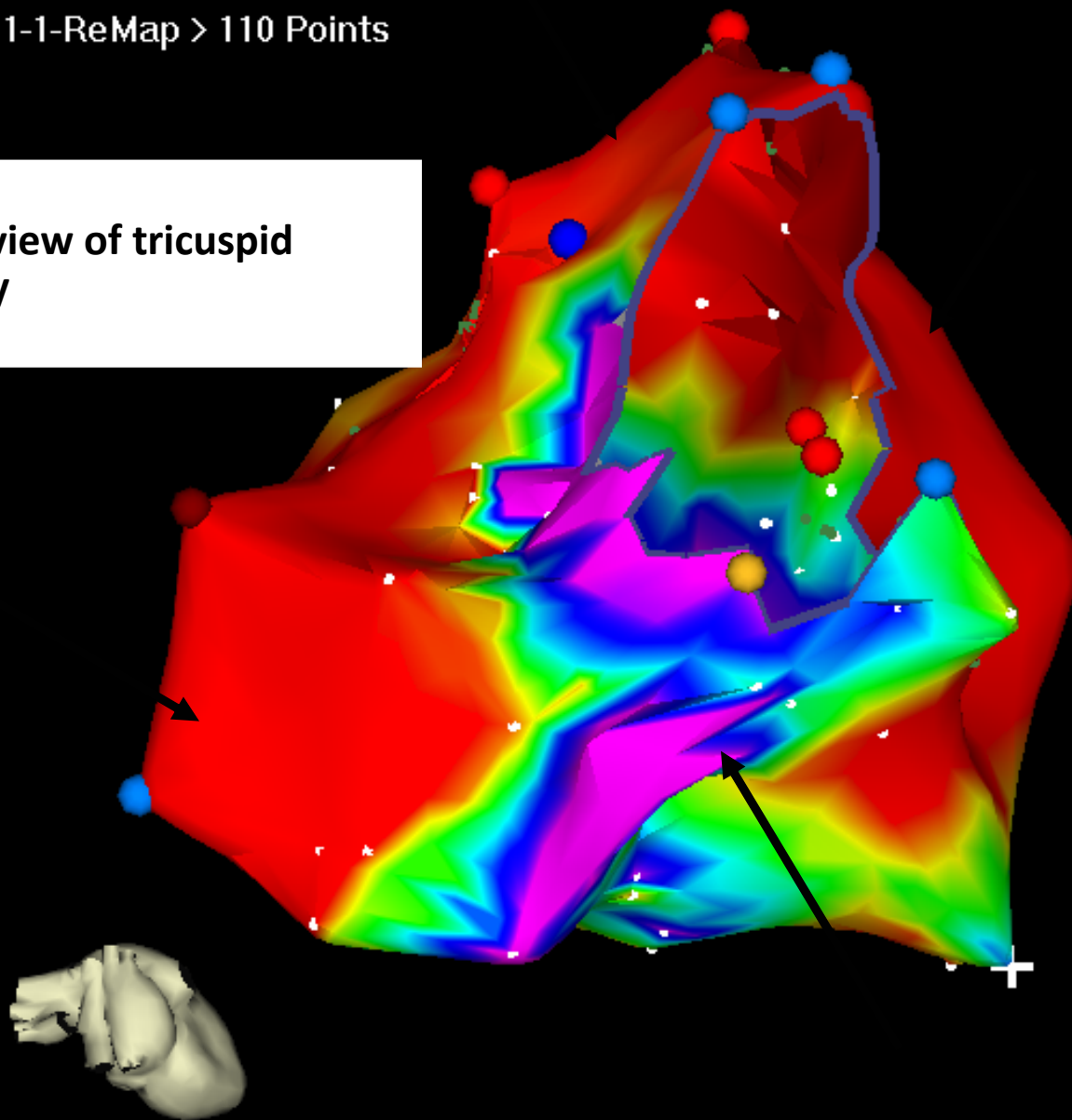


Modified RAO view of RV
free wall and RVOT

Bipolar

1-1-ReMap > 110 Points

LAO-Septal view of tricuspid valve and RV



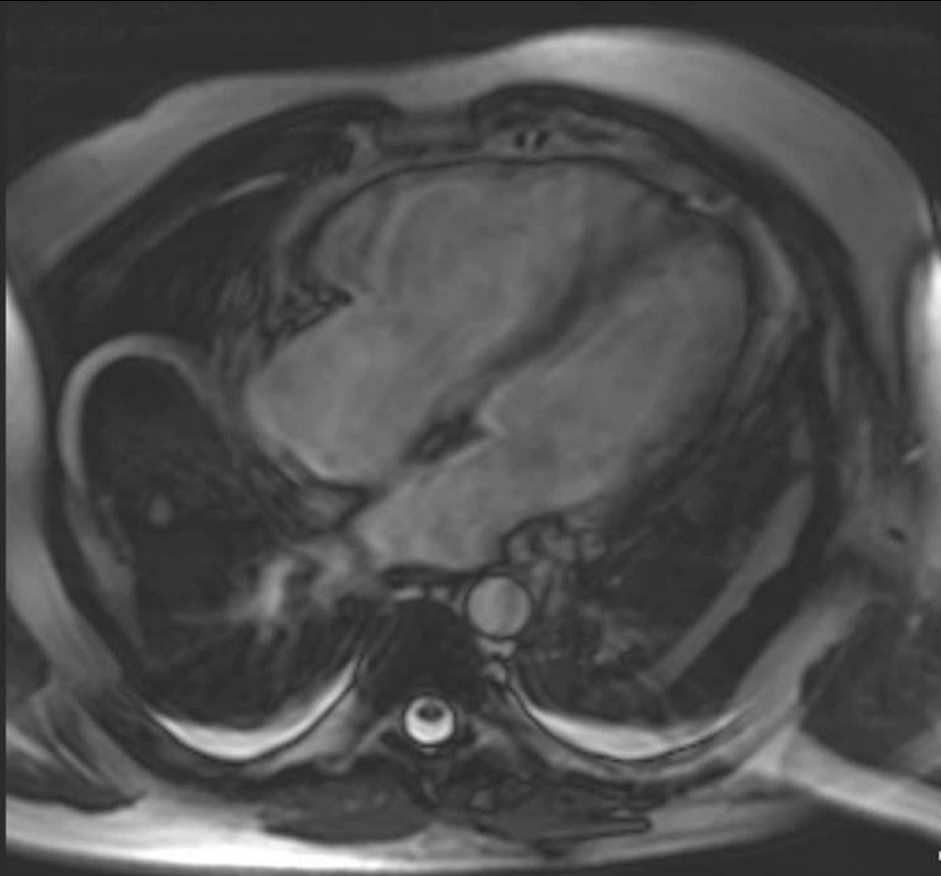
1.37 c



RV Substrate Map

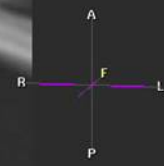
- Diffuse patchy scar
- Scar most dense at RV apex, basal free wall, and RVOT
- “Triangle of dysplasia”

Sc 4.1/1
B-TFE / FFE/M
Td 000 ms

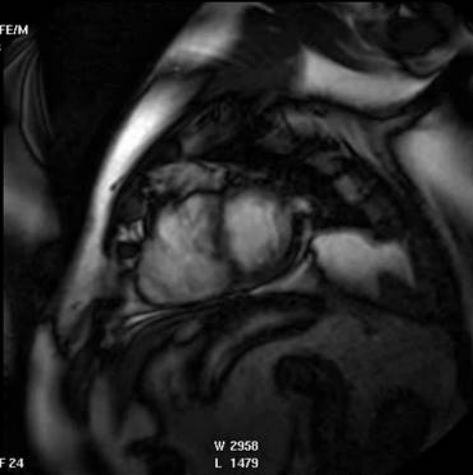


AP 11°
RL 10°
FH -2°
A 20 L 34 F 4

W 2000
L 651



Sc 5.1/4
B-TFE / FFEM
Td 000 ms



AP 21°
FH 42°
A 77 L 89 F 24

W 2958
L 1479



Sc 5.1/5
B-TFE / FFEM
Td 000 ms

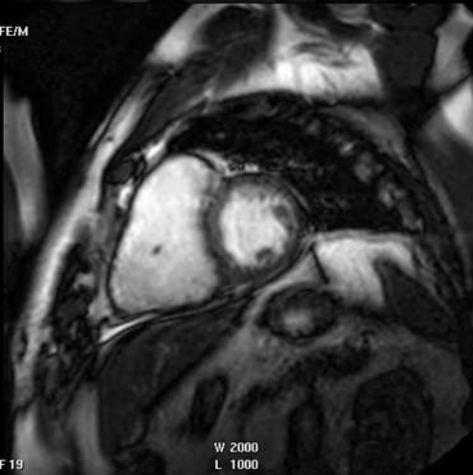


AP 21°
FH 42°
A 70 L 82 F 21

W 2077
L 1038



Sc 5.1/6
B-TFE / FFEM
Td 000 ms

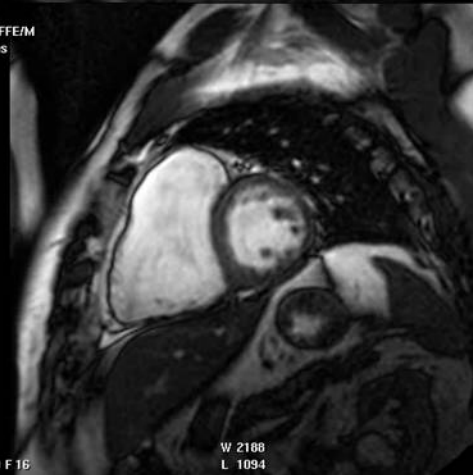


AP 21°
FH 42°
A 62 L 76 F 19

W 2000
L 1000



Sc 5.1/7
B-TFE / FFEM
Td 000 ms

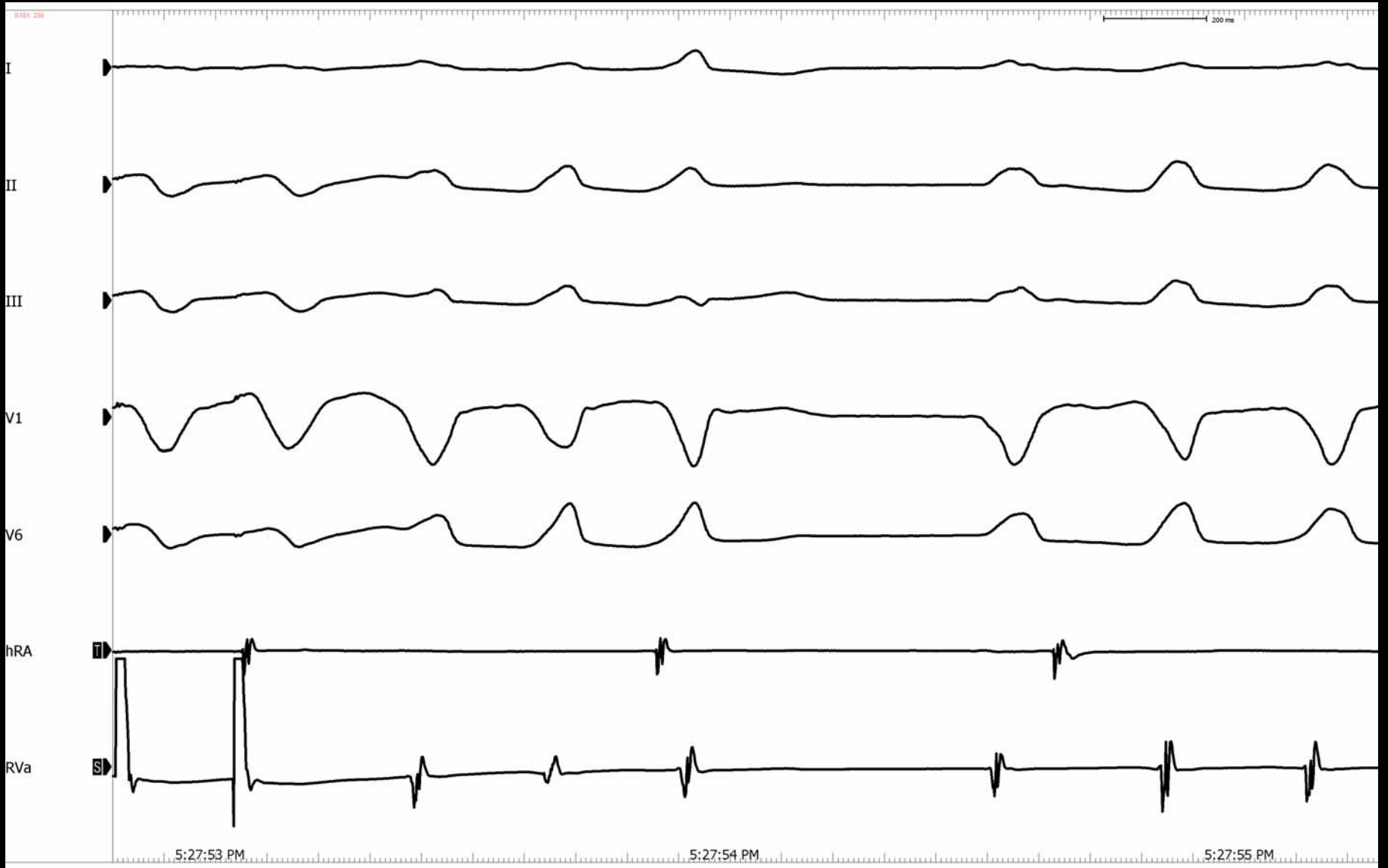


AP 21°
FH 42°
A 55 L 70 F 15

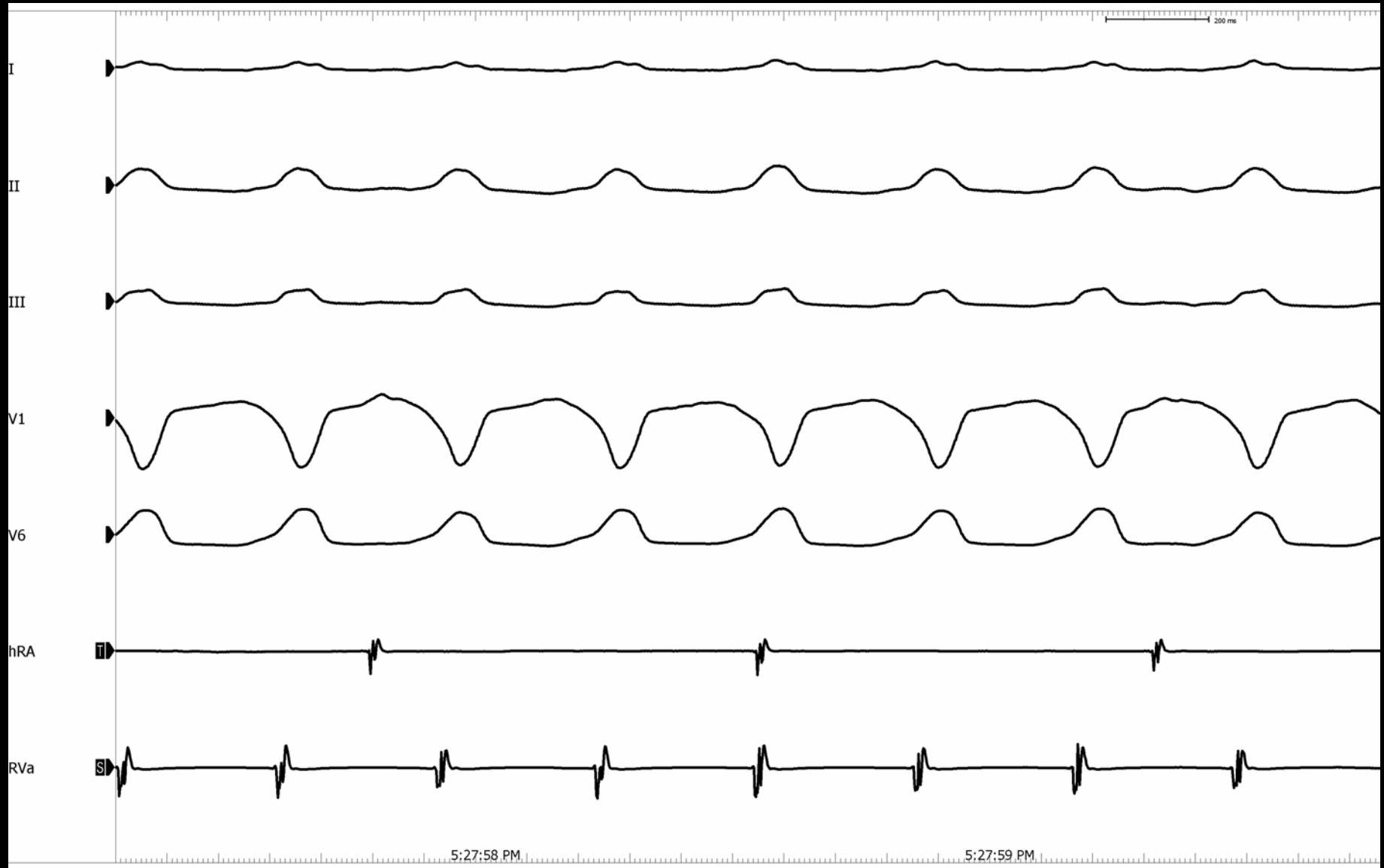
W 2188
L 1094



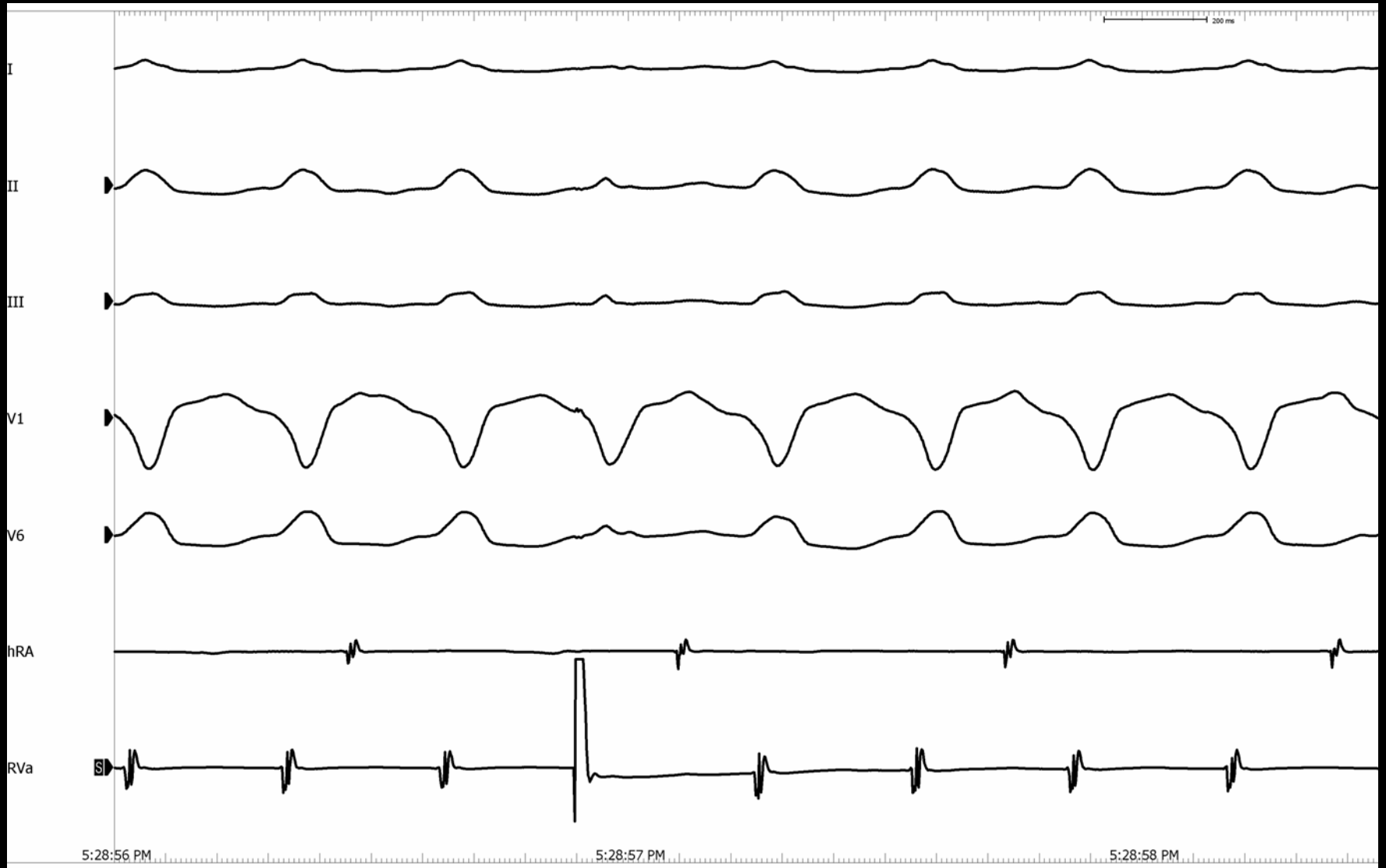
Induction 500-28--230



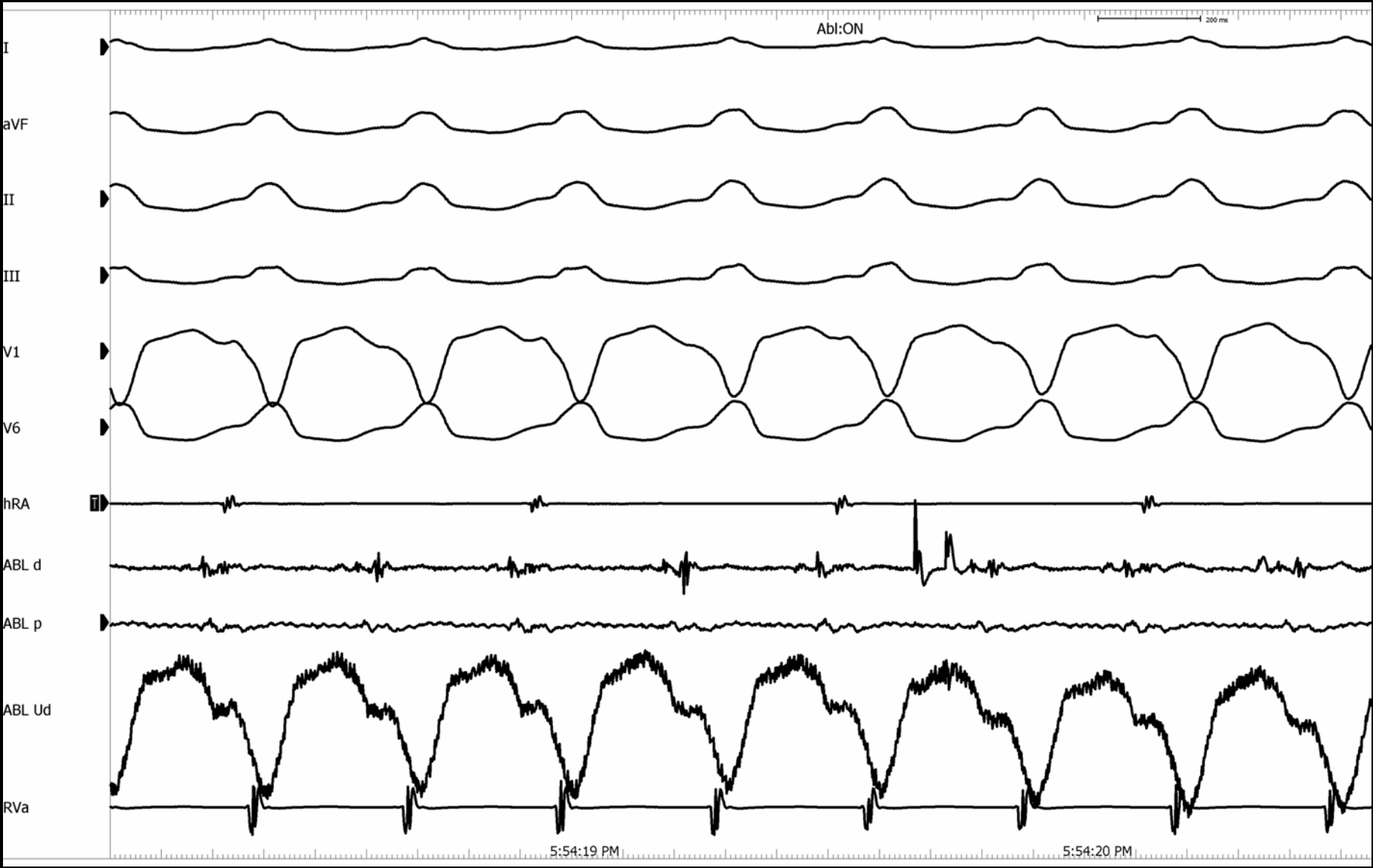
VT CL 305



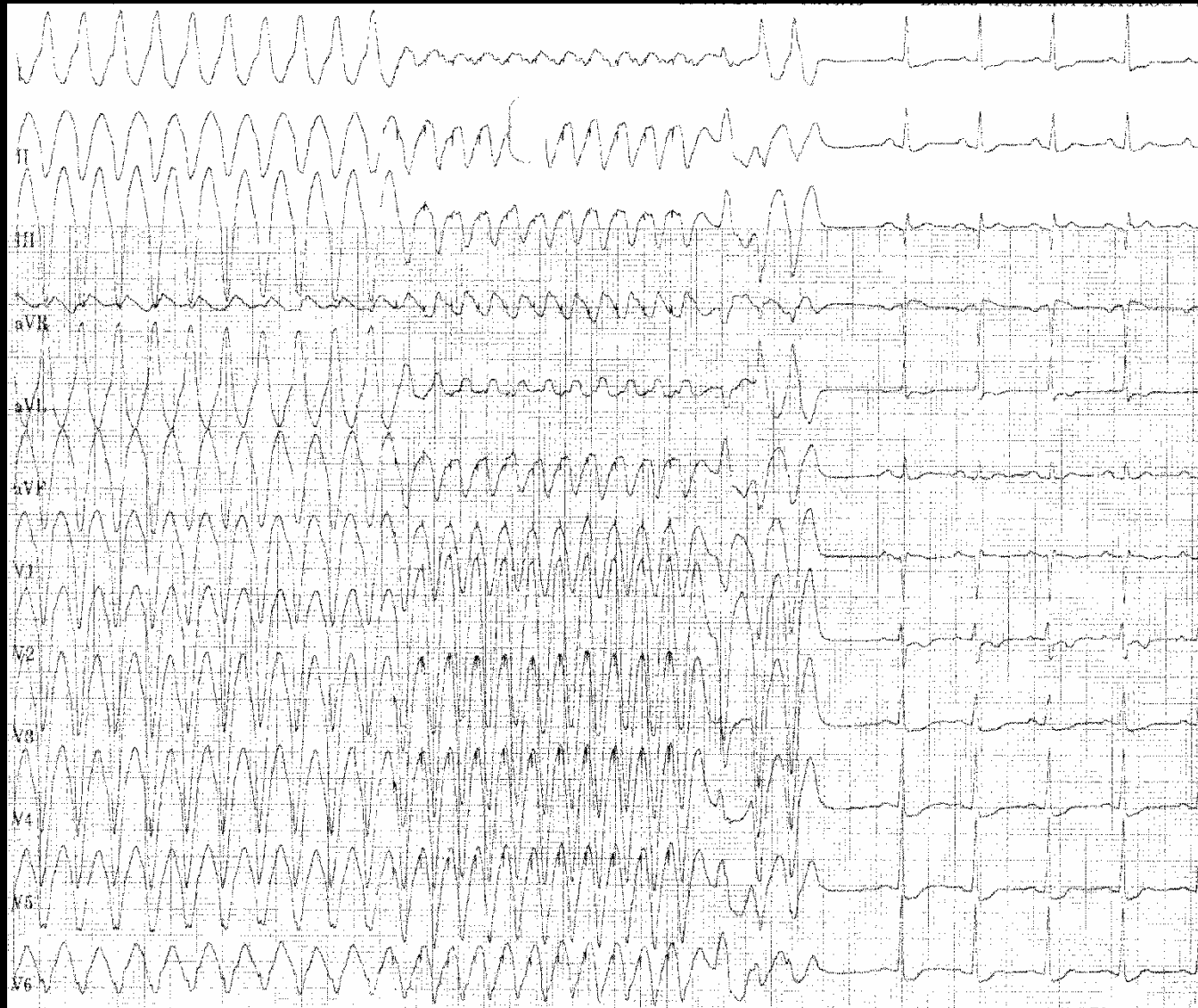
What does this prove?



Ablation in lateral RVOT terminates VT



VT still inducible, pacing induces second VT morphology which breaks with RV pacing



CMR

- LVEF was normal at 64%.
- Dilated right ventricle with severely depressed systolic function and thinning of the free wall.
- The RVEF was severely decreased at 10%.
- No MR evidence of right ventricular fatty infiltration/dysplasia.

Subsequent course

- After EPS, underwent implantation of a dual-chamber ICD
- Started on sotalol 120 mg bid for suppression of VT
- 3 months later developed dizziness and subsequent ICD shock
 - Interrogation revealed VT at 210bpm and appropriate shock with conversion to SR
 - Mexilitine added to sotalol for suppression of VT
 - NIPS performed with inducible VT (required aggressive stimulation) and able to be terminated with anti-tachycardia pacing
- Family was screened through Children's Hospital with cardiac MRIs – results unavailable



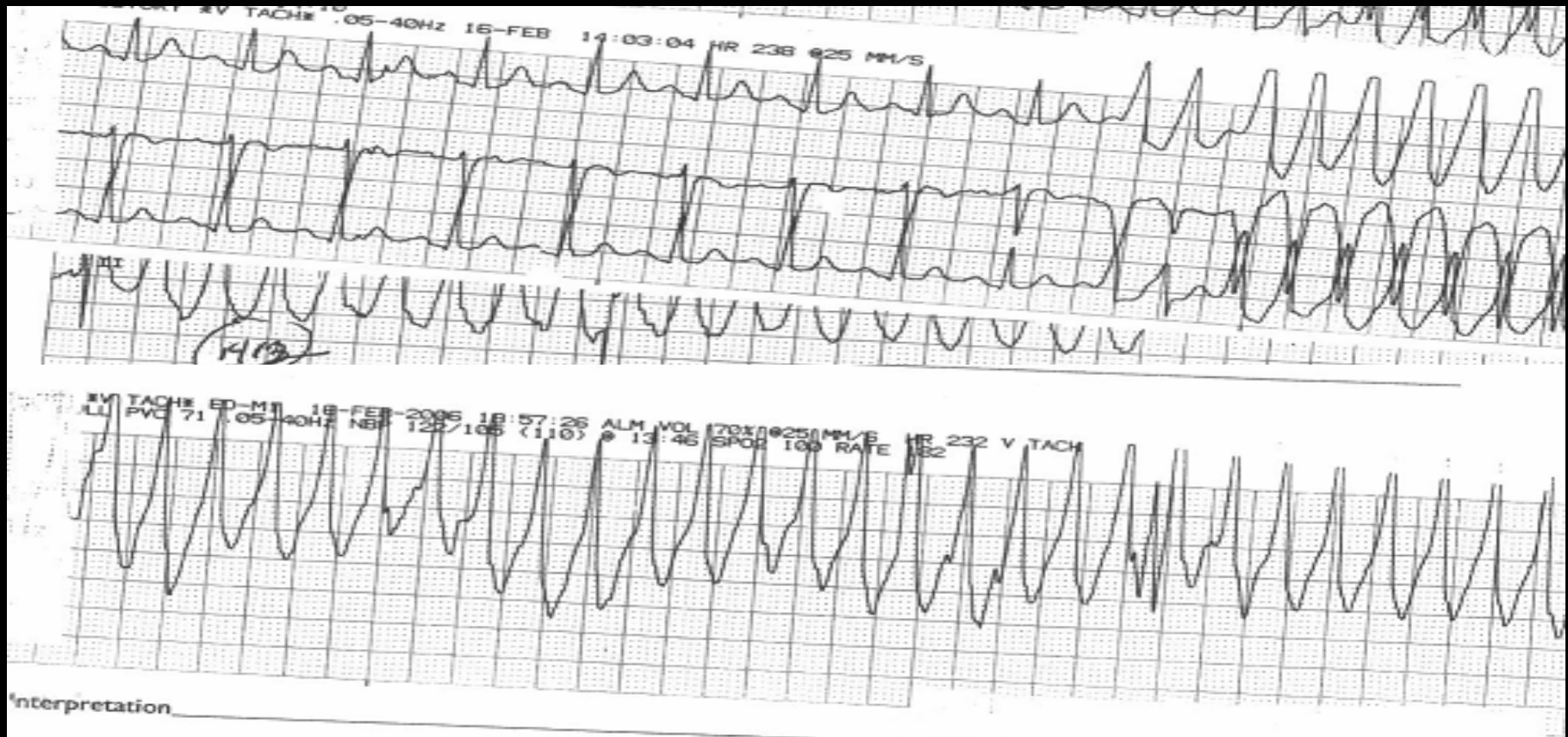
Let's see... tightness in chest... crushing sensation...
difficulty breathing... Good news! According to
my protocol book, we can authorize a cardiology consult!

- 40 year old French male with no significant PMH who has been suffering from palpitations for the past 2 years.
- His palpitations occurred suddenly at random with or without exertion and were often associated with sense of dizziness followed by heartburn sensation without any chest discomfort, dyspnea, nausea, presyncope or syncope.

- For these symptoms, he was seen by a local cardiologist and underwent a battery of tests
 - Holter: unremarkable
 - ETT stress test: unremarkable
 - Echocardiogram: unremarkable
- Seen by Neurology
 - EEG: negative
- Seen by Endocrinology to rule out undiagnosed hypoglycemic episodes
 - BG and insulin levels normal
- Seen by GI
 - EGD: mild gastritis
 - Colonoscopy: unremarkable

- After these tests, he was reassured by his PCP without additional therapy.
- He then did fine with only minimal symptoms that were never severe enough to cause syncope or chest pain.
- Beginning on February 2006, however, his symptoms began to worsen. Instead of tolerable palpitations, his symptoms now became more frequent (now occurring daily) and were described as washing machine on his chest followed by a head rush/dizziness, and heartburn like symptoms.

- On 2/16/06, while at work, he noted a sensation of “zoning out” with tunnel vision and felt as if he was about to pass out. While at home, his wife (RN) noted that he looked pale and checked his pulse that was “too high to count”
- Presented to Lowell General Hospital ED with the following rhythm on a telemetry with hemodynamic stability:



- He was 'expeditiously' transferred to Lahey Clinic where his blood work, ECG, and TTE were repeated and were again unremarkable.
- He was subsequently evaluated by EP service and underwent an EP study that revealed no evidence of inducible VT.
- He was then discharged home with Toprol XL 25mg qd and an event monitor.

Comments ?

- Choice of medication
- Additional studies or work up
- Would you have sent him home?

ID #: 025605506

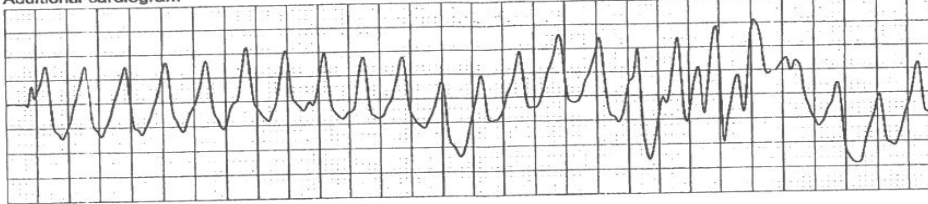
Page: 2

Event Date: 03/11/2006 08:29 am (EST)

Note: Listed Times are in (EST)

Additional cardiogram

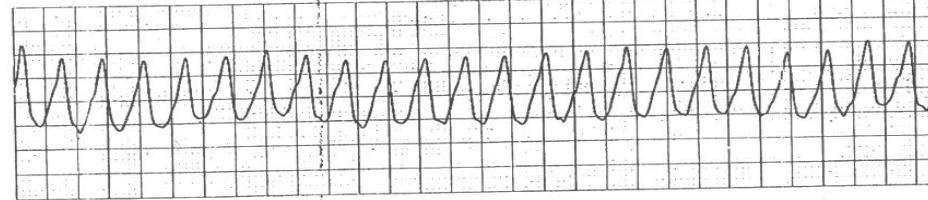
Received: 08:34:01 AM 25 mm/sec, 8 mm/mV Continues->



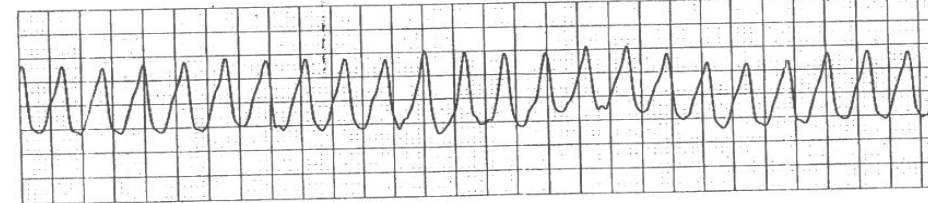
Received: 08:34:07 AM 25 mm/sec, 8 mm/mV Continues->



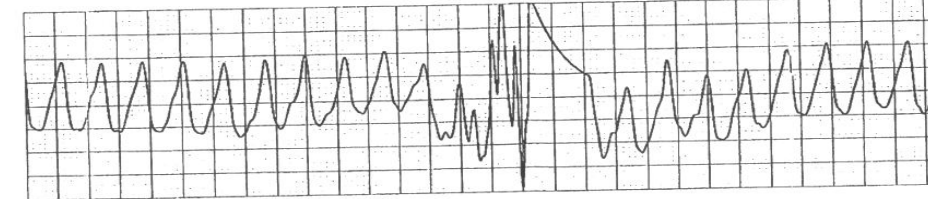
Received: 08:34:13 AM 25 mm/sec, 8 mm/mV Continues->



Received: 08:34:19 AM 25 mm/sec, 8 mm/mV Continues->



Received: 08:34:25 AM 25 mm/sec, 8 mm/mV Continues->



Work up in Lowell

- WBC 5.5, Hb 15.9, Plt 237
- Na 141, K 3.8, BUN/Cr 14/1.1
- TSH 1.57, Tnl 0.12
- Echocardiogram (3/13)
 - Global LV HK with EF 50%.
 - Normal RV function
 - No AI, MR
- Exercise MIBI
 - 13 minutes on Bruce with no sx or ECG changes
 - Non-sustained VT at 250 bpm for 10 seconds @ 4 minute recovery

ETT at Lowell

Berard Michael
145281

10-second Write Screen manual

HR 105
Target HR 136
Previous BP —/—

RPE —
METs(a) 14.8
BP —/—
HRxBP —

Baseline Wander Filter On
Muscle Artifact Filter On
Line Filter Off
40Hz Low Pass Filter Off

Protocol Bruce
Stage recovery 04:28
TM Speed 0.0 mph
TM Grade 0.0%

II



V2



V5



208-330-9036.

Plan: Transfer to BIDMC for
second opinion

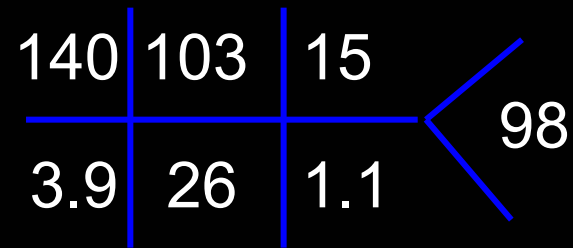
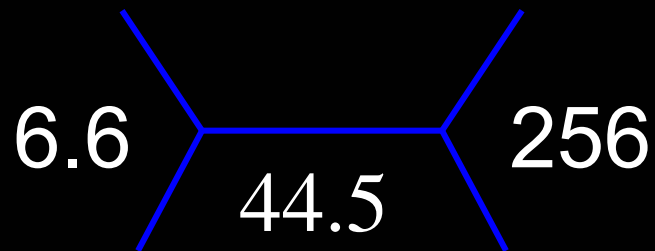
Other Pertinent Information

- PMH
 - GERD
- Medications upon transfer (meds at home=none)
 - ASA 325mg qd
 - Metoprolol 25mg bid
- Allergies: None
- Social History:
 - Married with two children. Wife is a nurse at Lowell
 - Works at a biotech company in Burlington, MA
 - Never smoked but drinks occasional beer
 - Denied any illicit drug use
- Family History
 - No sudden cardiac death
 - Baldness and Hyperlipidemia in family
 - Grandfather with CABG in 50s

Physical Exam

- BP 108/65, P 72, T 98, Pox 98% RA
- Gen: young, cheerful Caucasian male in NAD. A+Ox4
- Skin: normal
- HEENT: no elevated JVP or carotid bruits
- CV: RRR, normal S1, S2, no mgr
- Abd: +BS, NT, ND
- Ext: no edema. 2+ pulses on femoral, PT, and DP bilaterally

Labs upon transfer



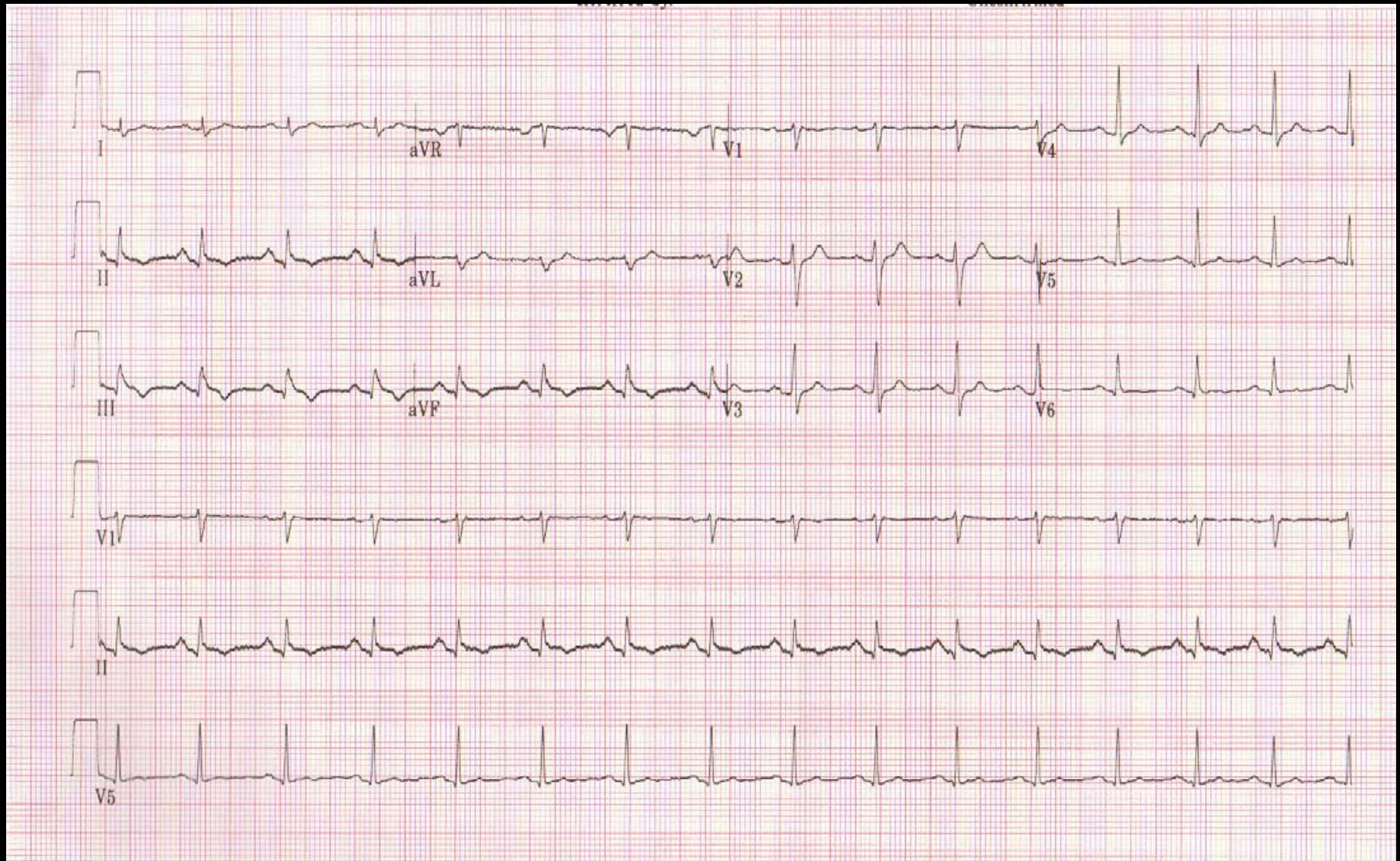
INR 1.2

Mg 2.1, Ca 9.1, P 3.9

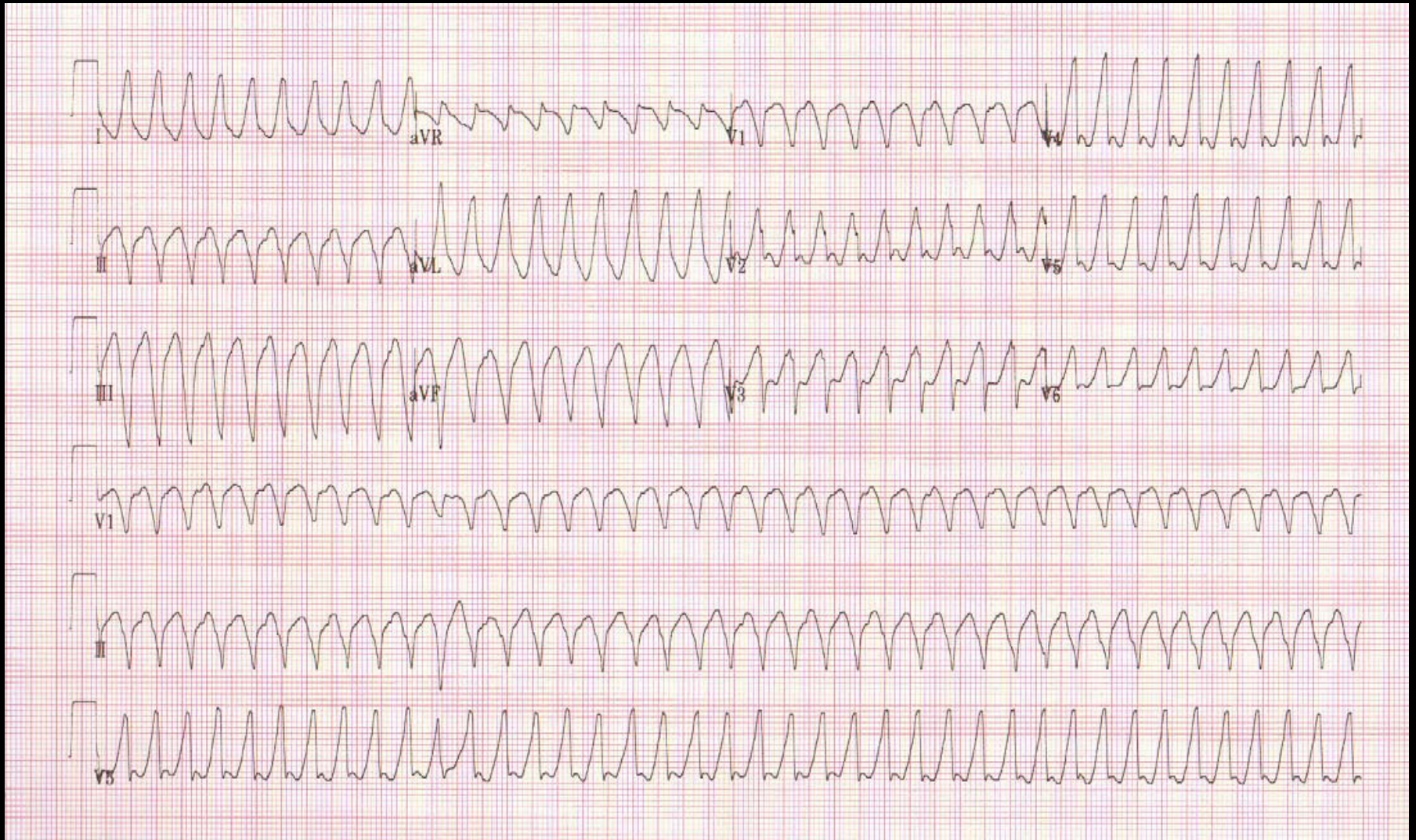
CXR



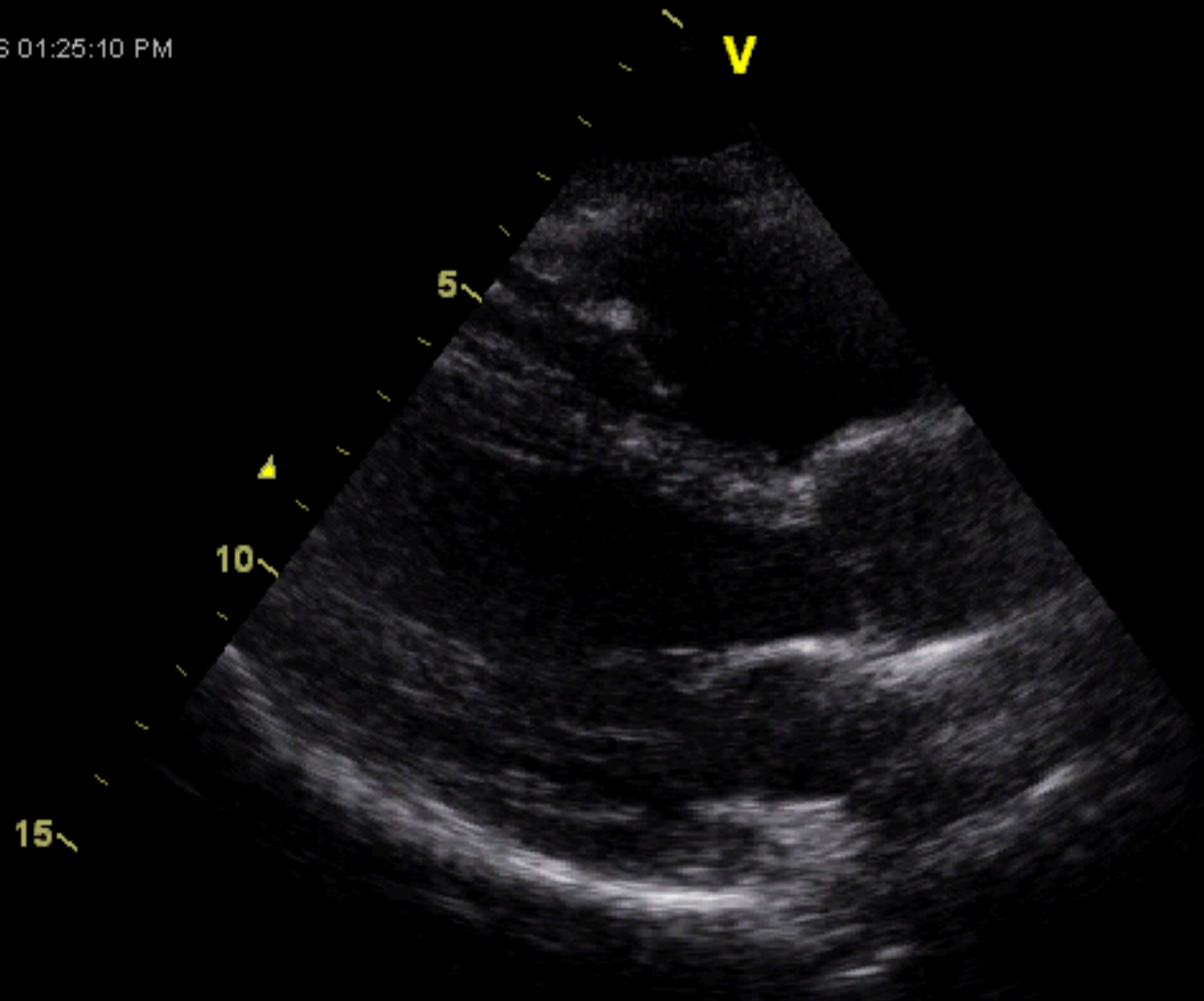
ECG on admission



His VT on 12 lead ECG

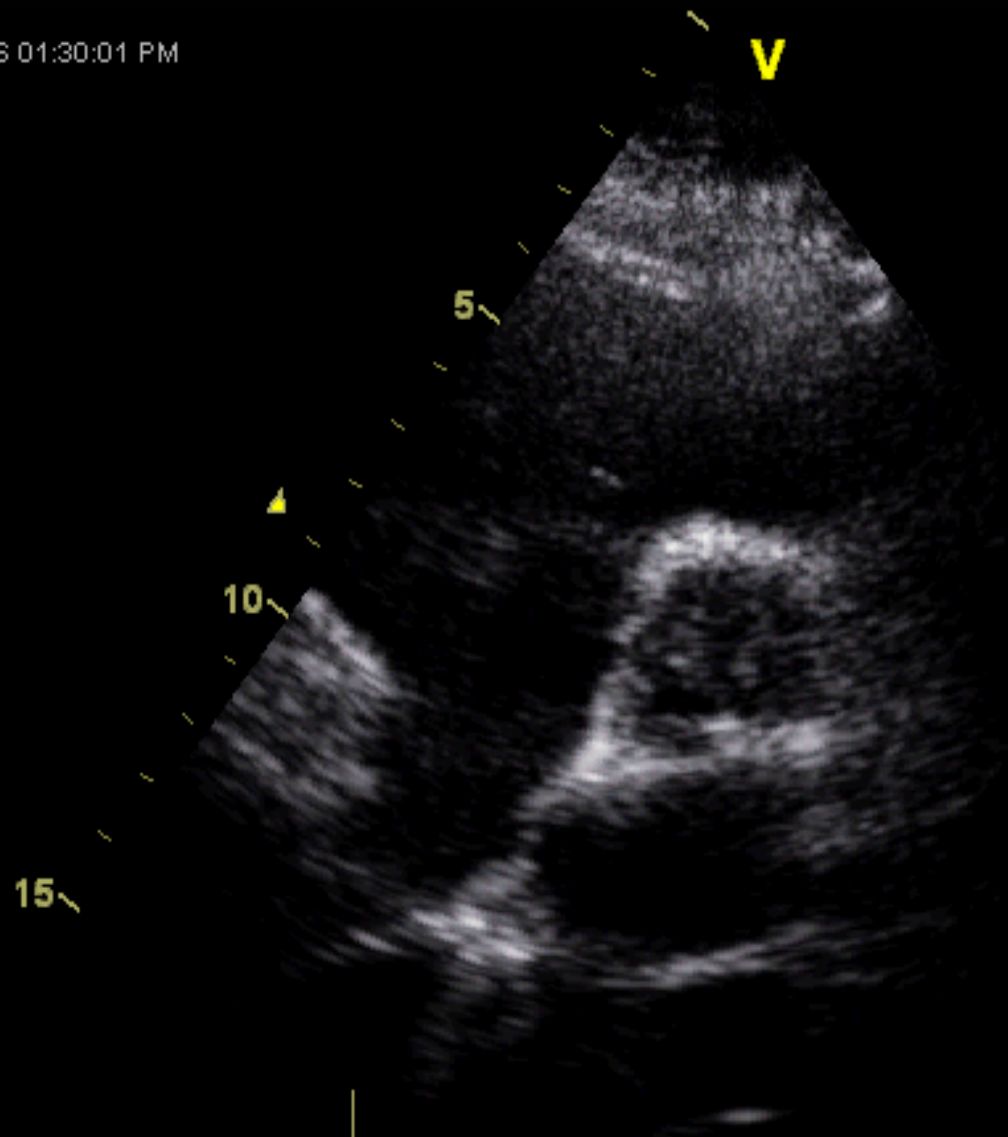


03/13/2006 01:25:10 PM



2:88 60 HR

03/13/2006 01:30:01 PM



68
1:54 HR

03/13/2006 01:31:27 PM

V

5

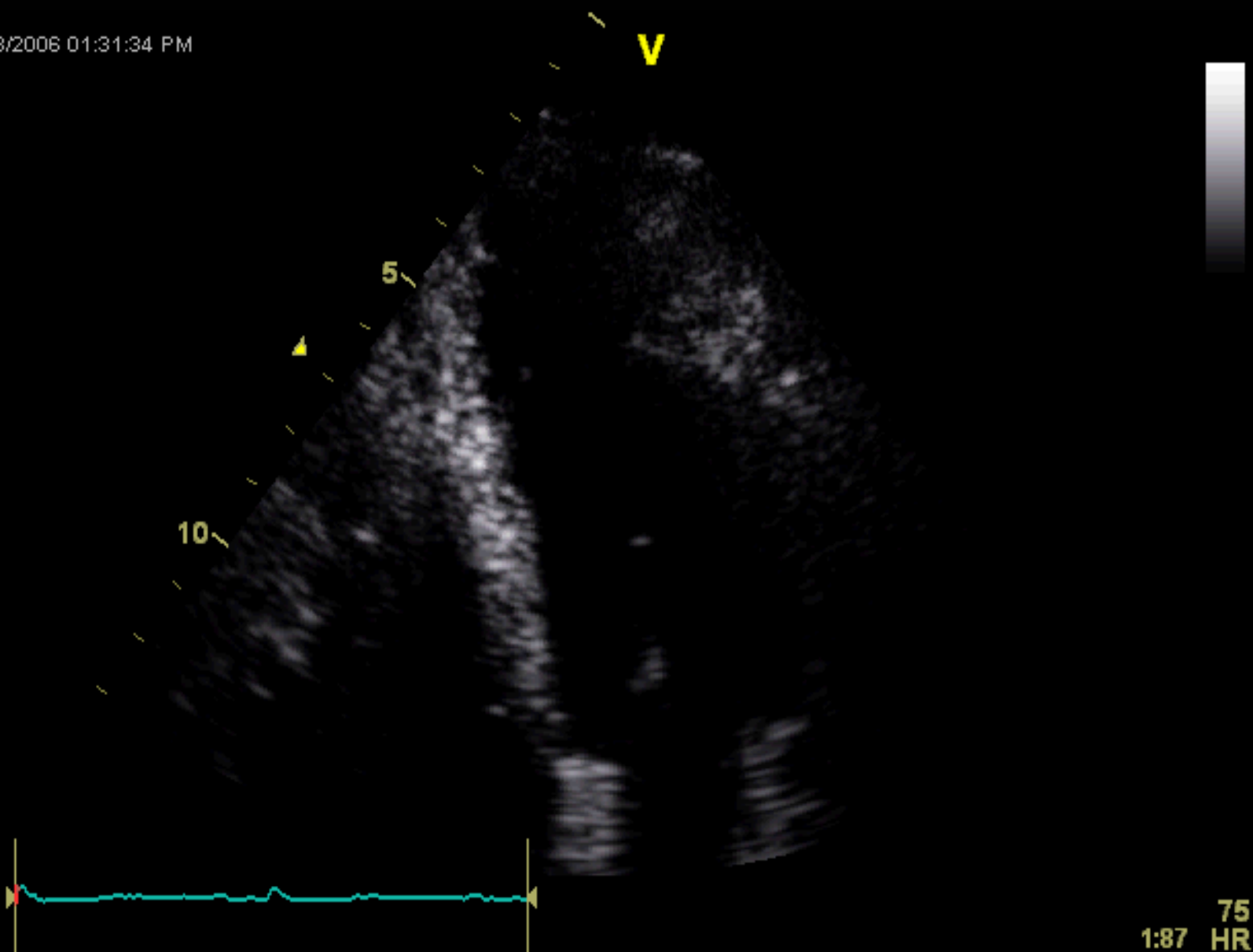
10

15



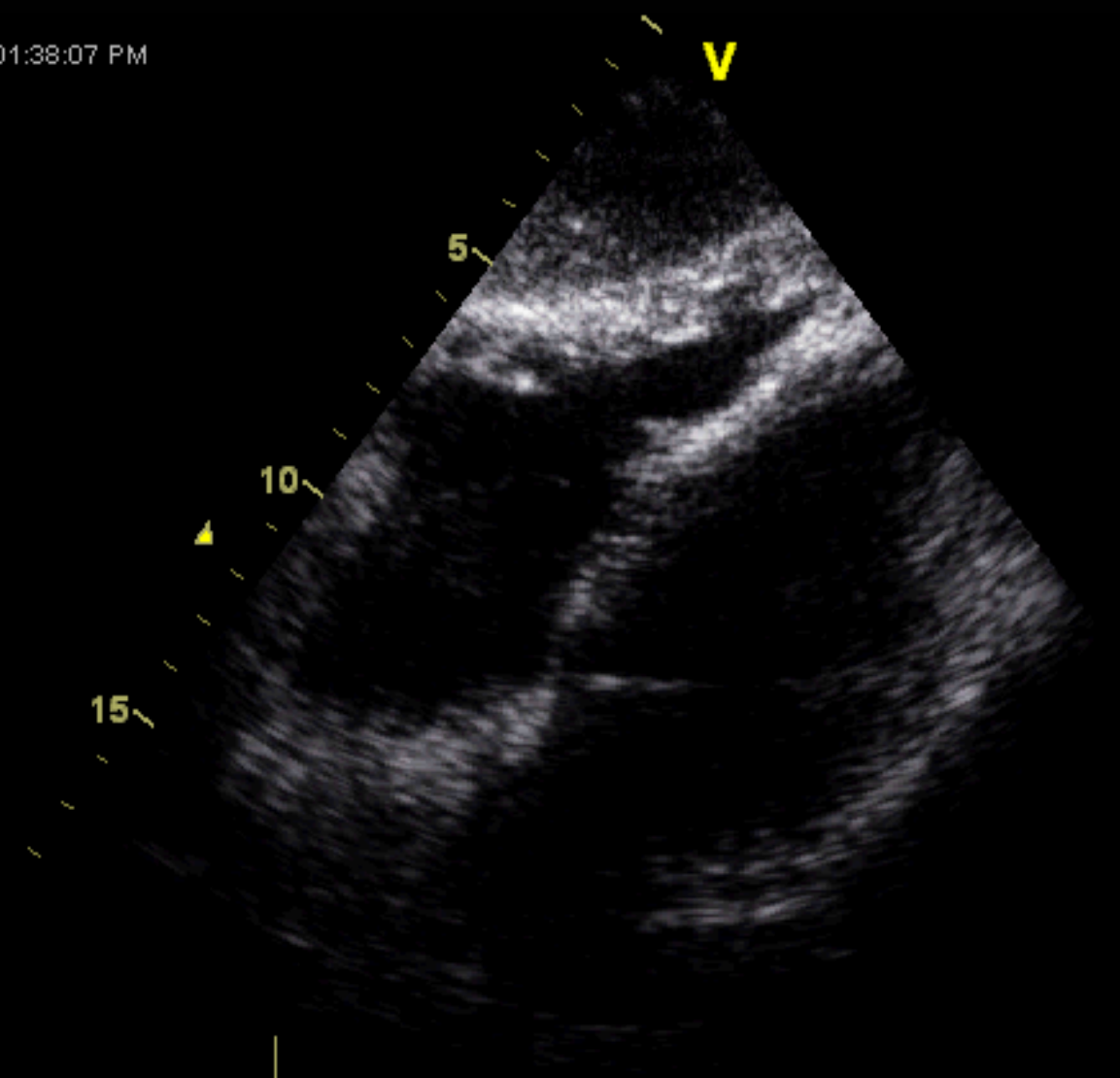
1:73 83 HR

03/13/2006 01:31:34 PM



75
1:87 HR

03/13/2006 01:38:07 PM

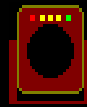


69
1:46 HR

Echocardiogram-Final

- LVEF > 55%
- No LV wall motion abnormalities
- Isolated, moderate global RV free wall hypokinesis
- Normal valvular function

Catheterization



Right Heart Catheterization

**HEMODYNAMICS RESULTS

BODY SURFACE AREA: 1.91 M2

HEMOGLOBIN: 15.9 gms %

FICK

**PRESSURES

RIGHT ATRIUM {a/v/m}	16/10/6
RIGHT VENTRICLE {s/ed}	27/10
PULMONARY ARTERY {s/d/m}	23/9/17
LEFT VENTRICLE {s/ed}	122/20
AORTA {s/d/m}	117/74/94

**CARDIAC OUTPUT

HEART RATE {beats/min}	65
RHYTHM	SINUS
CARD. OP/IND FICK {l/mn/m2}	5.68/2.96

**RESISTANCES

SYSTEMIC VASC. RESISTANCE	1239
CONTINUE <RETURN>	

**% SATURATION DATA (NL)

SVC LOW	77
PA MAIN	80
AO	99

Cardiac Catheterization-Final

- Normal coronary arteries
- Normal LV gram with EF>55%. No MR
- Normal filling pressures
- **Global RV hypokinesis**

Signal Averaged ECG

Analysis Filter : 40-250Hz
 Std. QRS Duration (unfiltered) : 111 ms
 Total QRS Duration (filtered) : 139 ms
 Duration Of HFLA signals < 40 μ V : 73 ms
 RMS Voltage in terminal 40 ms : 4 μ V
 Mean Voltage in terminal 40 ms : 3 μ V

Number Of Beats Averaged: 555
 Number Of Beats Detected: 642
 Noise Level (Std. Devn.) : 0.46 μ V

