SABATO 9 OTTOBRE

The mysteries of the Right Ventricle

Carlo Pappone, *San Donato Milanese - MI*

The mysteries of the Right Ventricle

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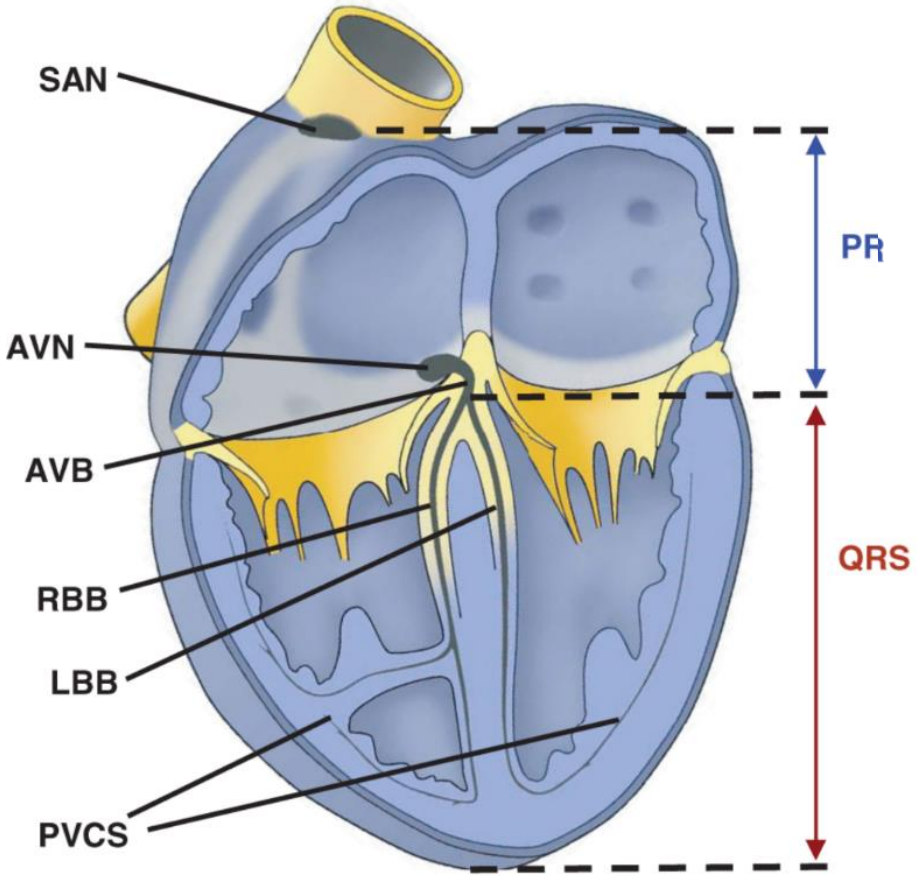
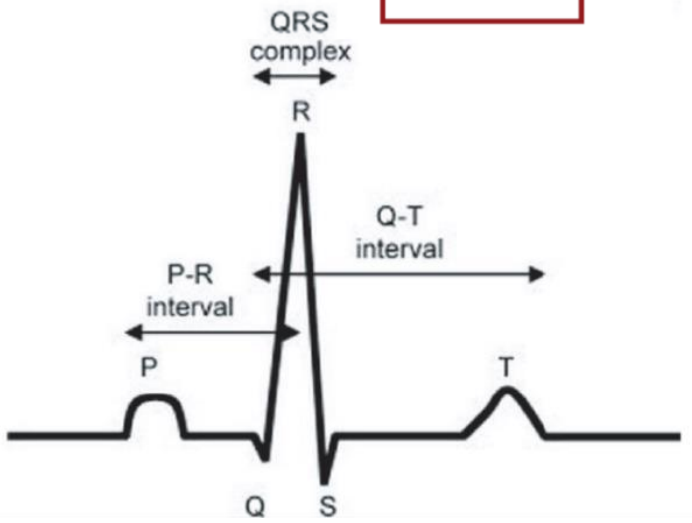
Gene Regulatory Networks in Cardiac Conduction System Development

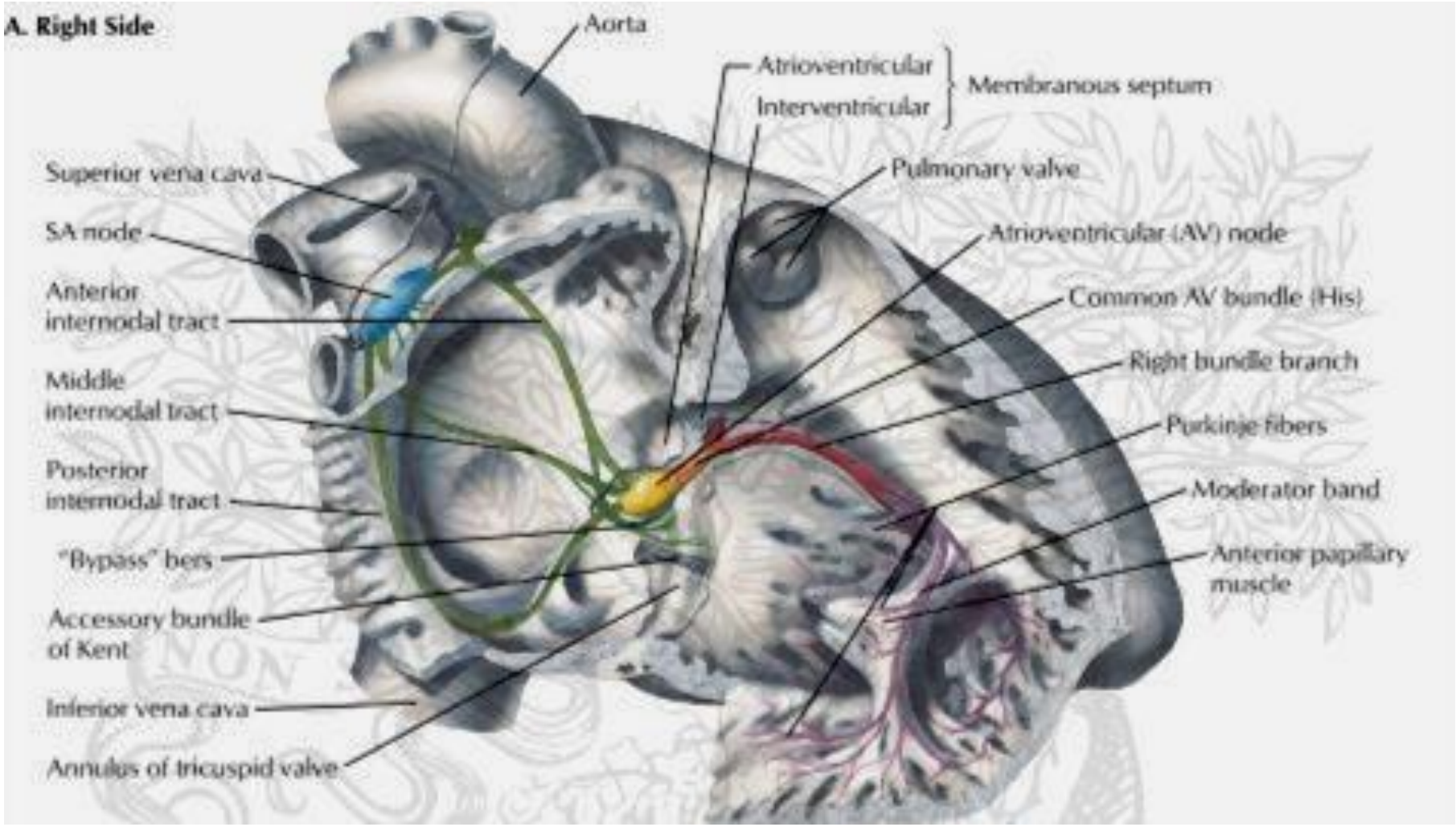


PR	PR & QRS	QRS	QT (repolarization)
CAV1	SCN10A	HAND1	SCN5A
MEIS1	SCN5A	TBX20	TBX5
NKX2-5	TBX5	CACNA1D	KCNQ1
SOX5	TBX3	DKK1	KCNE1
		CDKN1A	NOS1AP
		NF1A	

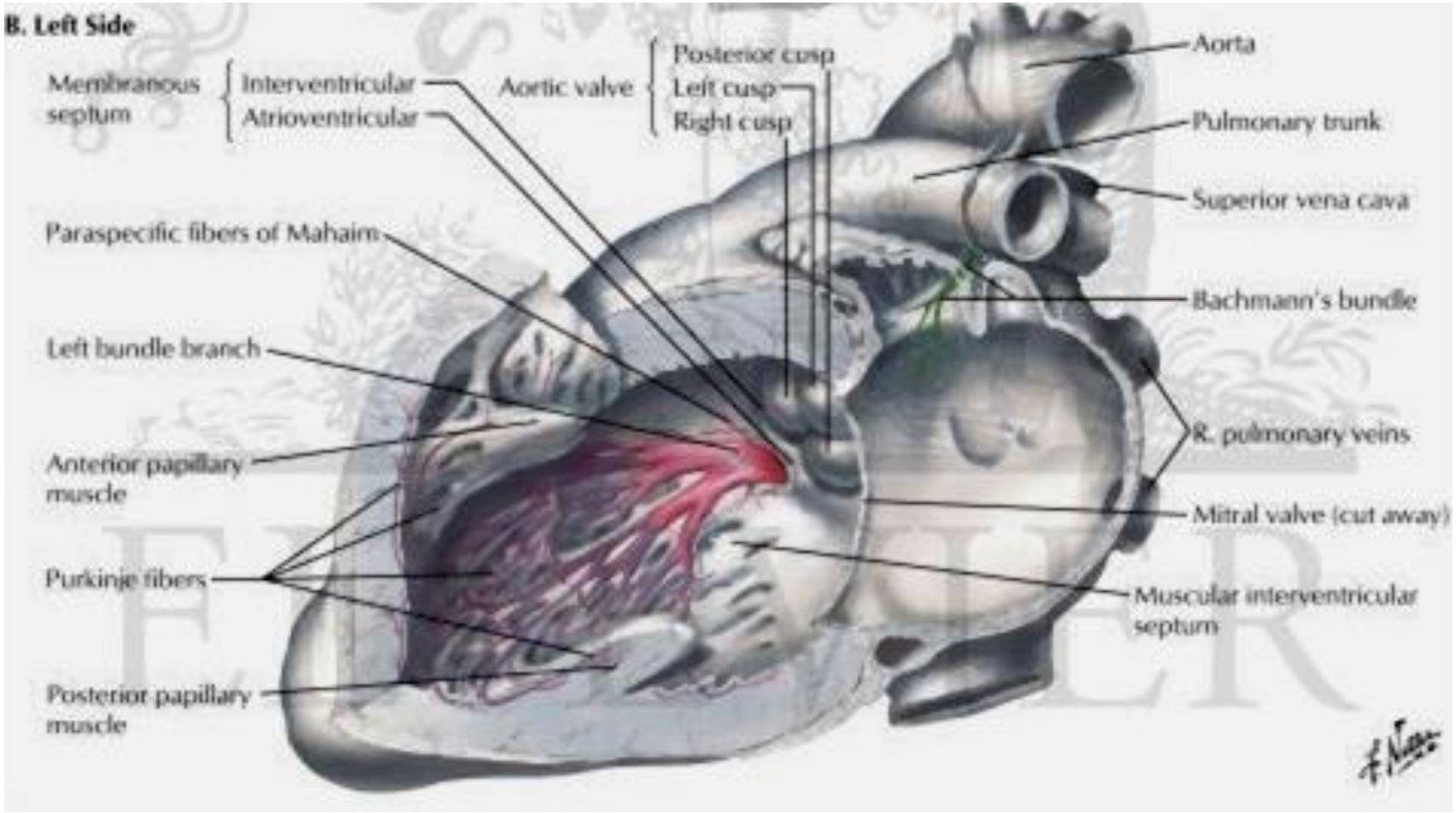


van Duijvenboden K. et al. Brief Funct Genomics. 2013

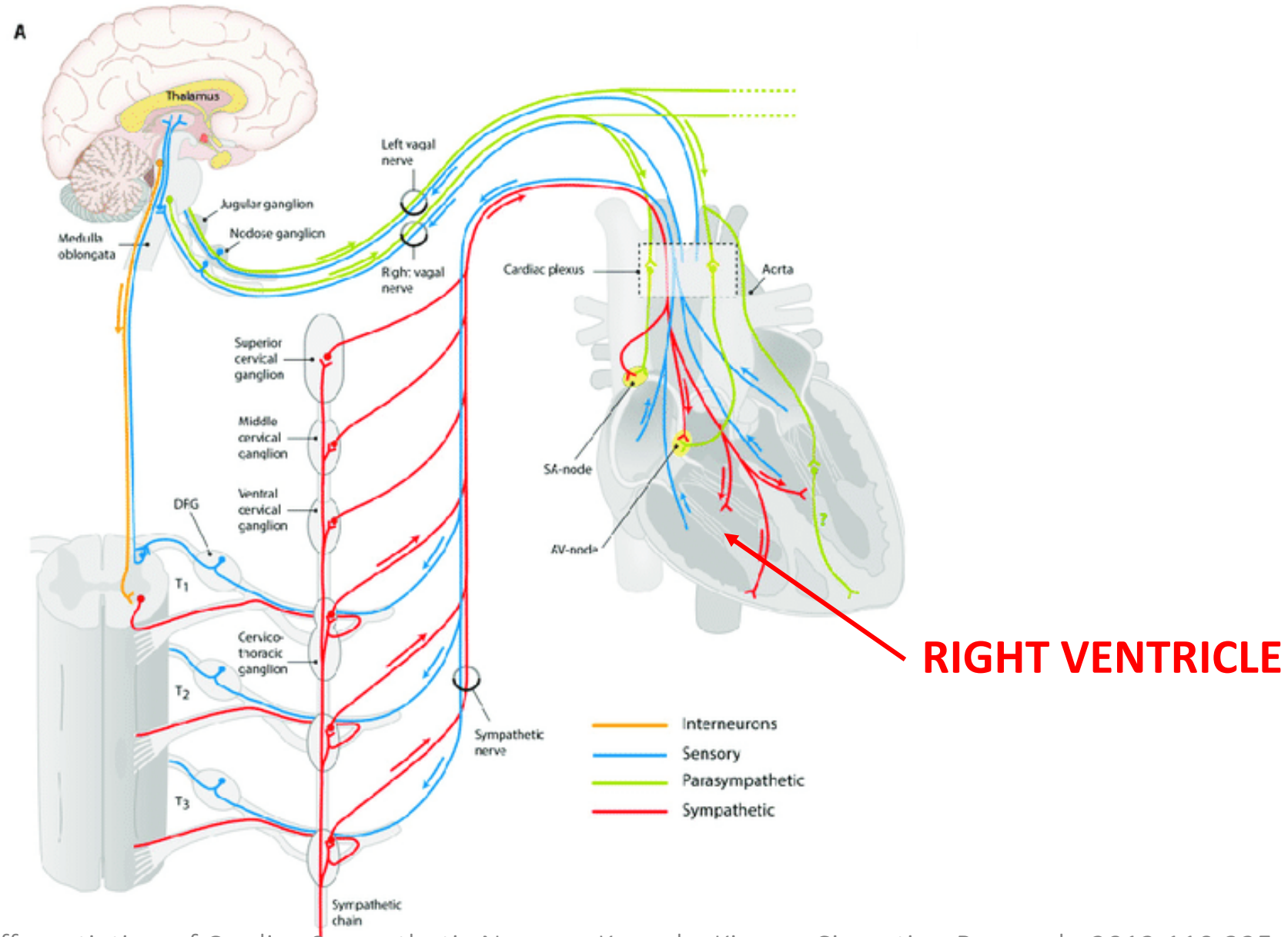
Right Side Heart Conduction System



Left Side Heart Conduction System



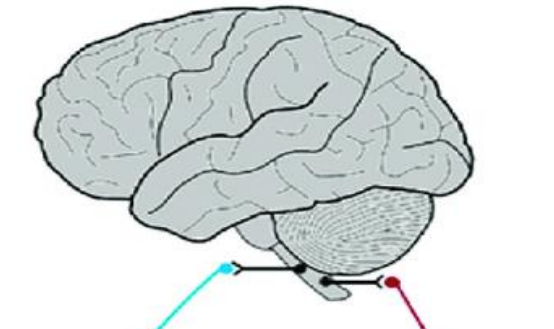
SYMPATHETIC AND PARA-SYMPATHETIC INNERVATION



RIGHT VENTRICLE

Development, Maturation, and Transdifferentiation of Cardiac Sympathetic Nerves - Kensuke Kimura Circuation Research. 2012;110:325–336

HEART AND ANS: electrophysiologic effect



Parasympathetic Stimulation (ACh)

- ↓ Heart Rate
- ↓ Cardiac inflammatory cytokines during ischemia
- ↑ Connexin 43
- ↑ Gap junctions
- ↓ AV node conduction
- ↑ Vagally derived nitric oxide
- ↓ Ventricular refractory period

Sympathetic Stimulation (NA)

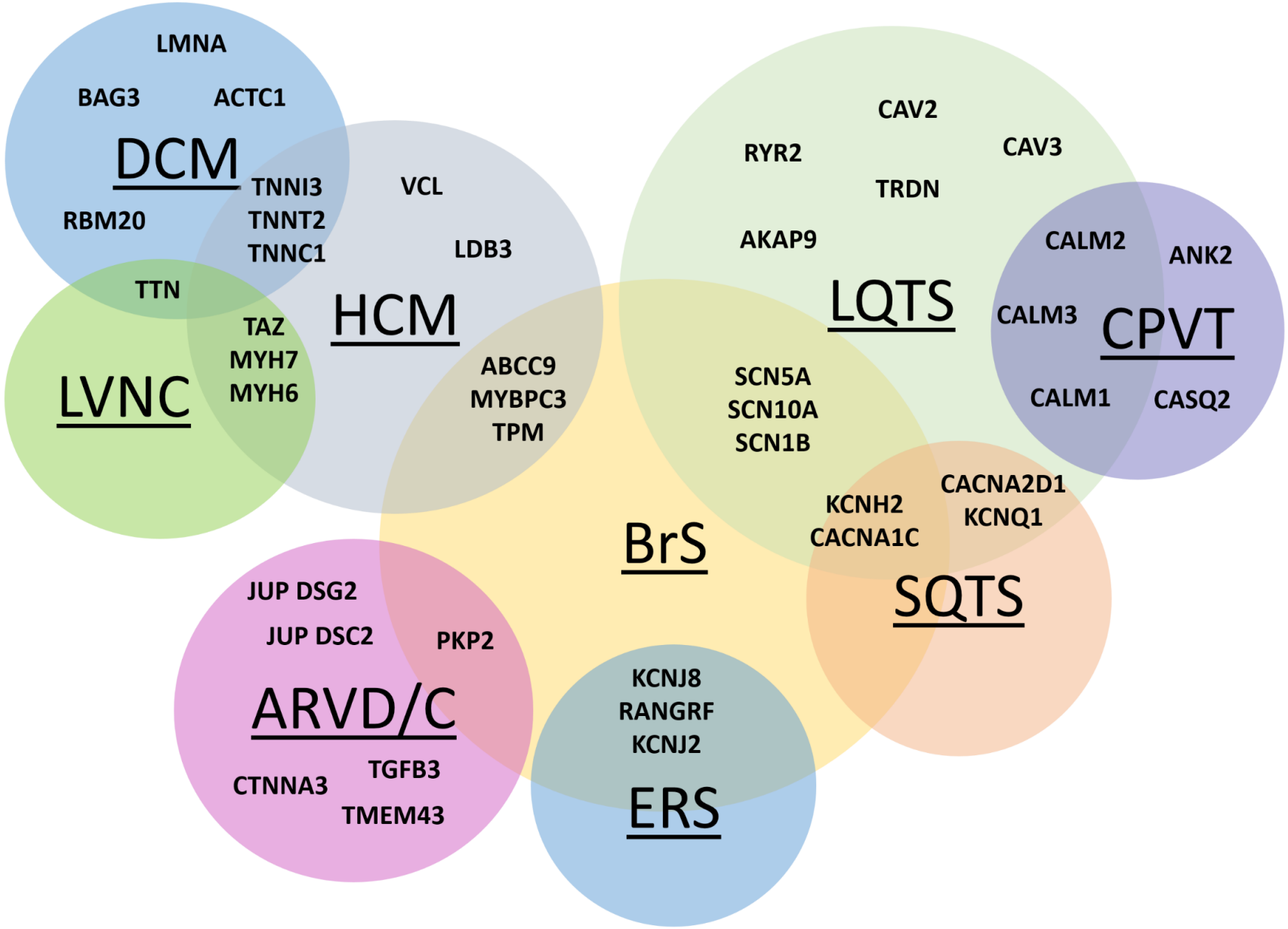
- ↑ Heart Rate
- ↑ Heterogeneity of refractoriness
- ↑ Abnormal direction of repolarization
- ↑ Action potential duration
- ↑ Contraction force
- ↑ Ventricular refractory period

\+ ↑ Triggered activity
↑ Re-entry
↑ Abnormal automaticity

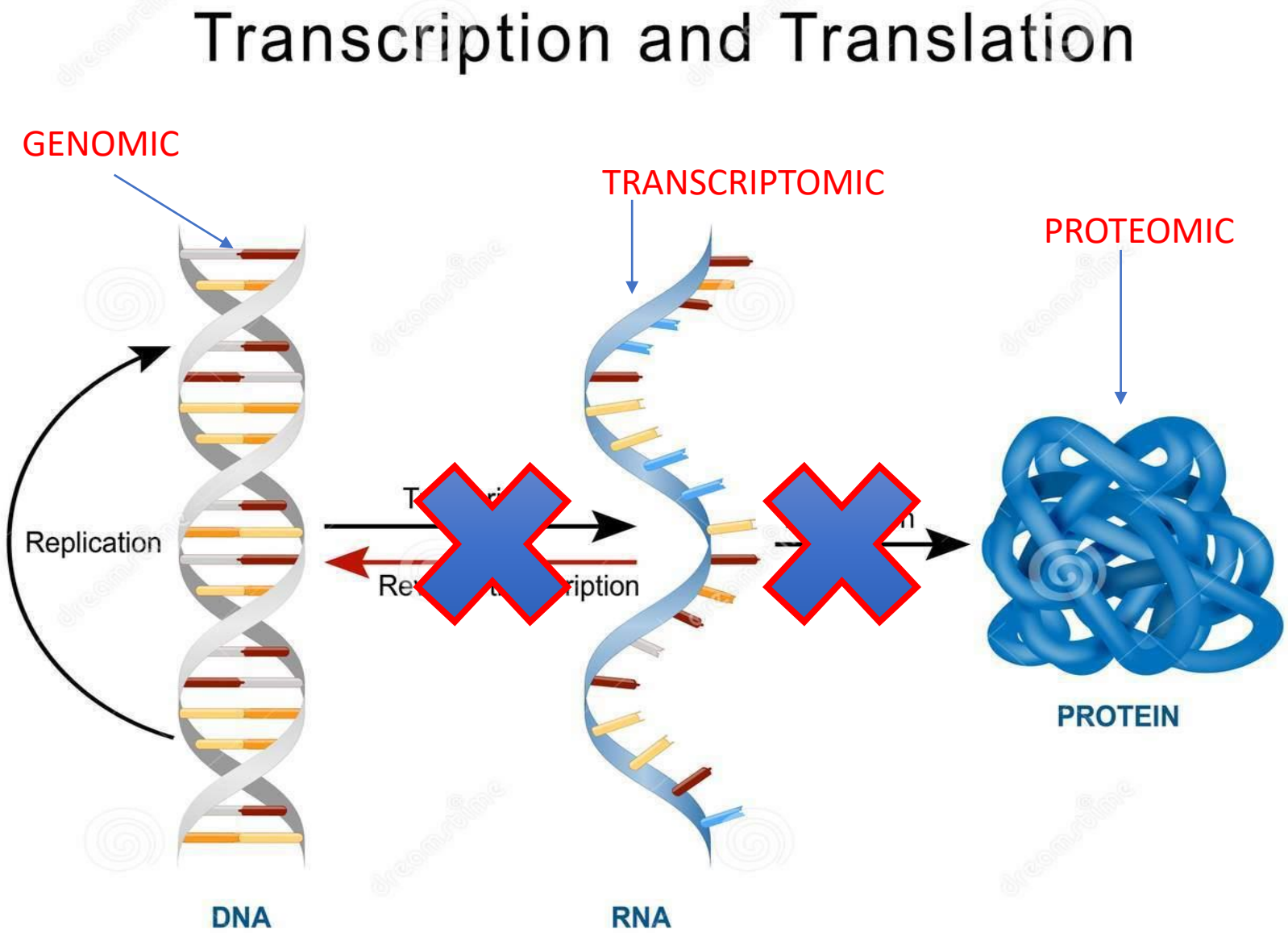
↑ VF inducibility
↑ Risk of ventricular arrhythmias

Epicardial fibrosys and voltage reduction

Oligogenic or Mendelian Disease?

