Recurrent VT Referred for Heart Transplantation

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50 yo male referred for cardiac transplantation

- 1 year ago presented palpitations and runs of VT
  - Coronary angiography no obstructive disease
  - Cardiac MR:
    - Borderline dilated LV with LVEF 32%.
    - Multiple regional wall motion abnormalities in a noncoronary distribution. Extensive subepicardial LGE.
  - Diagnosis: NICM
  - Amiodarone started and ICD implanted
  - Meds: carvedilol, sacubitril-valsartan,furosemide
- 7 months ago: recurrent VT - substrate guided VT ablation
- recurrent episodes of VT despite amiodarone
  - referred for transplantation evaluation

Episodes Summary

Episodes Last Cleared	Feb 28, 2019 3:42 pm	Last Read	Mar 4, 2019 2:03 pm
SEGMs Last Cleared	Feb 28, 2019 3:42 pm		

Therapy Summary

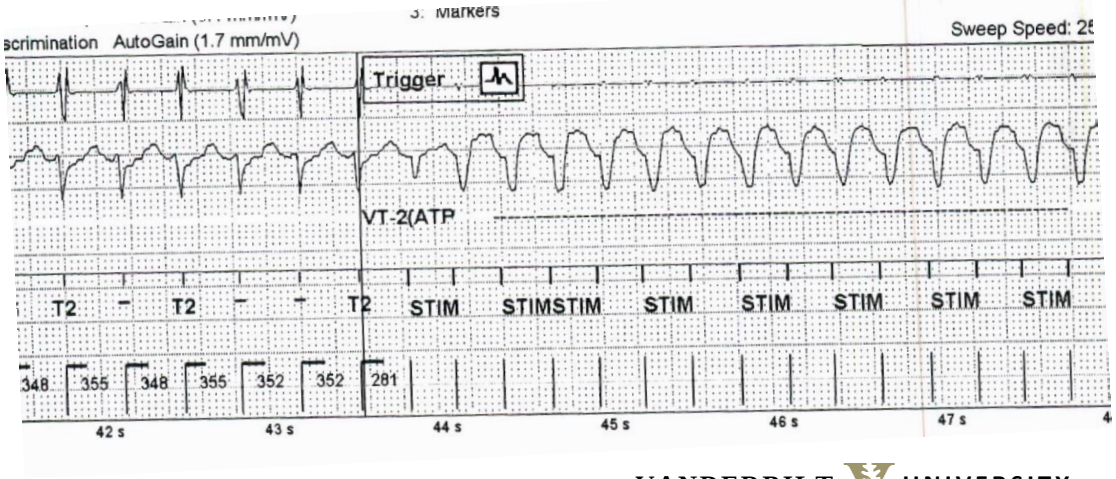
	VT-1	VT-2	VF
ATP Delivered		100	0
Shocks Delivered		0	0
Max Energy Shocks		0	0

VT-1 Zone is Monitor Only

Last HV Lead Impedance n/a

Results of ATP Delivery

	VT-1	VT-2	VF
Episodes Terminated	0	100	0
Episodes Not Terminated	0	4	0
Accelerations	0	0	



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*2019 HRS expert consensus statement on evaluation, risk stratification, and management of **arrhythmogenic cardiomyopathy***

J Towbin, W McKenna, et al

Arrhythmogenic cardiomyopathy (ACM):

an arrhythmogenic heart muscle disorder not explained by ischemic, hypertensive, or valvular heart disease.

- may present with atrial fibrillation, conduction disease, and/or right ventricular (RV) and/or left ventricular (LV) arrhythmia.

Etiologies include:

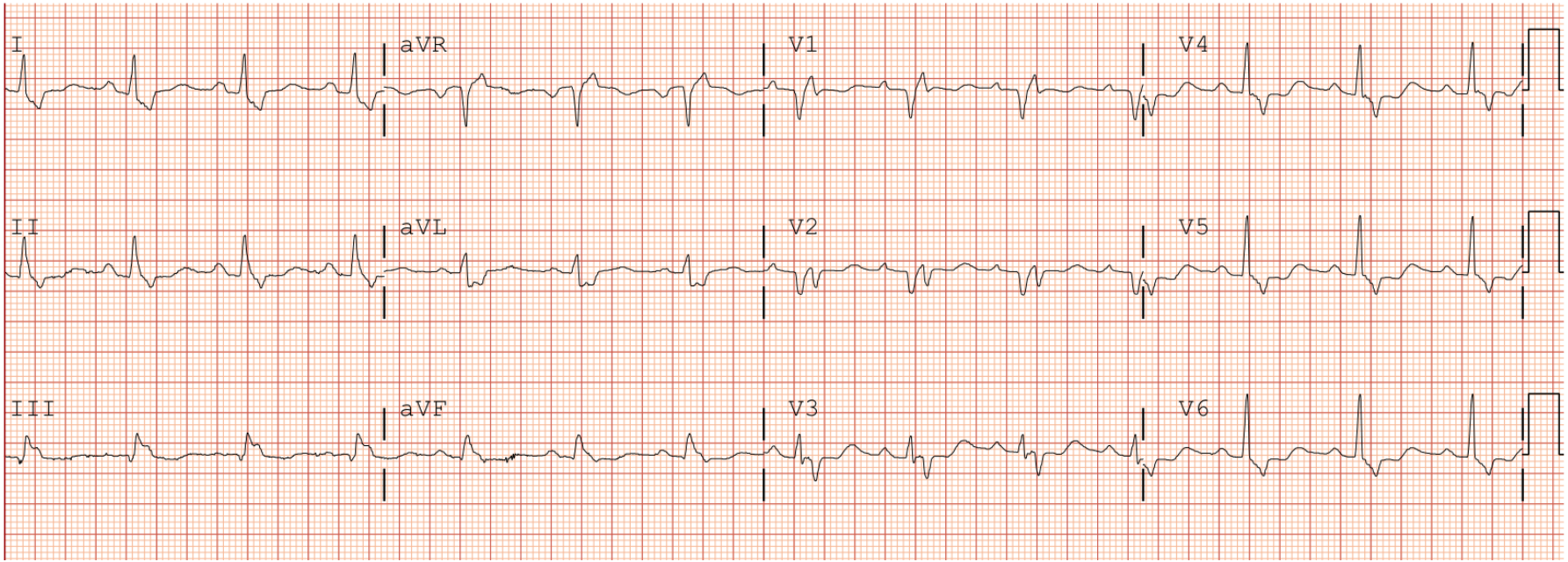
Genetic: 40% of NICM
Sarcoidosis
Amyloidosis
Chagas disease
Healed myocarditis

Heart Rhythm DOI: 10.1016/j.hrthm.2019.05.007

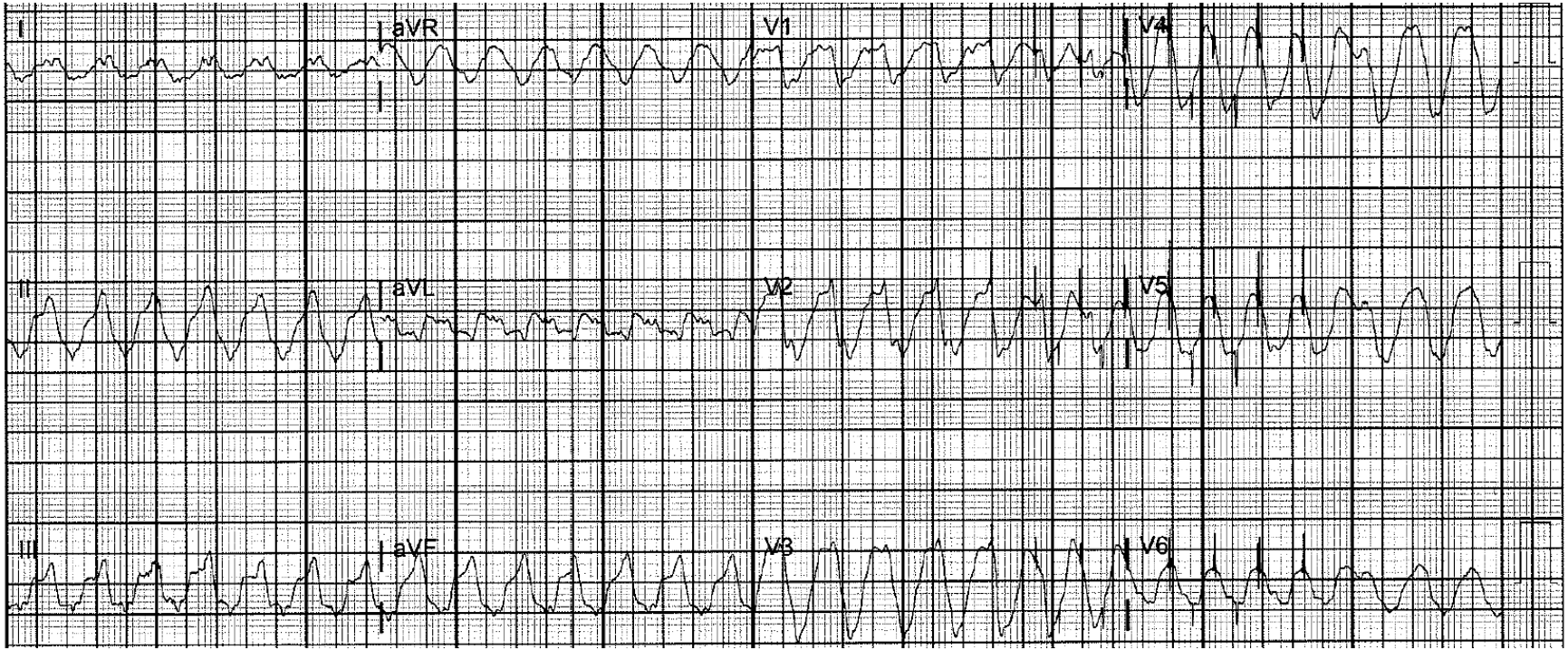
50 year old male referred for cardiac transplantation

- PMH:
  - hypertension, obesity (146 Kg)
- FH: no sudden death, cardiomyopathy
- Exam: unremarkable
- Transthoracic echo: LVEF 20 – 25%

50 year old male referred for cardiac transplantation



50 year old male referred for cardiac transplantation



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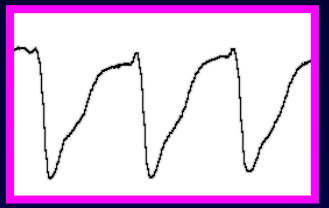
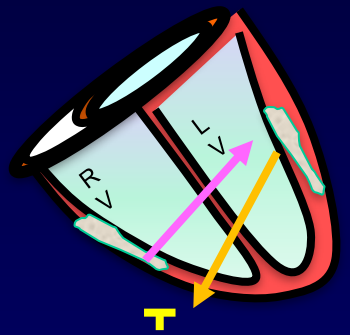
Monomorphic VT: QRS configuration suggests the origin and often the type of heart disease

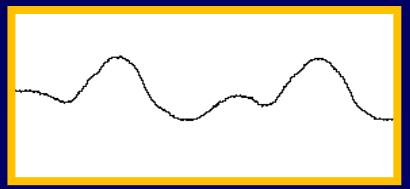
- **Left bundle branch block** VT configuration in V1
  - RV origin
    - Idiopathic outflow tract VT
    - bundle branch reentry
    - RV scar
      - Arrhythmogenic RV cardiomyopathy
      - Sarcoid
  - LV VT with a septal exit





V1

- **Right bundle branch block** VT configuration in V1
  - Usually LV origin



50 year old male referred for cardiac transplantation

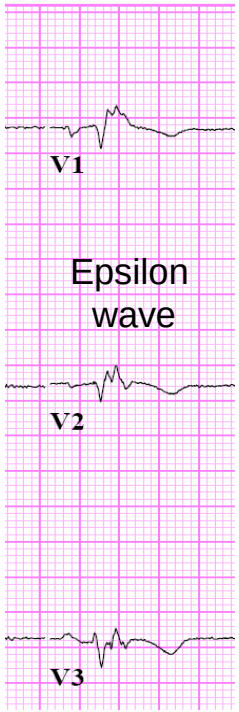
- PET scan for sarcoid:
  - Patchy LV FDG uptake sparing the lateral LV wall

- RV Biopsy at time of EPS
  - fibrosis with rare myocytes
  - no granulomas

Cardiac Sarcoidosis: Manifestations

- Conduction abnormalities
  - Most common cardiac manifestation
  - 19% of patients 18-55 yrs with complete heart block have sarcoid as the cause. (Kandolin et al, Circ AE 2011)
- Ventricular arrhythmias and sudden death
  - Reported in 23% of known cardiac sarcoidosis patients (Banba, Heart Rhythm 2007)
  - Sustained Monomorphic VT due to scar related reentry: LV, RV, septum
- Atrial arrhythmias
  - Less common (19%) than ventricular arrhythmias (Silverman Circulation 1978)
- MR imaging:
  - scar indicating delayed Gd in a noncoronary distribution with or without evidence of edema
  - Normal coronary perfusion

Sarcoid can Mimic ARVC



- Sarcoid is favored by:
  - Older age at presentation
  - PR prolongation or AV block
  - Septal delayed enhancement on MR imaging (or septal scar on mapping)
  - LV dysfunction
  - RV apical VT
  - Focal FDG uptake on PET

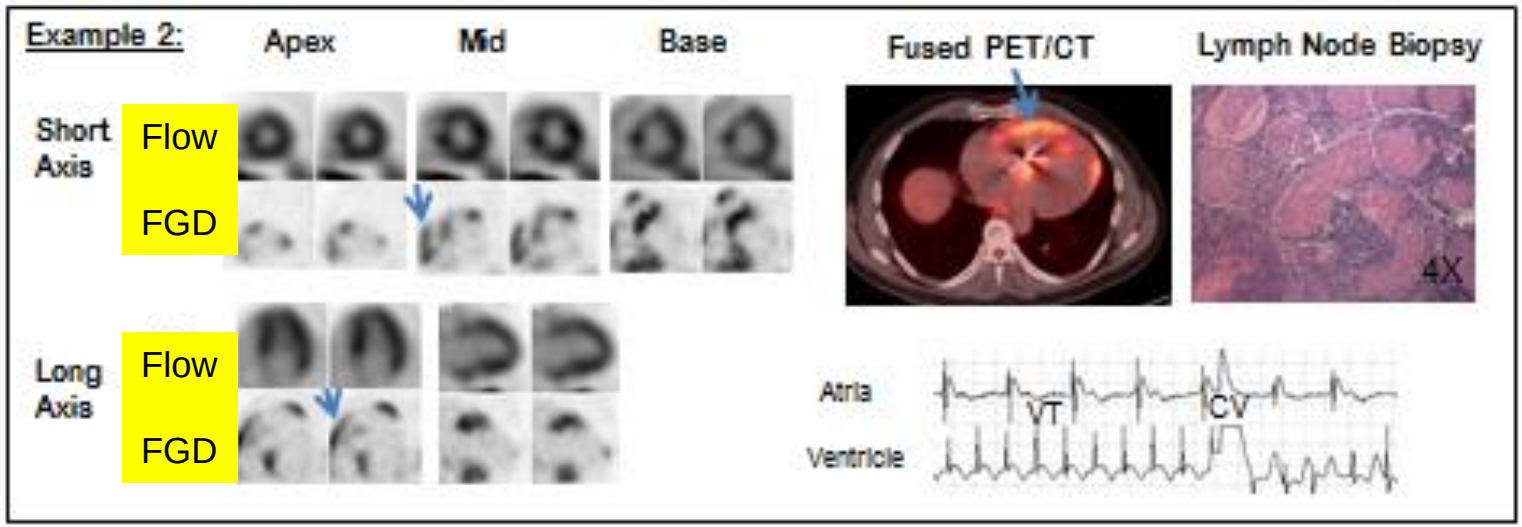
- ARVC is favored by family history of ARVC

Sarcoid mimicing ARVC

- 15 pts in Johns Hopkins ARVC registry found to have cardiac sarcoid - Philips et al Circ Arrh EP 2014
- 3 of 16 consecutive pts with LBBB VT suspected of having ARVC were found to have cardiac sarcoid - Vasaiwala et al JCE 2009
- 5 of 8 pts with cardiac sarcoid met TF criteria for ARVC - Dechering et al Heart Rhythm 2013