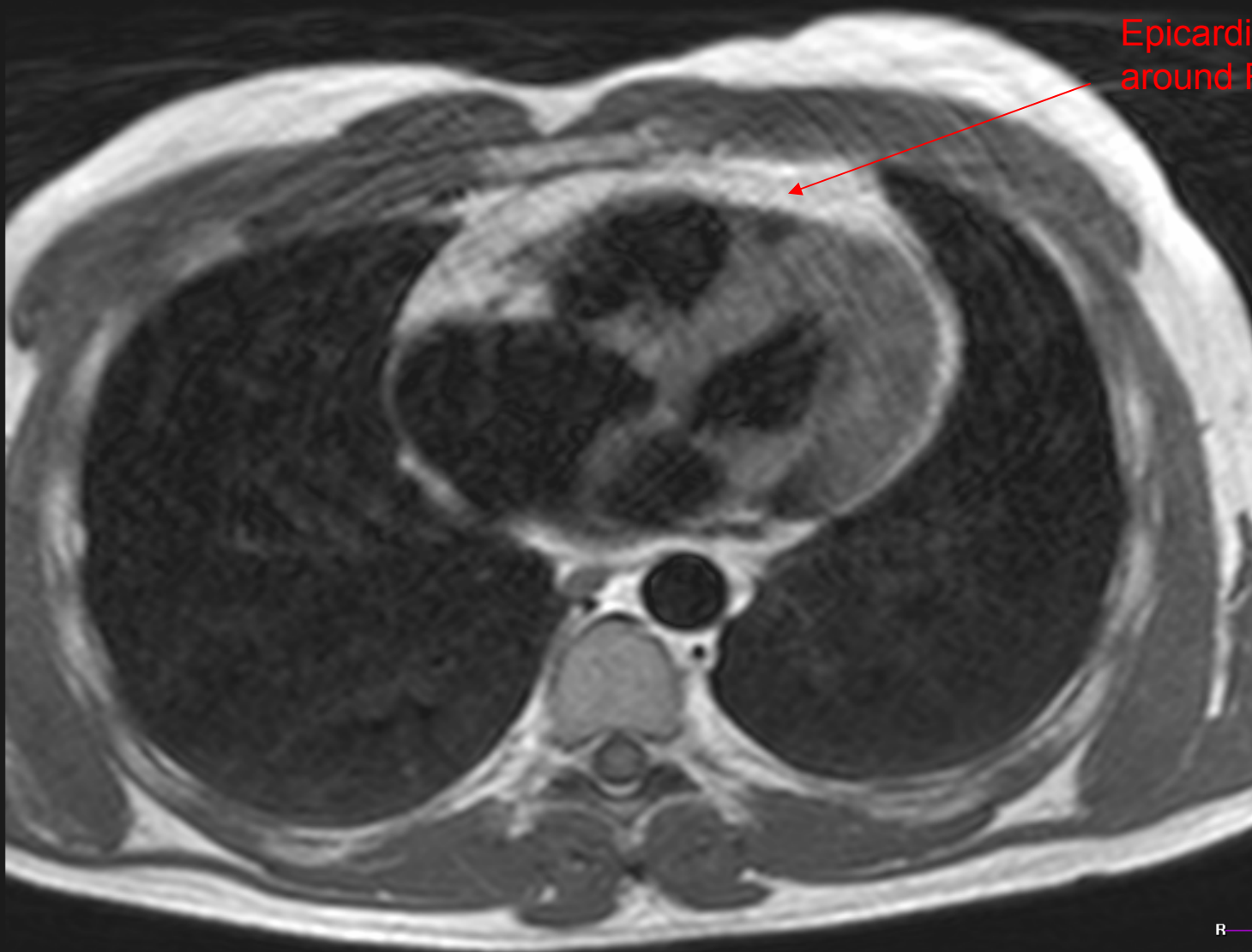


SAECG

- SAECG detects slow conduction or delayed depolarization potentials (as ventricular late potentials) through myocardium that are disrupted by scar, fibrosis using computerized averaging of multiple ECG complexes
 - Slowed propagation in the myocardium → delayed ventricular activation → substrate for reentrant arrhythmias
 - Used to risk stratify patients with ventricular arrhythmias and risk of sudden death
- Markers of high risk for sudden death (in pts with CAD) include (Gomes, circulation 2001. "MUSTT" study)
 - Filtered QRS duration (fQRS) >114ms (sens 91%, spec 90%)
 - Low Amplitude Signal duration (LAS40) ≥38ms
 - High frequency root mean square (RMS40) <20μV

Analysis Filter : 40-250Hz		Number Of Beats Averaged: 555
Std. QRS Duration (unfiltered) : 111 ms		Number Of Beats Detected: 642
Total QRS Duration (filtered) : 139 ms		Noise Level (Std. Devn.) : 0.46 μV
Duration Of HFLA signals < 40μV : 73 ms		
RMS Voltage in terminal 40 ms : 4 μV		
Mean Voltage in terminal 40 ms : 3 μV		

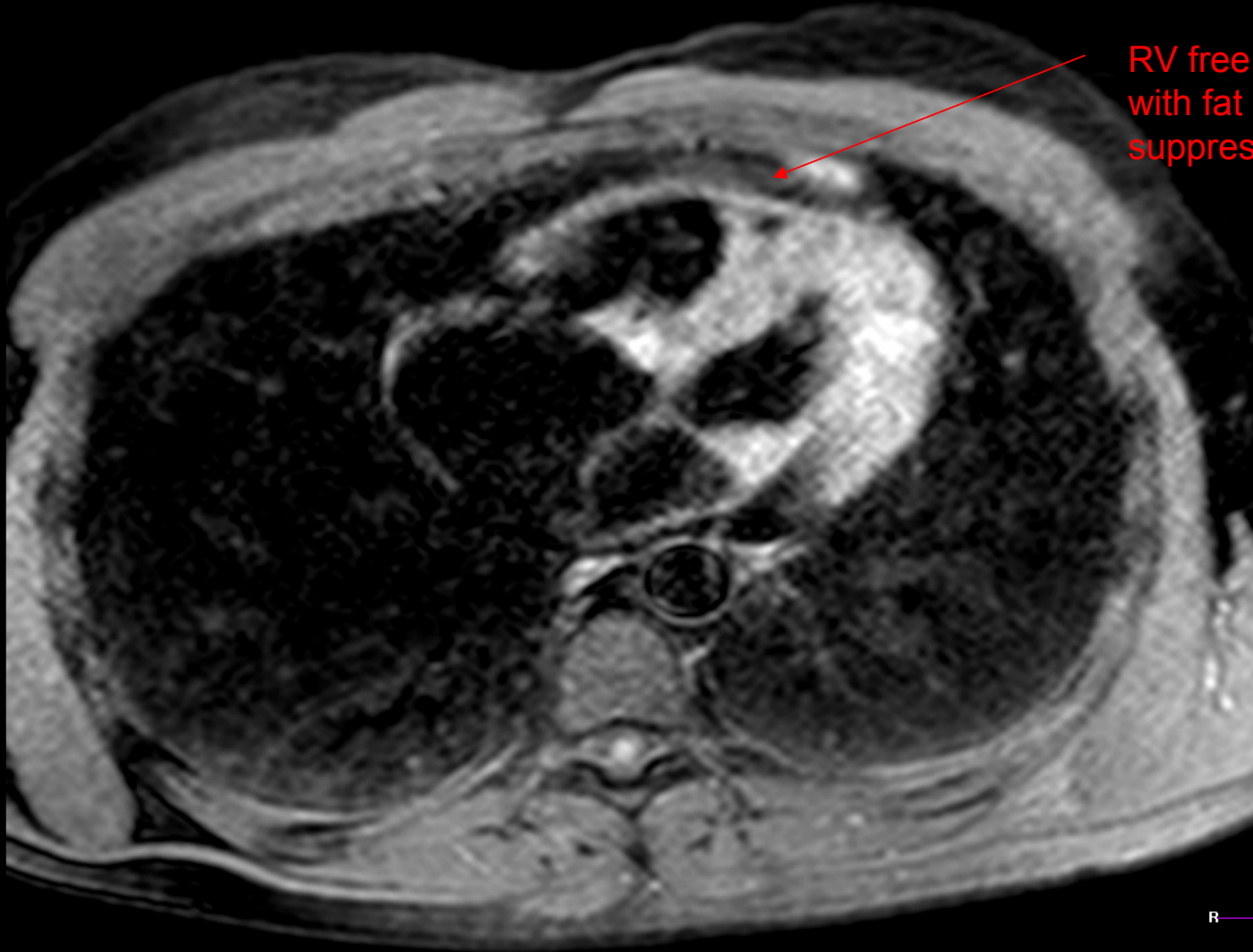
Thoracic axial TSE without fat saturation



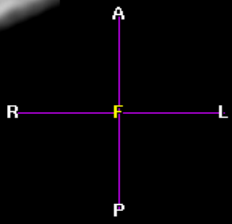
Epicardial fat
around RV



Thoracic axial TSE with fat saturation

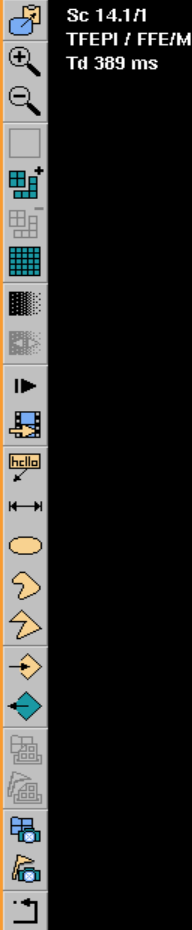


RV free wall
with fat
suppression



14.1: IR_TFE_LL_BH CLEAR: Image: 12

Sc 14.1/1
TFEPI / FFE/M
Td 369 ms

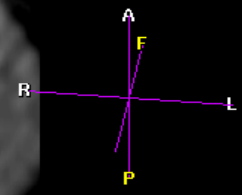


Four-chamber delayed enhancement



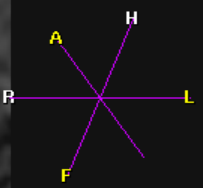
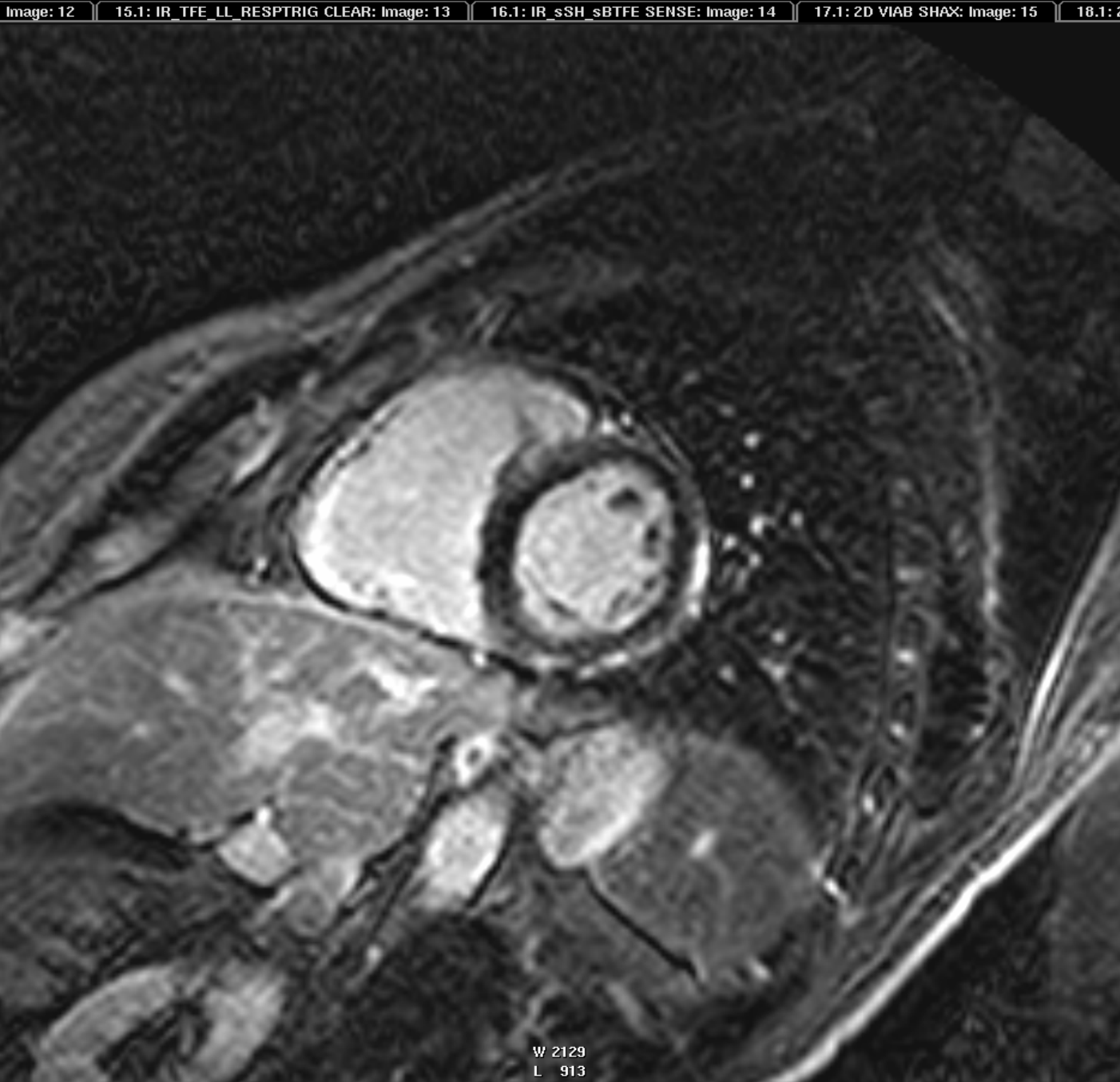
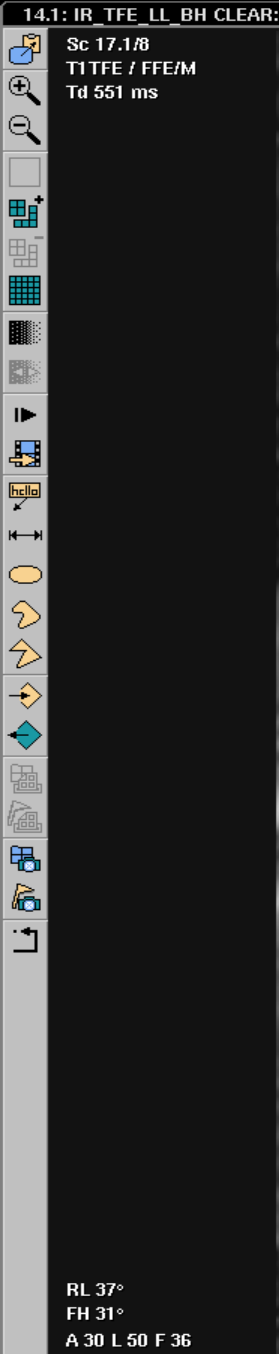
AP 6°
RL 33°
FH -4°
A 43 L 57 F 51

W 1220
L 904



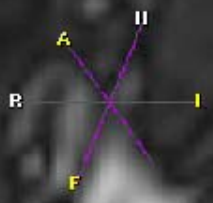
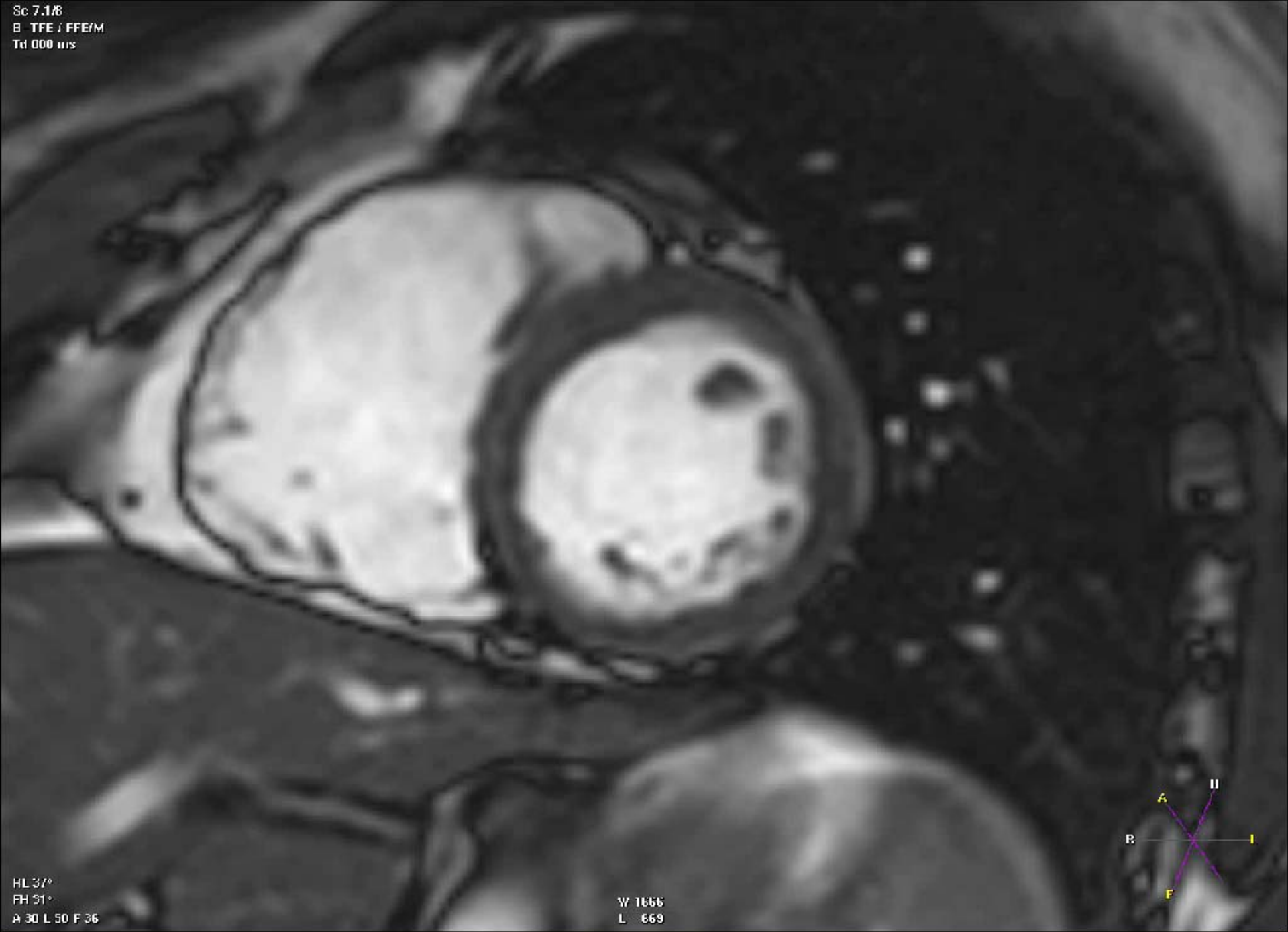
Sc 17.1/8
T1 TFE / FFE/M
Td 551 ms

RL 37°
FH 31°
A 30 L 50 F 36



W 2129
L 913

Sc 7.18
B TFE / FFE/M
Td 000 ms

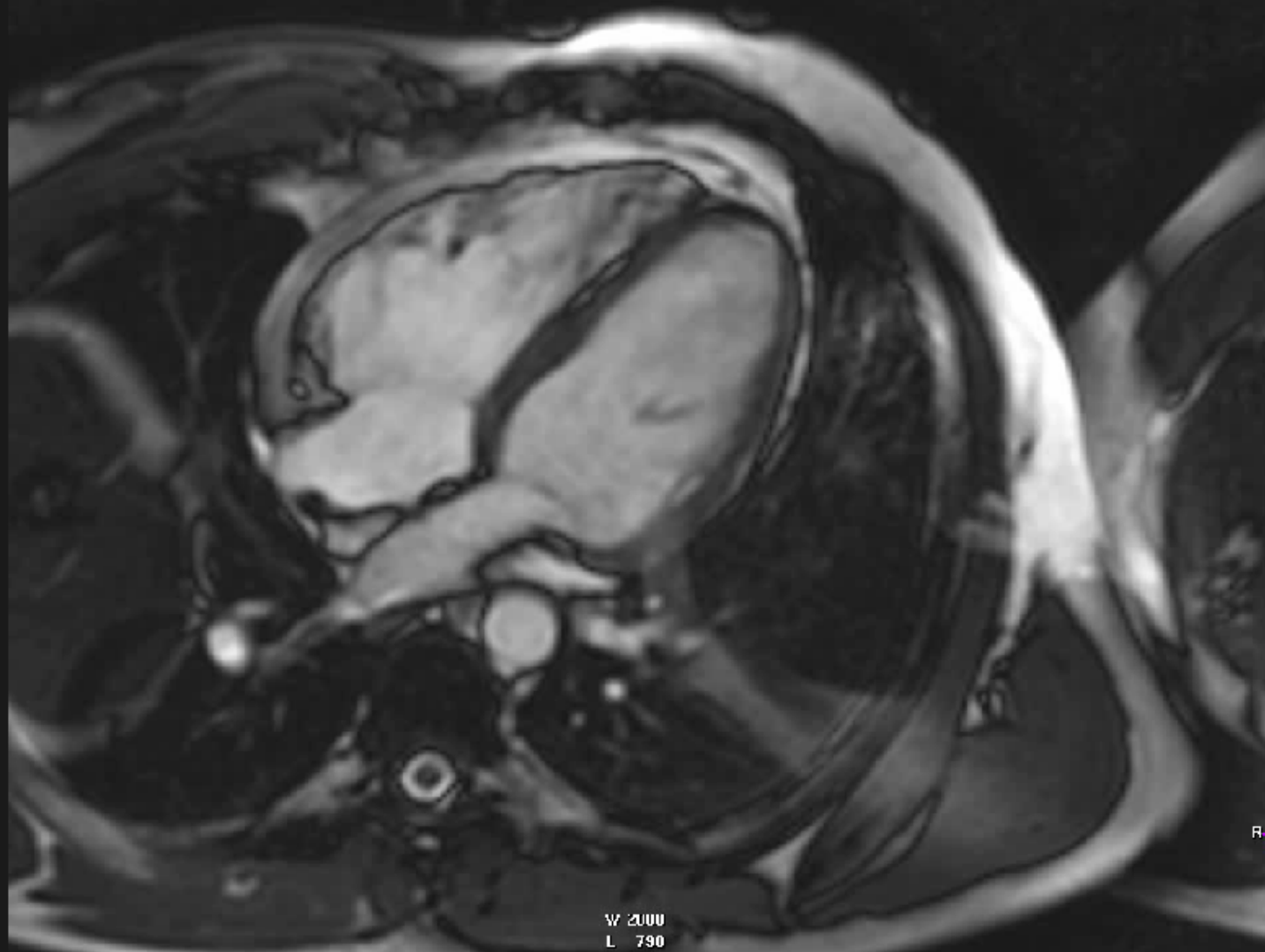


HL 37°
FH 51°
A 30 L 50 F 36

W 1666
L 669

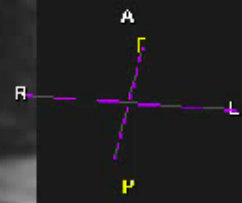
2157573_ARVC_SAX

Sc 8.1/1
B TFE / FFE/M
Tcl 000 ms

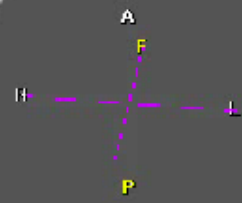
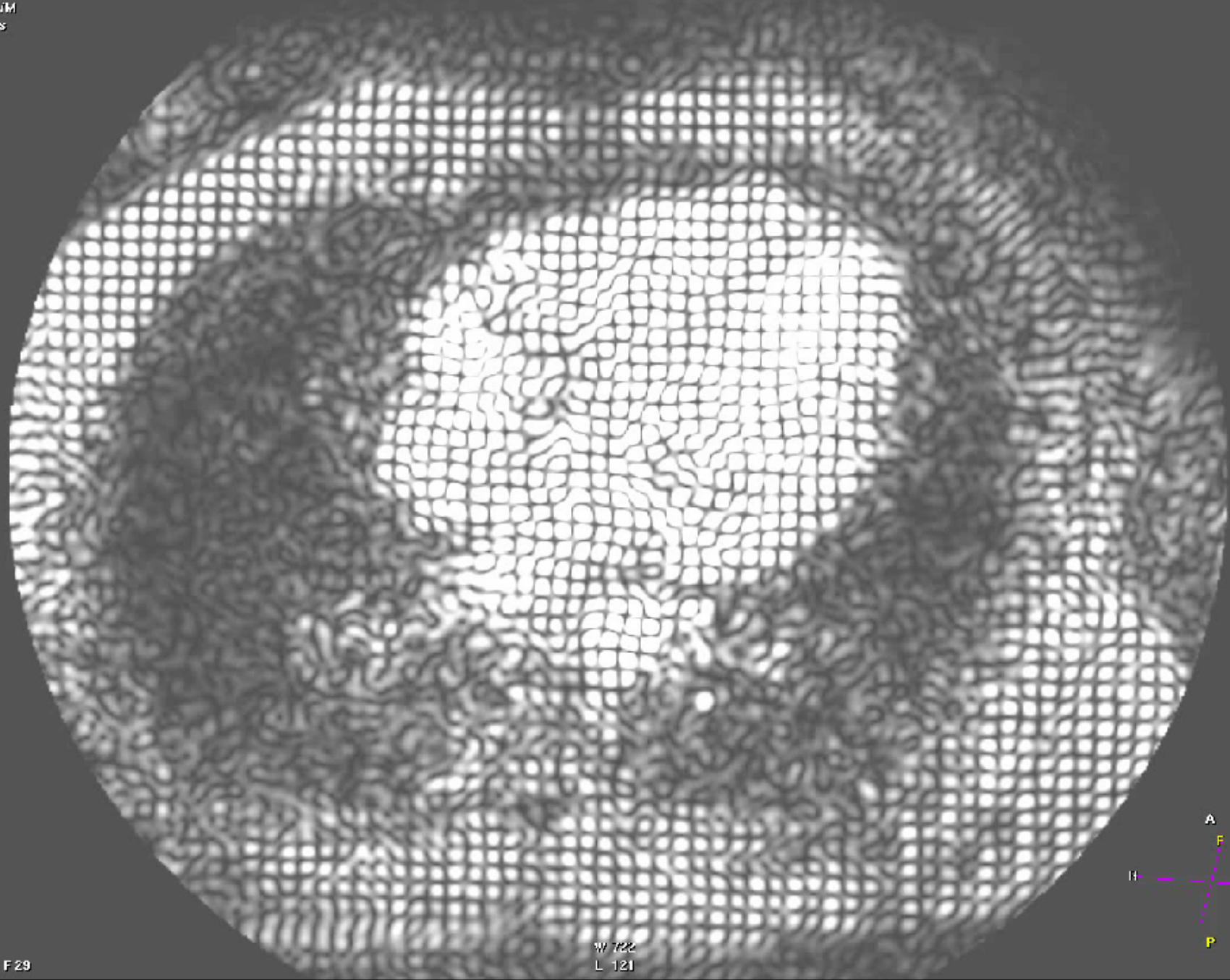


AP 0°
HL 33°
FH -4°
A 13 L 57 F 51

W 2000
L 790



Rn 23.171
FFE / FFEJM
Td 017 ms



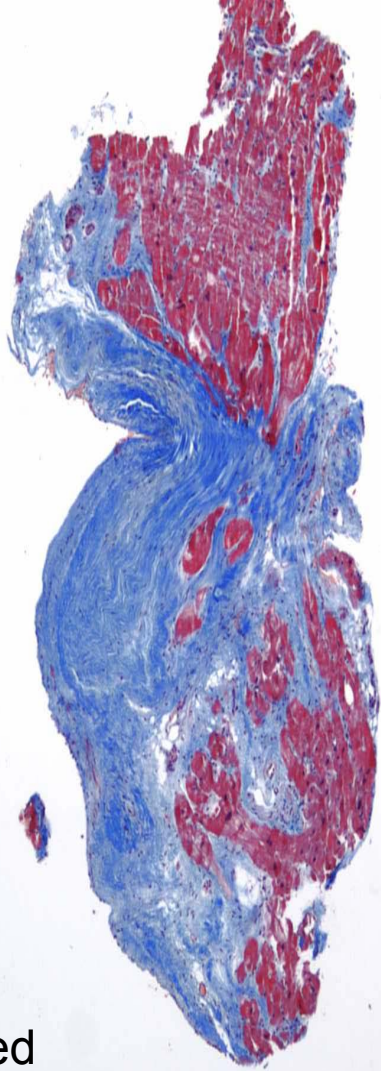
AP 0°
RL 33°
FH -4°
A 16 L 26 F 29

W 122
L 121

Cardiac MRI-Final

- Mildly increased right ventricular cavity size and mild global RV hypokinesis. The RVEF was mildly reduced at 43%. Possible MR evidence of right ventricular fatty infiltration/dysplasia. MR evidence of myocardial scarring/fibrosis/infiltration in the subendocardium of the RV free wall and inferior right ventricular septum
- Normal left ventricular cavity size with normal regional left ventricular systolic function. The LVEF was normal at 68%.
- MR evidence of myocardial scarring/fibrosis/infiltration in the mid-myocardium of the septum, and mid inferolateral wall.
- Mild mitral regurgitation.
- Moderate right and mild left atrial enlargement.
- These findings are consistent with probable arrhythmogenic right ventricular cardiomyopathy.

Endomyocardial Biopsy



Muscle=Red
Fibrosis=Blue



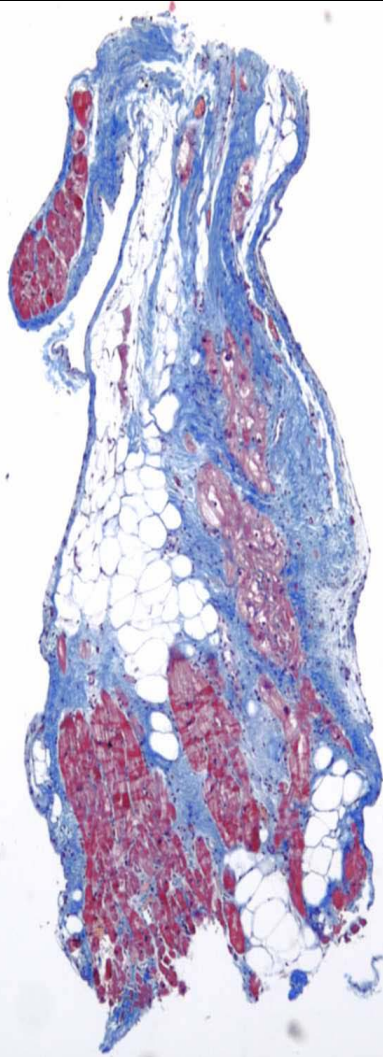
Fat=White
Muscle=Red
Fibrosis=Tan



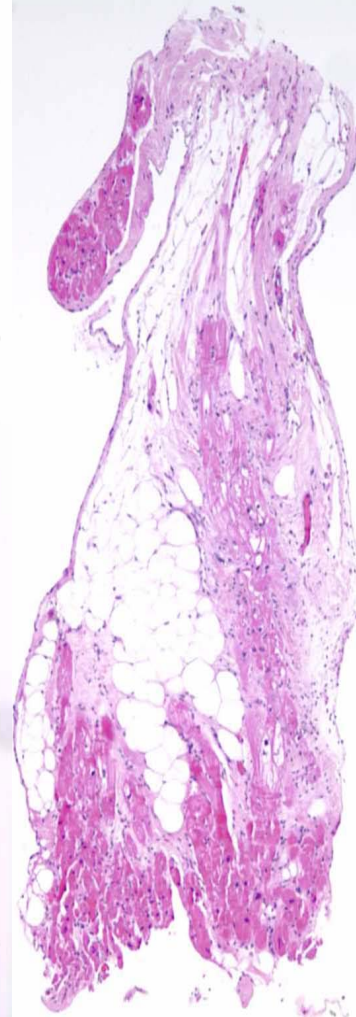
Normal

Trichrome

H&E



Muscle=Red
Fibrosis=Blue



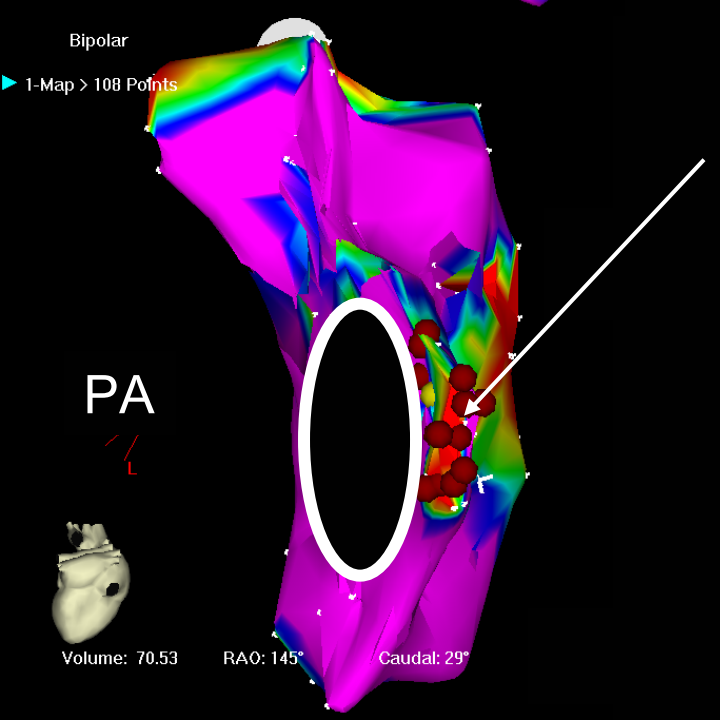
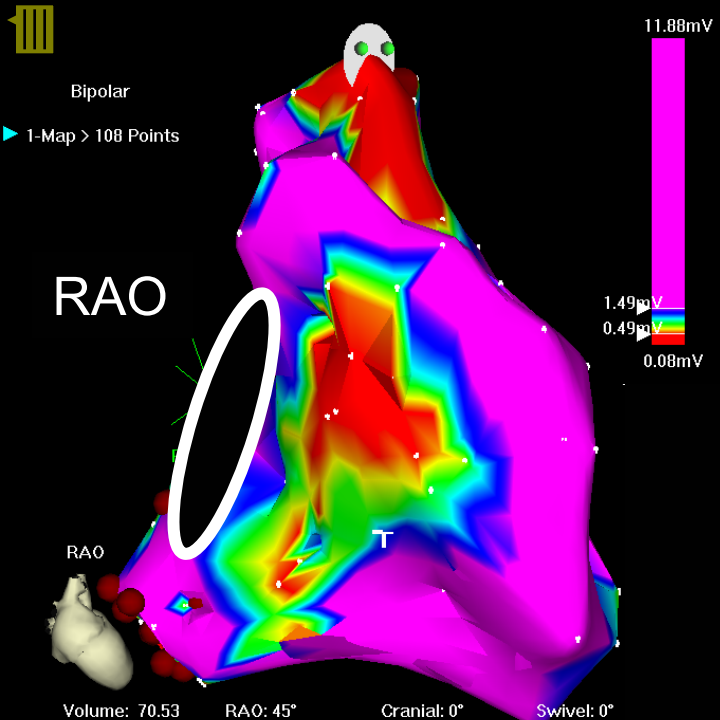
Fat=White
Muscle=Red
Fibrosis=Tan

Pathology-Final

- Three fragments of endomyocardium with focal fatty replacement and fibrosis.
- The findings are compatible with arrhythmogenic right ventricular cardiomyopathy in the appropriate clinical setting.
- Fibrosis is best demonstrated on the Trichrome stain.

EP Study

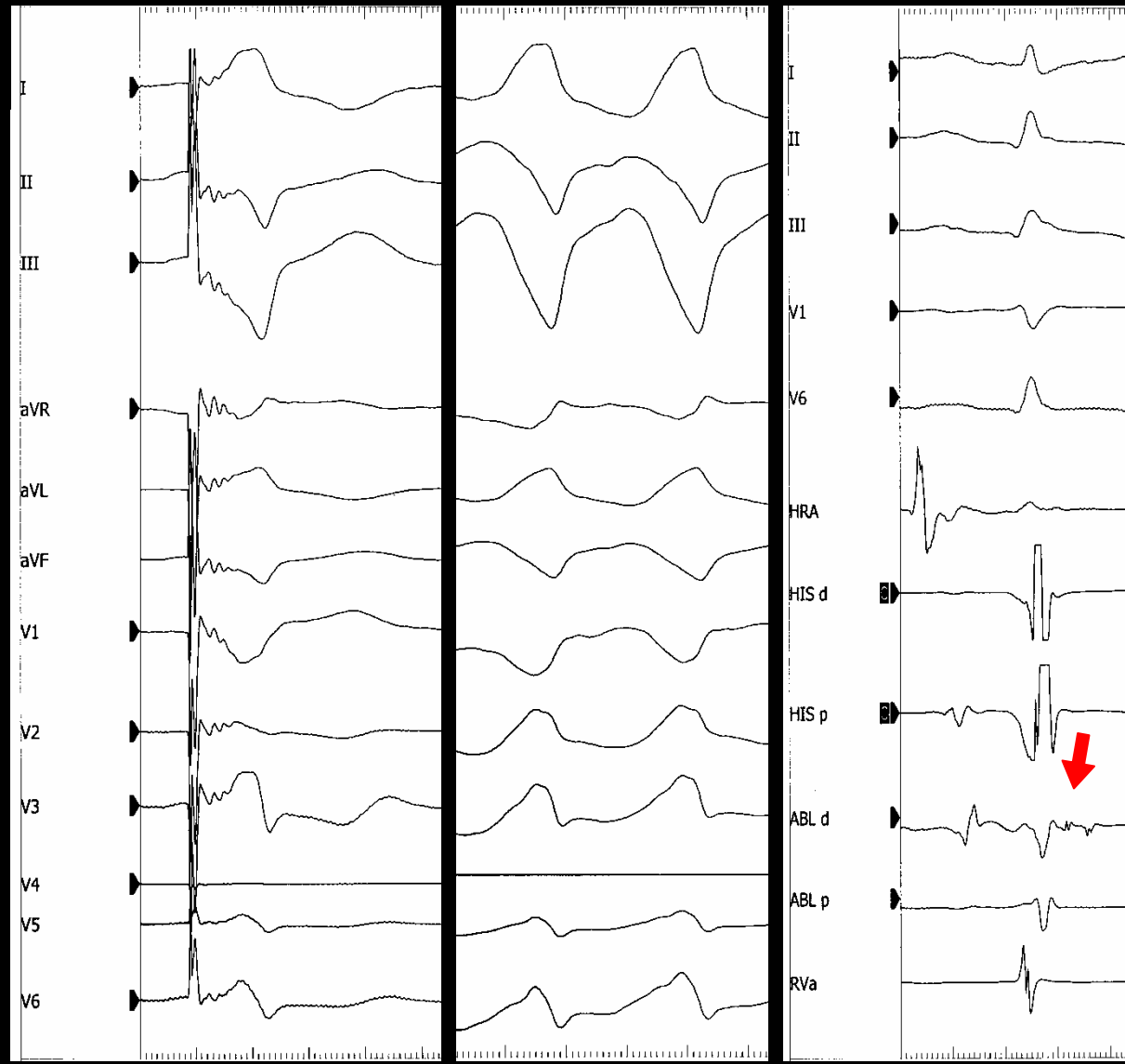
- Right ventricular mapping demonstrated significant abnormalities (areas of low voltage) in three regions
 - The right ventricular apex
 - The right ventricular inflow tract
 - The right ventricular outflow tract
- Inducible monomorphic ventricular tachycardia with double extrastimuli from the right ventricular apex.
 - The VT was a left bundle, left superior axis VT.
- Pace mapping demonstrated a perfect pace map match at the inferior base of the right ventricle right near the tricuspid valve. A series of RF lesions was made in this region.
- Following RF, there was no further inducible VT despite an aggressive stimulation protocol that utilizes up to 3 extra stimuli and burst pacing from two sites in the right ventricle.



Pace Mapping
during SR
PCL 500ms

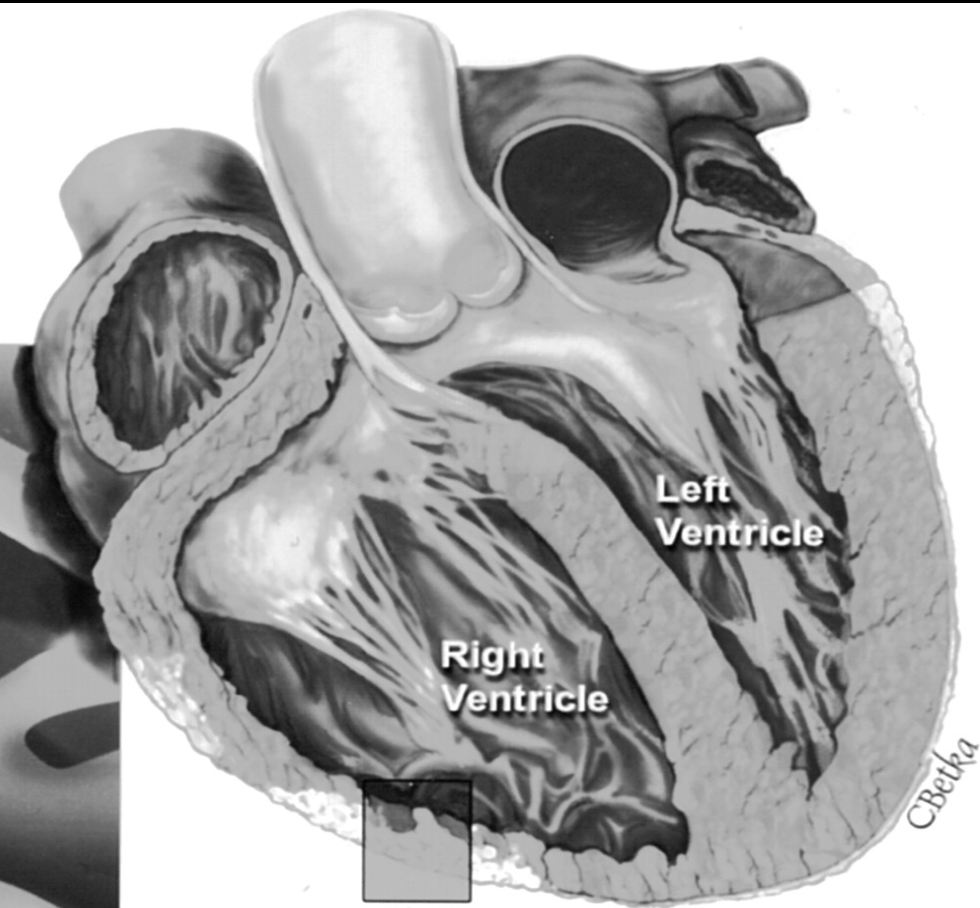
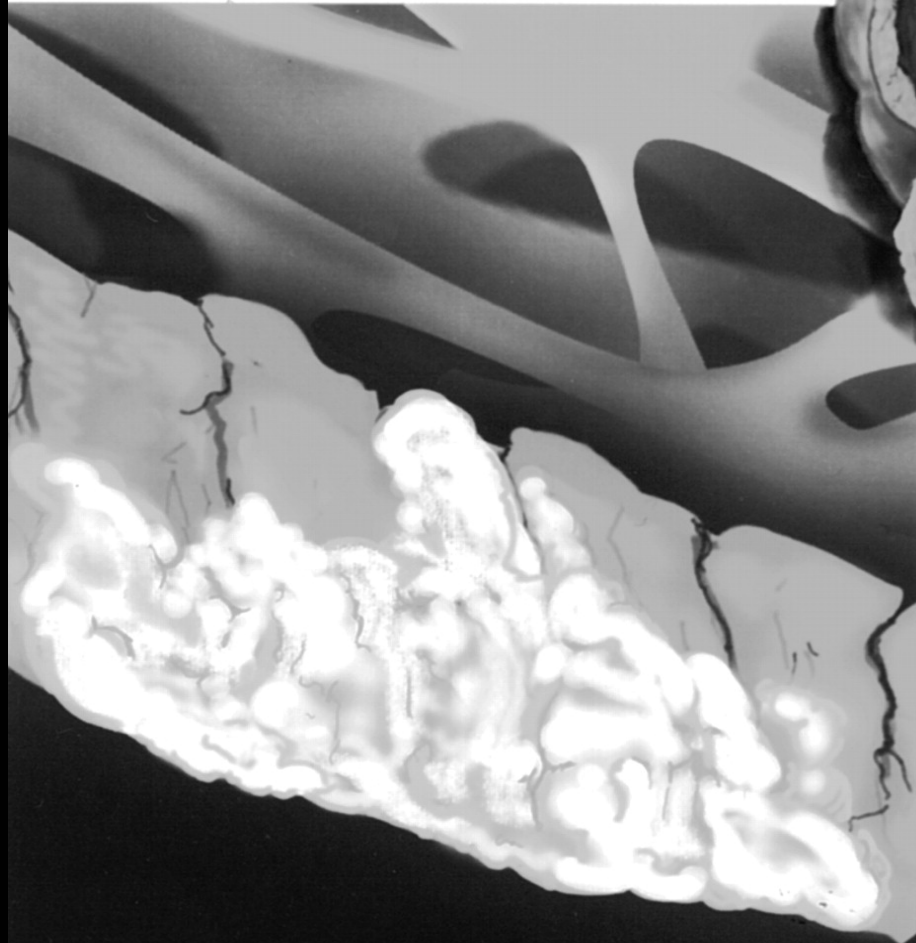
VT
TCL 240ms

Late
Potential



Arrhythmogenic Right Ventricular Dysplasia

- First described by Drs. Frank Marcus and Guy Fontaine in 1982
- Prevalence: 1/5000 with geographic variability (HOVM of Italy)
 - Important cause of SD in young adults in Northern Italy
 - 11% overall and 22% in young athletes with SD in Veneto regions of Northern Italy (vs. 4% United States)
 - In one U.S. series, accounted for 20% SCD in all < 35 yr age & 22% in young athletes (Thiene G, NEJM 1988)
- Fibrofatty infiltration of the RV myocardium, predominantly the free wall, providing a substrate for reentrant arrhythmias via interruption of electrical continuity (spares endocardium)
 - Due to apoptosis & focal myocarditis leading to scarring and fibrofatty infiltration (inflammatory theory)
- 30-50% familial often with dominant phenotype
 - 8 different ARVD genes discovered so far and all are autosomal dominant



**Fatty replacement
of myocardial muscle**

ARVD-Epidemiology

- Mean age at diagnosis: 31 yrs old
 - M > F (Forensic Autopsy in France in 1930s) 2.7:1
 - 3% while playing sports
- Symptoms (Hulot, Circulation 2004)
 - Palpitations 2/3
 - Syncope 1/3
 - Atypical CP ¼
- ECG
 - T-wave inversions on precordium 46%
 - ϵ wave in 1/3
 - Abnormal SAECG 2/3
- Three most commonly affected regions (“Triangle of Dysplasia”)
 - Anterior surface of pulmonary infundibulum
 - RV apex
 - Subtricuspid region
- Two morphologic types
 - Fatty (exclusive to RV)
 - Fibrofatty (may involve LV)

Diagnostic Criteria for ARVD

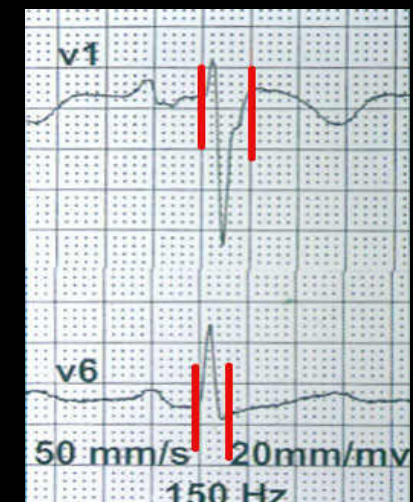
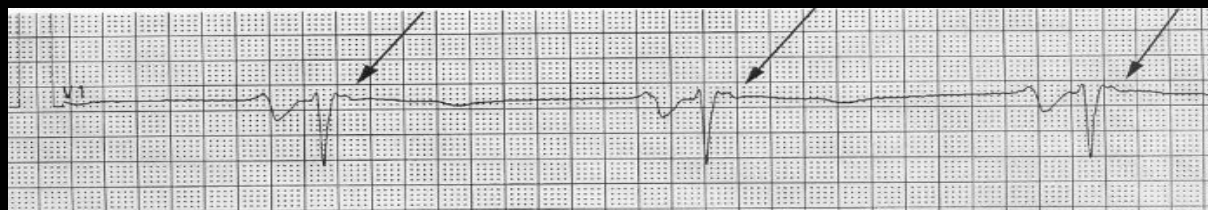
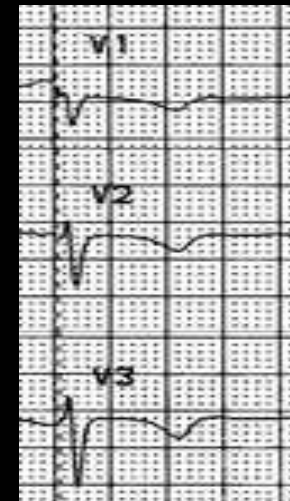
The diagnosis of ARVD is established by the presence of 2 major or 1 major and 2 minor or 4 minor criteria from these categories

	Major Criteria	Minor Criteria
Structural or Functional Abnormalities	<ol style="list-style-type: none"> 1. Severe dilation and reduction of RVEF with mild or no LV involvement 2. Localized RV aneurysm (akinetic or dyskinetic areas with diastolic bulging) 3. Severe segmental dilation of the RV 	<ol style="list-style-type: none"> 1. Mild global RV dilation and/or EF reduction with normal LV 2. Mild segmental dilation of the RV 3. Regional RV hypokinesis
Tissue Characterization	Infiltration of RV by fat with presence of surviving strands of cardiomyocytes	
ECG Depolarization/Conduction Abnormalities	<ol style="list-style-type: none"> 1. Localized QRS complex duration >110 msec in V₁, V₂, or V₃ 2. Epsilon wave in V₁, V₂, or V₃ 	Late potentials on signal-averaged ECG
ECG Repolarization Abnormalities		Inverted T-waves in right precordial leads (in V ₁ through V ₃ above age 12, in the absence of RBBB)
Arrhythmias		<ol style="list-style-type: none"> 1. LBBB VT (sustained or non-sustained) on ECG, Holter, or ETT 2. Frequent PVCs (>1,000/24 hours on Holter)
Family History	Family history of ARVD confirmed by biopsy or autopsy	<ol style="list-style-type: none"> 1. Family history of premature sudden death (< age 35) due to suspected ARVD 2. Family history of clinical diagnosis based on present criteria

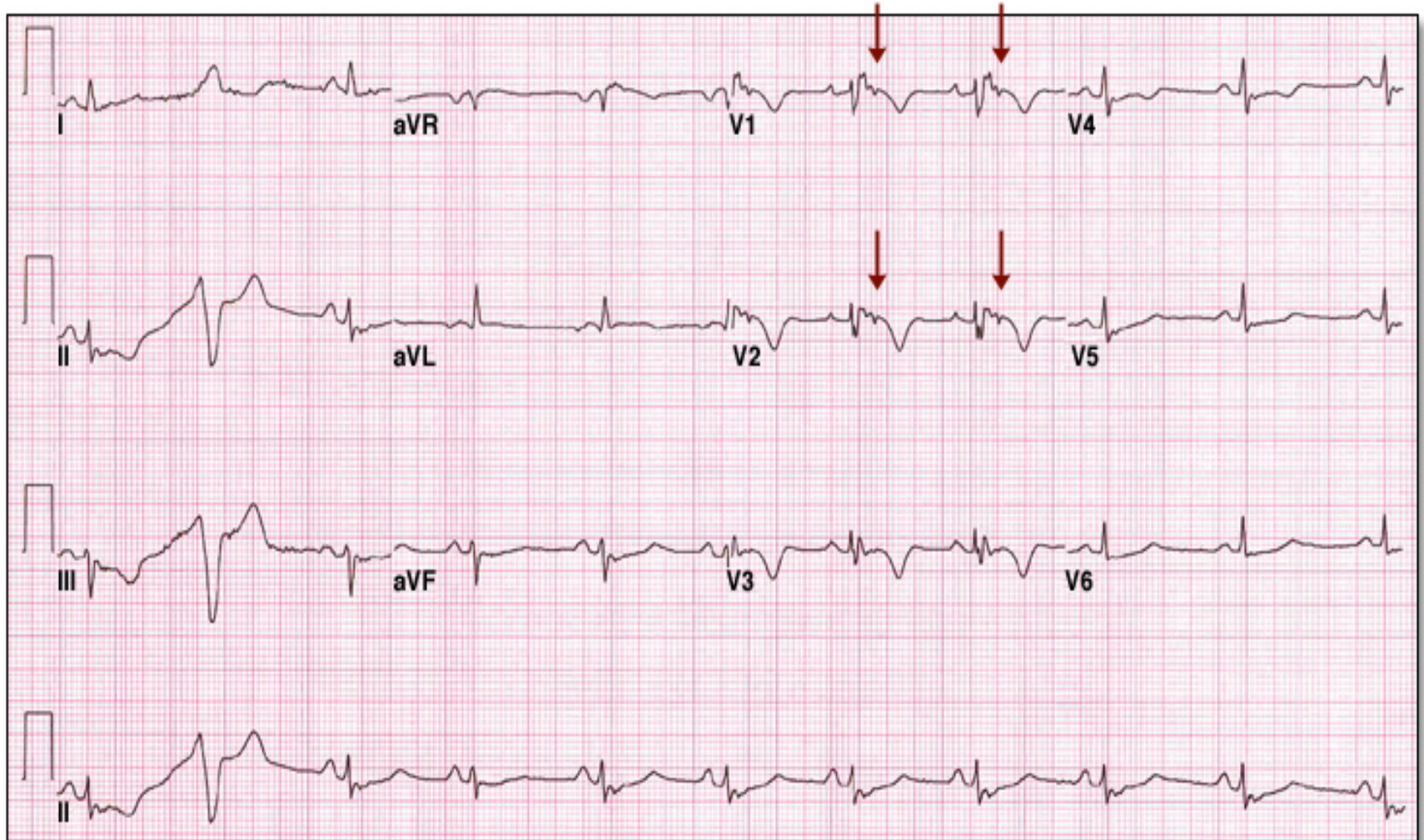
Append

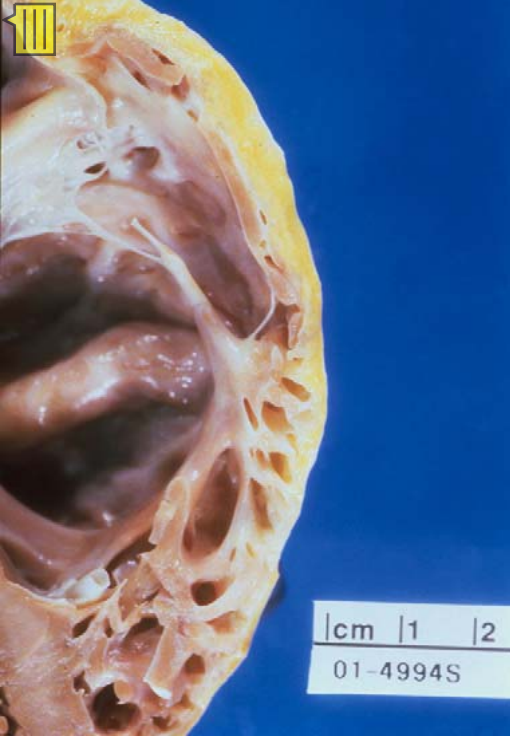
Diagnosis using ECG

- Double speed (50mm/sec) & amplitude (20mm/mV)
- 40-50% have abnormal ECG at presentation vs. 100% in 6yrs
 - Wide QRS (>100ms) on V1 (55% sens)
 - QRS>100ms in V1-3 & negative T in V2 (84% sensitivy) (Fontaine et al, Ann Cardiol Angeiol 1994)
 - 30% Epsilon Wave at the end of QRS on V1, V2
 - 50% T-inversion on right precordial leads
 - RV parietal block (Peters et al, Ann Noninvasive Cardiol 1996)
 - QRS duration V1-V3 longer than QRS V4-V6 by 1.2 (96% specificity, 98% PPV)
 - VT originates from RV so has LBBB pattern



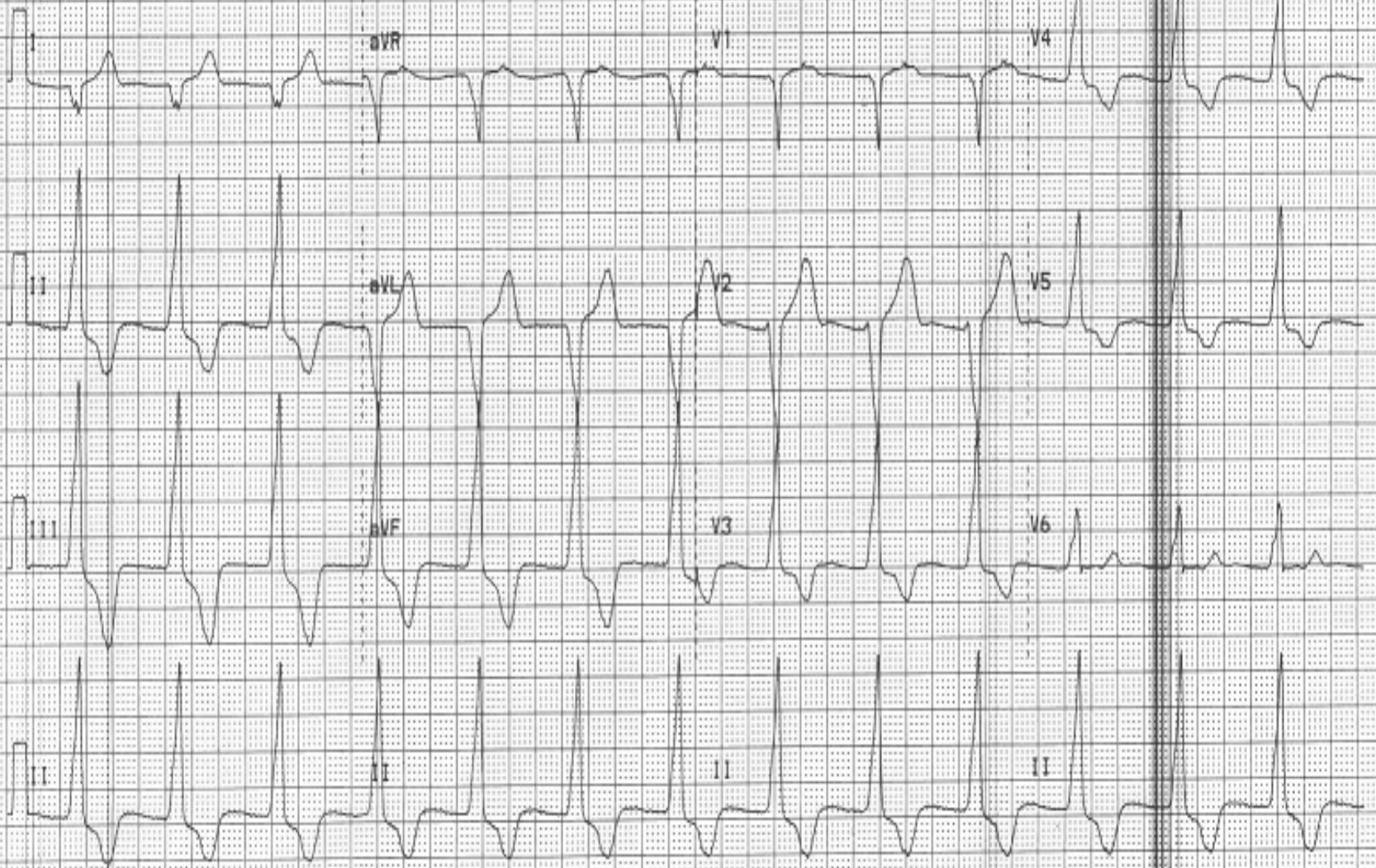
ECG of 48 yr old male s/p AICD and on Sotalol due to frequent ICD shocks from VT





RVOT VT vs. ARVD VT (O'Donnell, Eur Heart J, 2002)

- N=50 patients with LBBB VT with normal LV function, no CAD and scheduled to undergo EPS
- ECG changes
 - T wave inversion on right precordium V1-V3 in 36% ARVD vs. 6% RVOT
 - SAECG late potentials 78% ARVD vs. none in RVOT
 - 53% ARVD and 100% RVOT had inferior axis VT
- Major abnormalities on MRI & TTE
 - 6% RVOT vs. 88% ARVD (MRI)
 - 0% RVOT vs. 42% ARVD (TTE)
- EP study
 - Inducible VT 82% ARVD vs. 3% RVOT
 - Sustained VT 71% ARVD vs. 15% RVOT
 - Multiple morphology of VT 70% ARVD vs. 100% RVOT had single VT morphology
 - 42% RFA success rate in RVOT vs. 97% RVOT
 - 48% recurrence vs. 94% RVOT over 58 months follow-up
- Outcome
 - RVOT: excellent outcome vs. 2.5% yearly mortality in ARVD



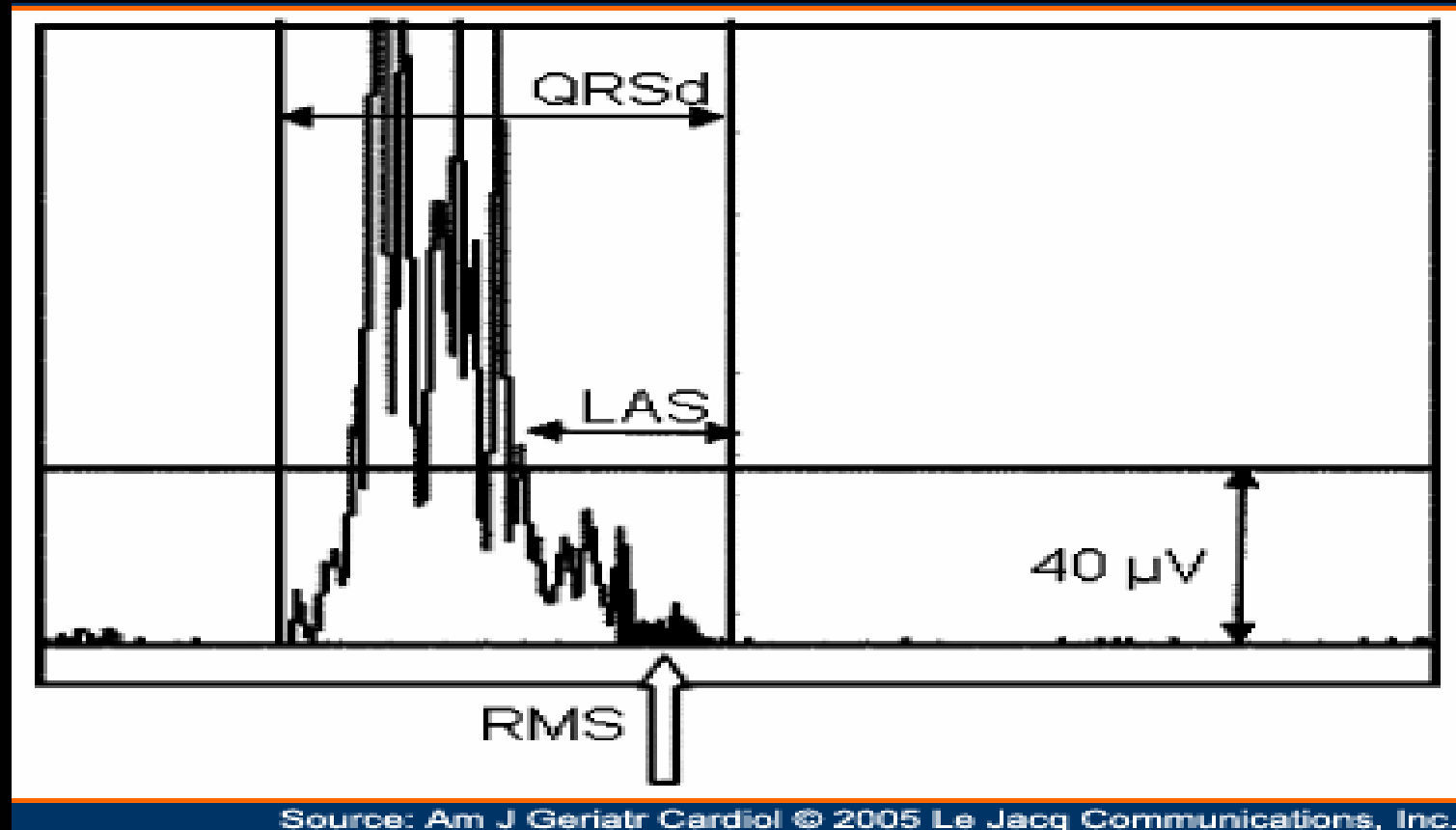
Limb: 10 mm/mV
Chest: 10 mm/mV

BETH ISRAEL DEACONESS MEDICAL

25 mm/s
*STABLE 40 Hz

SAECG

- Markers of high risk for sudden death (in pts with CAD & low EF) include (Gomes, circulation 2001. "MUSTT" study)
 - Filtered QRS duration (fQRS) $>114\text{ms}$ (sens 91%, spec 90%)
 - Low Amplitude Signal duration (LAS40) $\geq 38\text{ms}$
 - High frequency root mean square (RMS40) $<20\mu\text{V}$



SAECG in ARVD

- Data based primarily on pts with CAD s/p MI and LV dysfunction
 - MUSTT trial (n=1925): Chronic CAD, asx NSVT, low EF
 - fQRS duration >114 ms predicted a significantly greater risk of arrhythmic death or cardiac arrest, cardiac death, and total mortality
- Nasir et al from Johns Hopkins ARVD center reviewed 13 studies involving n=458 patients with ARVD subjected to SAECG (A.N.E., 2003)
 - Abnormal SAECG 47-100%
 - Correlation with extent of disease and presence of sustained VT
 - 57% mild ARVD vs. 87% severe ARVD
 - 48% vs. 76% sustained VT in familial VT (sens 66-88%; spec 80-100%)
 - fQRS \geq 110ms good predictor of inducible VT in ARVD (91% sens, 90% spec) (Nasir, Pacing Clin Electrophysiol, 2003)
- Keiko et al (Jpn Heart J, 2001) compared 7 patients with ARVD to controls and demonstrated positive SAECG 5/7 patients vs. 8% controls
- Abnormal particularly in LBBB VT with superior axis morphology
- Abnormal SAECG (Positive for late potential if 2/3 criteria met) (Oselladore, 1996)
 - 71% ARVD pts with VF
 - 71% ARVD pts with sustained VT
 - 40% ARVD pts with NSVT

ARVD Diagnostic tools (Cont)

- Echocardiogram
 - Ratio > 0.5 RV/LV end diastolic diameter (86% sensitivity and 93% specificity)-selection bias
 - TTE on asymptomatic pts with ARVD: 44% pts with abnormal TTE including enlarged RV or RV WMAs (Scognamiglio, Eur Heart J, 1989)
 - No studies done that compared to Gold Standard
- Right Ventricular Angiography
 - Historically the standard reference for ARVD diagnosis
 - 30° RAO and 60° LAO views
 - Look for trabeculations, aneurysms, and WMA
 - Bulging in the triangle regions
 - Problem: RV segments do not often contract equally in normal patients
- Endomyocardial Biopsy (Angellini, International J Cardiol 1993)
 - Highly specific (92%) with low sensitivity (67%) as biopsy often performed on septum and not on RV free wall due to high risk of perforation
 - Greater than 3% fat and less than 40% fibrous tissue replacing the myocardium often used as criterion for ARVD (Strain et al, Eur Heart J 1989)

Role of MRI (yellow is major criterion)

- MRI features consistent with ARVD (Tandri, J Cardiovasc Electrophysiol, 2003)
 - Intramyocardial hyperintense T-1 signal similar to fat on spin-echo images (sensitivity 22-67% with ARVD)
 - Alone has poor sensitivity and specificity
 - RV wall thinning or regional wall motion abnormalities (25-67%)
 - Secondary to fibrofatty infiltration
 - RV and RVOT dilatation/aneurysms
 - Diskinetic RVOT highly suggestive of ARVD in the absence of pulmonary hypertension
 - Heavy trabecular disarray (58%) & moderator band hypertrophy
 - Also seen in RVH and thus not specific
 - RV dysfunction in 75% of ARVD
 - Average RVEF 42% vs. RVEF of normal controls 57% ($p < 0.01$)
- ARVD should NOT be diagnosed based solely on presence of intramyocardial fat on MRI
 - RV wall is thin and epicardial fat often present in normal subjects (especially in apex and AV groove; near the triangle)
- Wide range of techniques and equipments without standard protocol
 - Study at Johns Hopkins ARVD center on 89 patients diagnosed with ARVD on MRI scans revealed that only 27% met the Task Force criteria for ARVD/C.