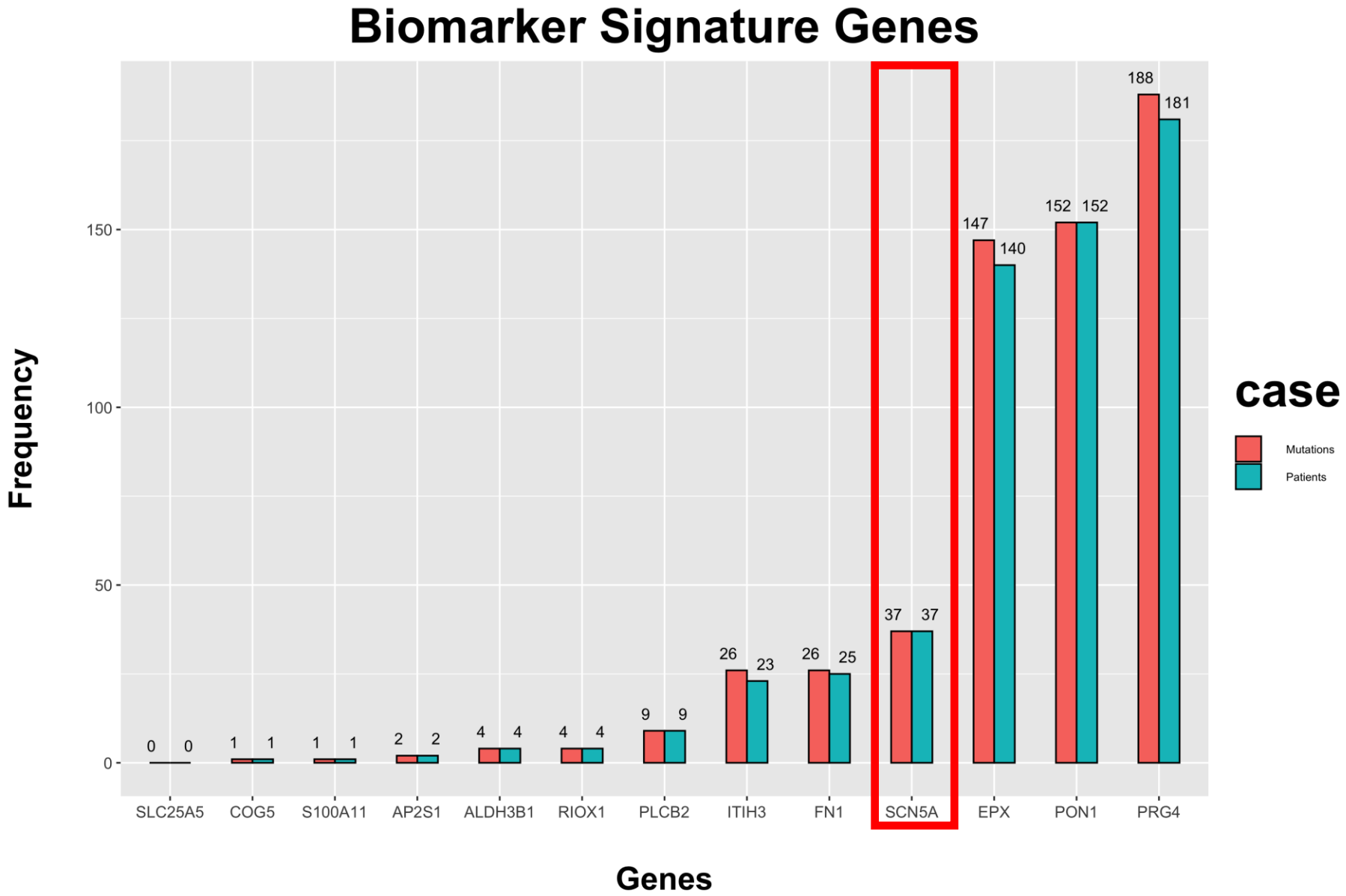


EFFECT OF A GENETIC ALTERATION (MUTATION OR DELETION) AGAINST



Mutational landscape for genes in the Biomarker Signature

(proteomics+metabolomis+lipidomics)



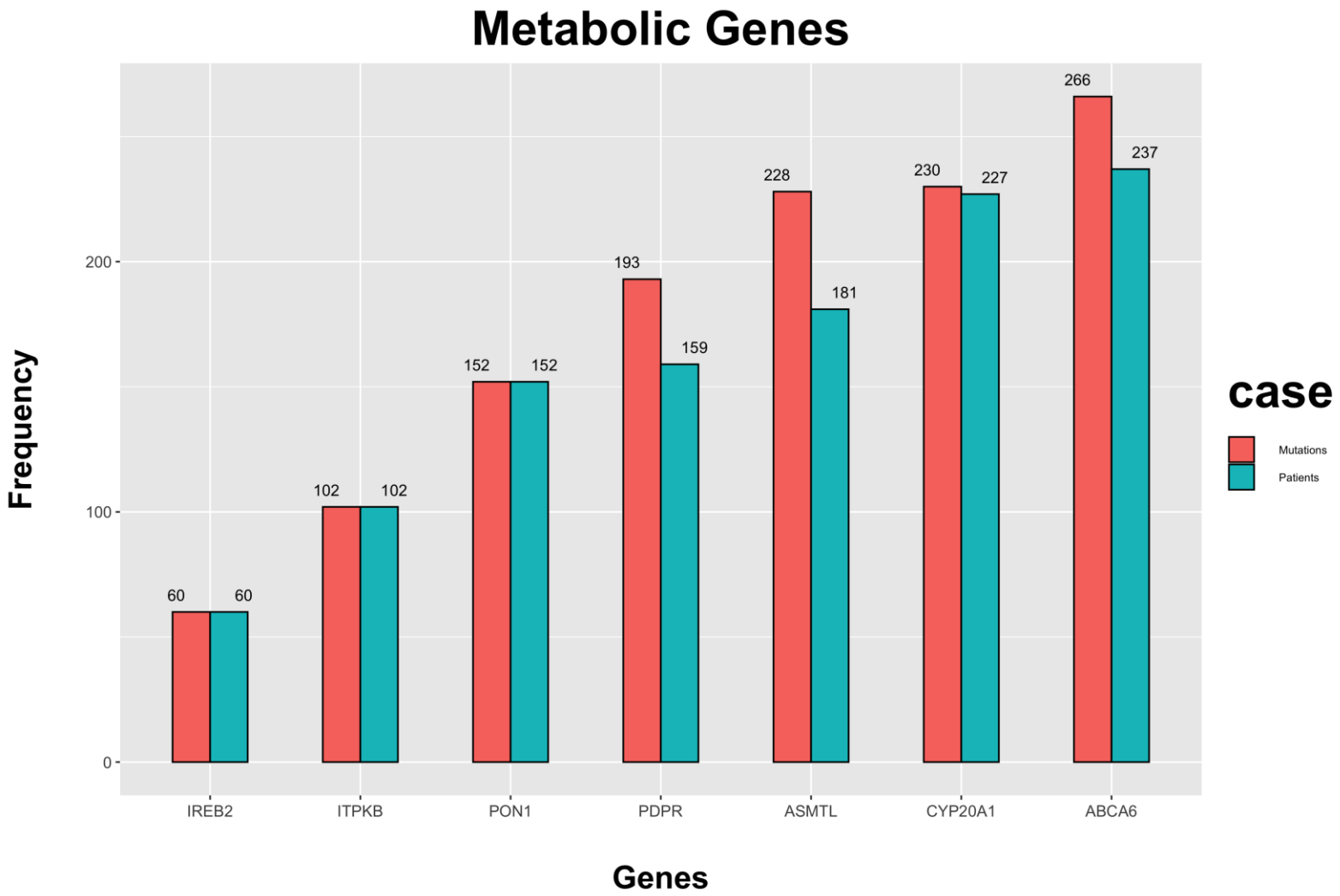
Assessment of the mutational landscape of a subset of Metabolic, Channel-related & Structural Genes

- Exploring the mutational landscape of:
  - ❑ All top-ranking and mid-ranking protein popping out in the biomarker feature selection process
  - ❑ Proteins associated with multiple metabolic processes also predicted as highly informative for BrS/CTL differentiation in the biomarker feature selection process
  - ❑ Channel proteins and channel-interacting proteins linked to the different Brugada phenotypes described so far
  - ❑ Structural proteins associated to Desmosome assembly, function and maintenance

❖ All the data presented here correspond to Single Nucleotide Variants classified as Deleterious/Damaging by deleteriousness predictors

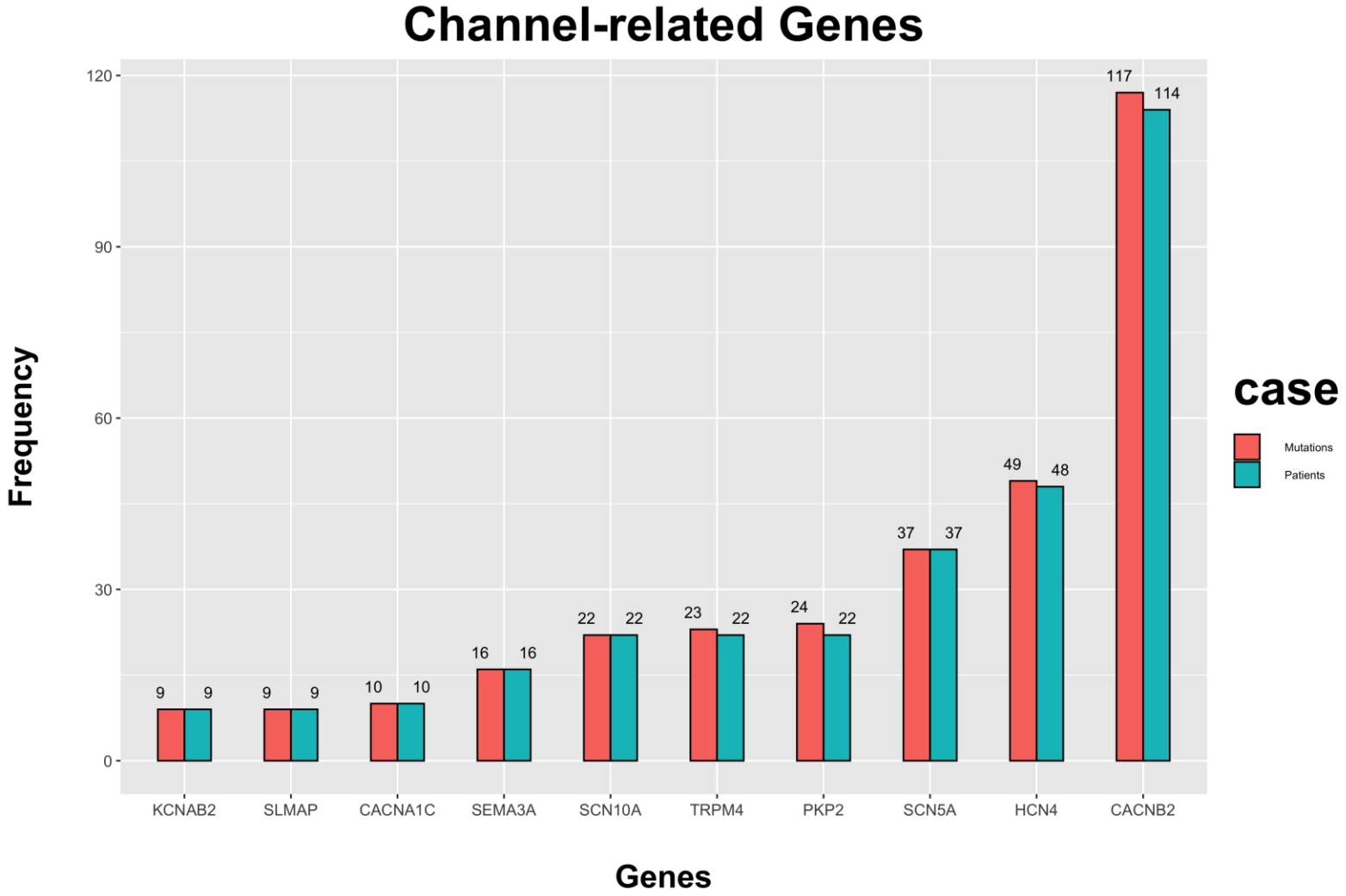
❖ In total 115 genes have been analyzed in detail so far to assess their mutational landscape across **259** Brugada patients (**whole BrS cohort**)

Mutational landscape for Genes involved in Metabolic Processes



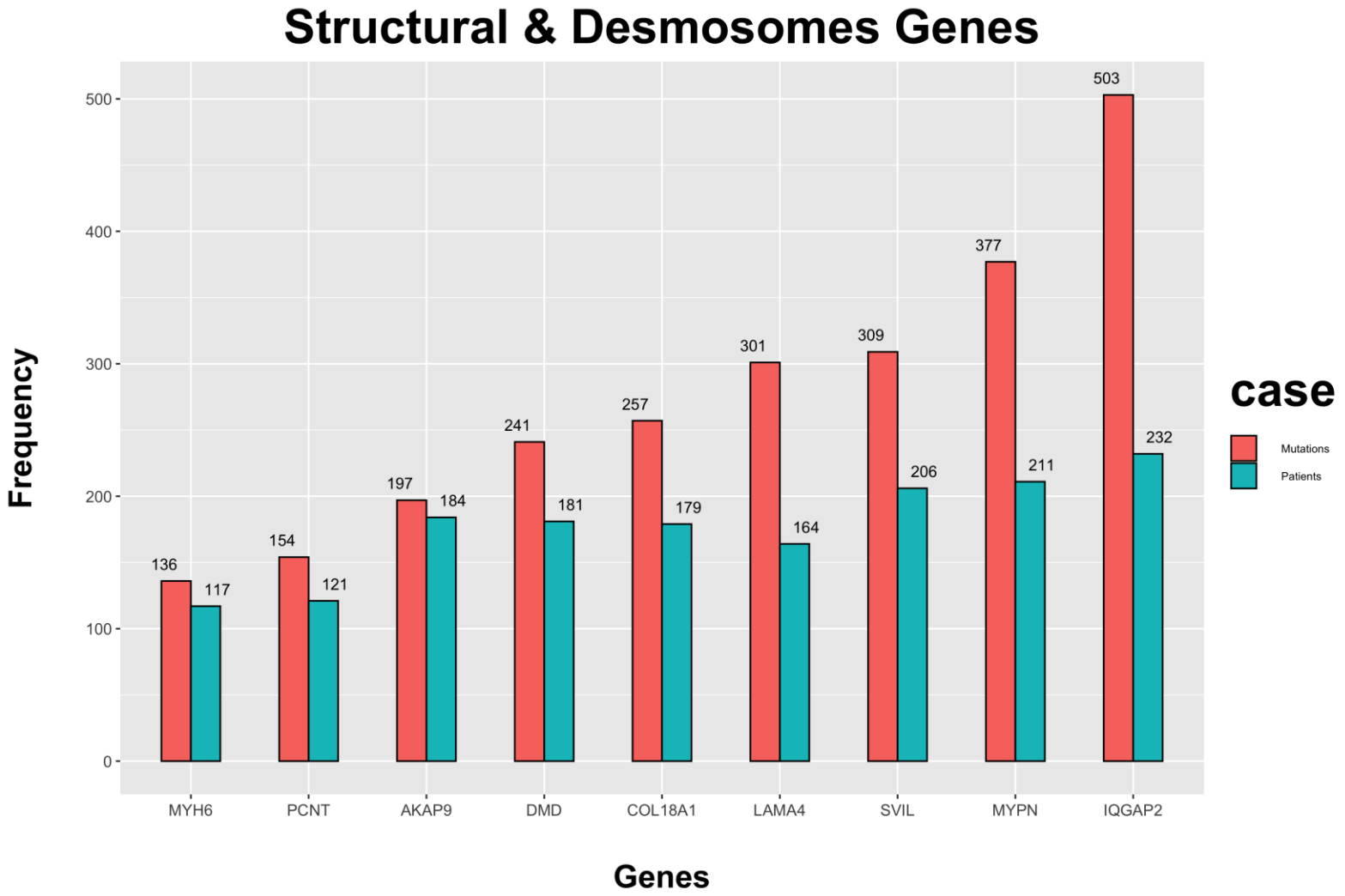
- **ITPKB:** Inositol-Trisphosphate 3-Kinase B. Regulates inositol phosphate metabolism by phosphorylation of second messenger inositol involved in cellular signalling.
- **IREB2:** Iron Responsive Element Binding Protein 2. RNA-binding protein that acts to regulate iron levels in the cells by regulating the translation and stability of mRNAs that affect iron homeostasis
- **ABCA6:** ATP Binding Cassette Subfamily A Member 6. Transport of glucose and other sugars, bile salts and organic acids, metal ions and amine compounds. ATPase activity, coupled to transmembrane movement of substances
- **CYP20A1:** Cytochrome P450 Family 20 Subfamily A Member 1. Involved in drug metabolism and synthesis of cholesterol, steroids and other lipids, iron ion binding and oxidoreductase activity, acting on paired donors, with incorporation or reduction of molecular oxygen
- **ASMTL:** Acetylserotonin O-Methyltransferase Like. Nucleoside triphosphate pyrophosphatase that hydrolyzes dTTP and UTP, dual role in cell division arrest and in preventing the incorporation of modified nucleotides into cellular nucleic acids
- **PDPRL:** Pyruvate Dehydrogenase Phosphatase Regulatory Subunit. Catalyzes the oxidative decarboxylation of pyruvate and links glycolysis to the tricarboxylic acid cycle and fatty acid synthesis. **Key regulatory protein in the Pyruvate metabolism and Citric Acid (TCA)**
- **PON1:** Serum Paraoxonase/Arylesterase 1. exhibits lactonase and ester hydrolase activity. **Synthesized in the kidney and liver**, is secreted into the blood, where it binds to high density lipoprotein (HDL) and Ca^{2+}. Diseases associated with PON1 include Microvascular Complications Of Diabetes and Amyotrophic Lateral Sclerosis 1

Mutational landscape for Channel or Channel Interacting Genes



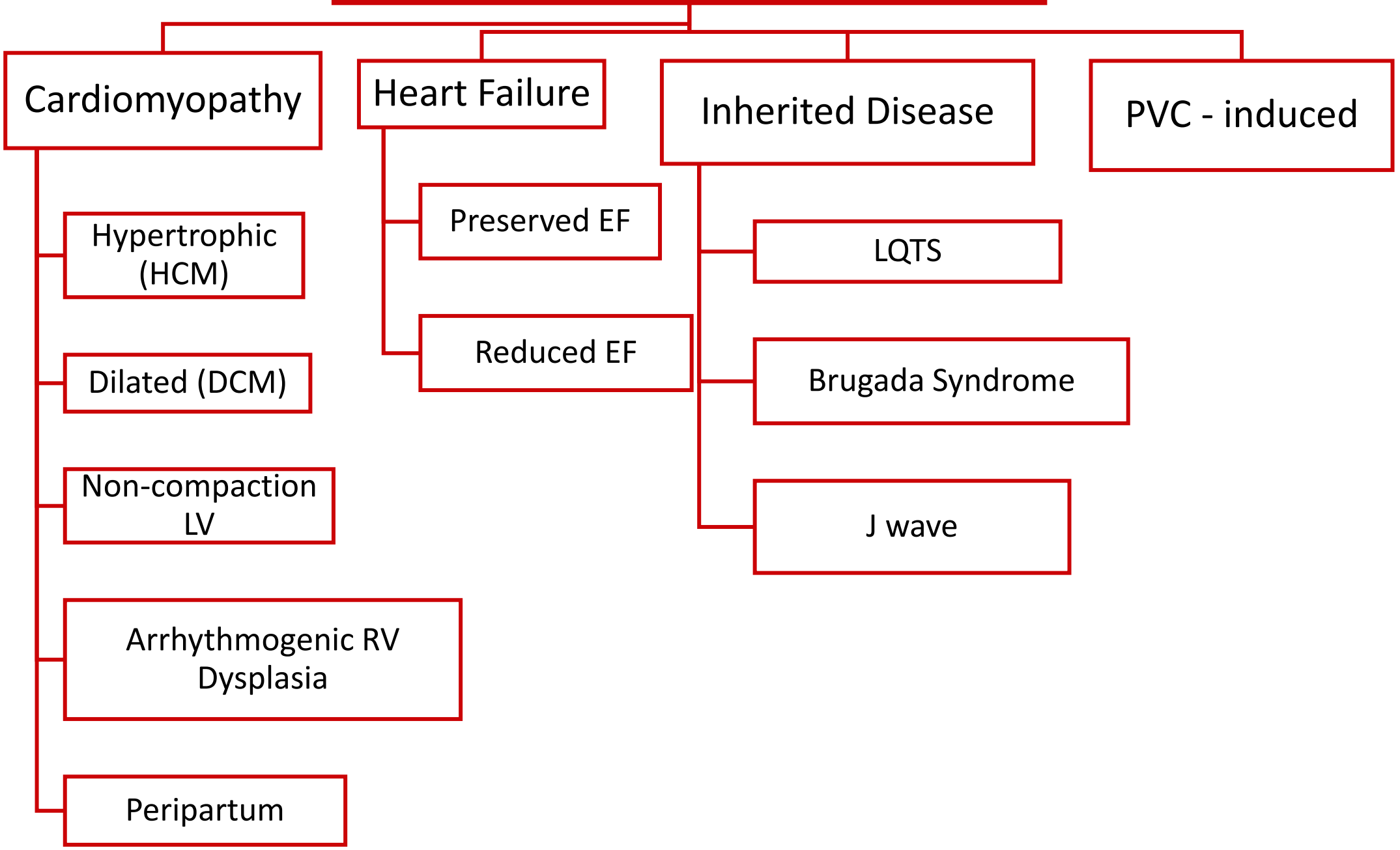
- **TRPM4:** Transient Receptor Potential Cation Channel Subfamily M Member 4. **calcium-activated nonselective ion channel** that mediates transport of monovalent cations across membranes, thereby depolarizing the membrane
- **KCNAB2:** Potassium Voltage-Gated Channel Subfamily A Regulatory Beta Subunit 2. functions **include regulating neurotransmitter release**, heart rate, insulin secretion, neuronal excitability, epithelial electrolyte transport, smooth muscle contraction, and cell volume
- **CACNB2:** Calcium Voltage-Gated Channel Auxiliary Subunit Beta 2. The beta subunit of voltage-dependent calcium channels contributes to the function of the calcium channel by increasing peak calcium current, **shifting the voltage dependencies** of activation and inactivation, **modulating G protein inhibition** and controlling the alpha-1 subunit membrane targeting
- **CACNA1C:** Calcium Voltage-Gated Channel Subunit Alpha1 C.
- **HCN4:** Hyperpolarization Activated Cyclic Nucleotide Gated Potassium Channel 4. Hyperpolarization-activated ion channel with very slow activation and inactivation exhibiting weak selectivity for potassium **over sodium ions**. Contributes to the **native pacemaker currents** in heart that regulate the rhythm of heart beat
- **SCN5A:** Sodium Voltage-Gated Channel Alpha Subunit 5. Responsible for action potential initiation and propagation in **excitable cells**, including nerve, muscle, and neuroendocrine cell types
- **SCN10A:** Sodium Voltage-Gated Channel Alpha Subunit 10.
- **PKP2:** Plakophilin 2. Plakophilin proteins contain numerous armadillo repeats, **localize to cell desmosomes and nuclei**, and participate in linking **cadherins to intermediate filaments in the cytoskeleton**