PET* in 118 patients with suspected cardiac sarcoidosis

- Areas of increased FDG uptake = inflammation or hibernation
- May coexist with diminished perfusion
- Noncoronary, RV distribution



* Cardiac PET for sarcoid must be preceded by fatty diet to suppress myocardial glucose uptake

Blankstein et al JACC 2014;63:329

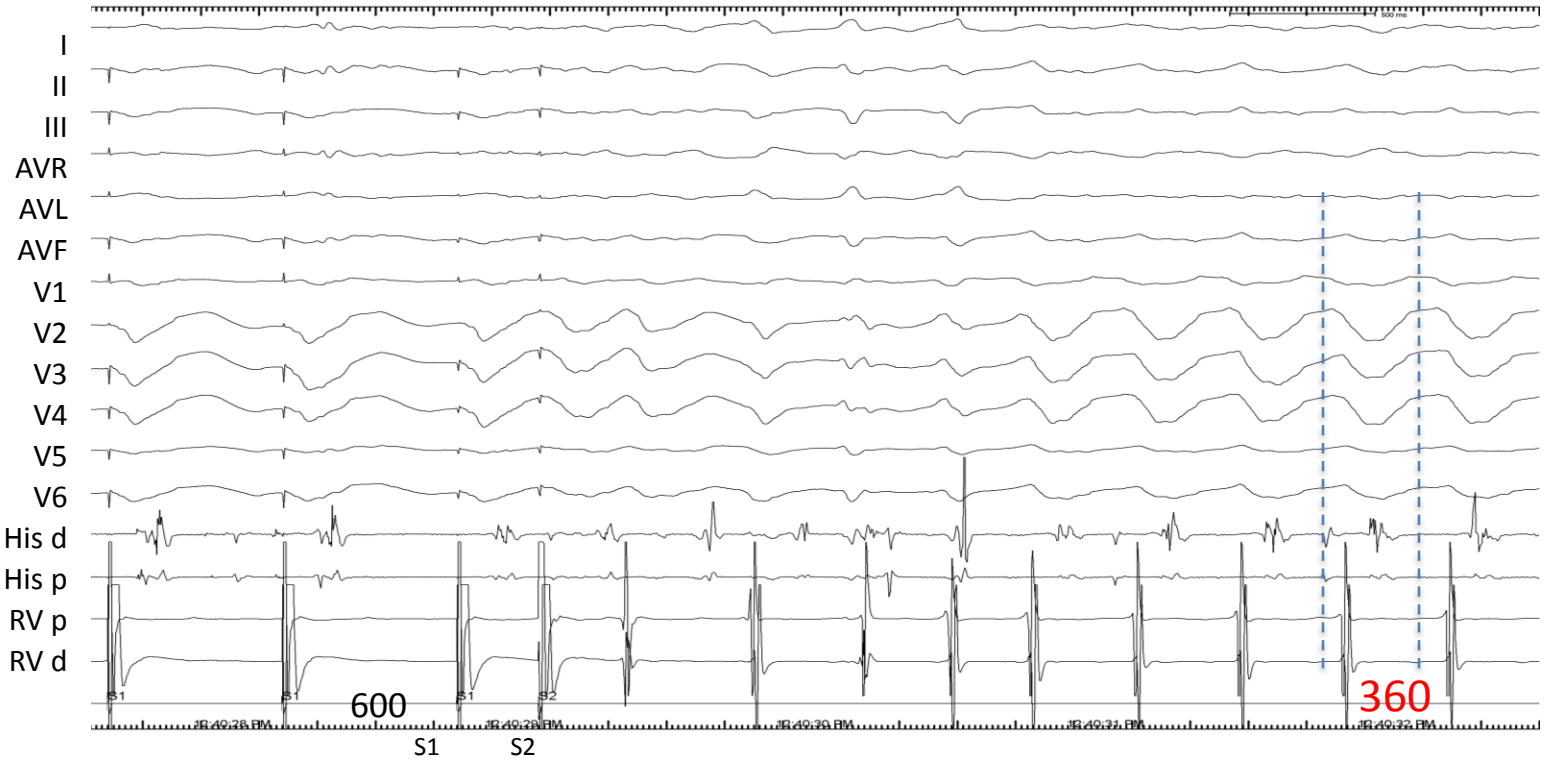
Managing Arrhythmias

- AV block may respond to immunosuppressive therapy
- PVCs and nonsustained VT – response to immunosuppression is variable
- Sustained monomophic VT usually is scar related reentry
  - the Purkinje system can be involved
  - immunosuppression does not usually prevent arrhythmias, although may have an impact, perhaps by reducing triggers
  - VT sometimes flares after initiation of immunosuppression

Approach to patients with multiple VTs and diffuse substrate

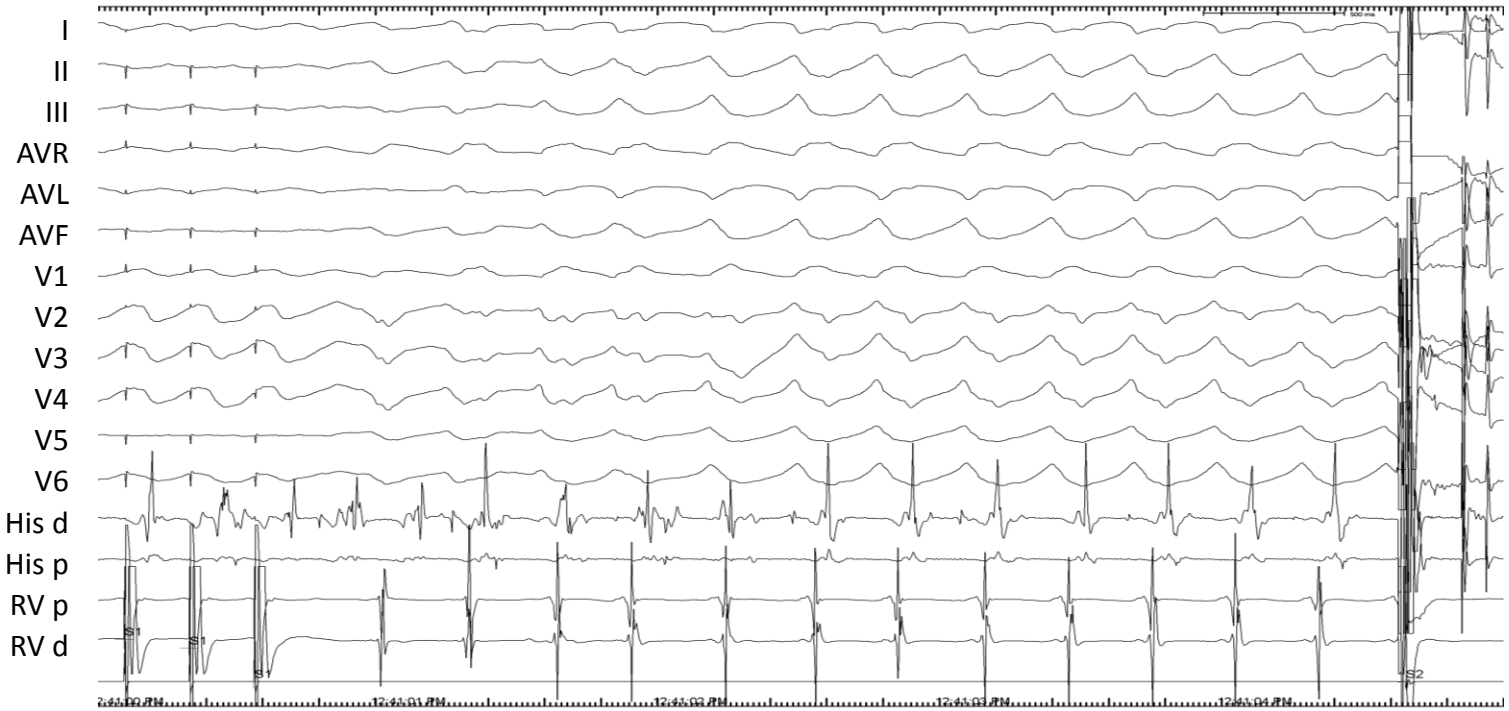
- Define the disease
- Identify the QRS morphology of spontaneously occurring and inducible VTs
- Define the substrate and regions associated with VT
- First ablation areas: target the scar areas that likely include the VT exit based on QRS morphology

EP Study: Initiation of VT1 by 600/1



Morphology stabilized, but difficult to interpret: RV mid sept earlier than basal septum

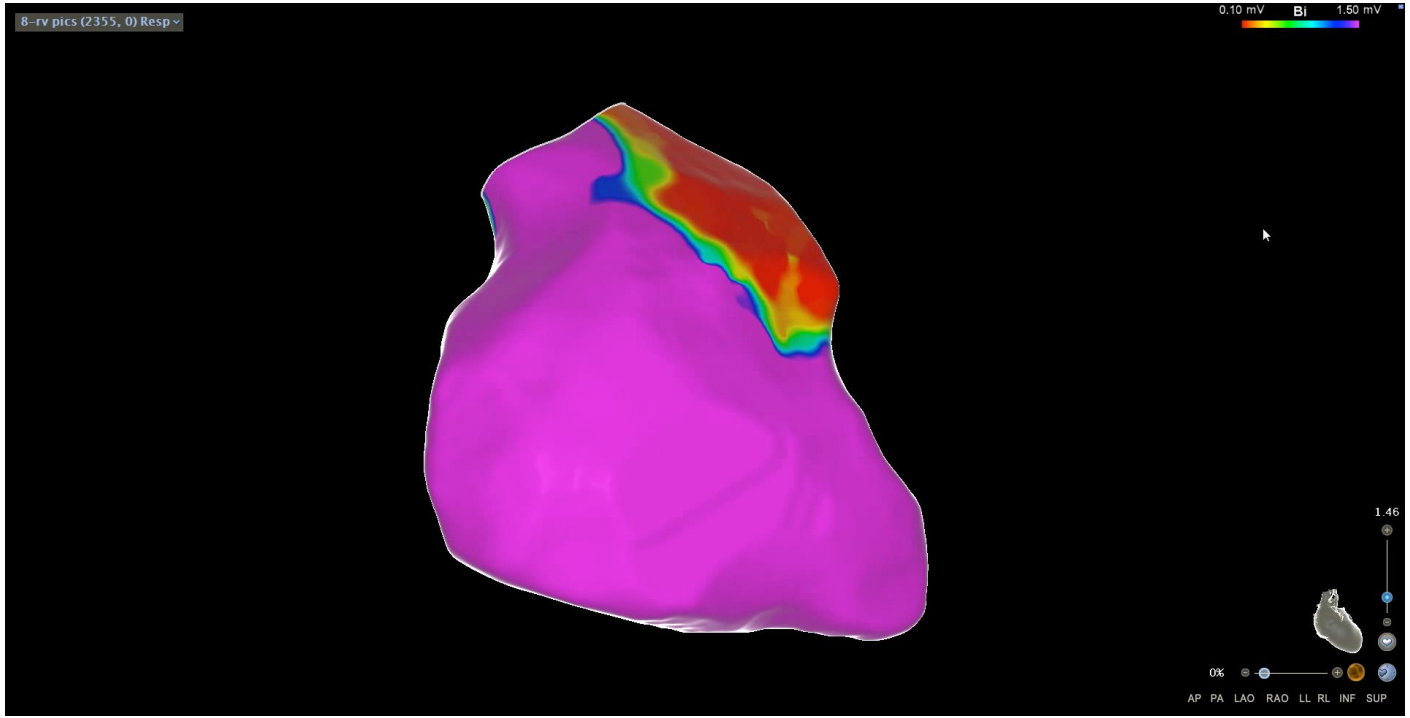
Burst pacing in VT 1 accelerates VT: cardioversion



VT2: induced by catheter manipulation

VT2: probably RB, inferior axis morphology; RV septum late

RV Voltage map: 0.1 – 1.5 mV



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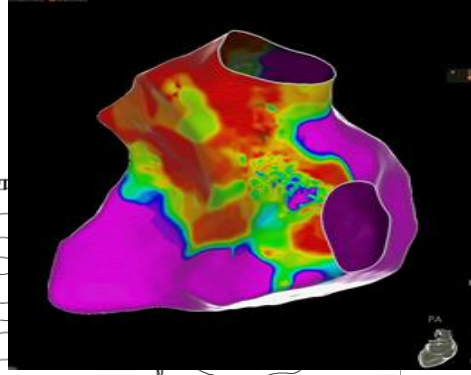
low voltage areas with split potentials in the RV basal septum



I
II
III
AVR
AVL
AVF
V1
V2
V3
V4
V5
V6
Map 1,2
Map 3,4
Map 5,6
Map 7,8
Map 9,10
RA
RV sept d
RV sept

S1
S1
S1

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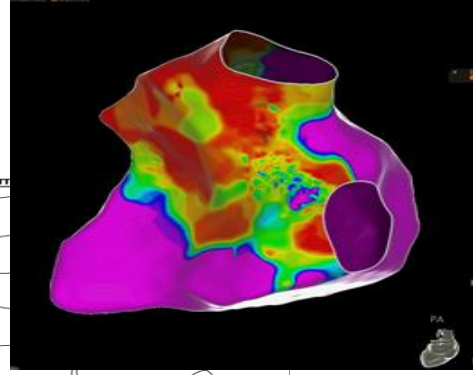
RV OT low amplitude electrograms pacing captures with S-QRS delay



I
II
III
AVR
AVL
AVF
V1
V2
V3
V4
V5
V6
Map 1,2
Map 3,4
Map 5,6
Map 7,8
Map 9,10
RV sept d
RV sept

S1 S1 S1

1:04:04 PM 1:04:05 PM 1:04:06 PM

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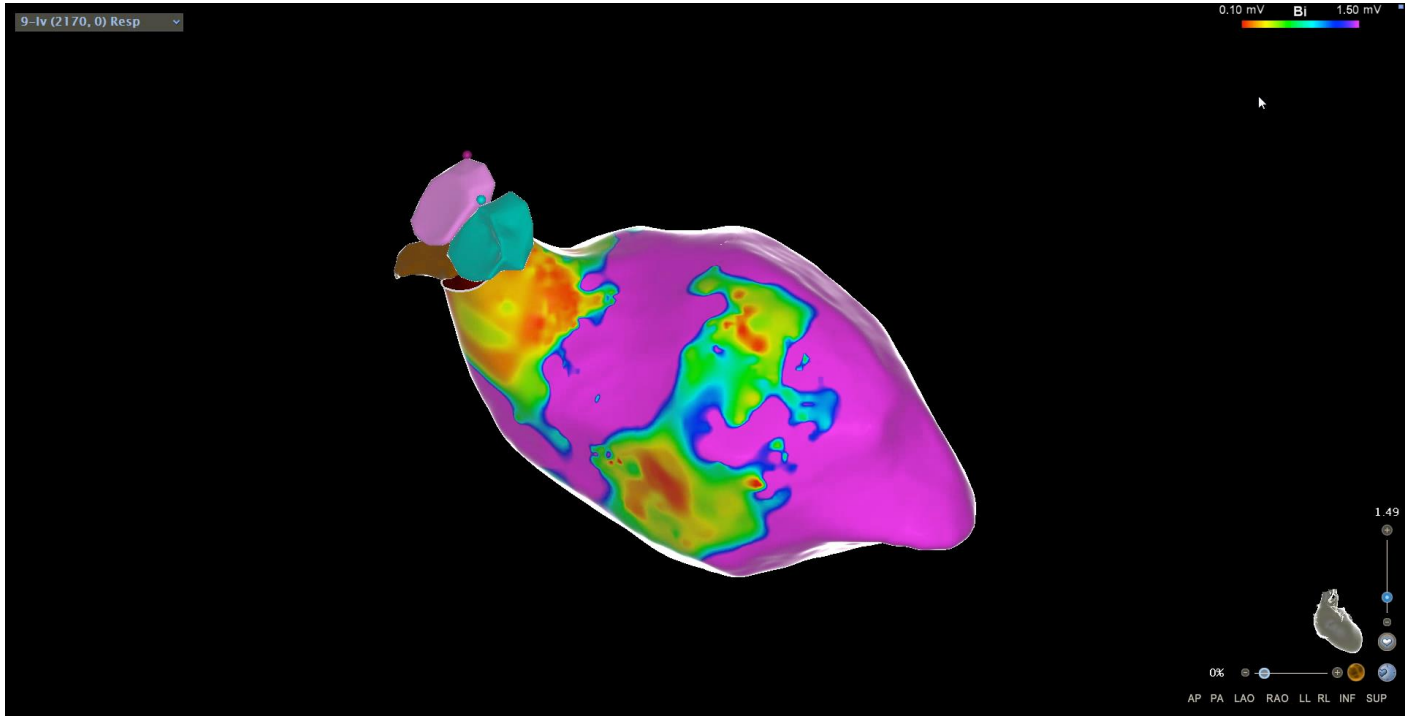
LV Voltage Map (0.1 – 1.5 mV)