Role of EBCT in ARVD

- Attenuation for epicardial adipose tissue is -65 ± 10 HU vs. 5 to -17 HU for intramyocardial fat
- Features of ARVD on EBCT (Tada et al, Circulation 1996)
 - Abundant epicardial fat 86%
 - Low attenuation trabeculations 71%
 - RV scalloping/enlargement 79%
 - Intramyocardial fat deposits 50%
 - Enlargement of RVOT 61% (Bomma et al, Pacing Clin Electrophysio 2003)
- Advantage
 - Can be used in ARVD pts with ICD
 - May evaluate mediastinum to r/o sarcoid that rarely mimics ARVD
- Disadvantage
 - Dye load
 - High radiation (not ideal for screening in pts with family history)

ARVD (Cont)

- Prognosis
 - 18.5% overall mortality in cohort of 15 patients followed over 10 yrs (2.3%/yr) (Hulot, Circulation 2004)
 - In Asx family cohorts, 10% progress to symptomatic disease in mean follow-up of 8.9 years with upto 50% having Sx VT (J Am Coll Cardiol 2000).
 - Only 1/49 pts with VT on antiarrhythmics (Sotalol or Amiodarone) died (8.9 yr follow-up)
 - Better prognosis than VT/VF from common structural disease as LV function often normal
- Predictors of Mortality (Indications for ICD)
 - Documented LBBB VT
 - Clinical signs of RV and/or LV failure
 - Polymorphic VT
 - EPS alone for risk stratification is poor: 49% PPV & 54% NPV

ARVD Management

- Treatment:
 - No competitive sports
 - Task Force on Sudden Cardiac Death (Eur Heart J 2001)
 - Primary Prevention
 - AICD (Class IIa)
 - Sotalol highly effective in NSVT vs. other agents (Beta-Blockers, Amiodarone)
 - Ablation: ARVD often diffuse and progressive with high recurrence rates of VT from other regions
 - » RFA however improves the efficacy of pharmacologic therapy by 70% (Movsowitz et al, Am Heart J, 1996)
 - Secondary Prevention
 - AICD (Class I) but may rarely be complicated by “electrical storm” & frequent AICD fire
 - All first degree relative should be screened

Variants of ARVD

- Naxos disease (ARVD+palmoplantar keratoderma)
 - AR, 100% penetrance by adolescence
 - Ectodermal signs present within the first years of life
 - 17q21 Plakoglobin (component of desmosomes), Type II Keratin
 - Hyperkeratosis of palms/soles with woolly hair
 - “diagnosed by shaking their hands” – Melvin Scheinman, MD
 - Annual Sx arrhythmia 3% & mortality 2.3%
- Carvajal Syndrome
 - Variant of Naxos with earlier onset and LV involvement
 - Worse prognosis
 - Found in India and Ecuador

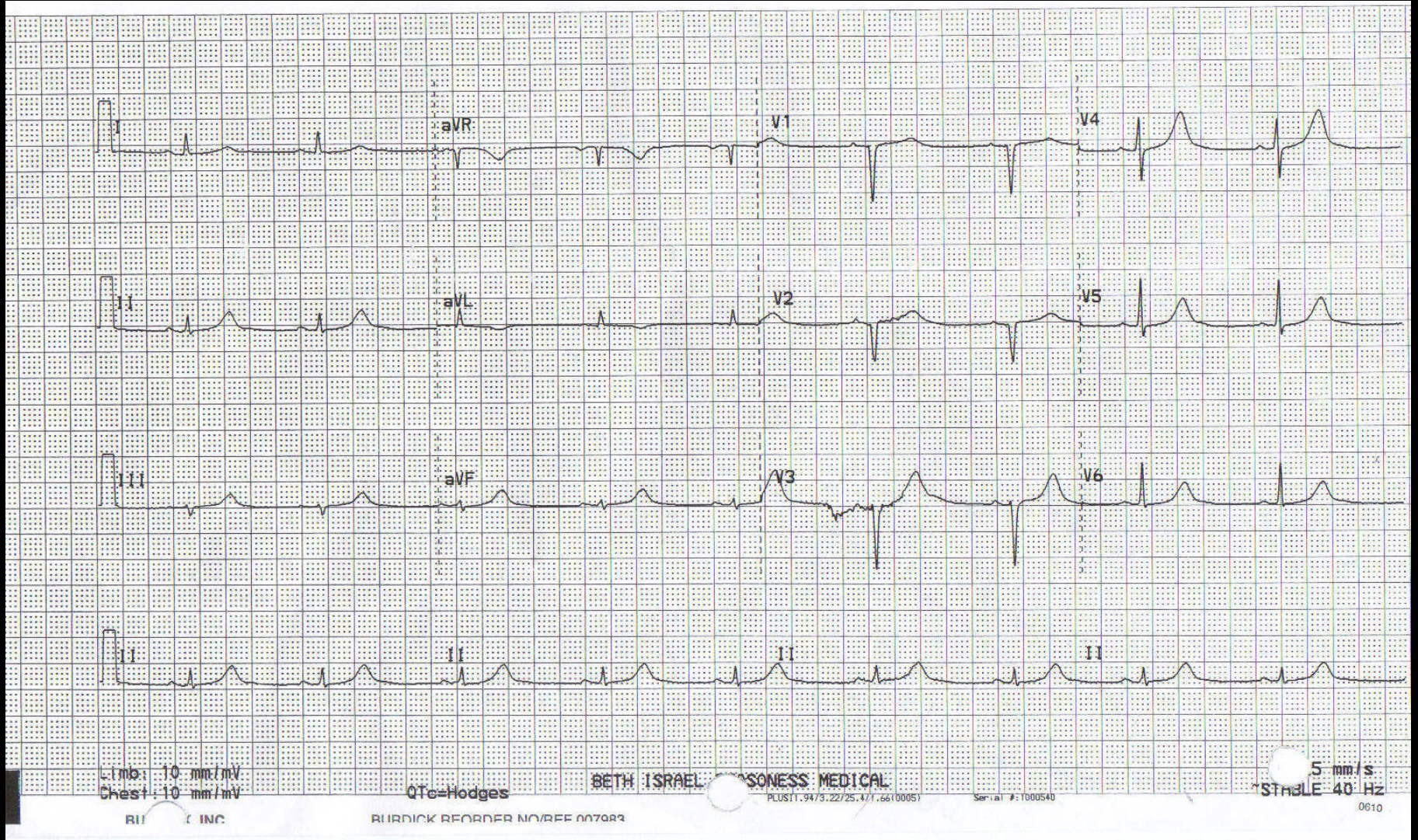


ARVD variants

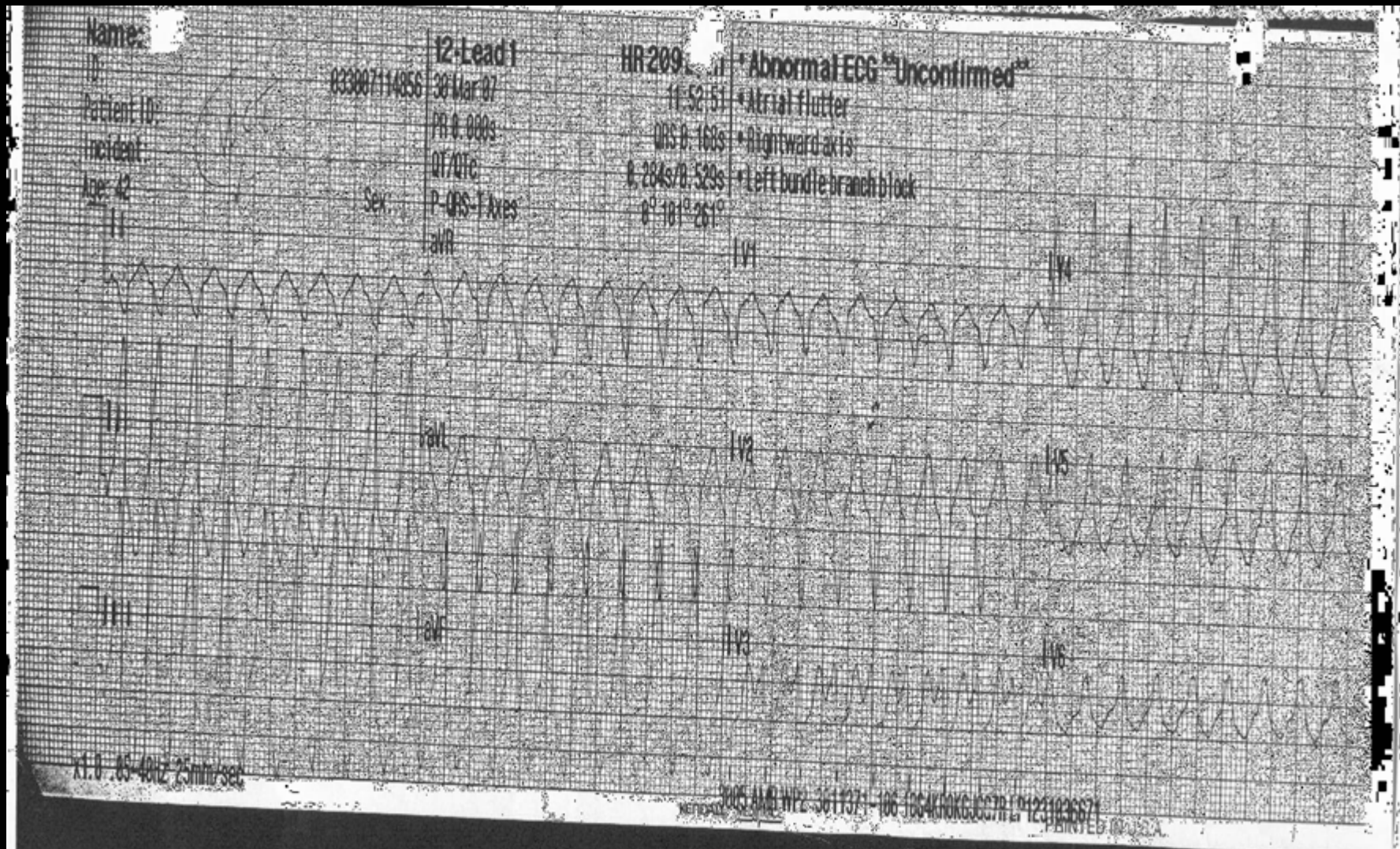
- RYR2 mutations (AD)
 - Cardiac ryanodine receptor gene on chromosome 1
 - Found in several Italian families with ARVD and exertional polymorphic VT
 - Polymorphic or bidirectional VT
 - Only involve the apex
 - High penetrance
 - Equally affect males and females
- Uhl's anomaly
 - Congenital absence of RV myocardium
 - No gender predilection and non-familial
 - Age at presentation: infancy & childhood
 - Exercise induced death, sudden death, arrhythmia rare
 - Common mode of presentation is congestive heart failure

Outflow tract VTs

Baseline ECG

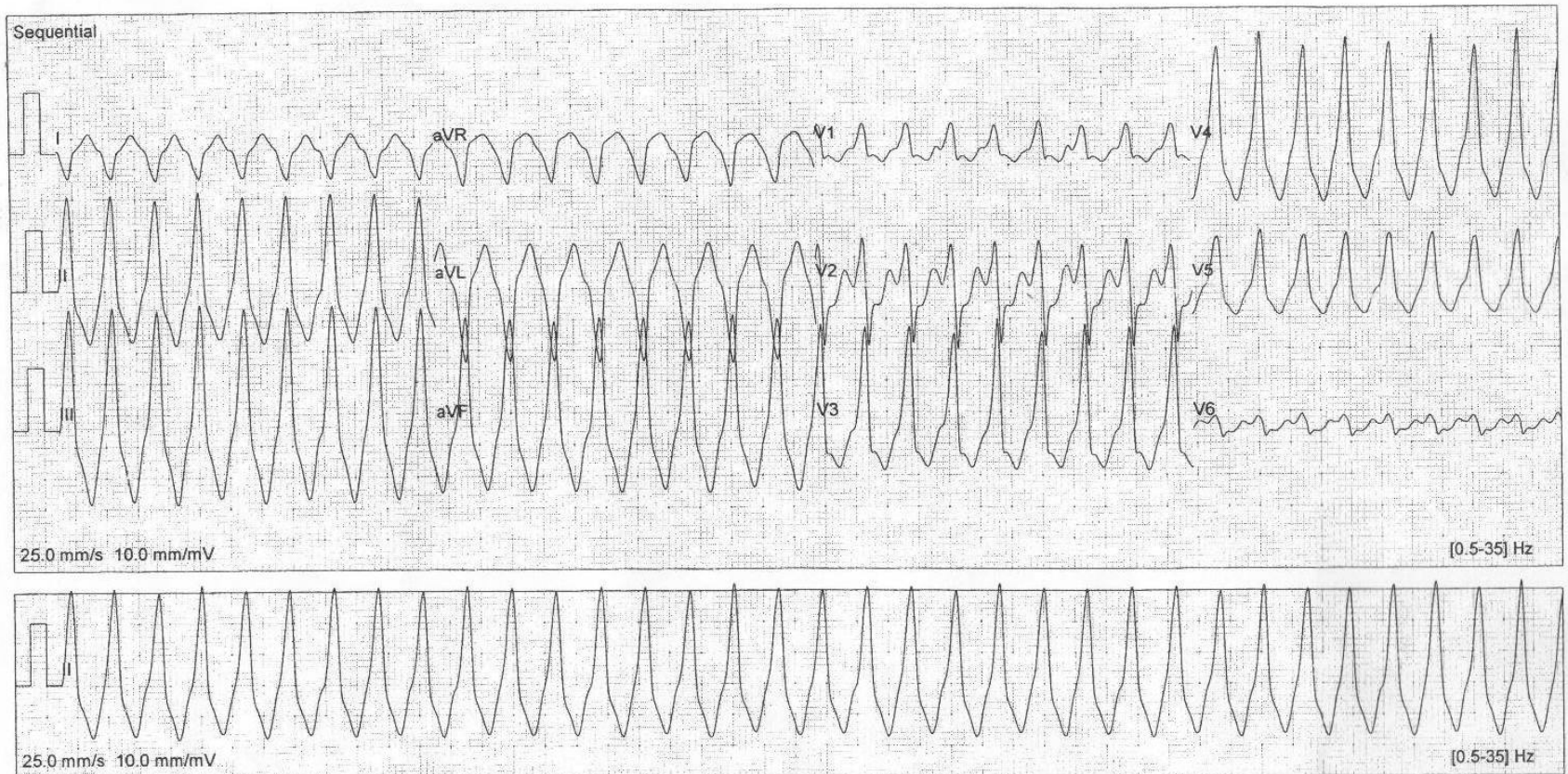


Clinical VT 1



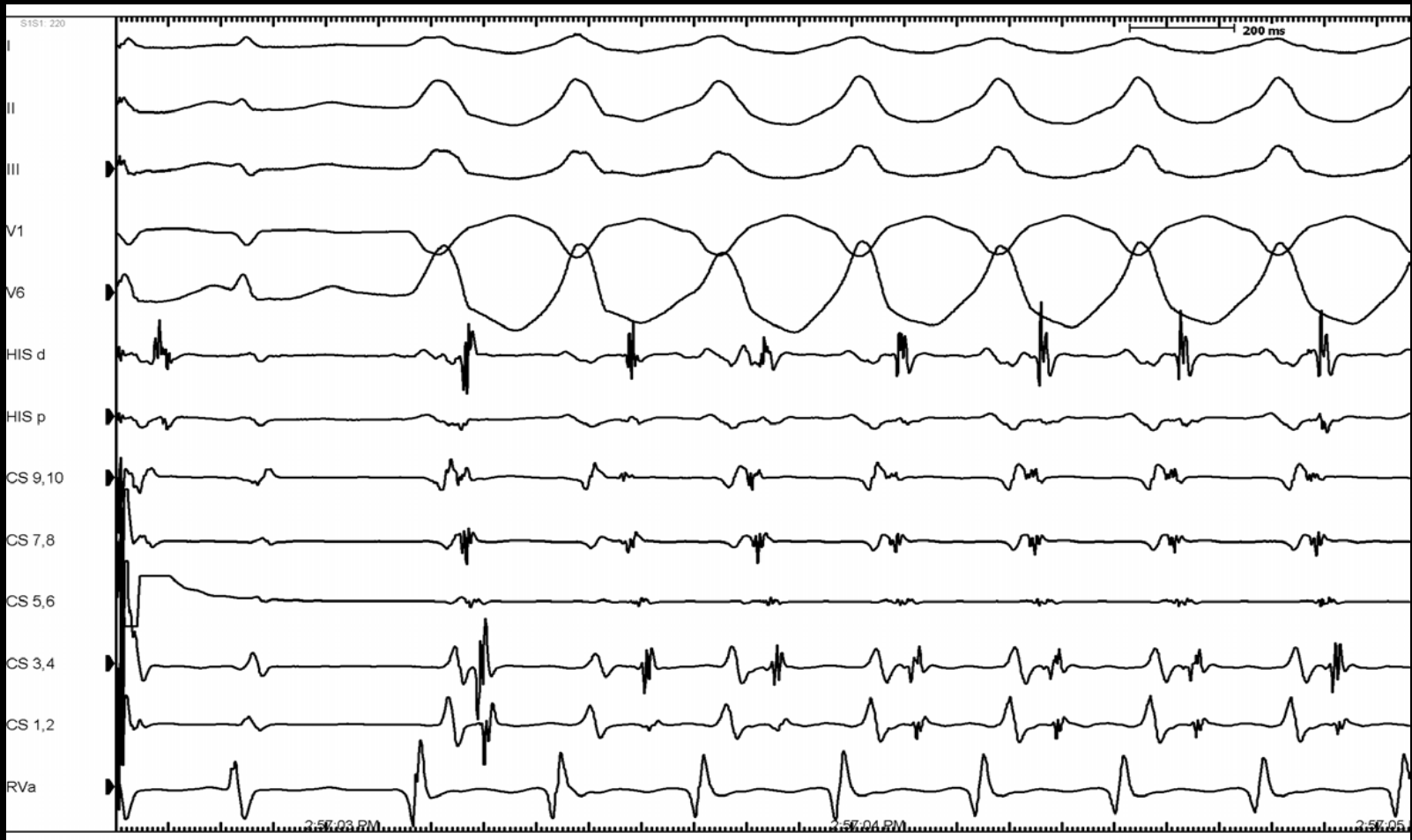
RVOT antero-septal

Clinical VT

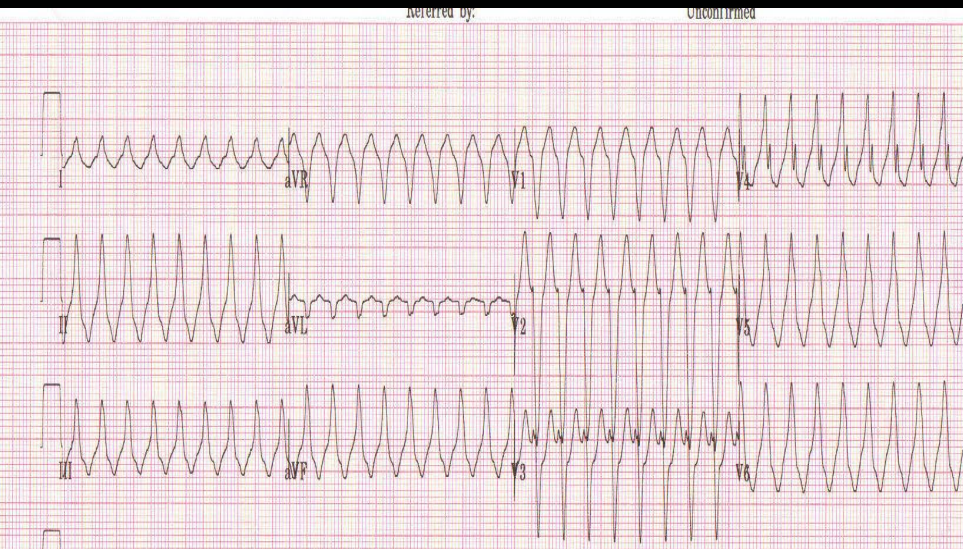


LV Basal VT- SUPERIOR Lateral Mitral Annulus

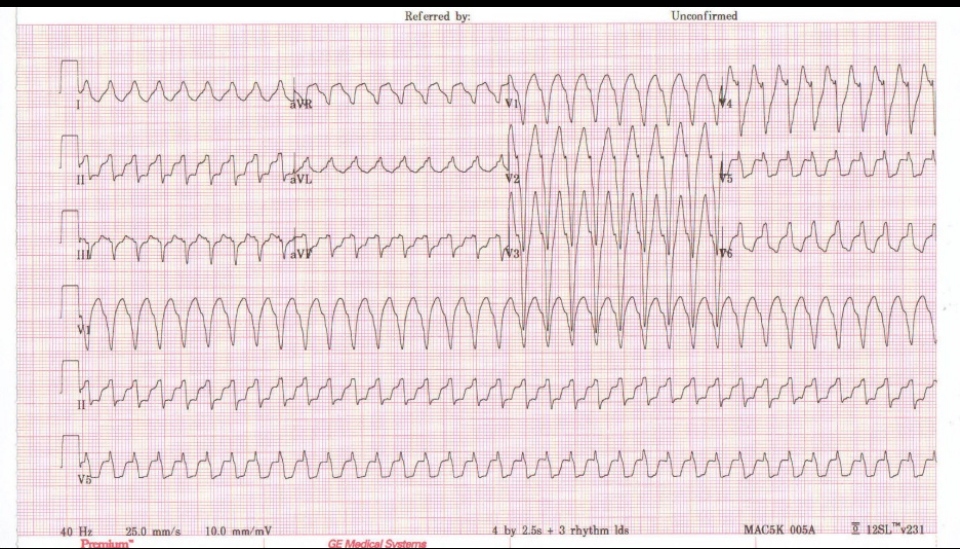
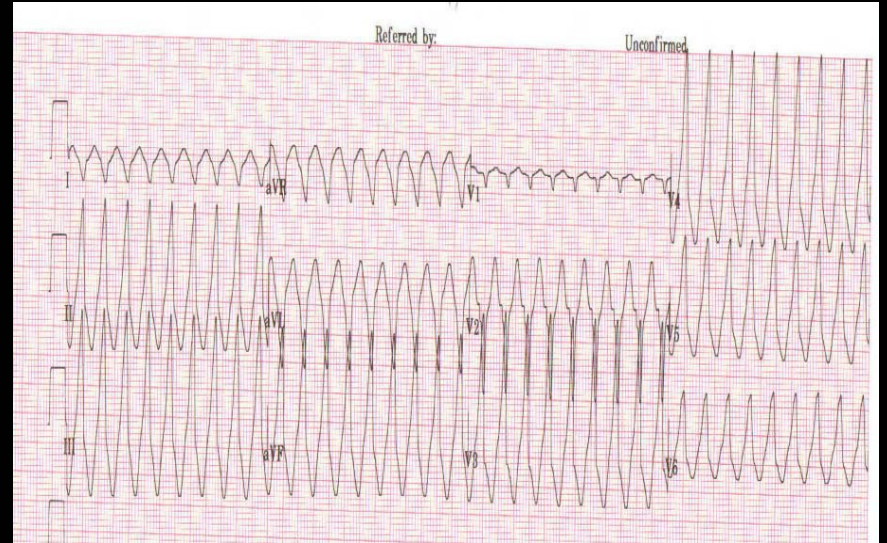
VT1 spontaneous induction



VT1



VT2



VT3

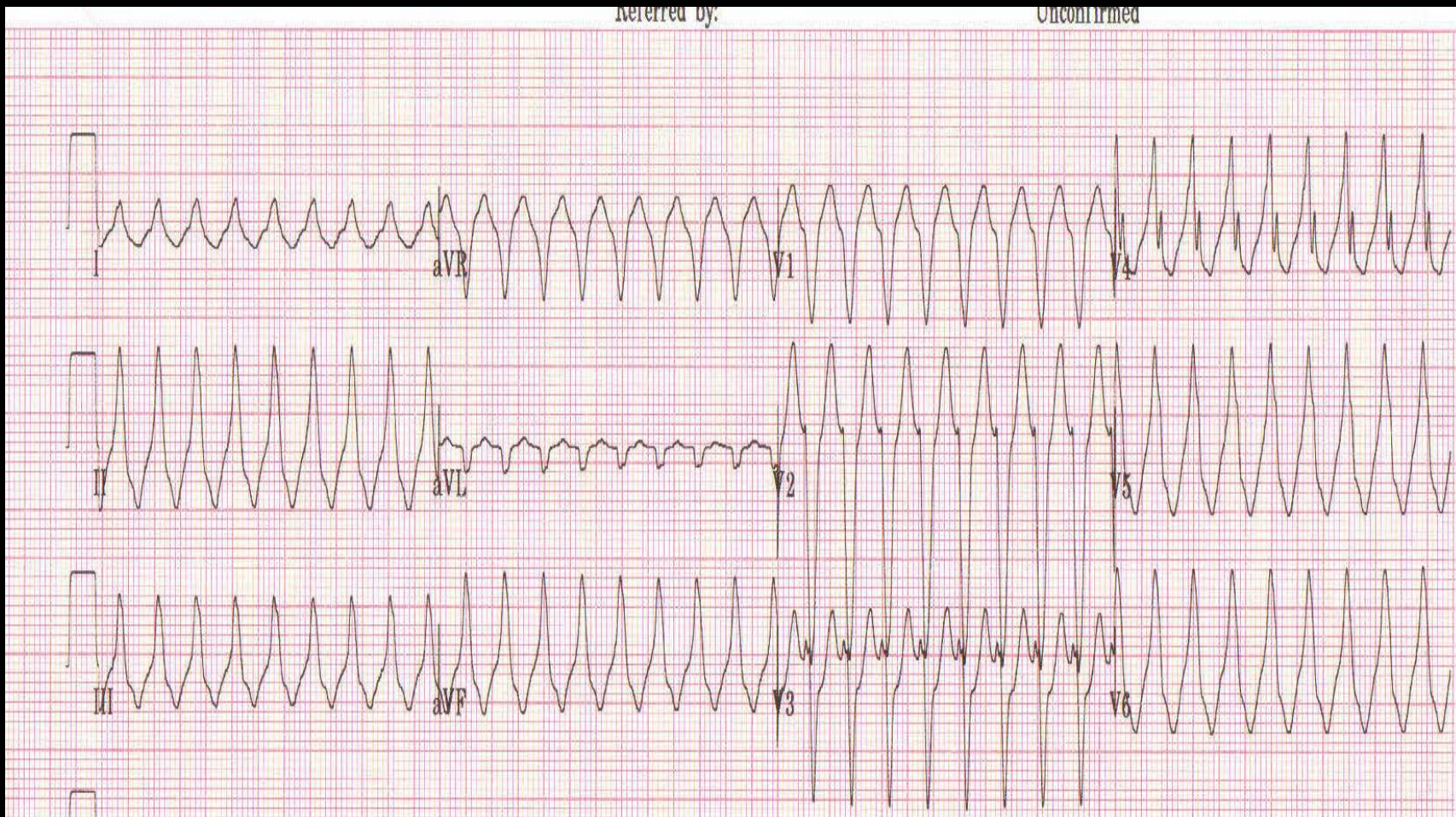


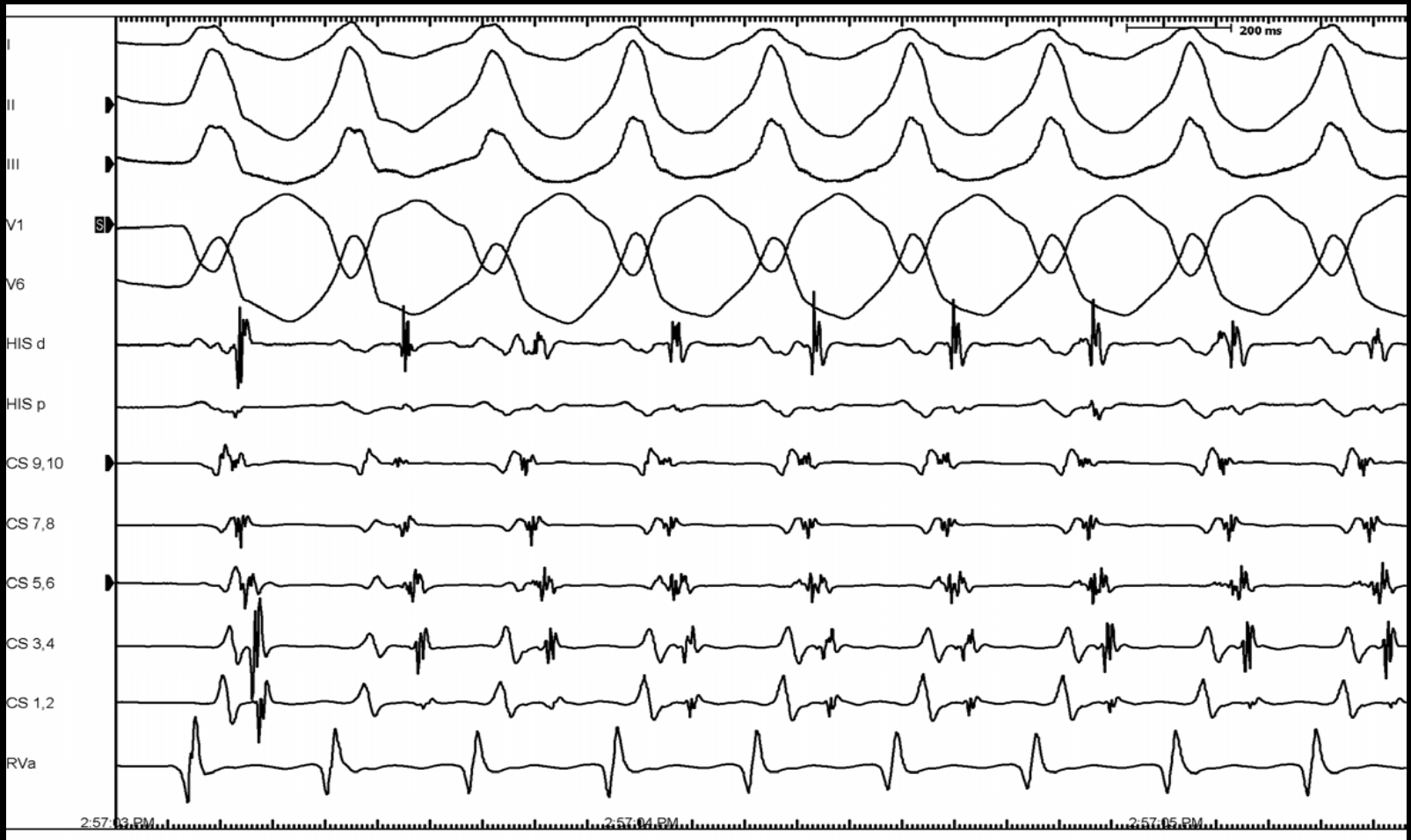
VT4

VT1 Postero-septal VT?

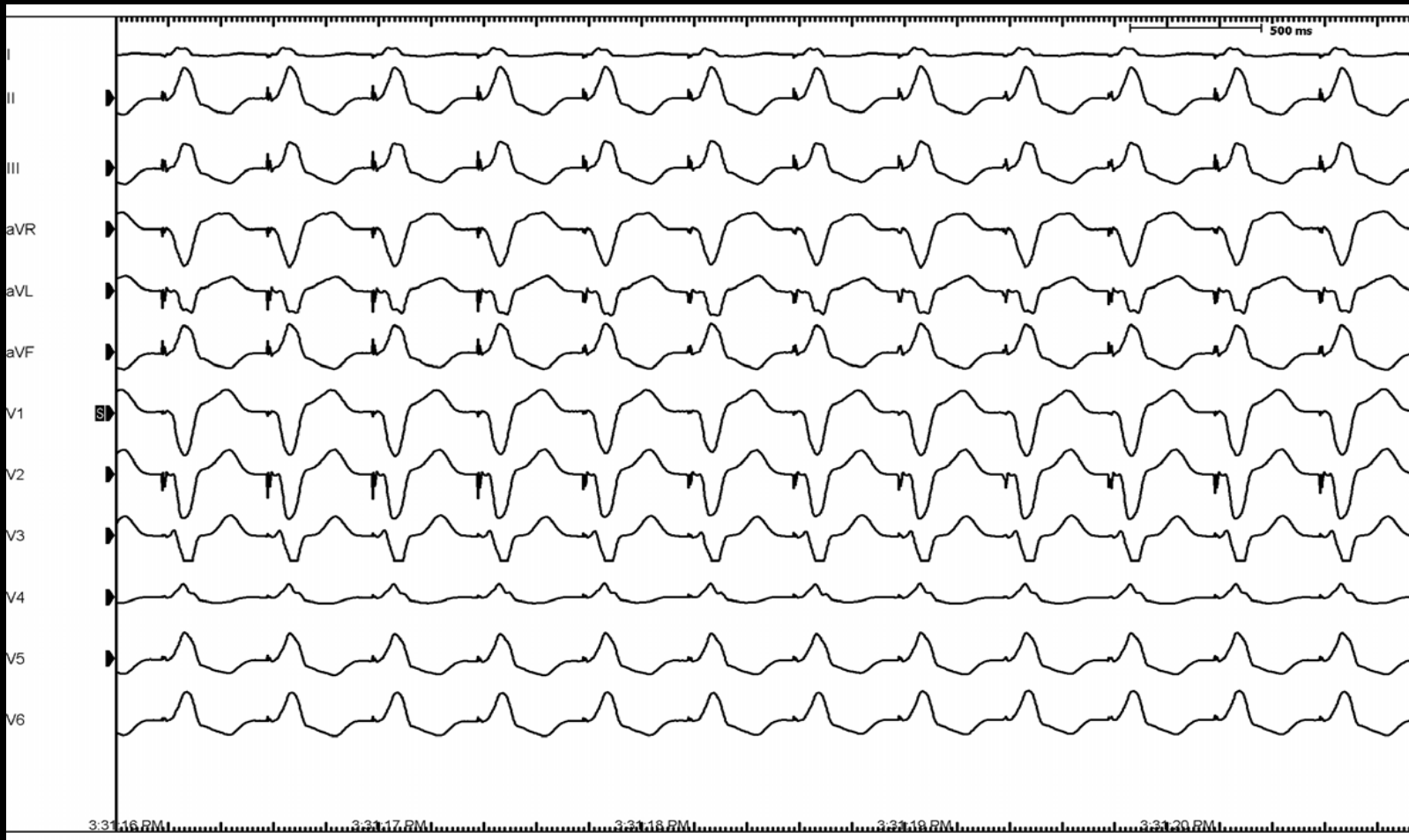
Referred by:

Unconfirmed

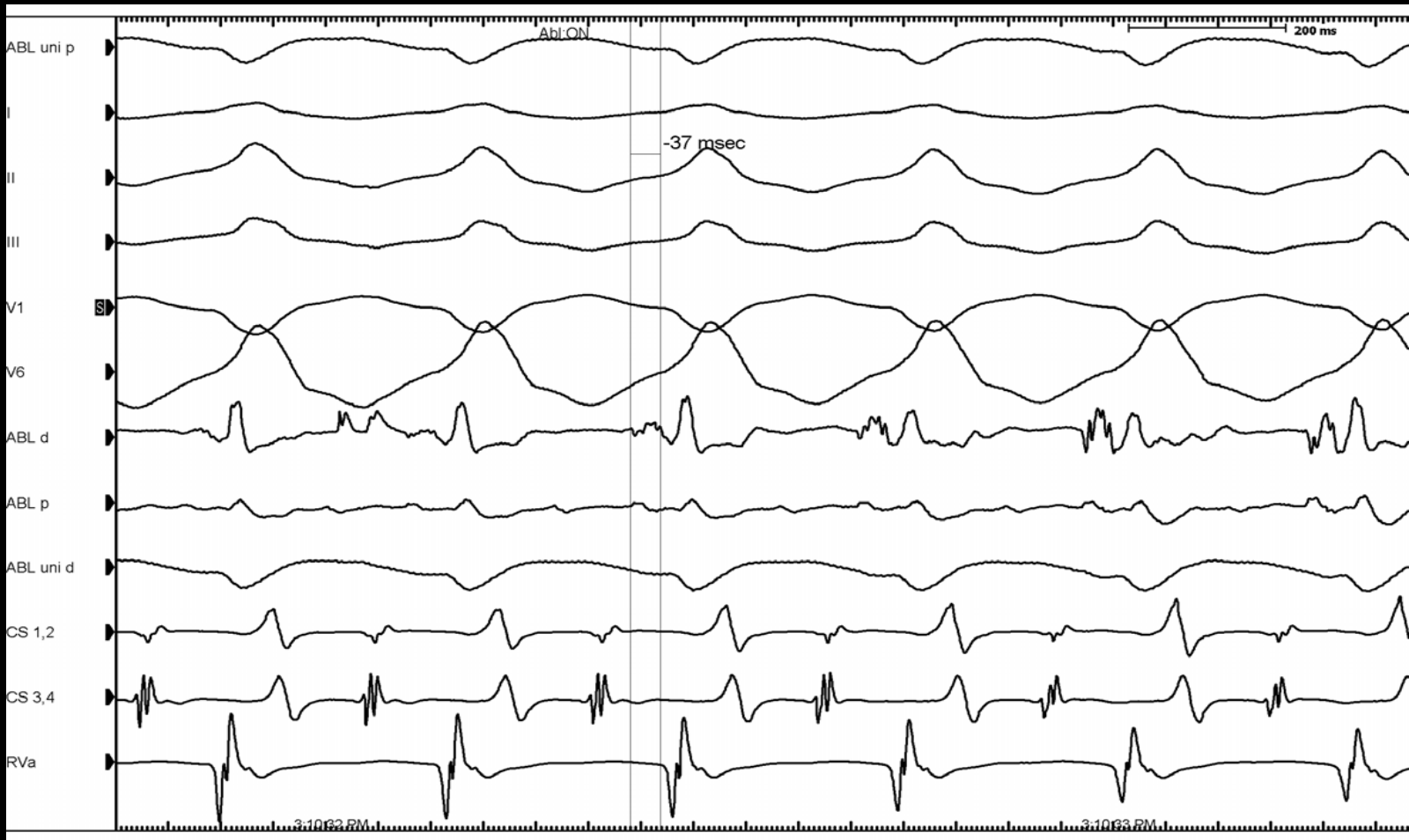




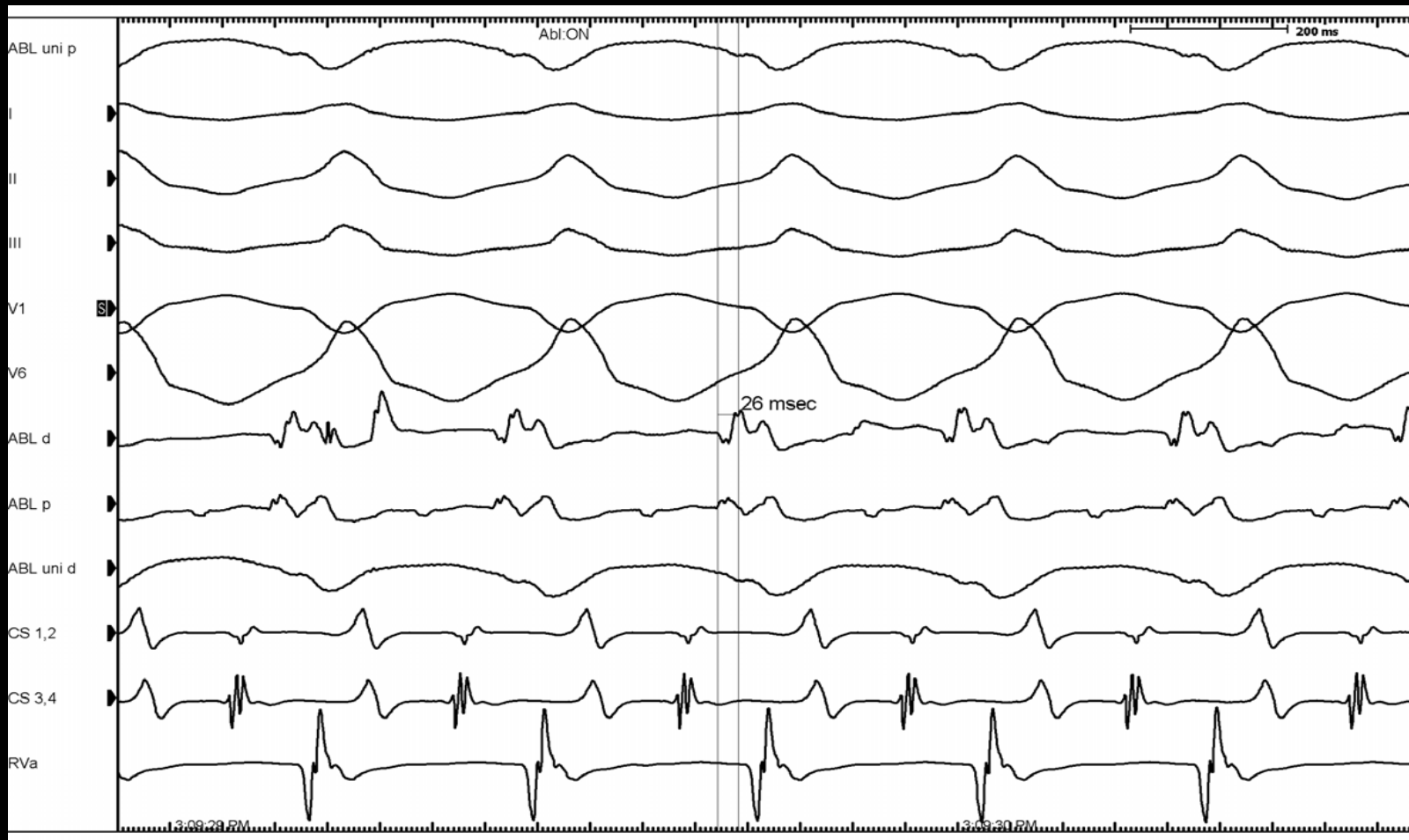
Paec map resembles VT1 from free wall



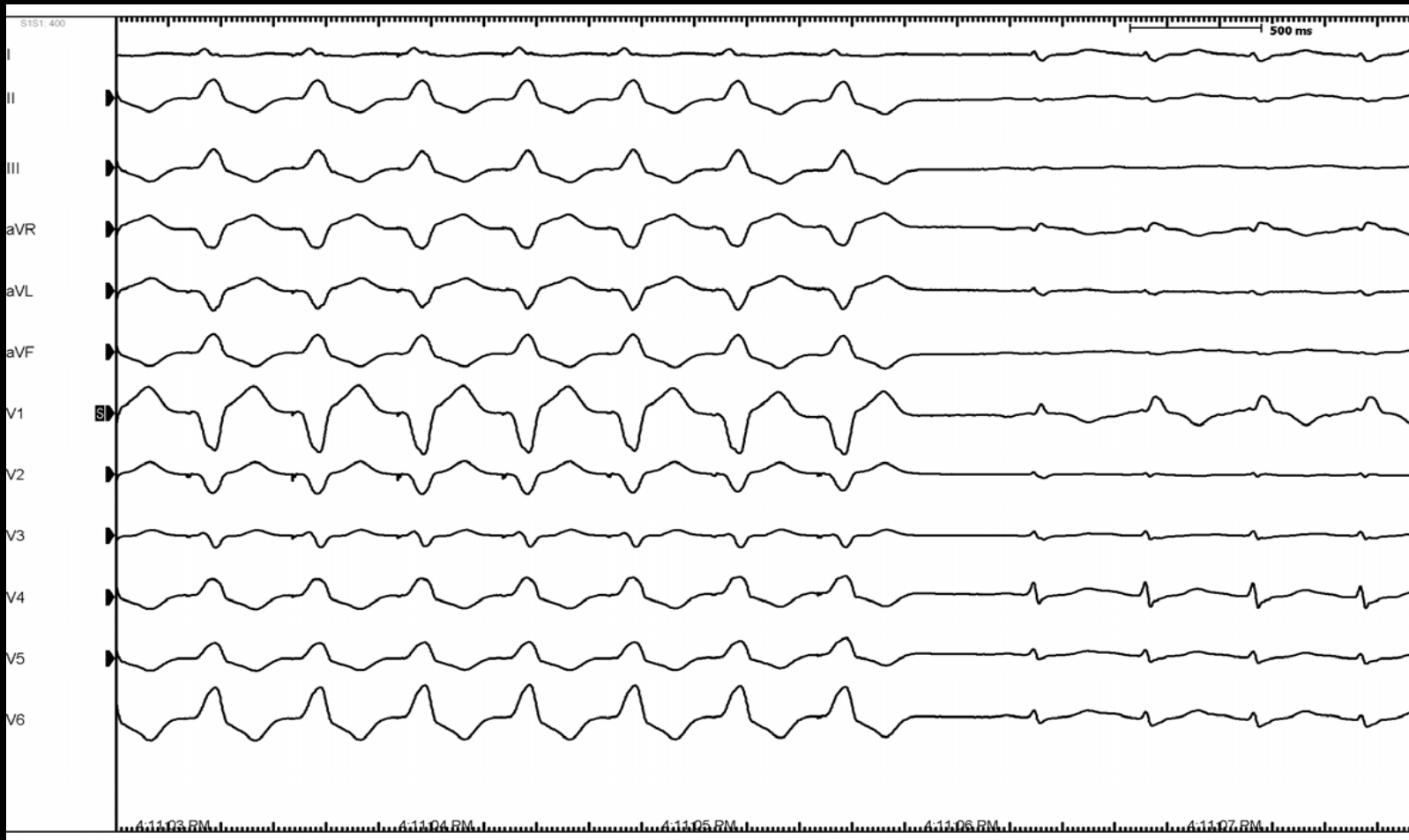
Vt1 ABLATION SITE



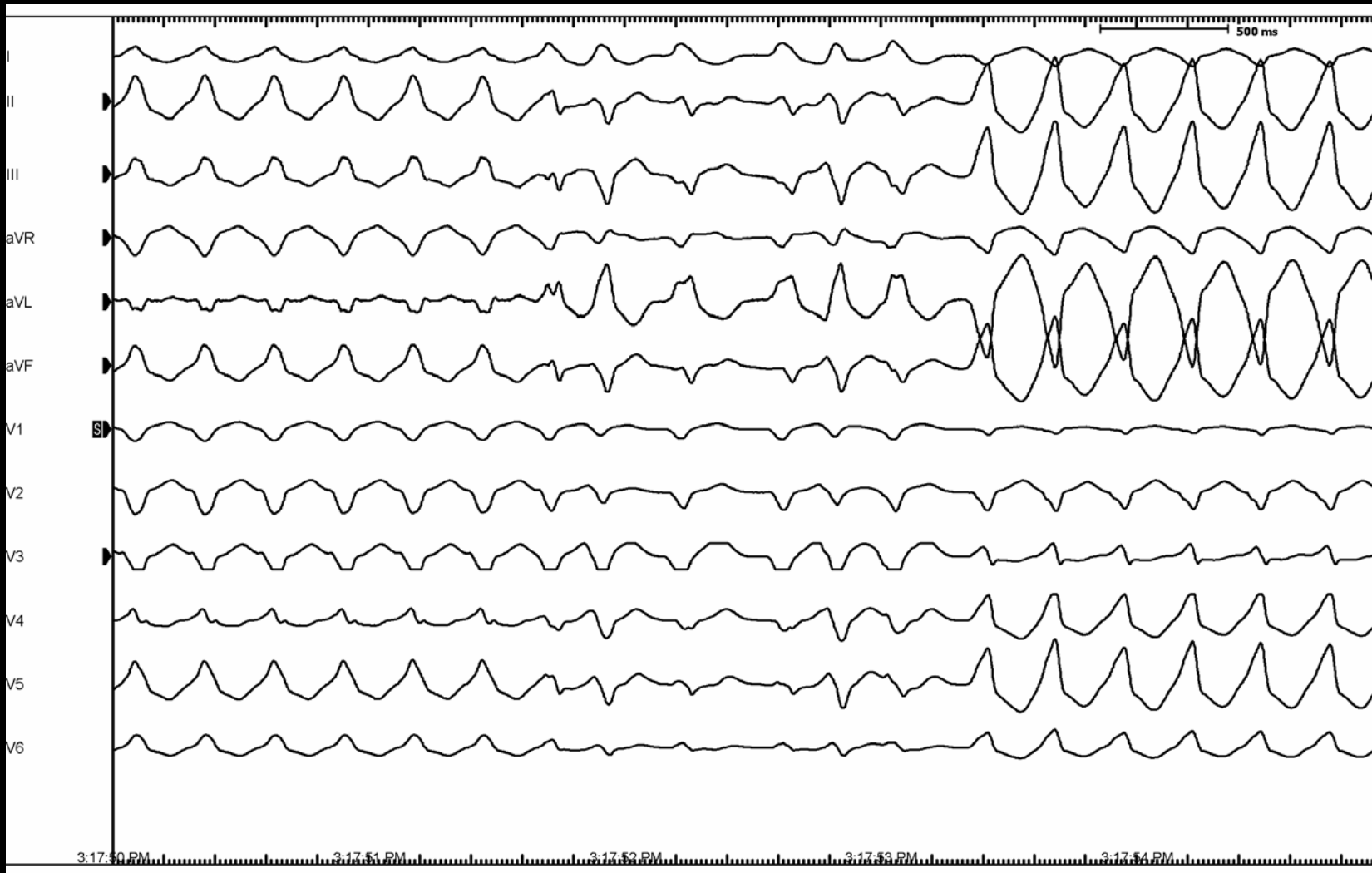
Another Site for VT1



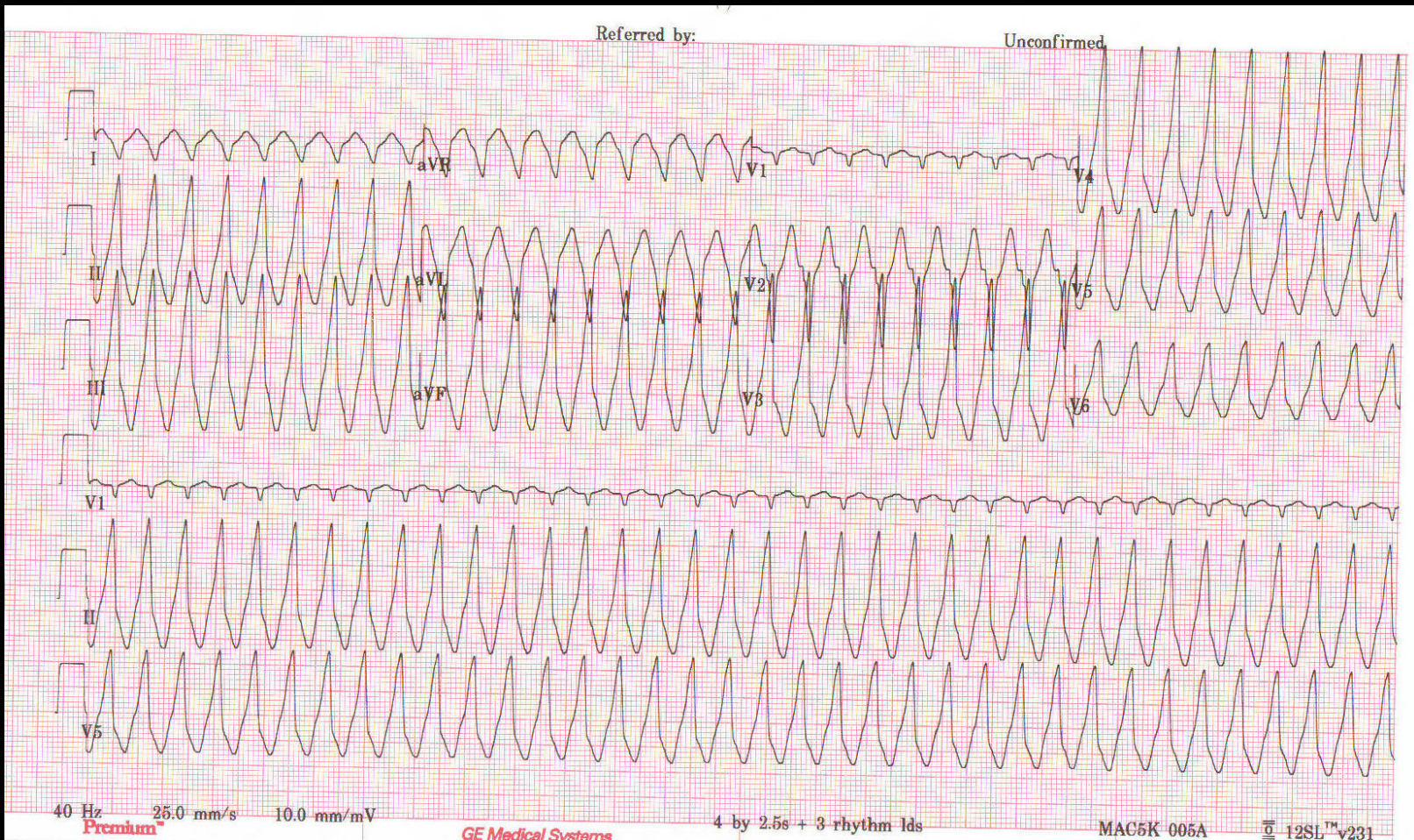
Pace map in Rt coronary Cusp Resembles VT1 ?

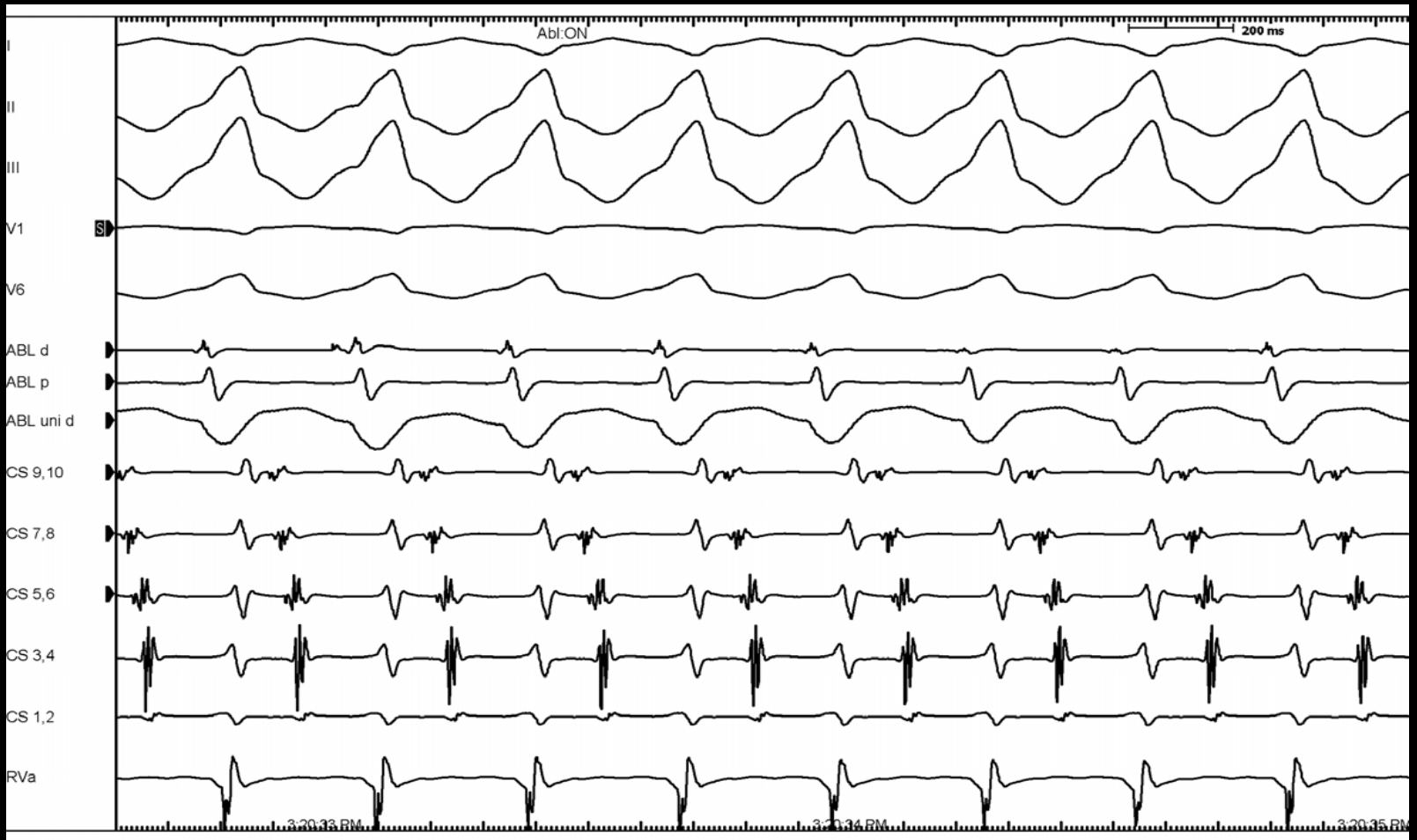


VT1 breaks with VPCs to VT2

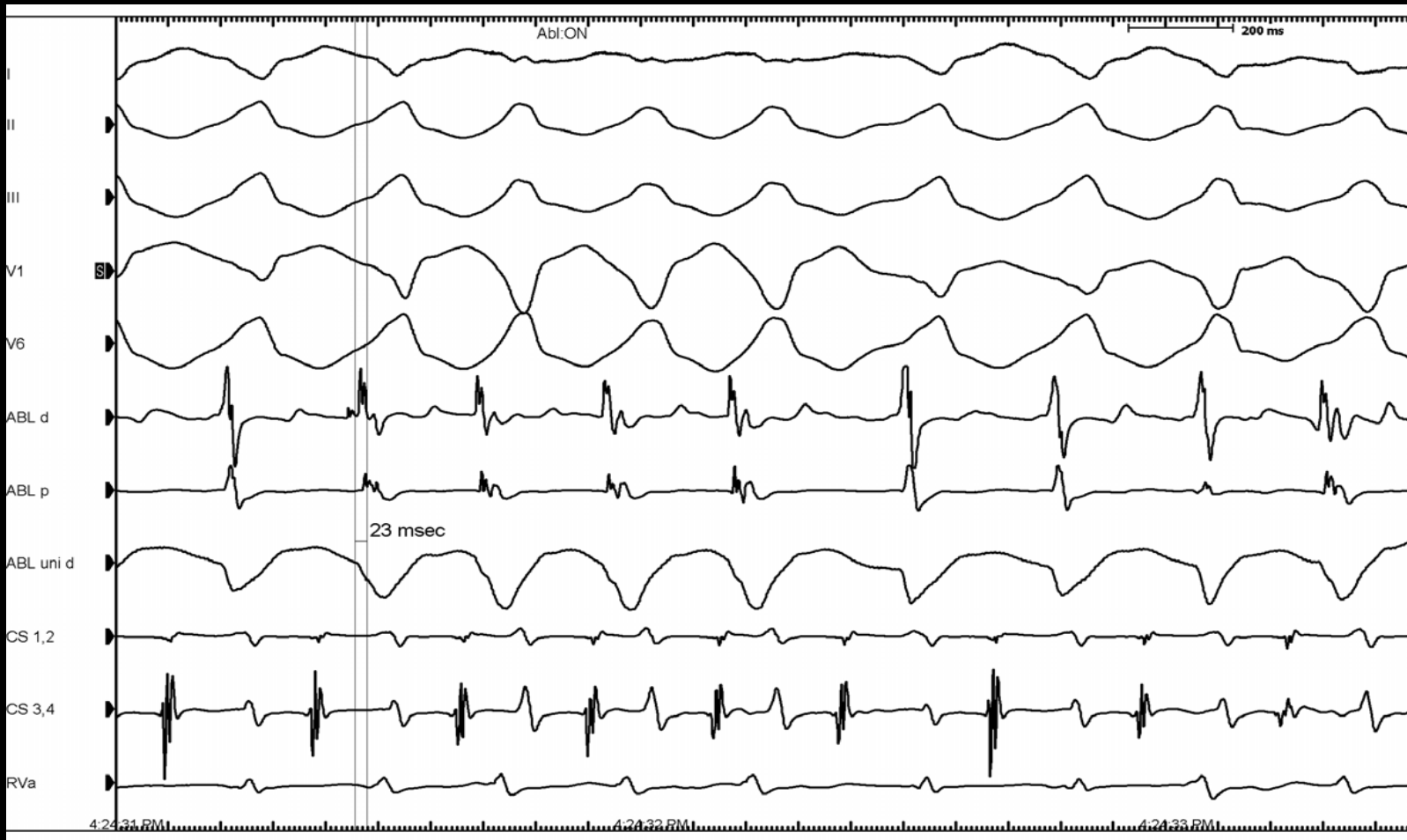


VT2- Antero-septal (rt cusp?)