Mutational landscape for Structural & Desmosome Genes



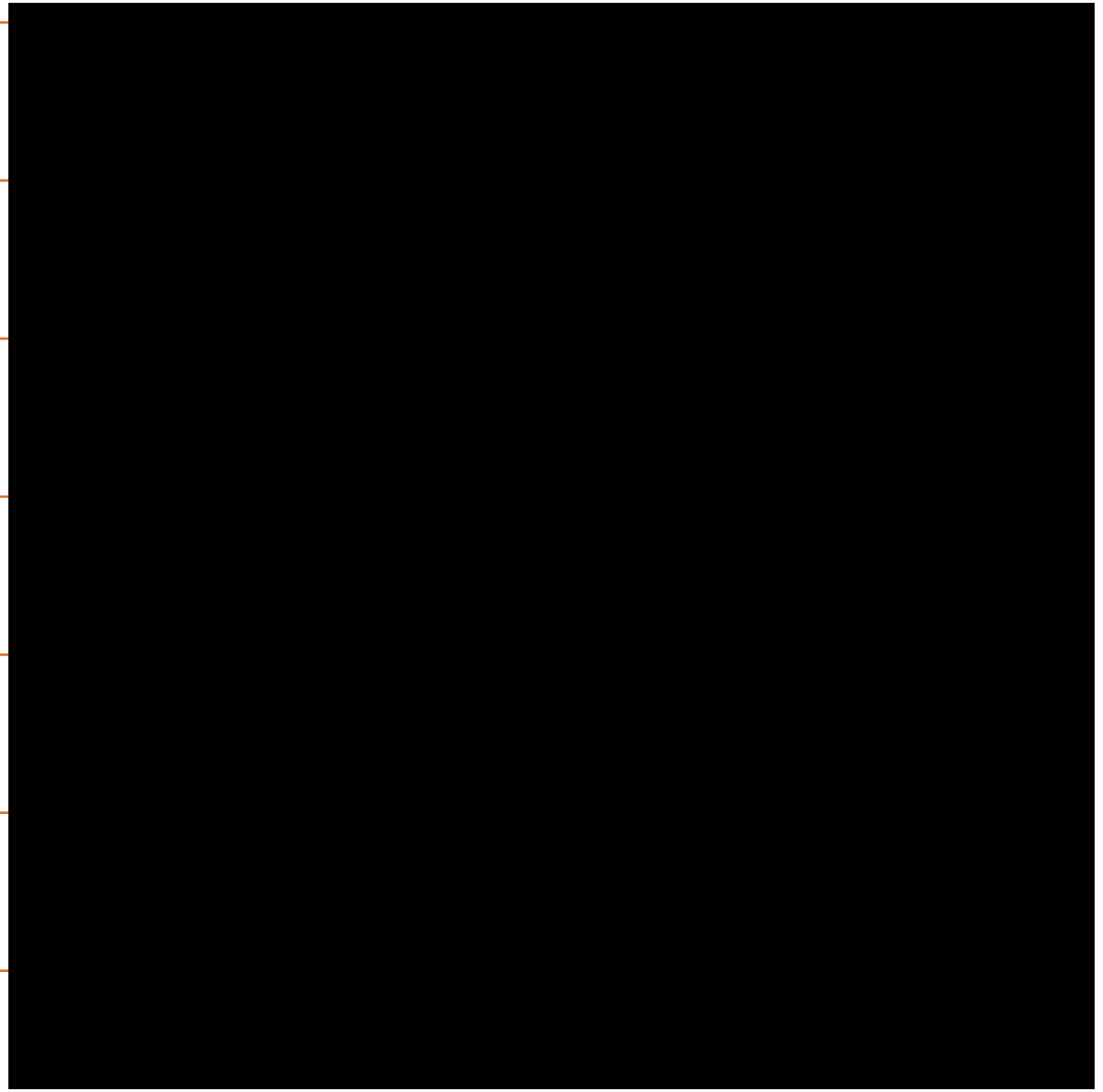
- **DMD:** Dystrophin. **Bridges the inner cytoskeleton and the extracellular matrix**. Component of the dystrophin-associated glycoprotein complex which accumulates at the neuromuscular junction (NMJ) and at a variety of synapses in the peripheral and central nervous systems and has a structural function in stabilizing the sarcolemma
- **PCNT:** Pericentrin. **Interacts with the microtubule** nucleation component gamma-tubulin and is likely important to normal functioning of the centrosomes, cytoskeleton, and cell-cycle progression
- **IQGAP2:** IQ Motif Containing GTPase Activating Protein 2. **Interacts with components of the cytoskeleton**, with cell adhesion molecules, and with several signaling molecules to regulate cell morphology and motility.
- **MYPN:** Myopalladin. Interacts with **nebulin in skeletal muscle or nebulette in cardiac muscle and alpha-actinin**. In addition, this gene product can interact with a protein with the I-band indicating it has a regulatory as well as structural function
- **SVIL:** Supervillin. **Tightly associated with both actin filaments and plasma membranes**, with a role as a high-affinity link between the actin cytoskeleton and the membrane. The encoded protein appears to **aid in both myosin II assembly during cell spreading and disassembly of focal adhesions**
- **LAMA4:** Laminin Subunit Alpha 4. Binding to cells via a high affinity receptor, laminin is thought to mediate the attachment, migration and organization of cells into tissues during embryonic development by interacting with other extracellular matrix components
- **COL18A1:** Collagen Type XVIII Alpha 1 Chain. **regulate extracellular matrix-dependent motility** and morphogenesis of endothelial and non-endothelial cells; the function requires homotrimerization and implicates MAPK signaling
- **AKAP9:** A-Kinase Anchoring Protein 9. Scaffolding protein that **assembles several protein kinases and phosphatases on the centrosome.** Required for association of the centrosomes with the poles of the bipolar mitotic spindle during metaphase

Ventricular Fibrillation Causes

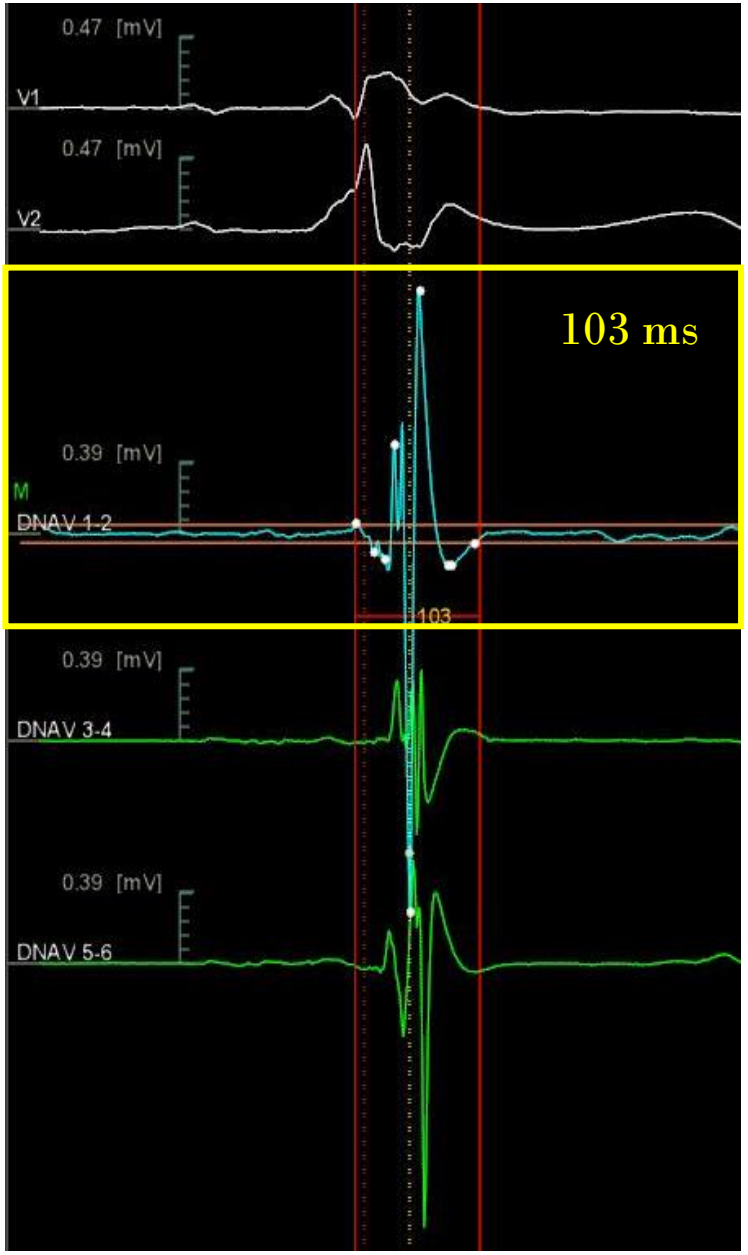


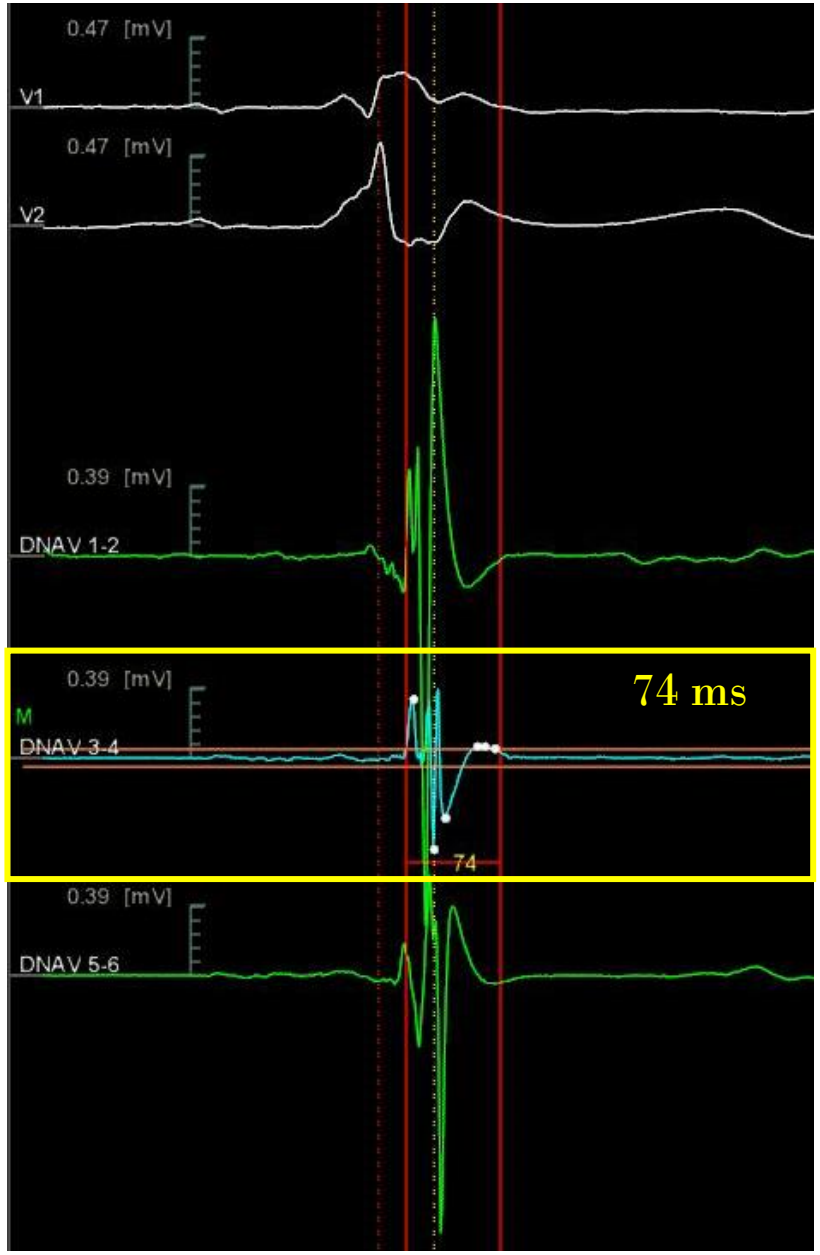
CATH-LAB SETTINGS

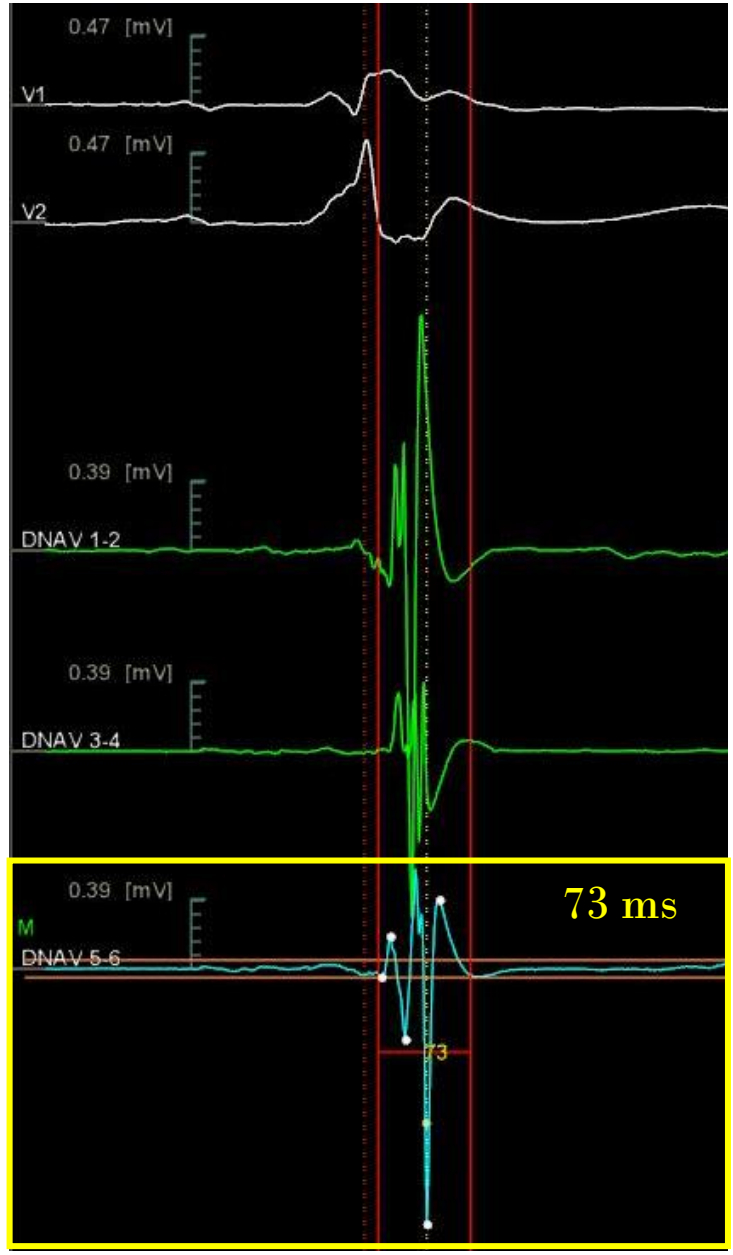
General anesthesia and mechanical ventilation with invasive arterial line for blood pressure monitoring
Diagnostic quadripolar catheter in RV apex
Epicardial access
ThermoCool SF 4 mm irrigated tip mapping and ablation catheter
ECG filtered: 0,05-200 Hz
Signals filters: 16-500 Hz
30 minutes of total ablation on a 16,7 cm^2 area