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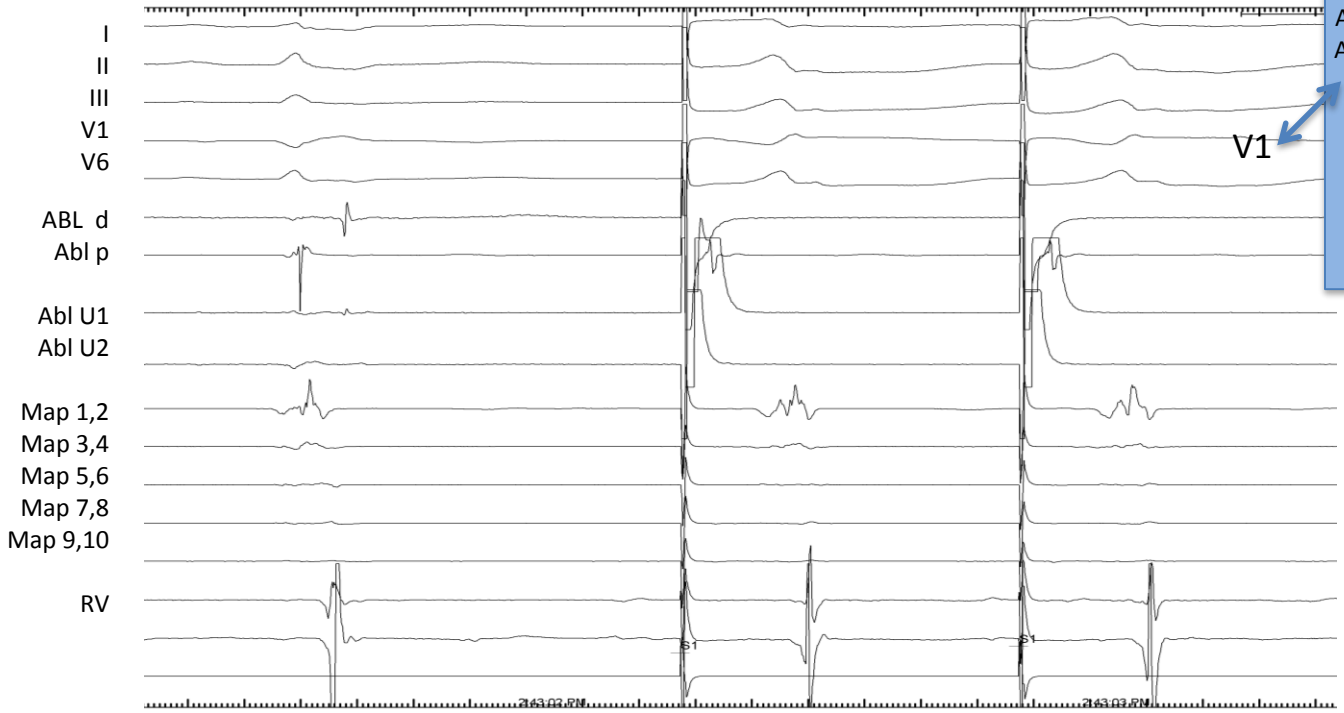
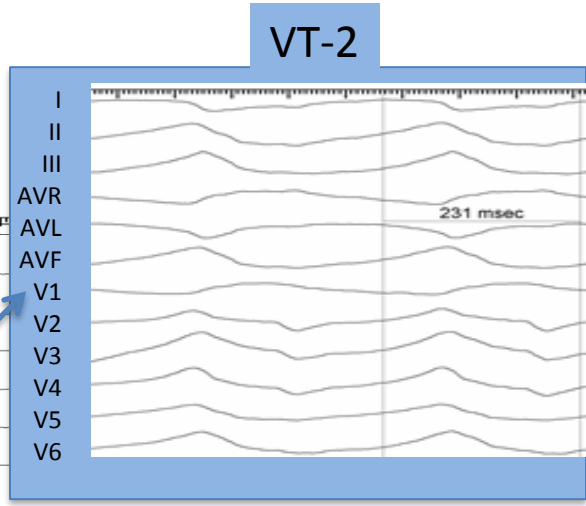
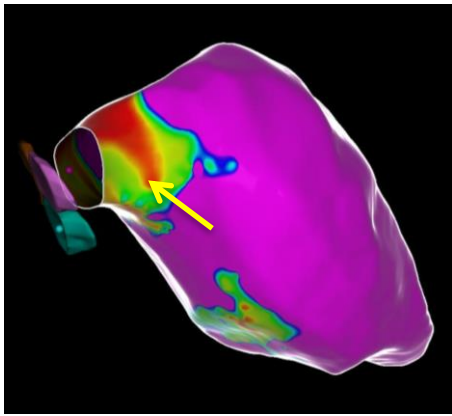
Below aortic valve: abnormal electrograms
S-QRS delay with pacing







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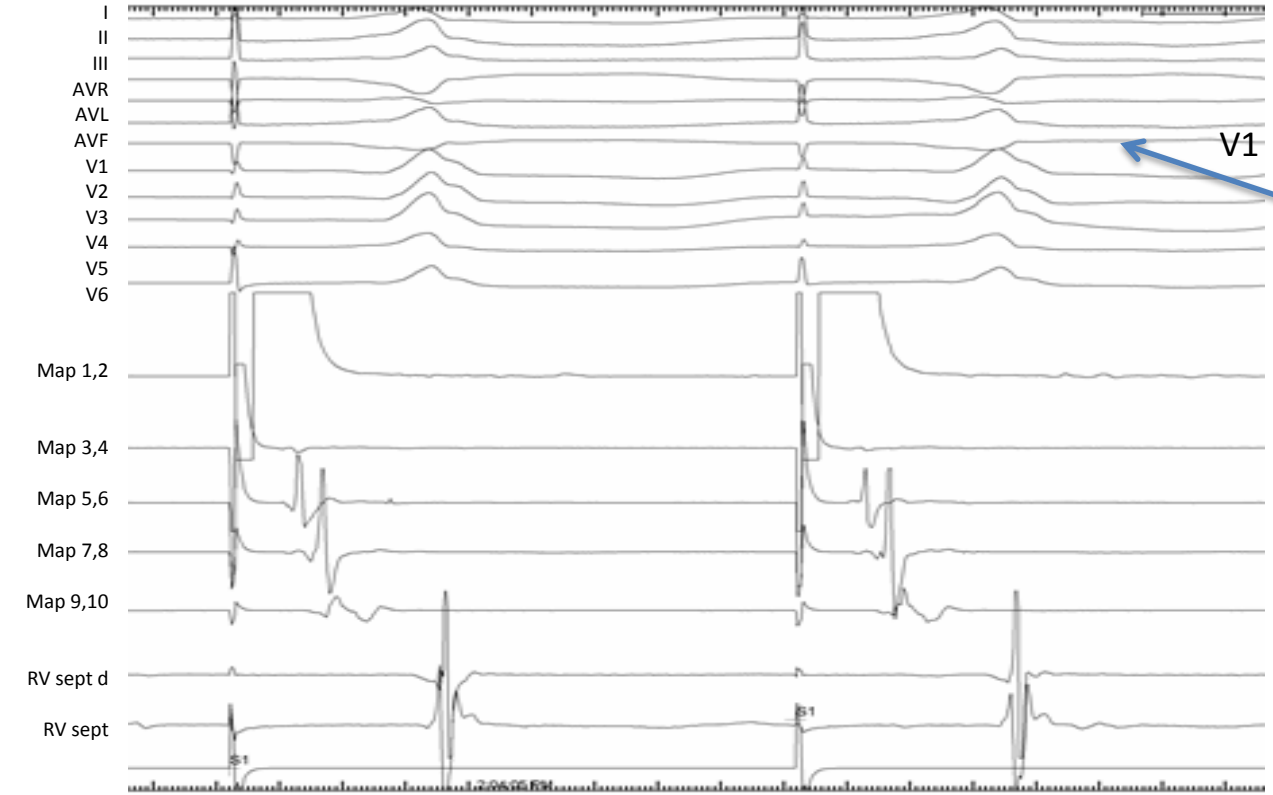
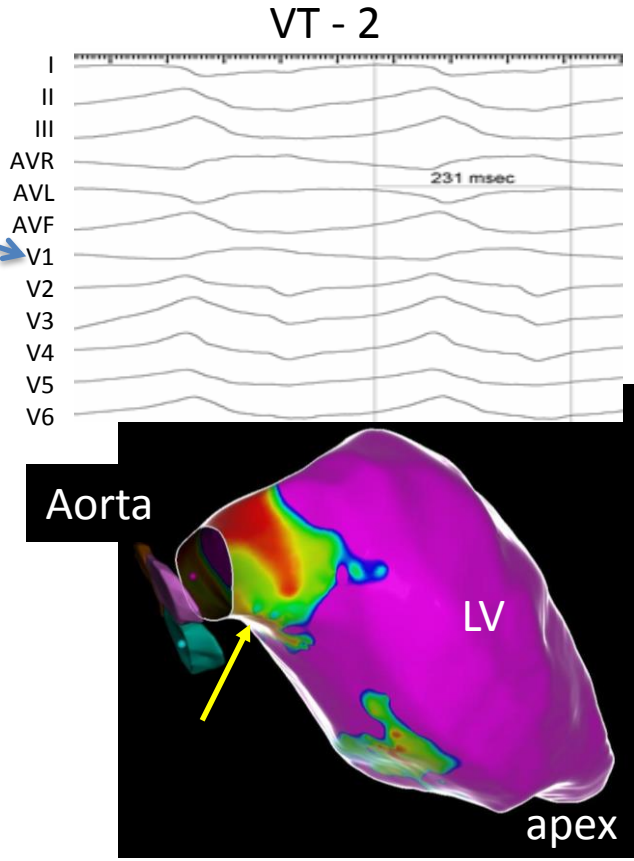
Pace-map under right coronary cusp





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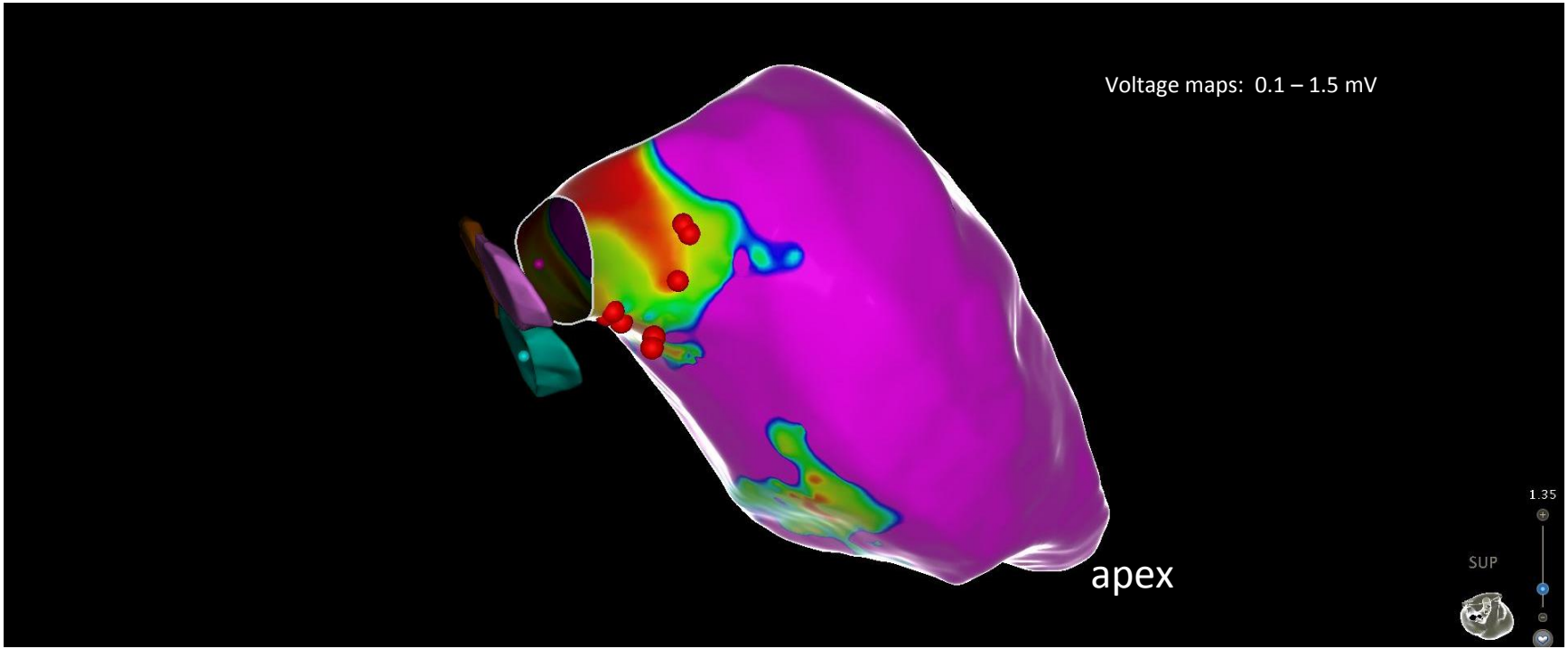
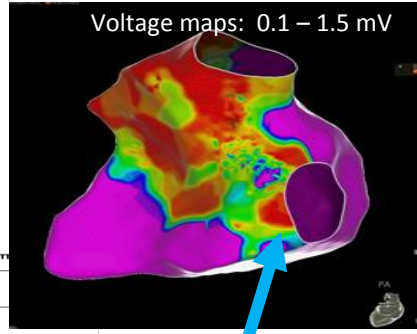
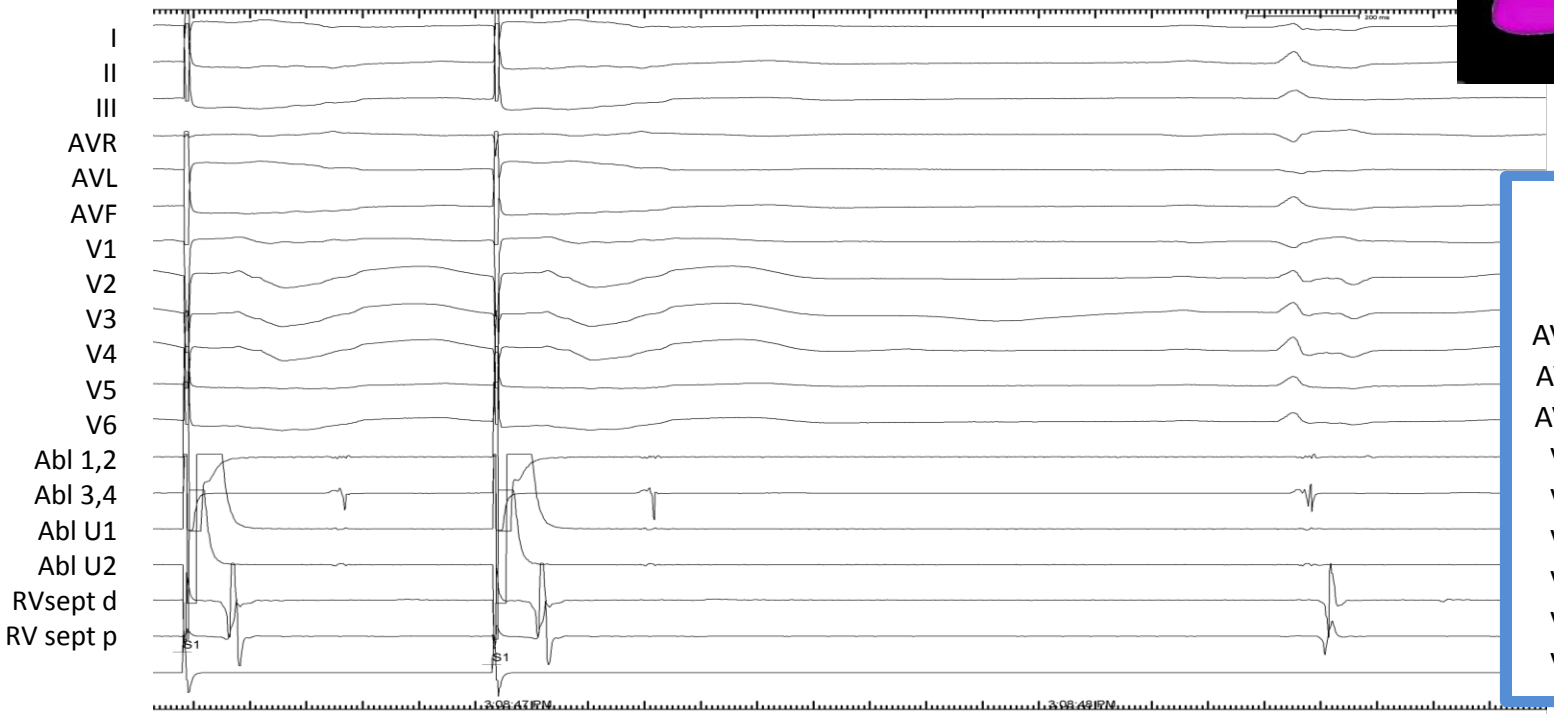
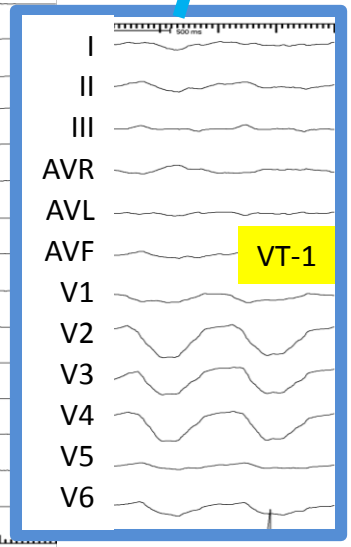
LV periaortic ablation sites