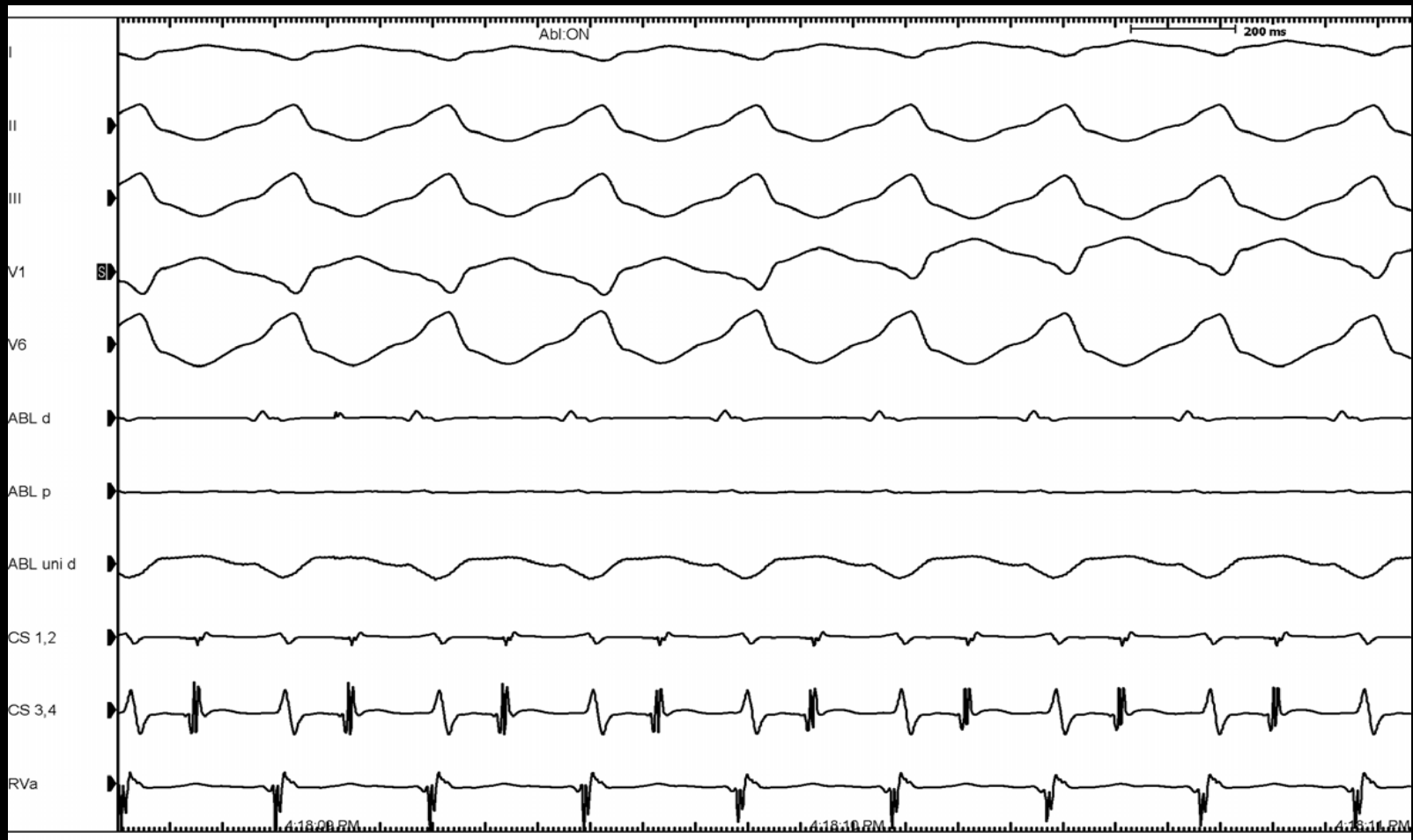




Ablation site in RVOT



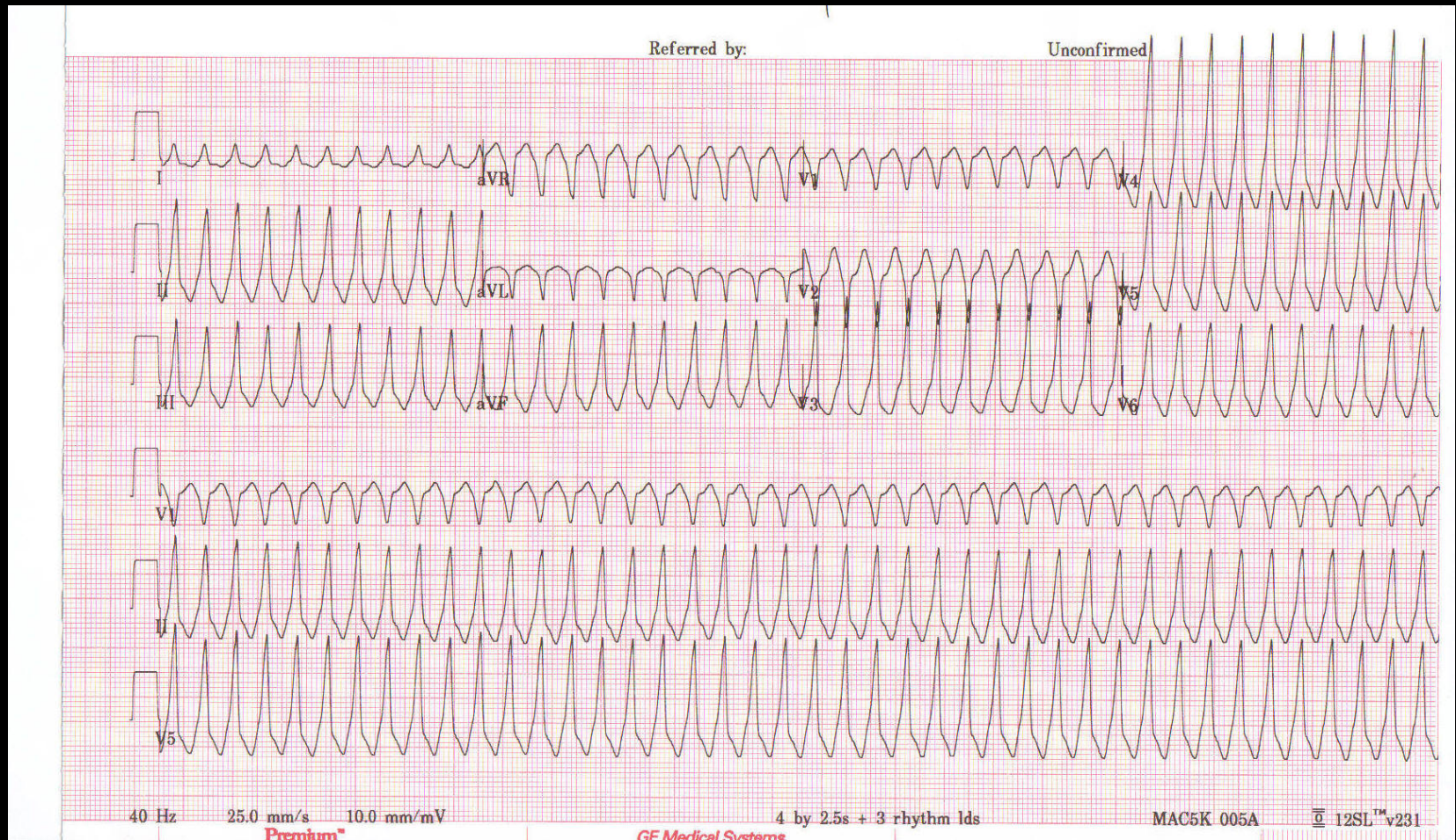
Ablation site in Rt. Coronary Cusp

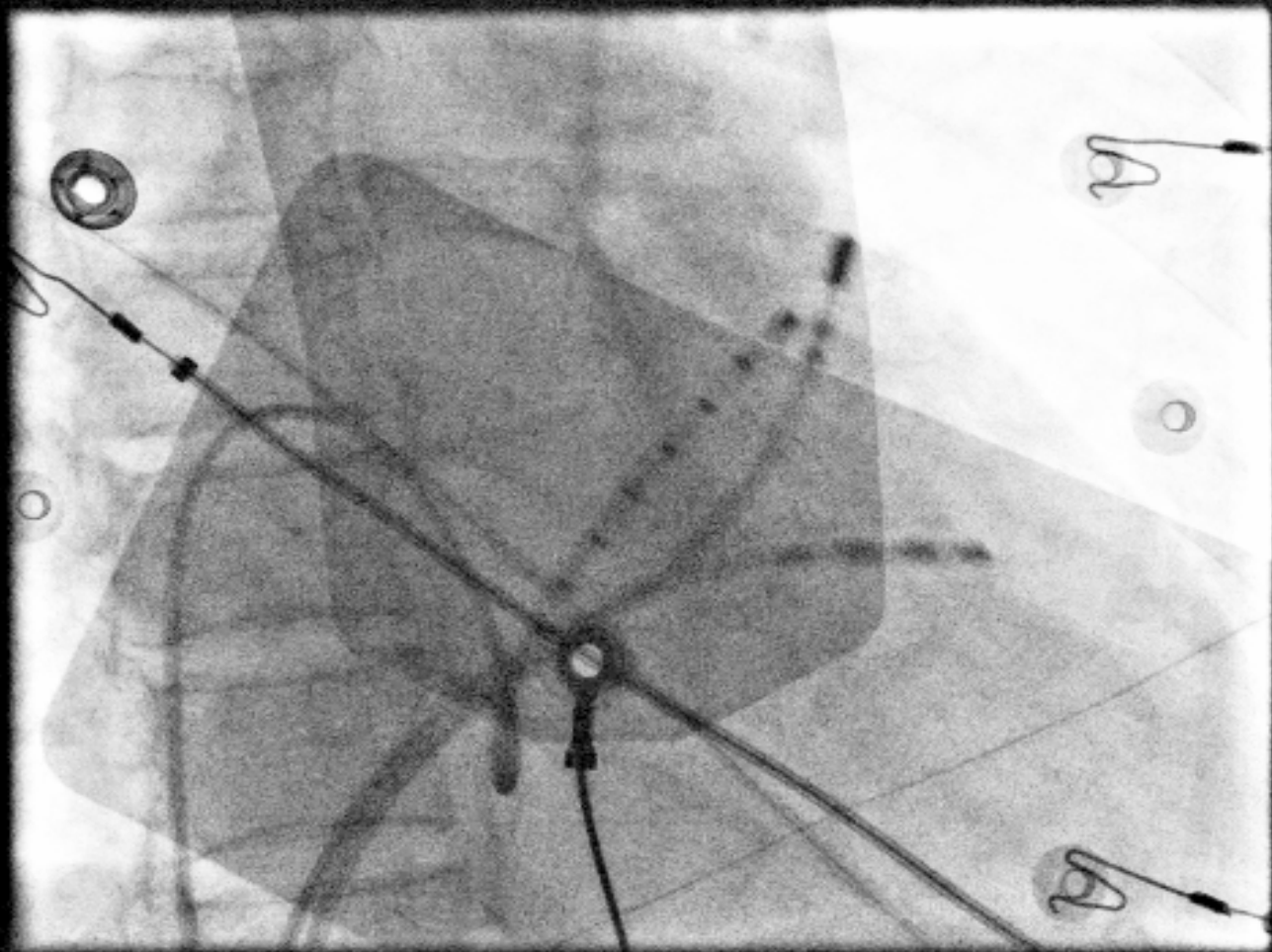


VT 3 inferior RV septal apex ?



VT4 Postero septal RVOT (Rt Cusp?)



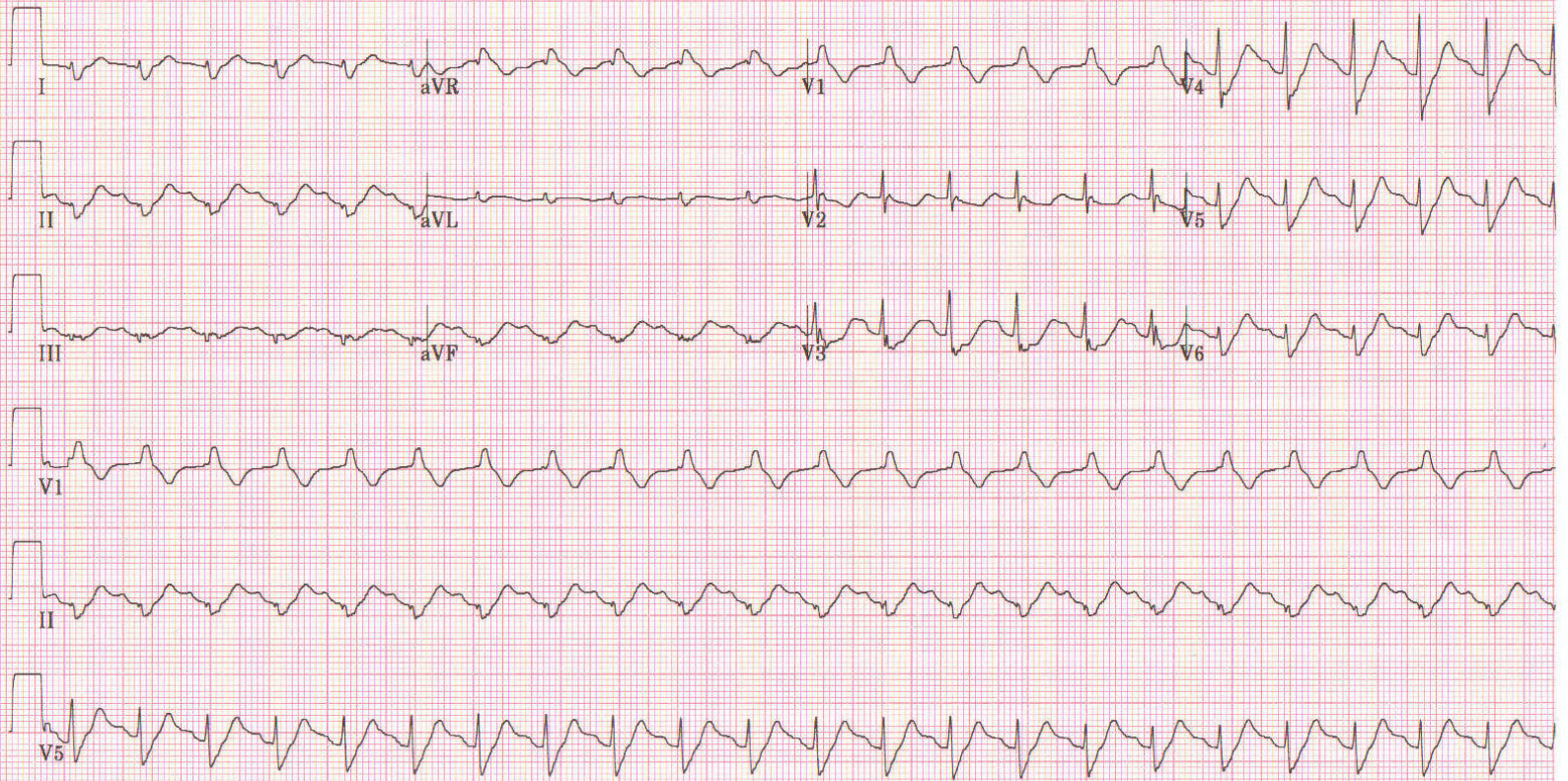




SVT

Referred by:

Unconfirmed



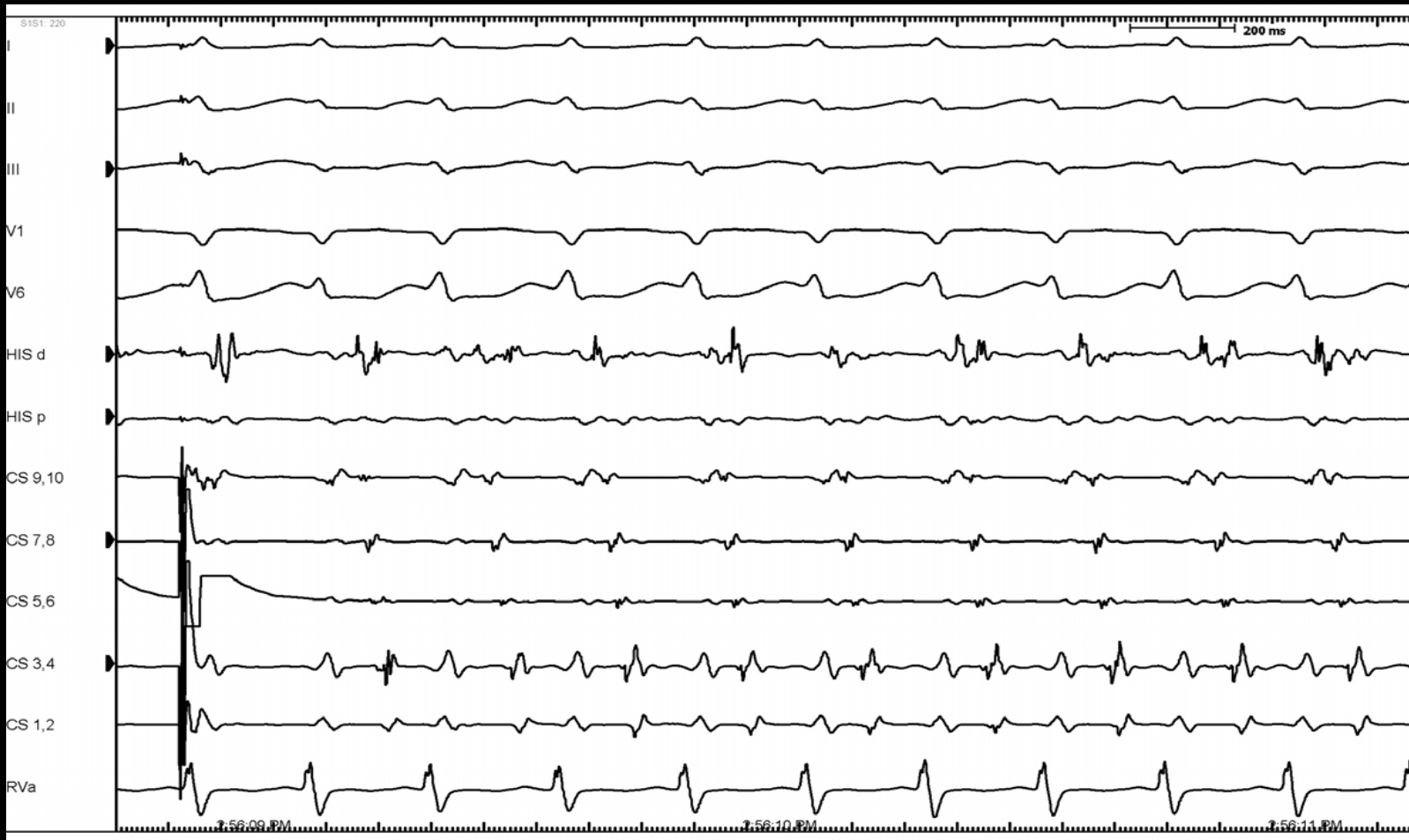
40 Hz 25.0 mm/s 10.0 mm/mV

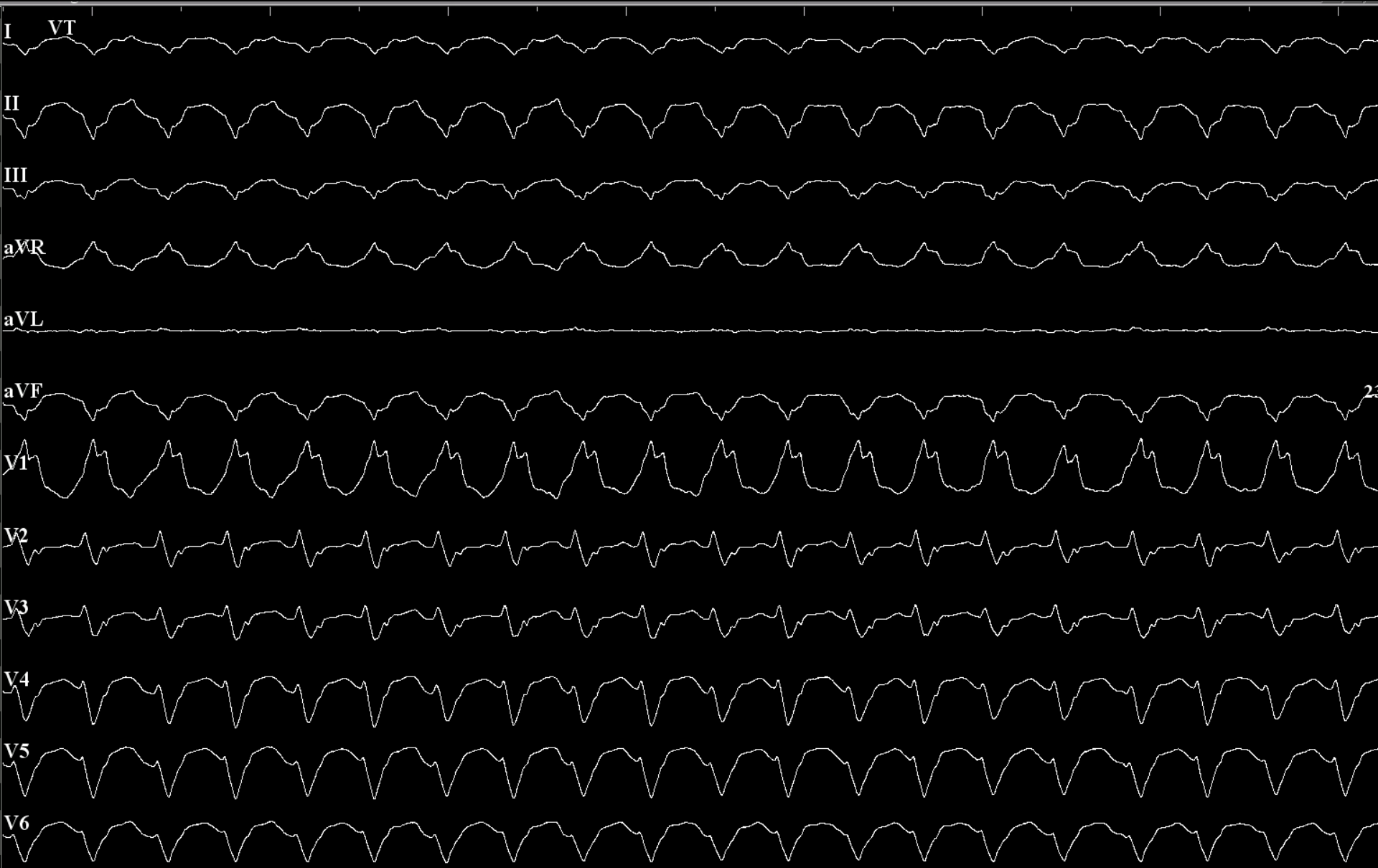
4 by 2.5 cm 2 calibration 1ds

MAC5K 005A

0-12SI™ 221

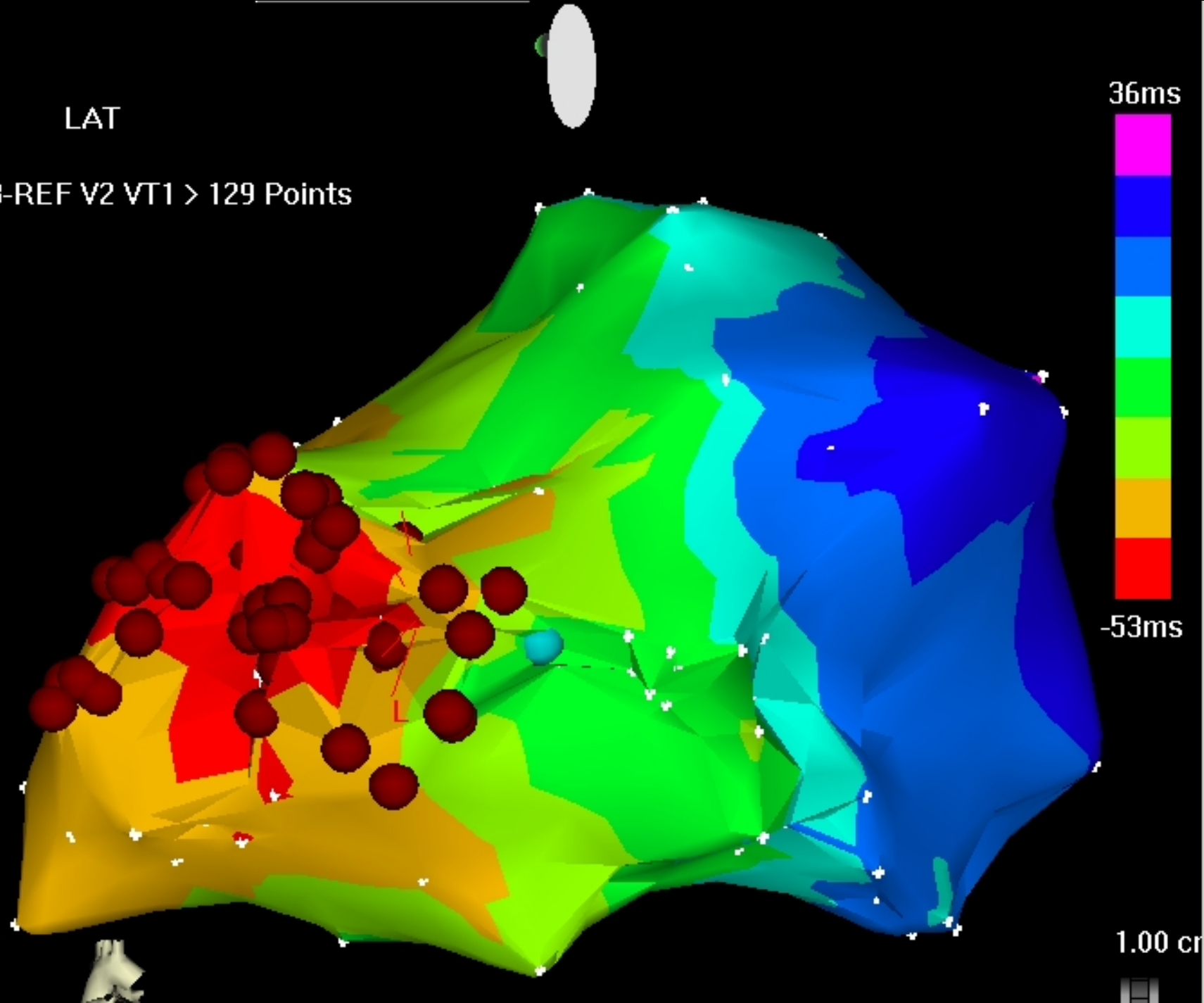
Induction of AVNRT





LAT

3-REF V2 VT1 > 129 Points



Breaks During Ablation



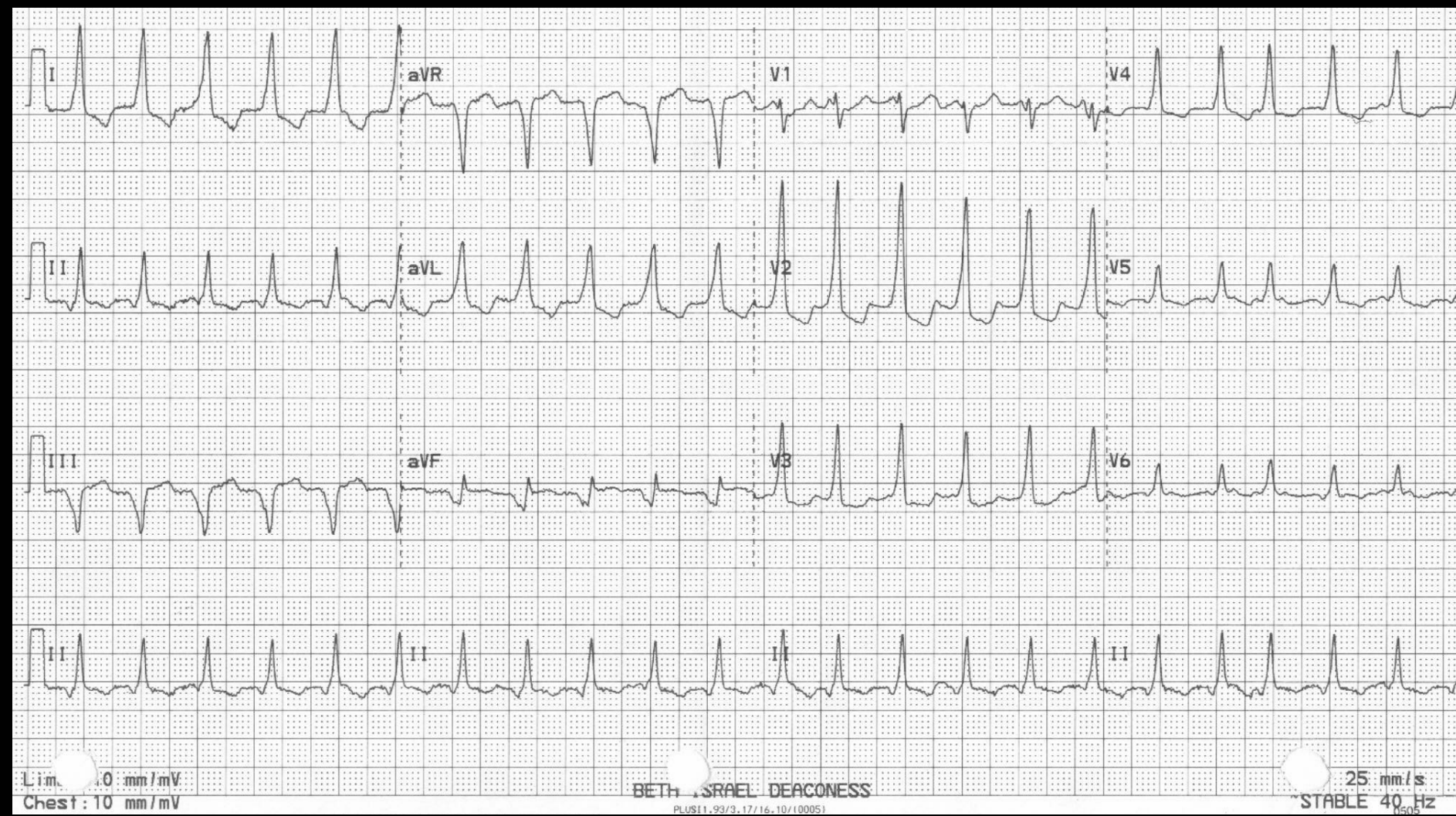
WCT



History

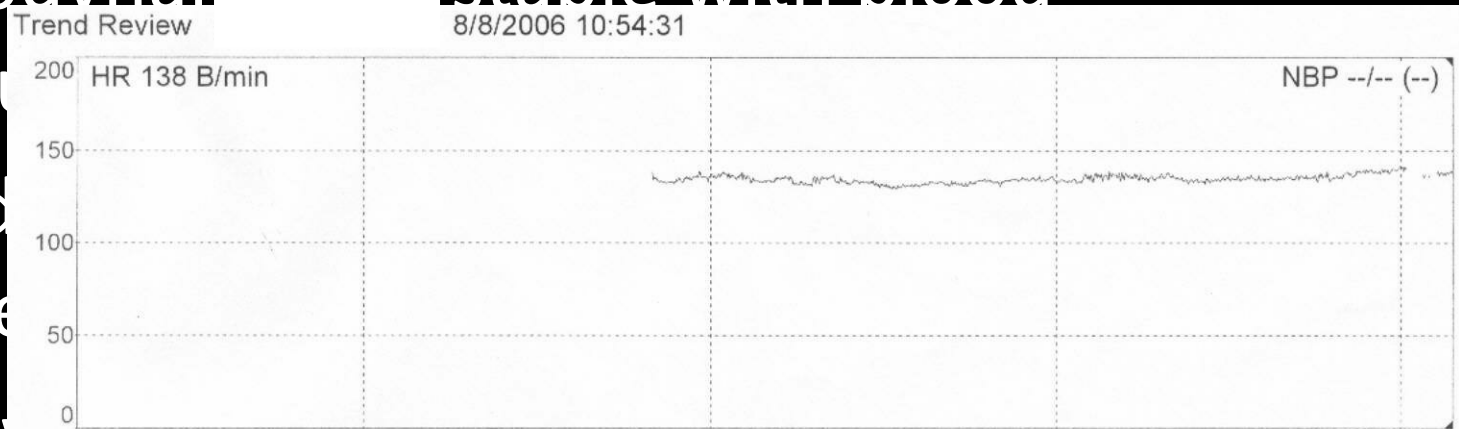
- 58 year-old man with HTN, COPD, CAD
 - Reports a history of MI in his 20's requiring a month-long hospitalization
 - Stress test in 2001 showed a severe fixed inferior and inferolateral defect and ejection fraction of 35%
 - No chest pain or CHF symptoms in the past
- Several months of worsening fatigue and dyspnea
- Over 2-3 weeks, fatigue and dyspnea markedly worse

Presenting ECG



Hospital Course

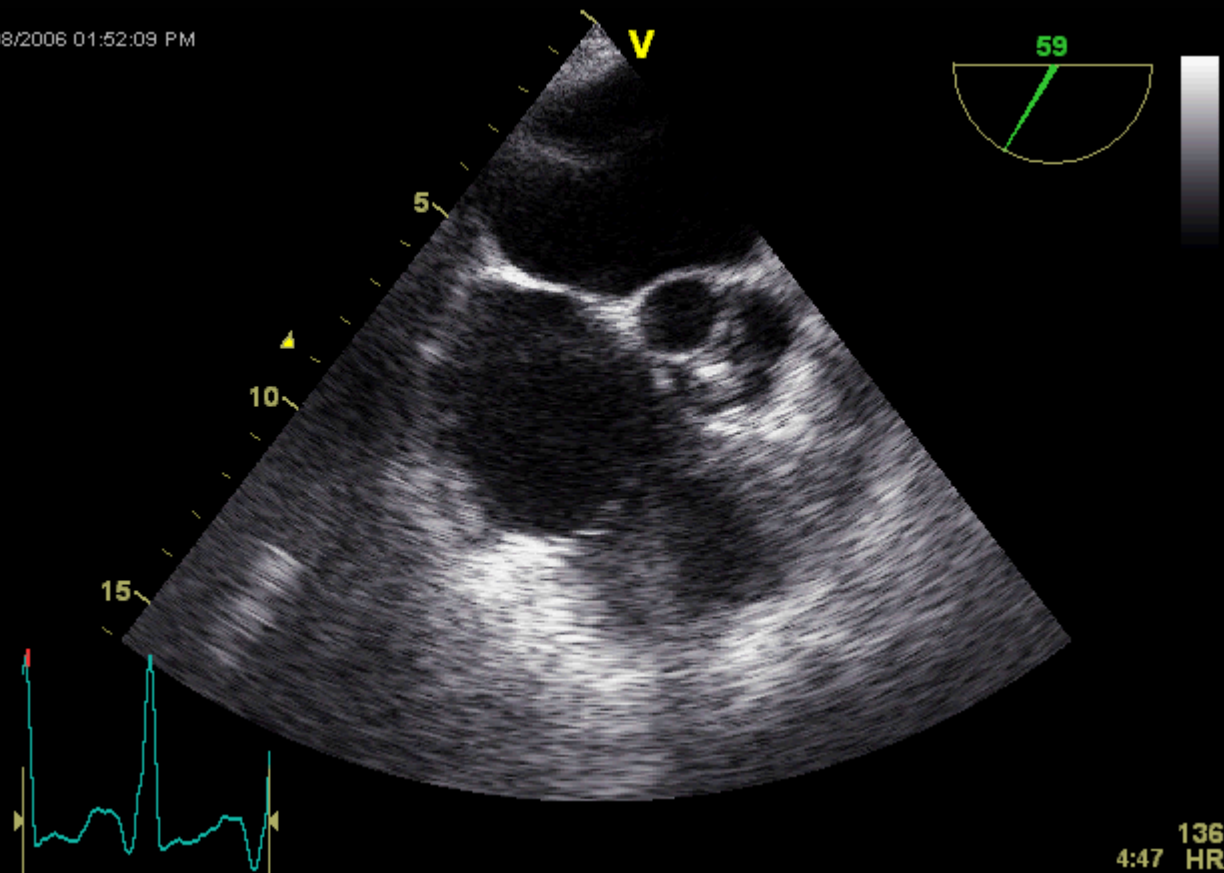
- Diagnosed as atrial fibrillation with rapid ventricular response
 - Treated with rate control medications – metoprolol and diltiazem
- No effect on ventricular rate
- Hemodynamically stable with blood



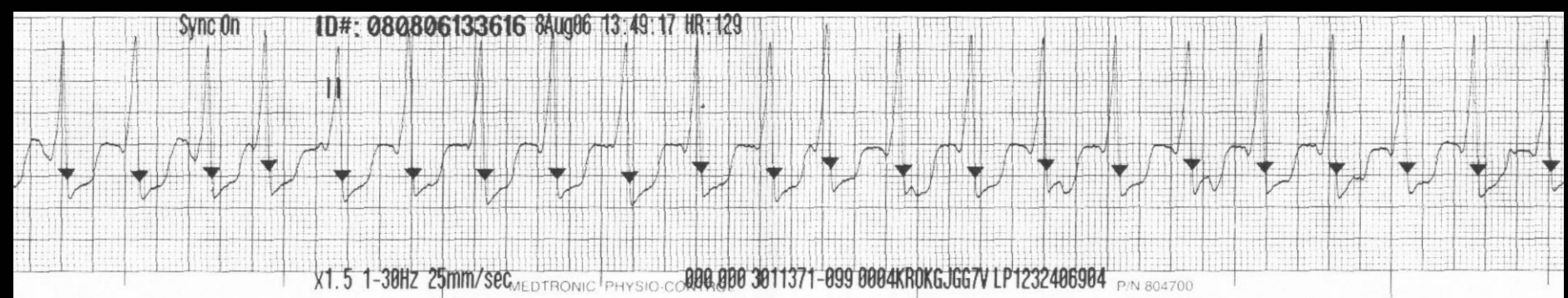
- Scheduled for surgery
- the next day
- Review of telemetry.

TEE

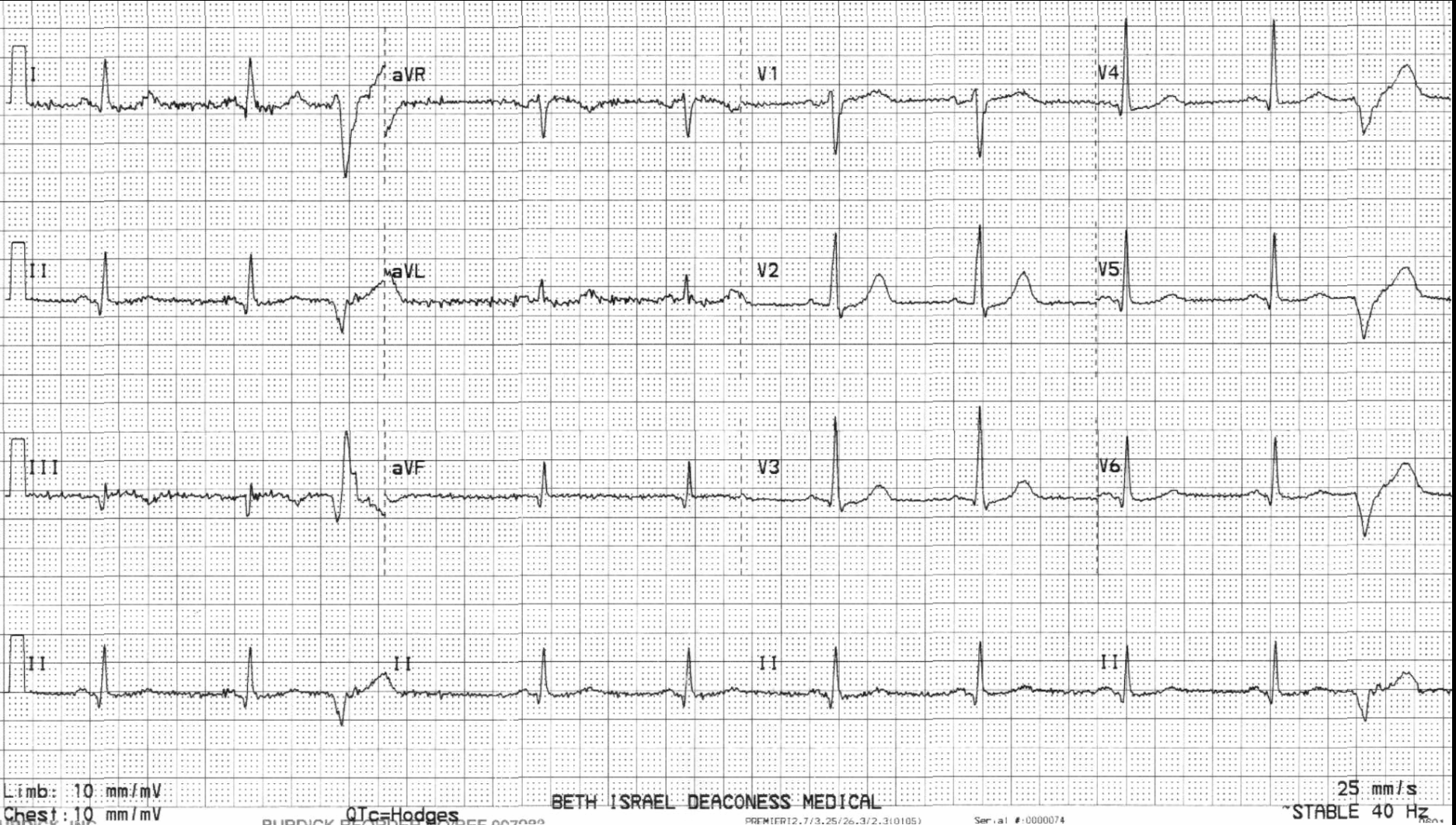
08/08/2006 01:52:09 PM



Telemetry strip prior to DCCV



Post-DCCV ECG



Limb: 10 mm/mV
Chest: 10 mm/mV

BETH ISRAEL DEACONESS MEDICAL

25 mm/s
STABLE 40 Hz

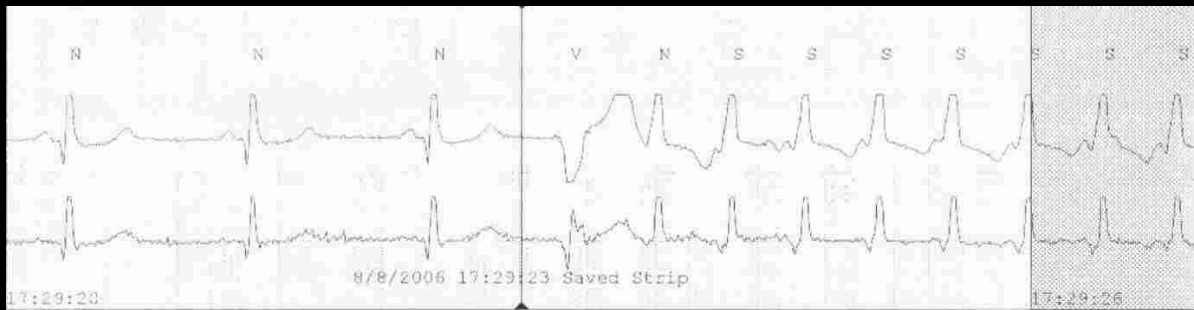
QTC=Hodges

PREMIER12.7/3.25/26.3/2.3(0105)

Serial #: 0000074

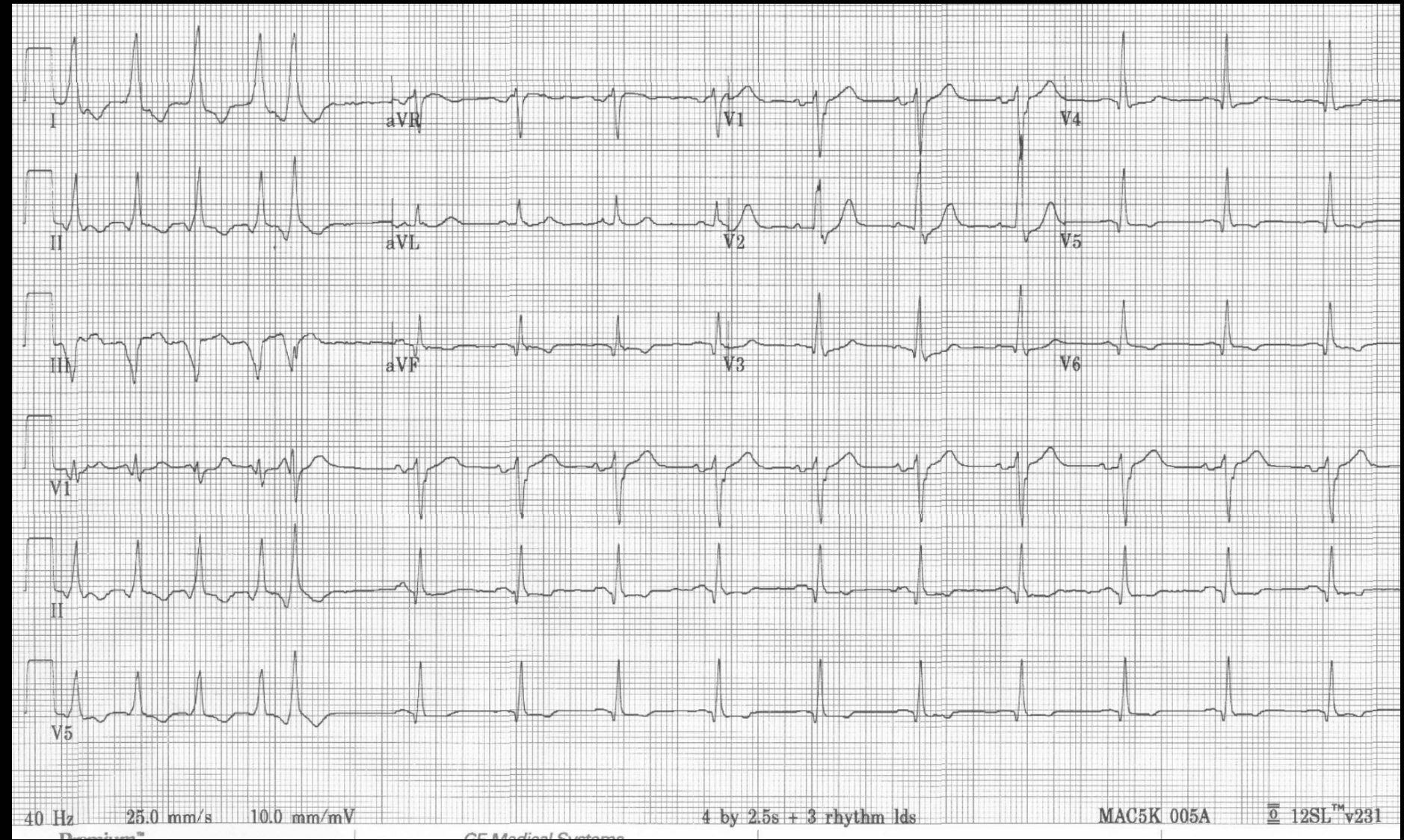
Hospital Course (cont.)

- Recurrent episodes of WCT after DCCV

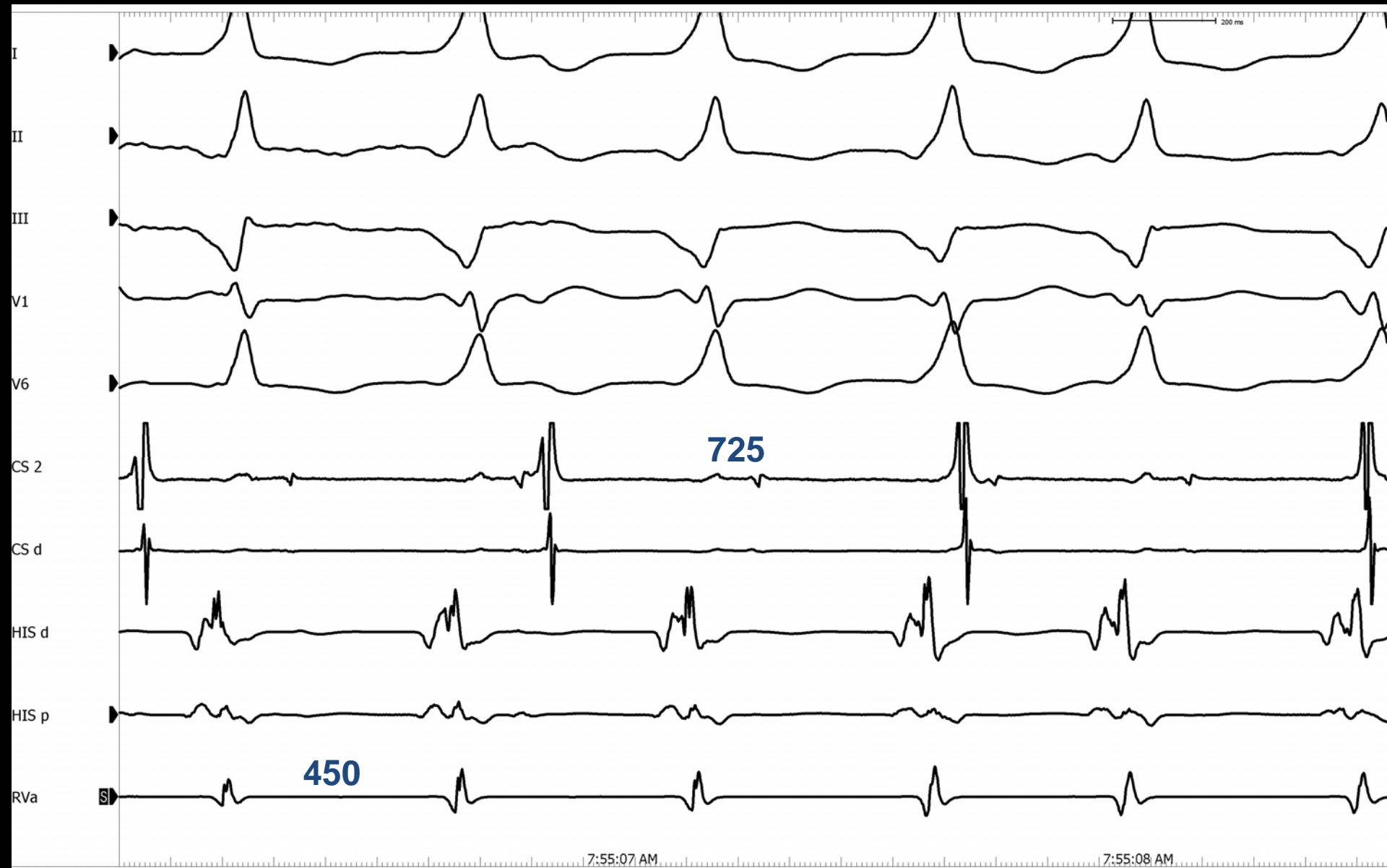


- Referred for EPS

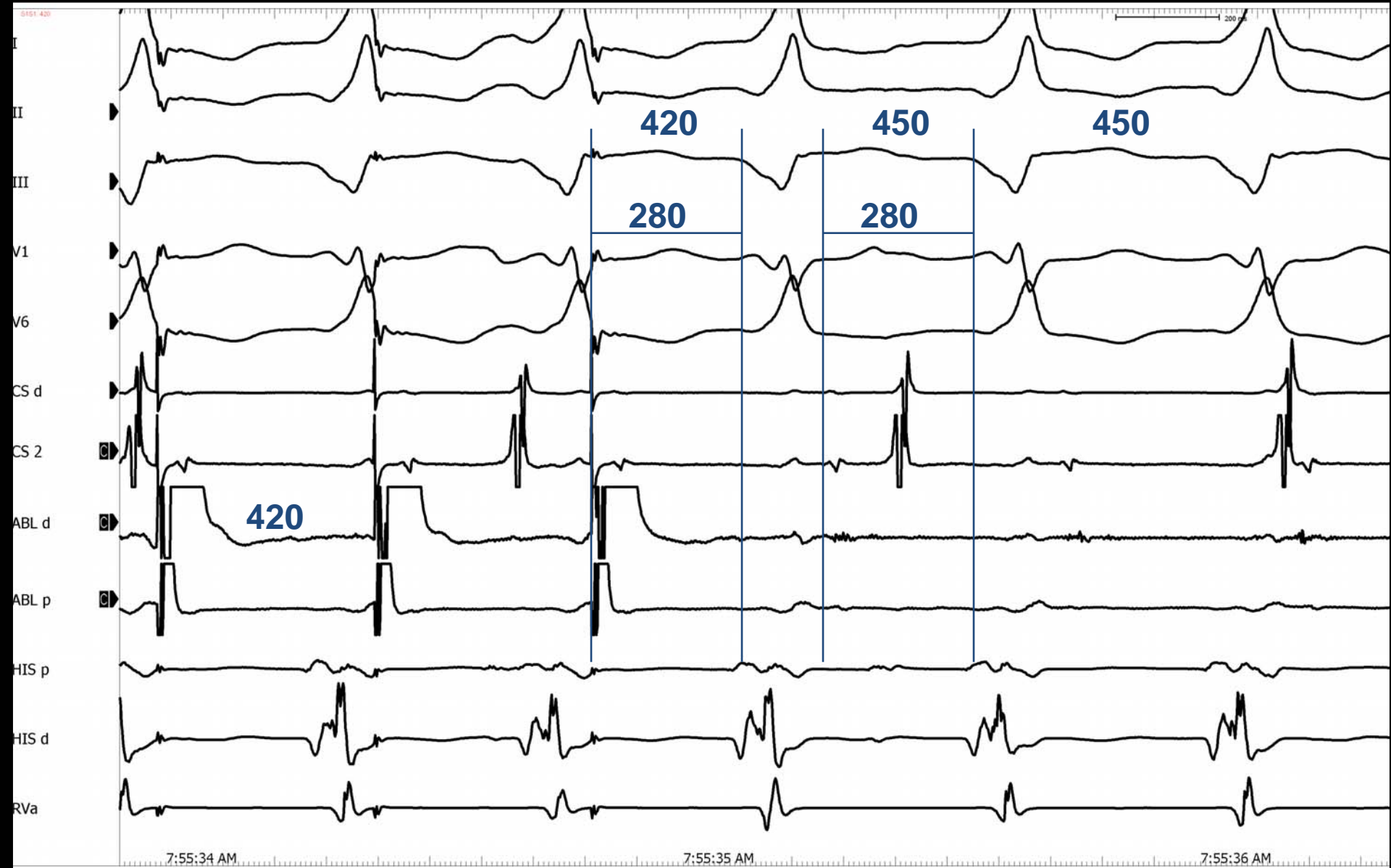
Spontaneous termination



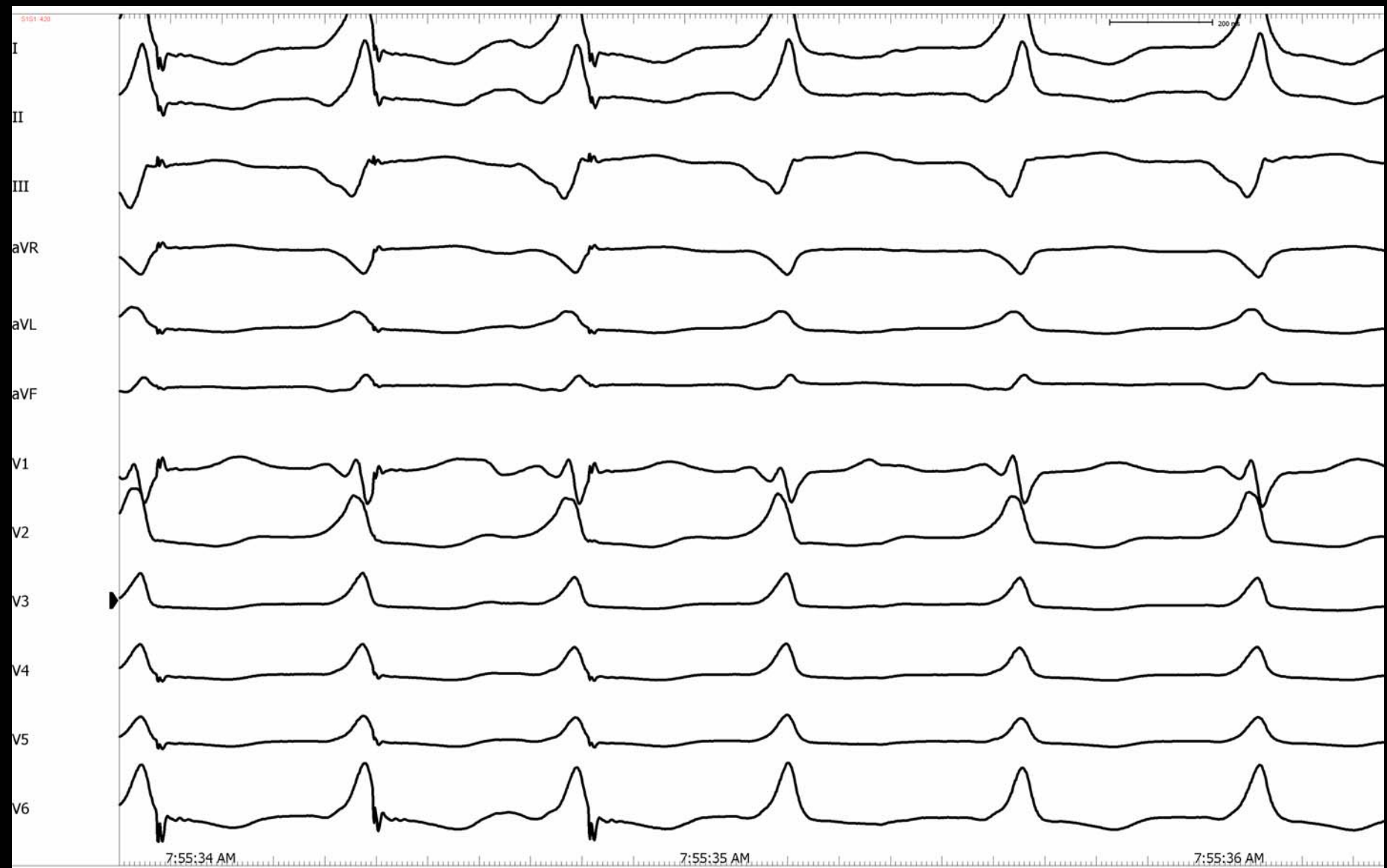
Baseline



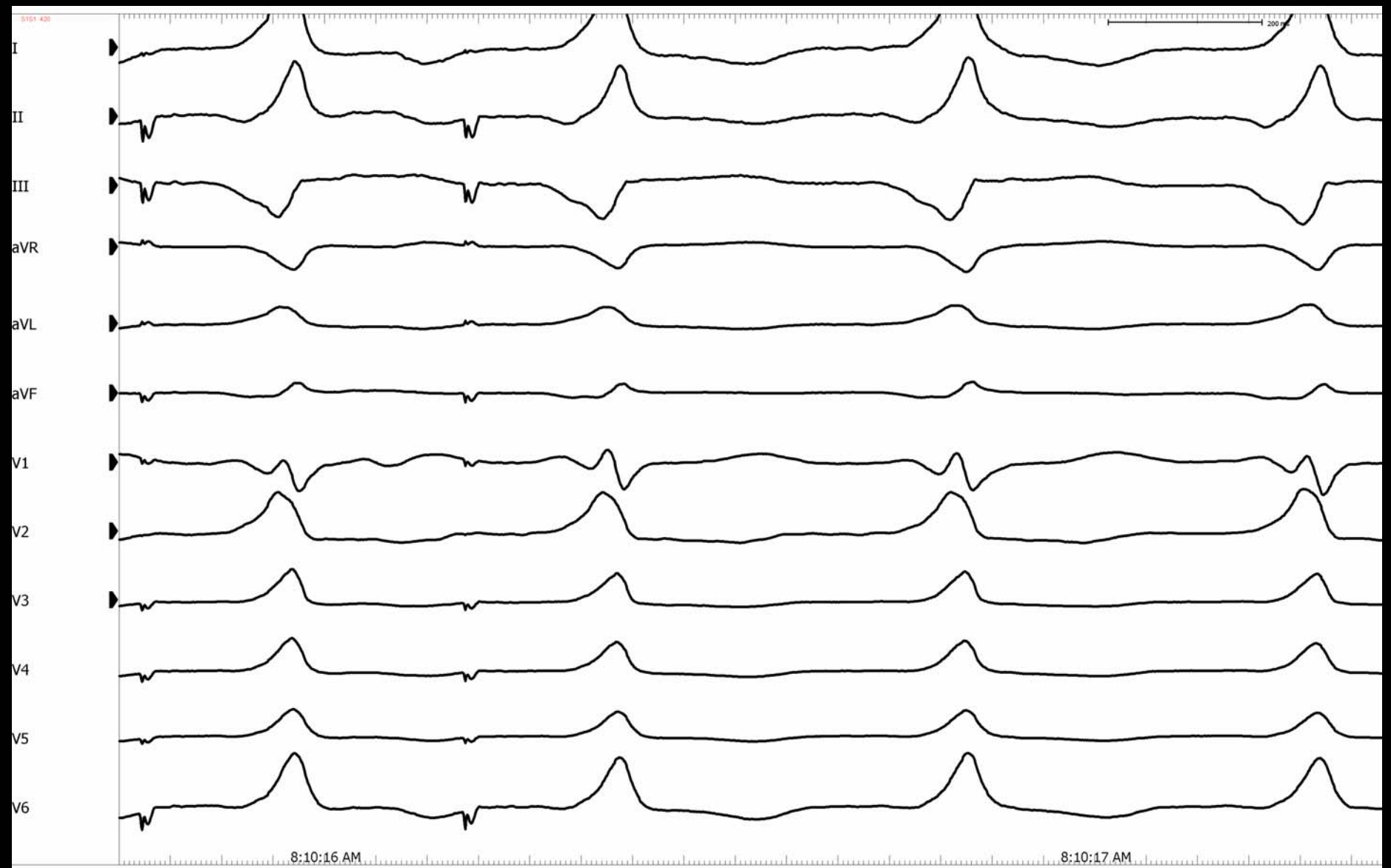
Pacing from basal inferior wall 420ms



Pacing from basal inferior wall 420ms



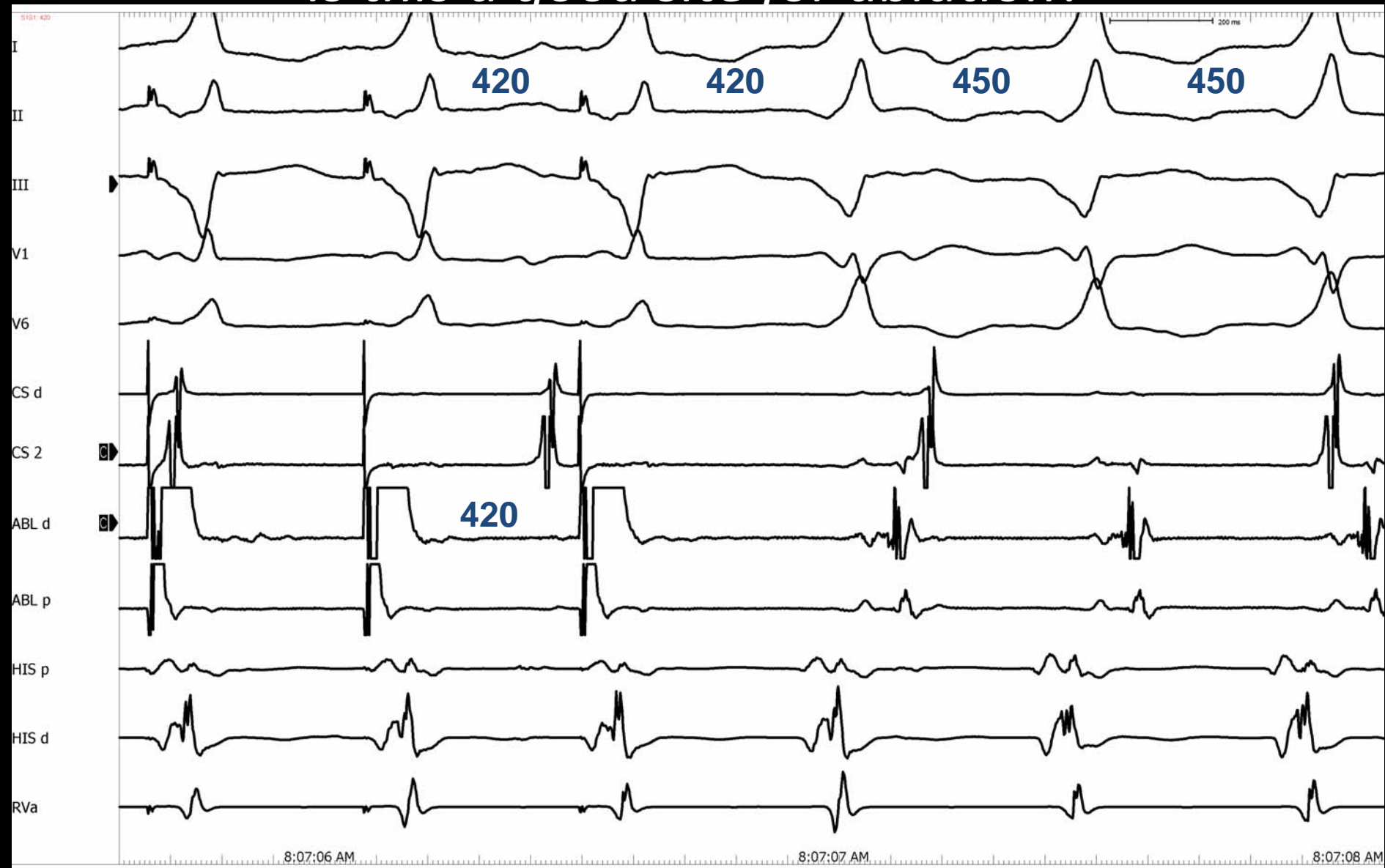
Pacing from basal septal inferior wall 420ms





Pacing from the basal inferolateral wall 420ms

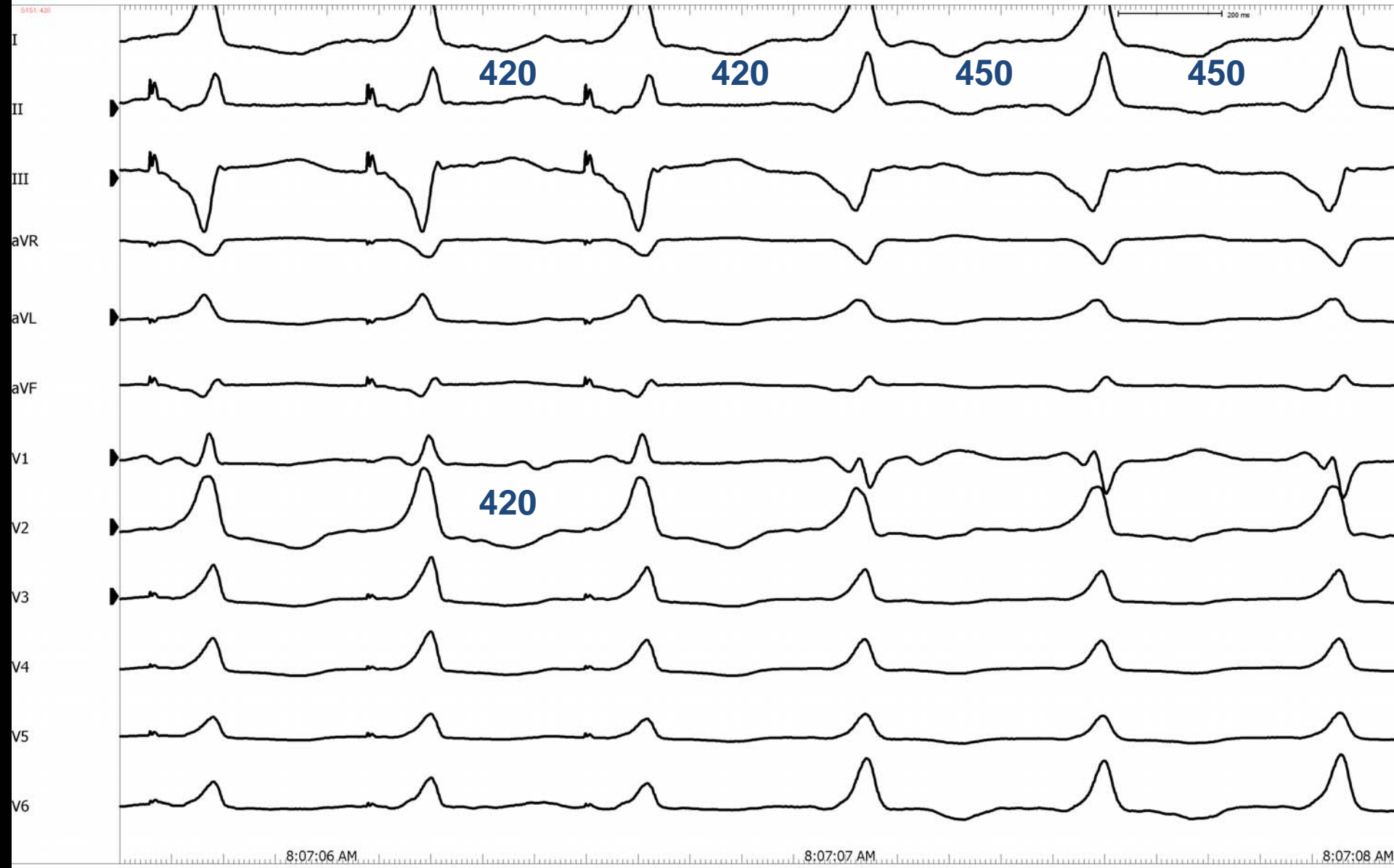
Is this a good site for ablation?