NORMAL EPICARDIAL VENTRICULAR ELECTROGRAMS

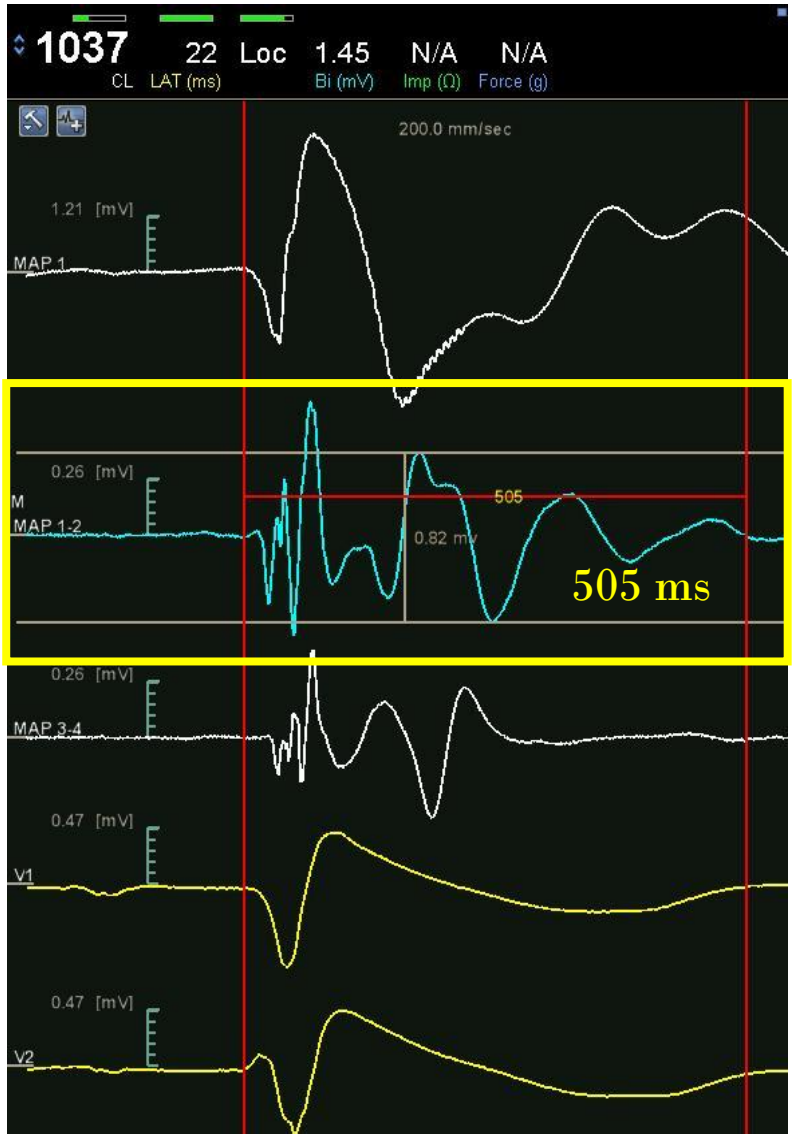




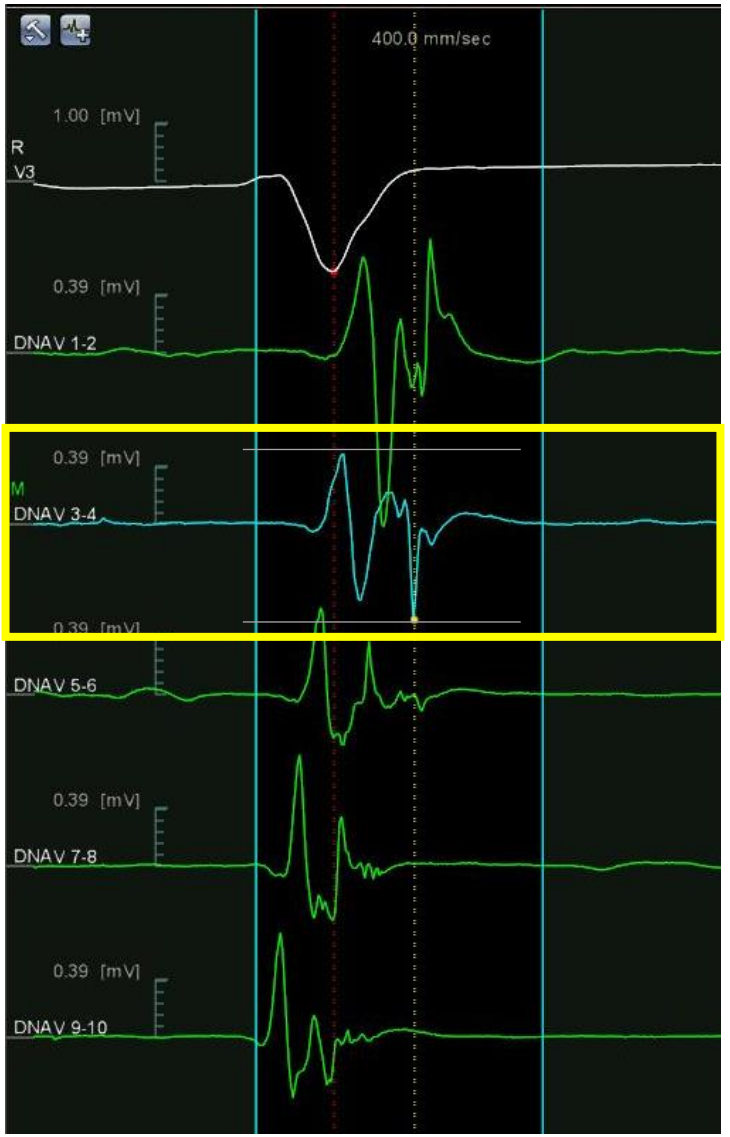


ABNORMAL EPICARDIAL VENTRICULAR ELECTROGRAMS

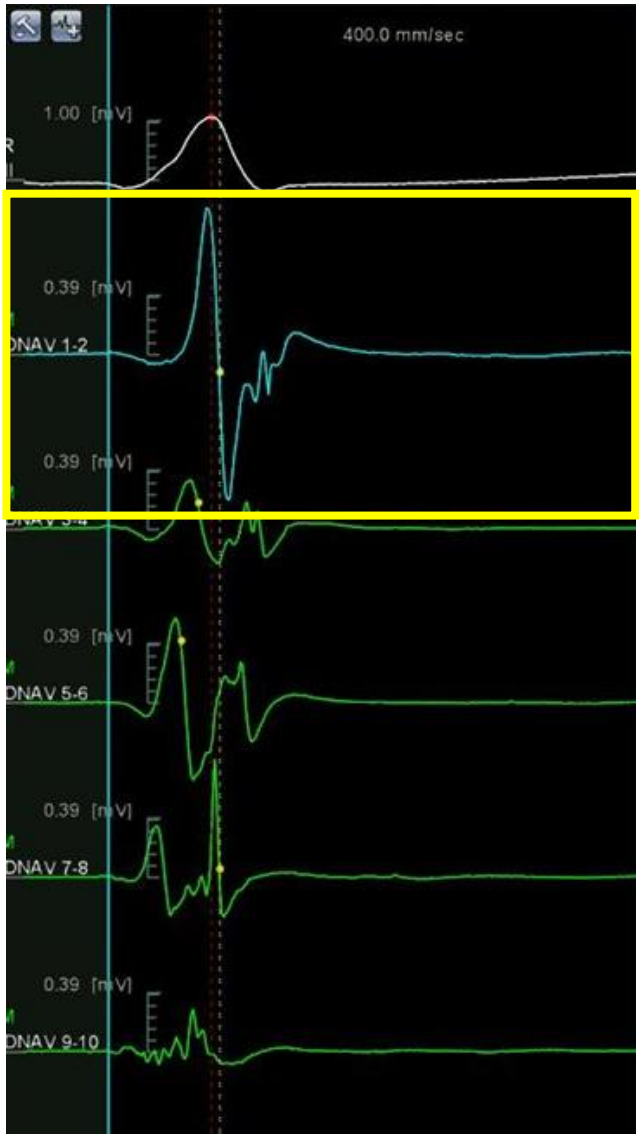
Brugada Syndrome



Long QT Syndrome



J-Wave Syndrome



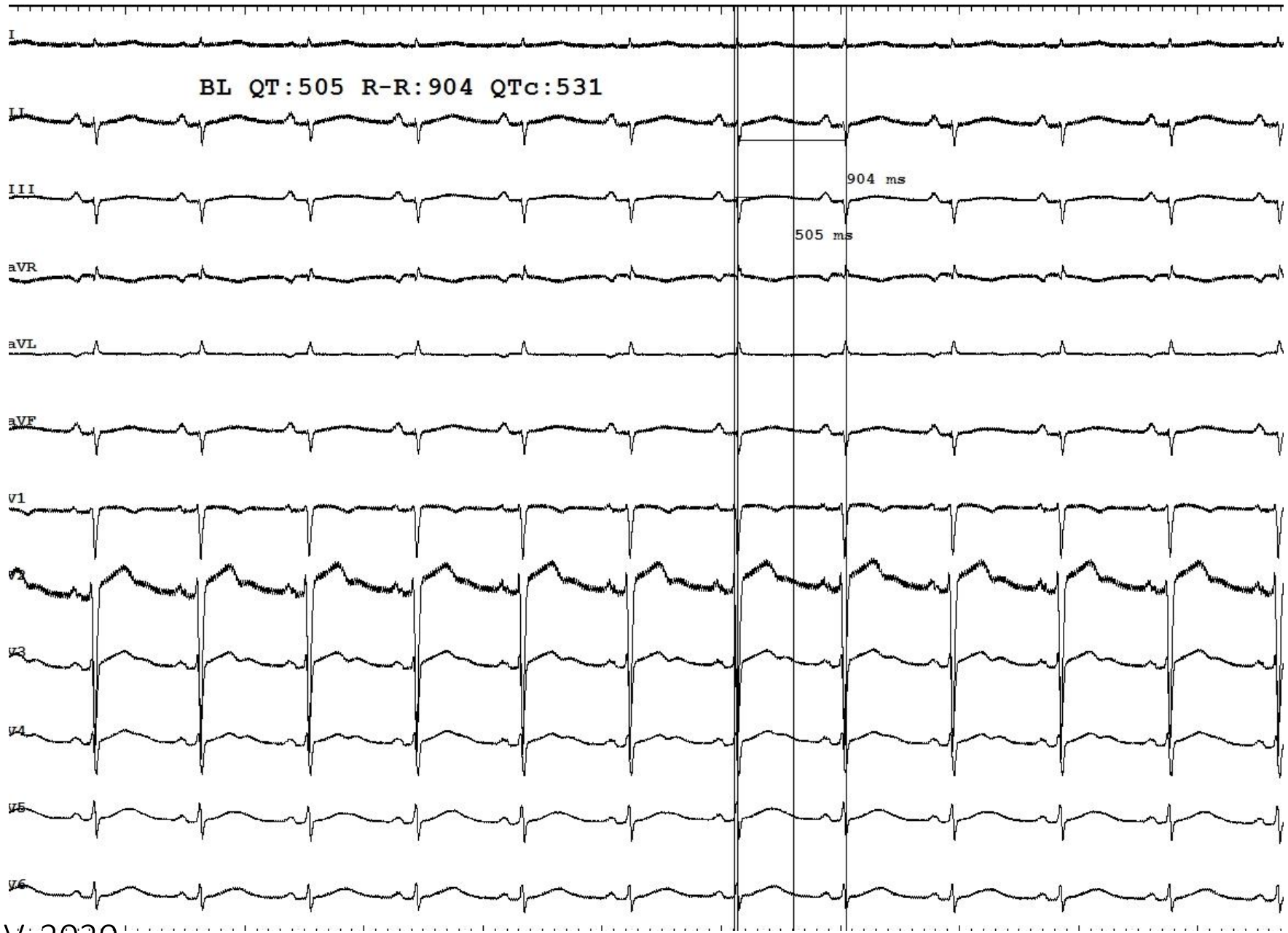
Long QT syndrome

SUMMARY

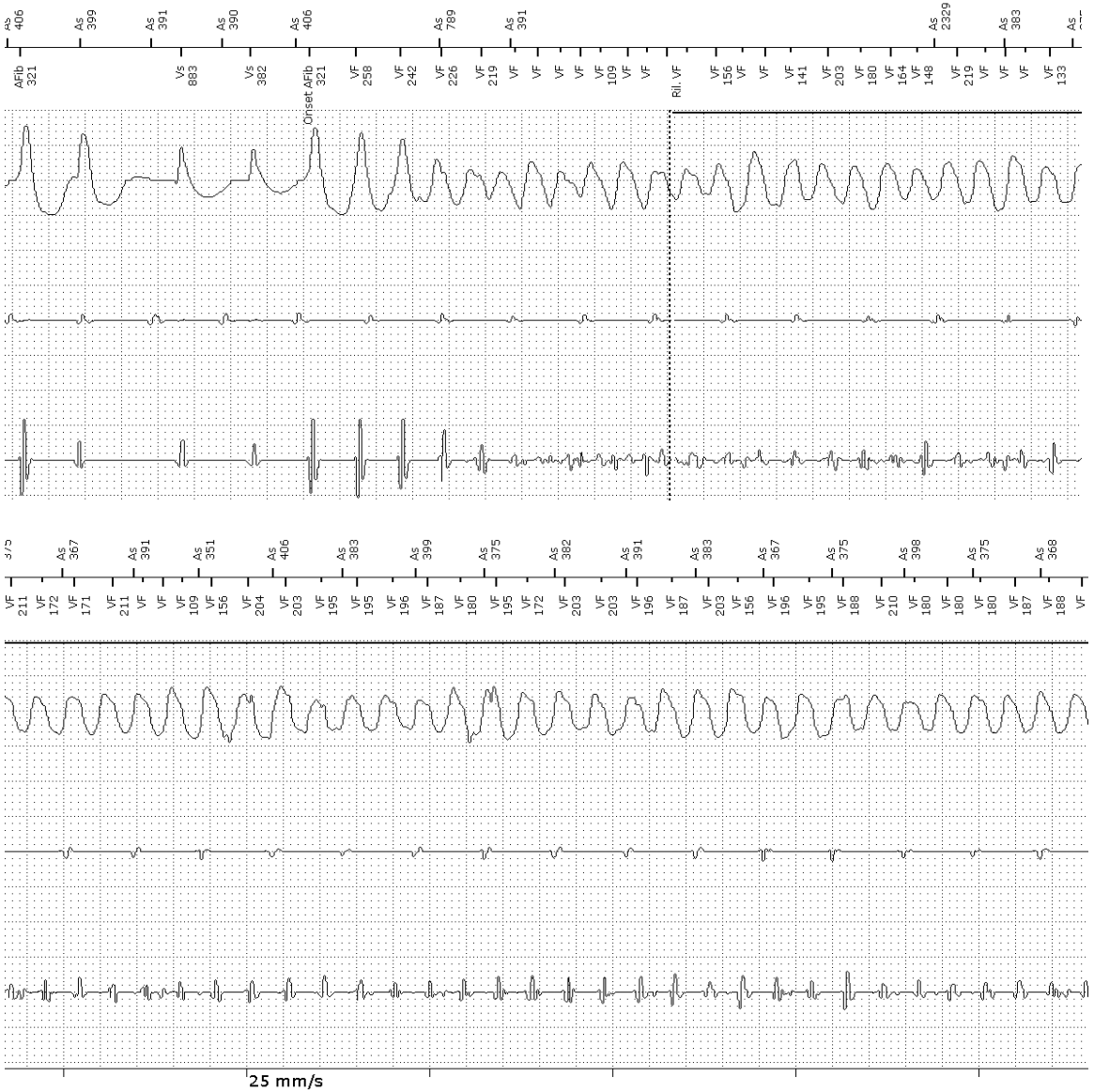
- Male 44 YO
- 2011: lipotimic episodes, II-degree AV block type I and type II
- 2012: lipotimic episodes due to polymorphic ventricular tachycardia, Holter episodes of ventricular trigeminy, negative flecainide test, severe nodal disfunction, ICD implantation
- 2015, 2016: VT and TdP episodes
- 2017: negative ajmaline test (NO Brugada sign), positive adrenaline test for LQTS
- KCNE1 gene mutation
- 2020: Several episodes of VF (admission reason)

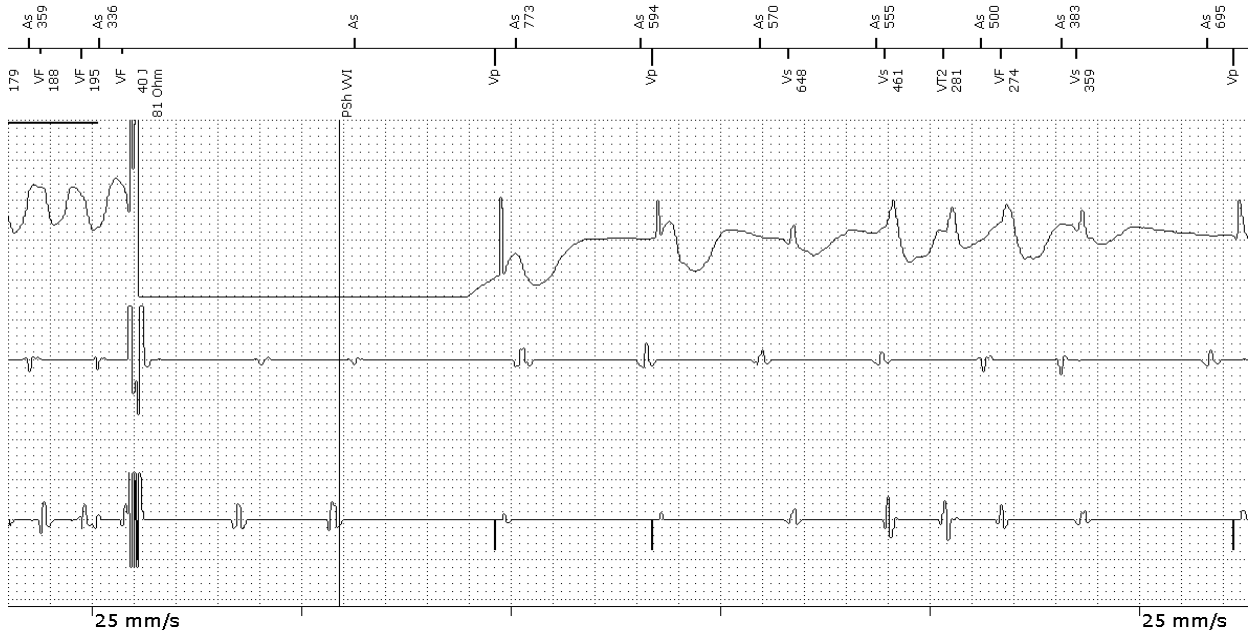
LQTs ECG

QT = 531ms



VF EPISODE (ICD registration, 06/12/2020)





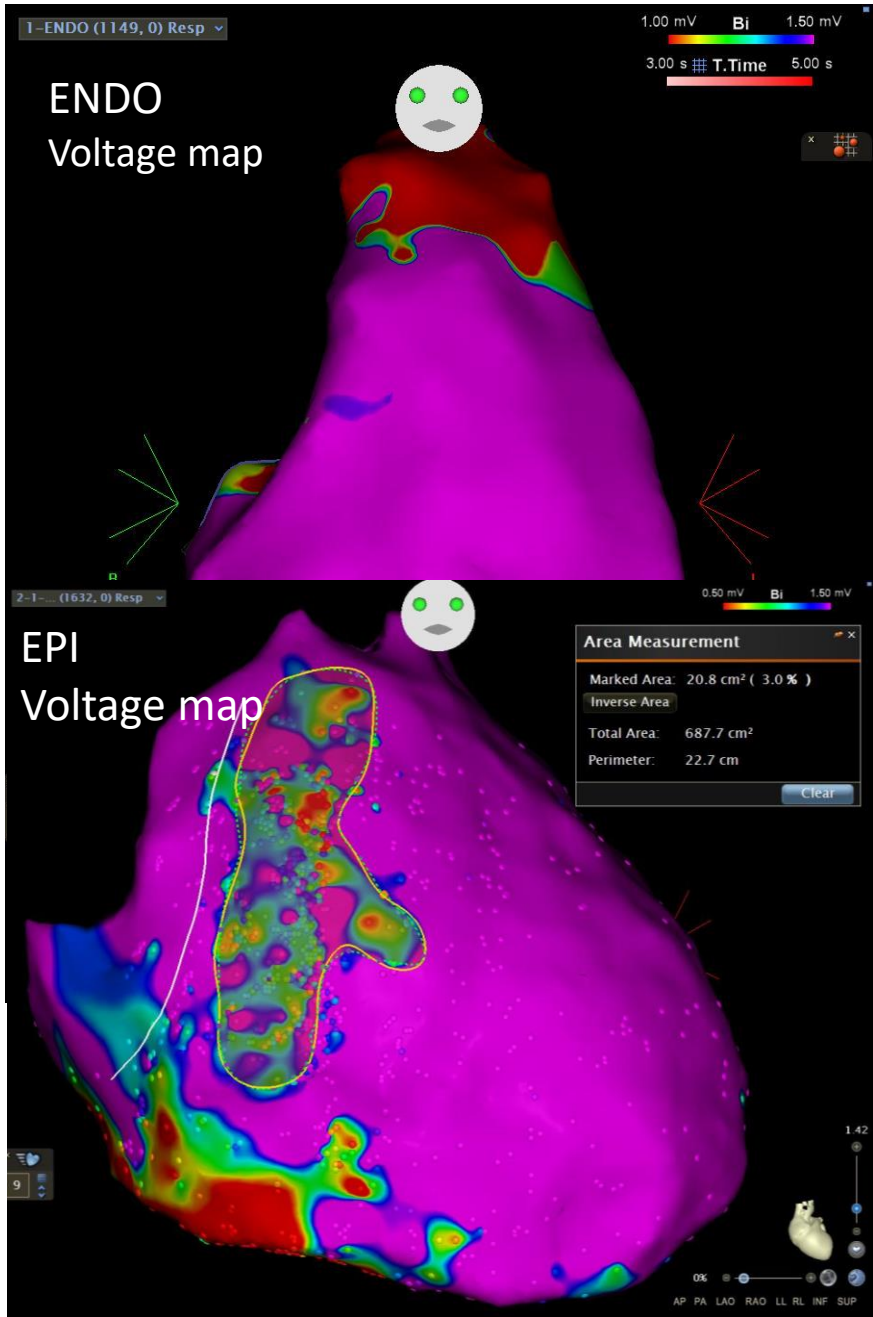
MAPPING DETAILS

Ablation date 09-12-2020

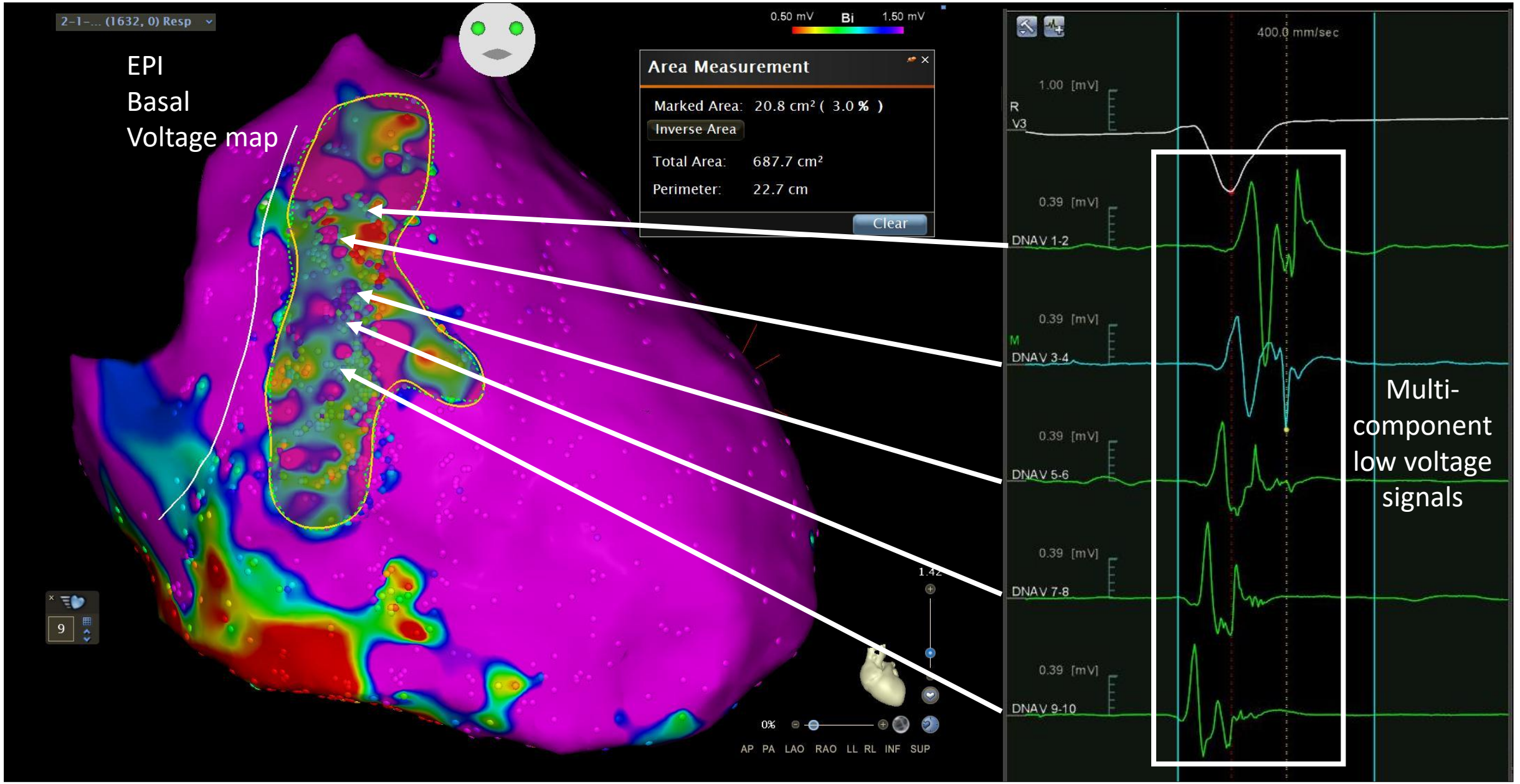
DecaNAV substrate mapping catheter

Endocardial mapping - Healthy RV

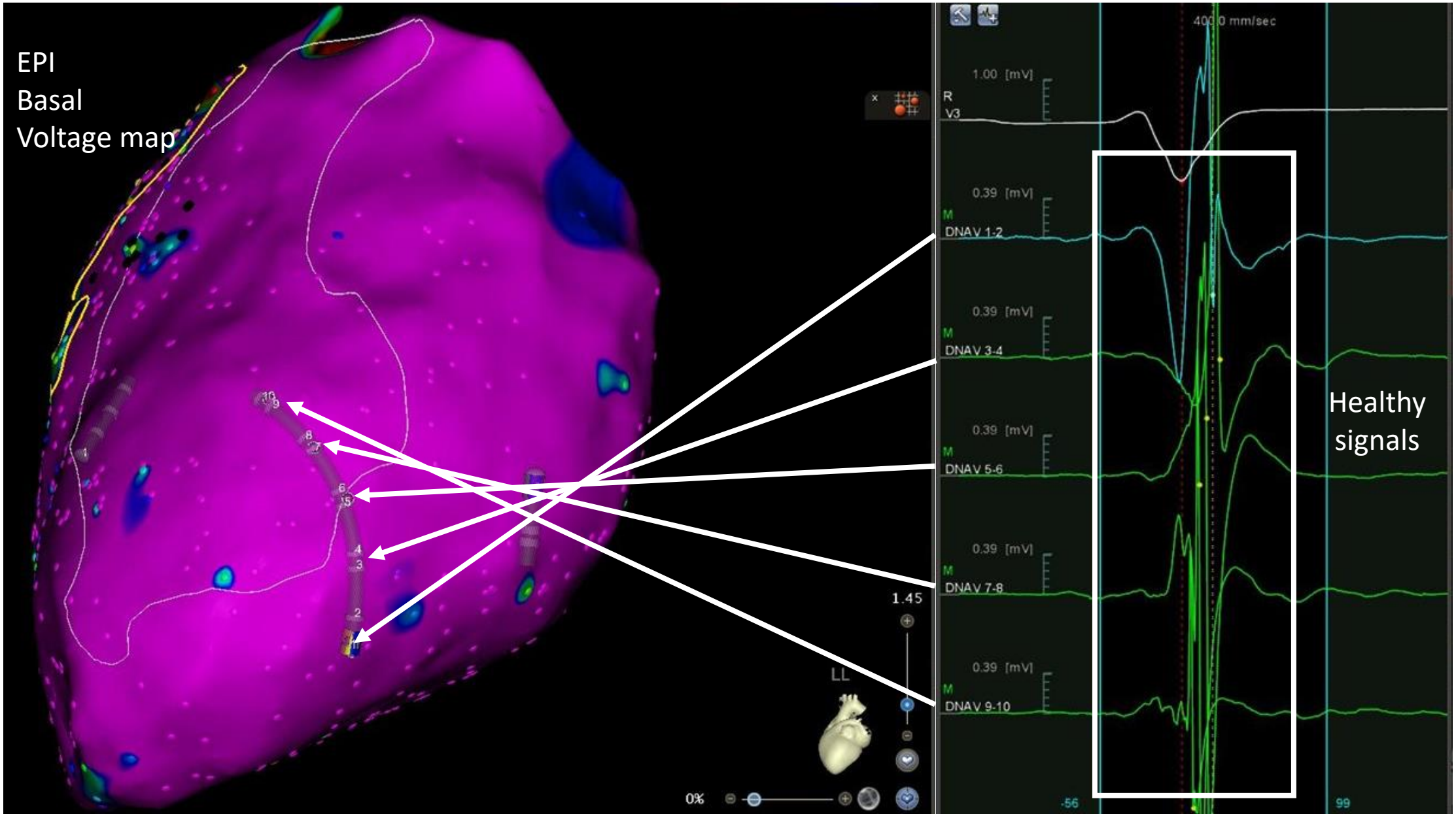
Epicardial mapping - Low voltage area on anterolateral wall



SIGNAL ANALYSIS – Target area



SIGNAL ANALYSIS – Outside the target area

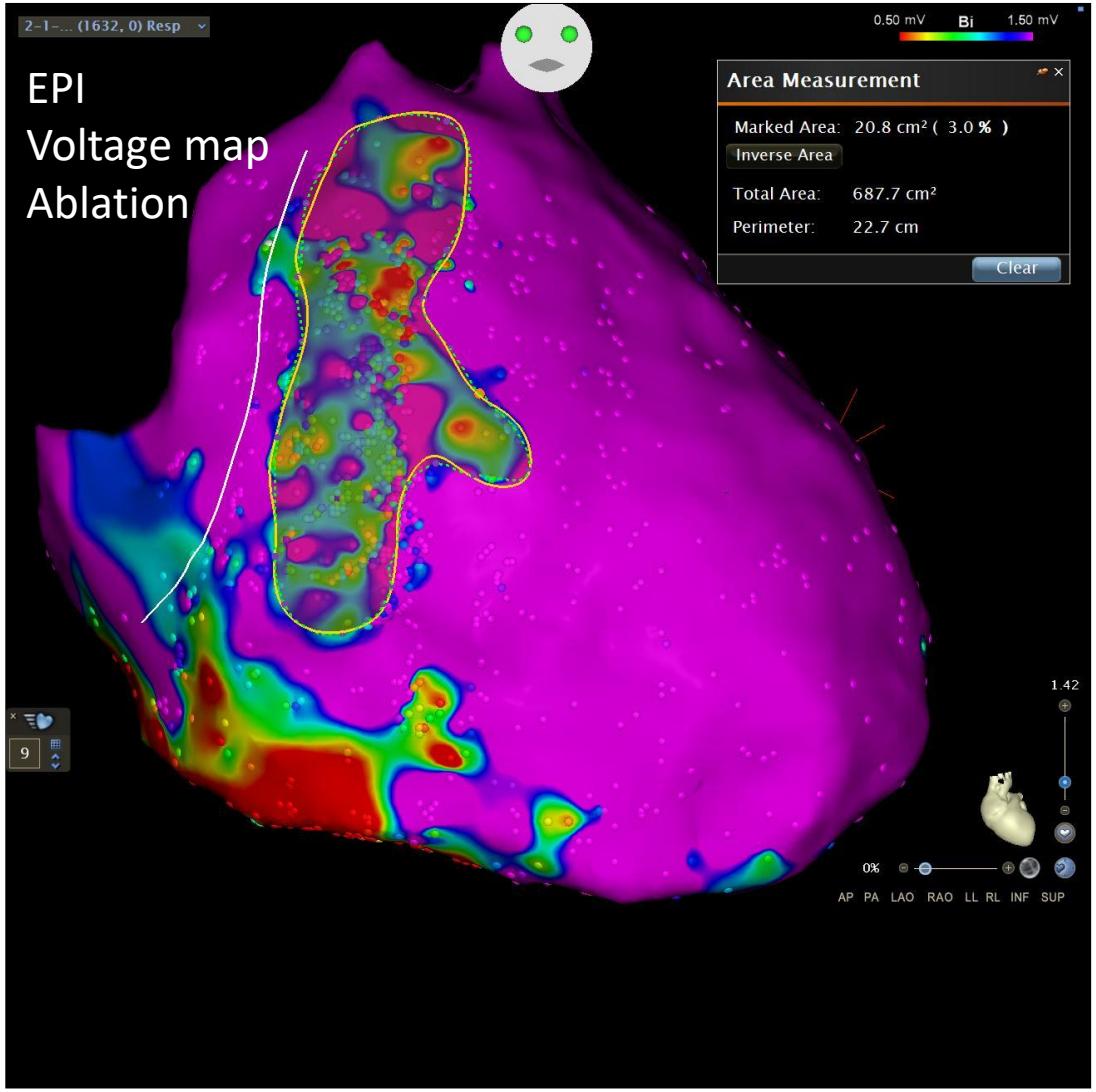


ABLATION STRATEGY

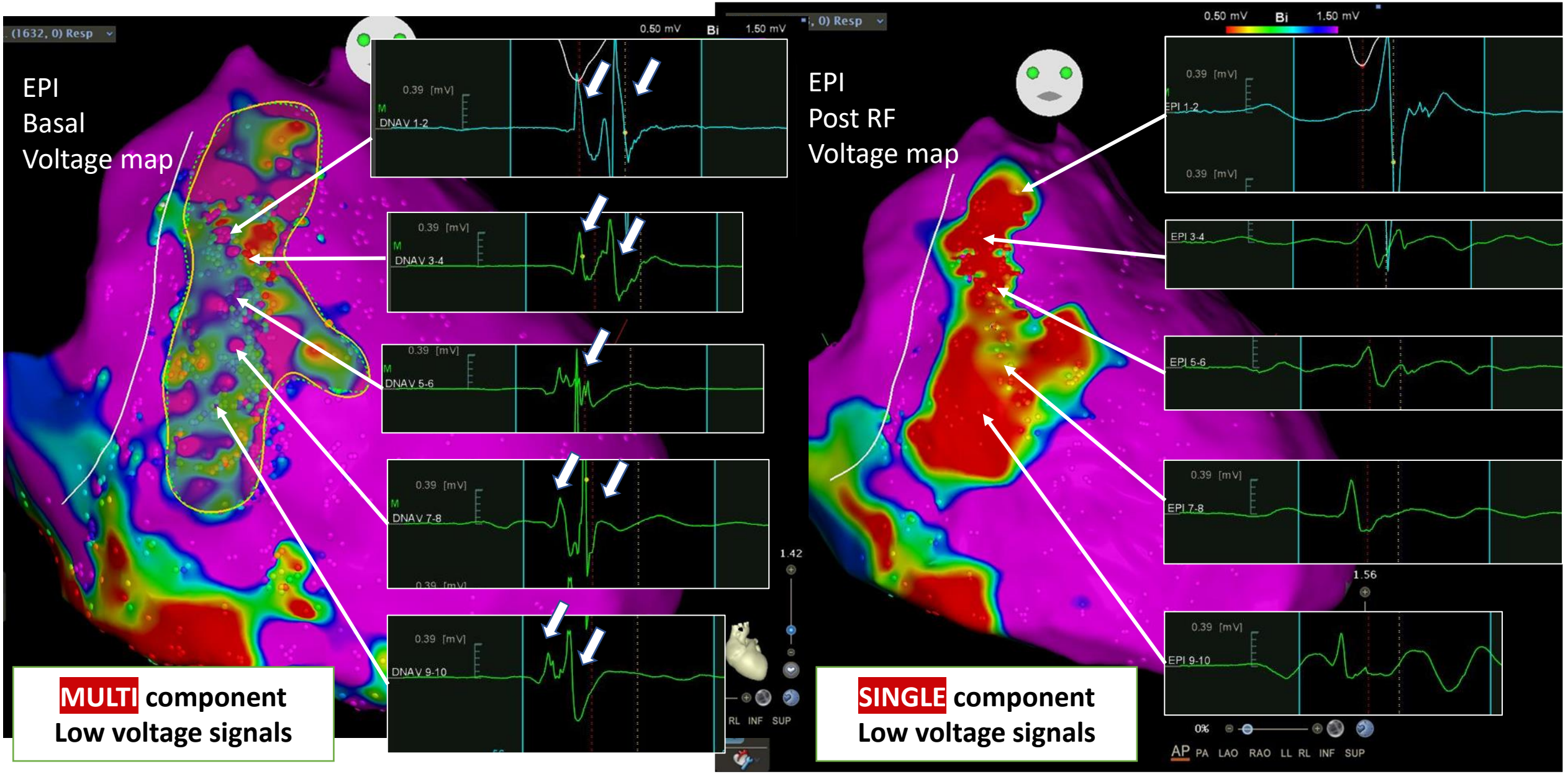
Homogenization of fragmented low voltage potentials area with SmartTouch SF catheter