Inferior septal RV near Tricuspid Annulus
Pace-map similar to VT-1







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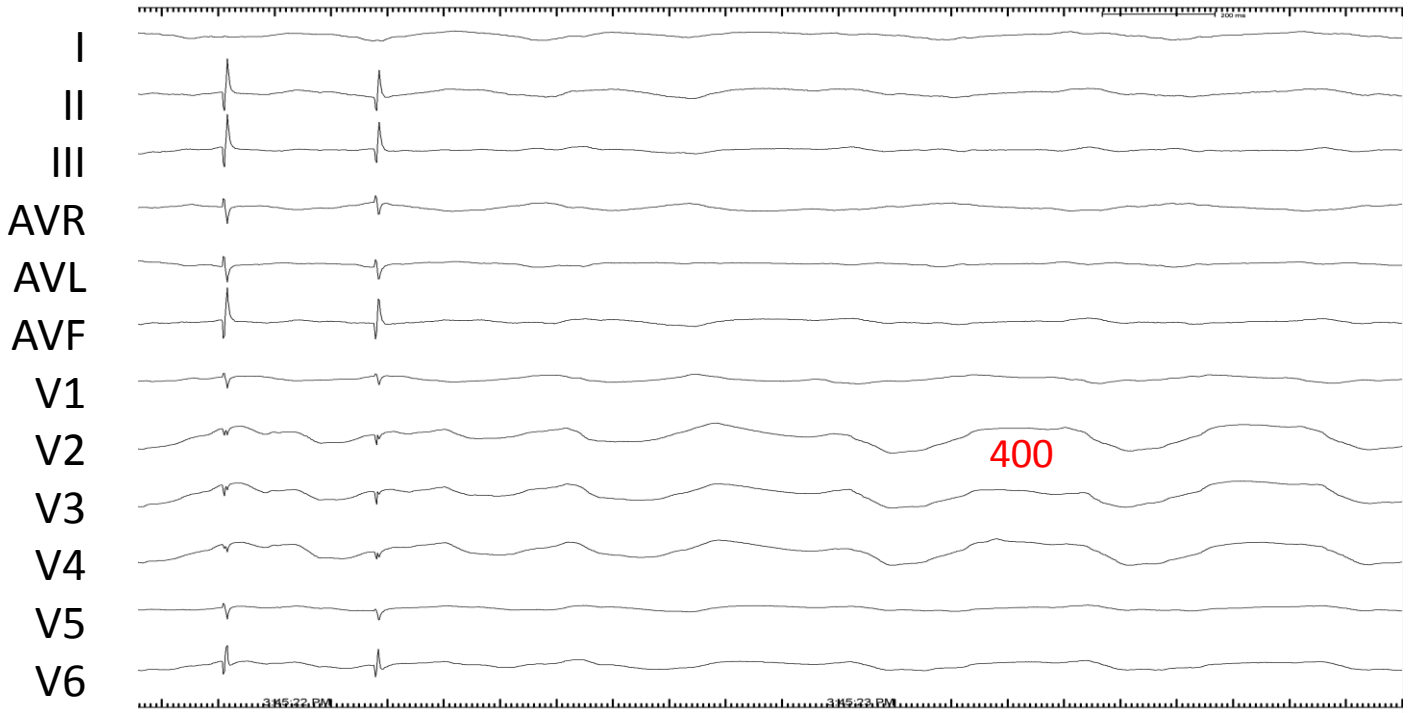
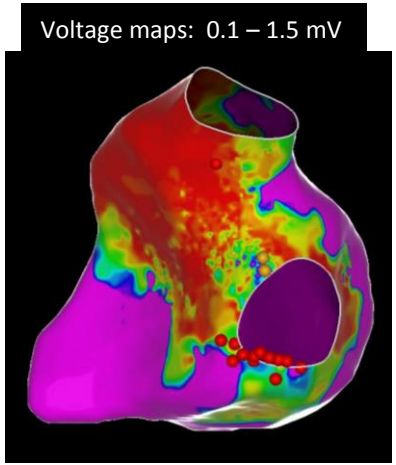
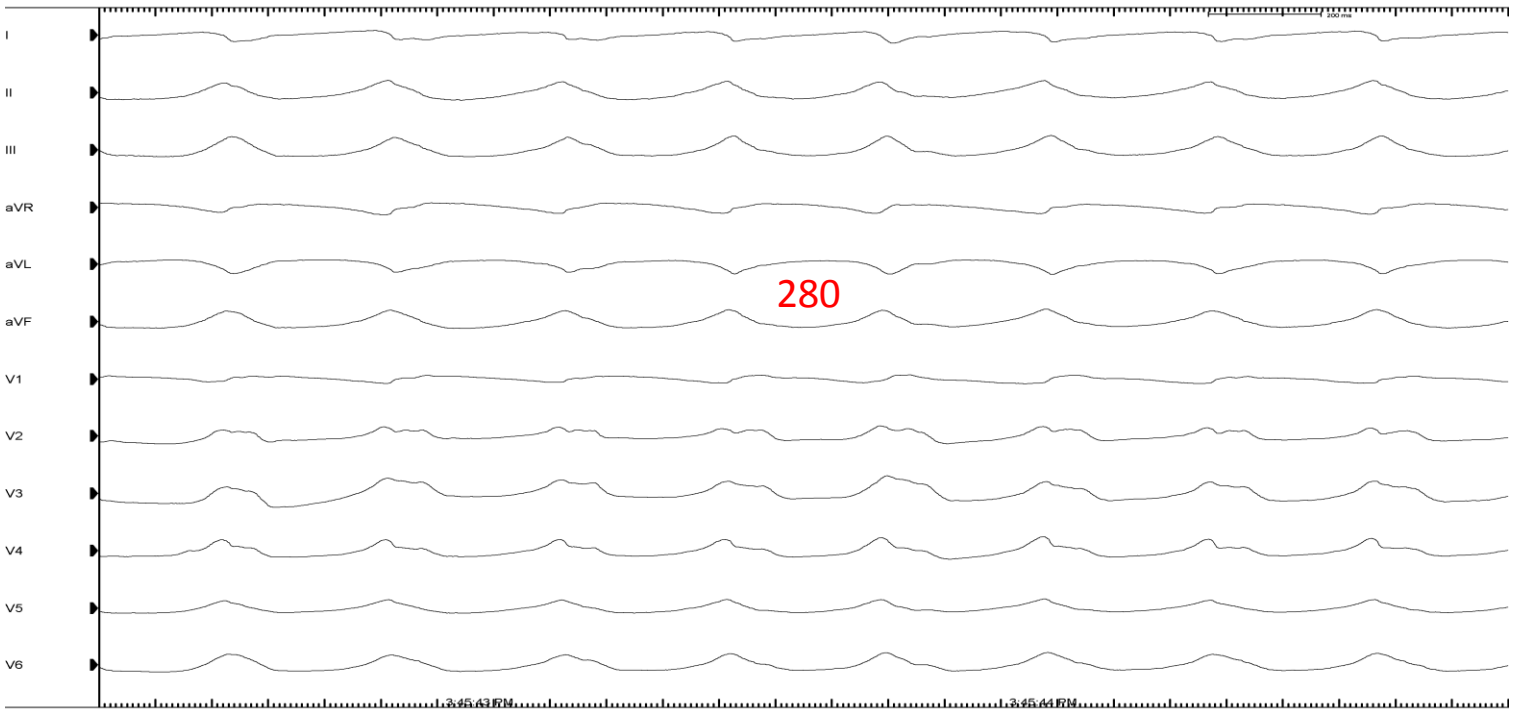
Post RF 18: 600/2 induces VT3 similar to VT1 but slower

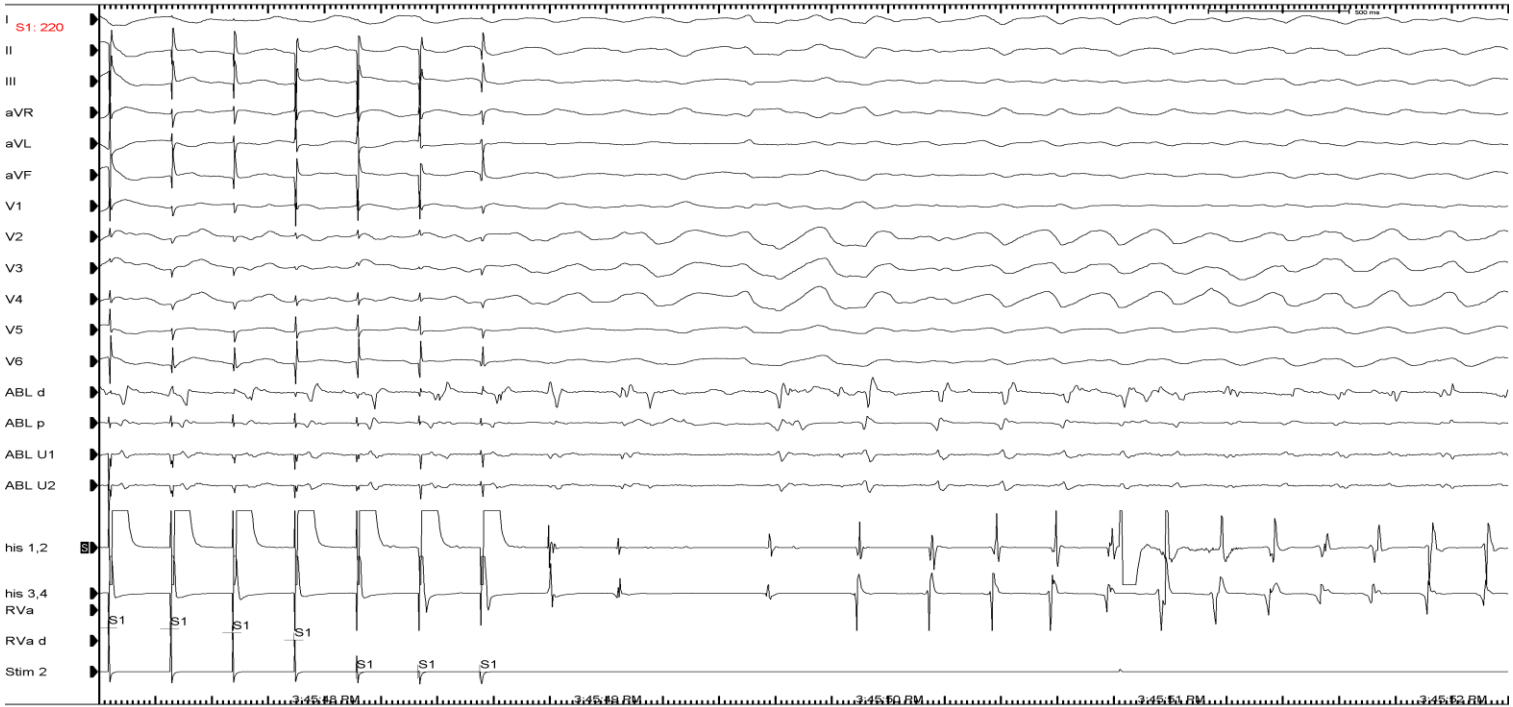




Burst in VT 3 induces VT4

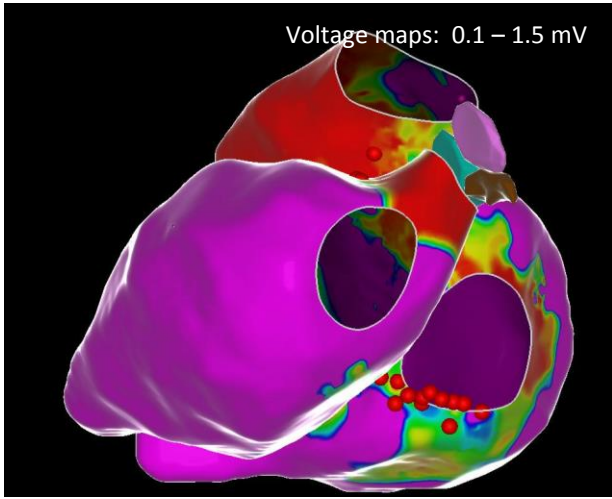


Burst in VT4 accelerates to VF

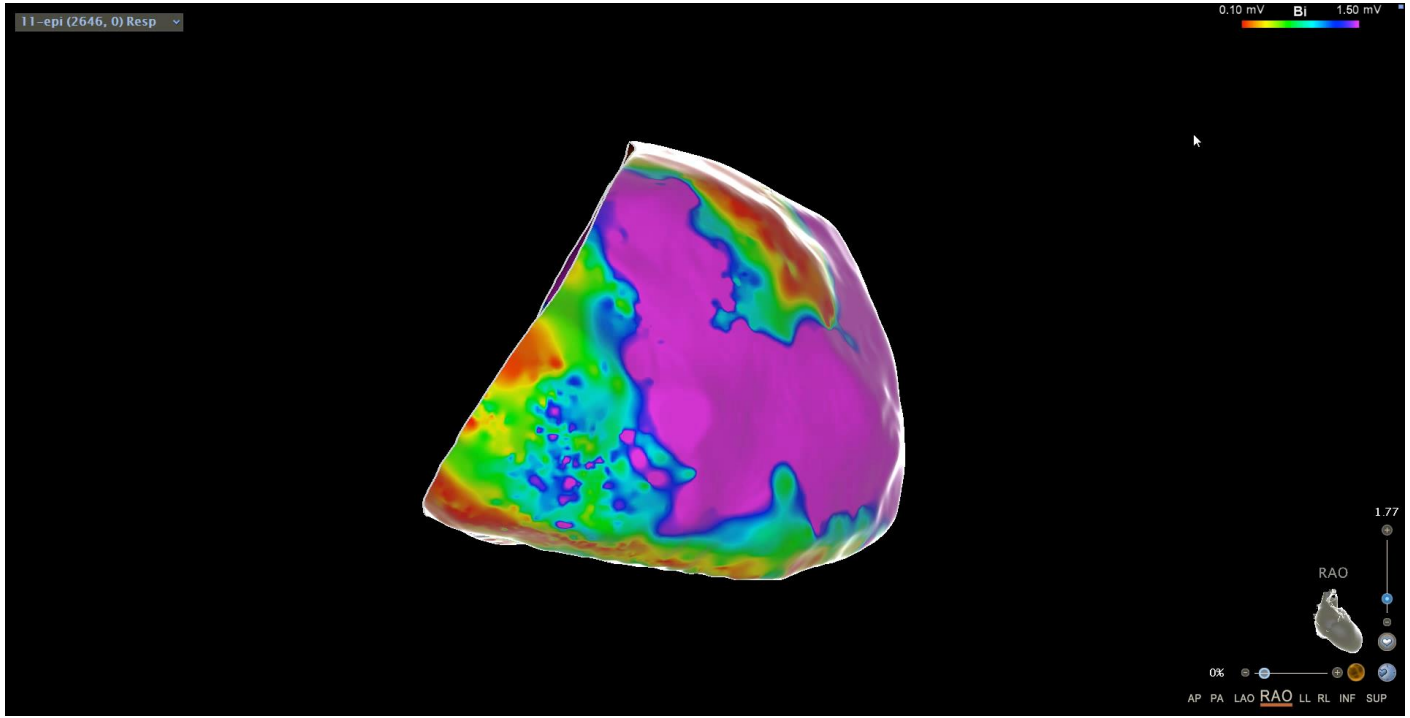


- Ablation at LVOT scar where pacing resembled VT2
- Ablation at RV scar areas in basal RV, RV septum
- VTs with morphologies similar to VT 1 and 2 remain inducible

What next?