Pacing from the basal inferolateral wall 420ms

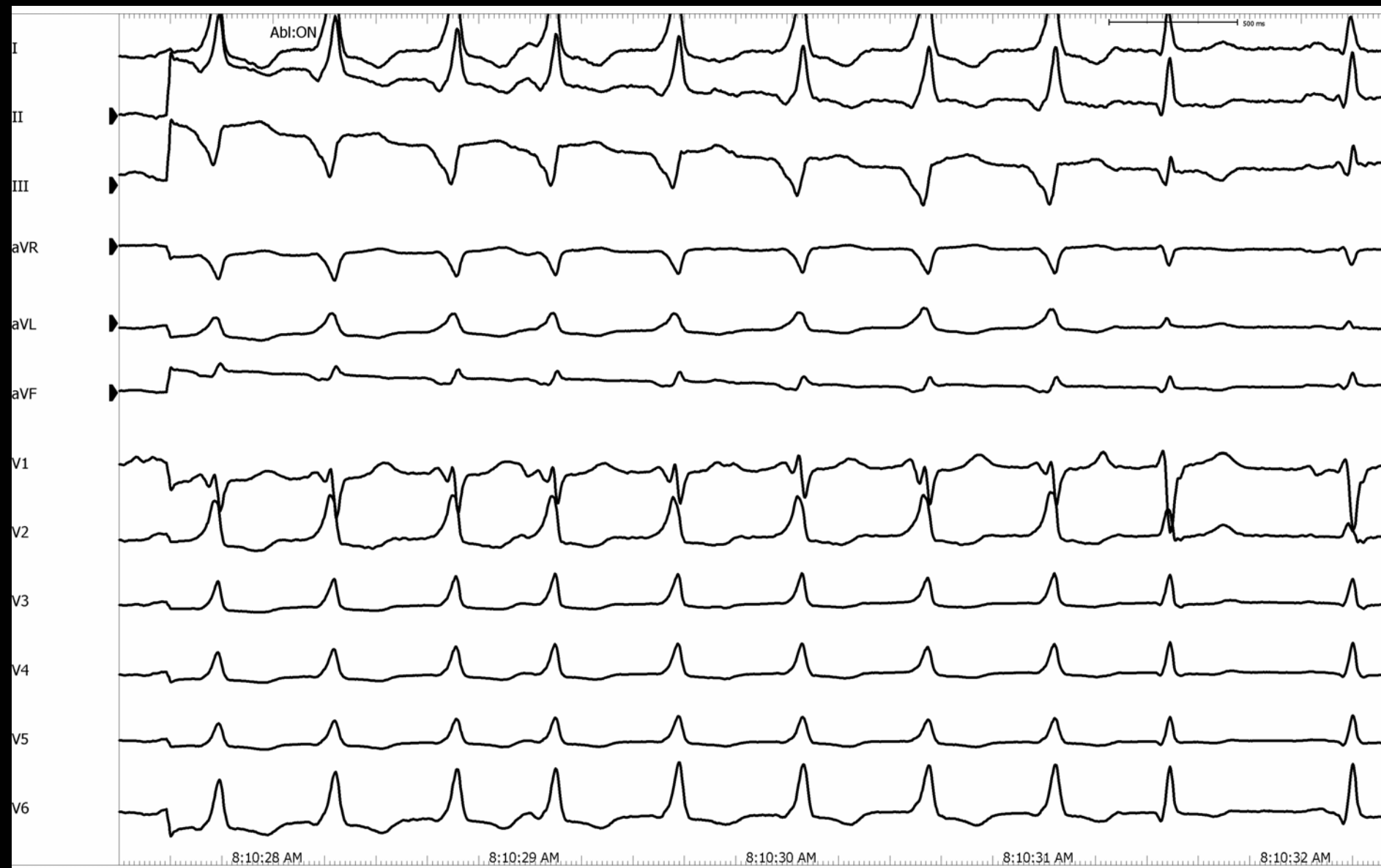
Is this a good site for ablation?



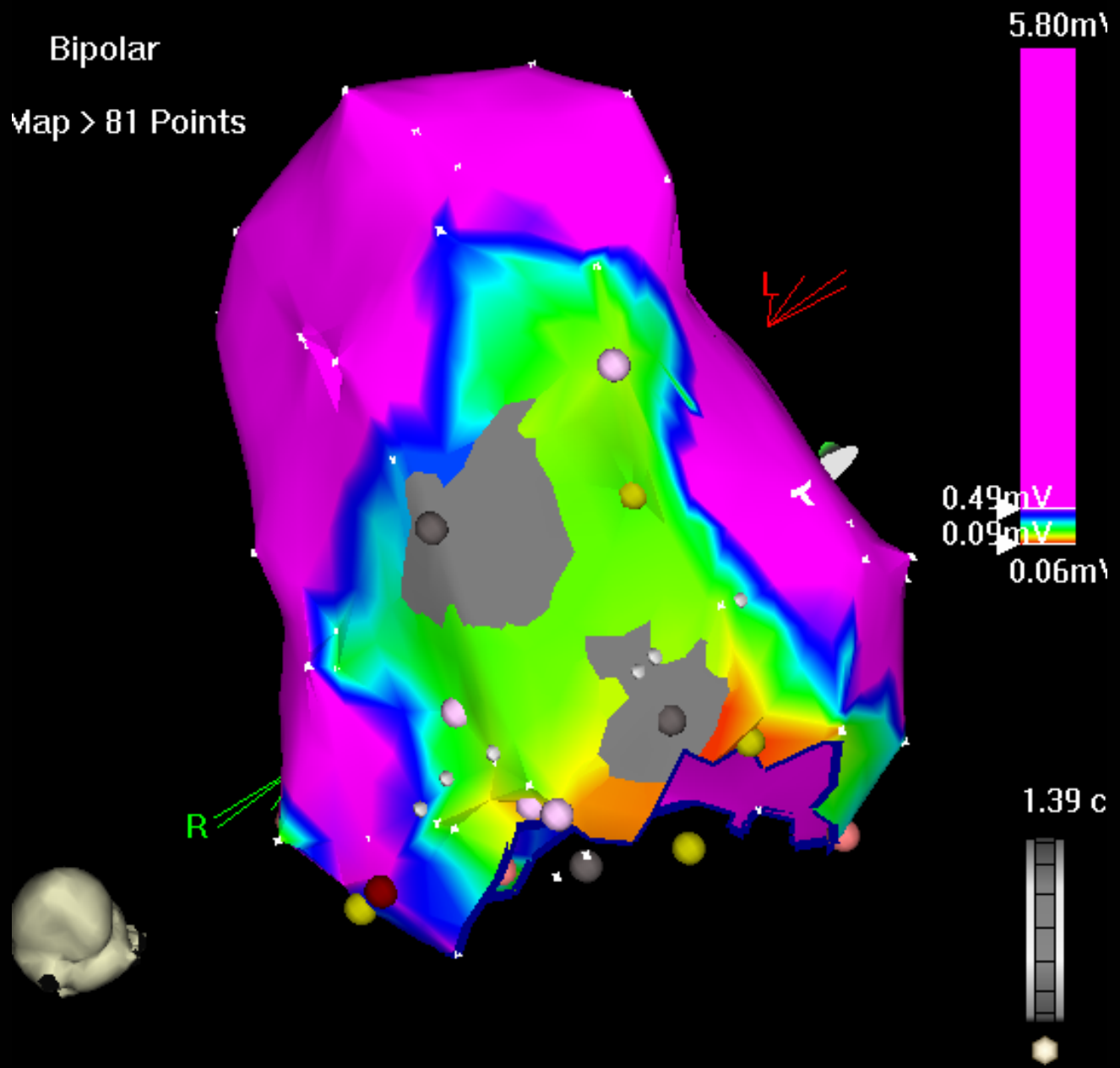
During ablation #3 near exit site

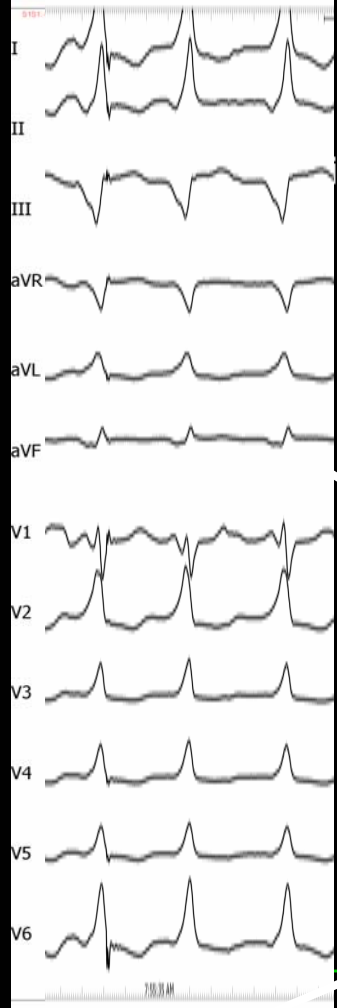


During ablation #3 near exit site

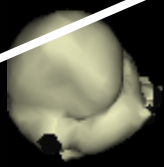
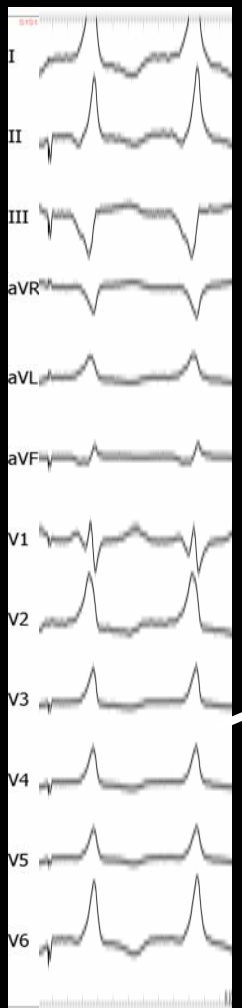
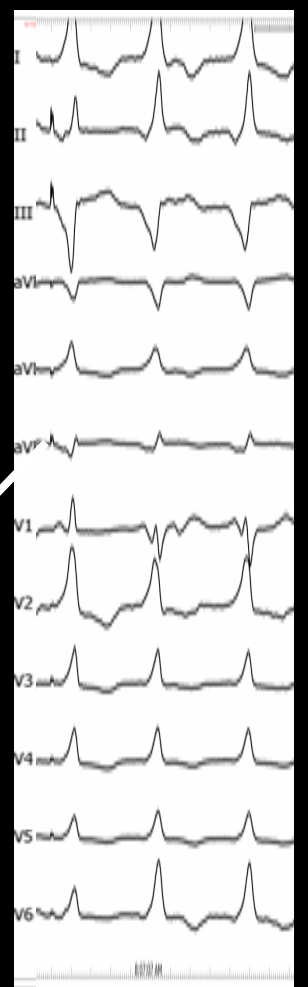
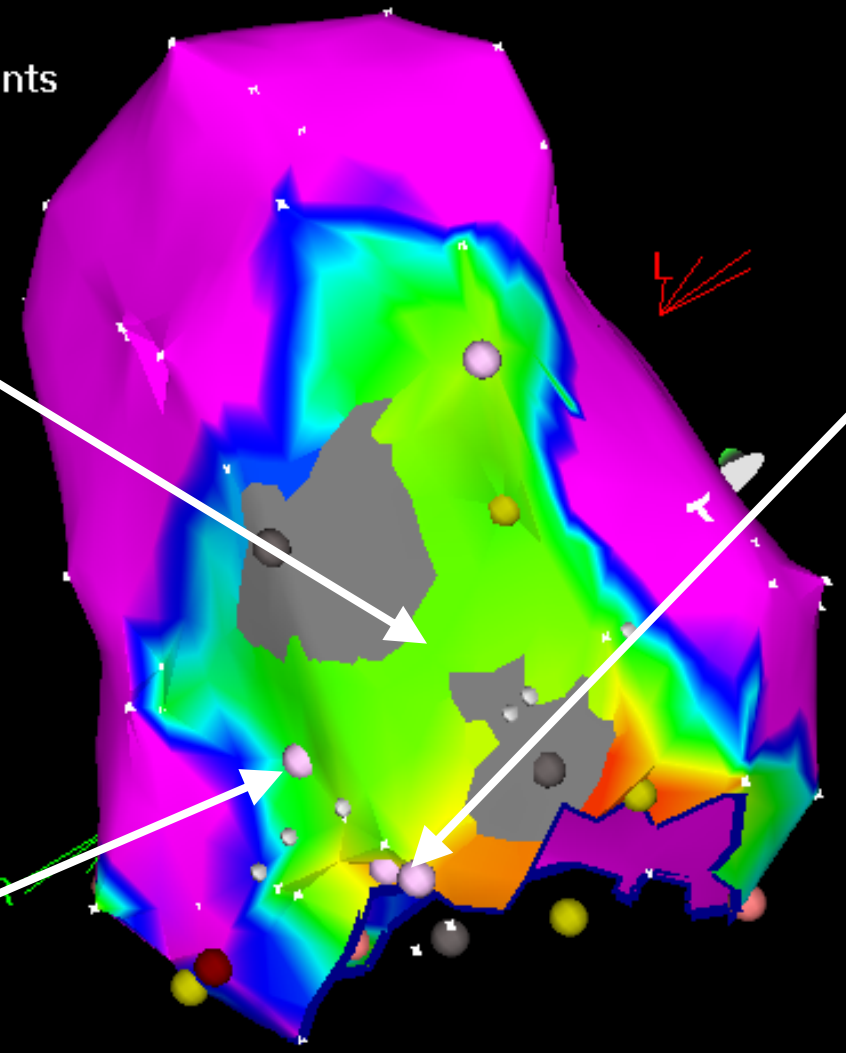


Bipolar
Map > 81 Points





nts

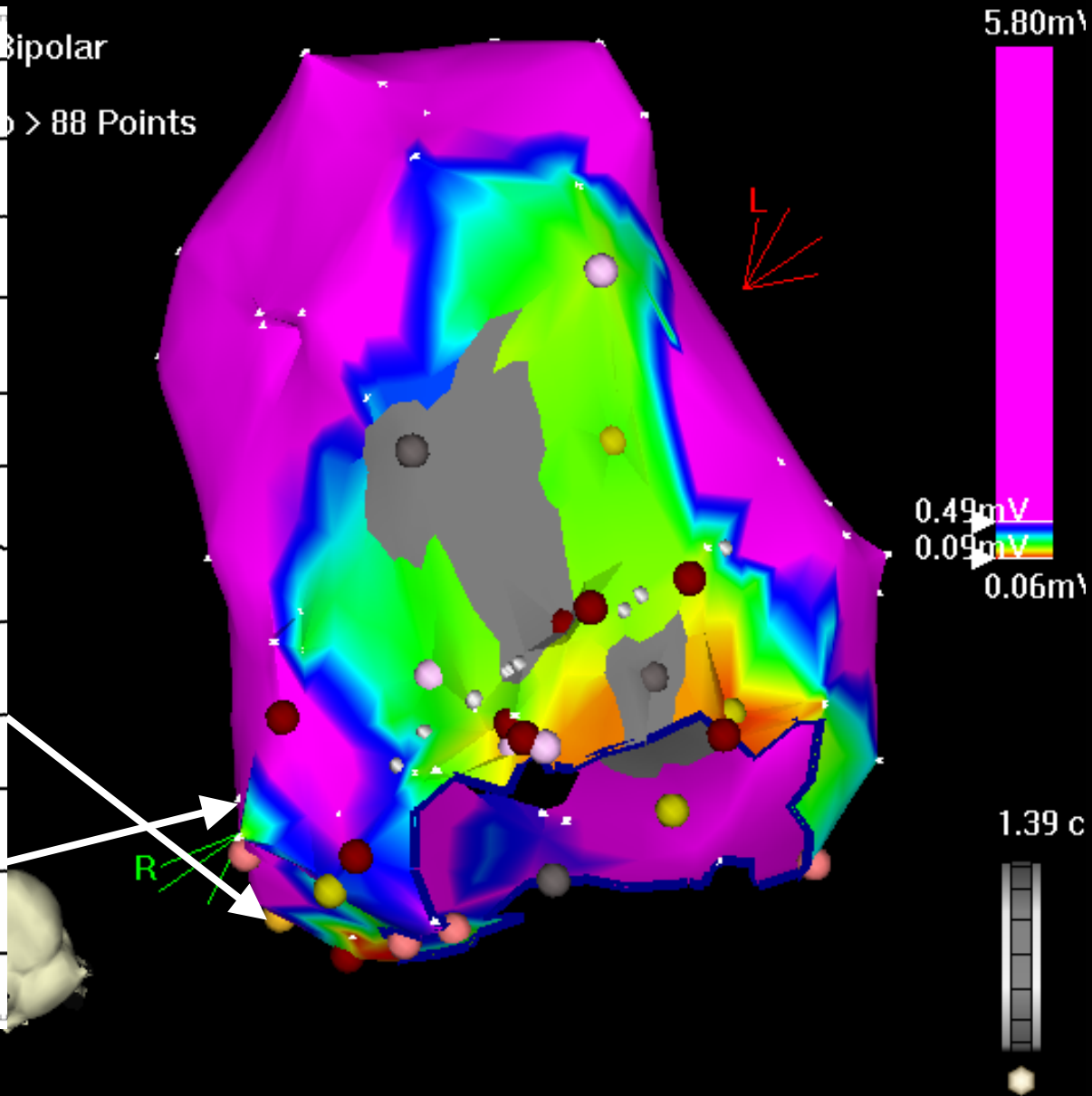




Bipolar

> 88 Points

8:41:07 AM



5.80mV

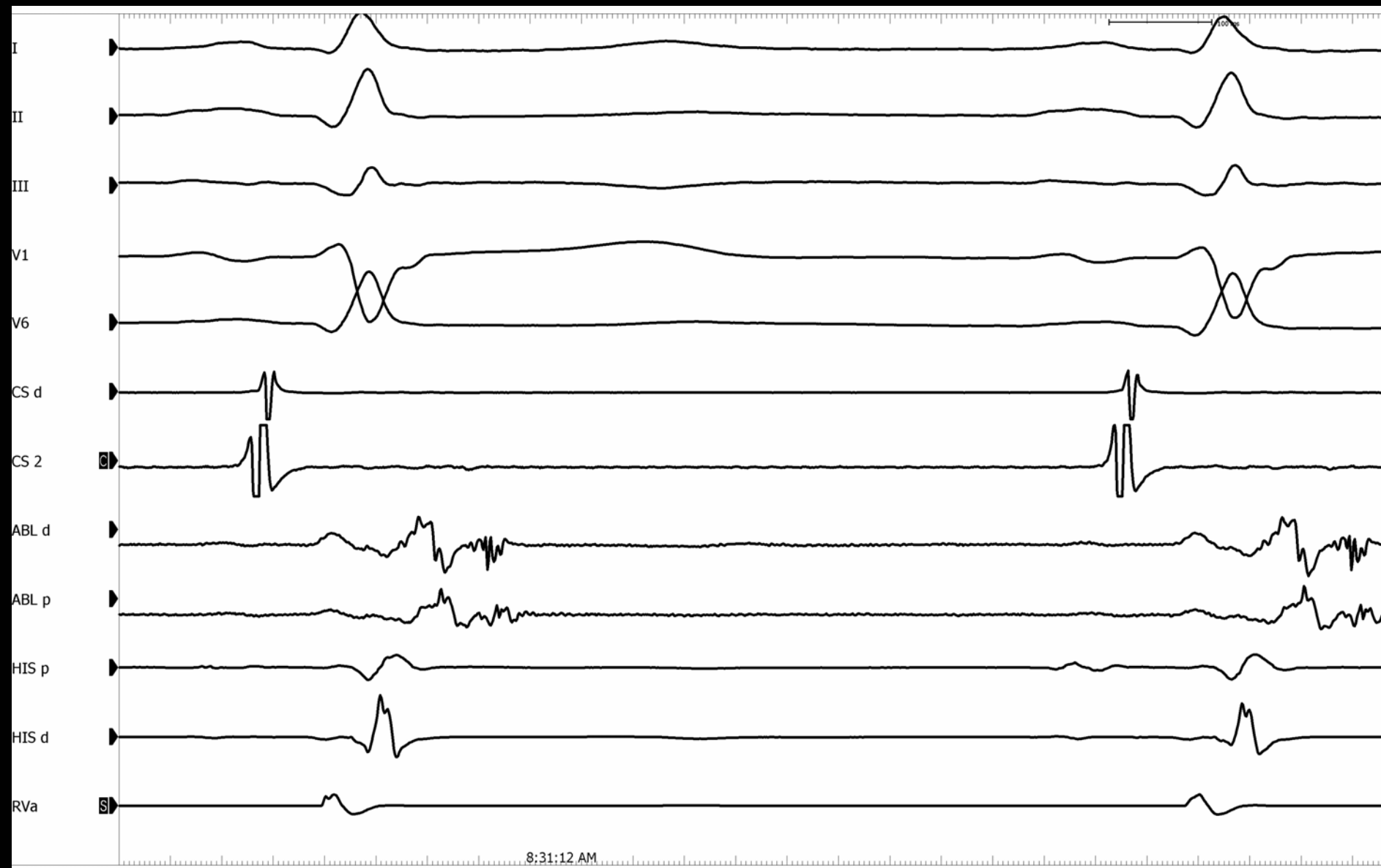
0.49mV

0.09mV

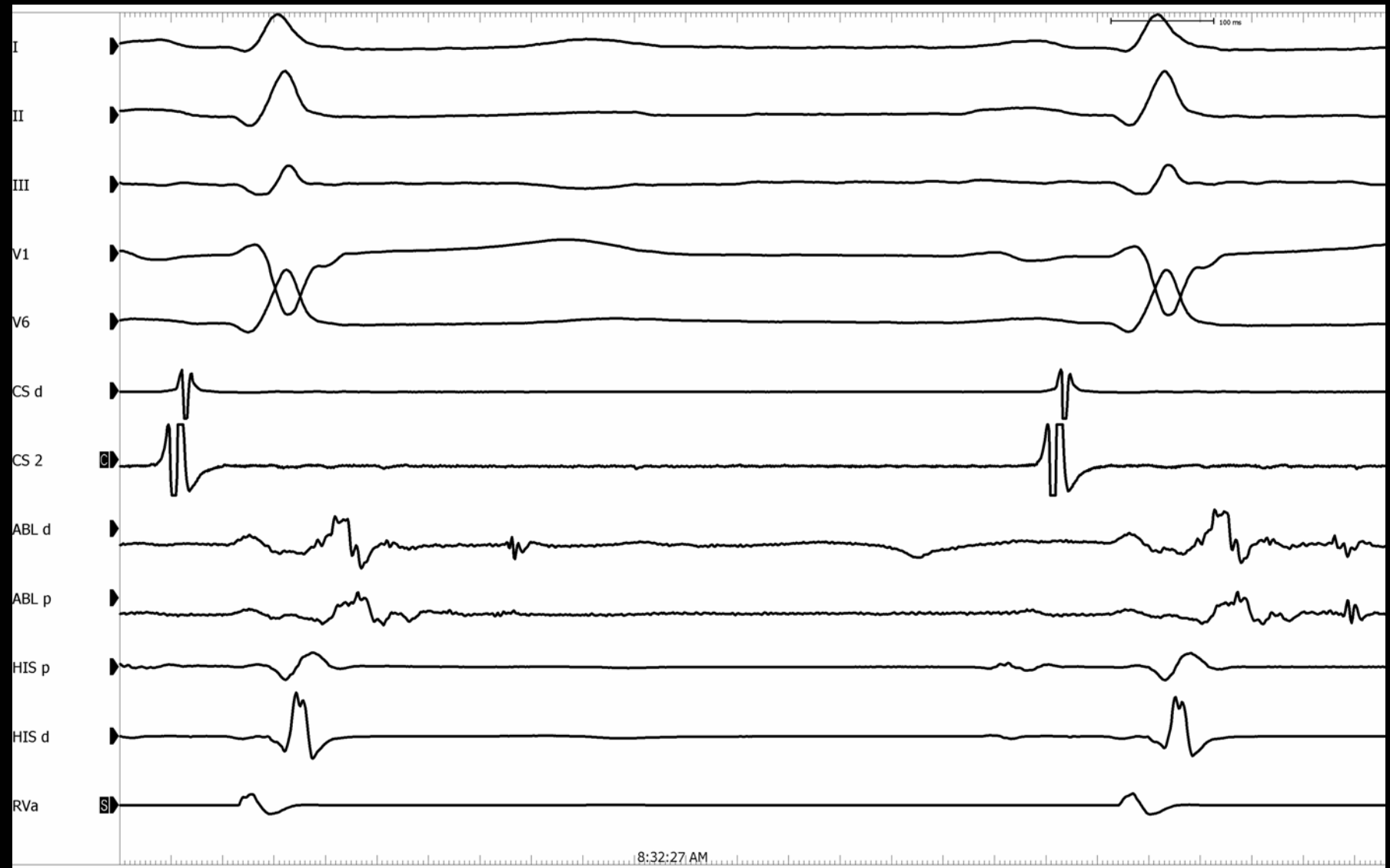
0.06mV

1.39 c

Ablation site #9 – diastolic potentials



During ablation #9 activation pattern of diastolic potentials change



Fusion?



Follow-up

- Echocardiogram performed after the EPS revealed
 - EF 25% with LV diastolic diameter 6.4cm
 - Akinetic inferior and inferolateral wall
 - Inferior aneurysm
- Patient offered ICD for prevention of SCD given low ejection fraction and history of MI
- Patient did not want ICD implant and was discharged after treatment of mild CHF and up-titration of beta blocker and ACE-I therapy

51 yo man with WCT

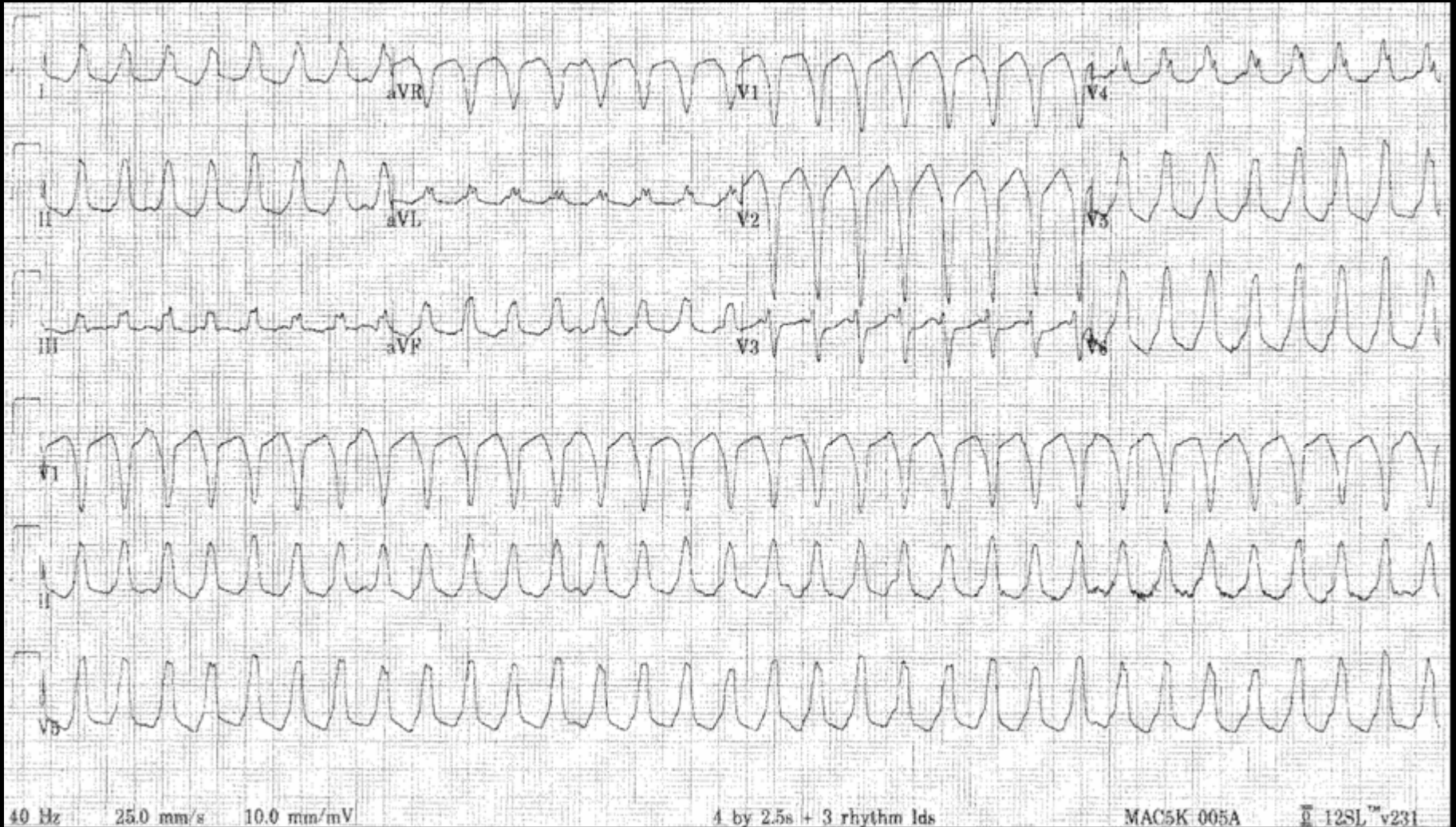
History

- 51 yo man with h/o HTN, no cardiac hx
- Several days of fatigue, malaise
- Worked up with stress test and echo, both unremarkable
- Developed sudden onset of fatigue, weakness, and presented to a local ED

Ancillary History

- Brother died suddenly at age 47, unknown cause
- Father developed heart failure age 53, died of a “heart attack”
- Social History
 - Rare alcohol consumption
 - No tobacco or drug use
 - Physically active

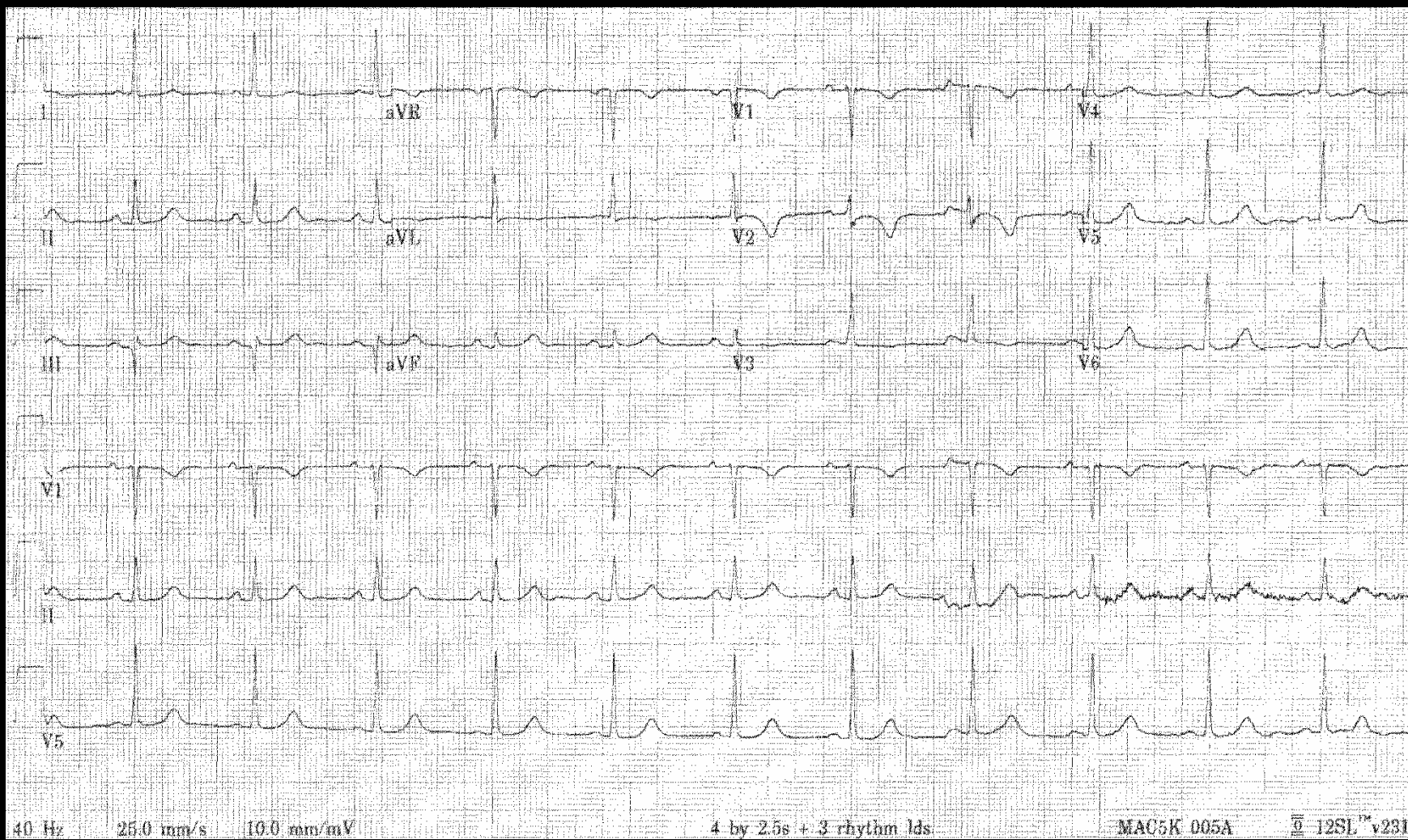
ECG on presentation



Course

- DC CV at 100J failed to convert from WCT
- Adenosine IV administered with no effect
- DCCV at 100J converts to SR

Baseline ECG



Course

- Transferred to BIDMC for further evaluation and EPS
- SAECG done prior to EPS

Analysis Filter : 40-250Hz

Std. QRS Duration (unfiltered) : 98 ms

Total QRS Duration (filtered) : 127 ms

Duration Of HFLA signals < 40 μ V : 51 ms

RMS Voltage in terminal 40 ms : 7 μ V

Mean Voltage in terminal 40 ms : 5 μ V

Number Of Beats Averaged: 94

Number Of Beats Detected: 97

Noise Level (Std. Devn.) : 0.30 μ V

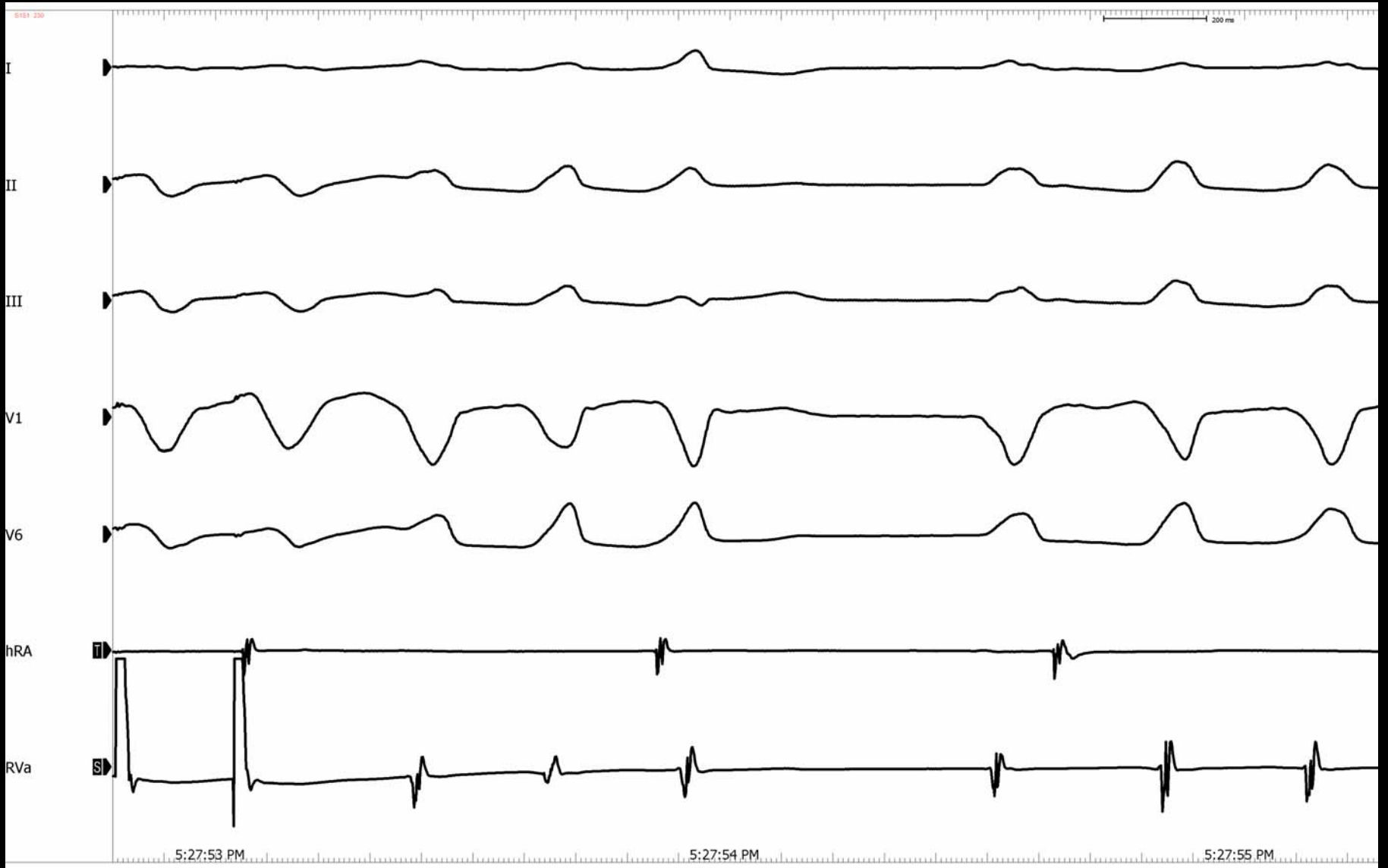
Referred by:

ALTIMETER RMS VOLTAGE 5 400V

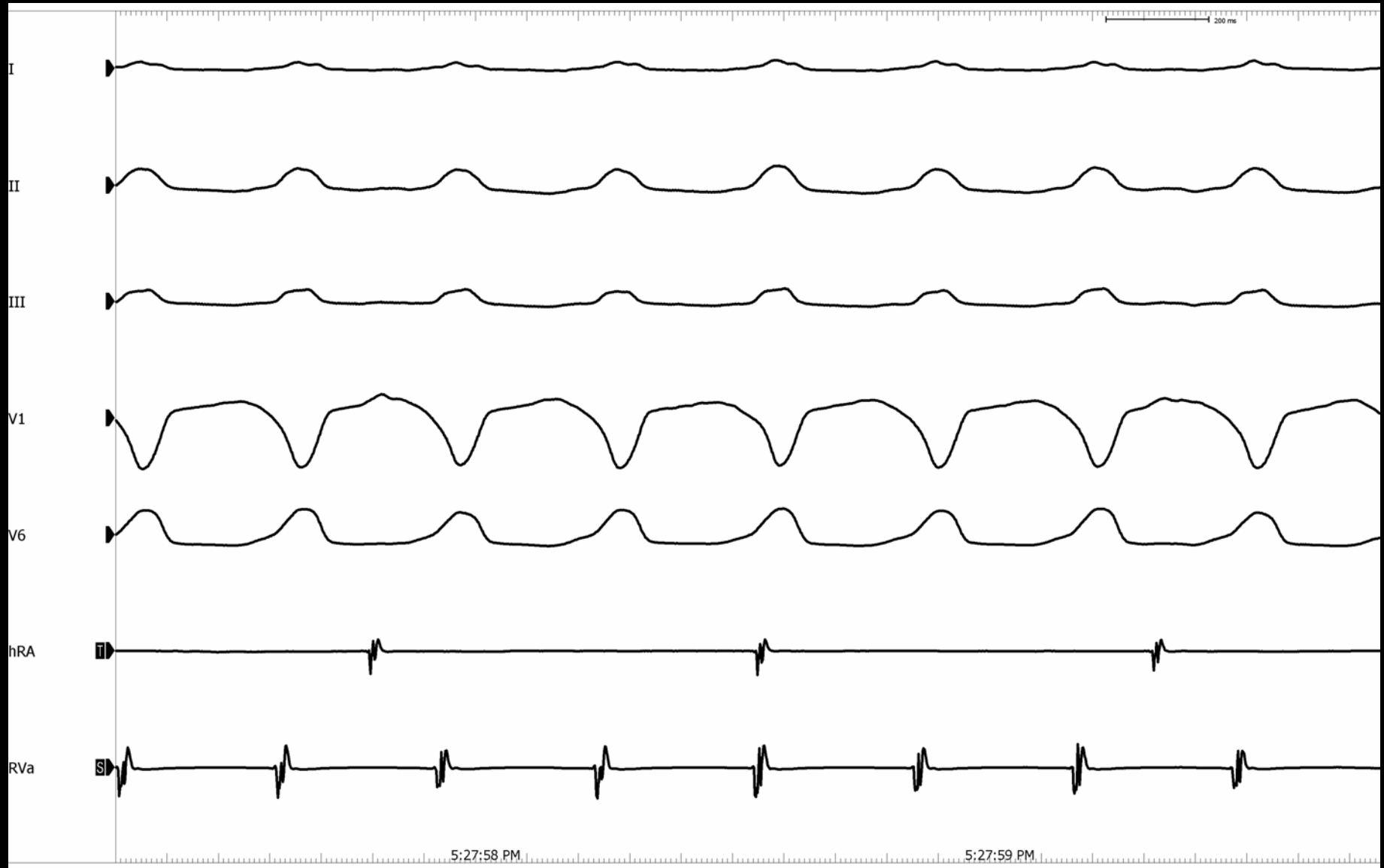
Circuit 2000



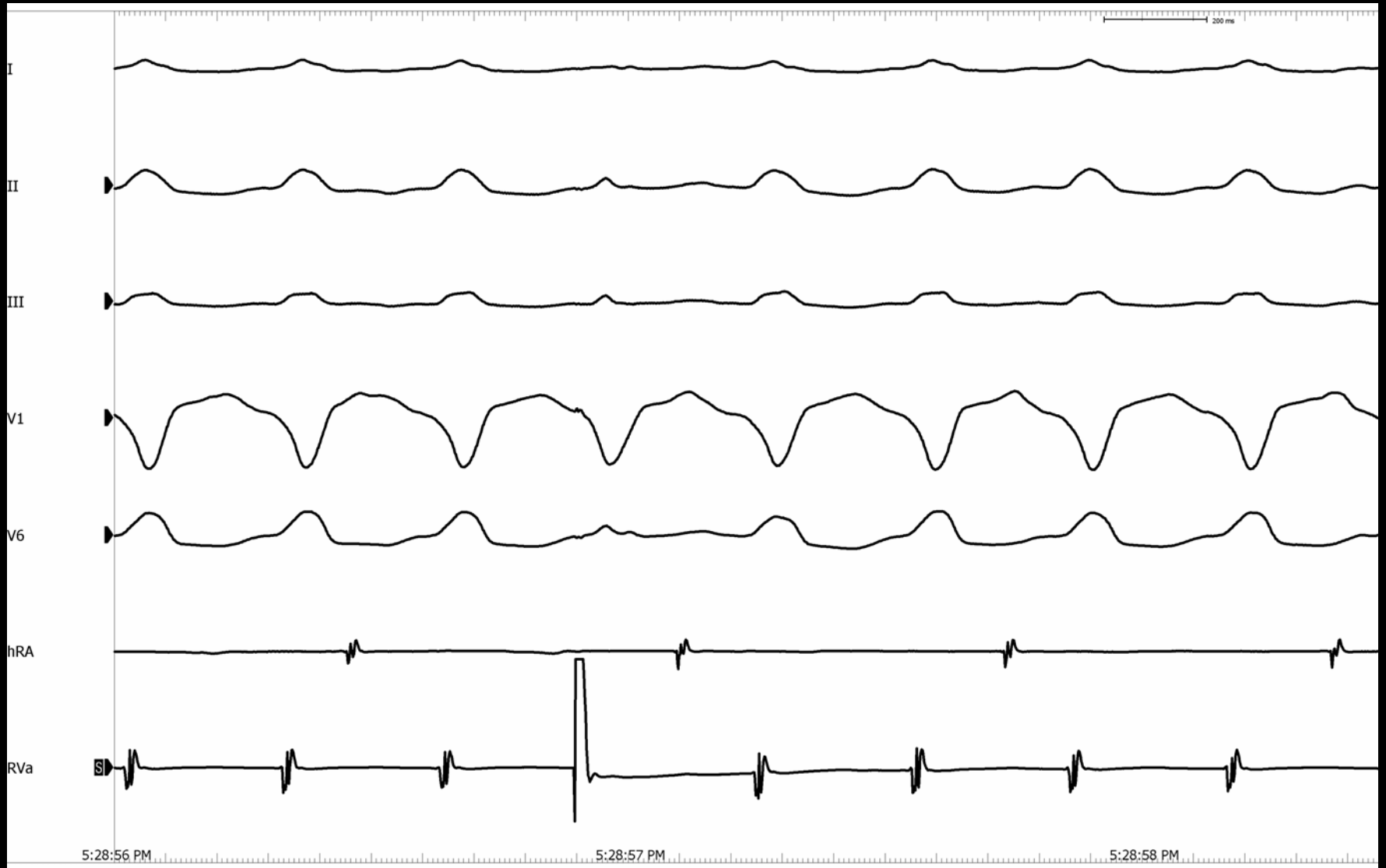
Induction 500-28--230



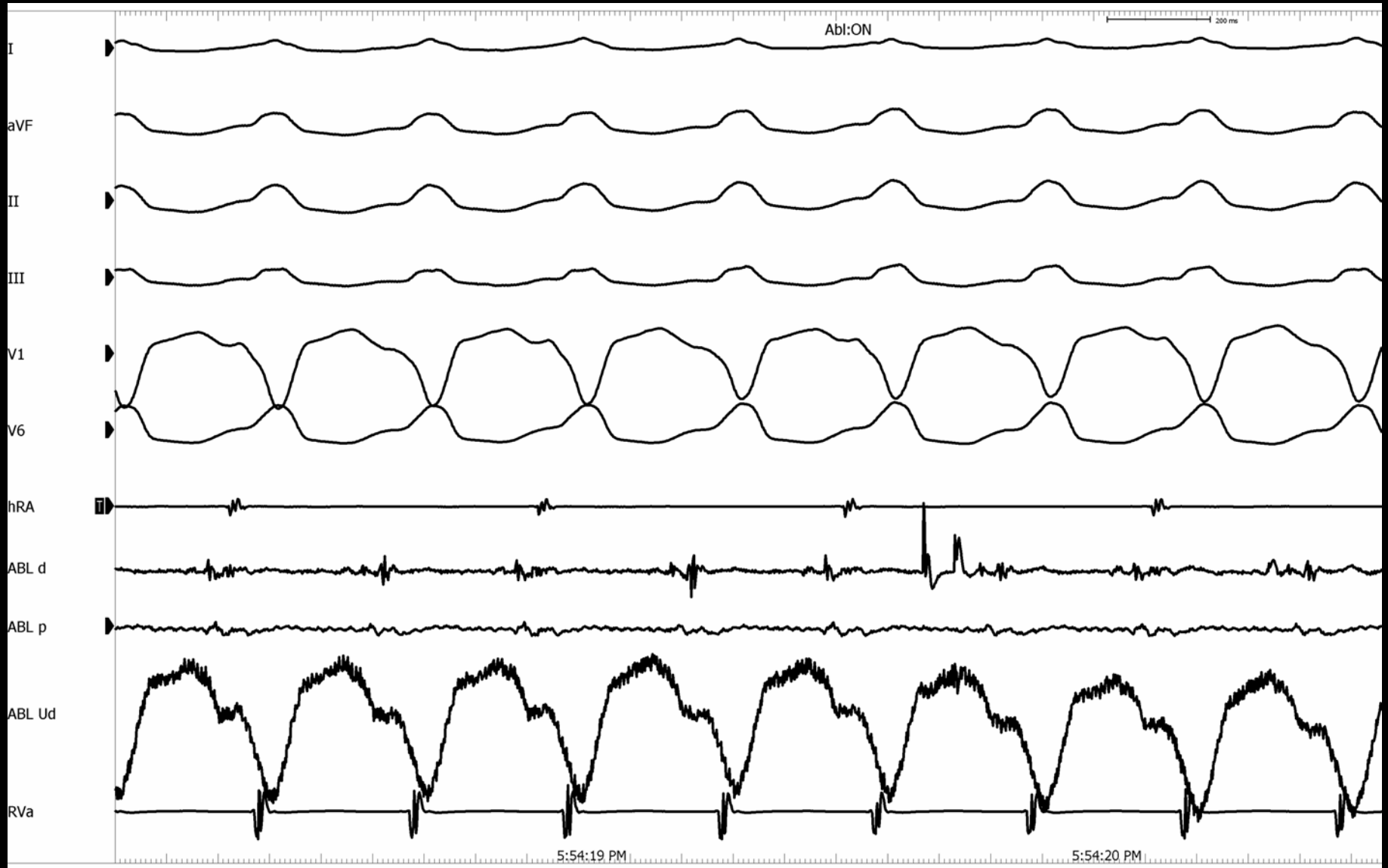
VT CL 305



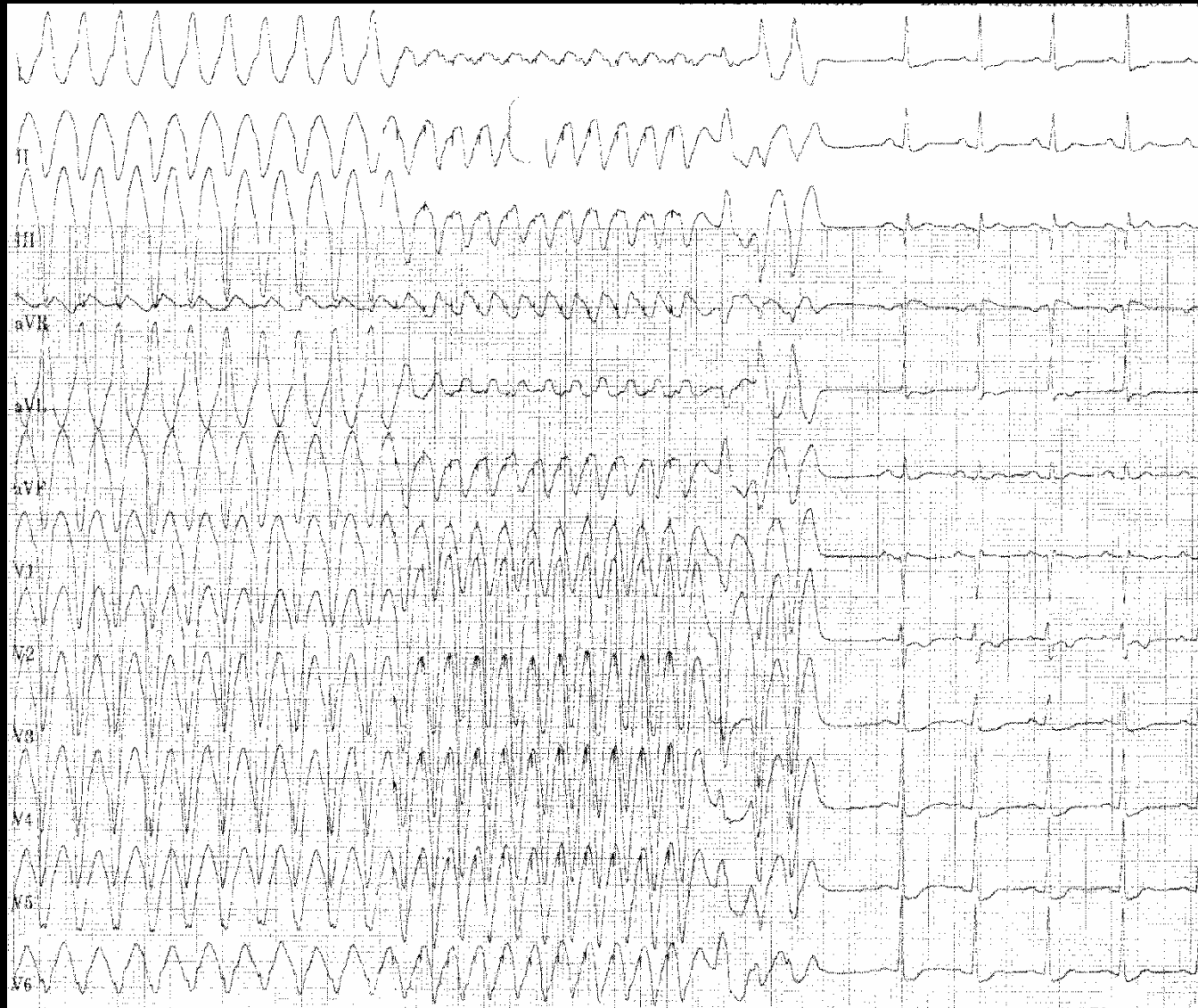
What does this prove?



Ablation in lateral RVOT terminates VT

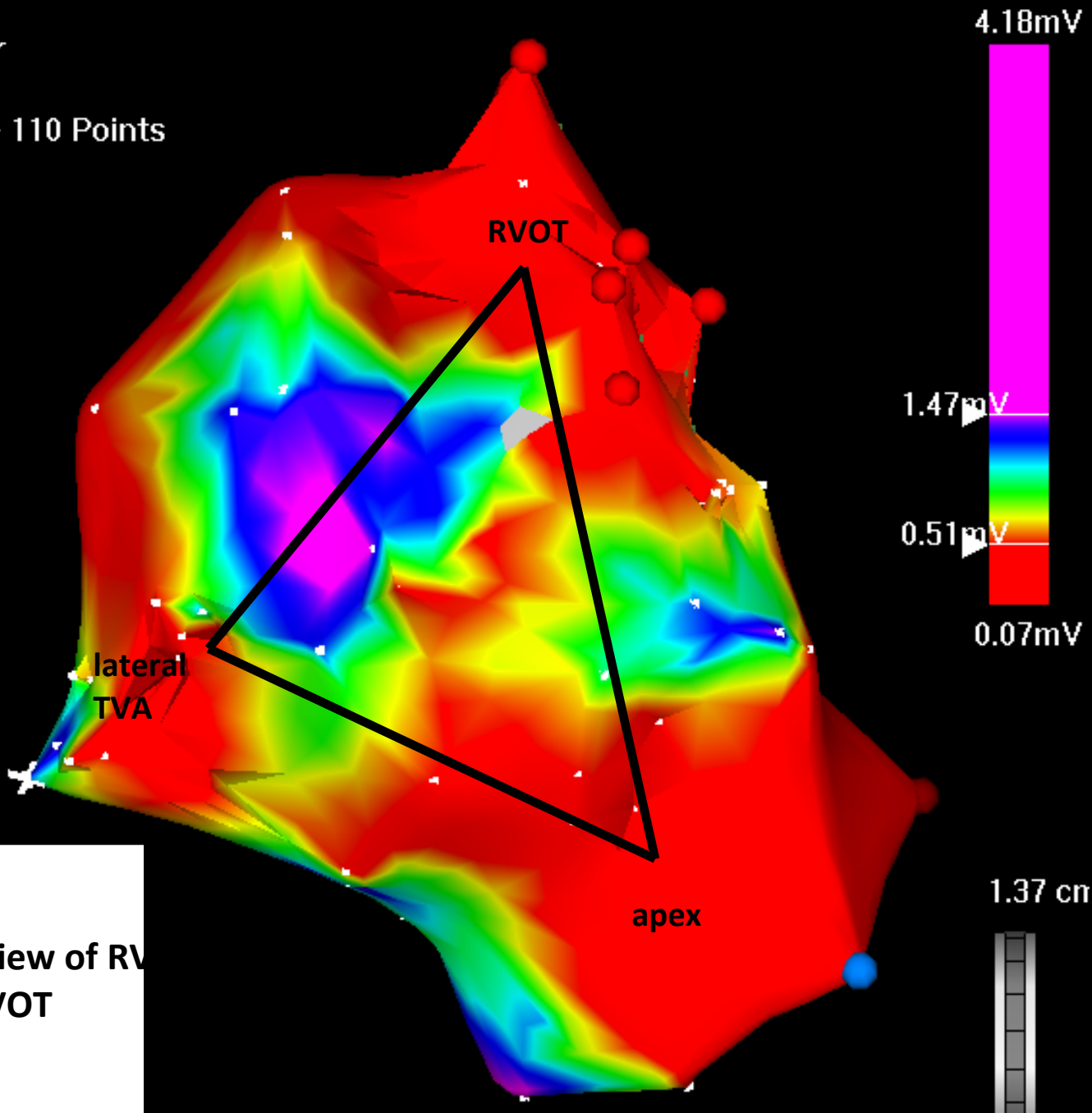


VT still inducible, pacing induces second VT morphology which breaks with RV pacing



Bipolar

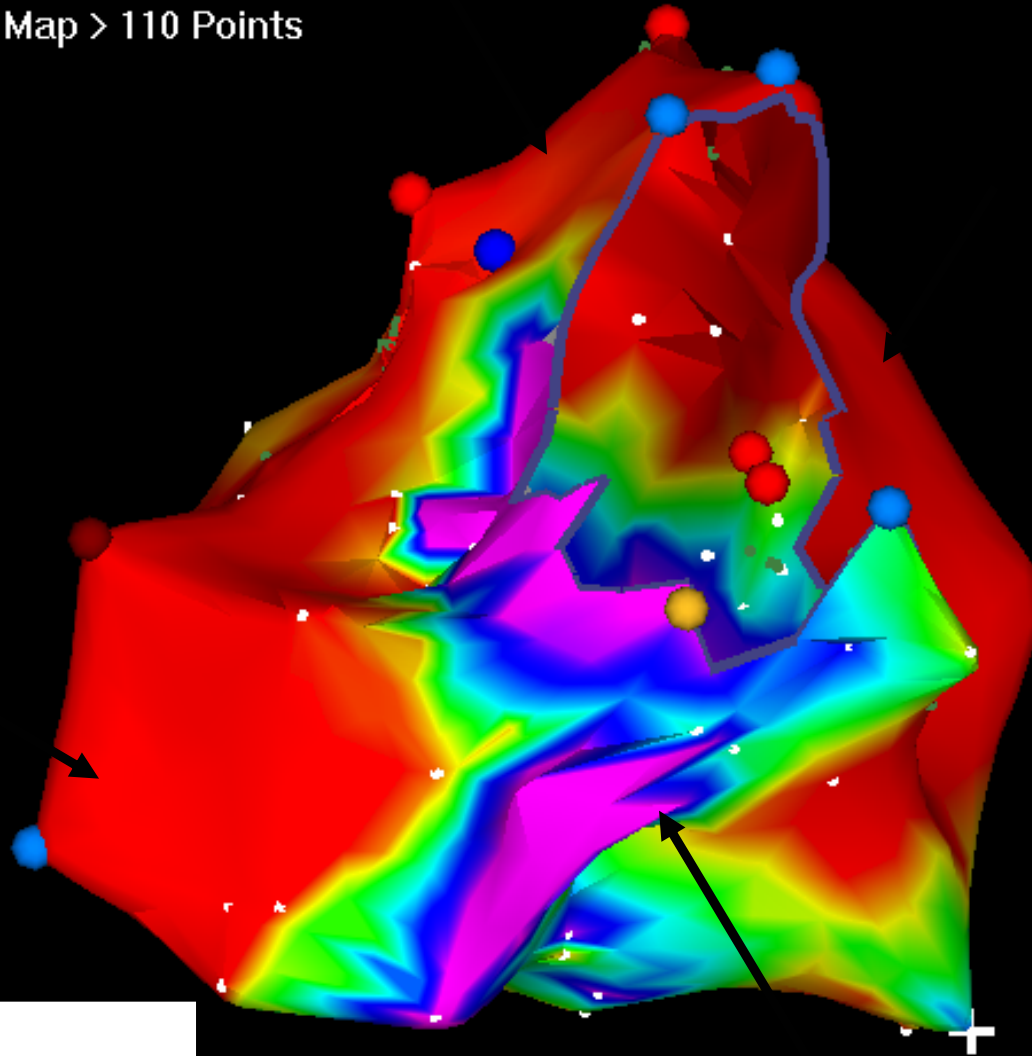
1-1-ReMap > 110 Points



Modified RAO view of RV
free wall and RVOT

Bipolar

1-1-ReMap > 110 Points



4.18mV

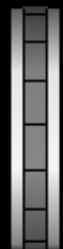
1.47mV

0.51mV

0.07mV

1.37 c

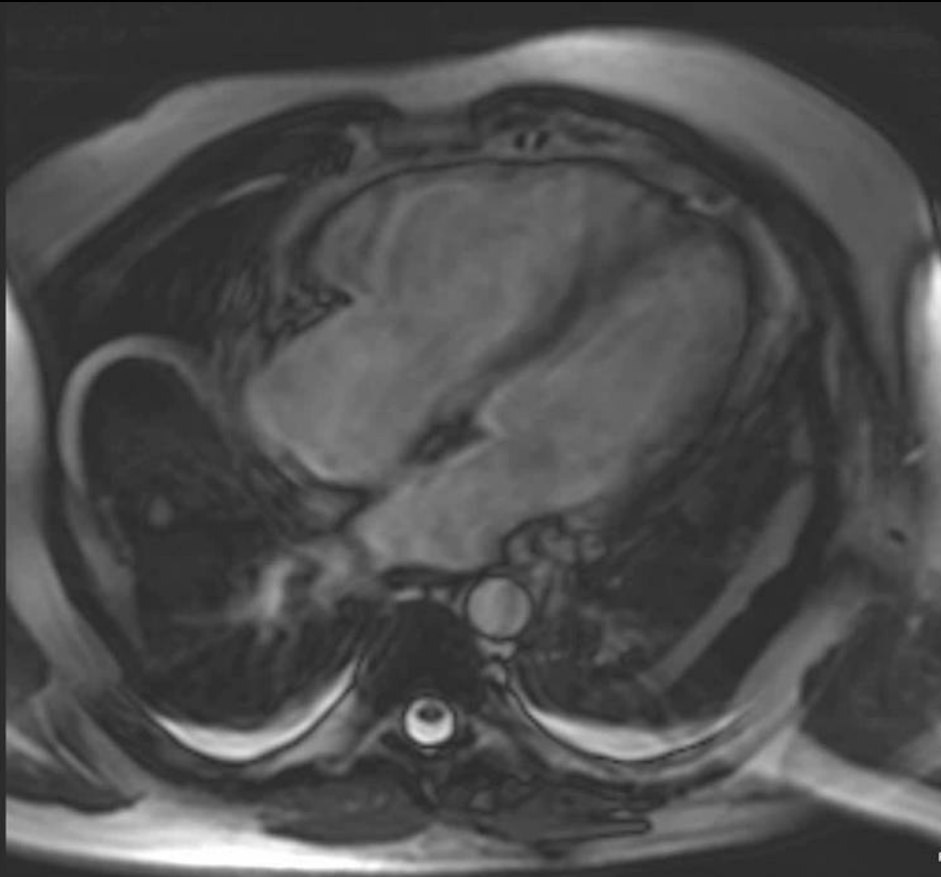
Septal view of tricuspid
and RV



RV Substrate Map

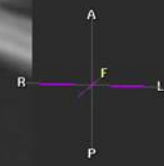
- Diffuse patchy scar
- Scar most dense at RV apex, basal free wall, and RVOT
- “Triangle of dysplasia”

Sc 4.1/1
B-TFE / FFE/M
Td 000 ms

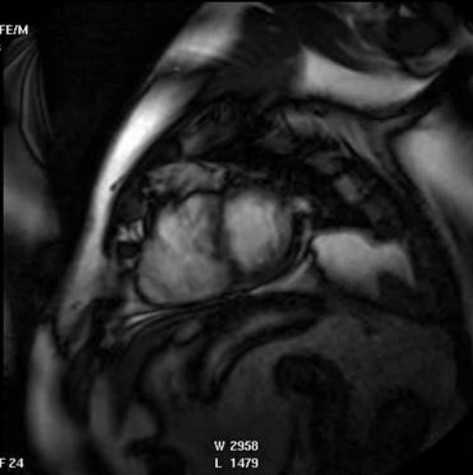


AP 11°
RL 10°
FH -2°
A 20 L 34 F 4

W 2000
L 651



Sc 5.1/4
B-TFE / FFEM
Td 000 ms



AP 21°
FH 42°
A 77 L 89 F 24

W 2958
L 1479



Sc 5.1/5
B-TFE / FFEM
Td 000 ms

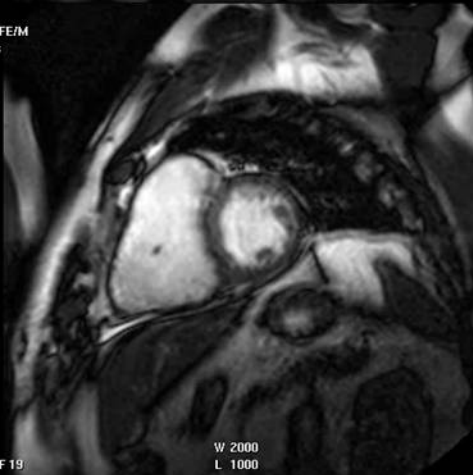


AP 21°
FH 42°
A 70 L 82 F 21

W 2077
L 1038



Sc 5.1/6
B-TFE / FFEM
Td 000 ms

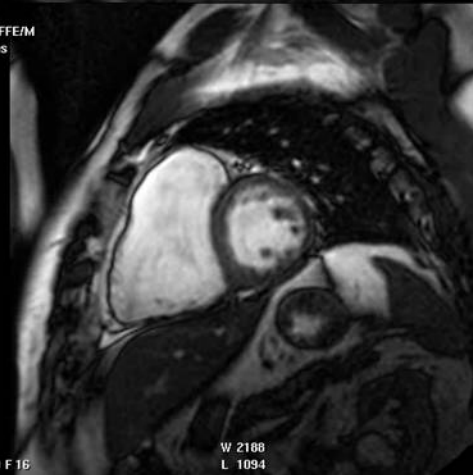


AP 21°
FH 42°
A 62 L 76 F 19

W 2000
L 1000



Sc 5.1/7
B-TFE / FFEM
Td 000 ms



AP 21°
FH 42°
A 55 L 70 F 15

W 2188
L 1094



CMR

- LVEF was normal at 64%.
- Dilated right ventricle with severely depressed systolic function and thinning of the free wall.
- The RVEF was severely decreased at 10%.
- No MR evidence of right ventricular fatty infiltration/dysplasia.