40-45 watts with fast dragging technique, to reach superficial lesion

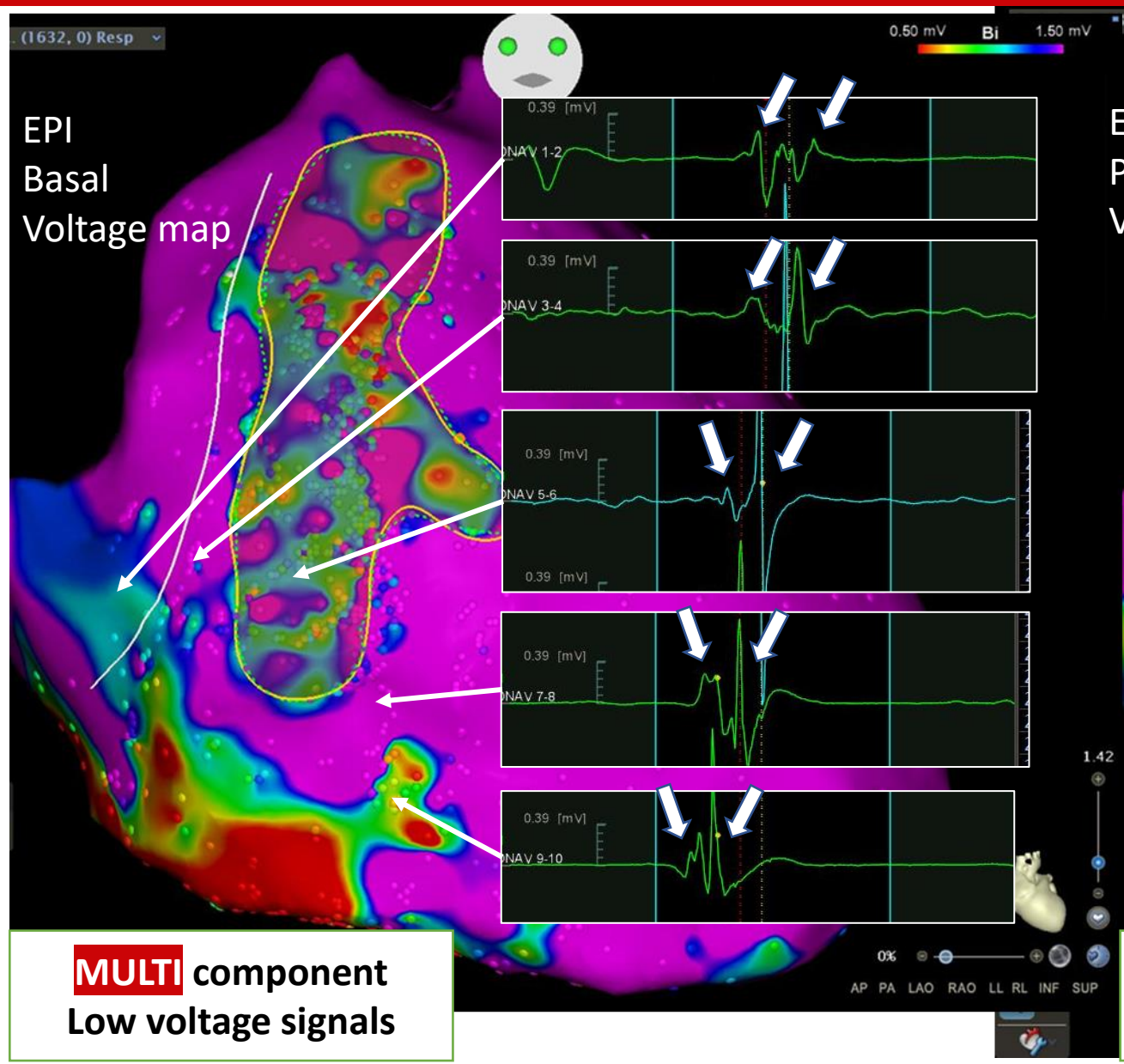
42 minutes of total ablation for a 21.2 cm^2 area



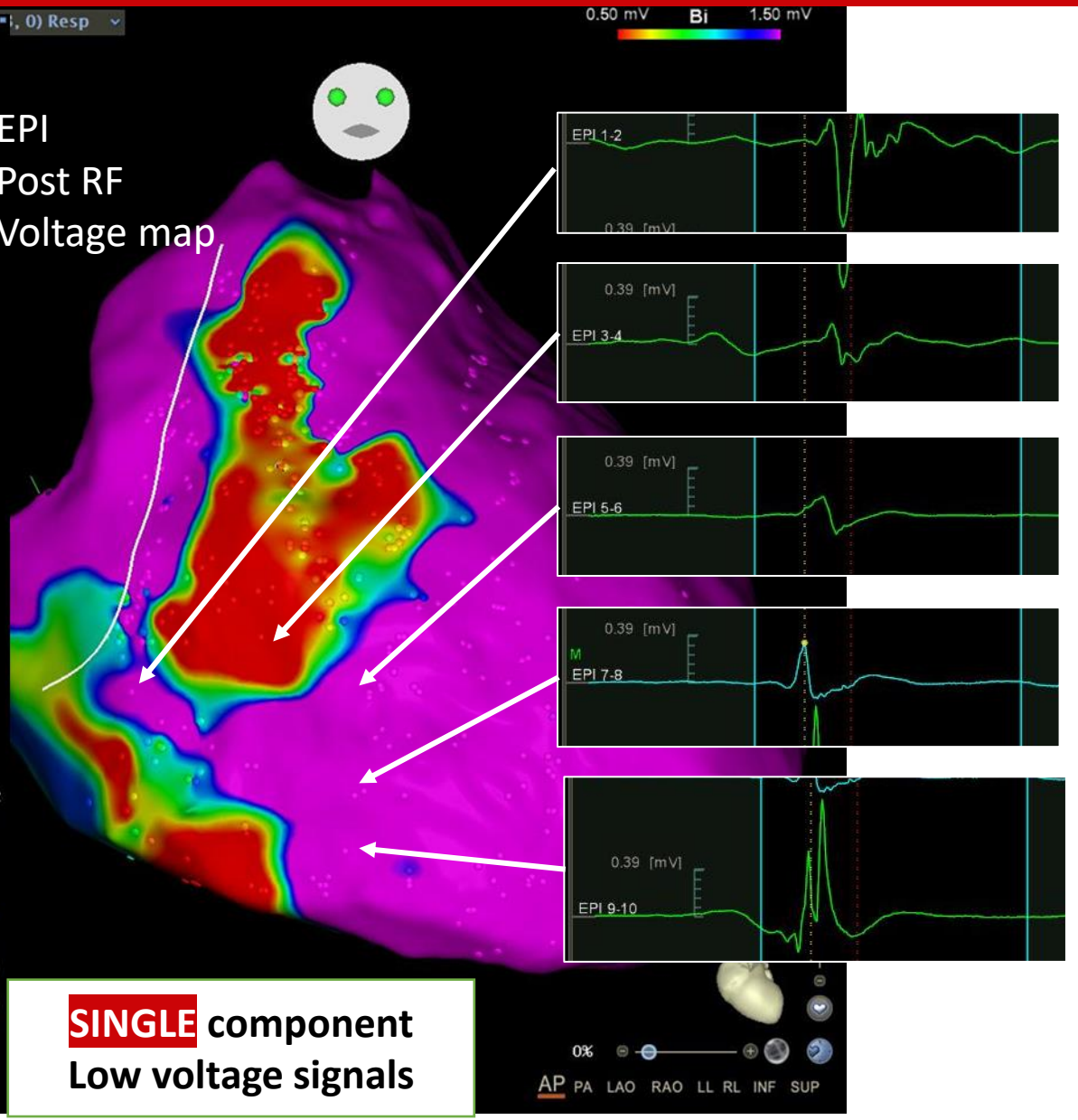
SIGNAL COMPARISON in the target area: PRE AND POST ABLATION



SIGNAL COMPARISON in the target area: PRE AND POST ABLATION



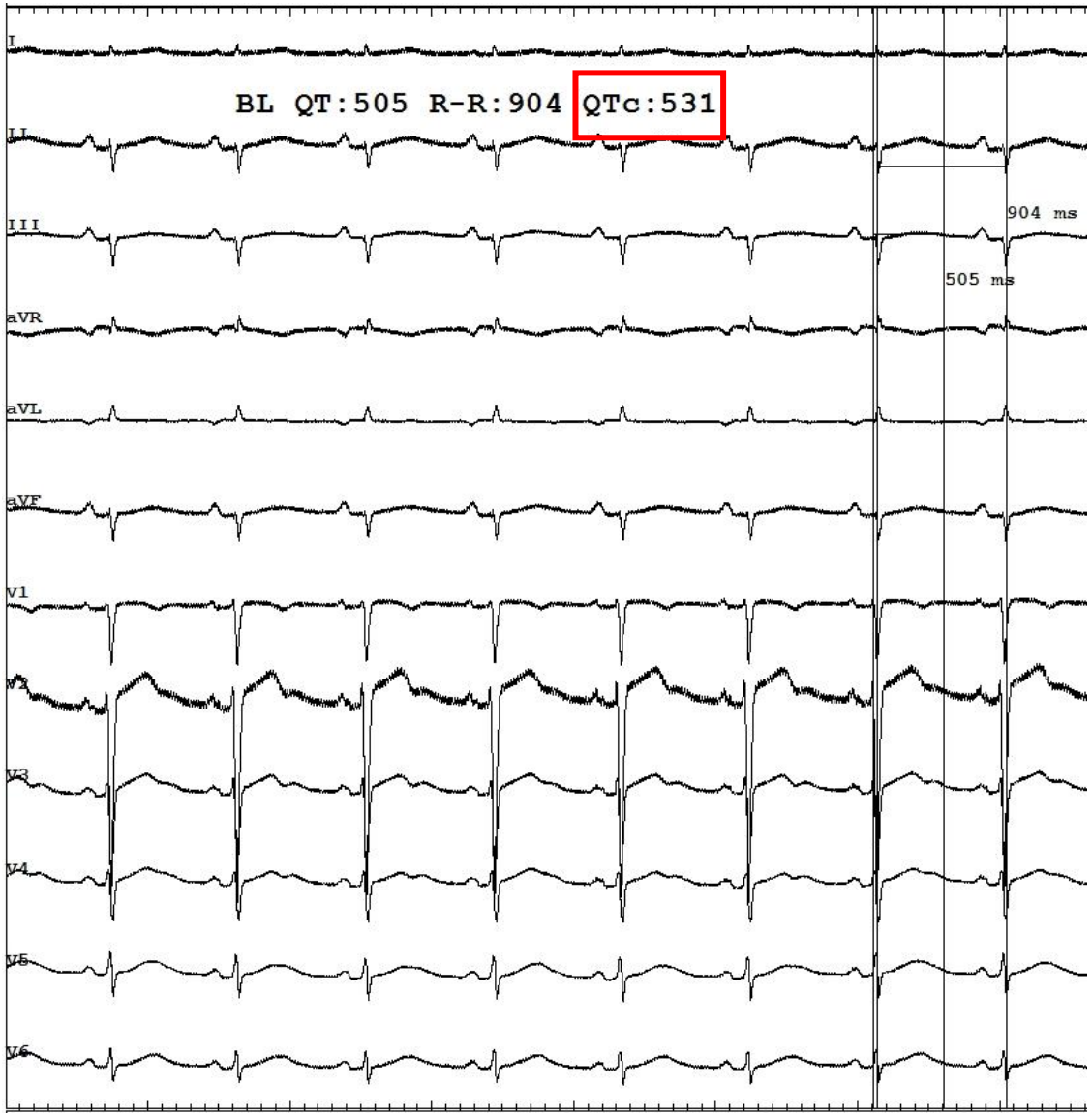
MULTI component
Low voltage signals



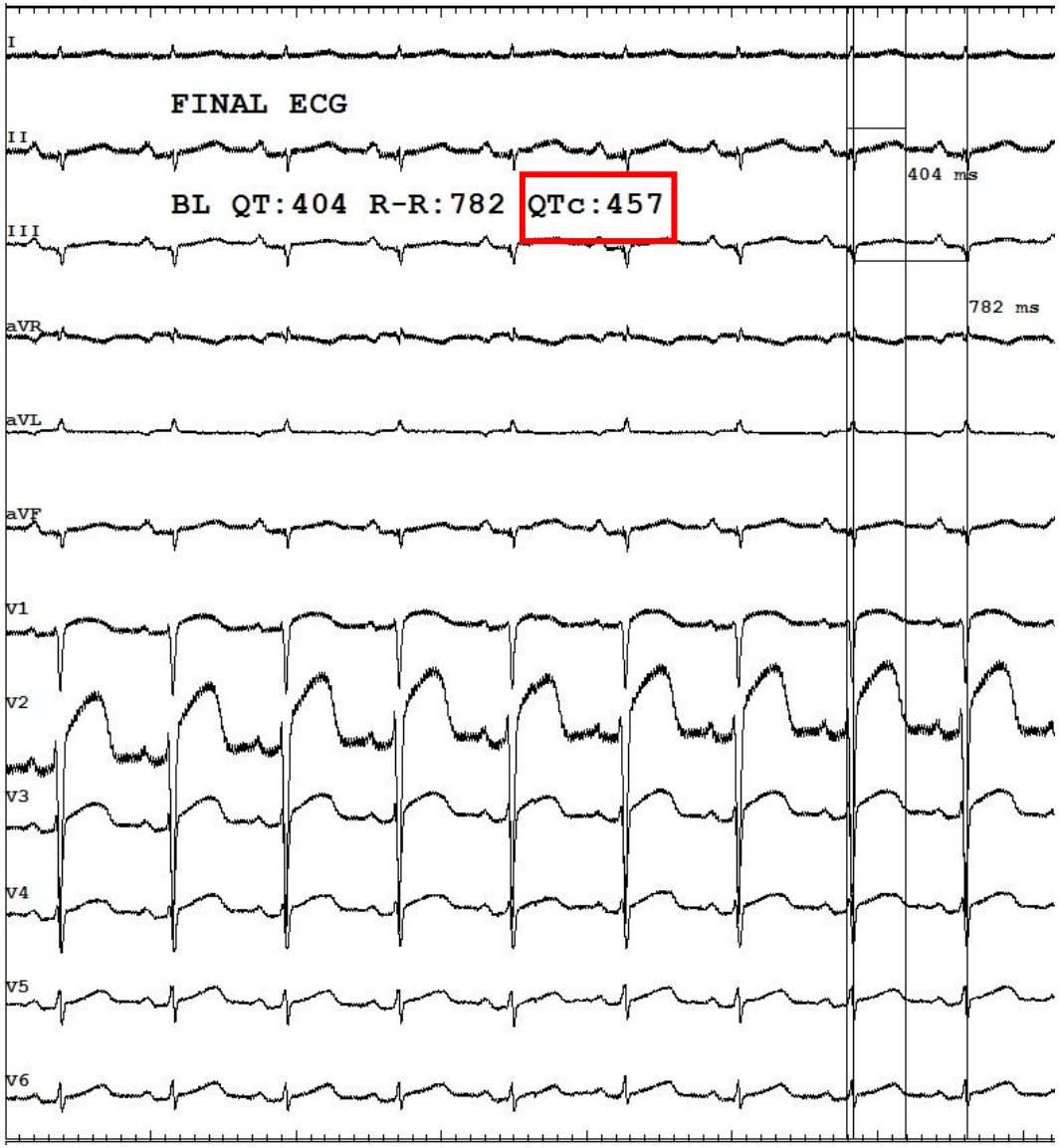
SINGLE component
Low voltage signals

ECG MODIFICATION POST RF: shortening of QT

INITIAL ECG



FINAL ECG



HOME MONITORING

NO MORE VF EPISODES AFTER RF ABLATION

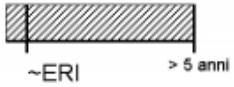
ST. JUDE MEDICAL

29 apr 2021
12:24 (CEST)
Remoto

Sommario FastPath™

1 Avviso

Batteria
Longevità: 5.9-6.7 anni



Data Impianto:	8 gen 2020
Ultima Carica Max.	3.3 sec (Terapia)
Corrente Batteria	12 uA
Capacità residua a ERI	78%

Risultati test 29 apr 2021 — Automatico

	Cattura	Sensing	Impedenza Elettrocatetere
V	**Non eseguito** 1.0V @ 1.0ms (Bi) 20 gen 2021	**2.7mV** (Bi) 2.8mV (Bi) 20 gen 2021	**330Ω** (Bi) 330 Ω (Bi) 19 gen 2021
HV			**45Ω** (VD-SVC e Can) 47 Ω (VD-SVC e Can) 19 gen 2021

Riepilogo Diagnostica Dal 13 apr 2021

VP <1 %

Episodi VT/VF: 0 Dal 13 apr 2021

	VT-1	VT-2	VF
Episodi	0	0	0
ATP erogati		0	0
Shock erogati		0	0

Episodi SVT: 0
Episodi non sostenuti: 0

J-wave Syndrome

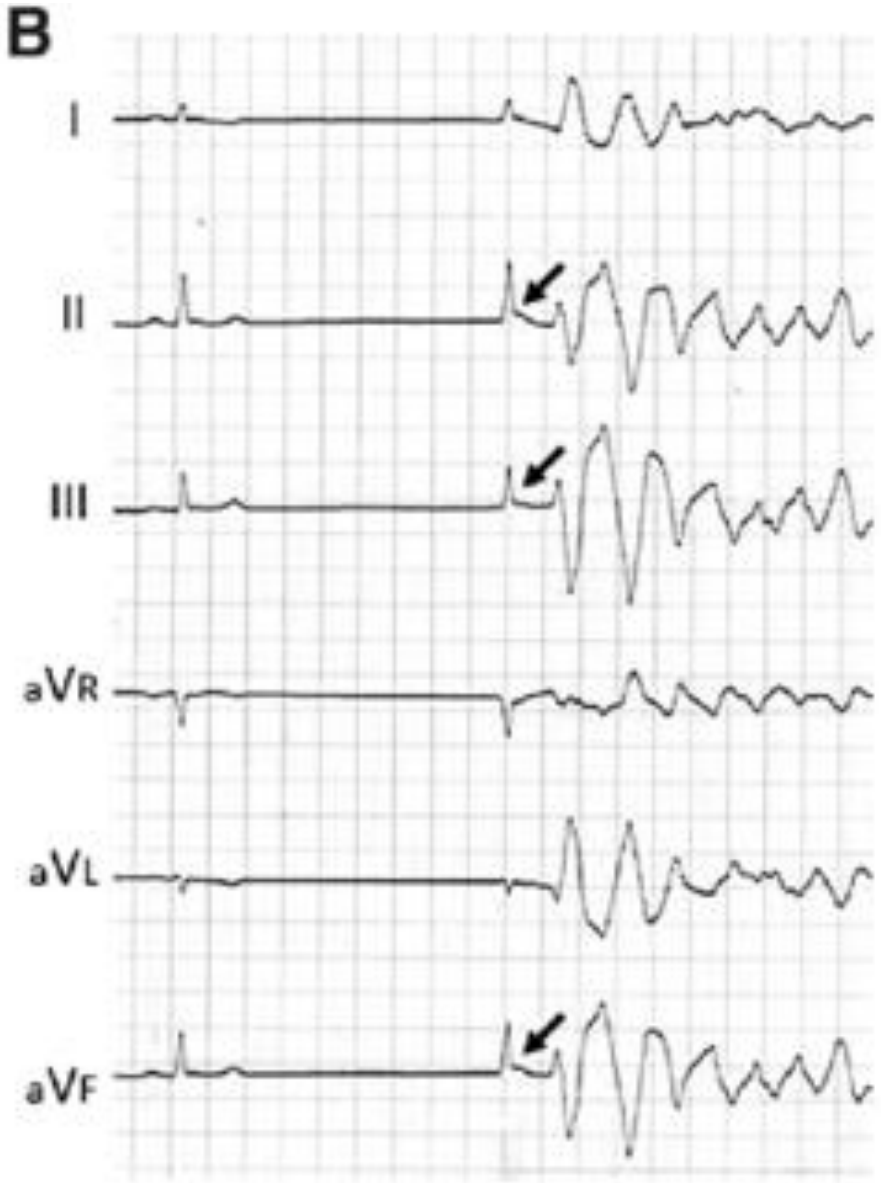
WHAT IS J-WAVE/EARLY REPOLARIZATION SYNDROME?

In humans, the normal J wave often appears as a J-point elevation, with part of the J wave buried inside the QRS. An early repolarization pattern (ERP) in the ECG, consisting of a distinct J-wave or J-point elevation, or a notch or slur of the terminal part of the QRS with and without an ST-segment elevation, has traditionally been viewed as benign . The benign nature of an ERP was challenged in 2000 based on experimental data showing that this ECG manifestation predisposes to the development of polymorphic ventricular tachycardia (VT) and ventricular fibrillation (VF) in coronary-perfused wedge preparations.

(…) risk for development of life-threatening arrhythmic events and sudden cardiac death (SCD) among patients presenting with an ERP, particularly in the inferior and inferolateral leads.

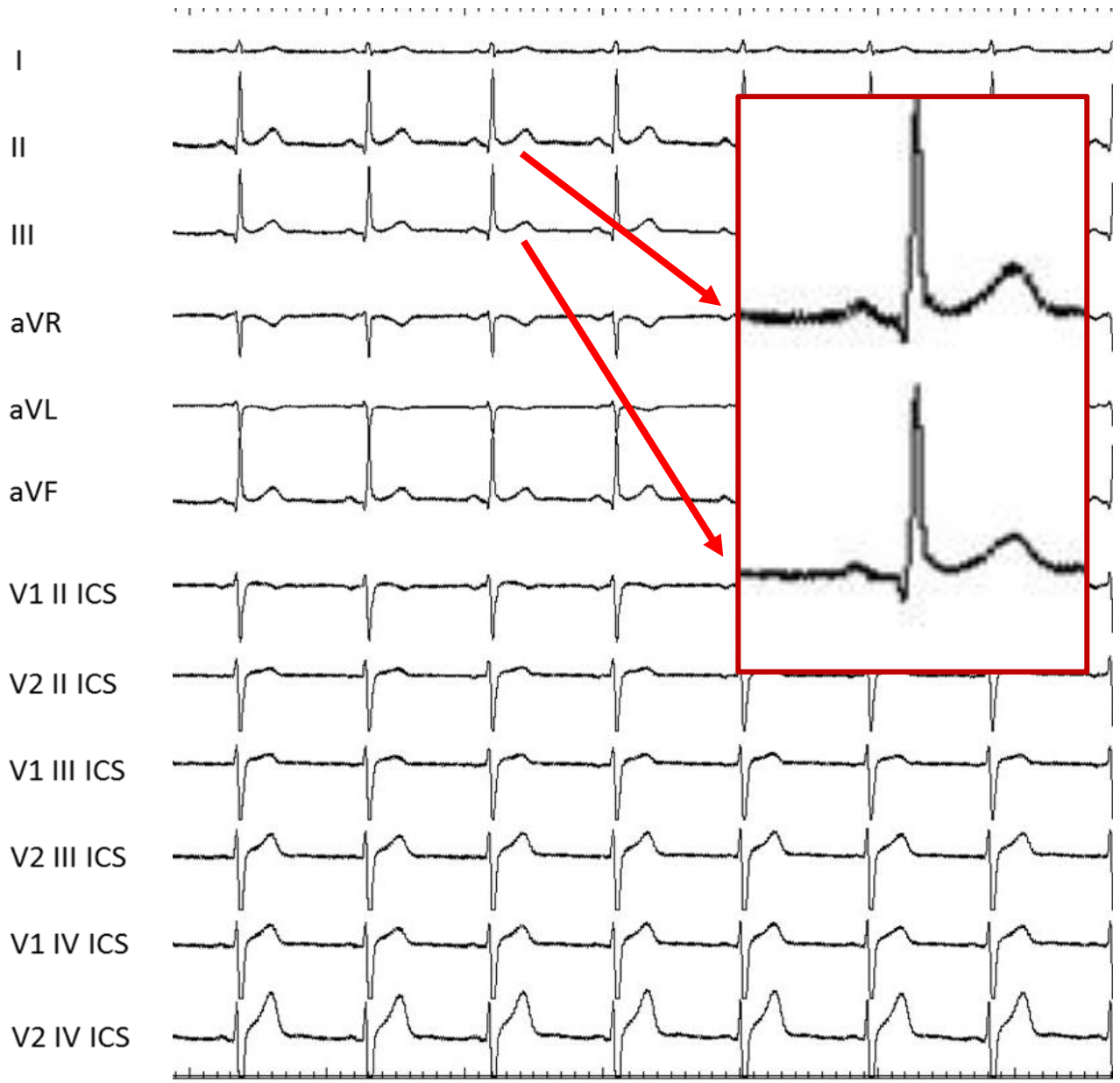
The **prevalence of an ERP** in the inferior and/or lateral leads with a J-point elevation **≥0.1 mV** ranges **between 1% and 24%** and for J-point elevation **≥0.2 mV** ranges **between 0.6% and 6.4%**.