Epicardial Voltage map (0.1 – 1.5 mV)



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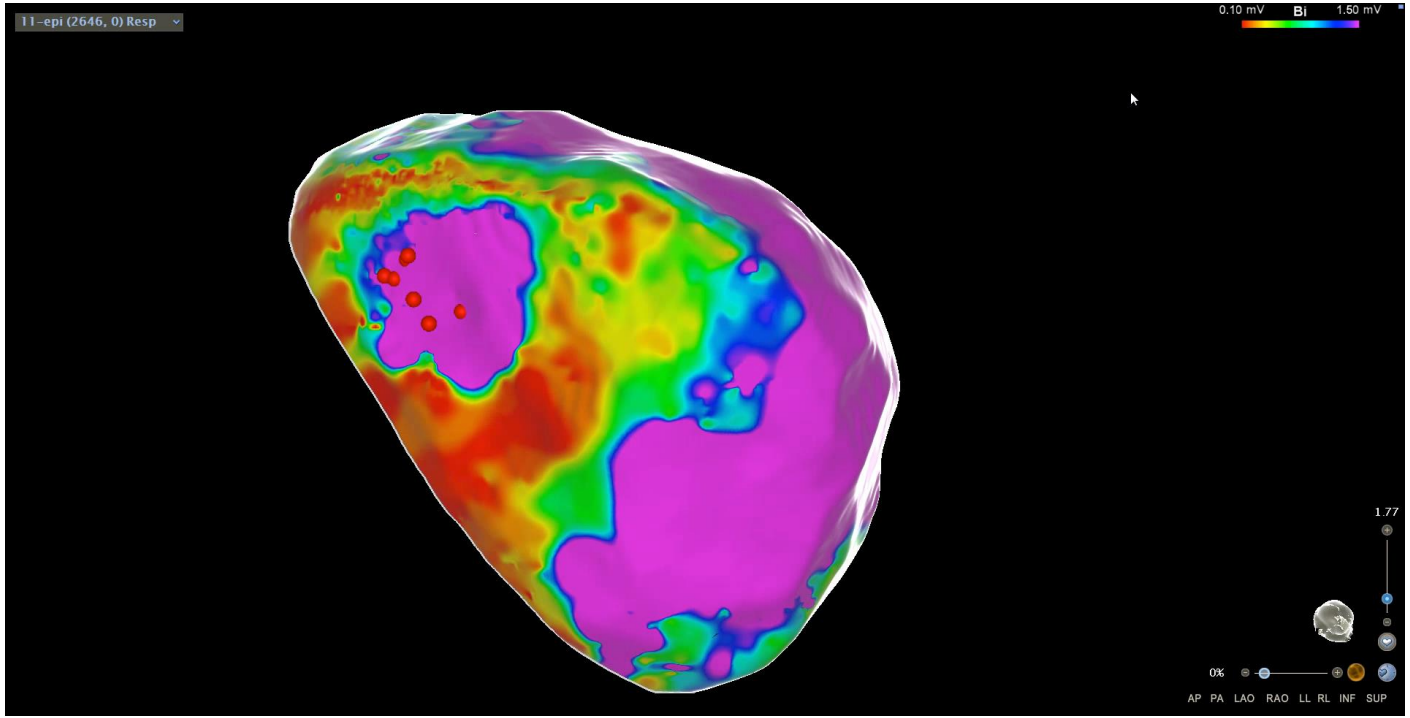
Epicardial lesions over inferior RV



Voltage maps: 0.1 – 1.5 mV

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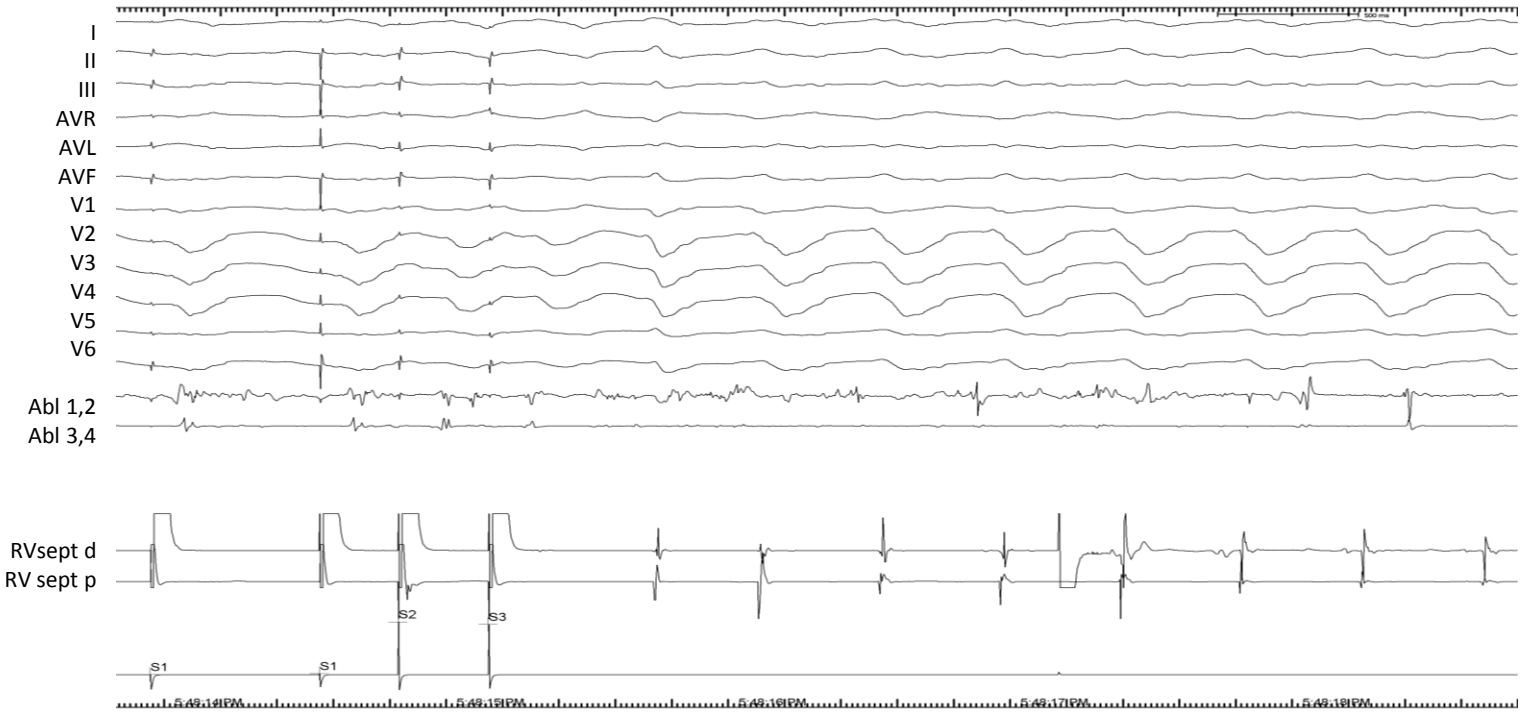
After epicardial mapping and more RV endocardial ablation VT inducible with 600/2 ES



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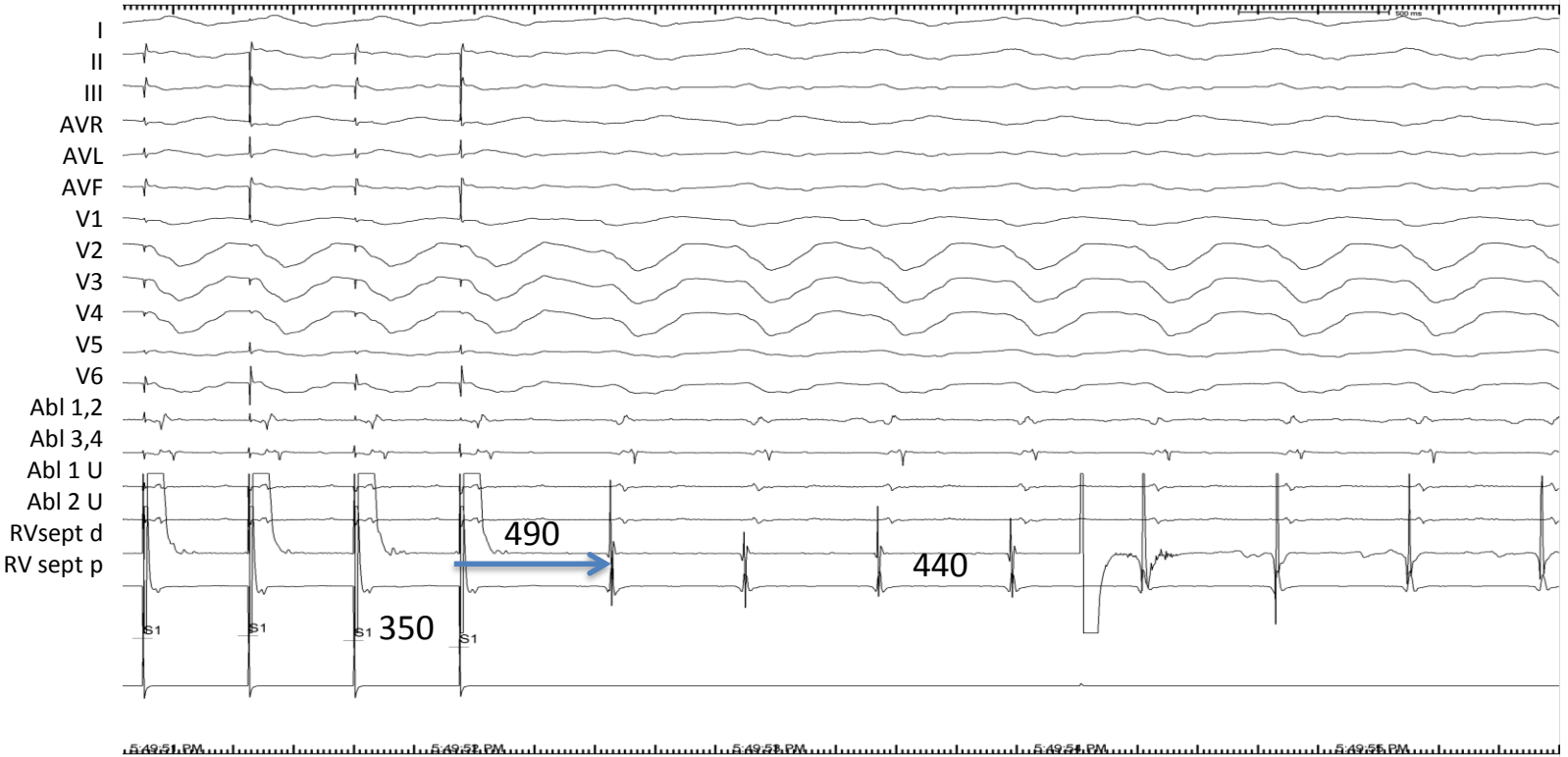
Entrain from RV septum



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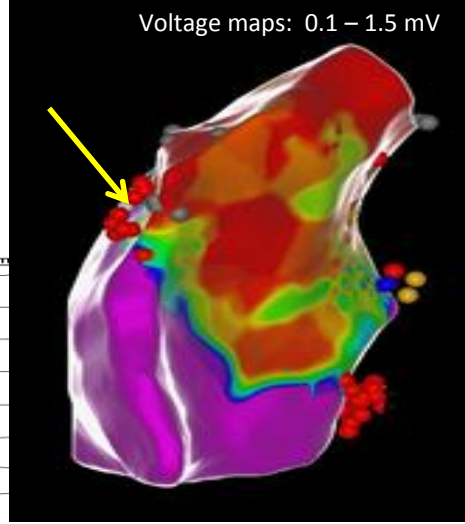
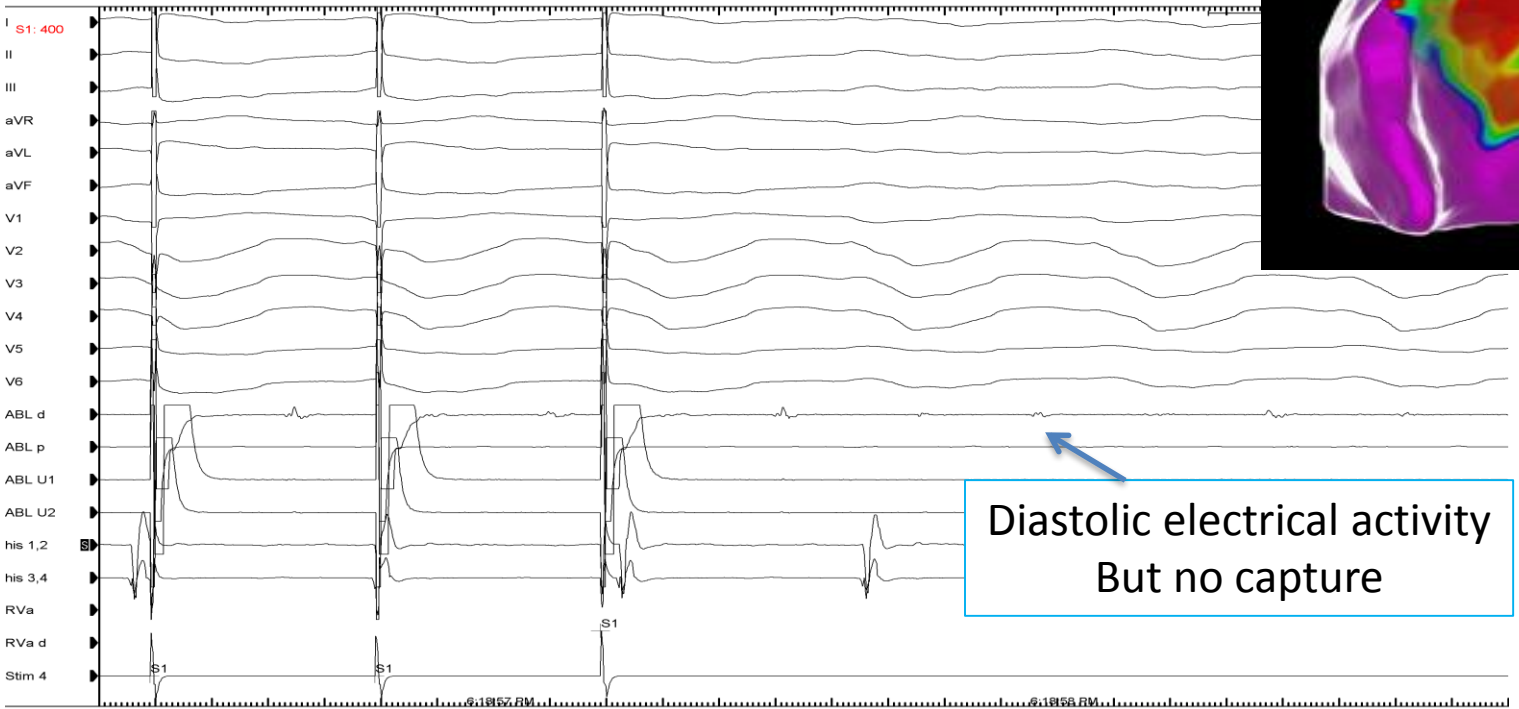
Pacing fails to terminate - CV