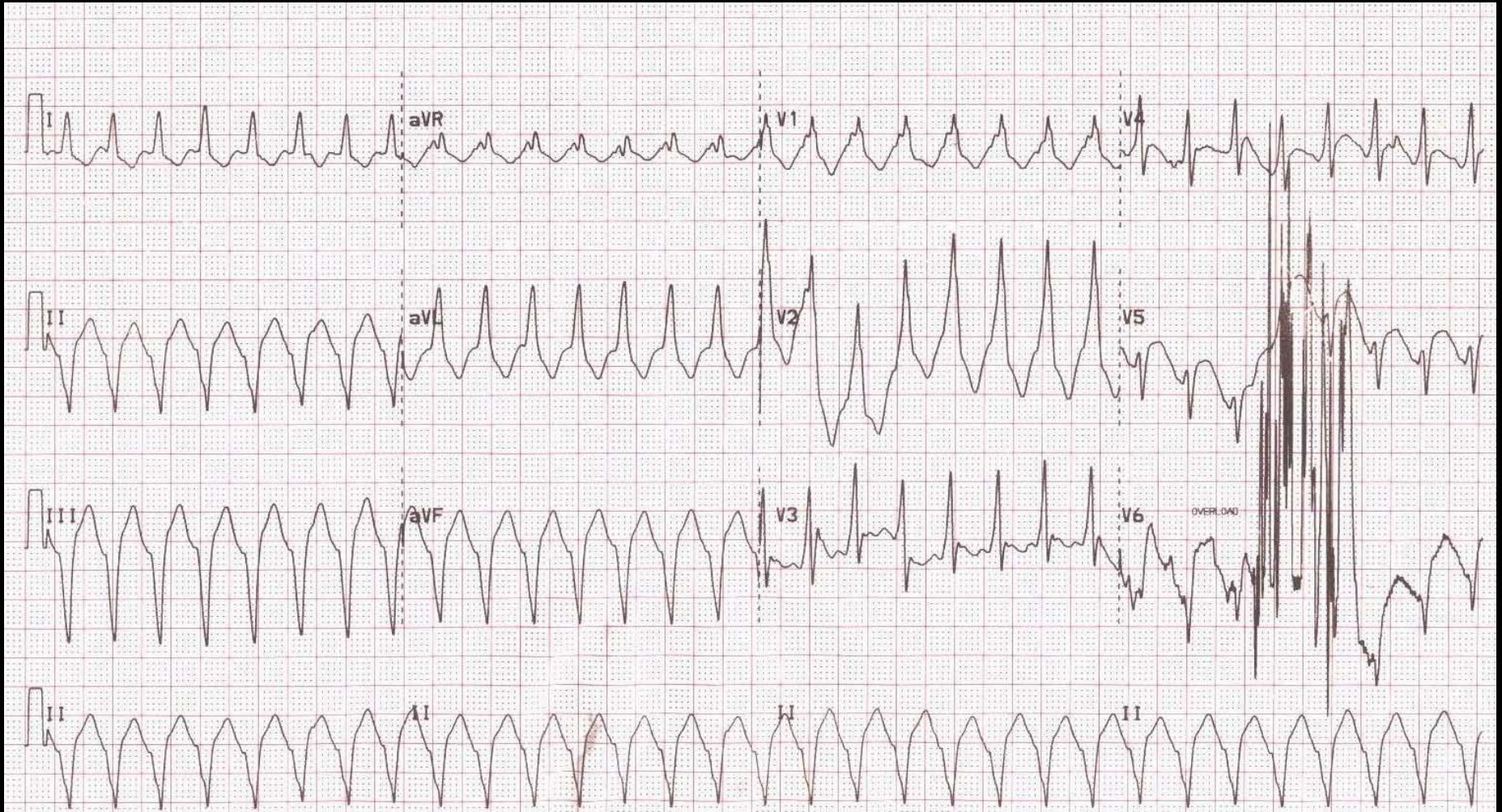
Subsequent course

- After EPS, underwent implantation of a dual-chamber ICD
- Started on sotalol 120 mg bid for suppression of VT
- 3 months later developed dizziness and subsequent ICD shock
 - Interrogation revealed VT at 210bpm and appropriate shock with conversion to SR
 - Mexilitine added to sotalol for suppression of VT
 - NIPS performed with inducible VT (required aggressive stimulation) and able to be terminated with anti-tachycardia pacing
- Family was screened through Children's Hospital with cardiac MRIs – results unavailable

WCT

Presentation

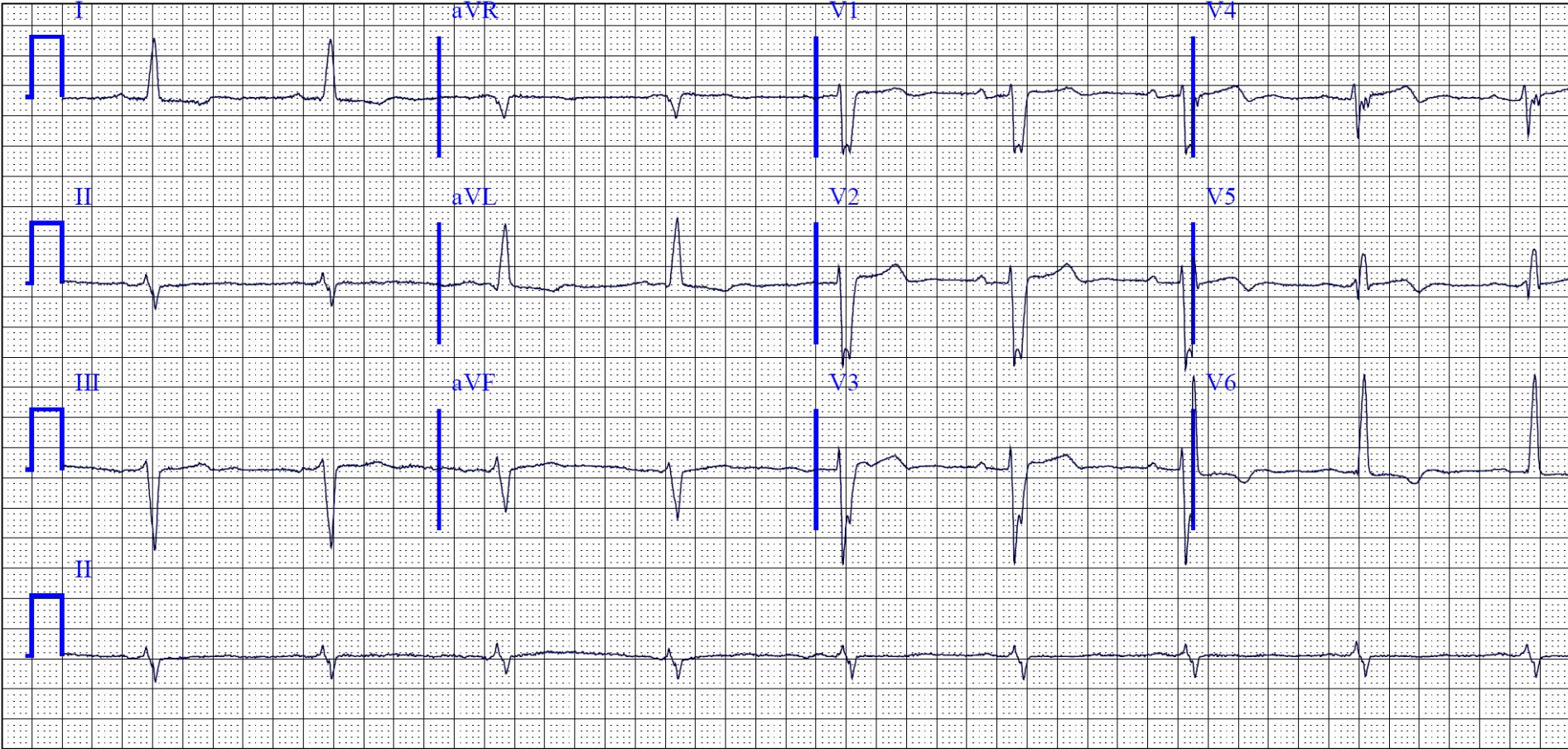
- 67M admitted with L>R lower extremity claudication for an elective angiogram
- Lying in bed he felt palpitations and lightheaded
- The nurse found him with a HR in 180s and systolic BP in 60s



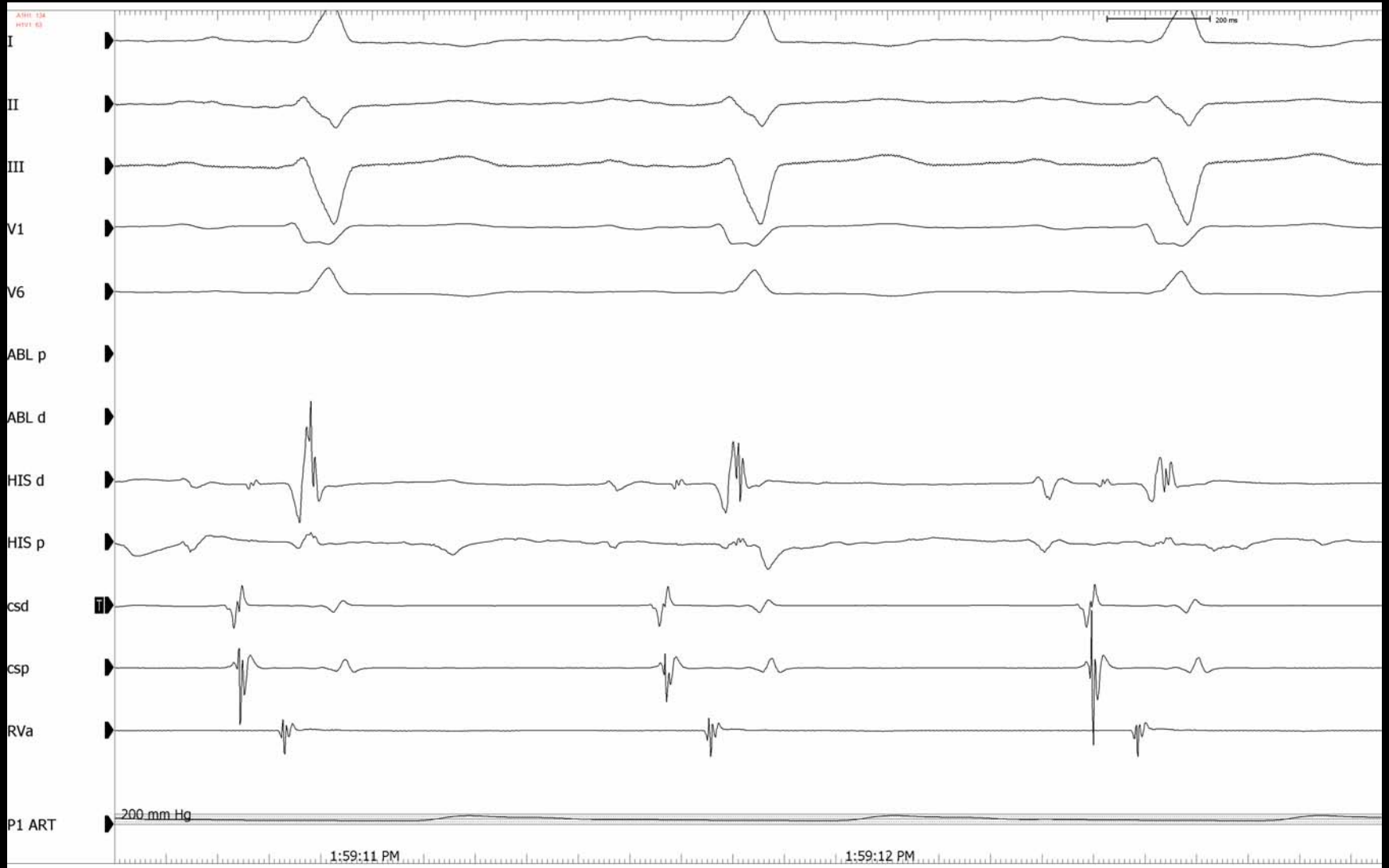
Past History

- Hypertension
- Tobacco
- Hyperlipidemia
- CAD with prior MI, prior CABG, and depressed LVEF (23% on 01/31/02, nuclear)
- PAD (Rutherford class II-III, prior RLE stenting)
- COPD (not on home oxygen, unknown FEV1)

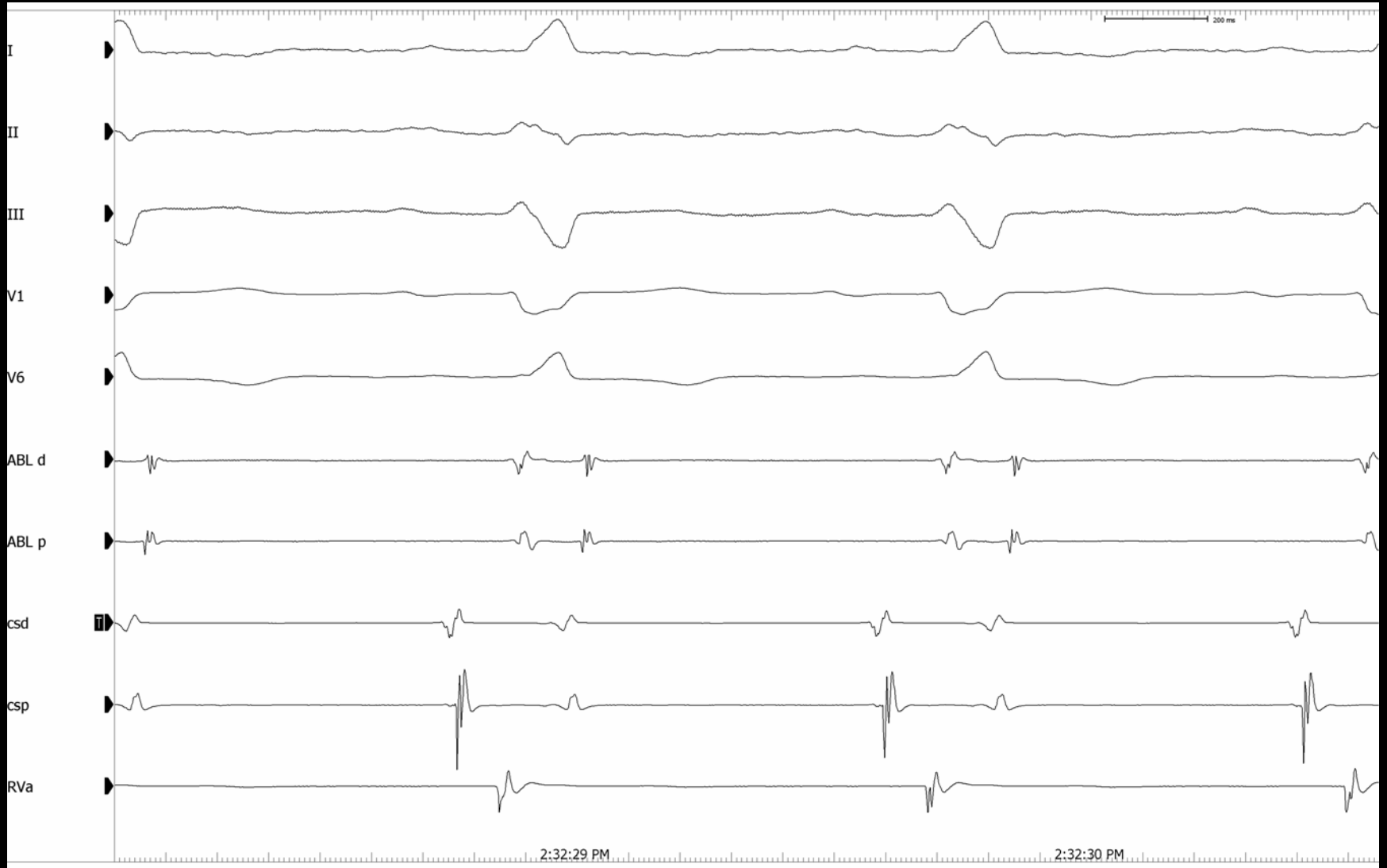
Baseline



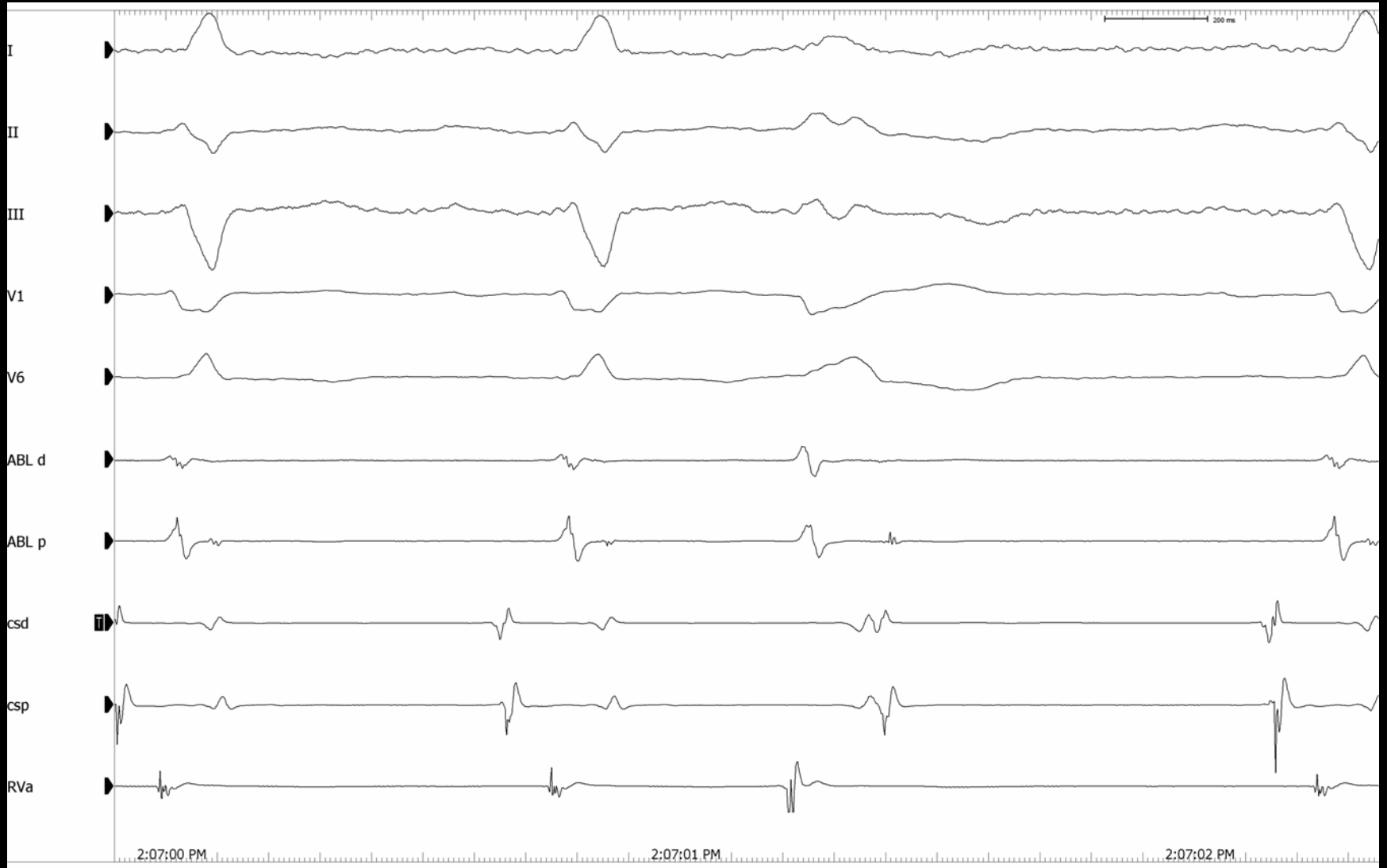
Baseline 800/135/63



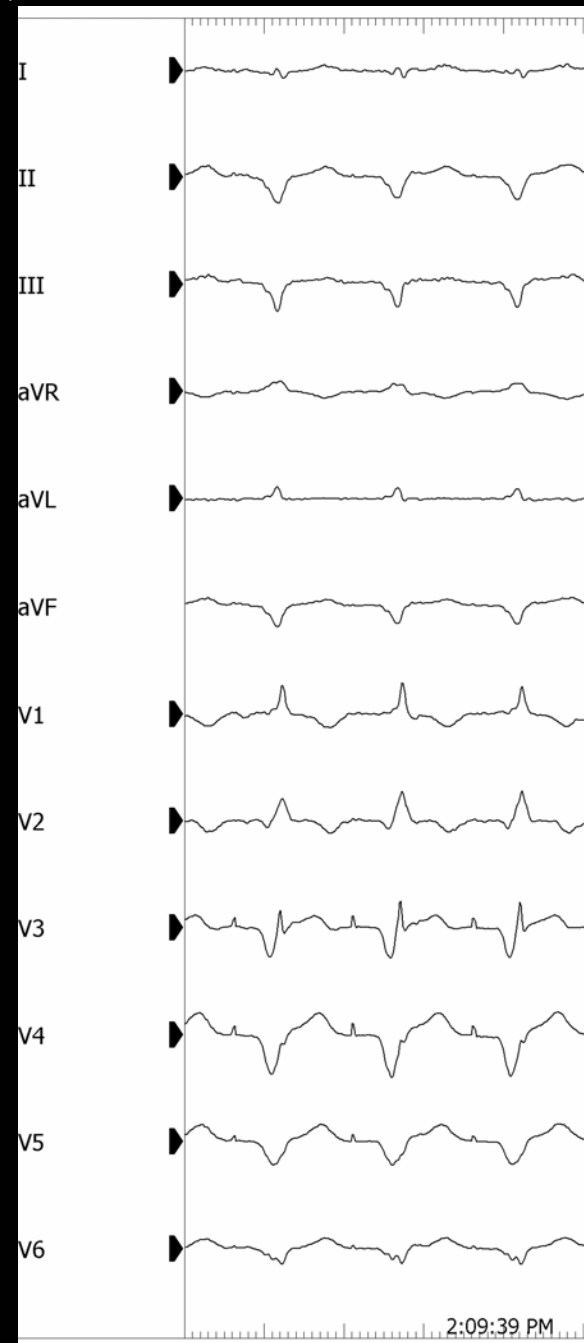
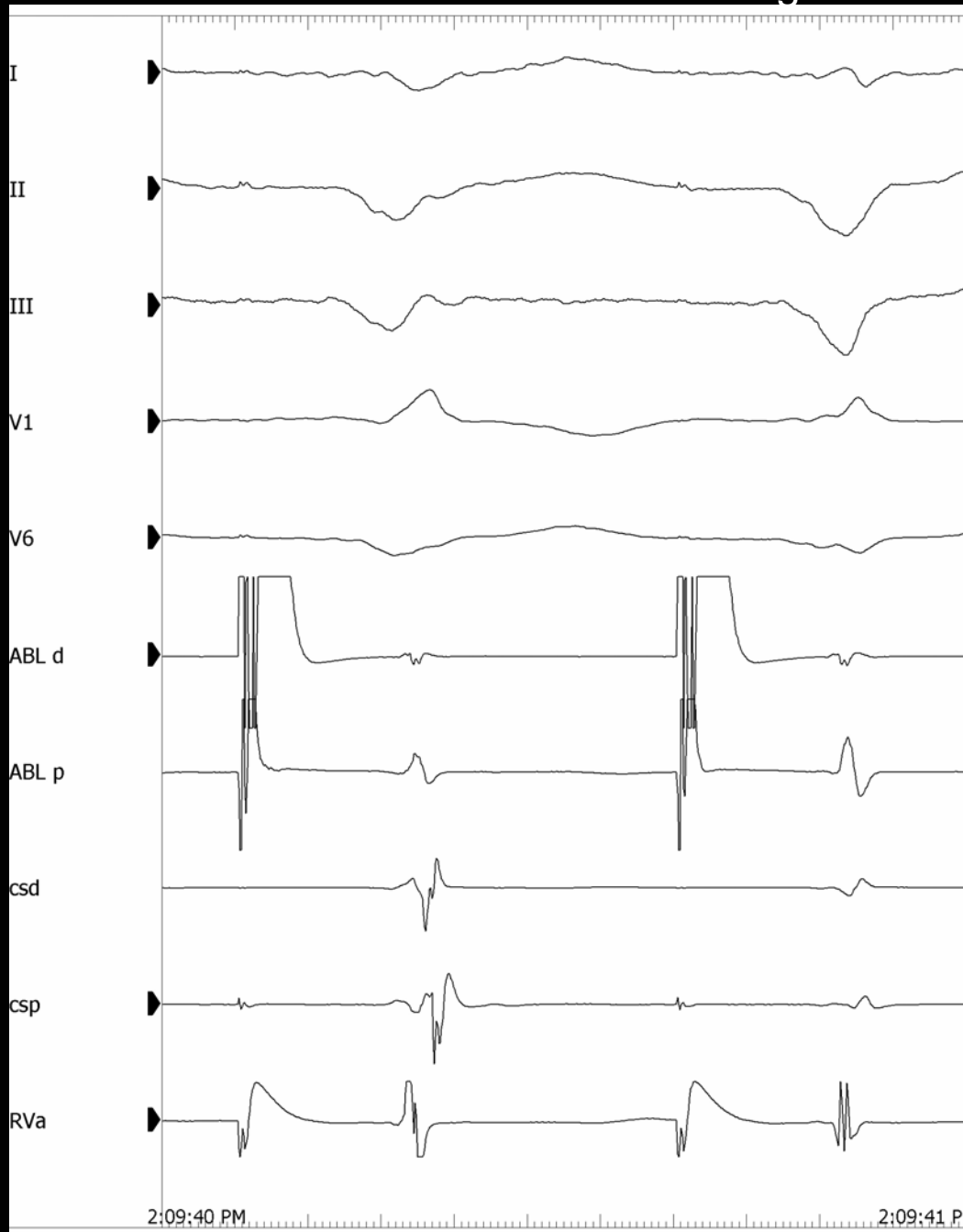
Late Potentials: Sinus Rhythm

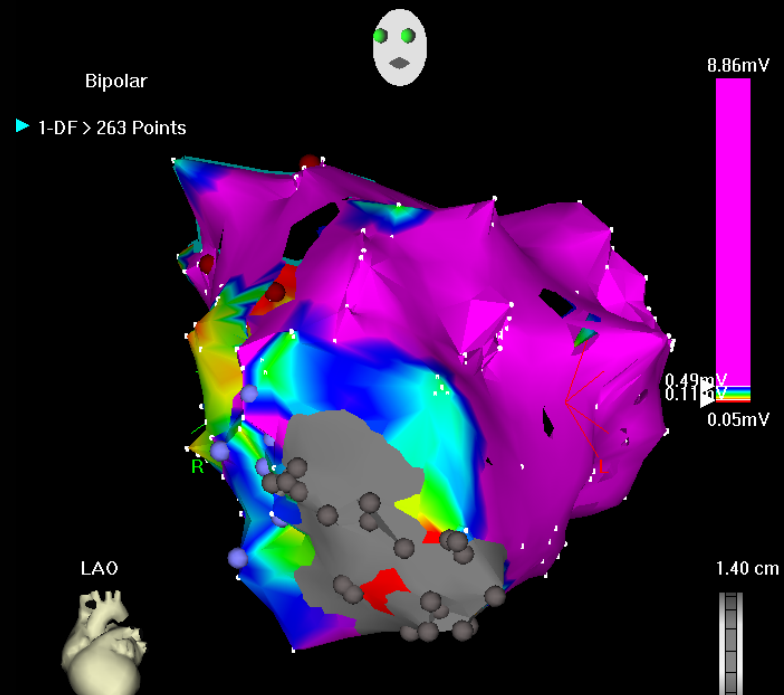
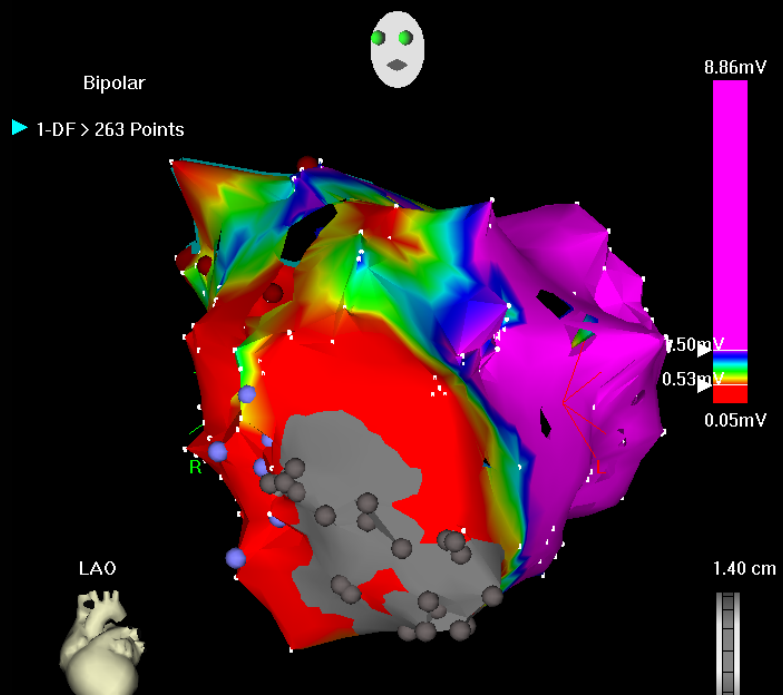
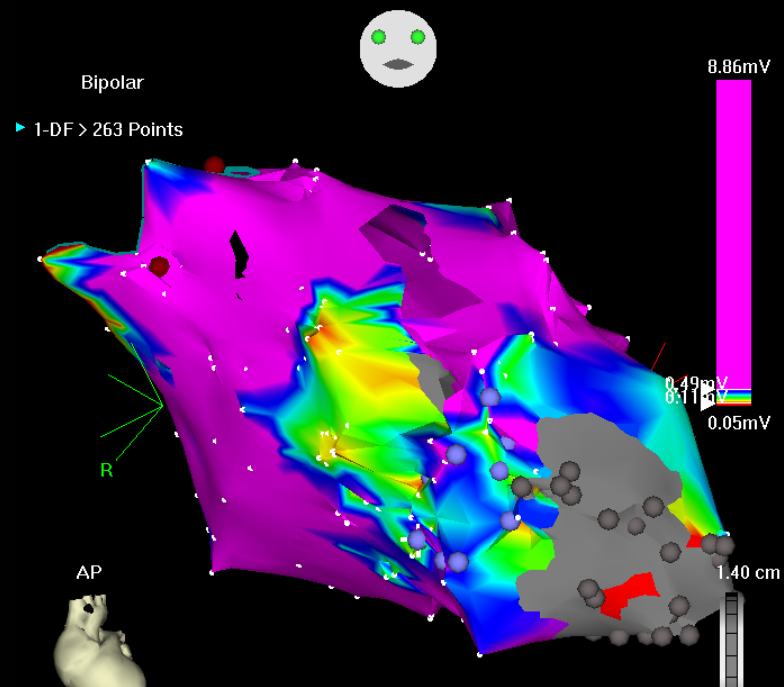
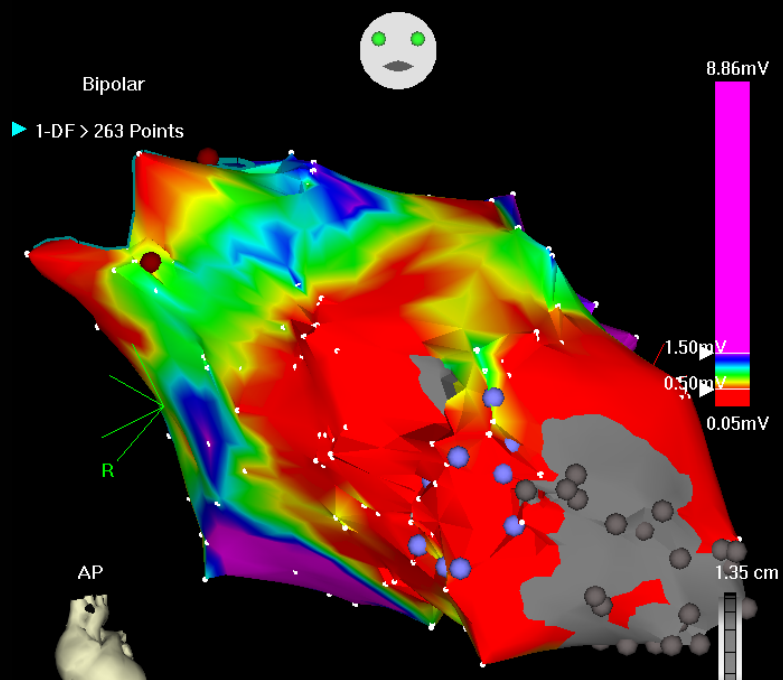


Late Potential: PVC

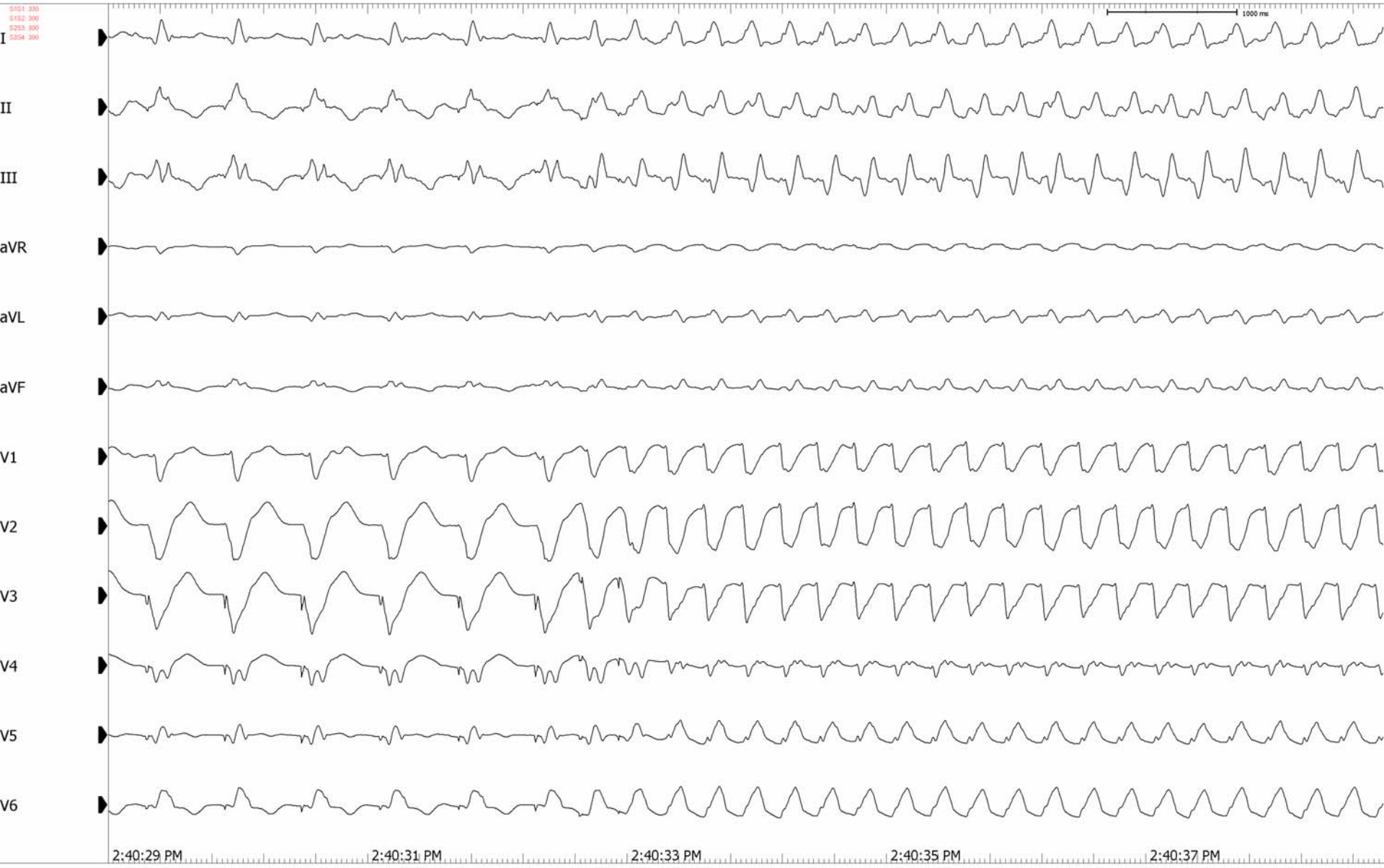


Long Stimulus to QRS

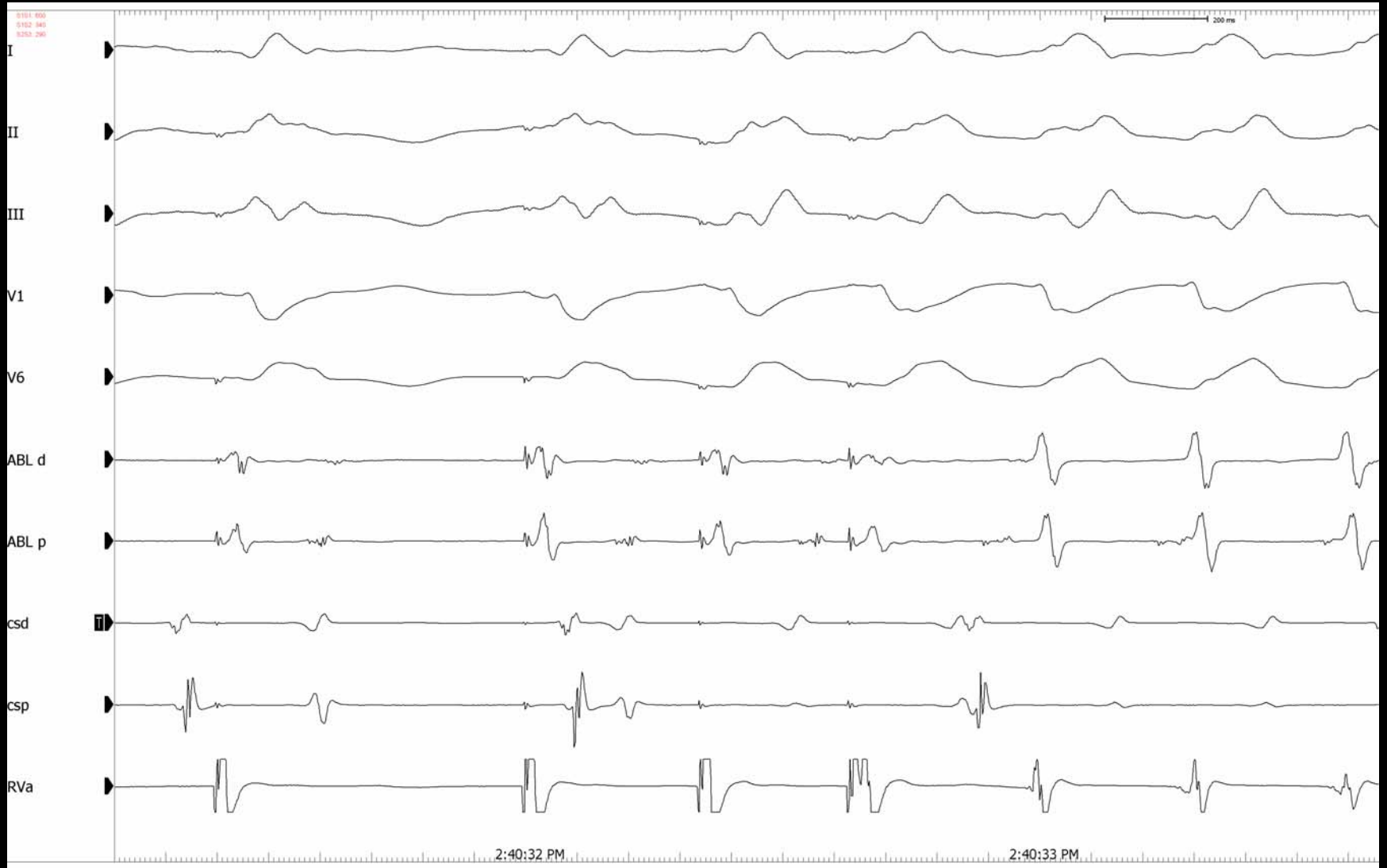


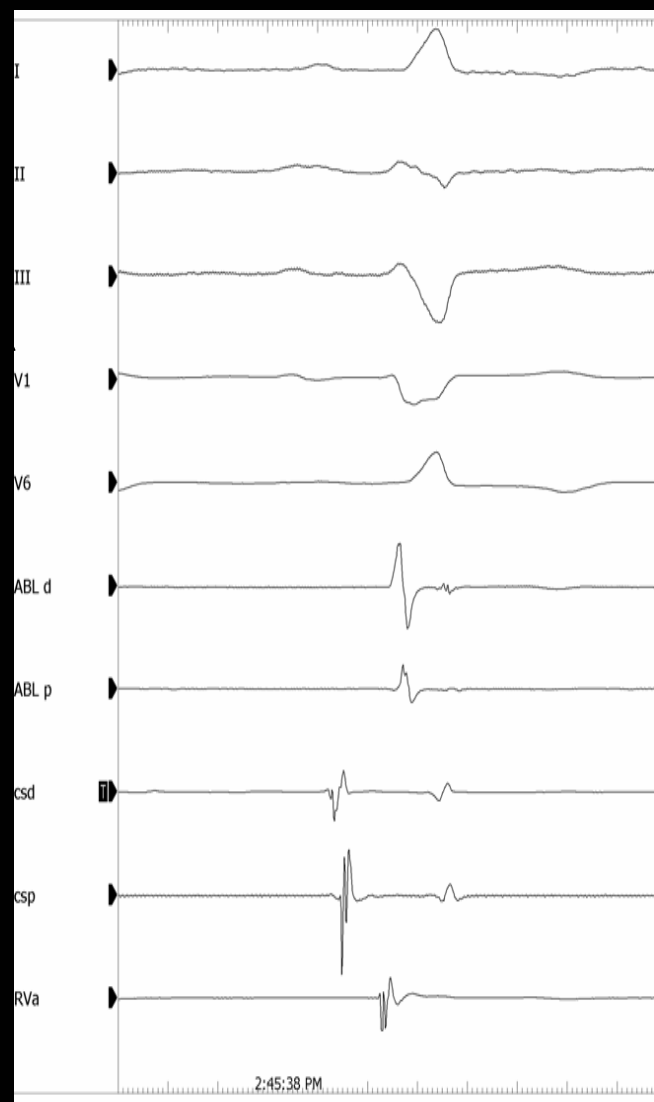
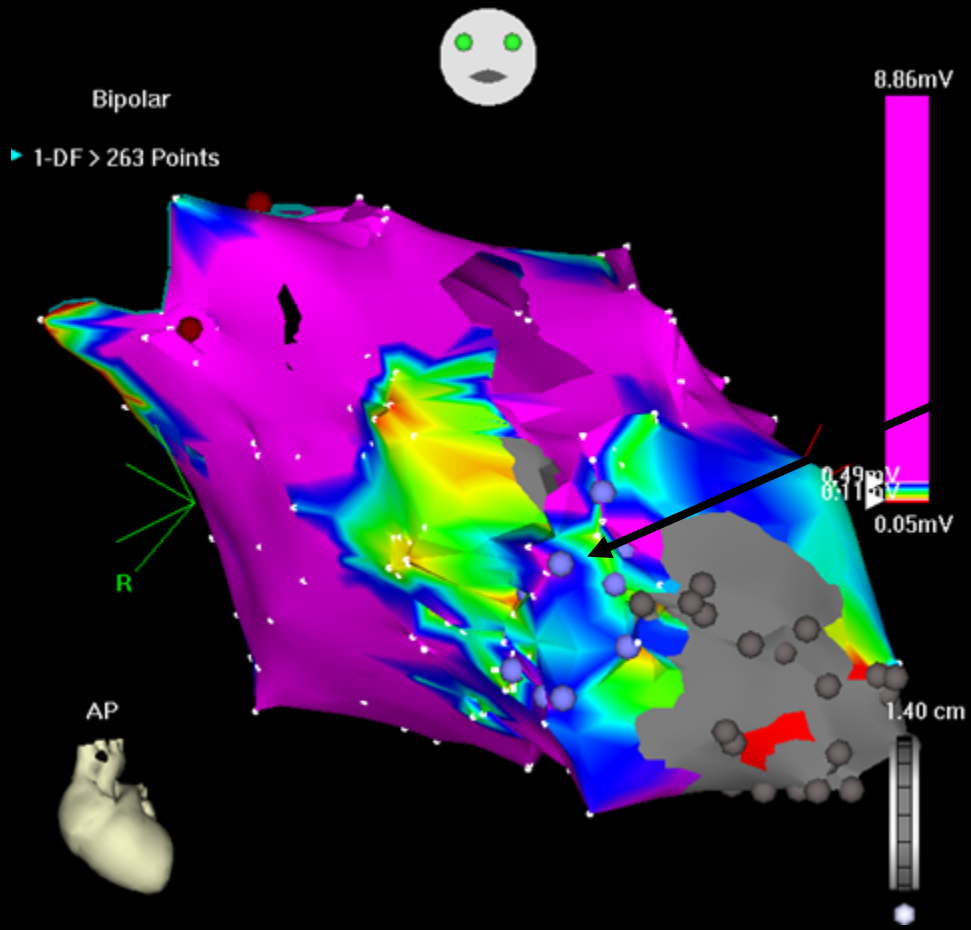


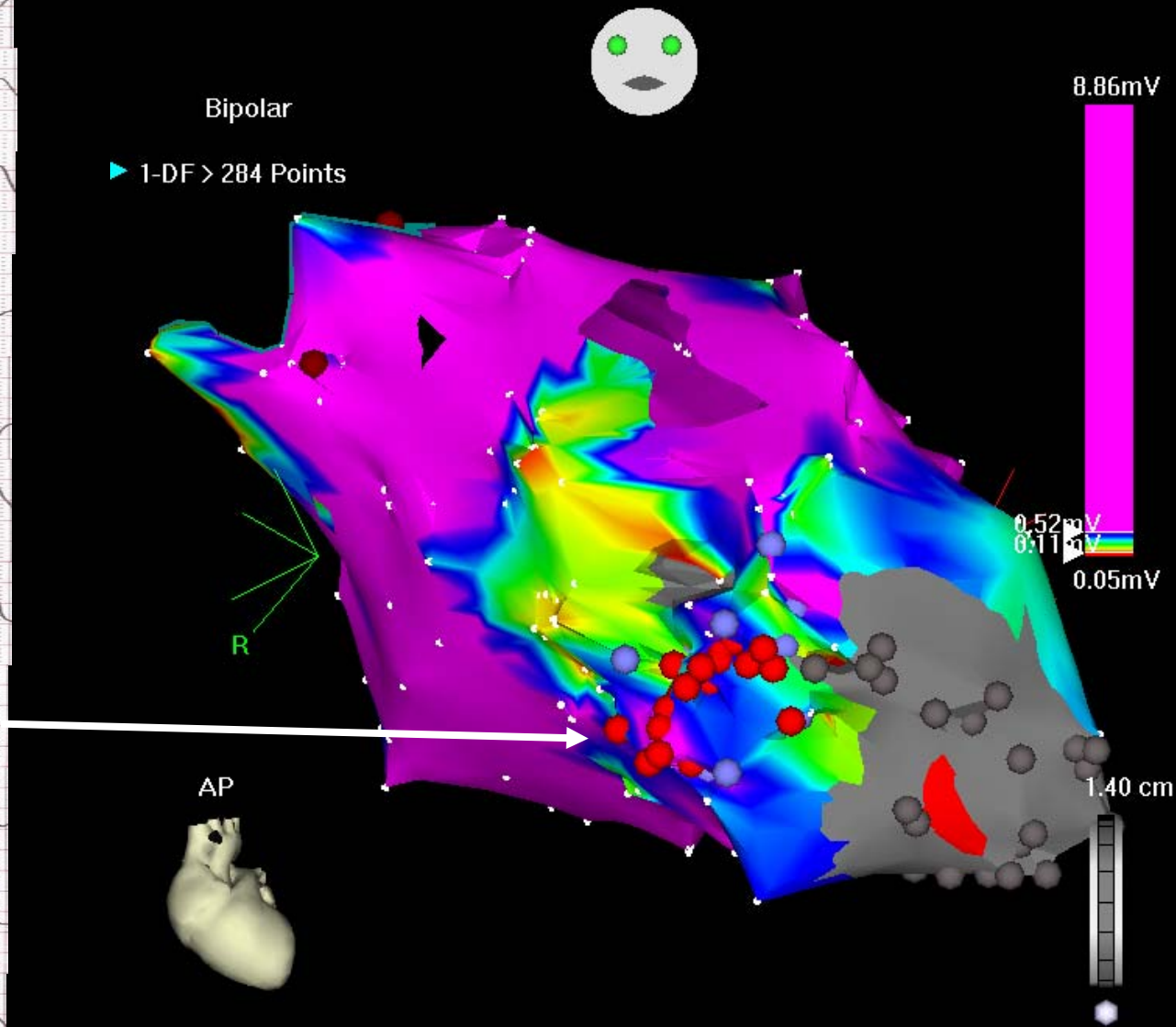
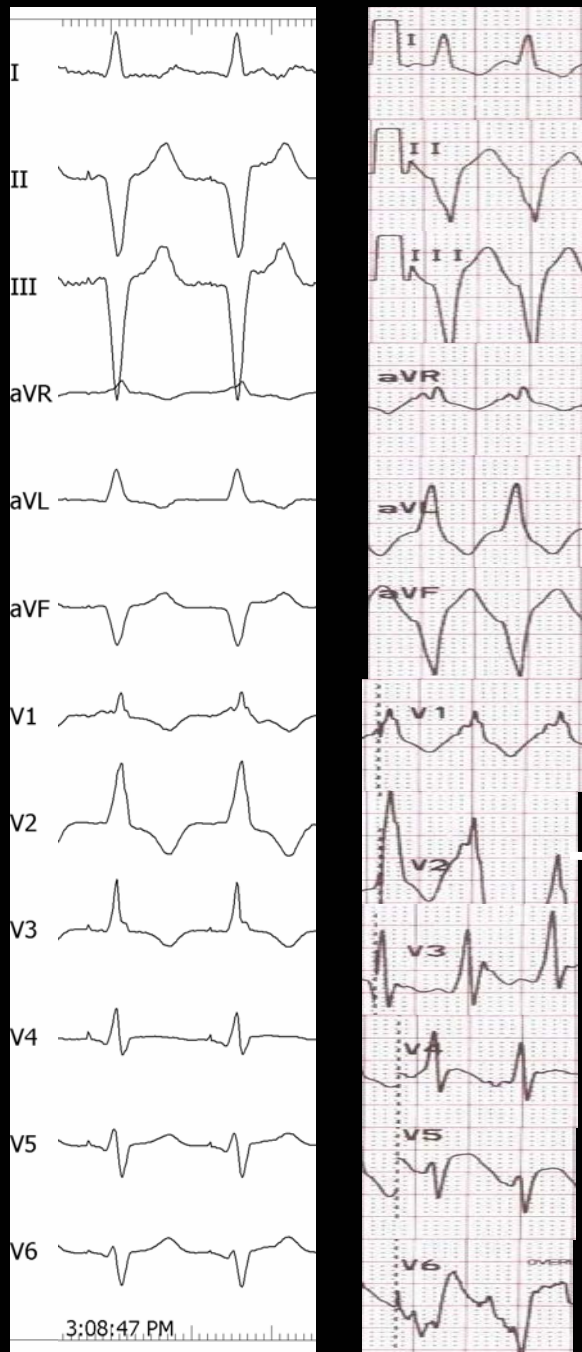
VT#1 Initiation