Antzelevitch, Charles et al. “J-Wave syndromes expert consensus conference report: Emerging concepts and gaps in knowledge.” *Journal of arrhythmia* vol. 32,5 (2016): 315-339. doi:10.1016/j.joa.2016.07.002

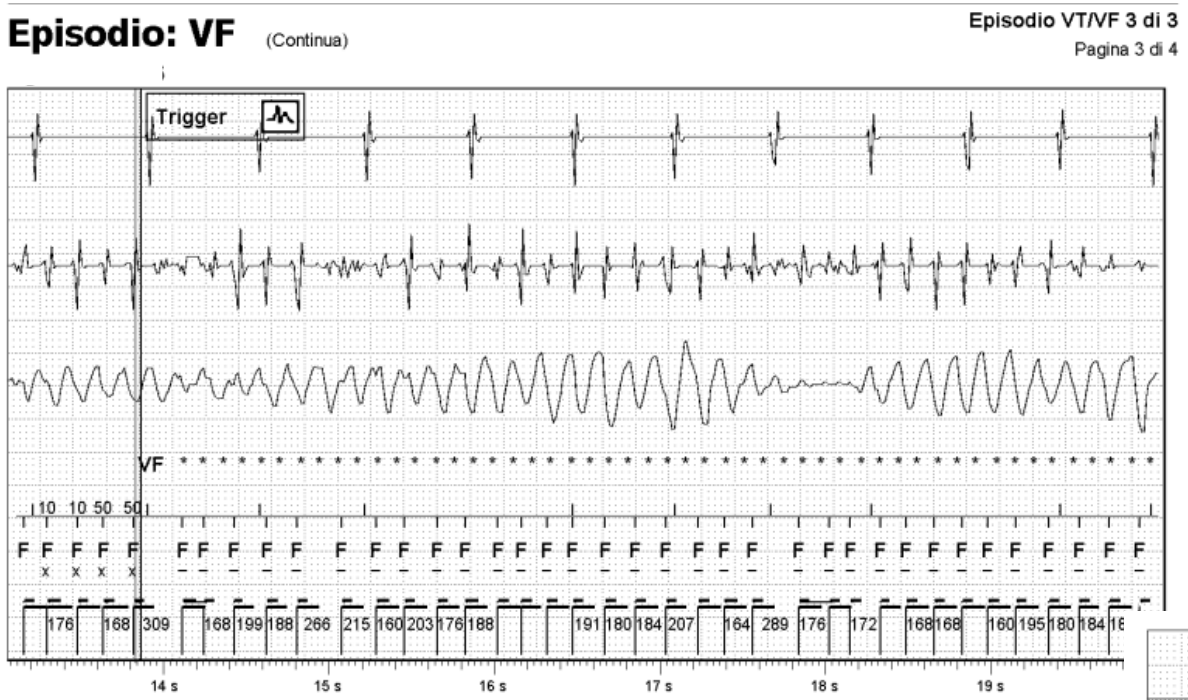


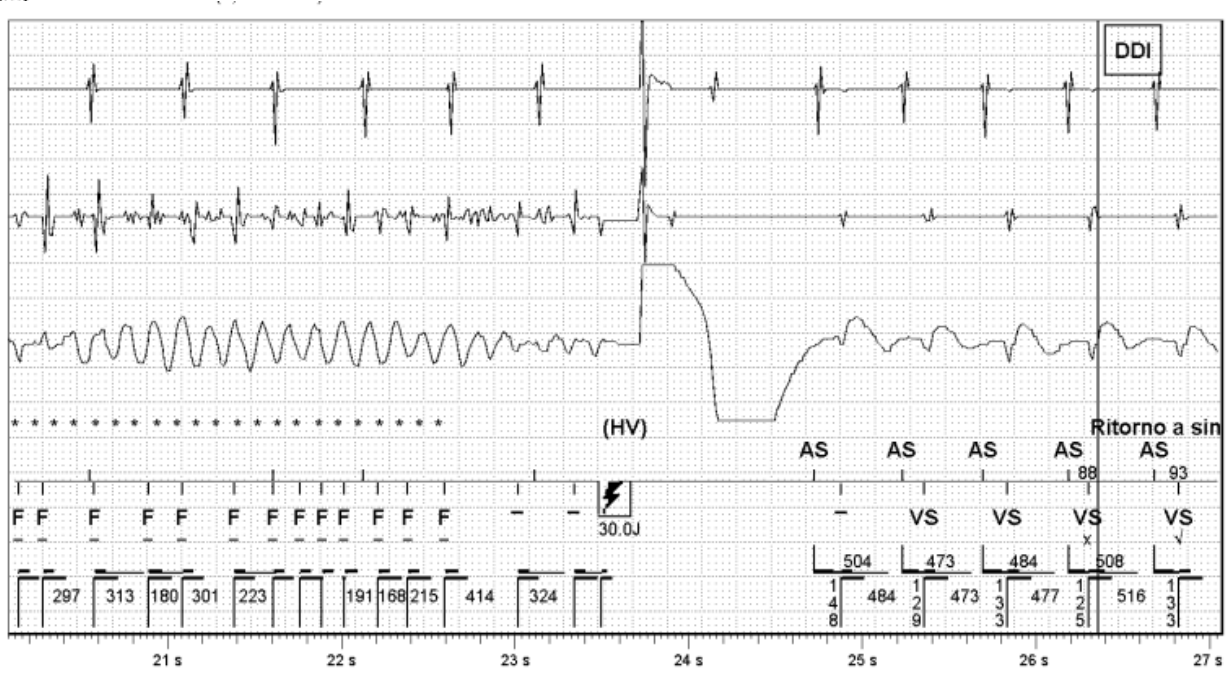
SUMMARY

- Male 35 YO
- Family history of HCM and SCD (brother and grandfather)
- 2020: negative ajmaline test (No Brugada syndrome), J wave diagnosis, ICD implantation
- Jan 2021: ICD intervention on VF episode
- 2021: endocardial and epicardial mapping (admission reason)