VT recurs with catheter manipulation at the anterior RV



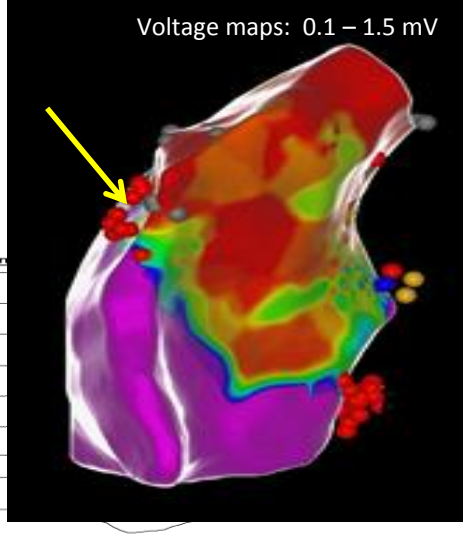


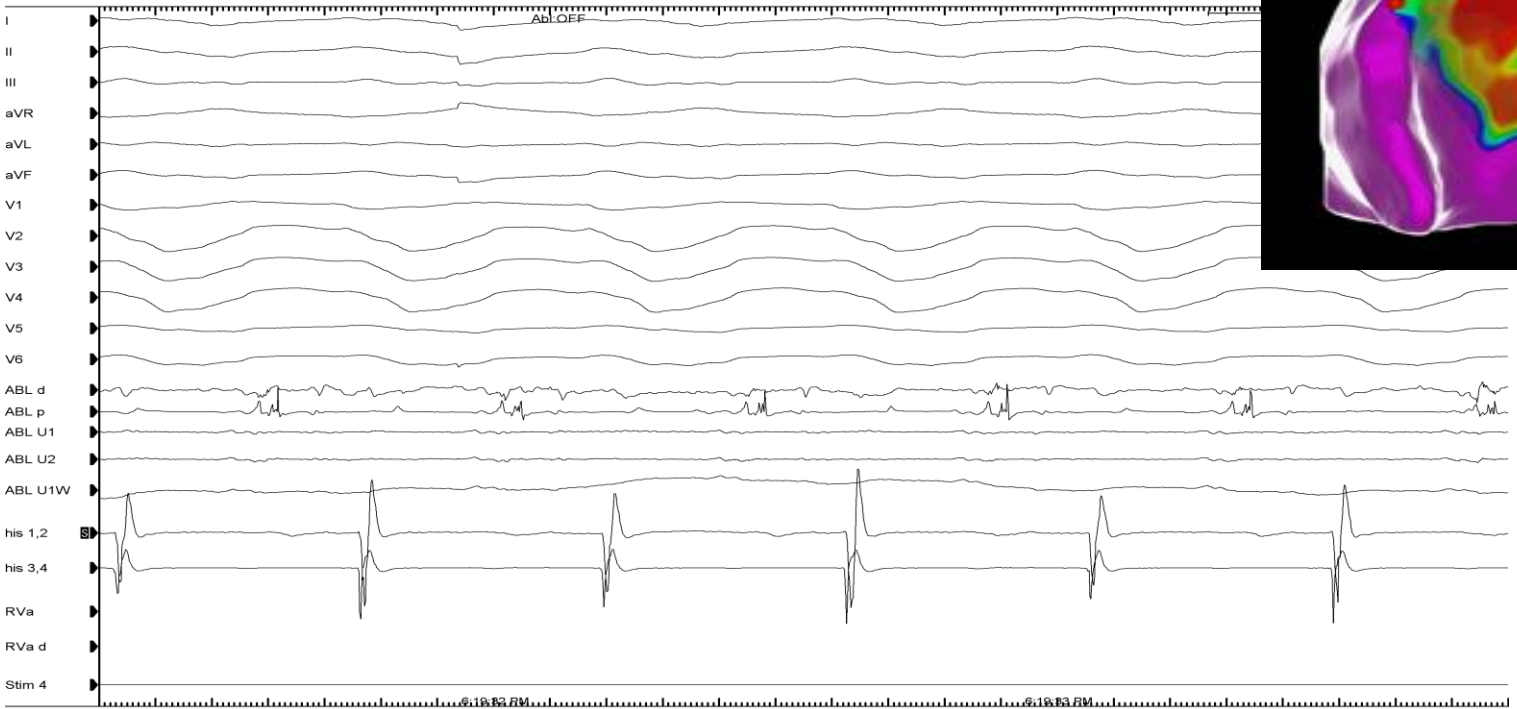
Diastolic electrical activity
But no capture