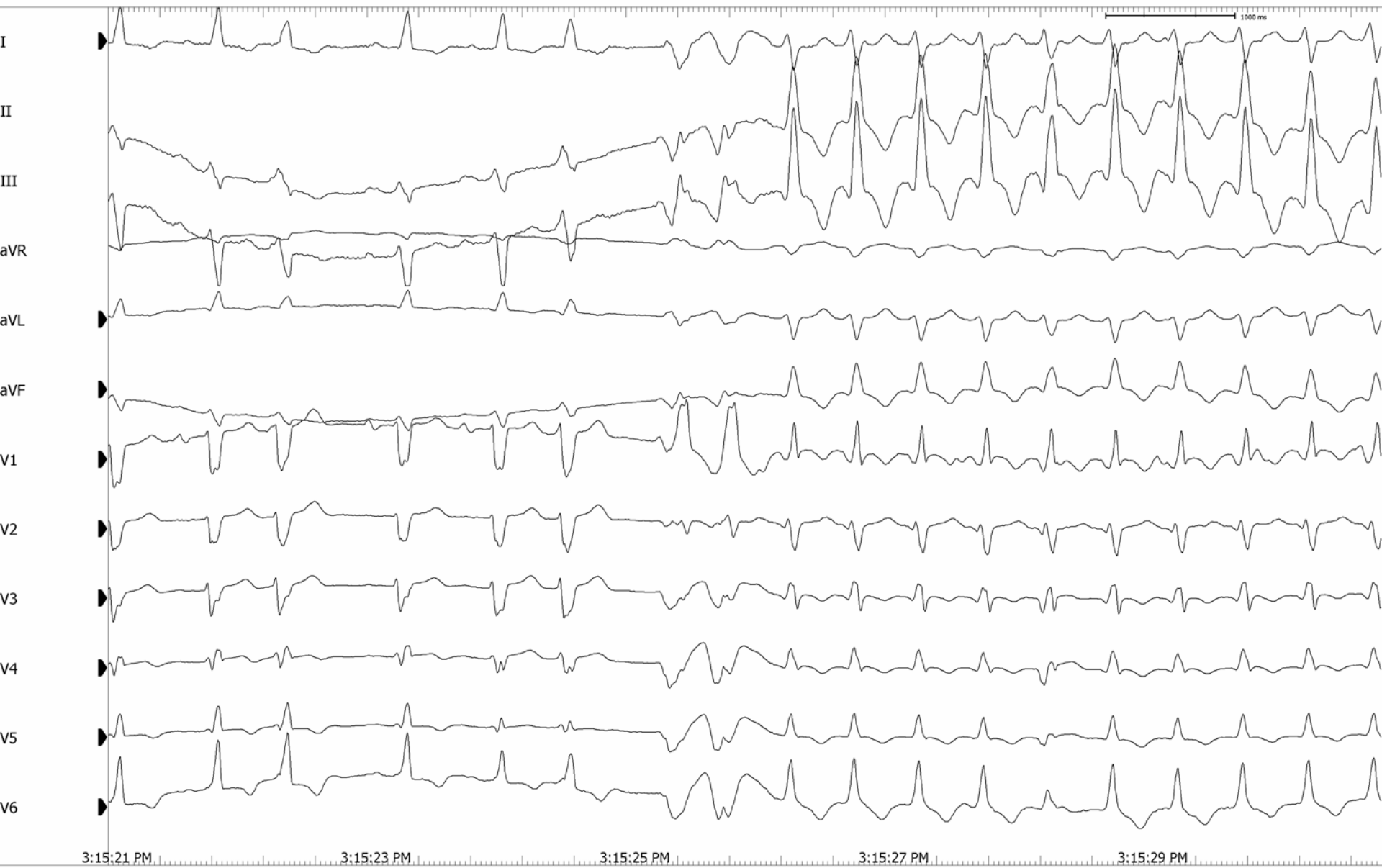
VT#1 Initiation

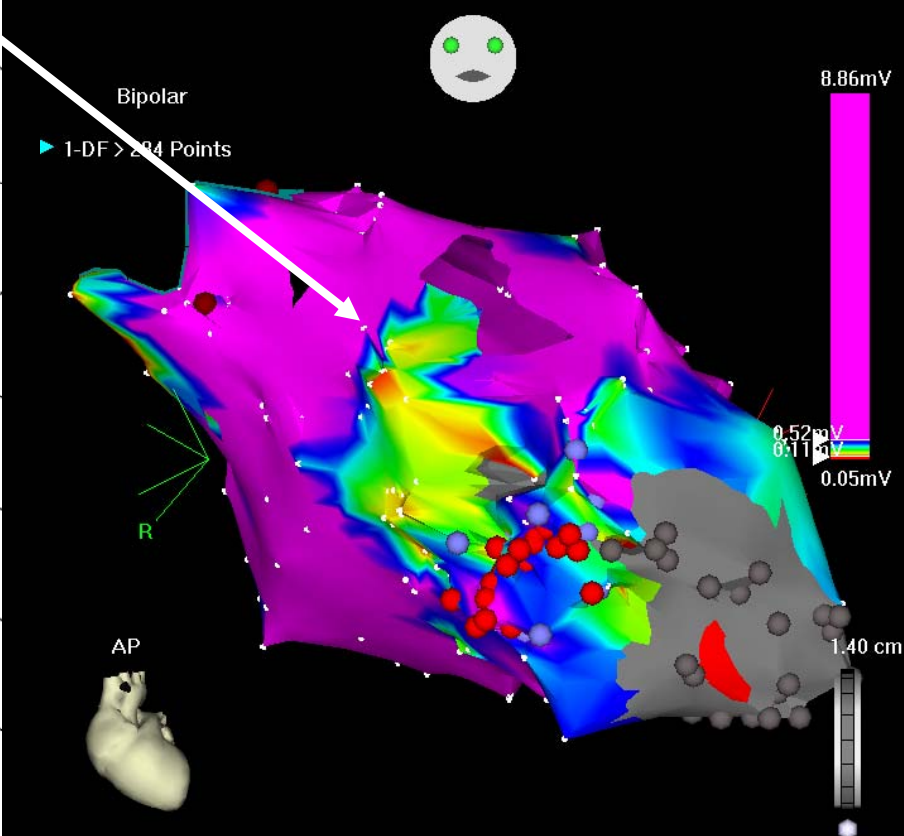


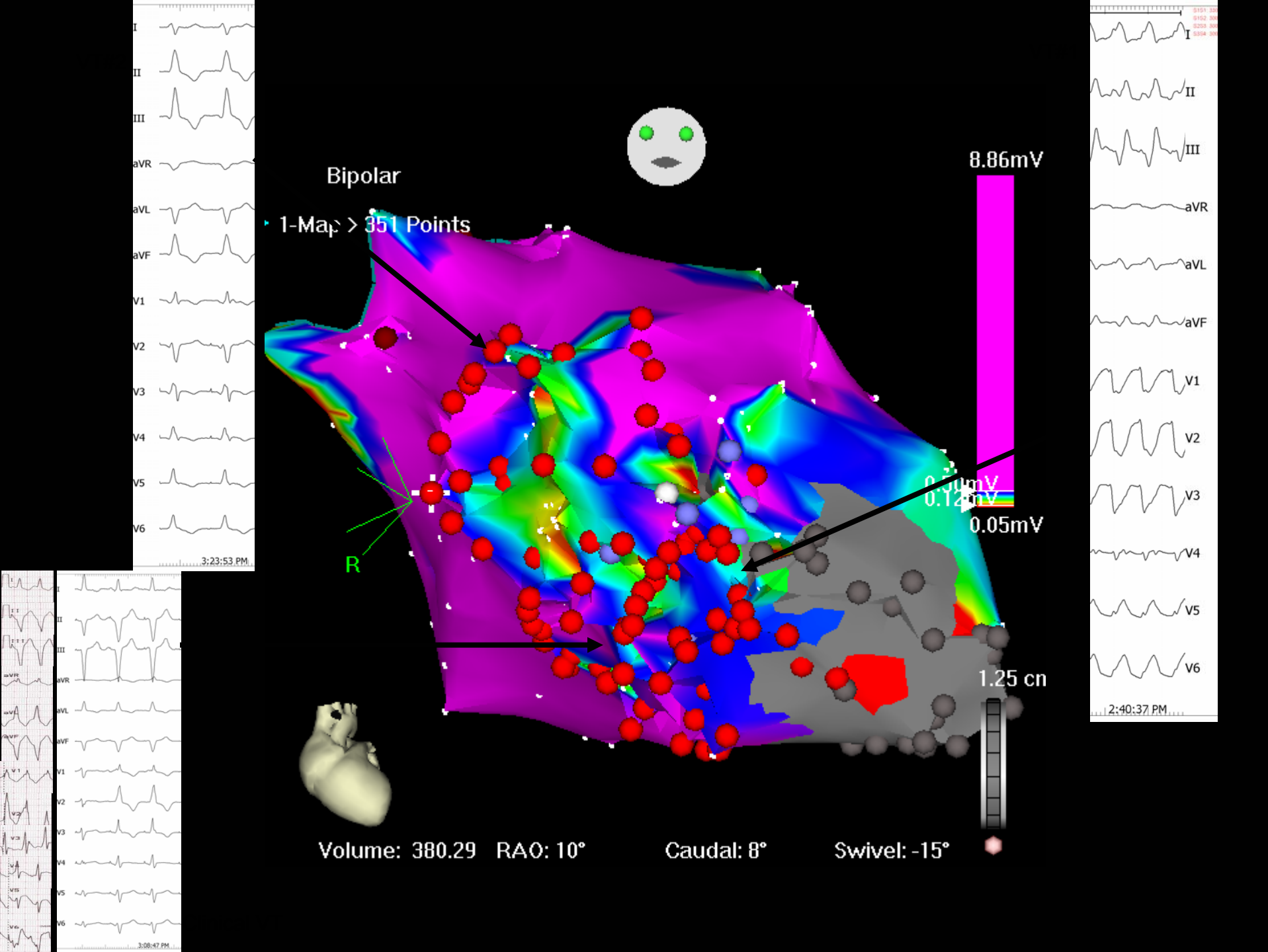


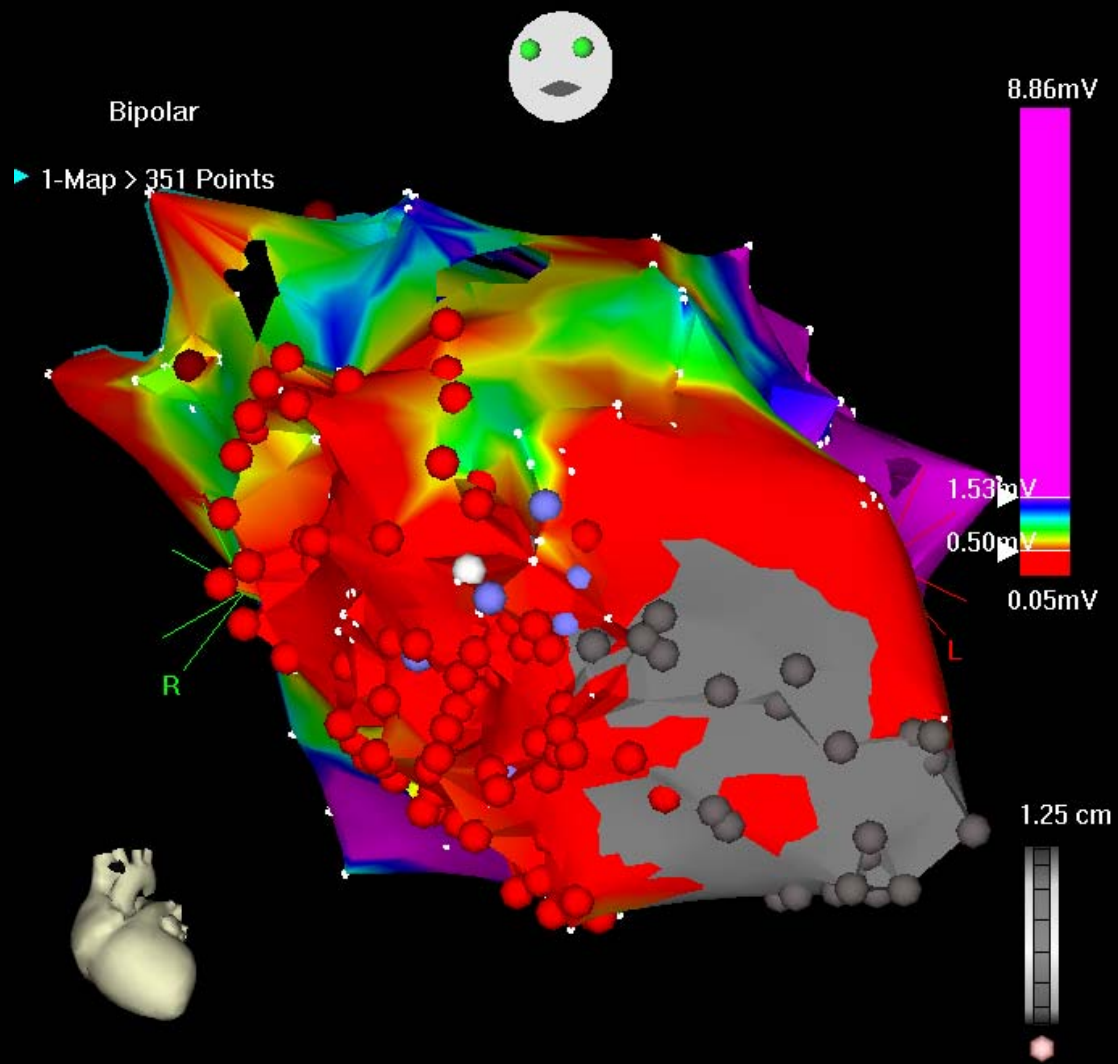


VT#2 Initiation





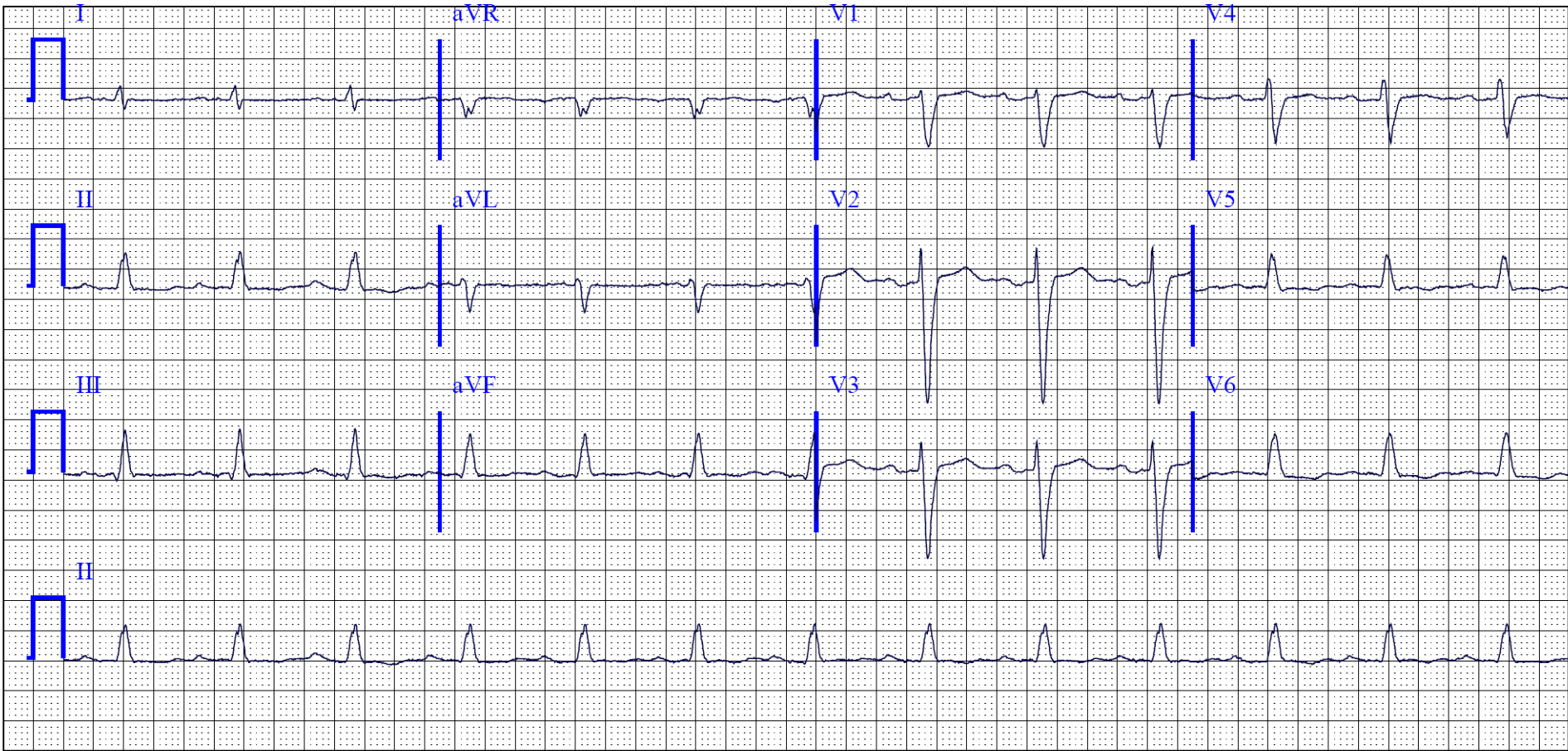


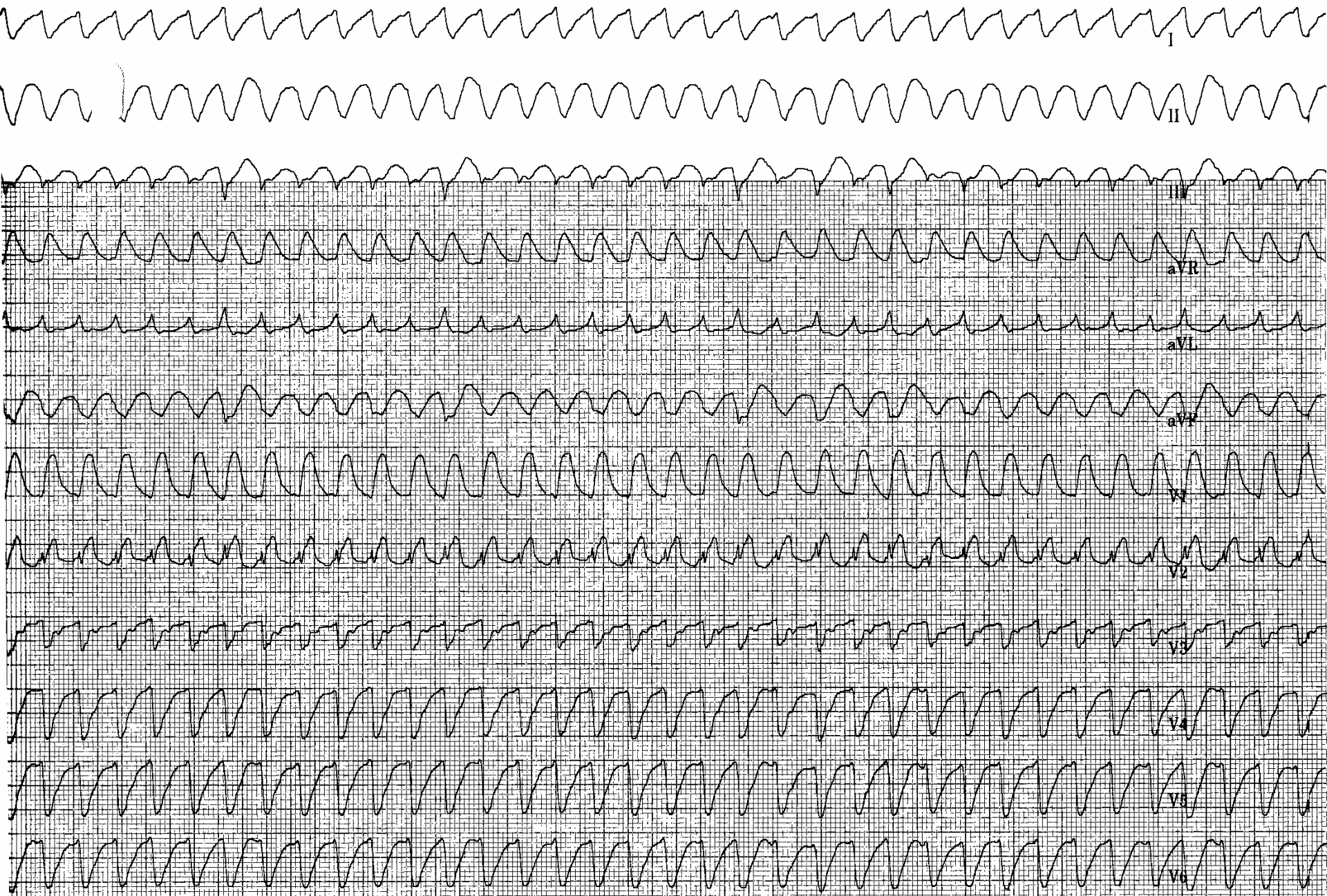


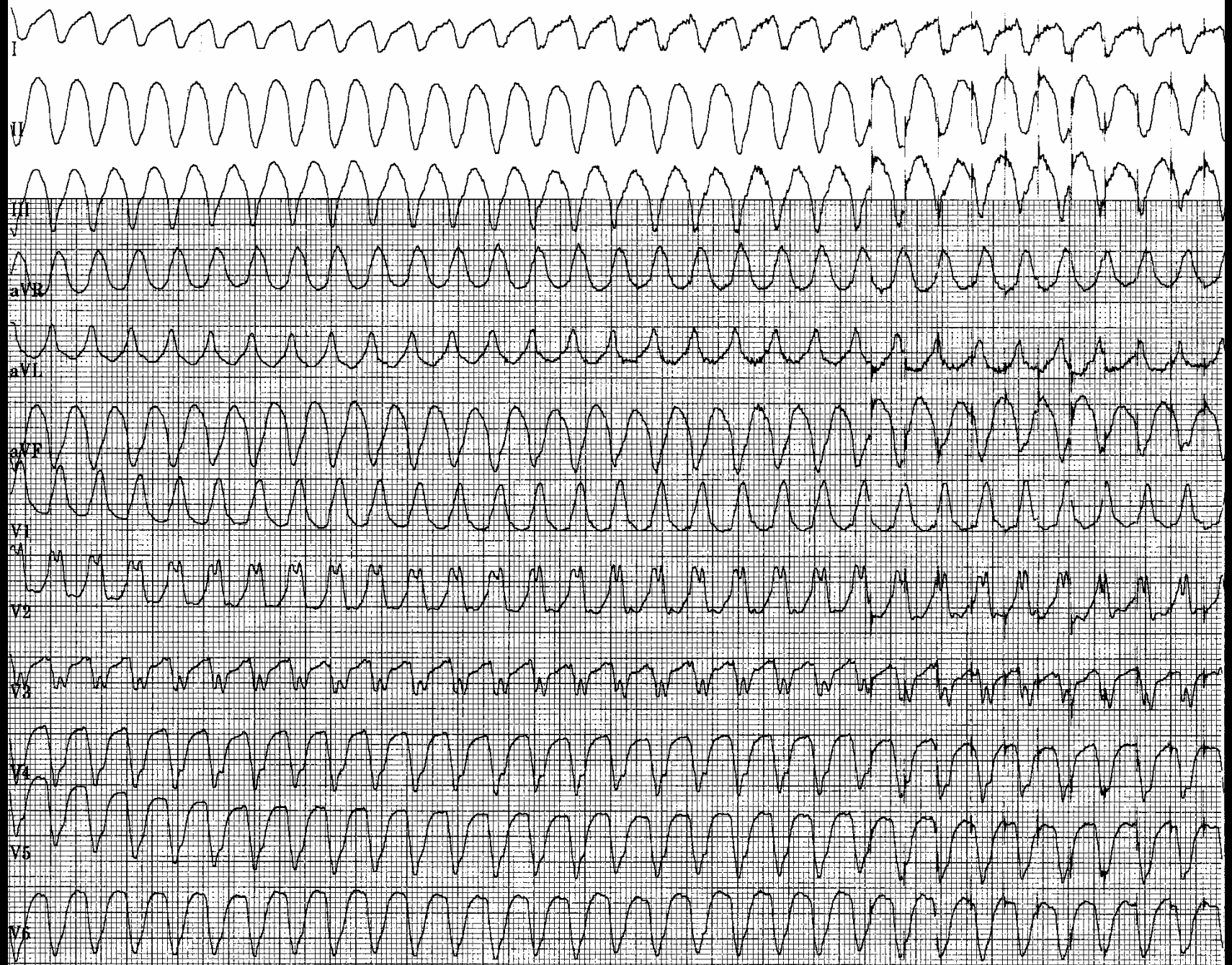
Clinical

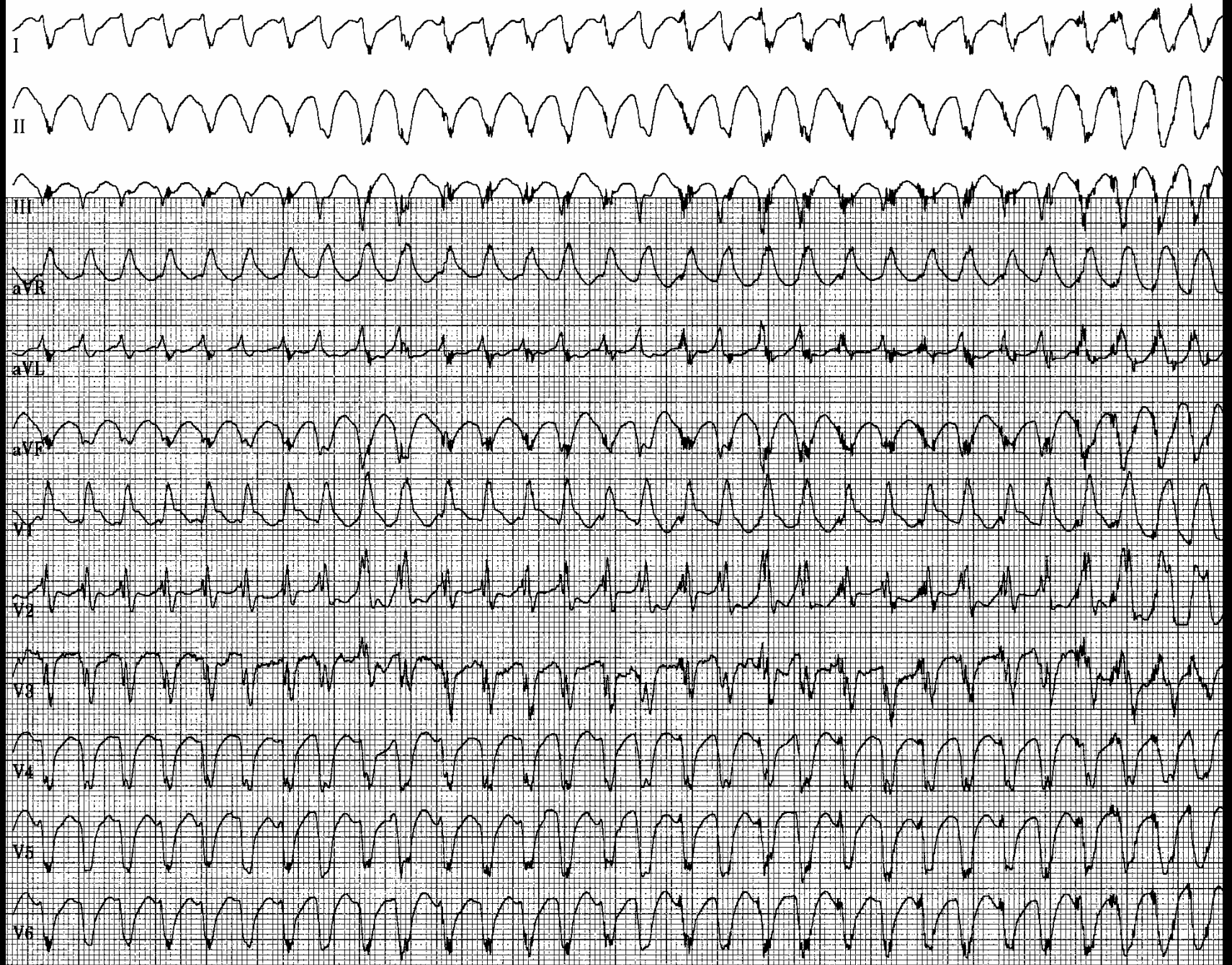
- 58M with morbid obesity, cardiomyopathy (LVEF 0.30), atrial fibrillation/flutter on amiodarone at home, COPD, OSA, DM, tonsillar CA s/p chemo/XRT complicated by need for a PEG
- Was witnessed falling to his bathroom floor in on 12/30/06. His son performed CPR until EMS arrived
- Pt was found in VF (down <5 min). He was shocked x 1, intubated, and brought to OSH where he was given lidocaine 100mg x 1 and started on amiodarone drip. Cardiac enzymes were not elevated and electrolytes were not deranged.
- On 12/31/06 he had VT (RBBB, superior axis, CL 280 ms) and was cardioverted and kept on amiodarone and given additional lidocaine 100mg x 1 and started on lido drip at 2mg. He then had another VT event which was resolved by repeat lidocaine bolus and increasing drip to 3mg
- He was referred for VT ablation.

	Intervals			Axes		
Rate	PR	QRS	QT/QTc	P	QRS	T
78	234	136	426/457	52	86-123	

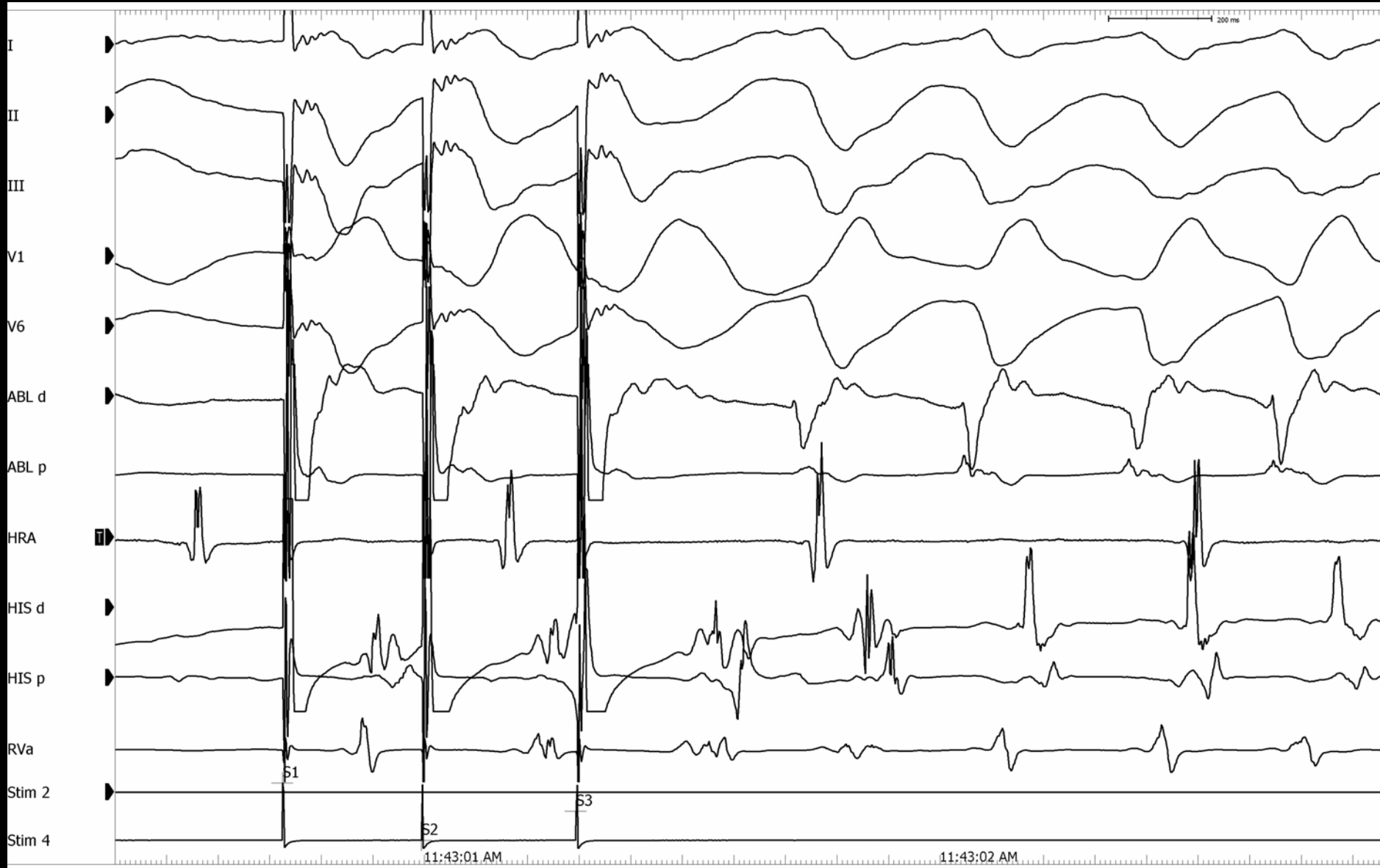








VT1 Induced



VT1



RVA pacing 270 ms

during VT1



RVA pacing 260 ms

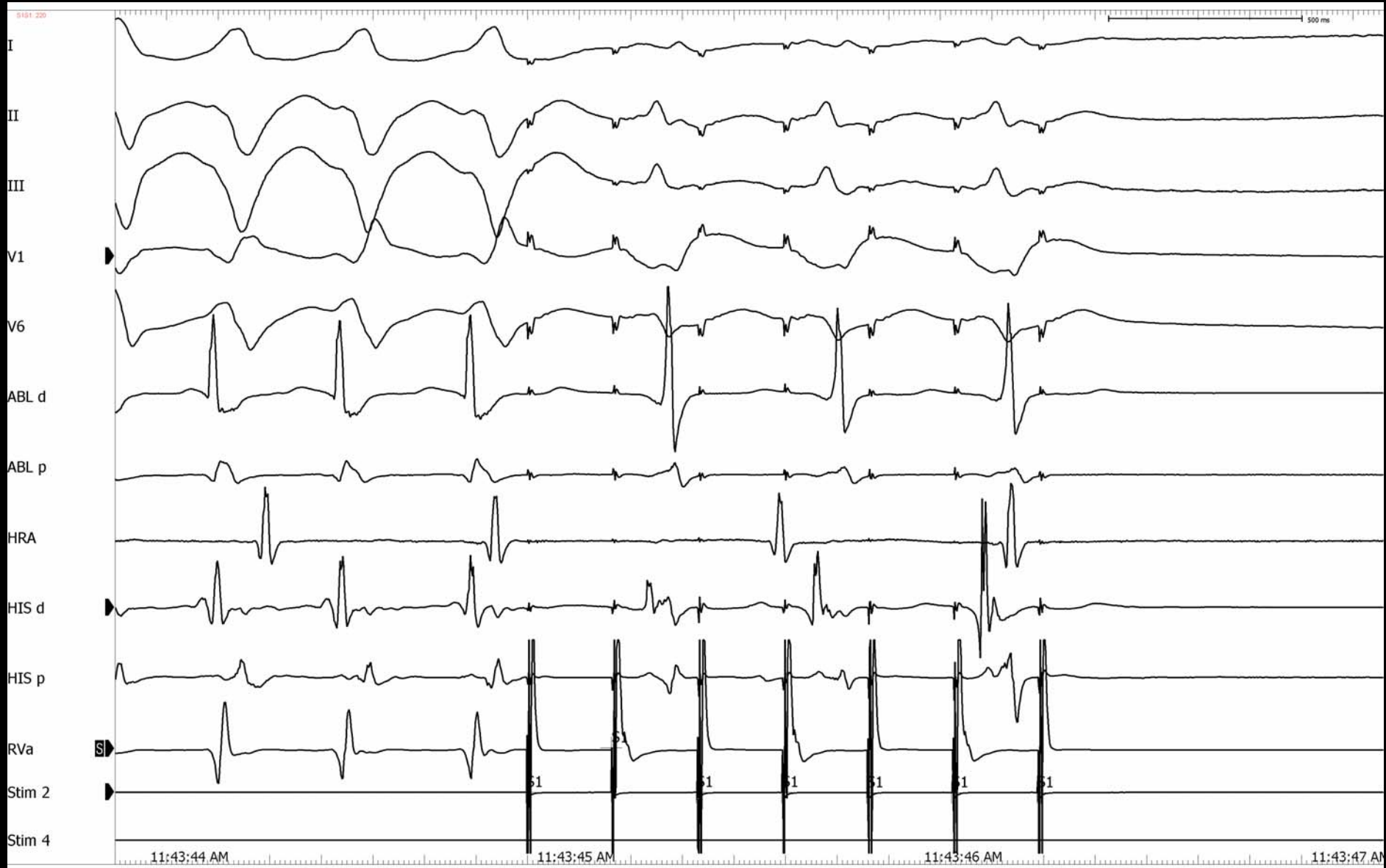
during VT1



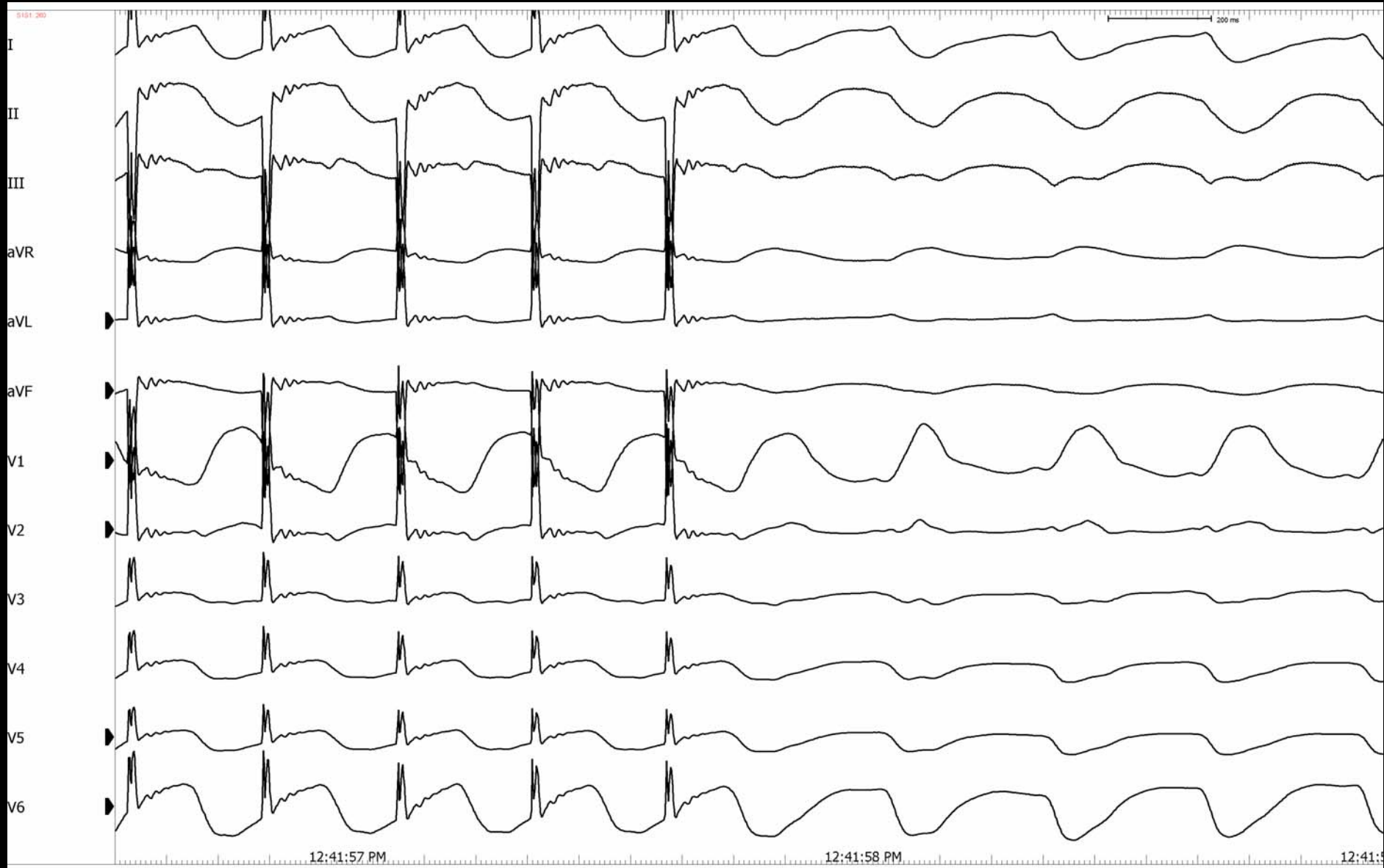
RVA pacing 300 ms

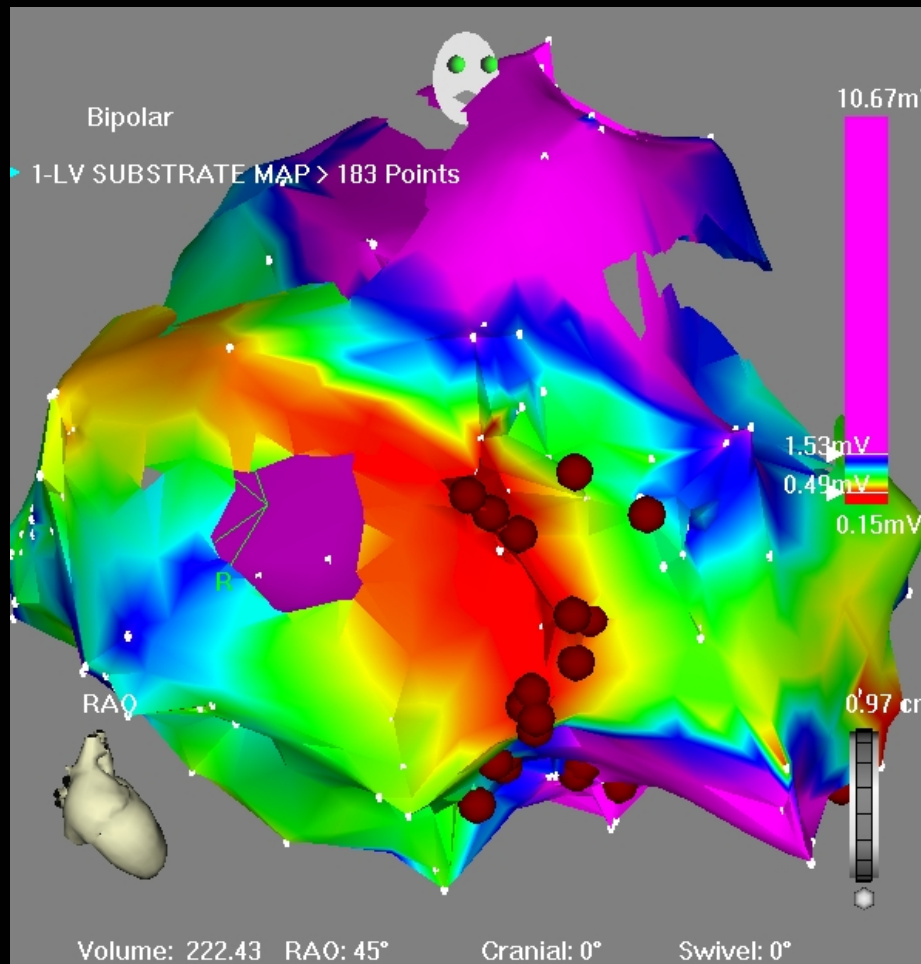


RVA pacing 220ms during VT#1

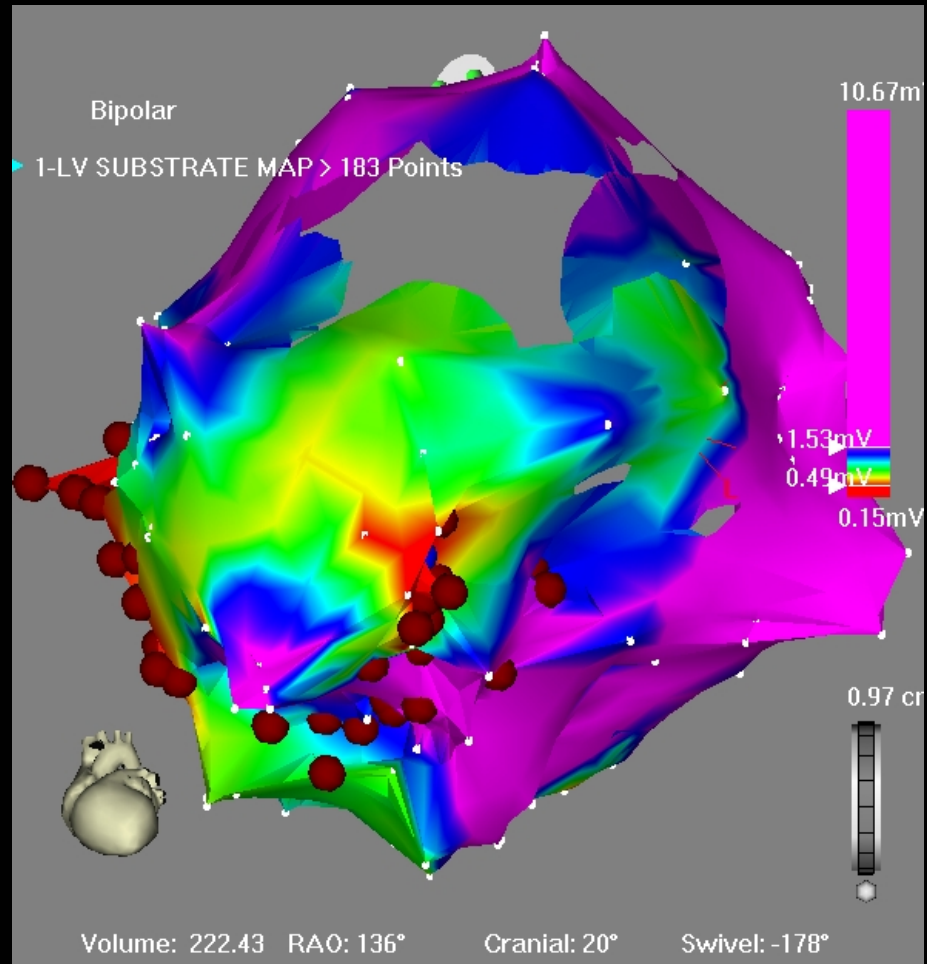


ABL pacing during VT#1





RAO

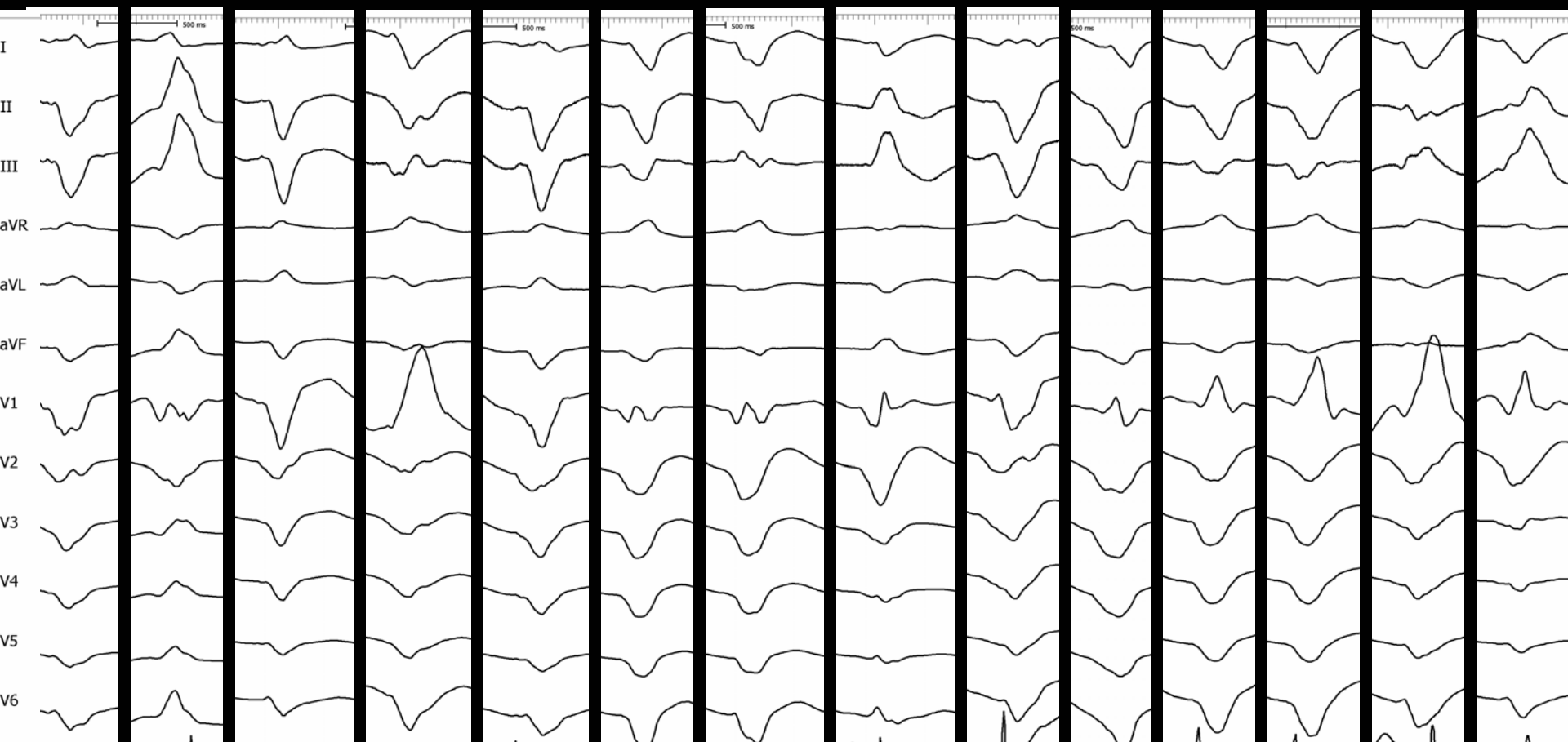


LAO

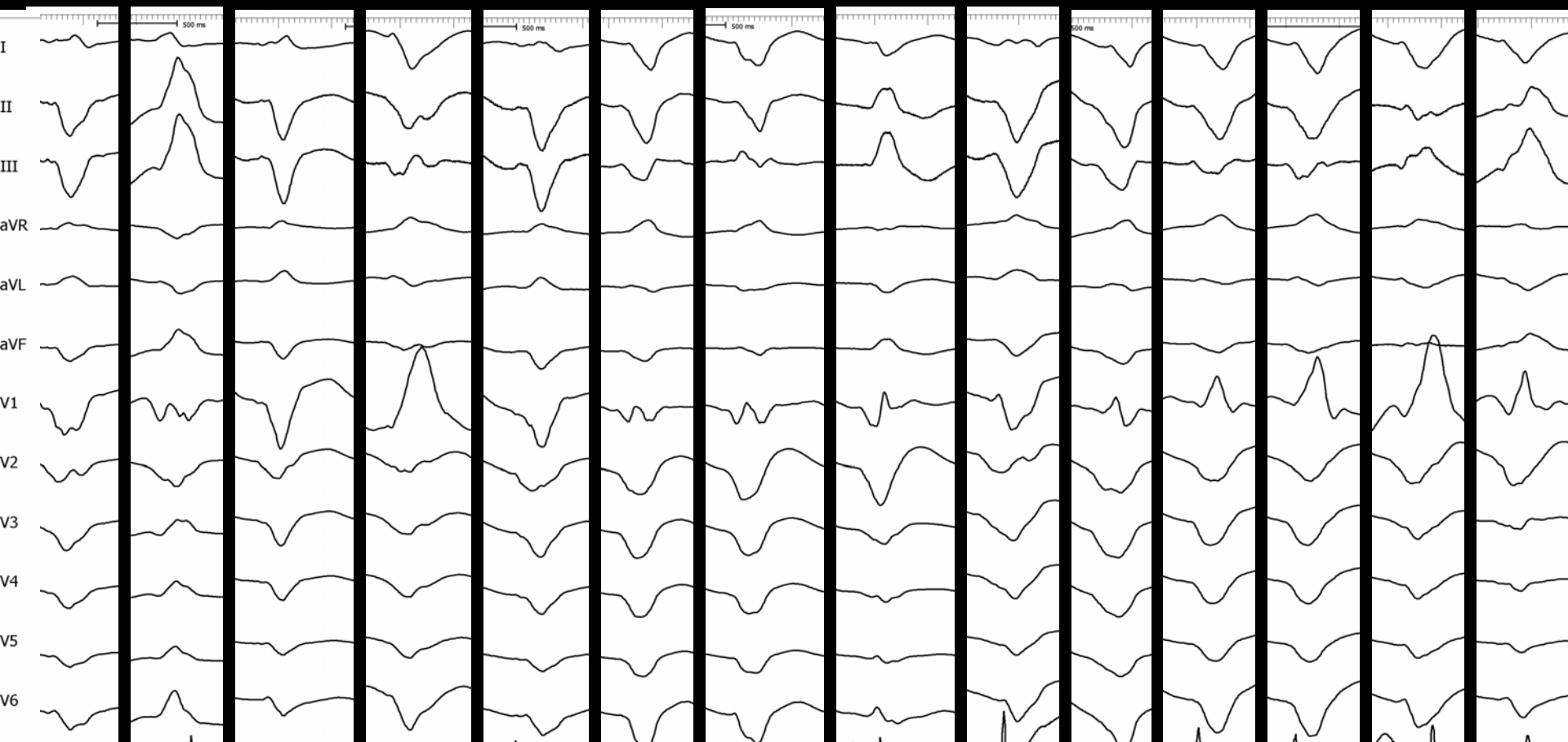
Clinical

- Recurrent VT despite ablation
- Sent for angiography where proximal LAD was found to be stenotic and dominant LCX was proximally occluded

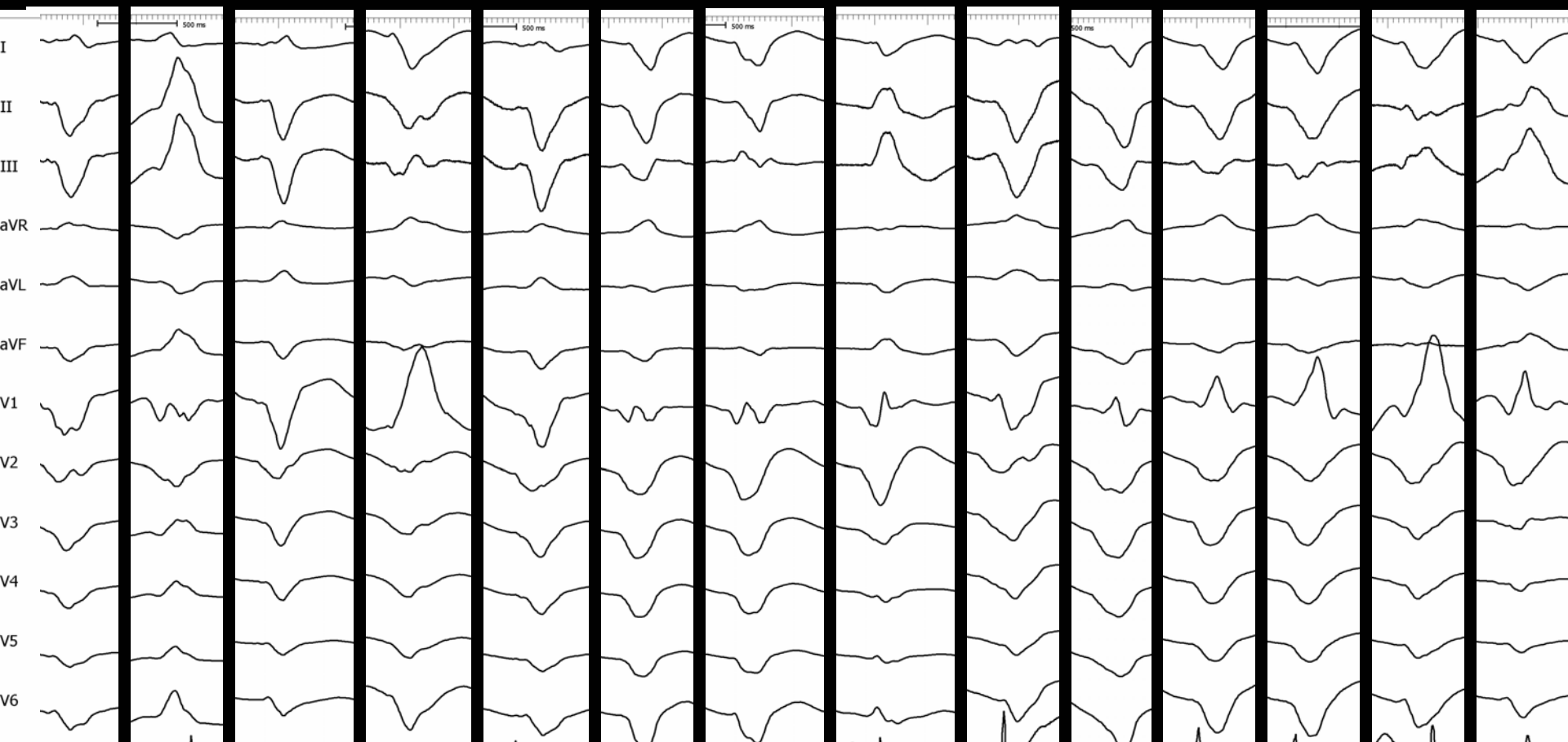
VT1 VT2 VT3 VT4 VT5 VT6 VT7 VT8 VT9 VT10 VT11 VT12 VT13 VT14



VT1 VT2 VT3 VT4 VT5 VT6 VT7 VT8 VT9 VT10 VT11 VT12 VT13 VT14

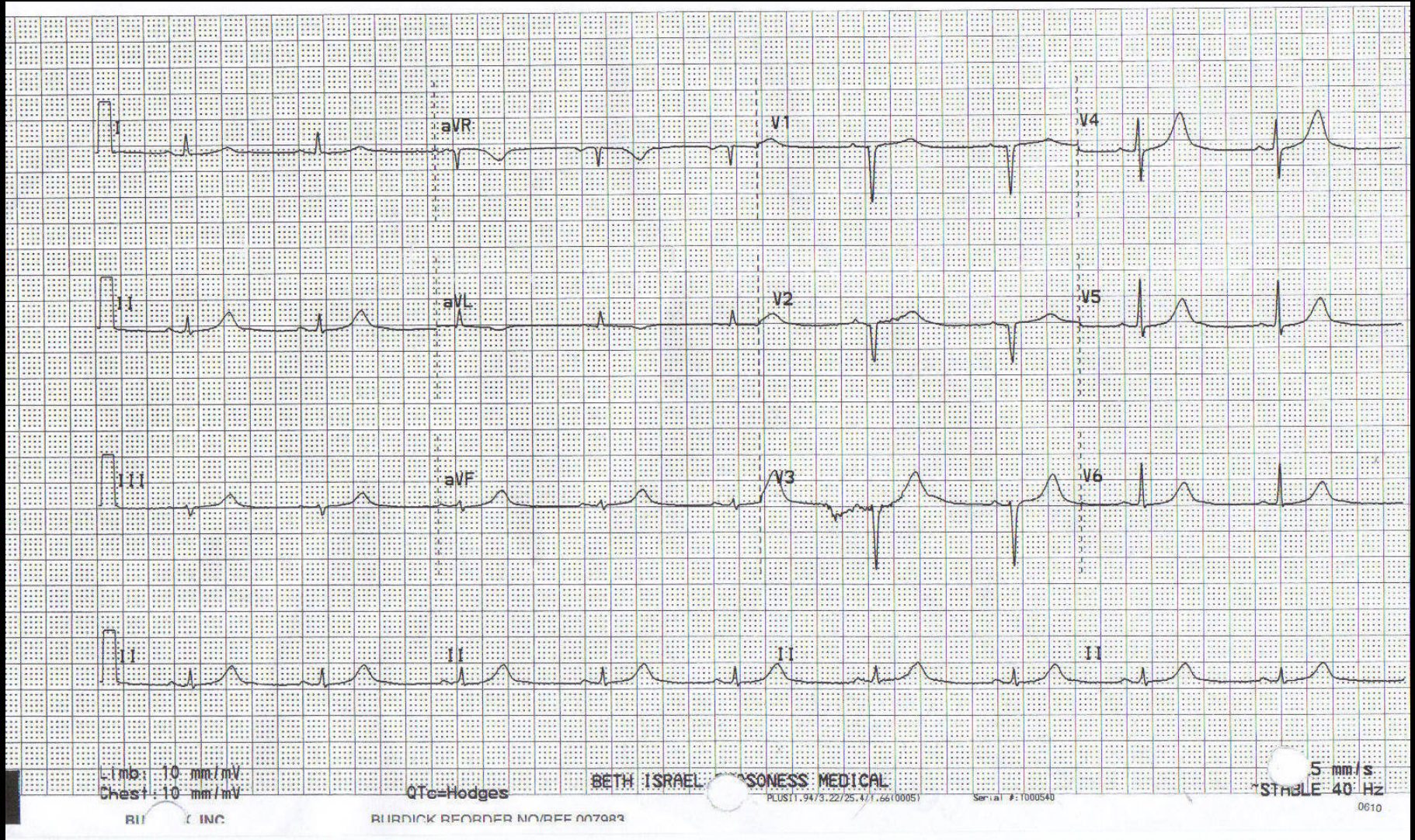


VT1 VT2 VT3 VT4 VT5 VT6 VT7 VT8 VT9 VT10 VT11 VT12 VT13 VT14

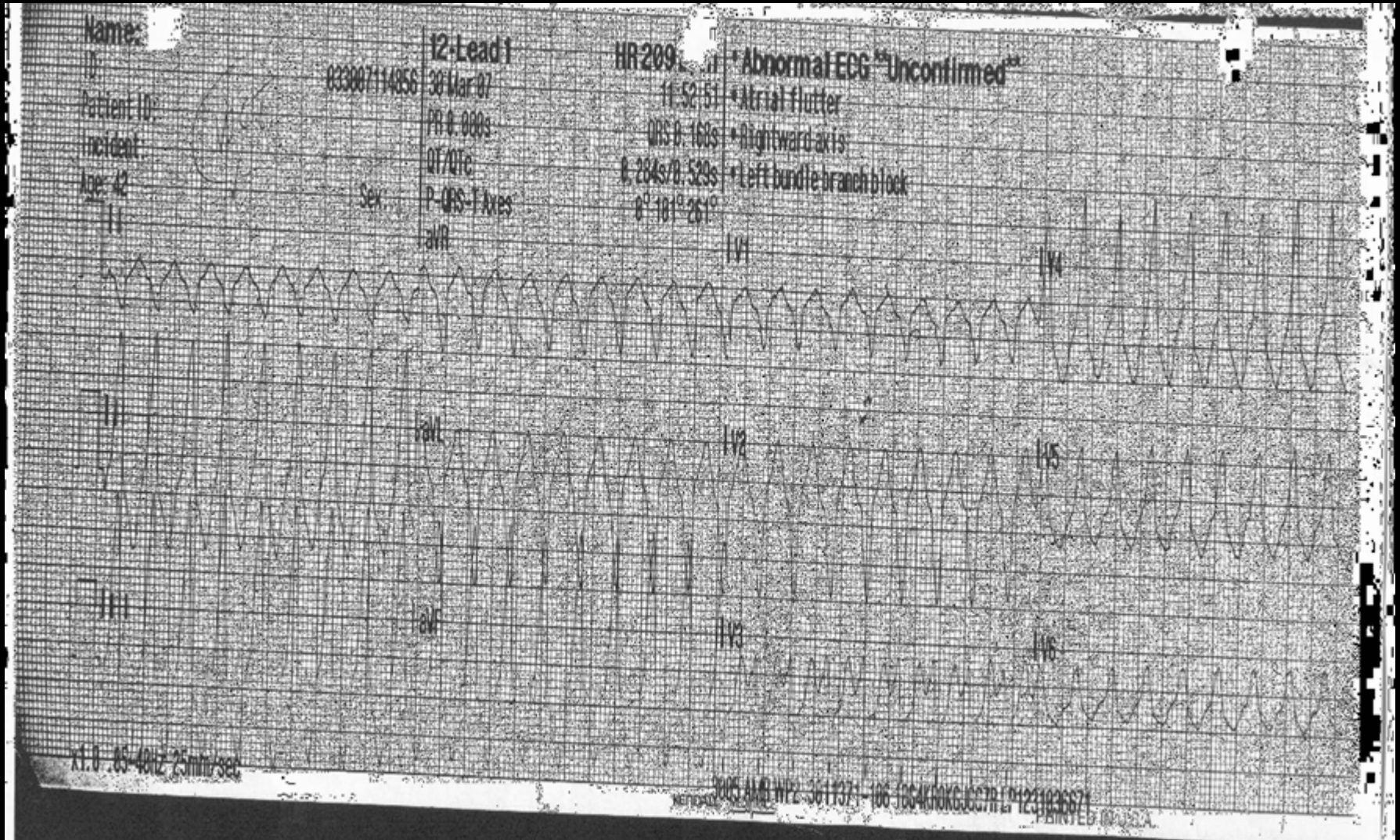


Outflow tract VTs

Baseline ECG

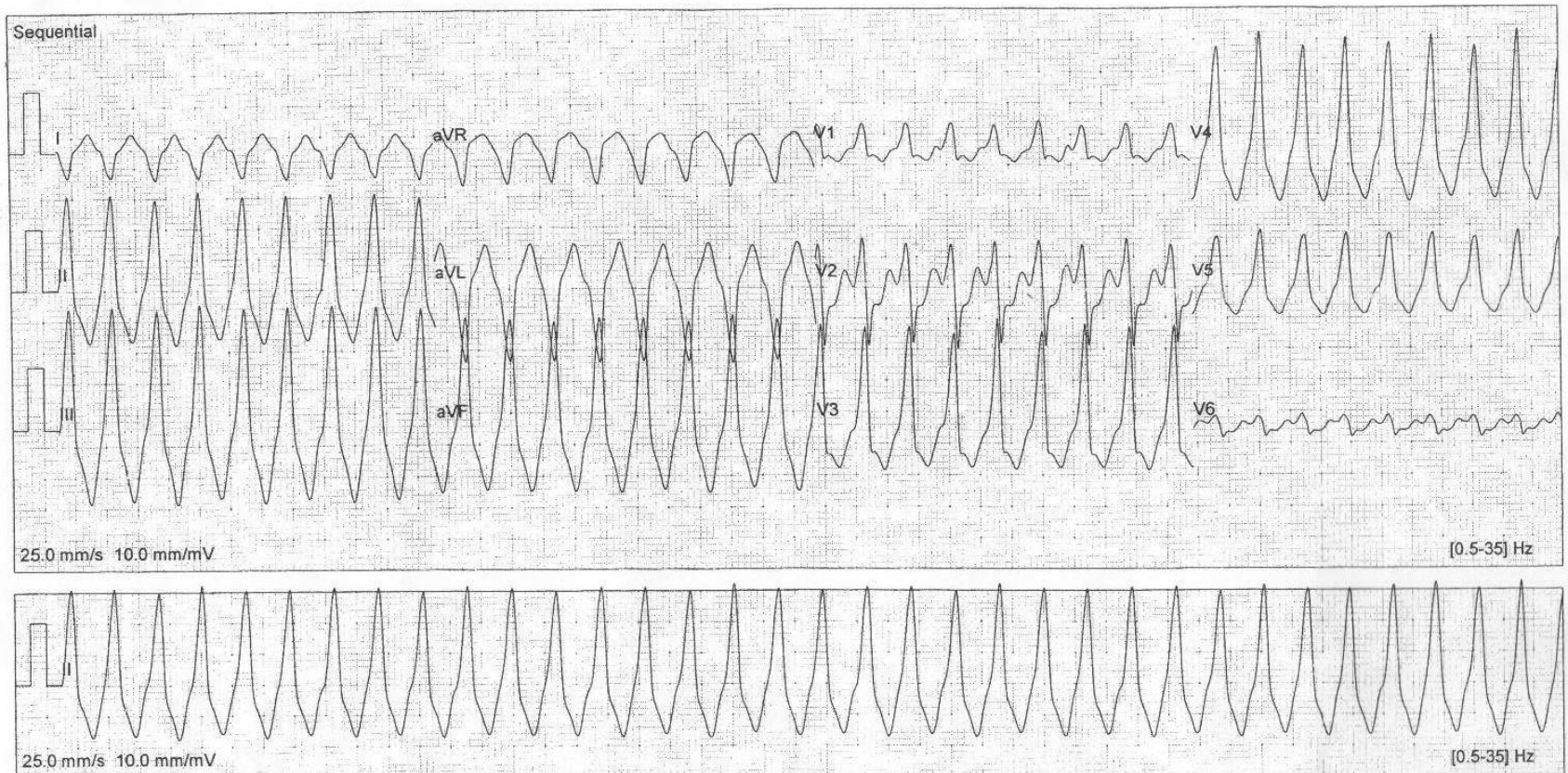


Clinical VT 1



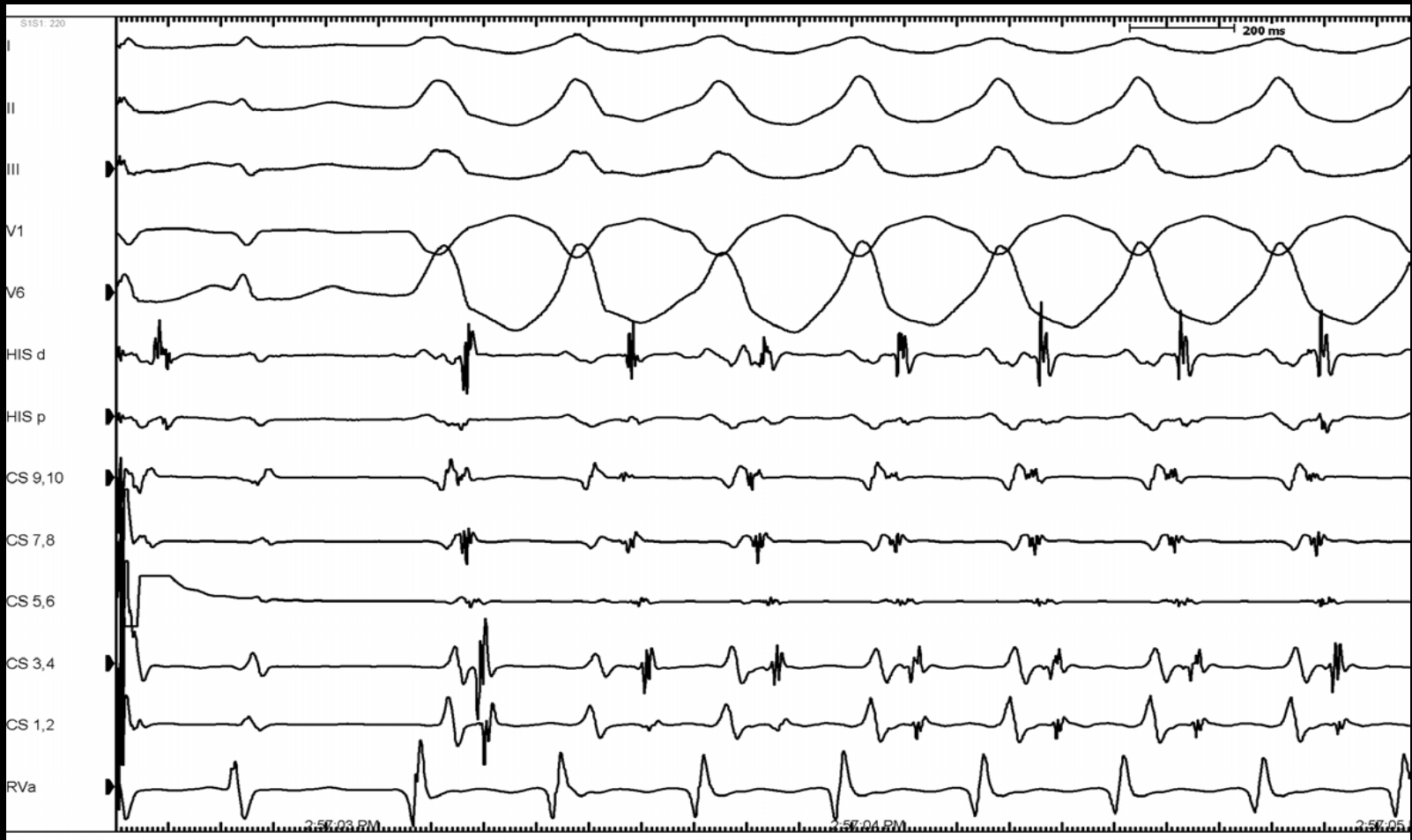
RVOT antero-septal

Clinical VT



LV Basal VT- SUPERIOR Lateral Mitral Annulus

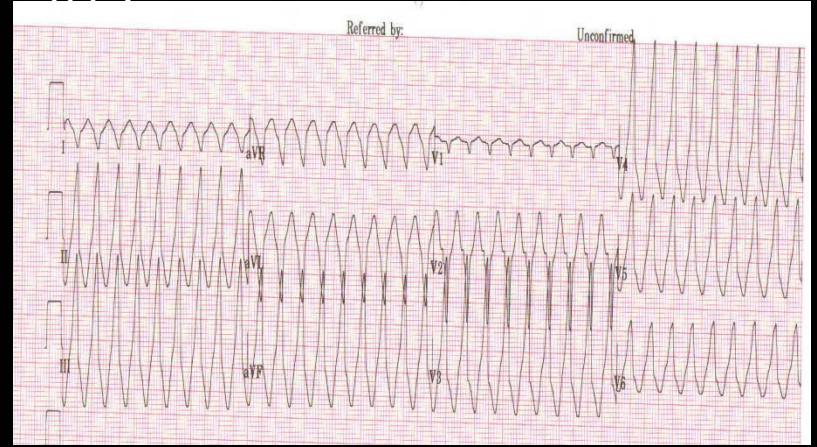
VT1 spontaneous induction



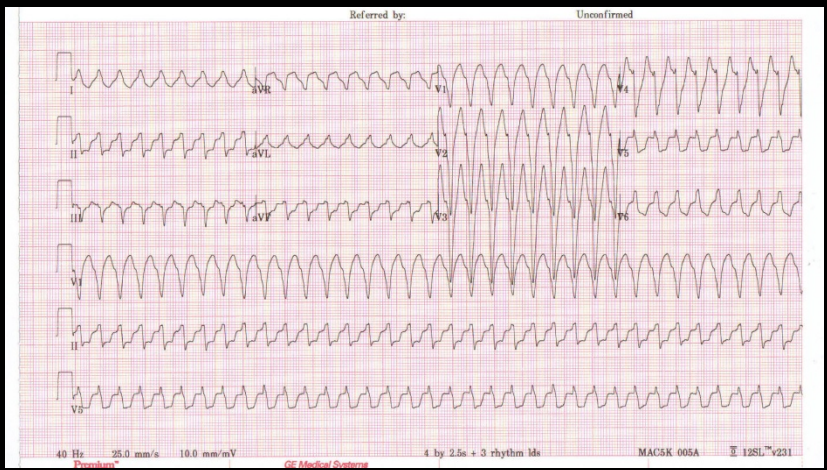
VT1



VT2

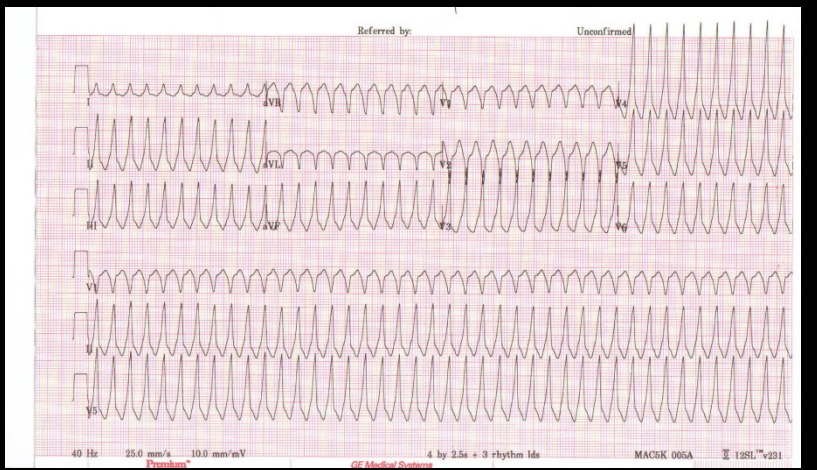


Referred by: Unconfirmed



VT3

Referred by: Unconfirmed



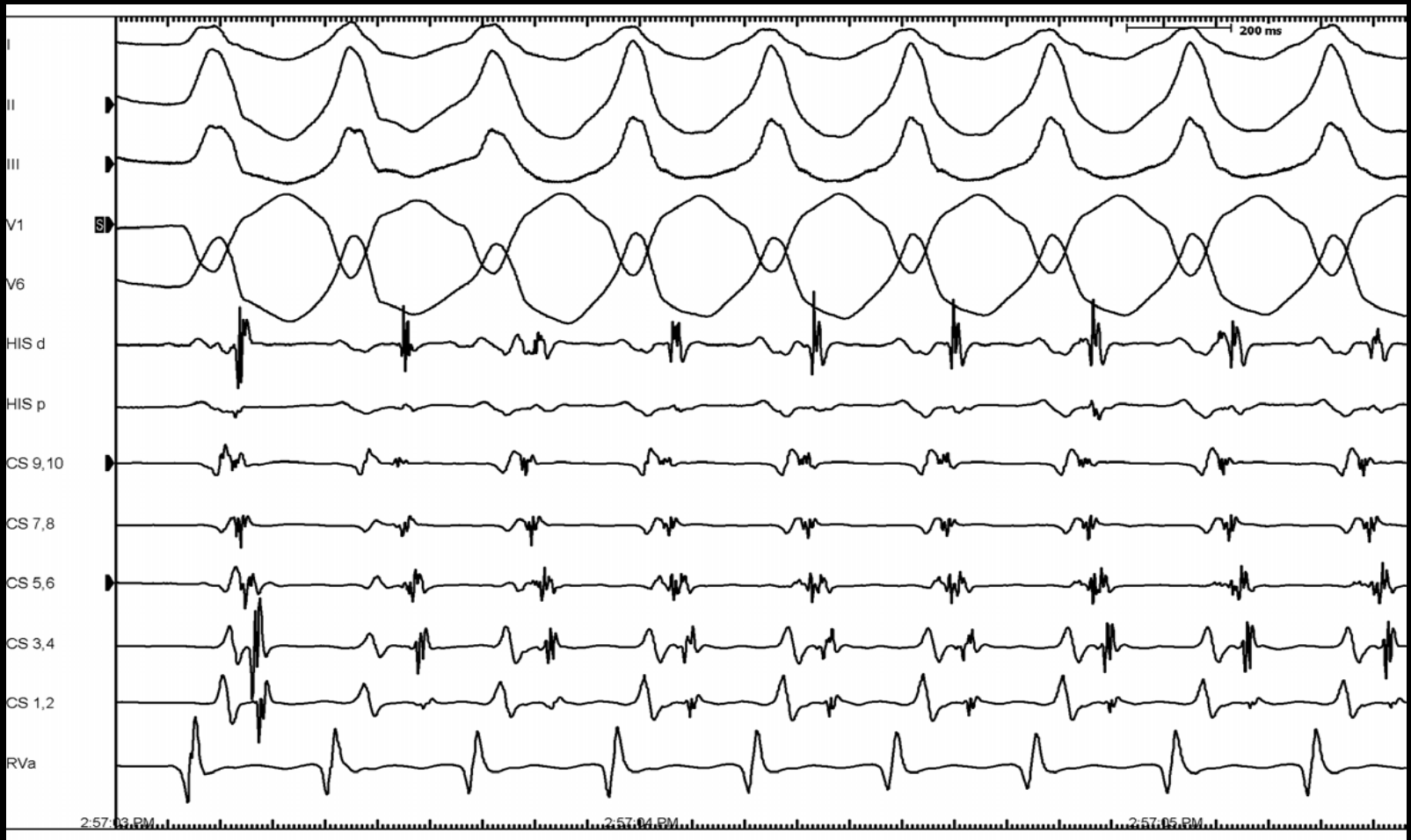
VT4

VT1 Postero-septal VT?