2021 VF EPISODE (ICD registration)





J wave – Case 1 – R.L. 2021

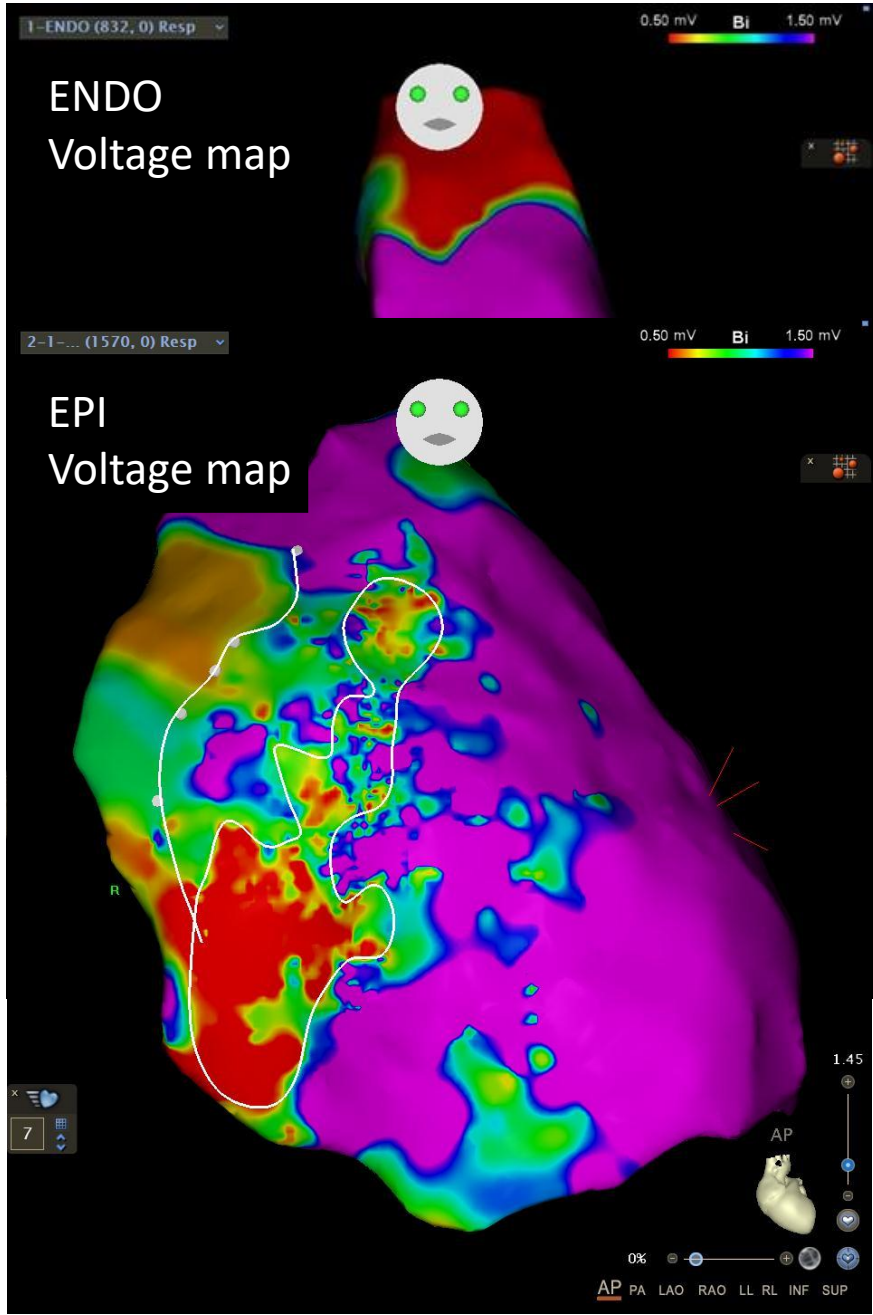
MAPPING DETAILS

Ablation date 19-01-2021

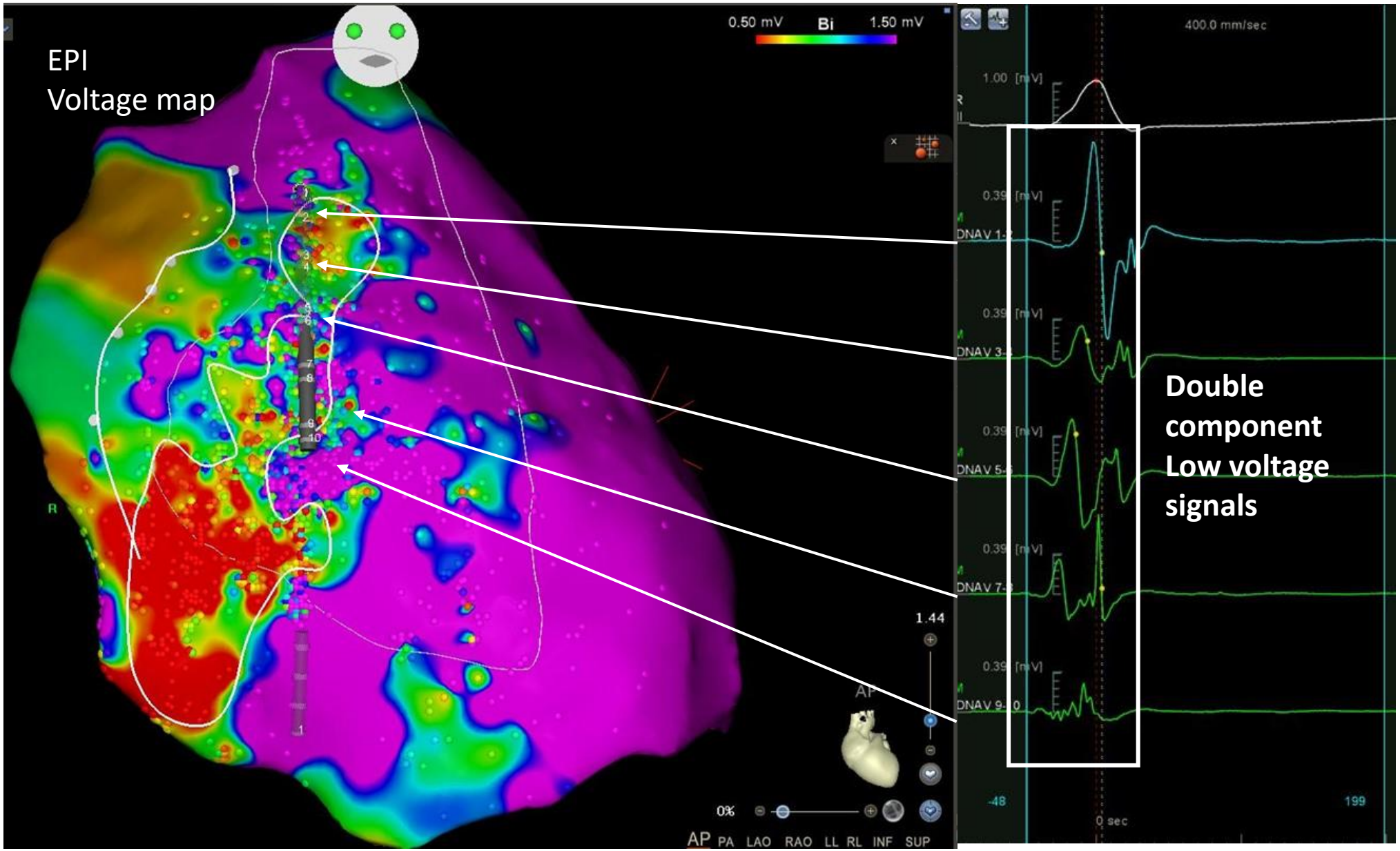
DecaNAV substrate mapping catheter

Endocardial mapping - Healthy RV

Epicardial mapping - Low voltage area on anterolateral tricuspid valve

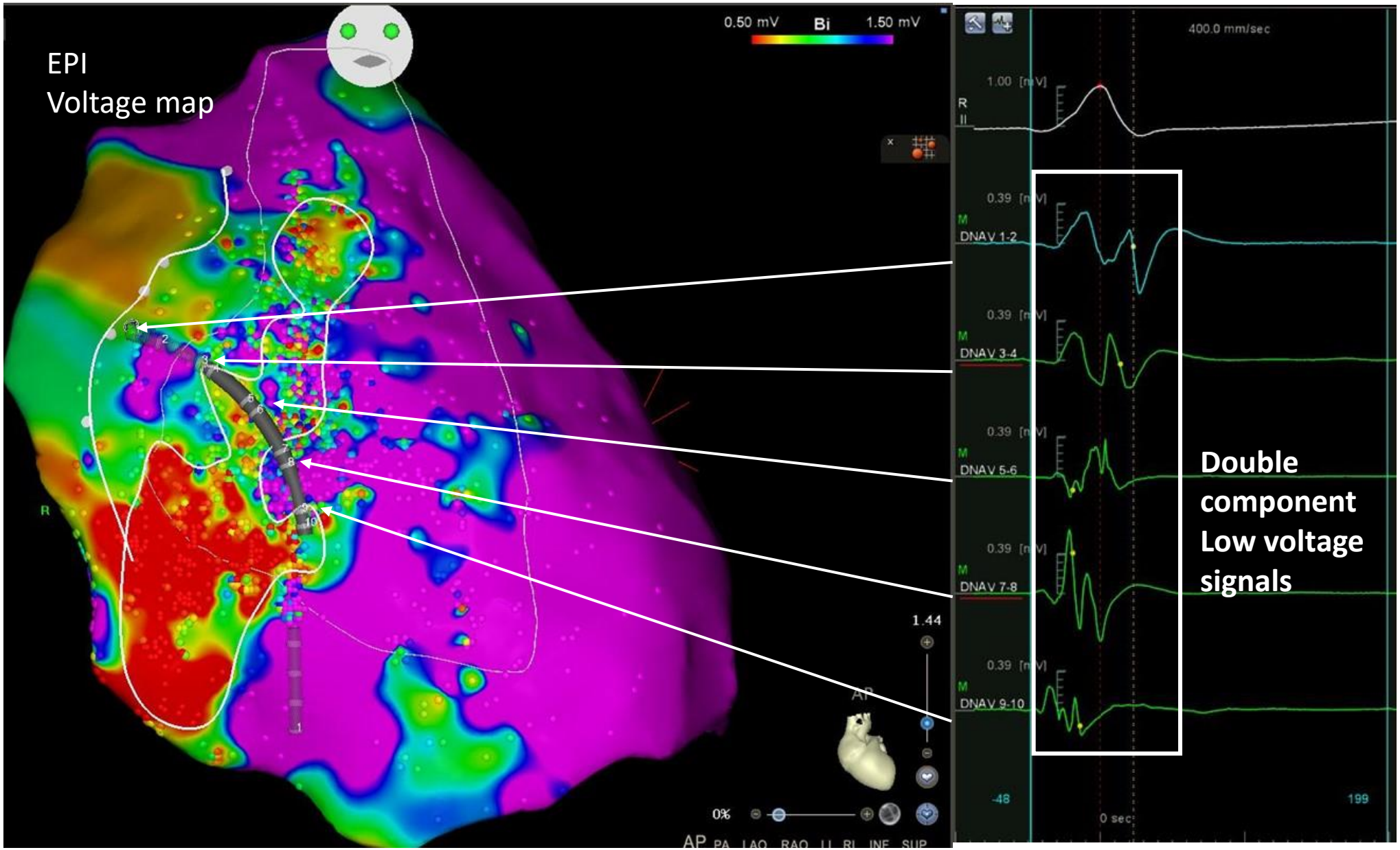


SIGNAL ANALYSIS – Target area

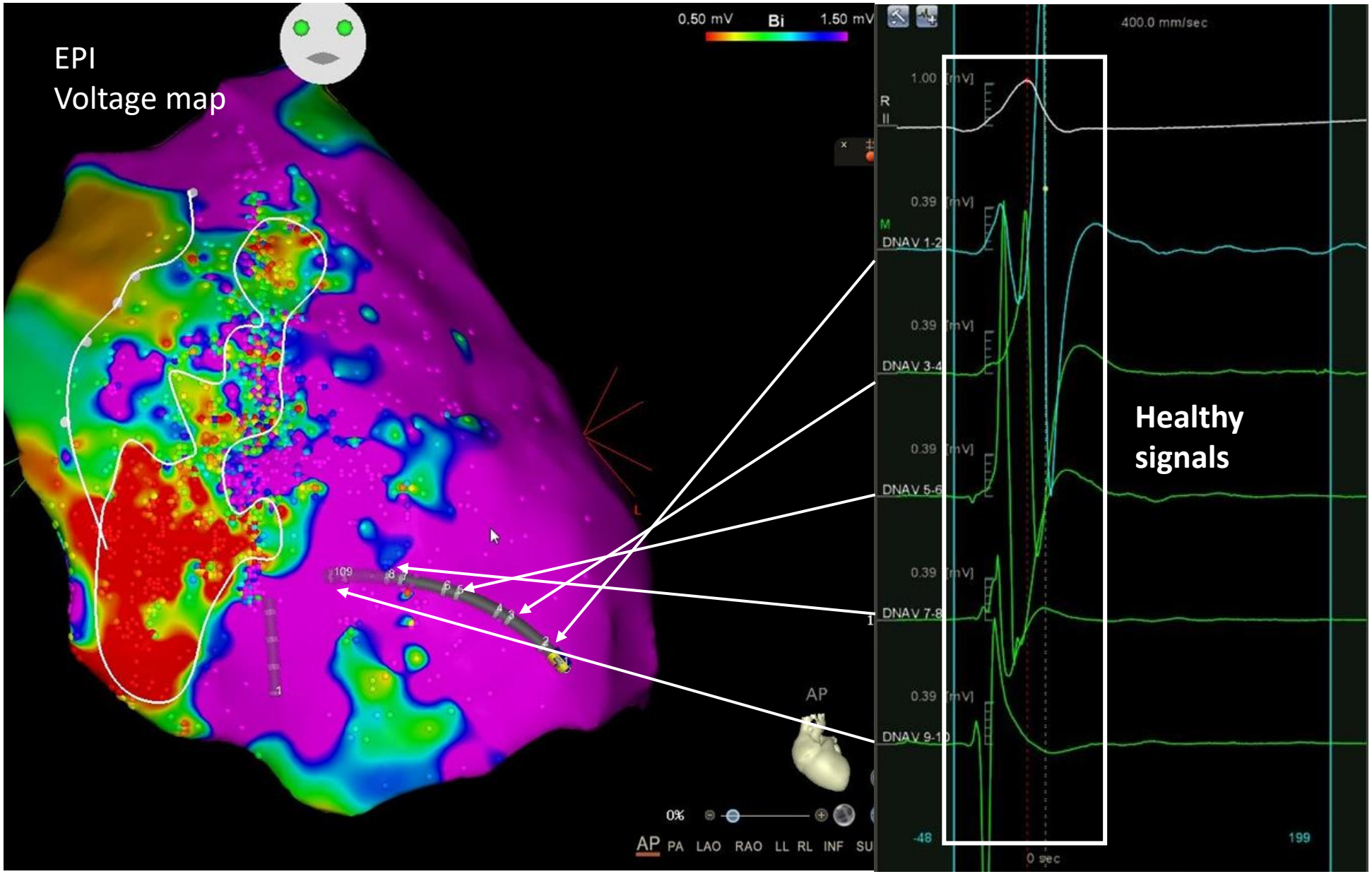


J wave – Case 1 – R.L. 2021

SIGNAL ANALYSIS – Target area



SIGNAL ANALYSIS – Outside the target area



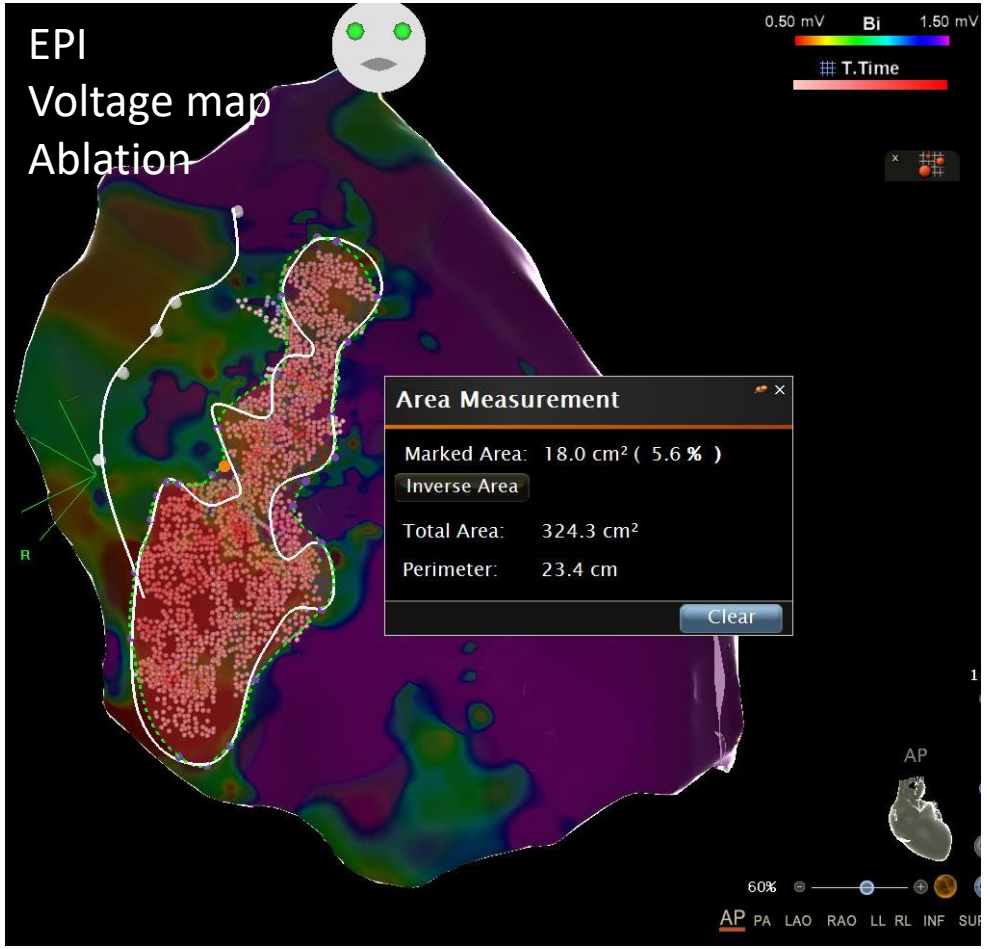
J wave – Case 1 – R.L. 2021

ABLATION STRATEGY

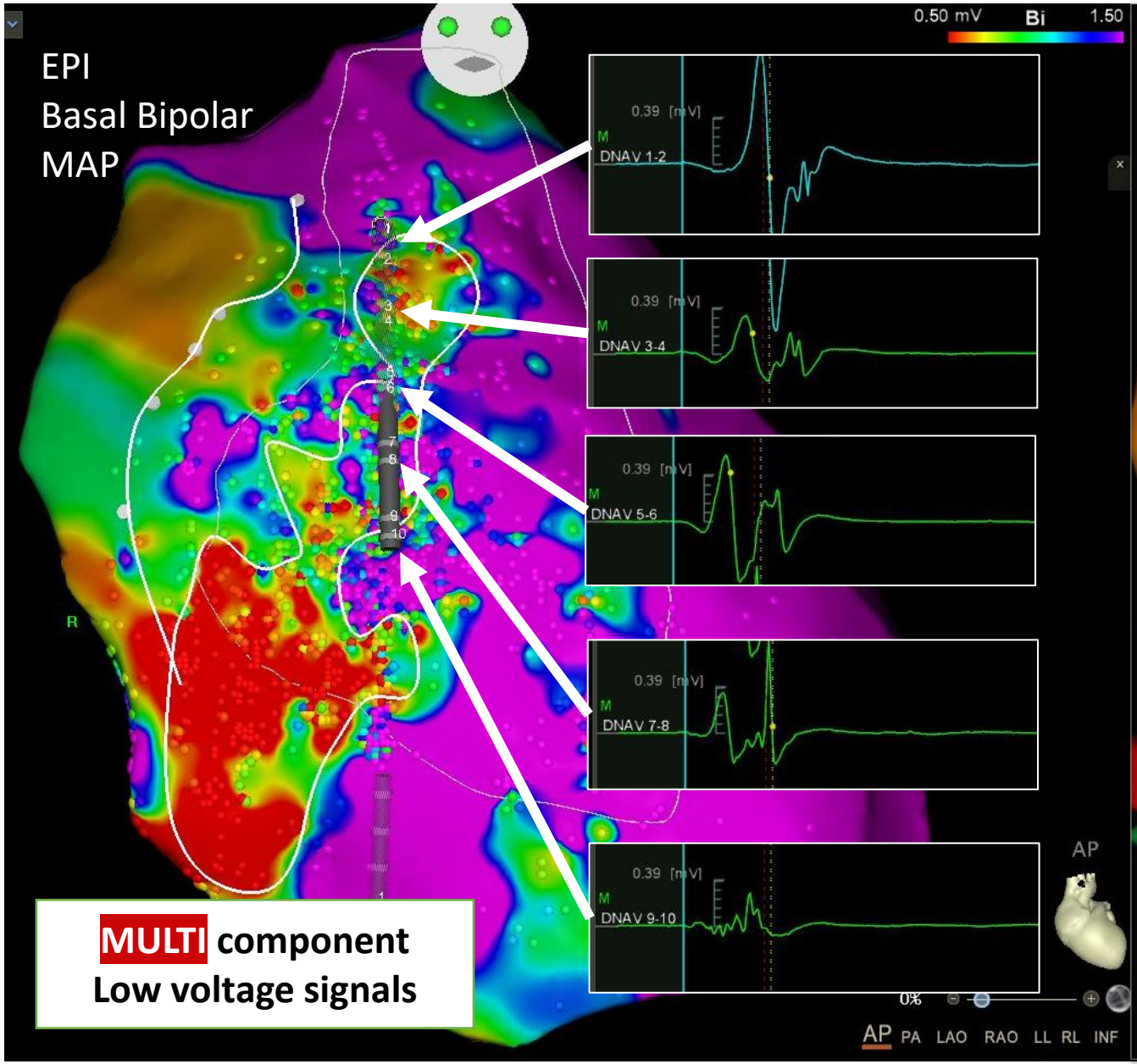
Homogenization of fragmented low voltage potentials area with SmartTouch SF catheter

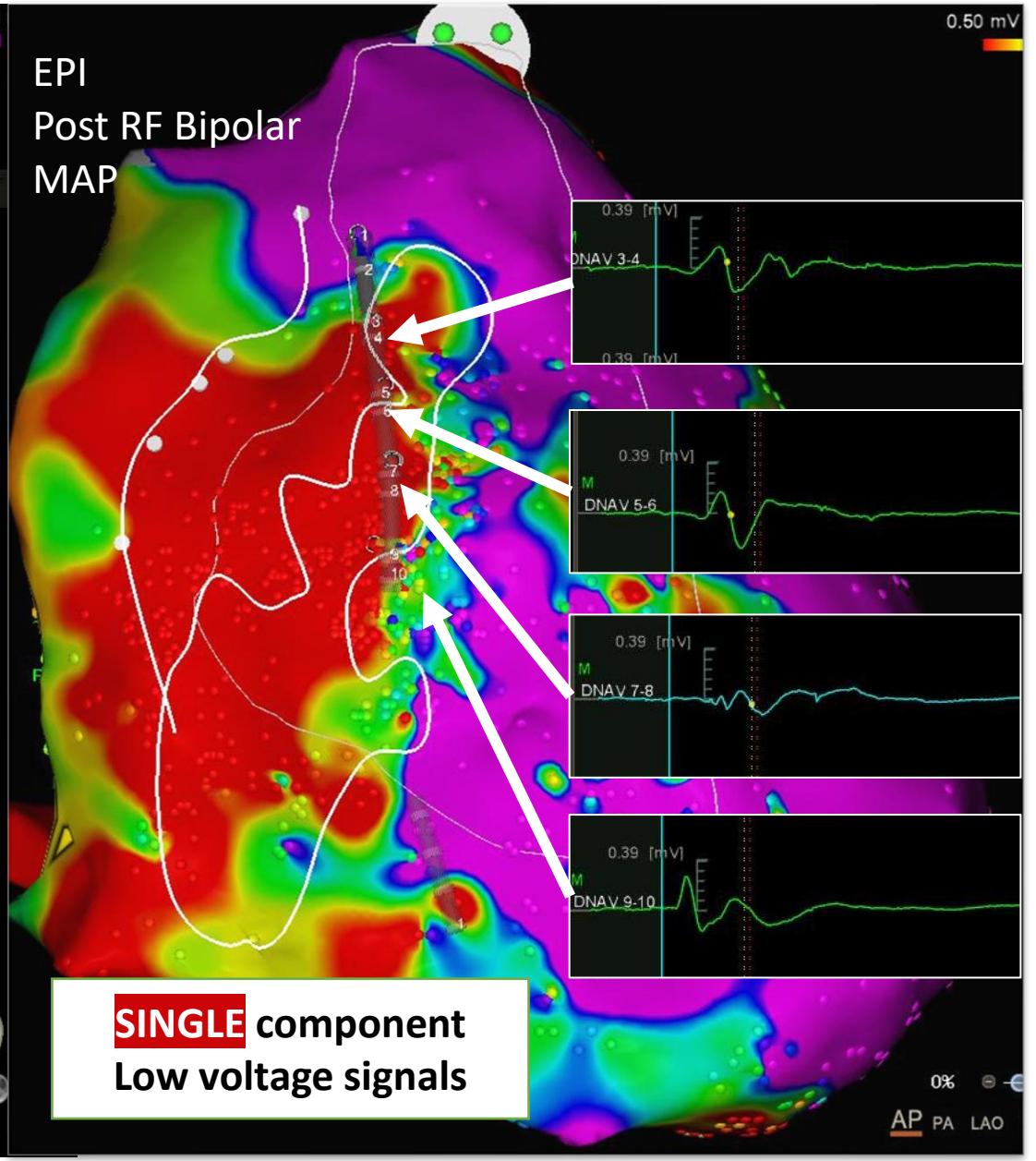
40-45 watts with fast dragging technique, to reach superficial lesion

25 minutes of total ablation over a area of 18.0 cm^2

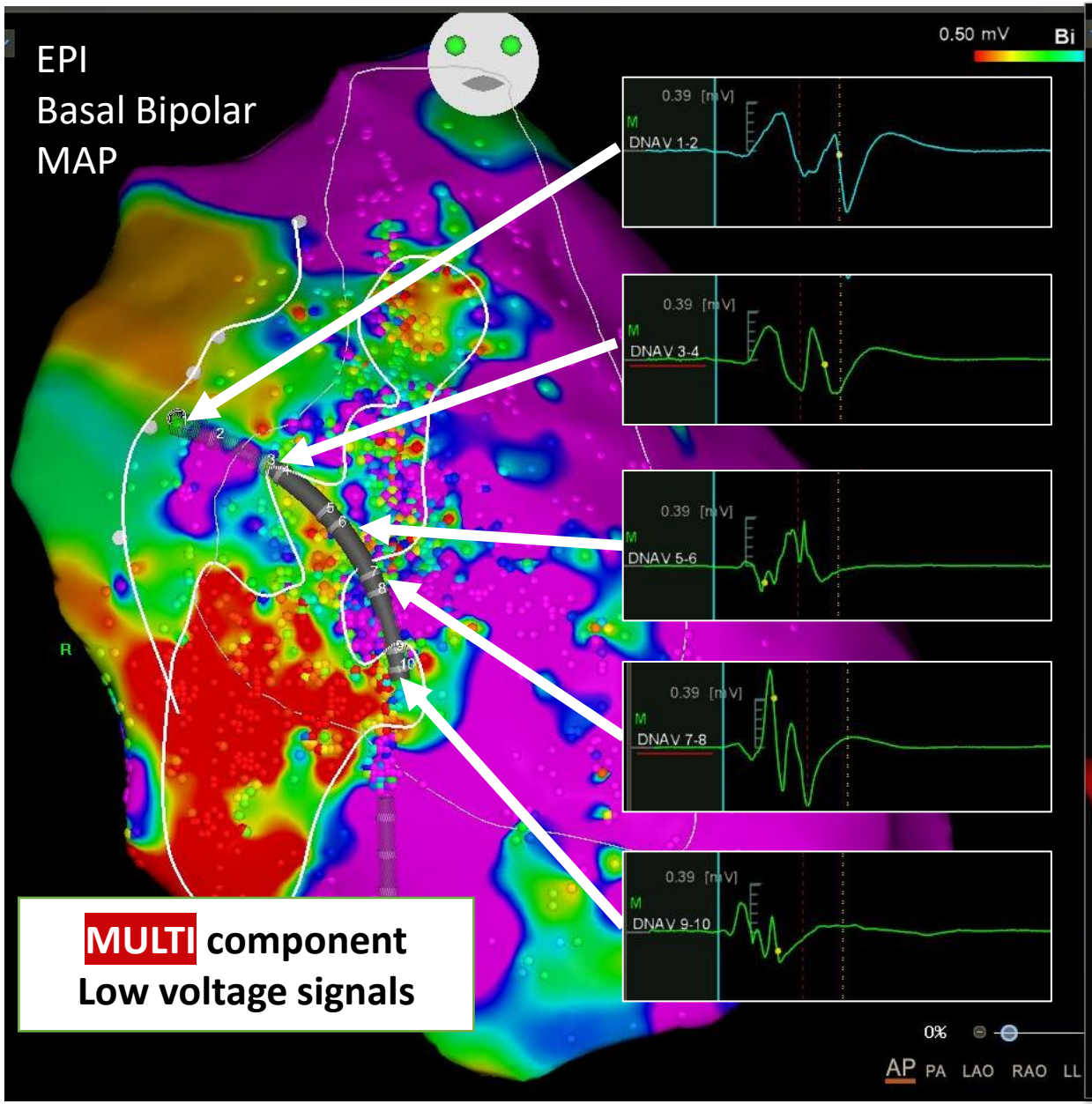


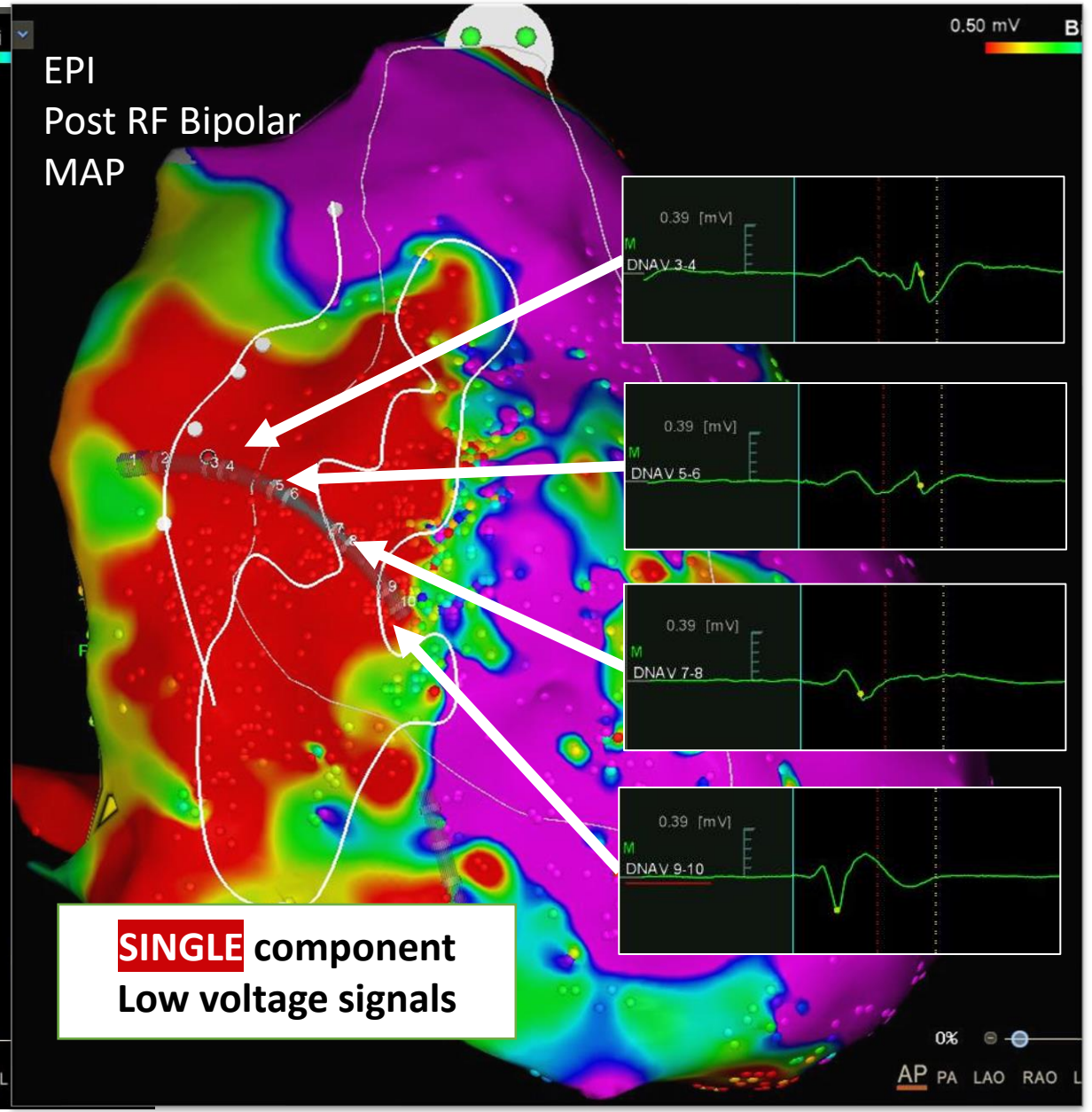
SIGNAL COMPARISON in the target area: PRE AND POST ABLATION





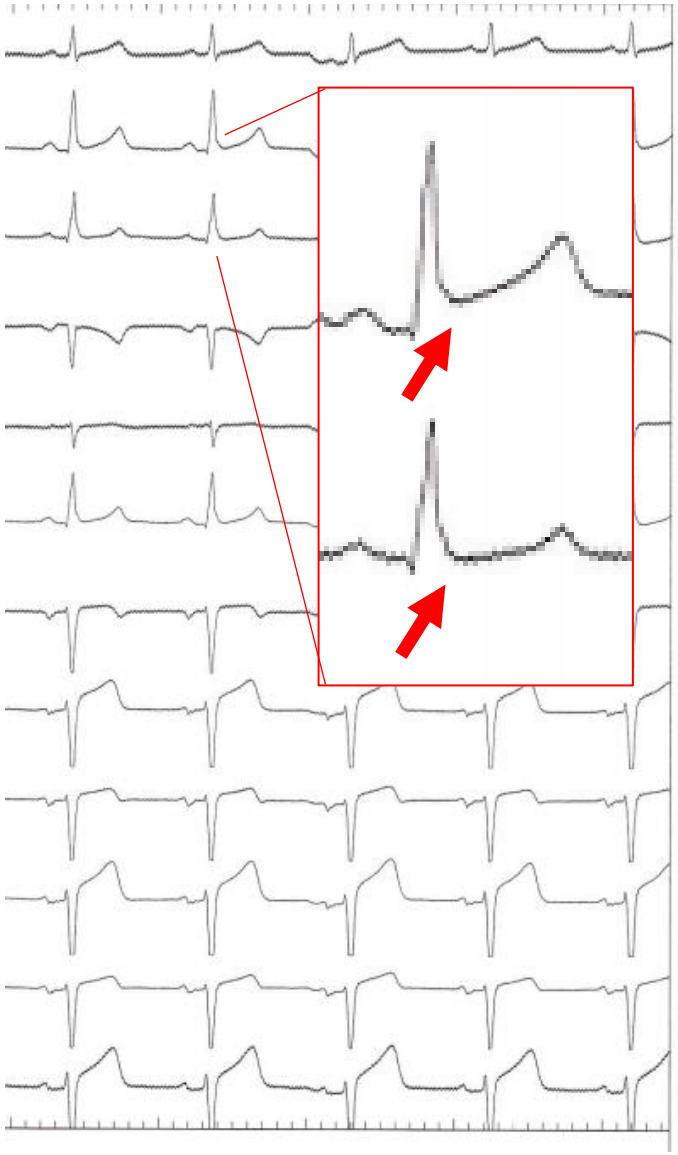
SIGNAL COMPARISON in the target area: PRE AND POST ABLATION