Pacing: No capture

RV free wall

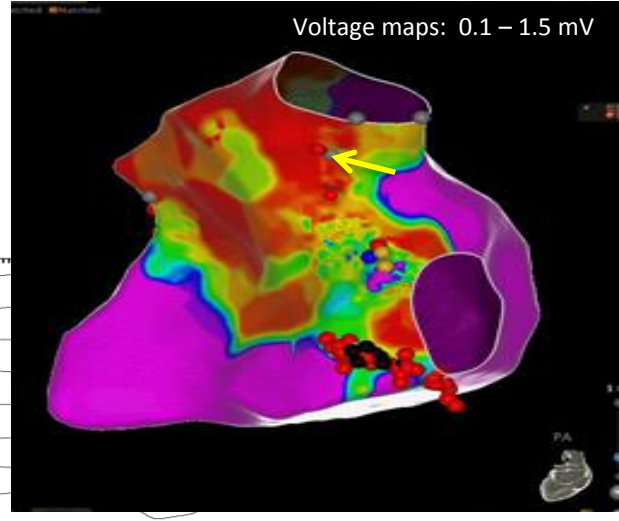
RF off no effect

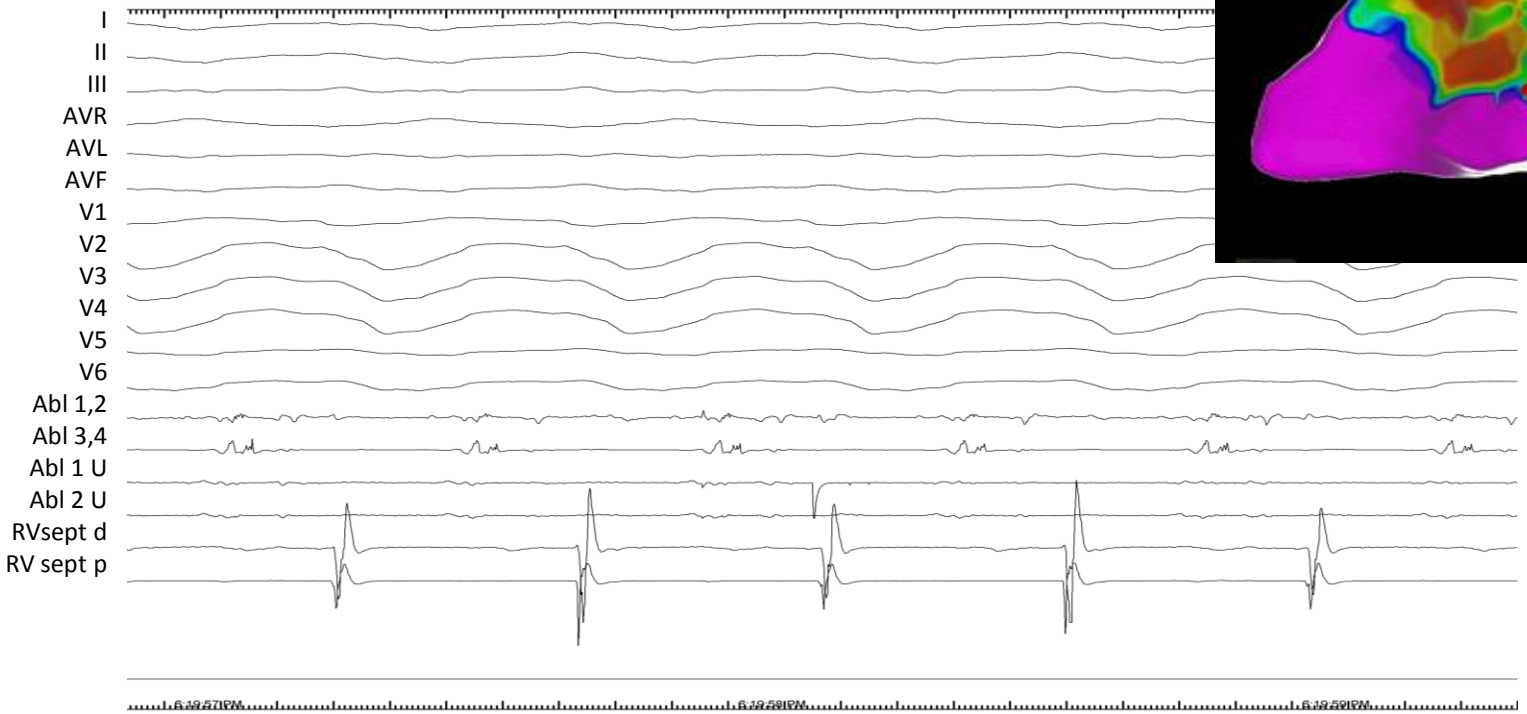




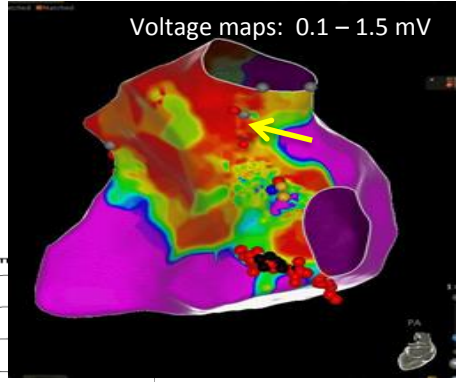
VT recurs with catheter manipulation

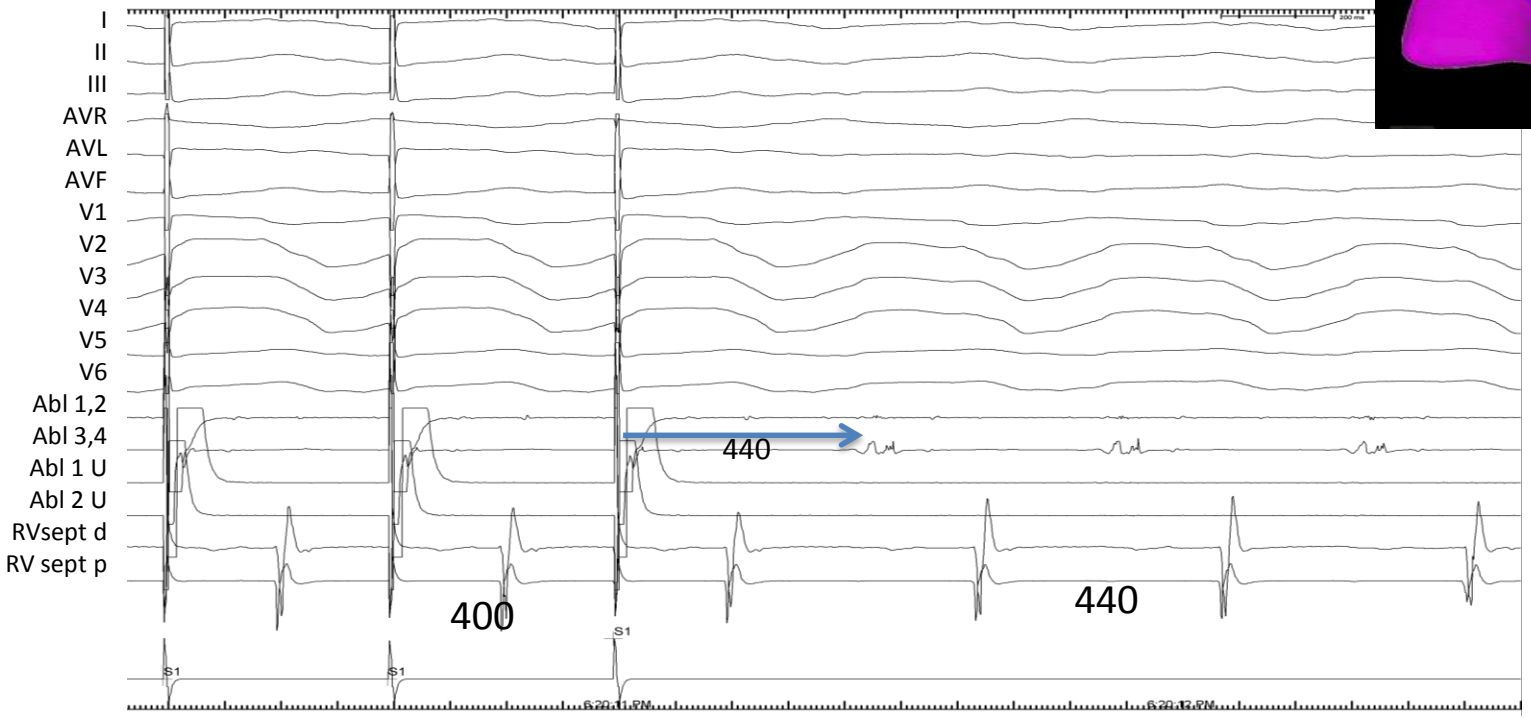
Mapping Site: Infundibular septum



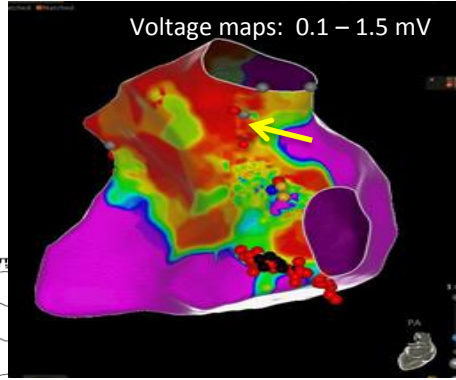


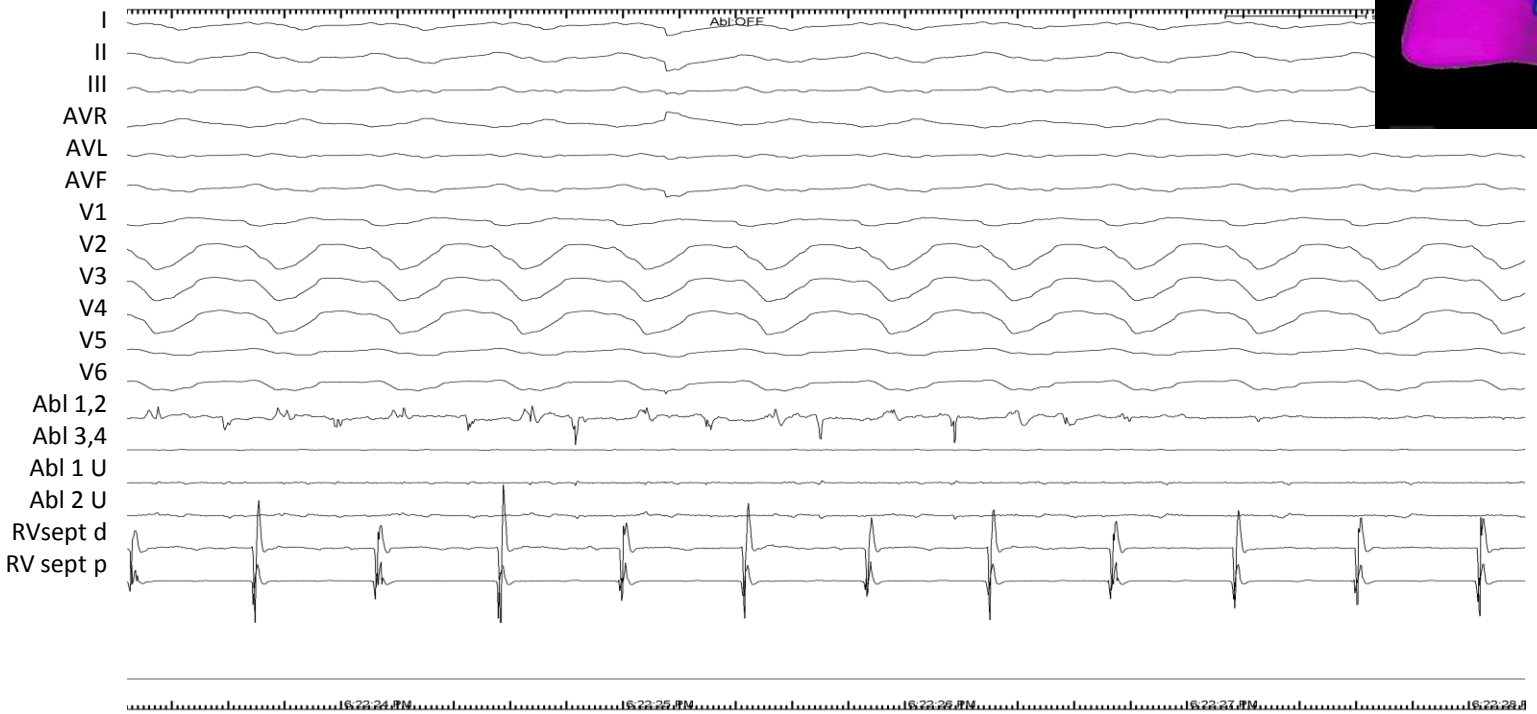
Infundibular septum





Infundibular septum - RF
No effect on VT





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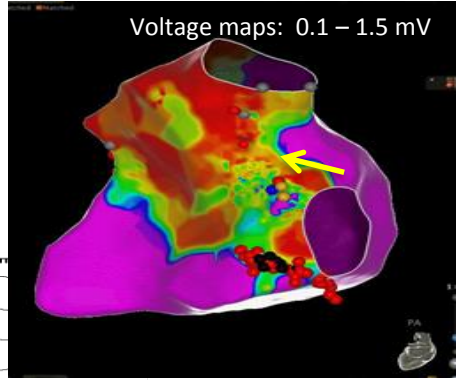
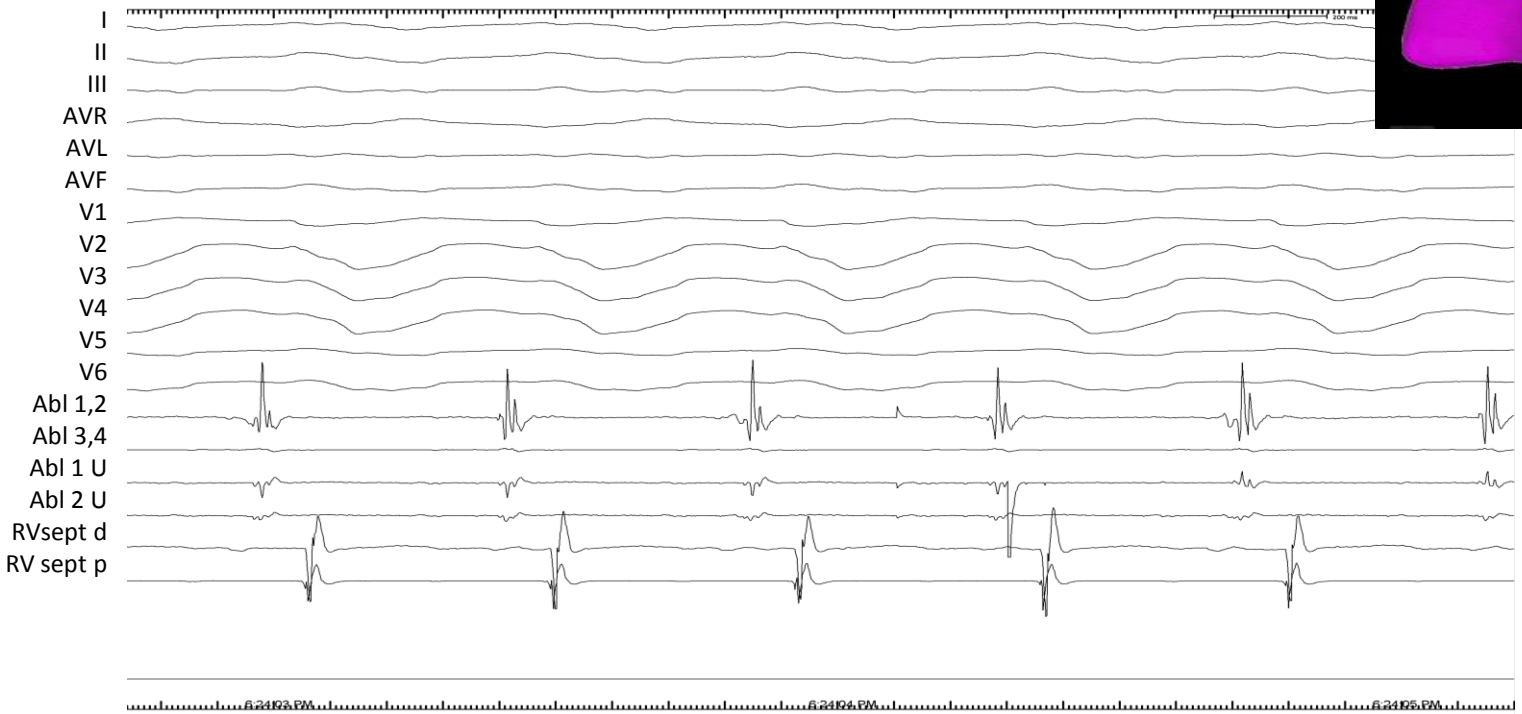
Infundibular septum above the His





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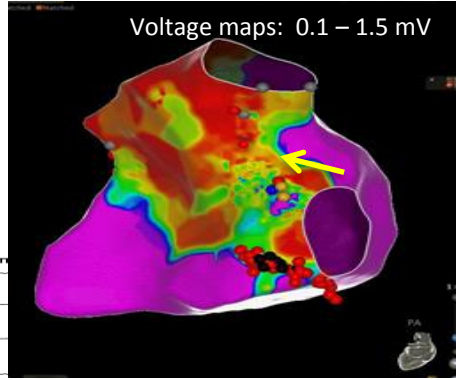
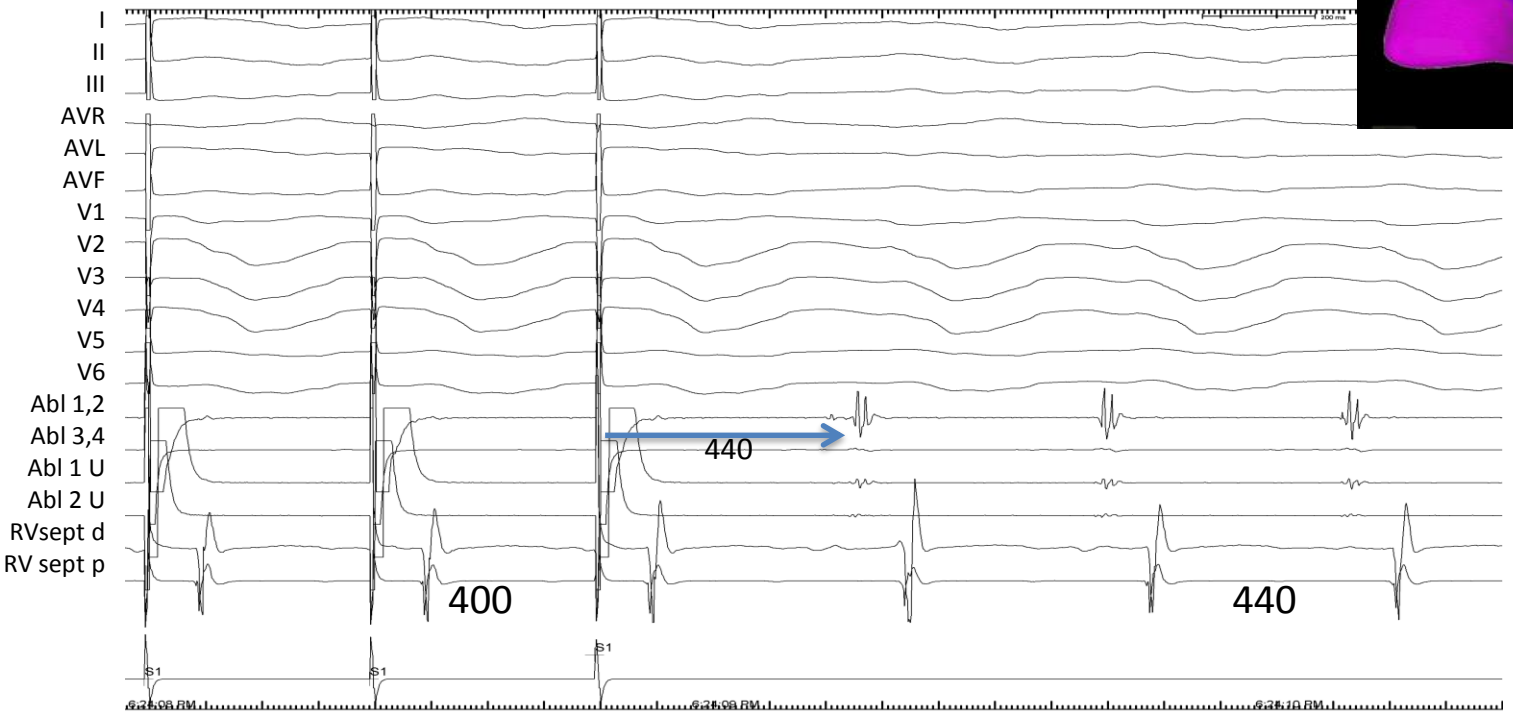
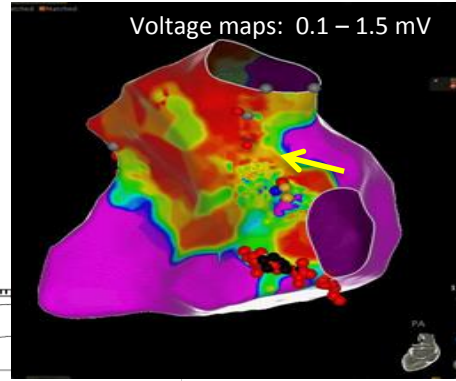
Infundibular septum above the His