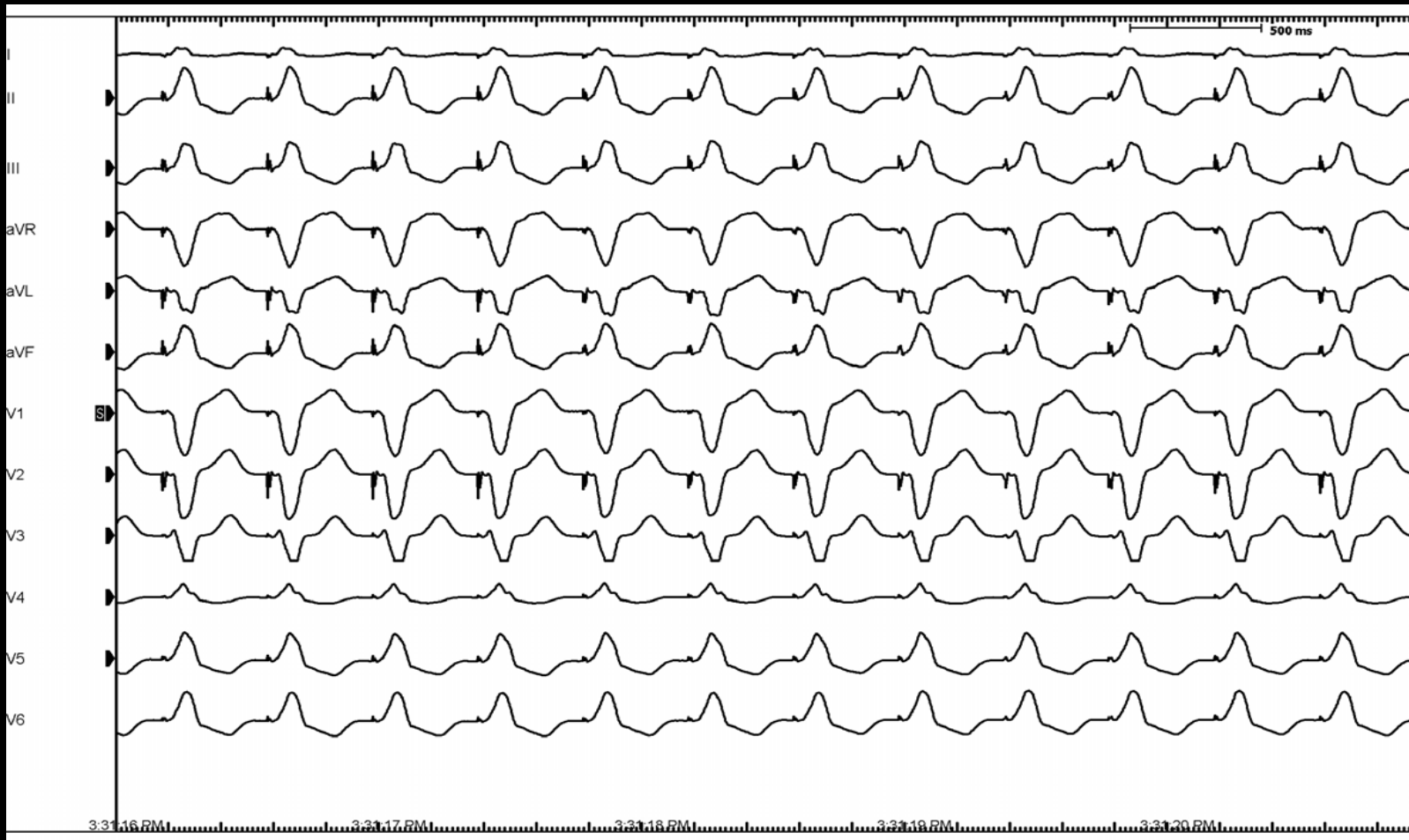
Referred by:

Unconfirmed

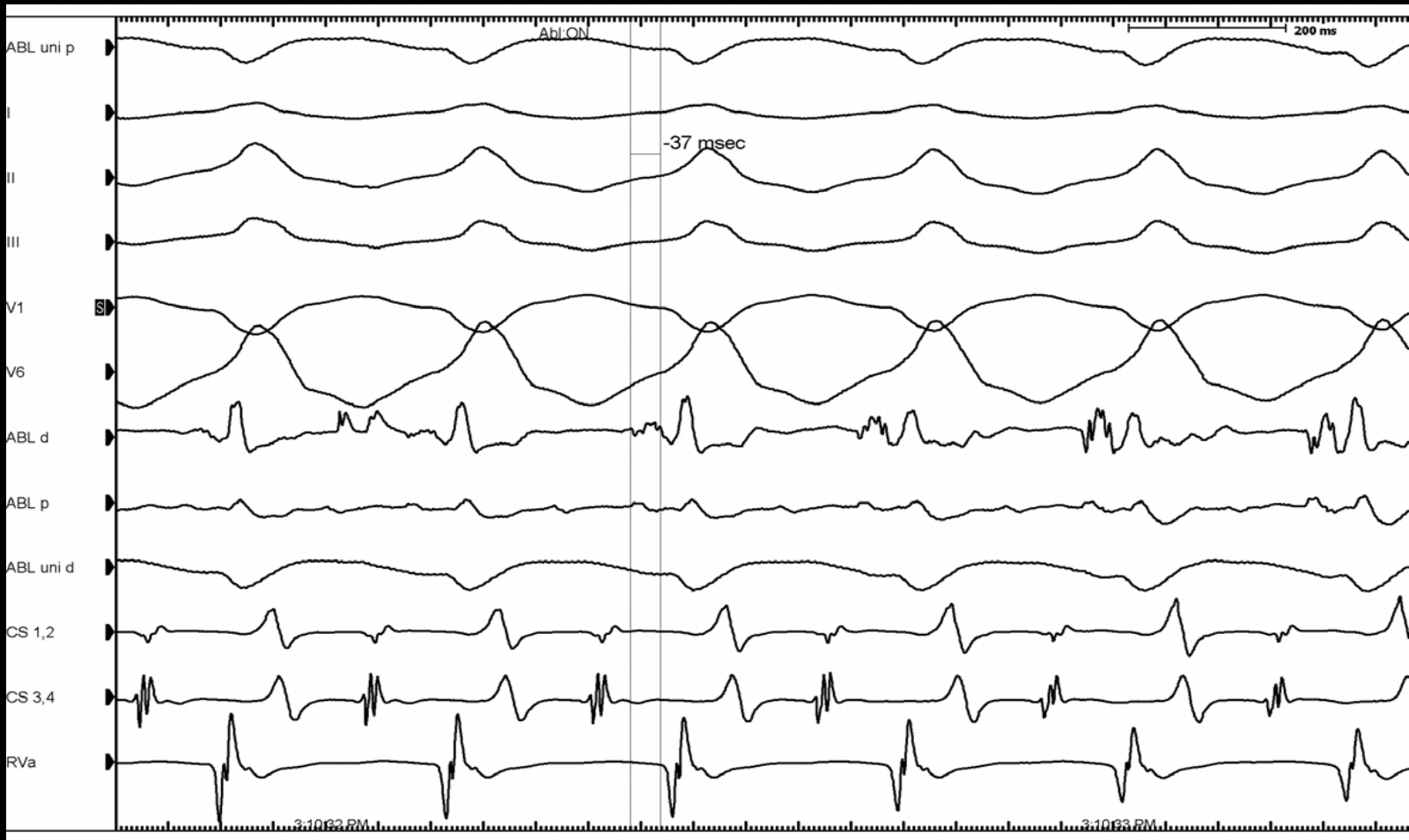




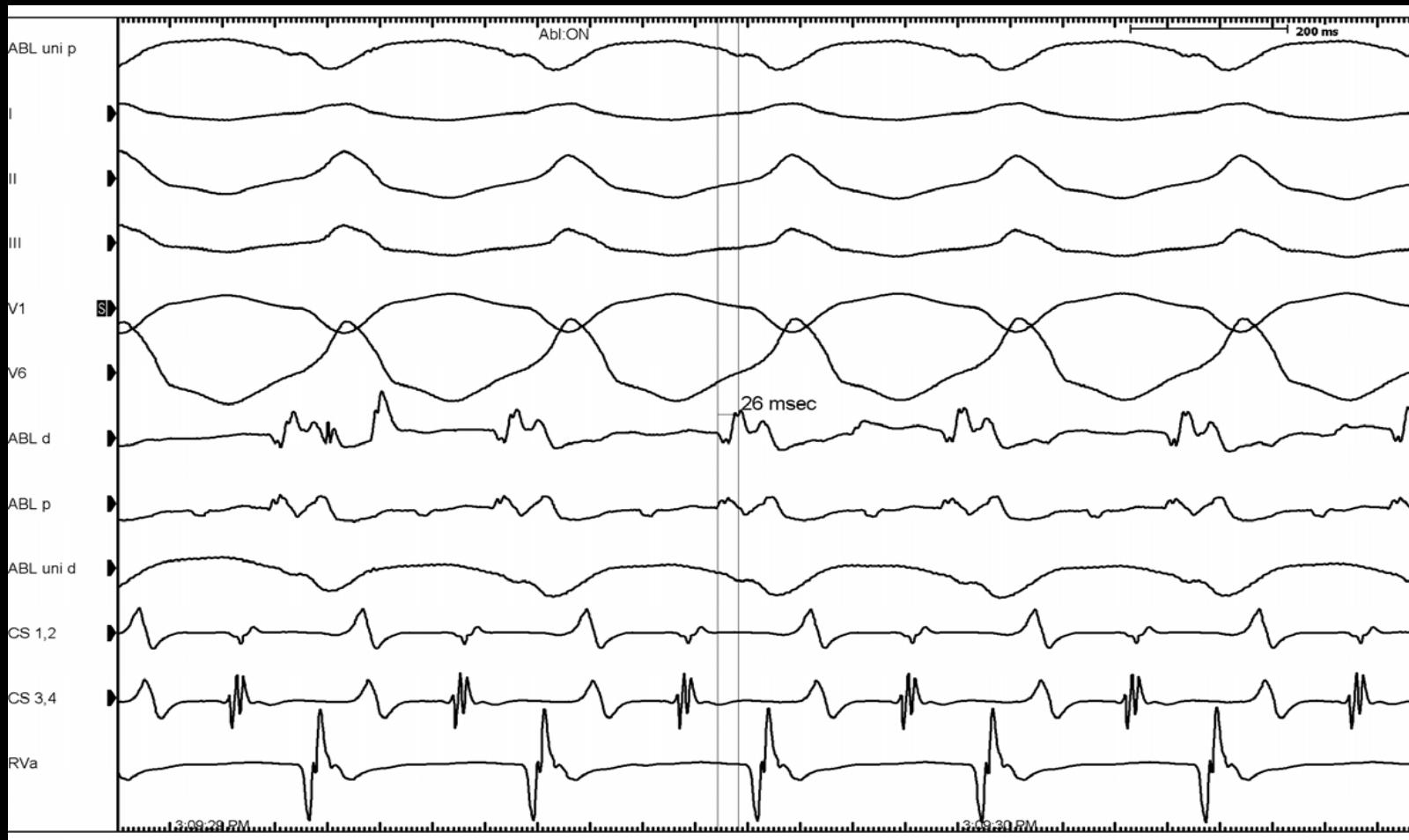
Paec map resembles VT1 from free wall



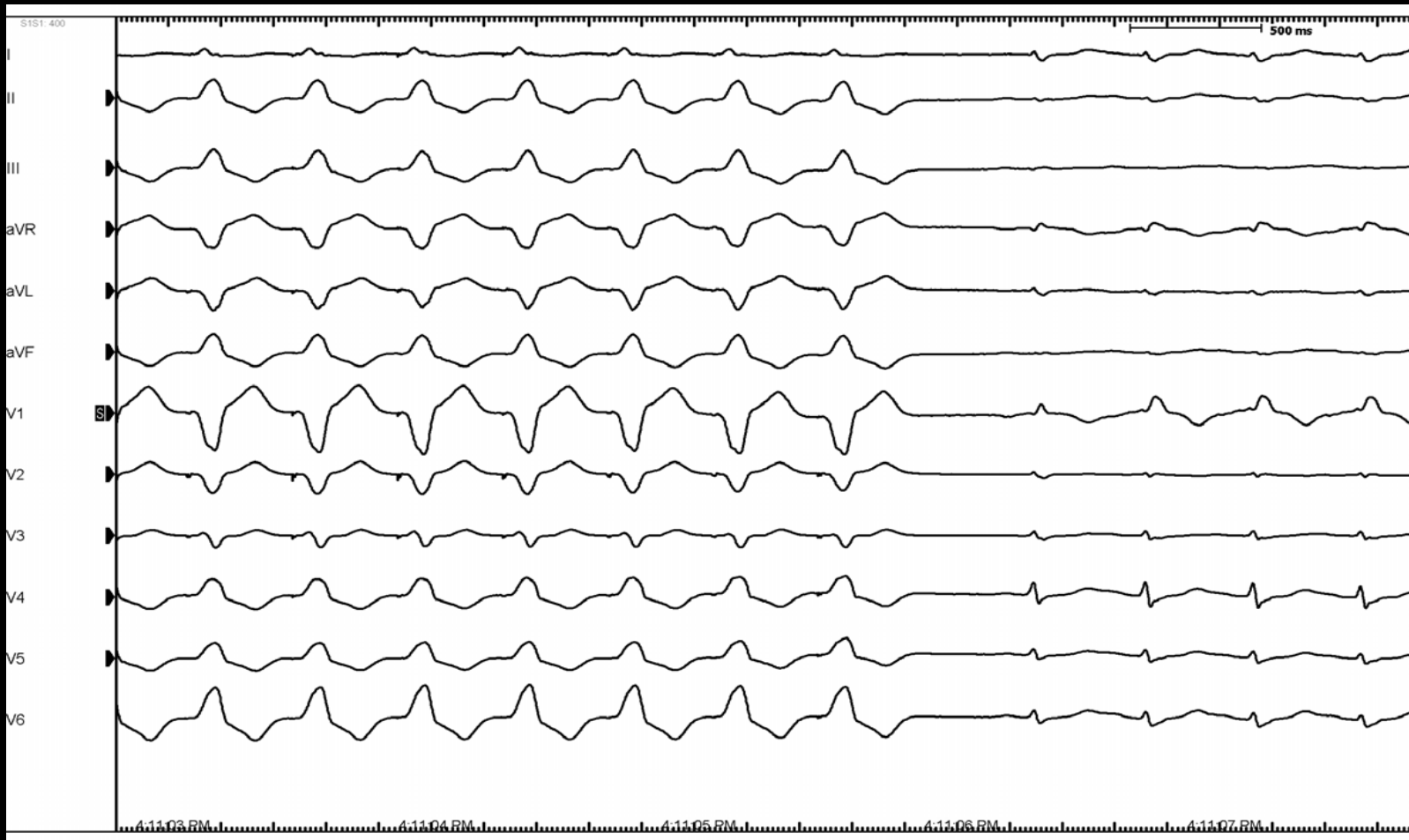
Vt1 ABLATION SITE



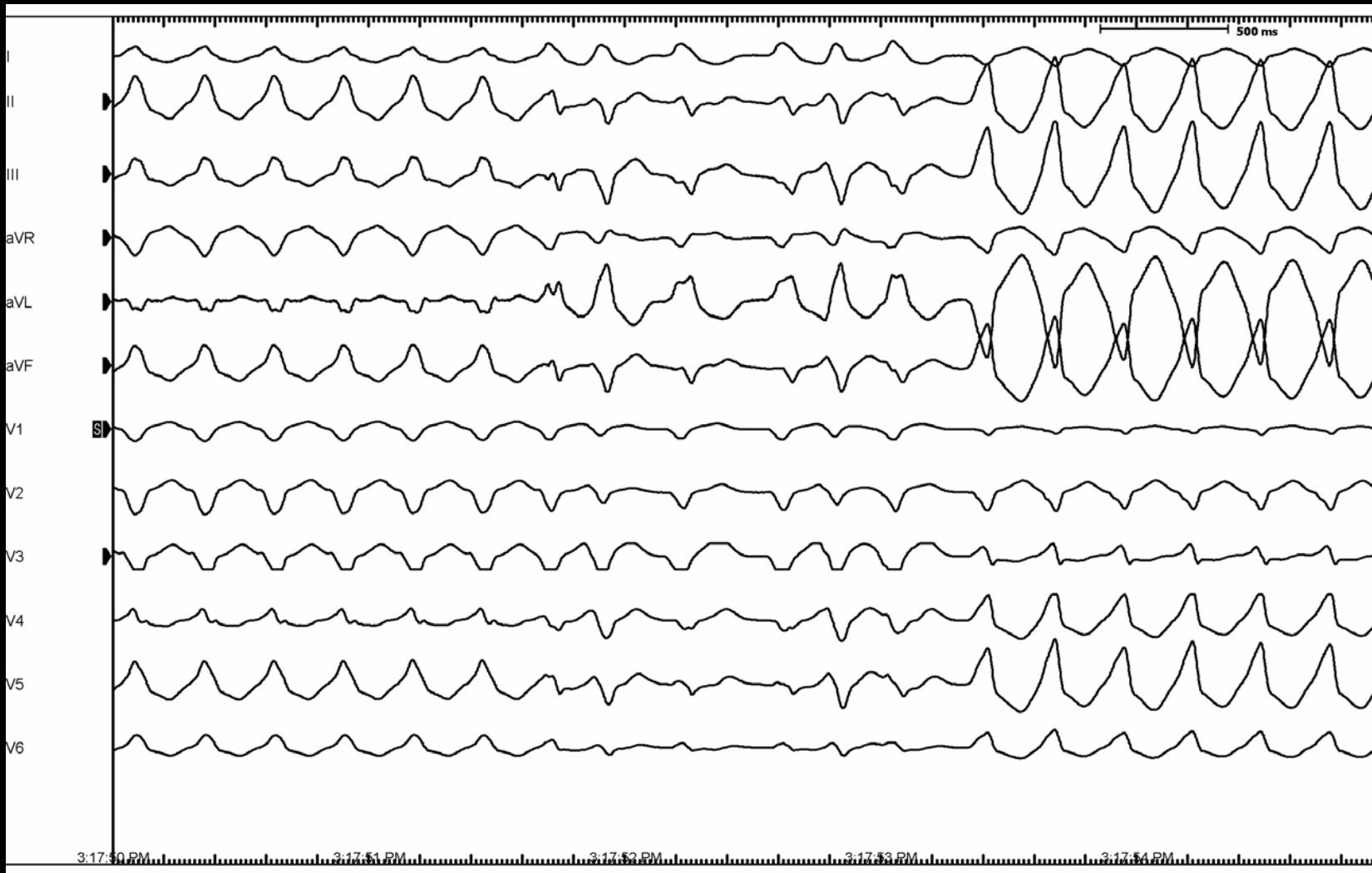
Another Site for VT1



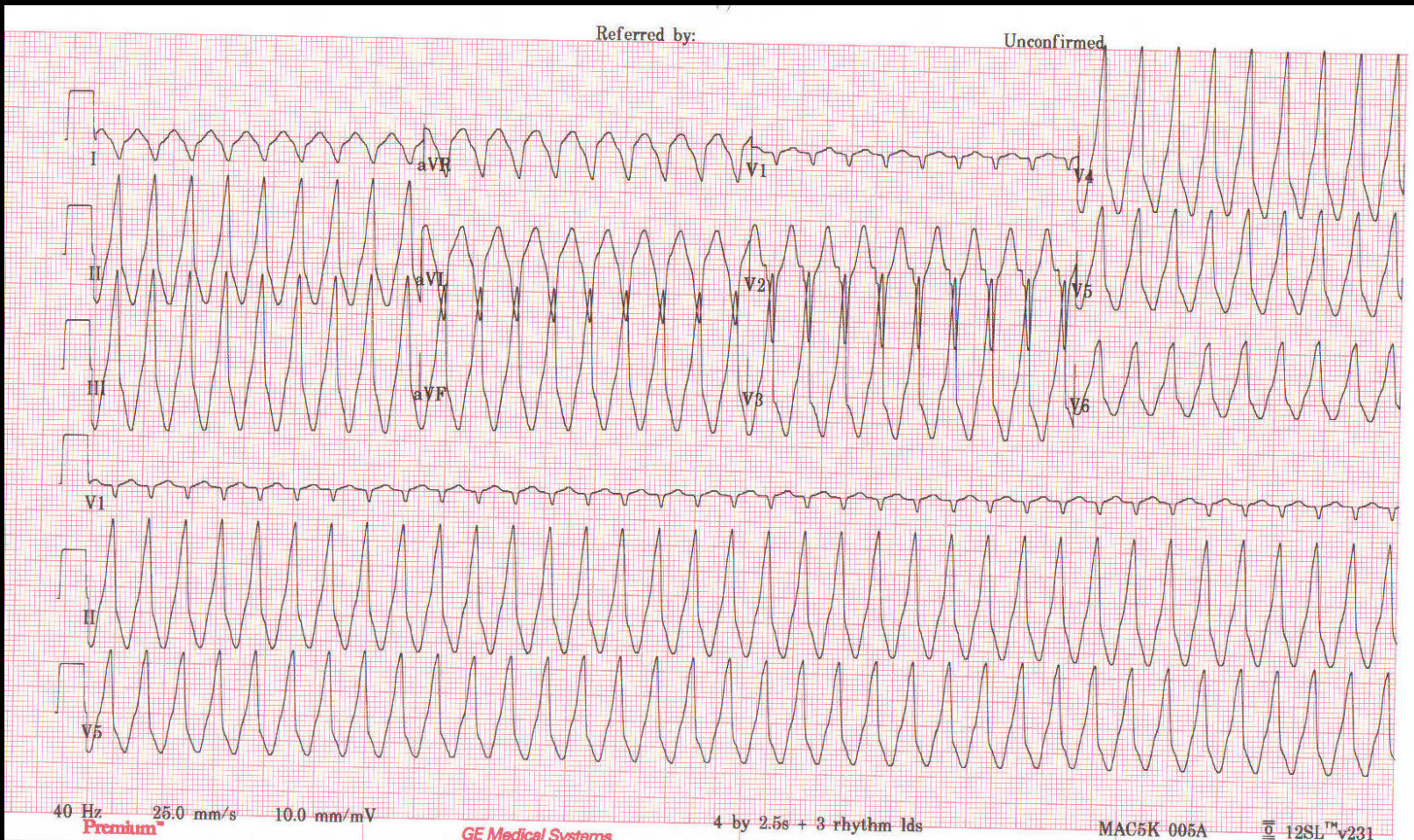
Pace map in Rt coronary Cusp Resembles VT1 ?

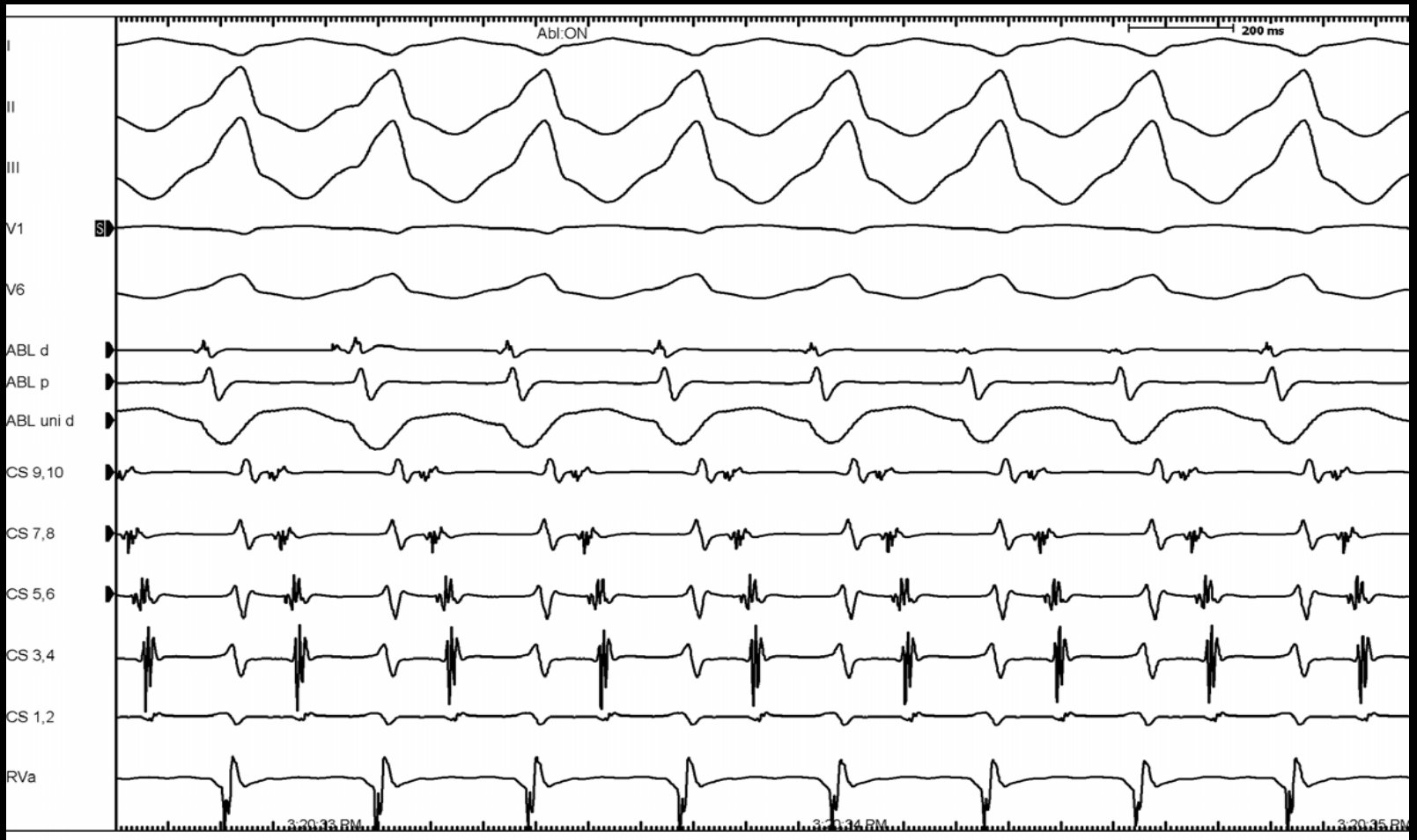


VT1 breaks with VPCs to VT2

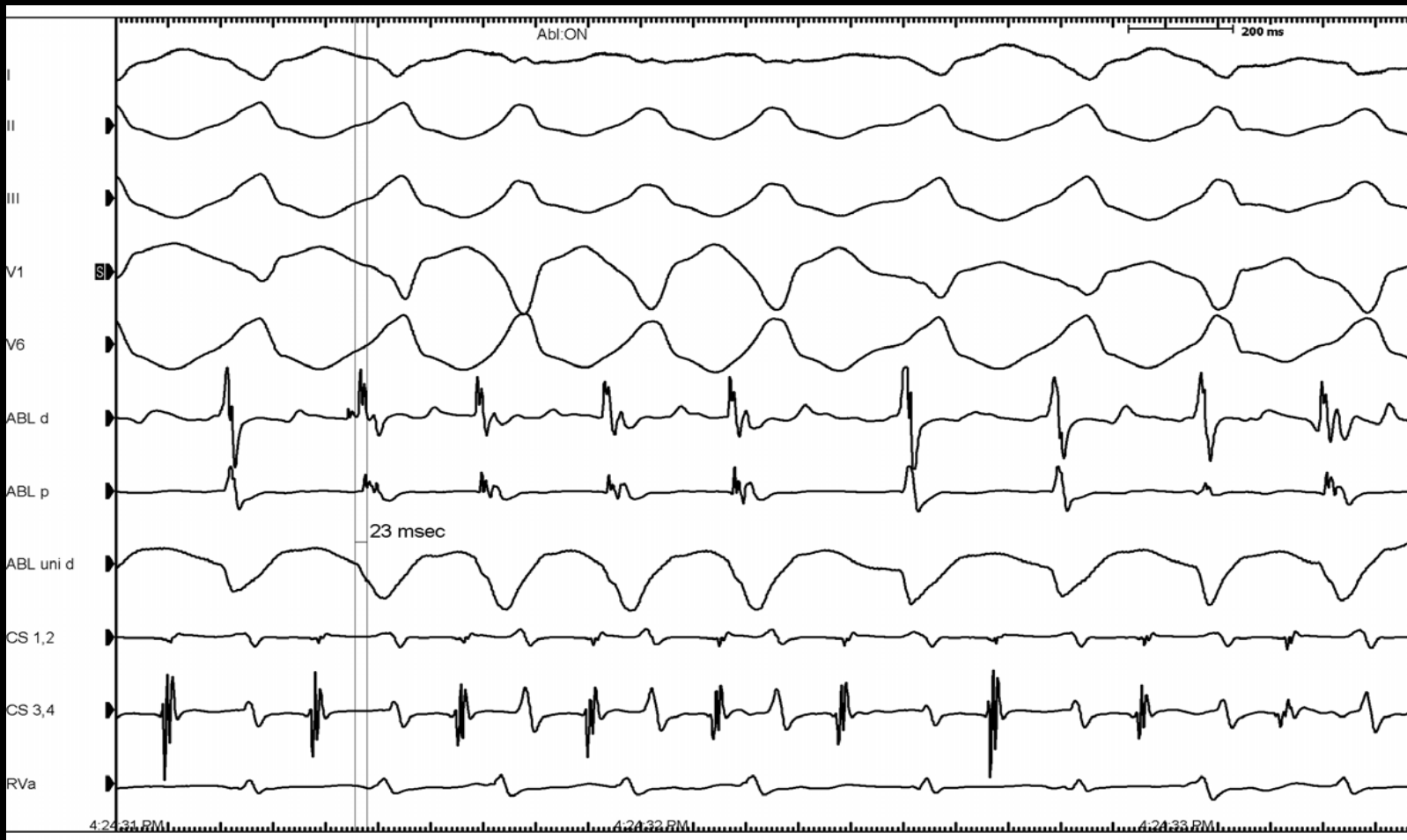


VT2- Antero-septal (rt cusp?)

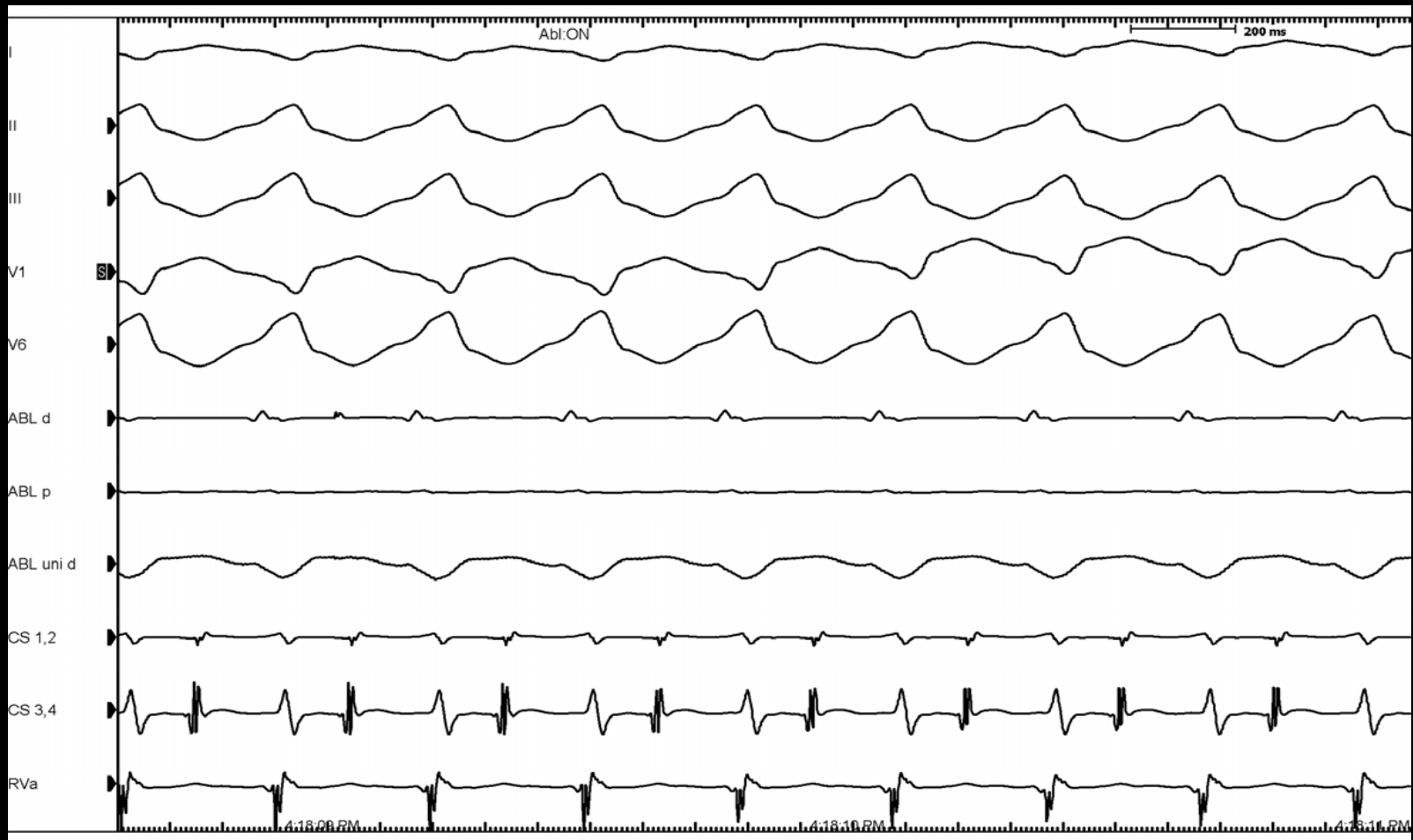




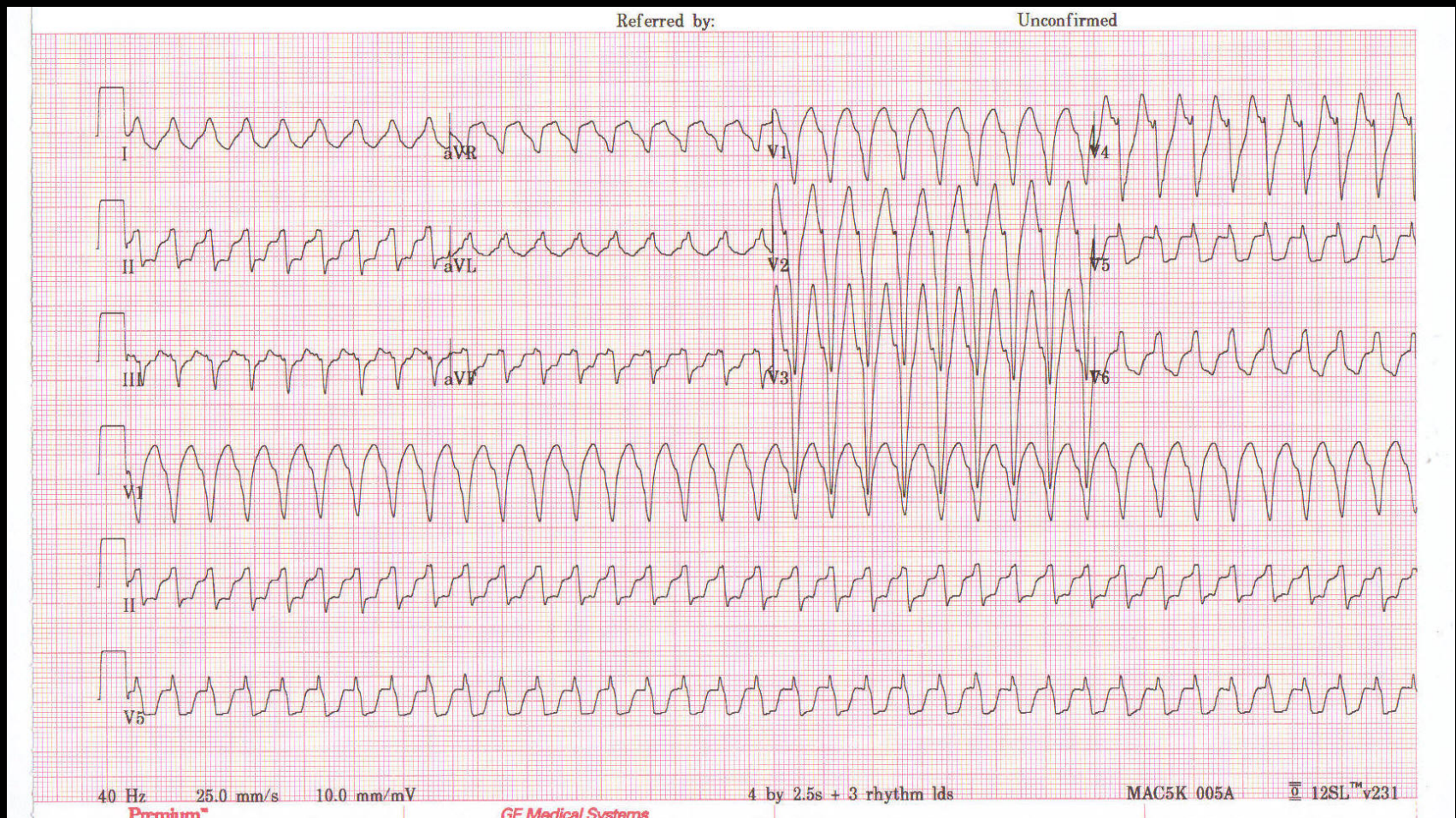
Ablation site in RVOT



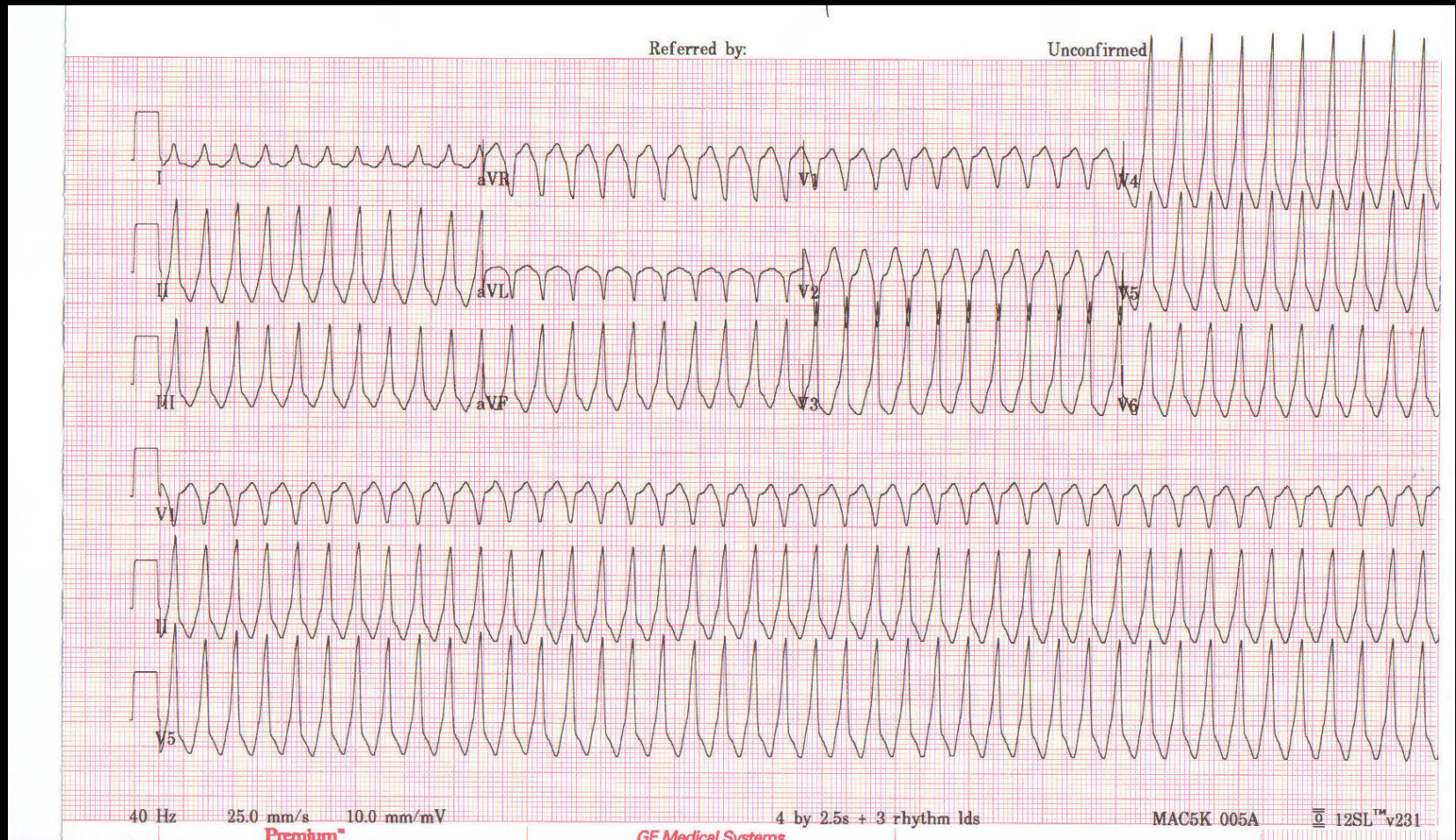
Ablation site in Rt. Coronary Cusp

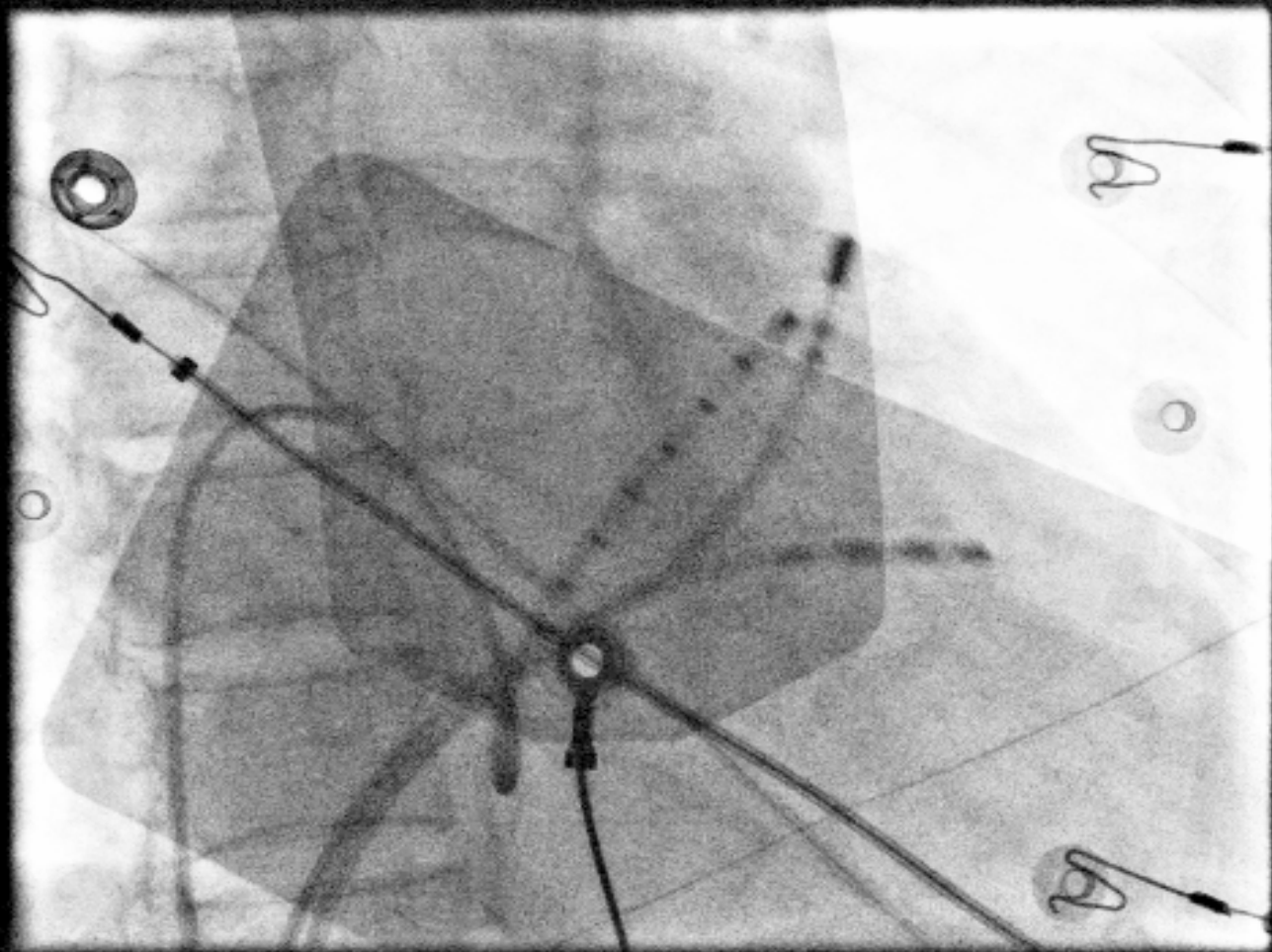


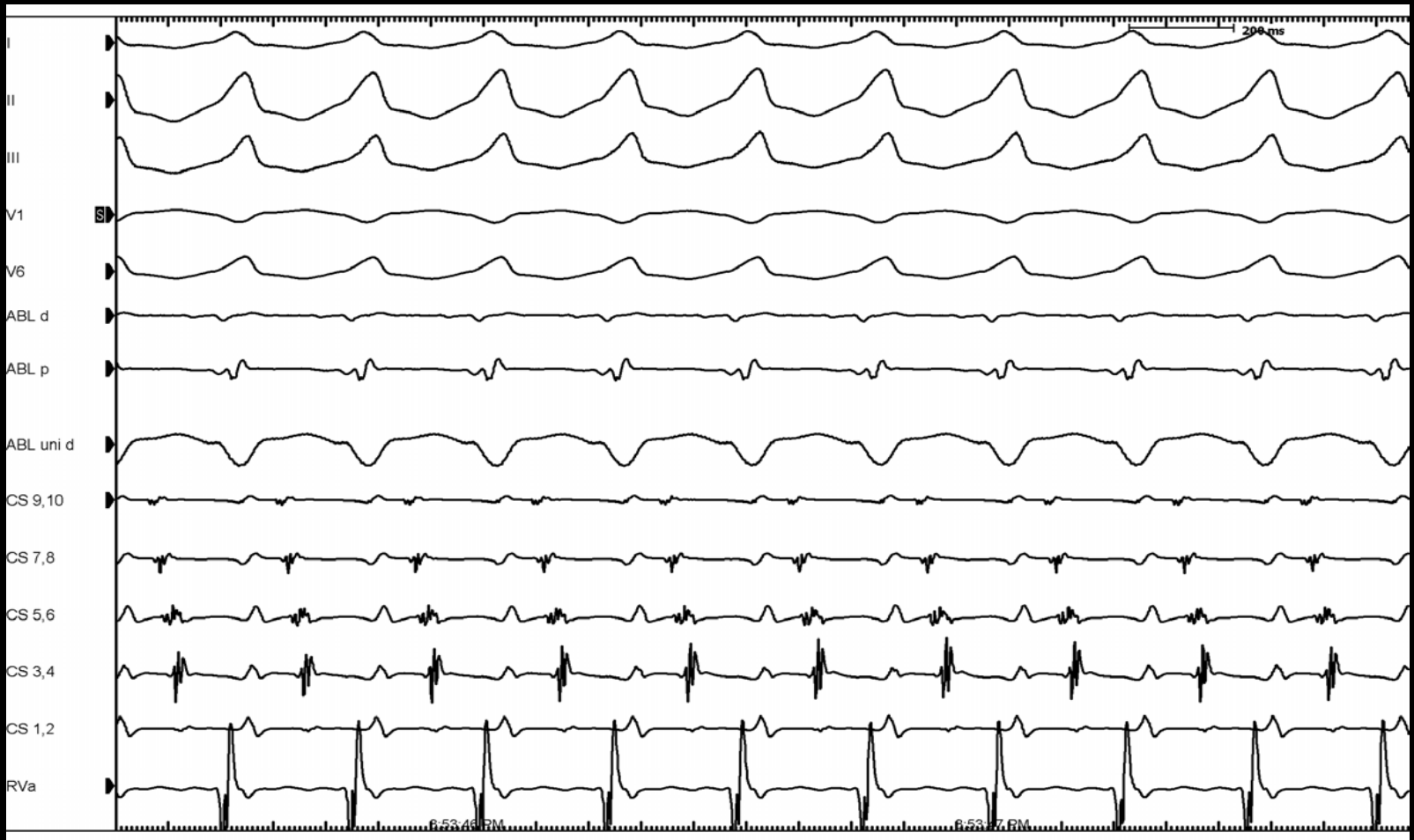
VT 3 inferior RV septal apex ?



VT4 Postero septal RVOT (Rt Cusp?)



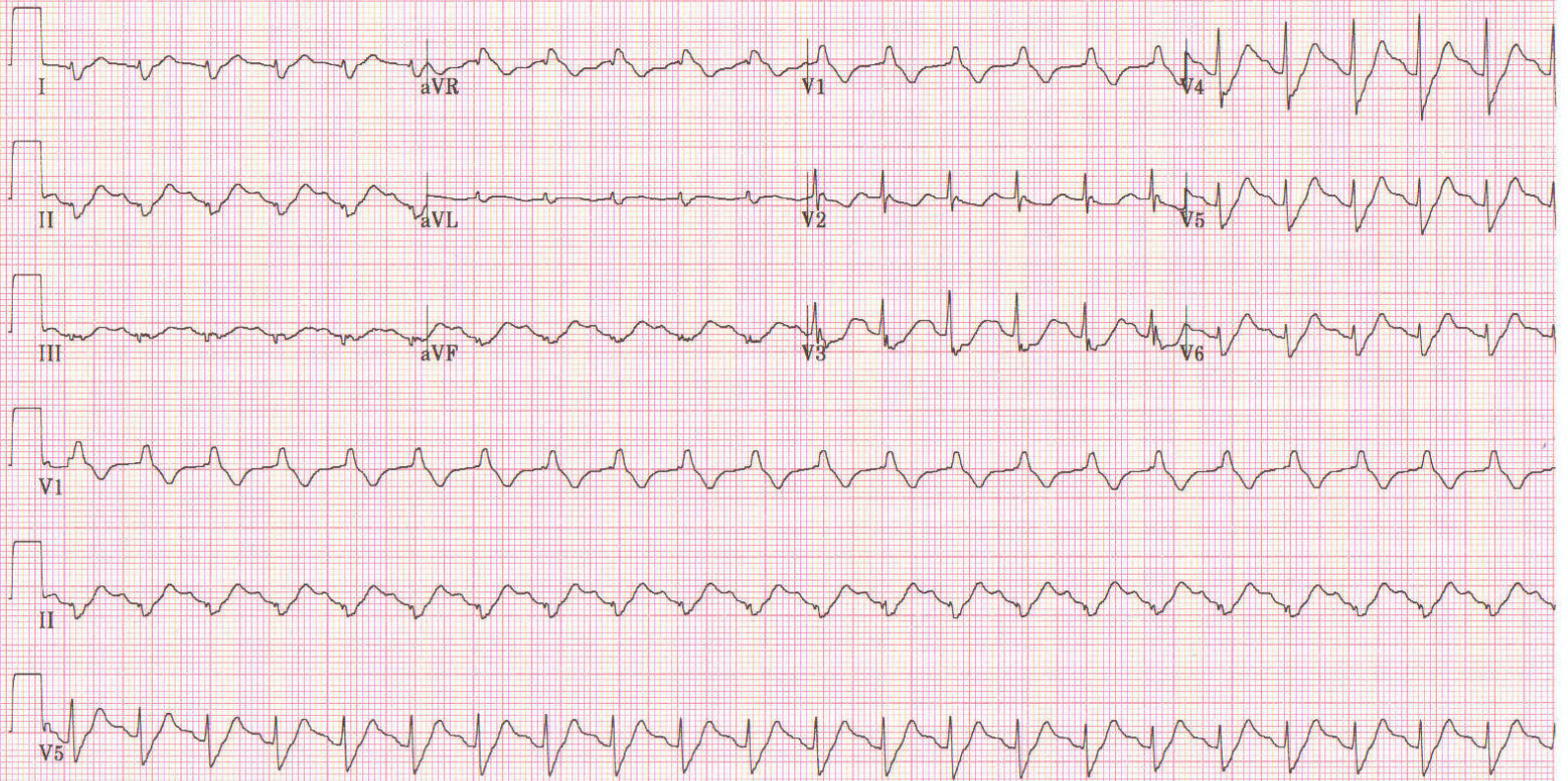




SVT

Referred by:

Unconfirmed



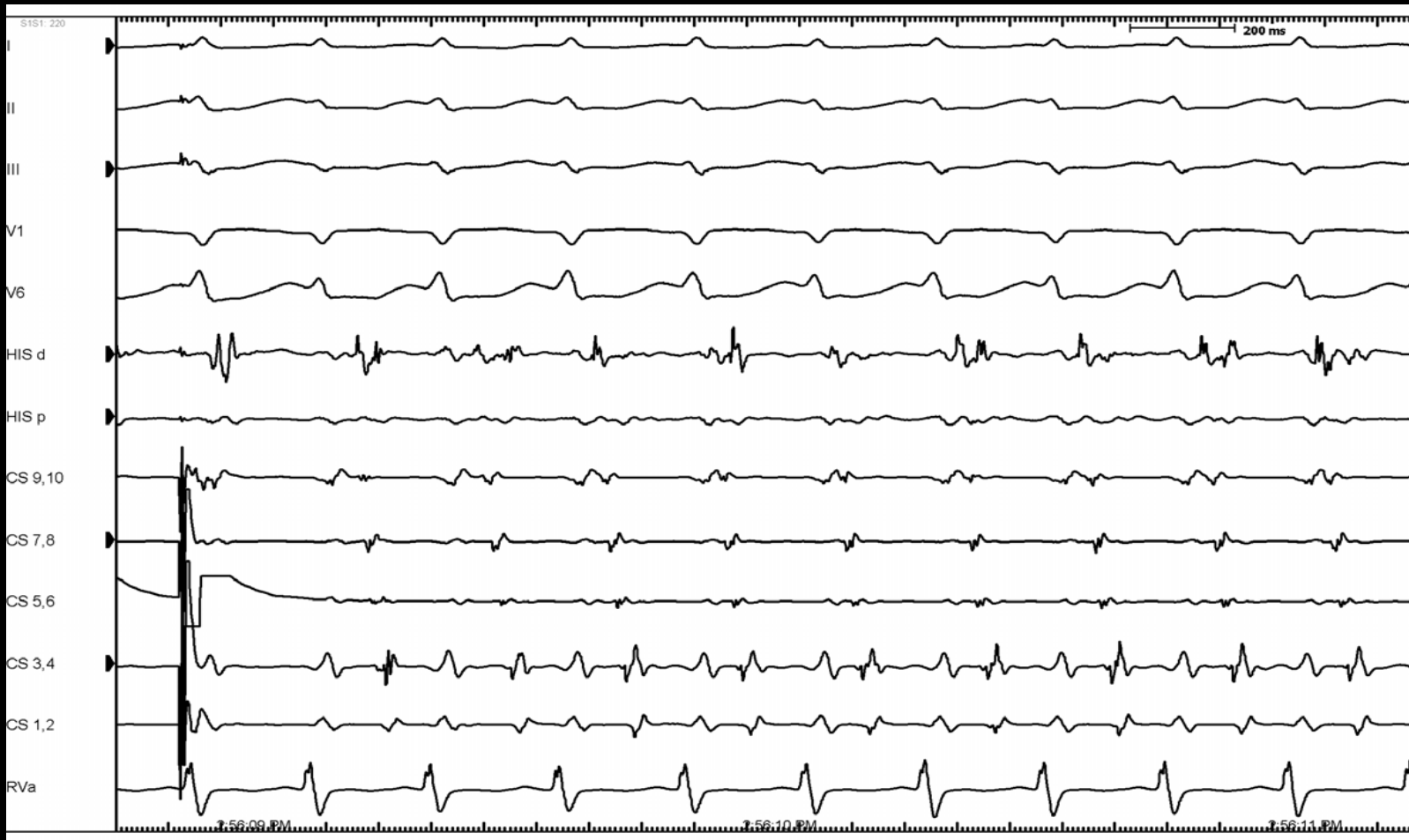
40 Hz 25.0 mm/s 10.0 mm/mV

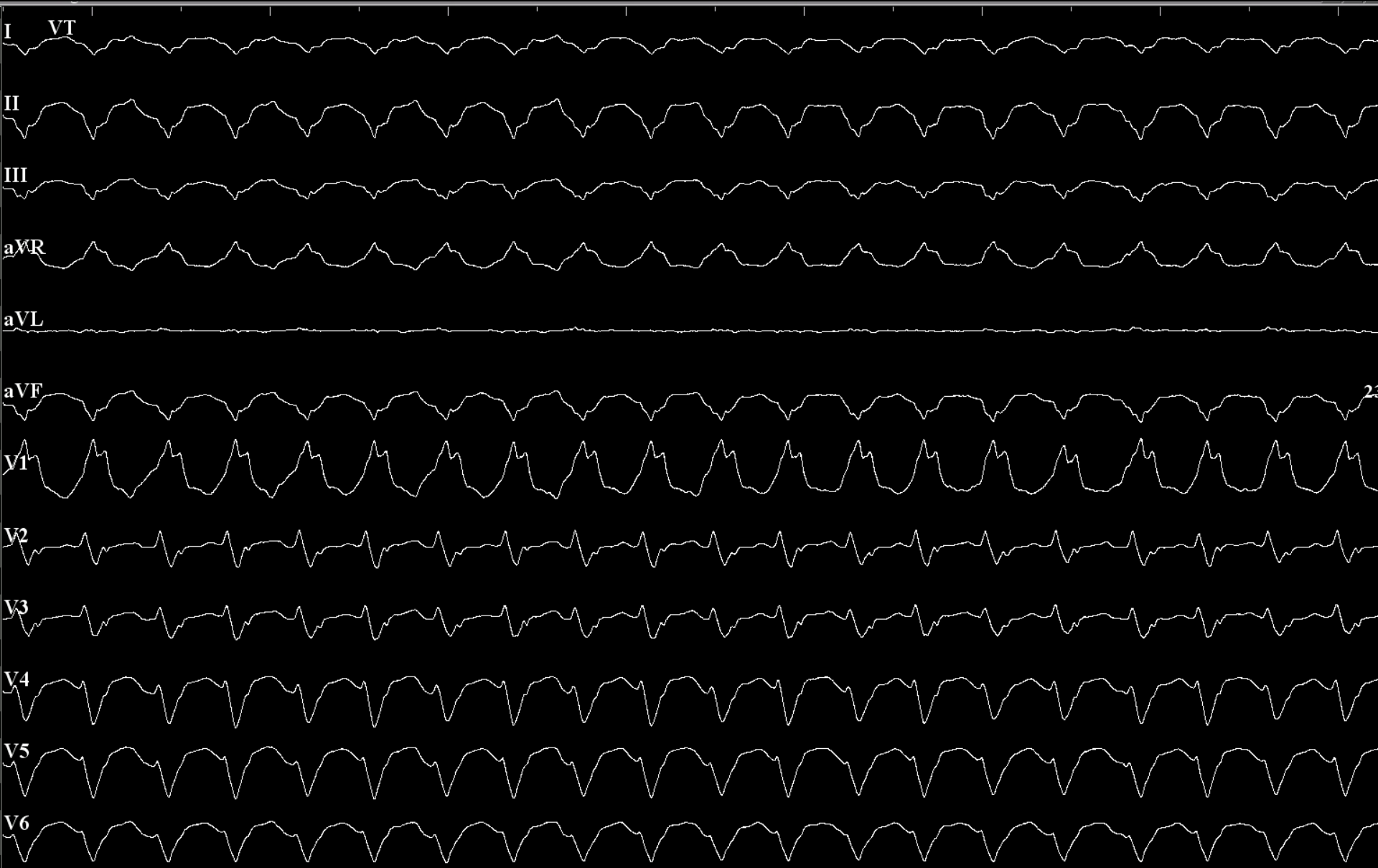
4 by 2.5 cm 2 abstr 1 de

MAC5K 005A

0-12SI™ 221

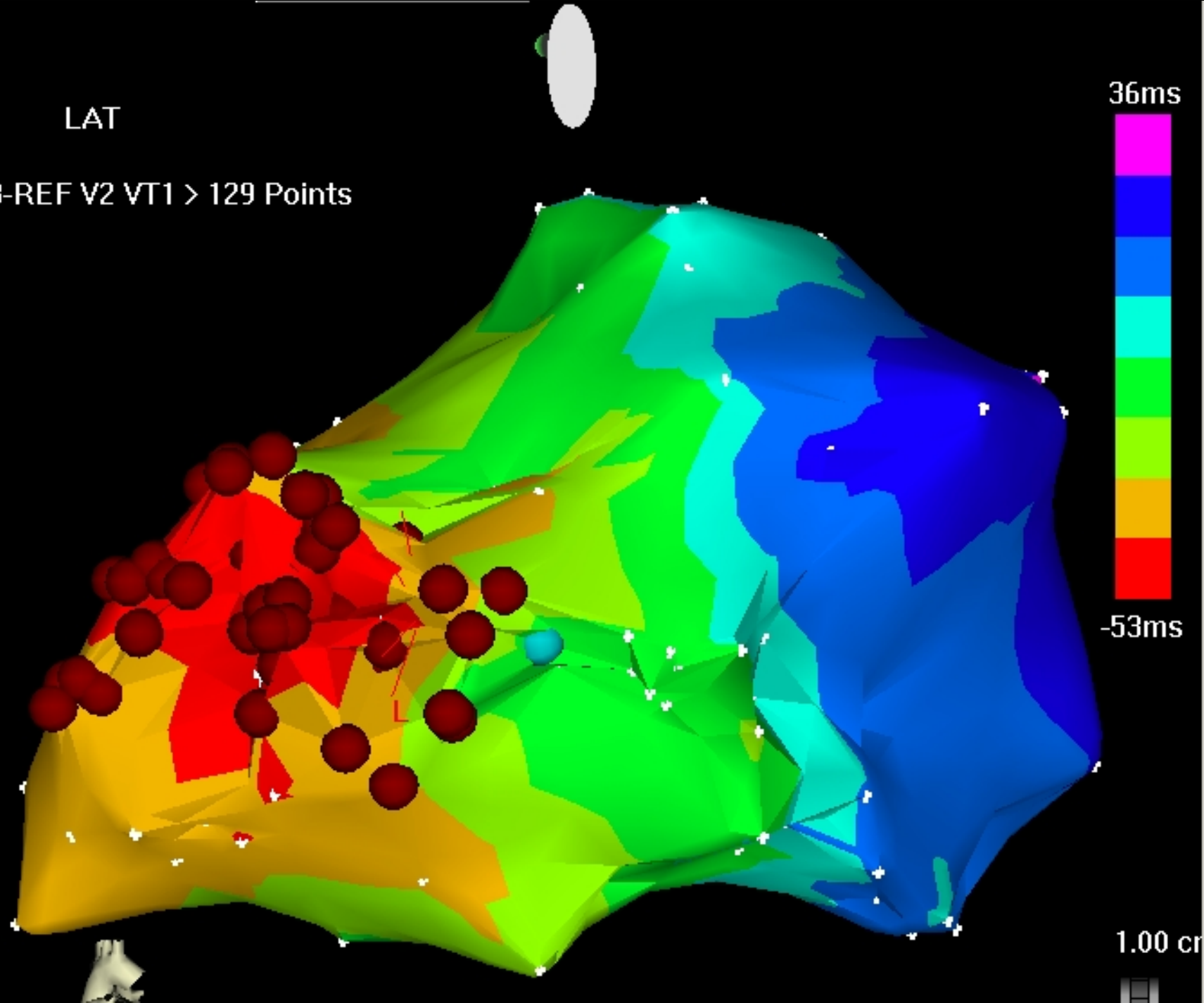
Induction of AVNRT





LAT

3-REF V2 VT1 > 129 Points



Breaks During Ablation