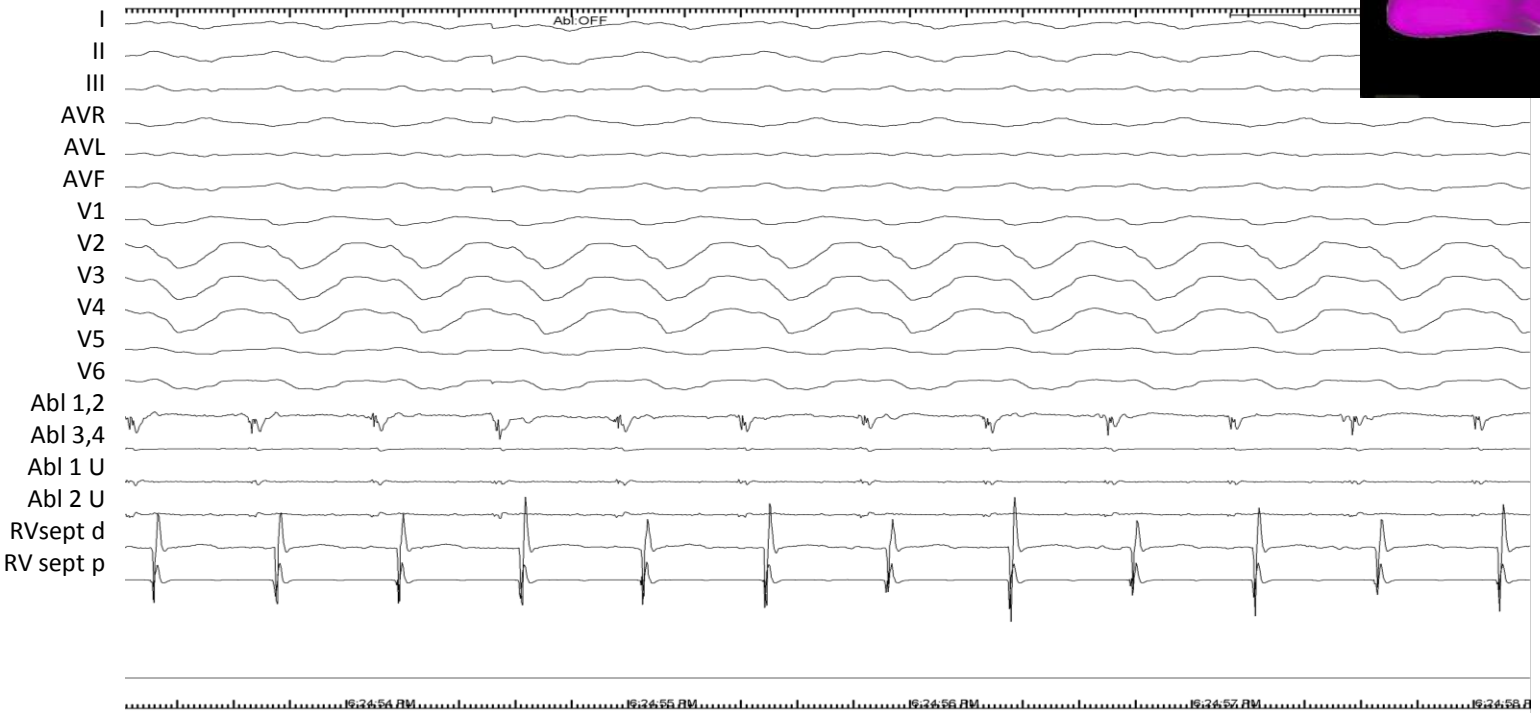




Infundibular septum above the His

RF fails to terminate VT





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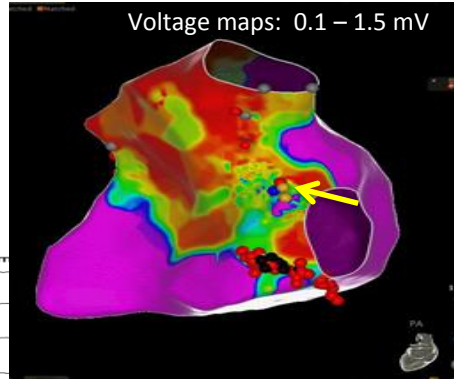
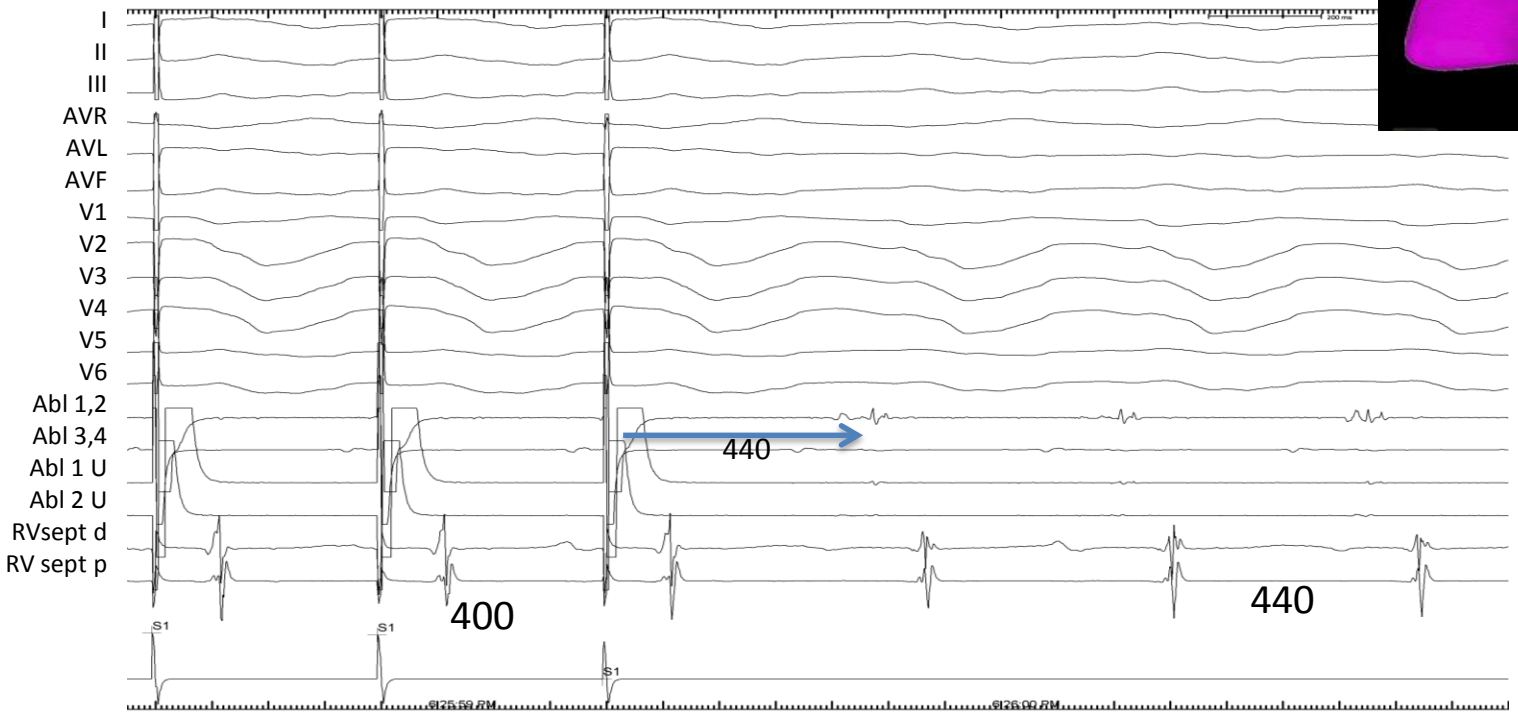
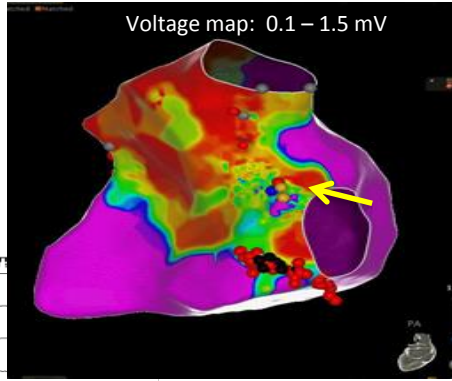
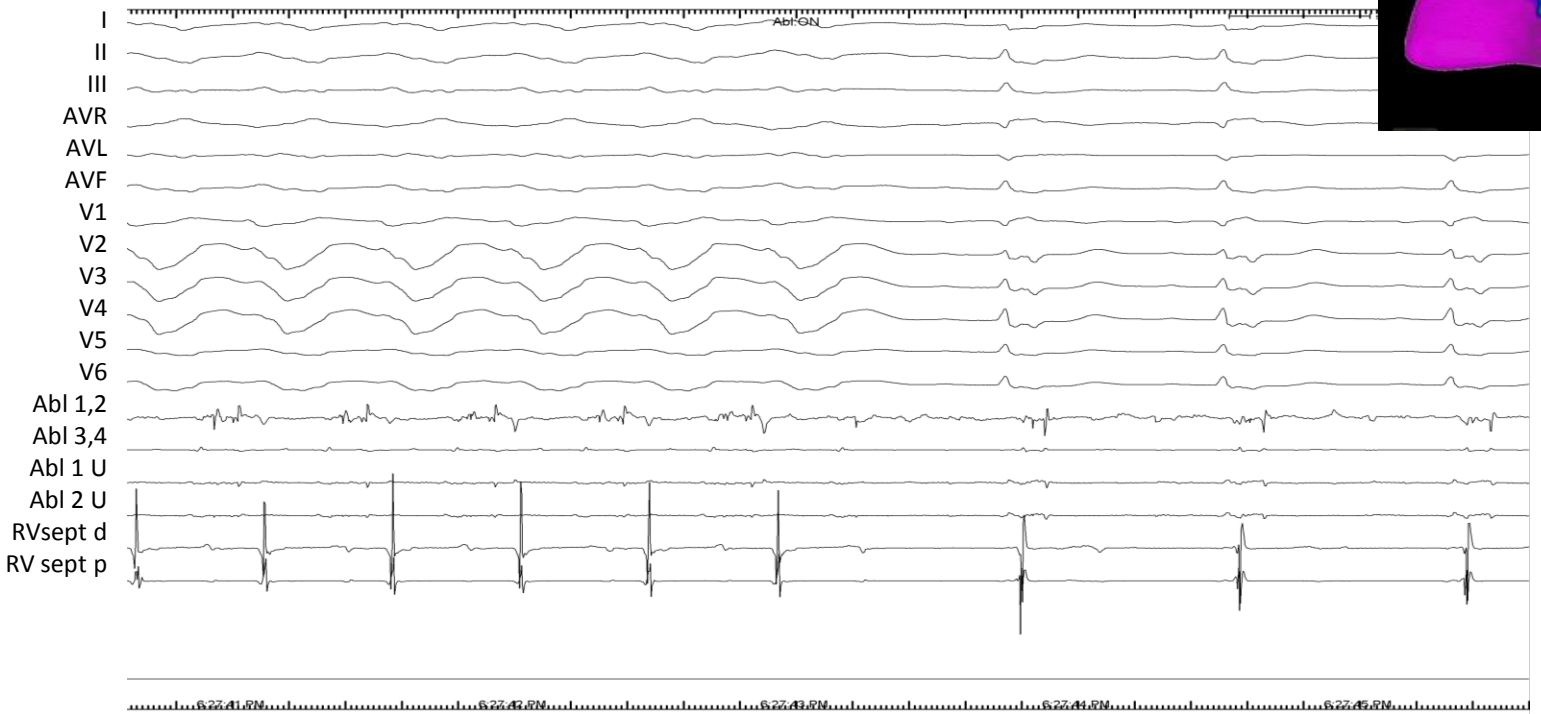
Infund septum above the His at the tricuspid annulus



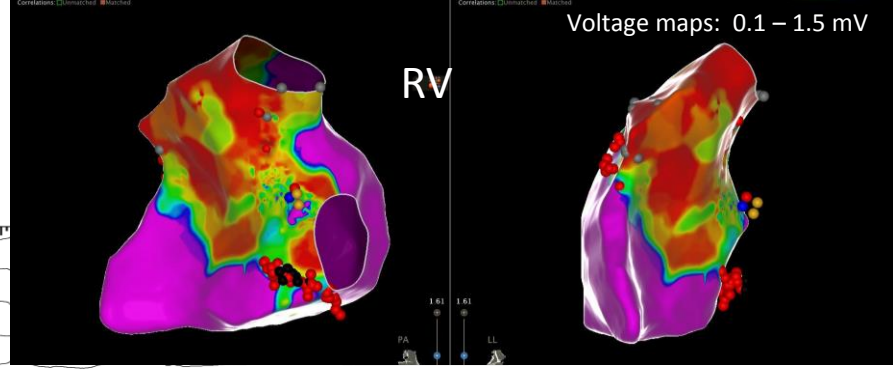


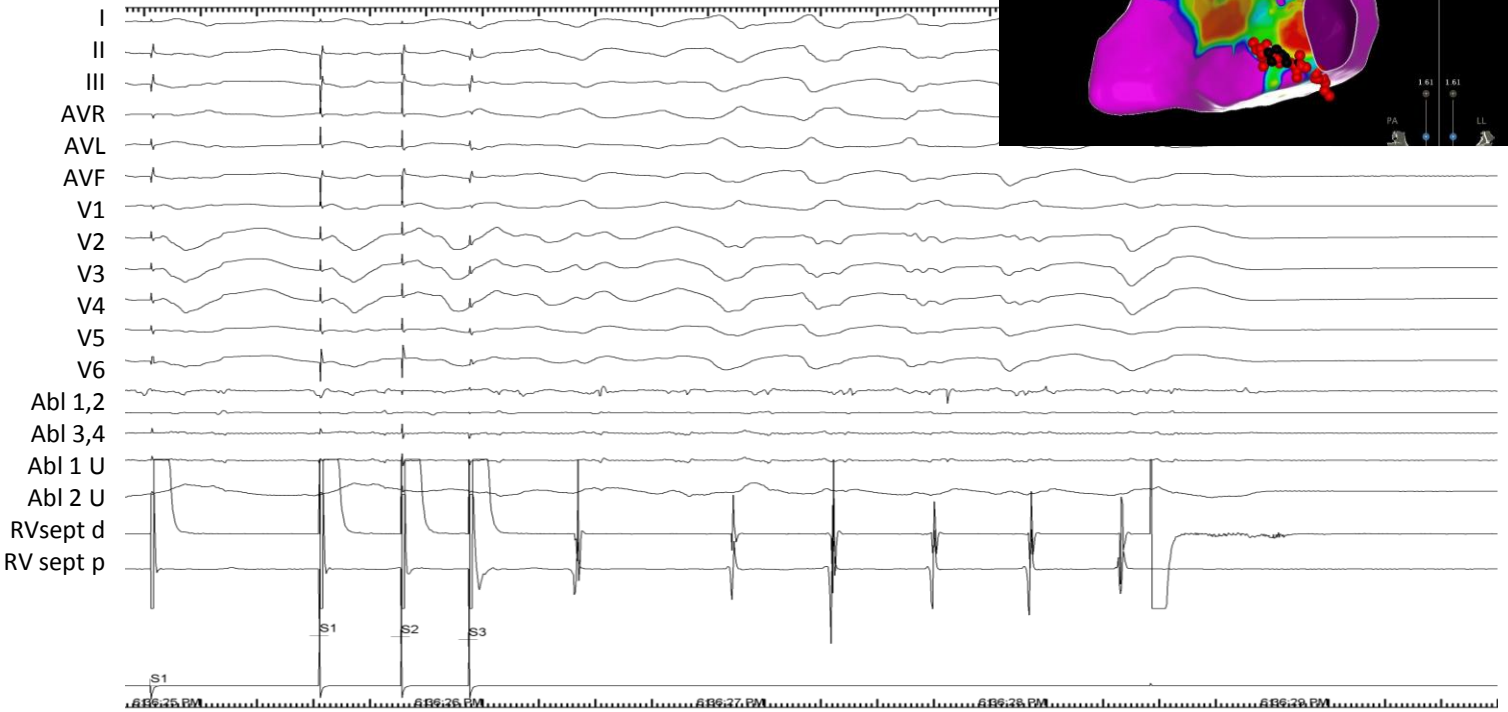
Infund septum above the His at the tricuspid annulus





Maximum response to programmed stimulation





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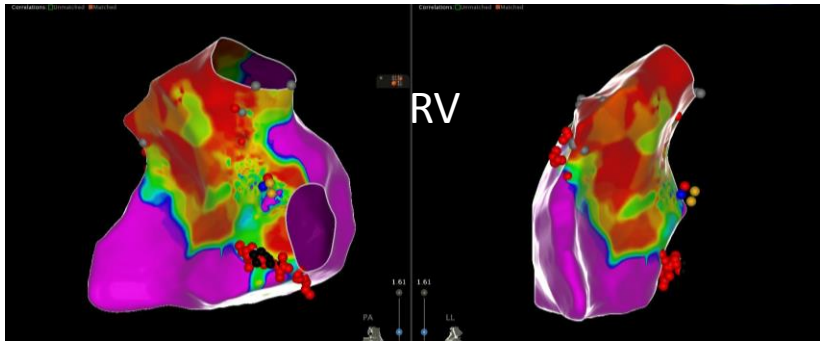
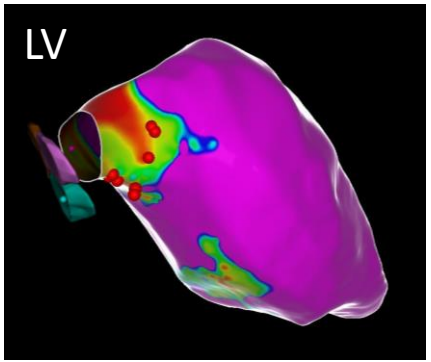
Follow - up

- Amiodarone reduced to 200 mg daily
- Treated with predinsone followed by taper
- Follow-up 7 months:
  - Ambulatory and active
  - No VT
  - LVEF 20 – 30%
  - Transplantation deferred

Cardiac Sarcoid with VT

- RV > LV involvement
- Prominent septal involvement
- VTs from infundibular septum and likely periaortic region



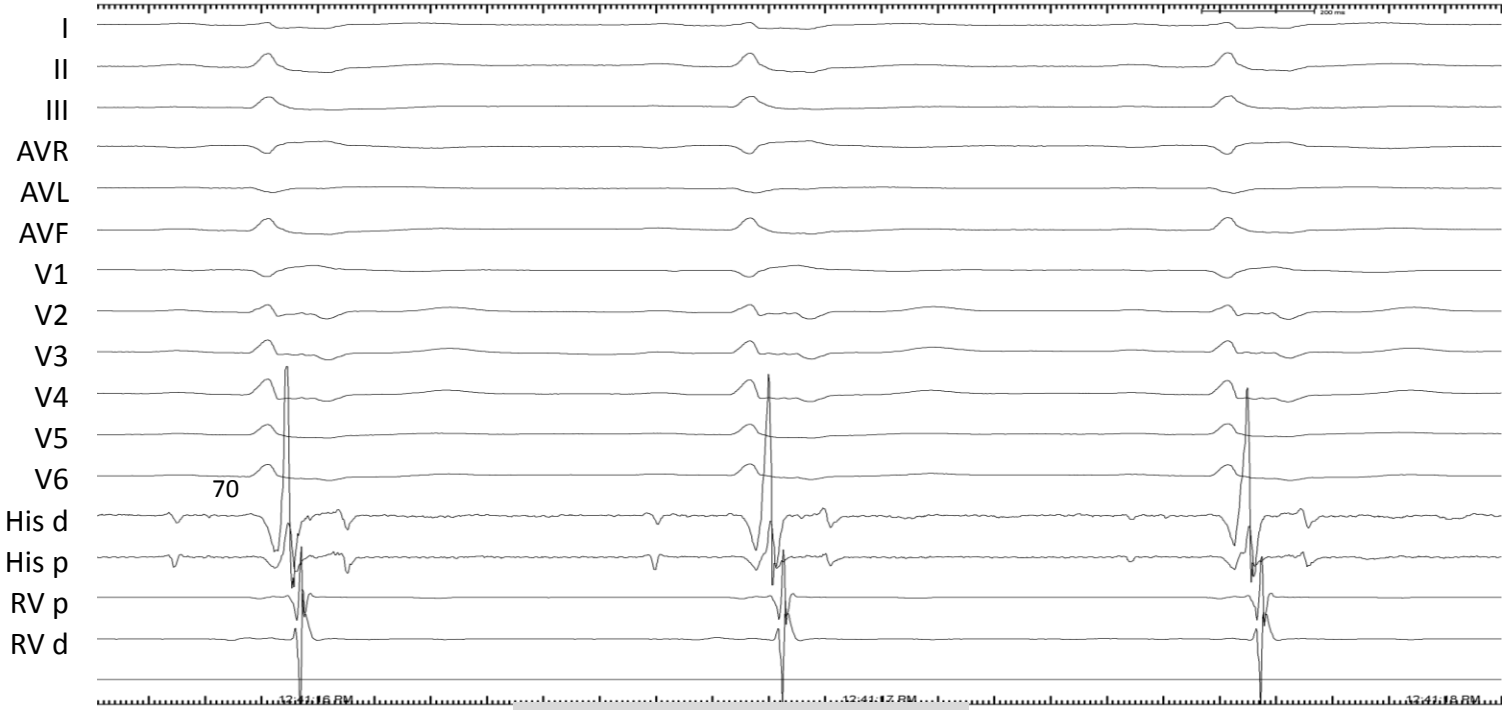


Voltage maps: 0.1 – 1.5 mV

Thank You

EP Study

Baseline



Wide RBBB
HV 70
possible LP in his region

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