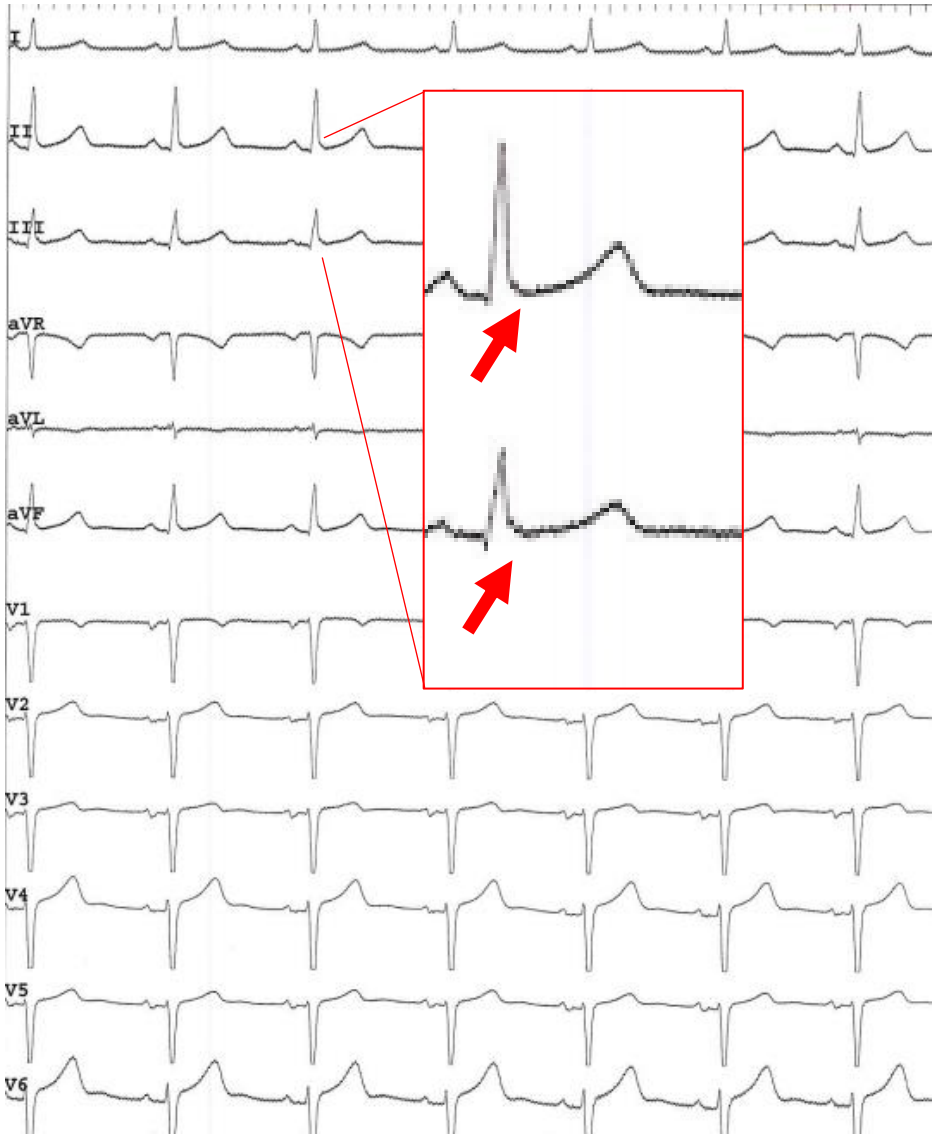


ECG MODIFICATION POST RF: J wave reduction

INITIAL ECG



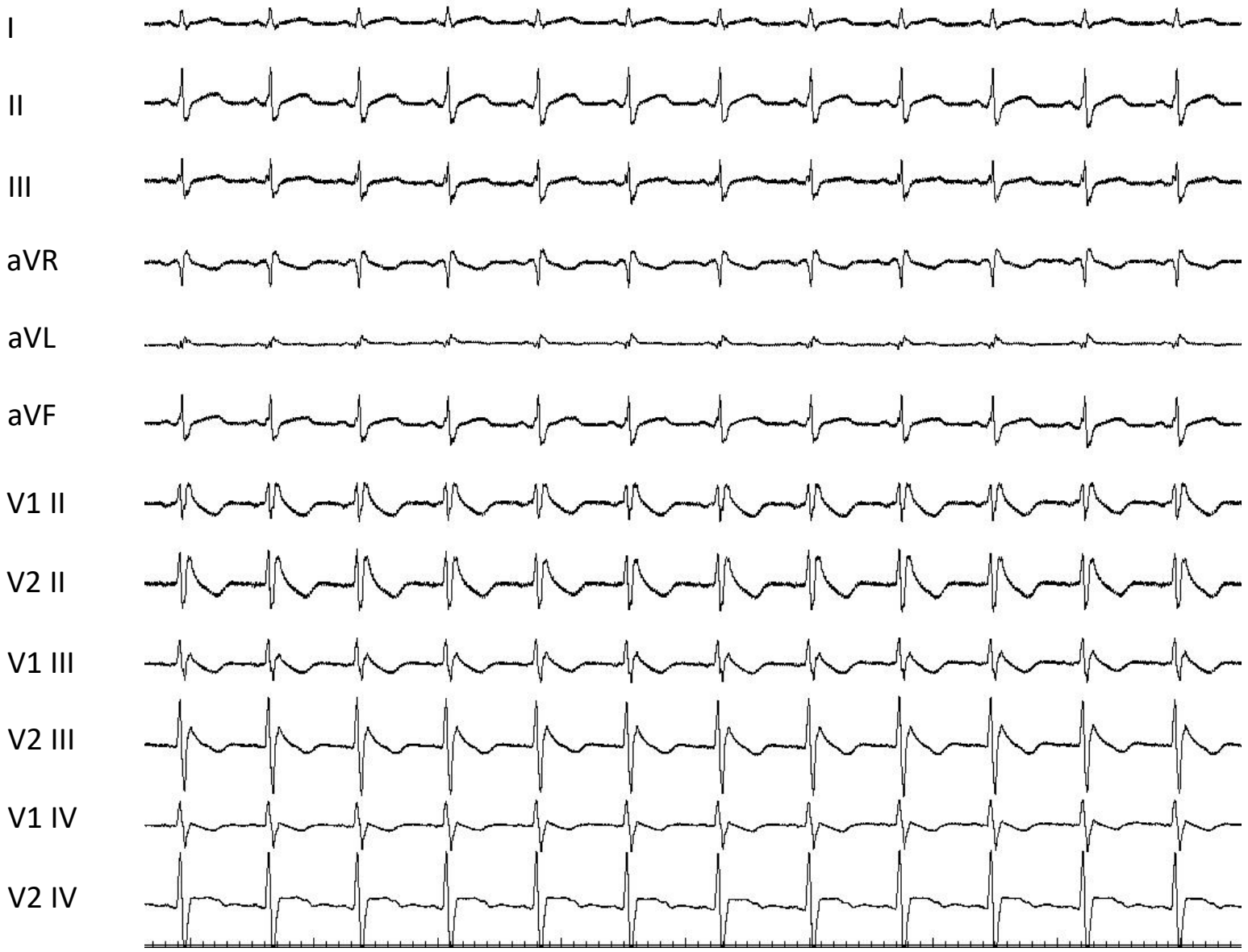
ECG POST RF



J wave – Case 1 – R.L. 2021

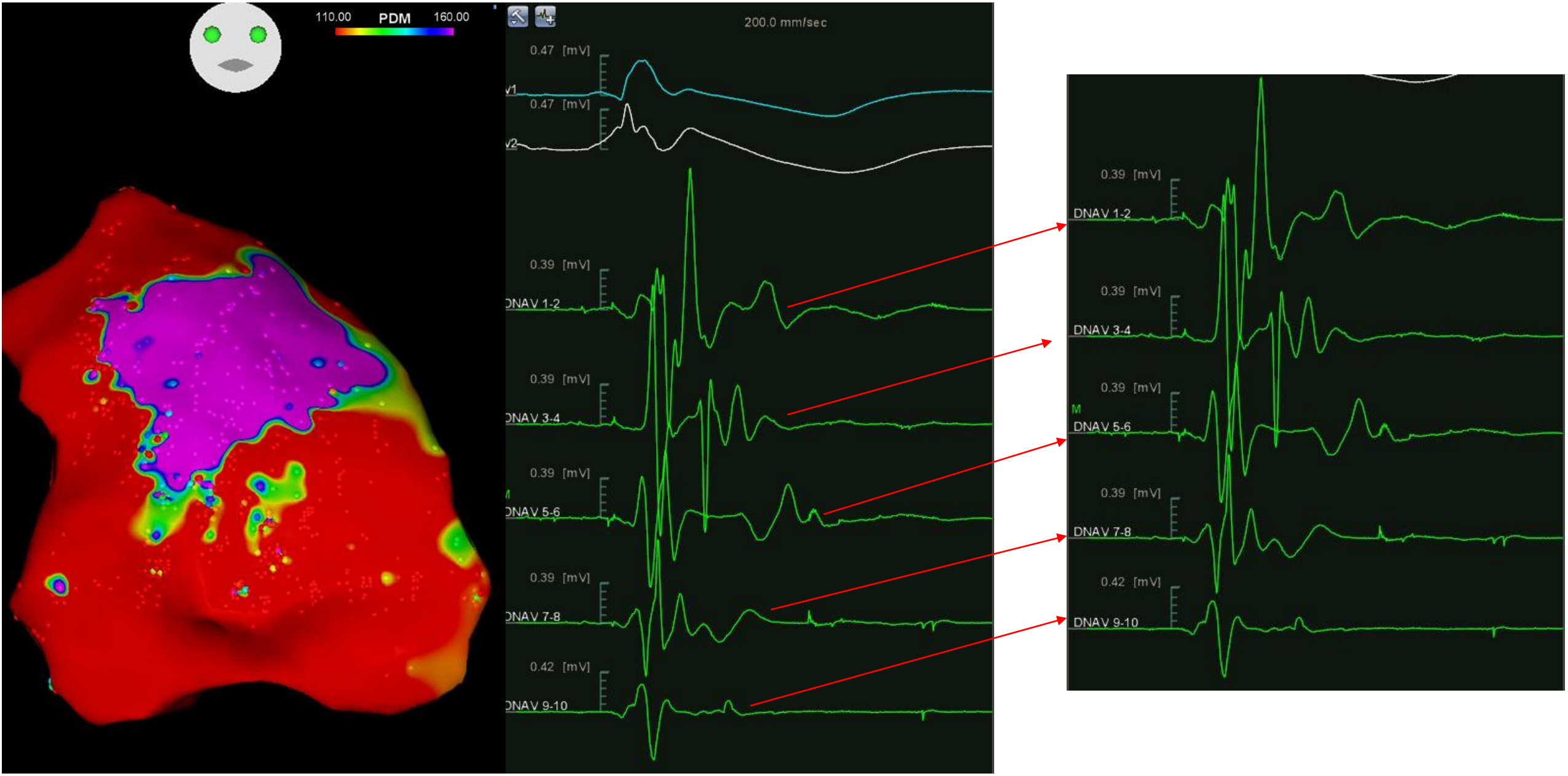
Brugada Syndrome

Type 1 ECG, family history SD, Syncope

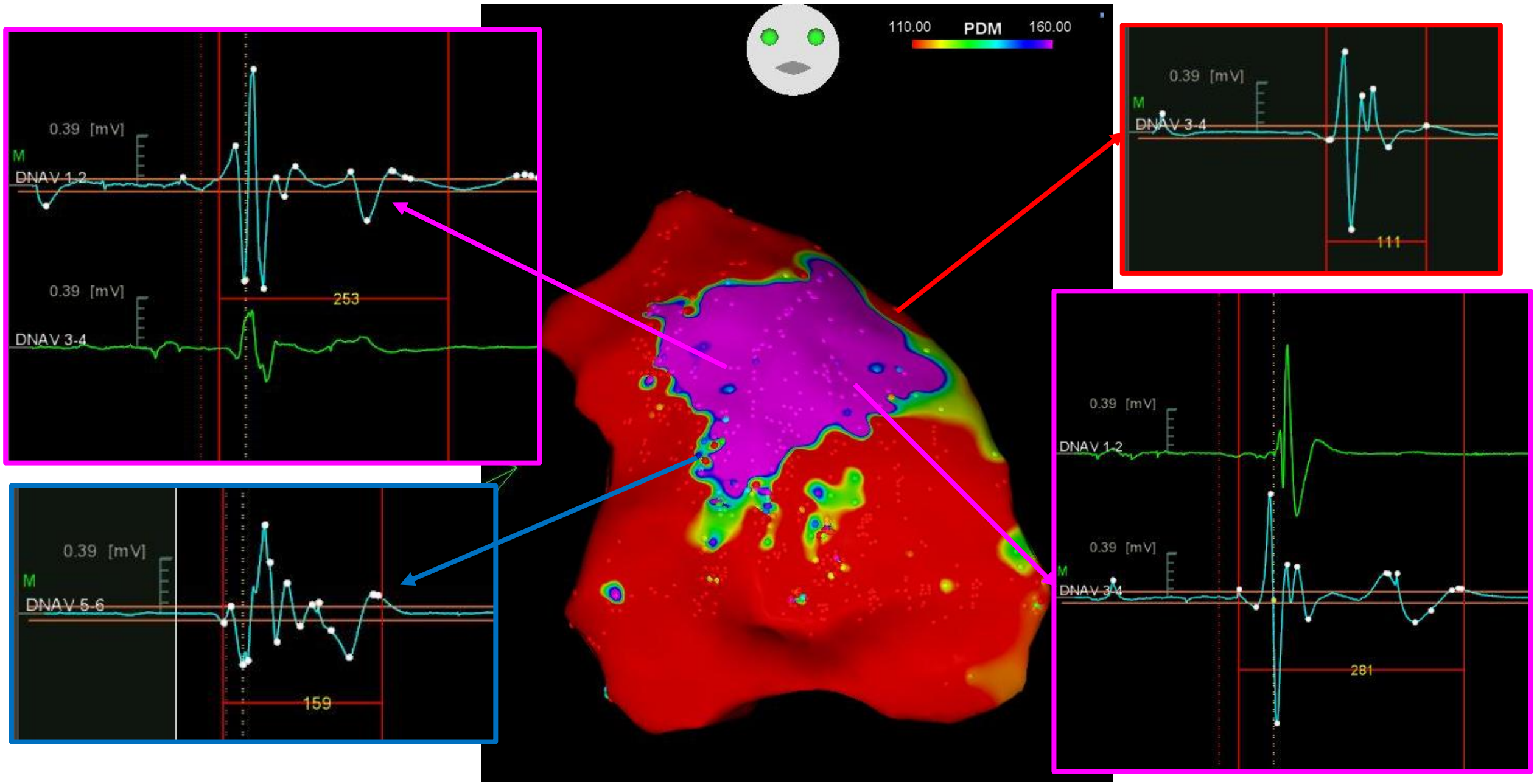


- **Male, 22 years**
- **Family history of BrS**
- **Family history of SD (father and brother)**
- **Syncope**
- **EPS + for VF induction**
- **ICD**
- **SCN5A Mutation**

AUTOMATIC POTENTIALS ANNOTATION

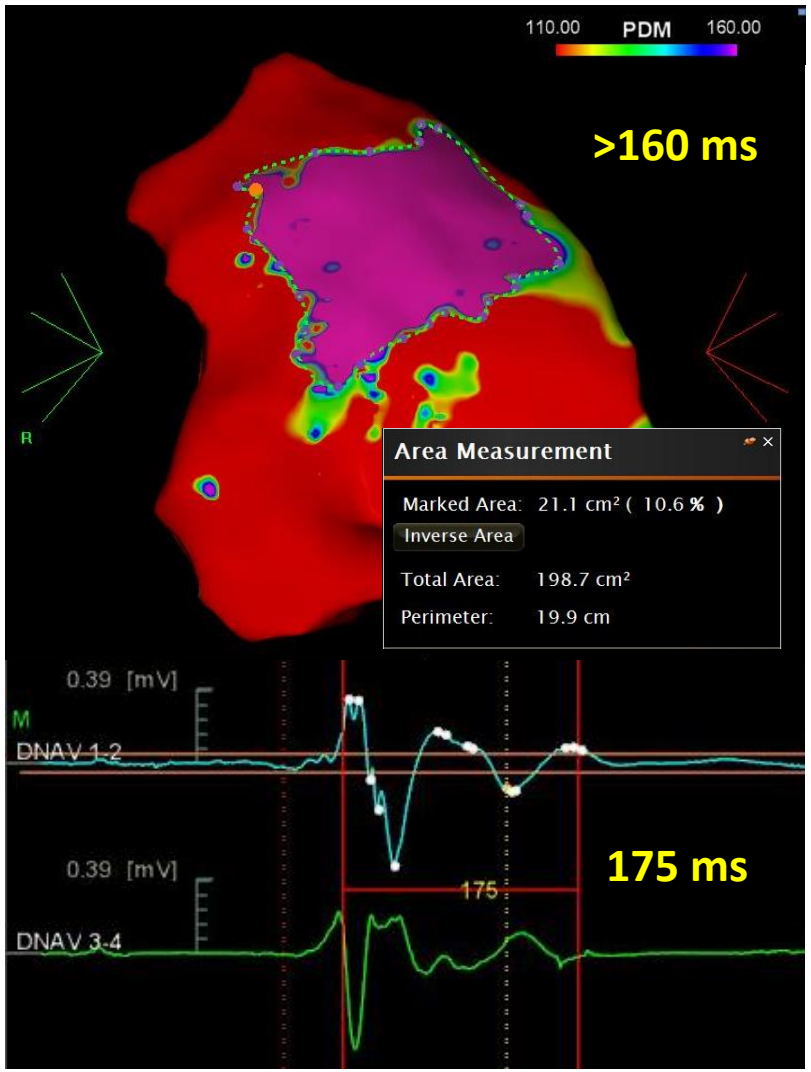


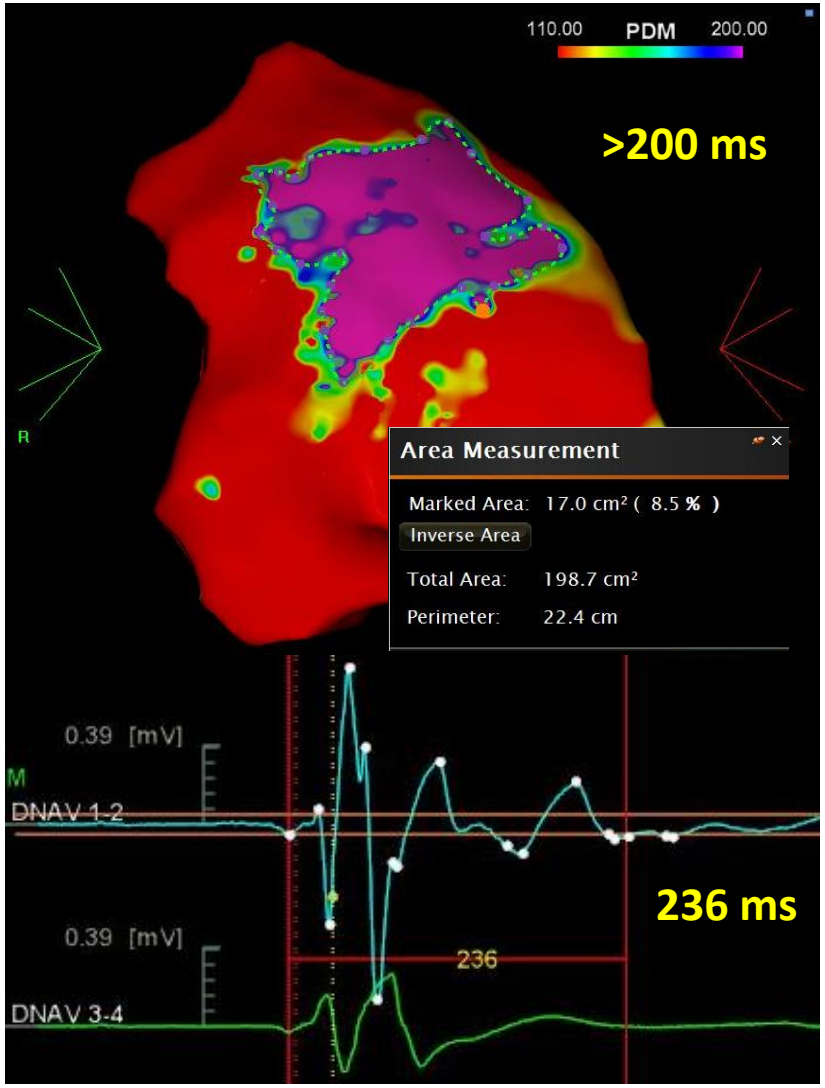
COLOR-CODED POTENTIAL DURATION MAP (PDM)

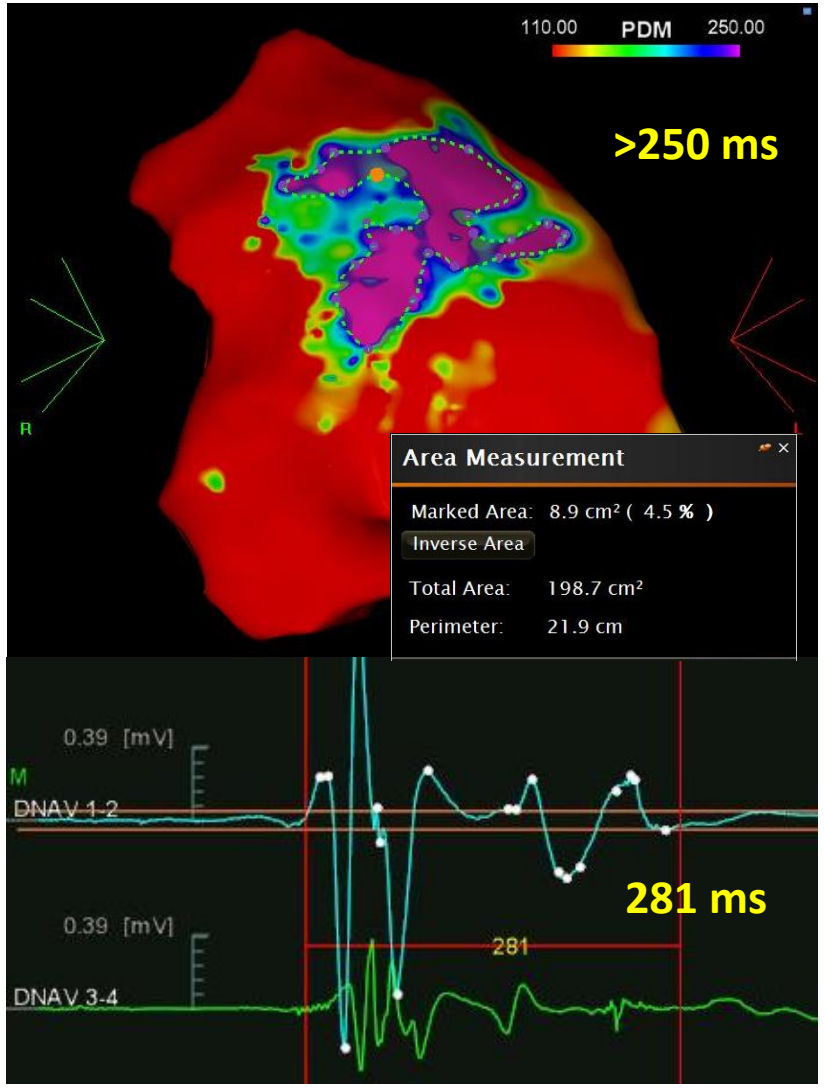


POTENTIAL DURATION MAP (PDM)

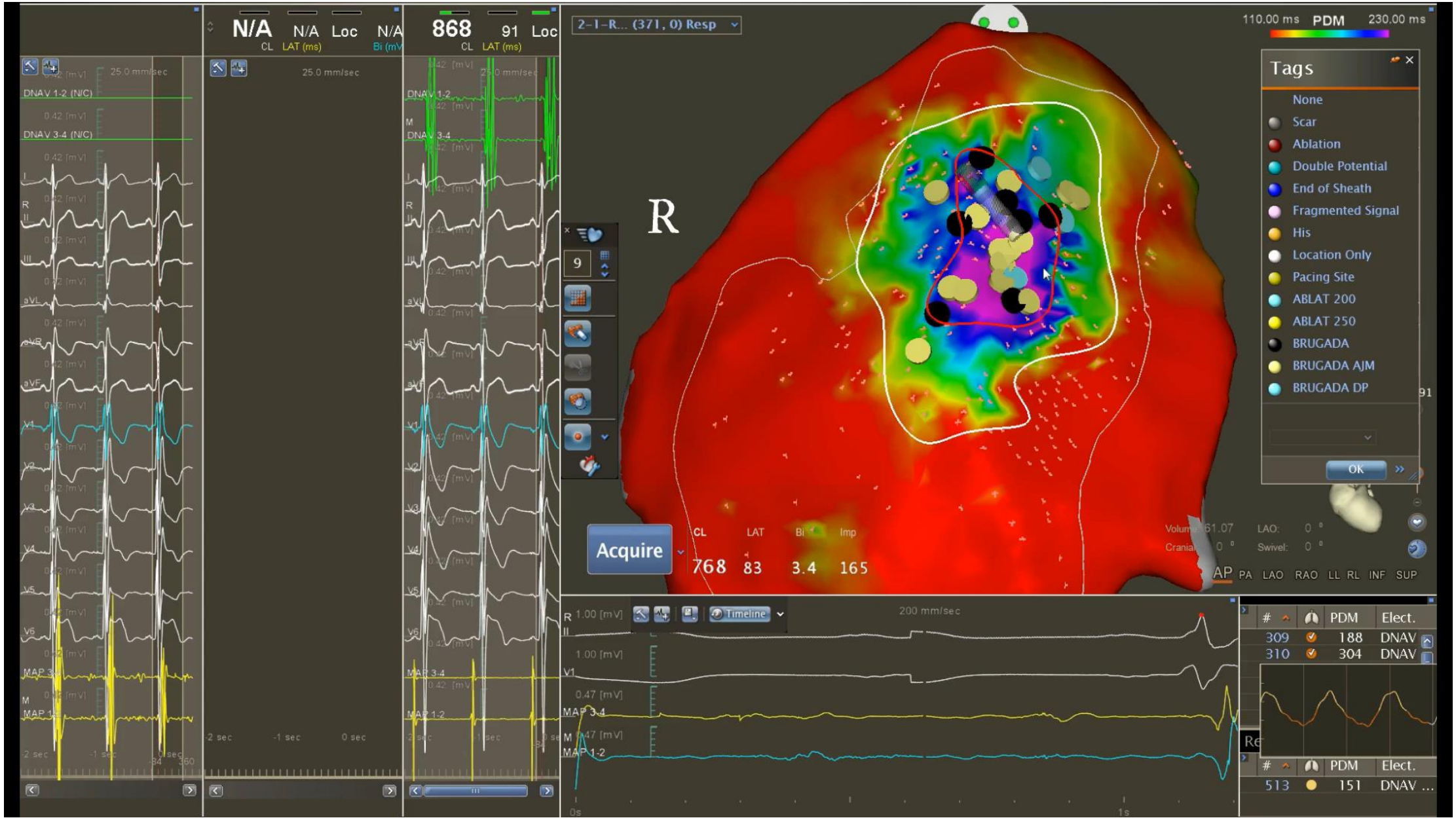
Electrograms with longer duration, low voltage and late or consistent and discrete component are shown in purple color.





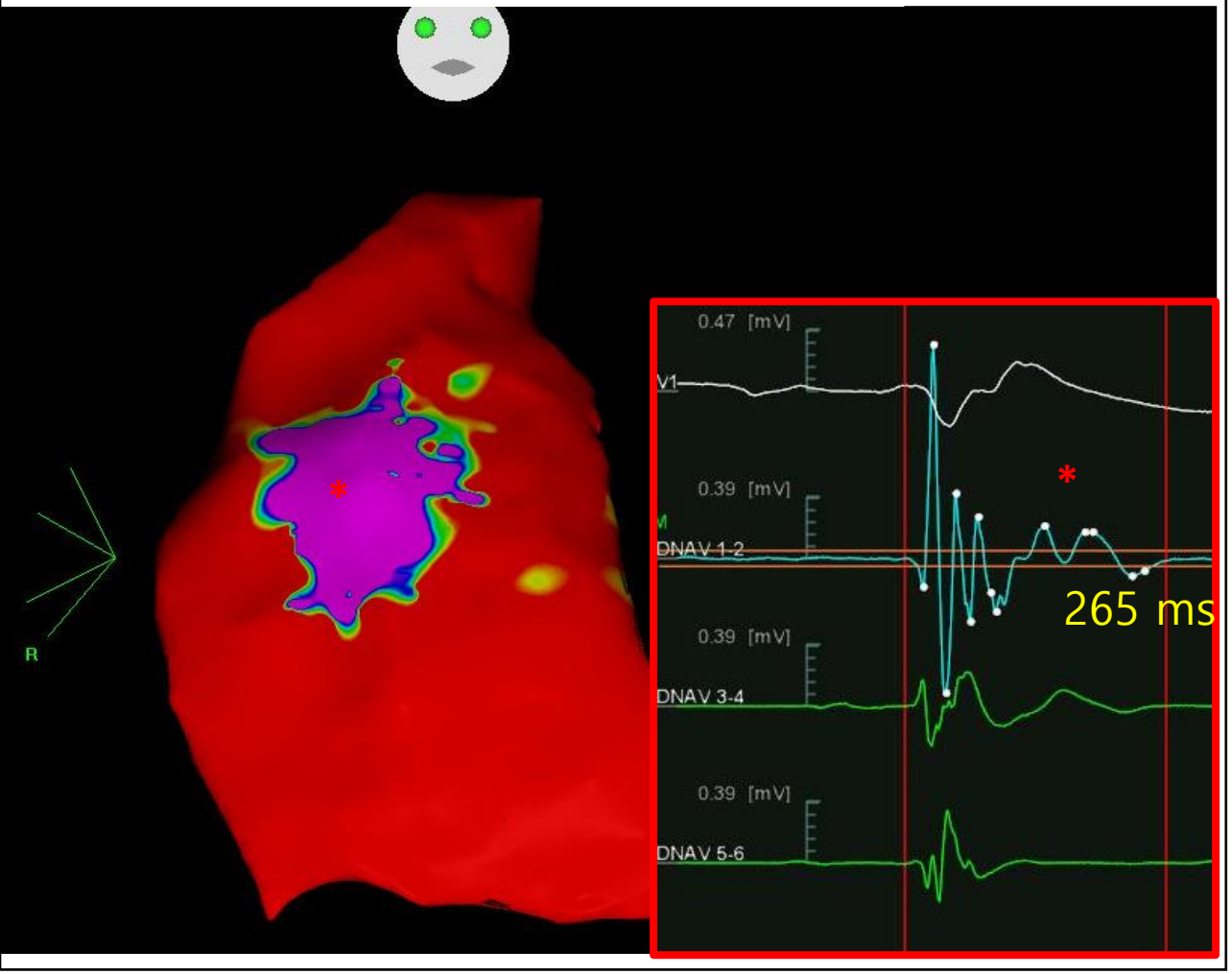


Example of ECG pattern during epicardial RF ablation

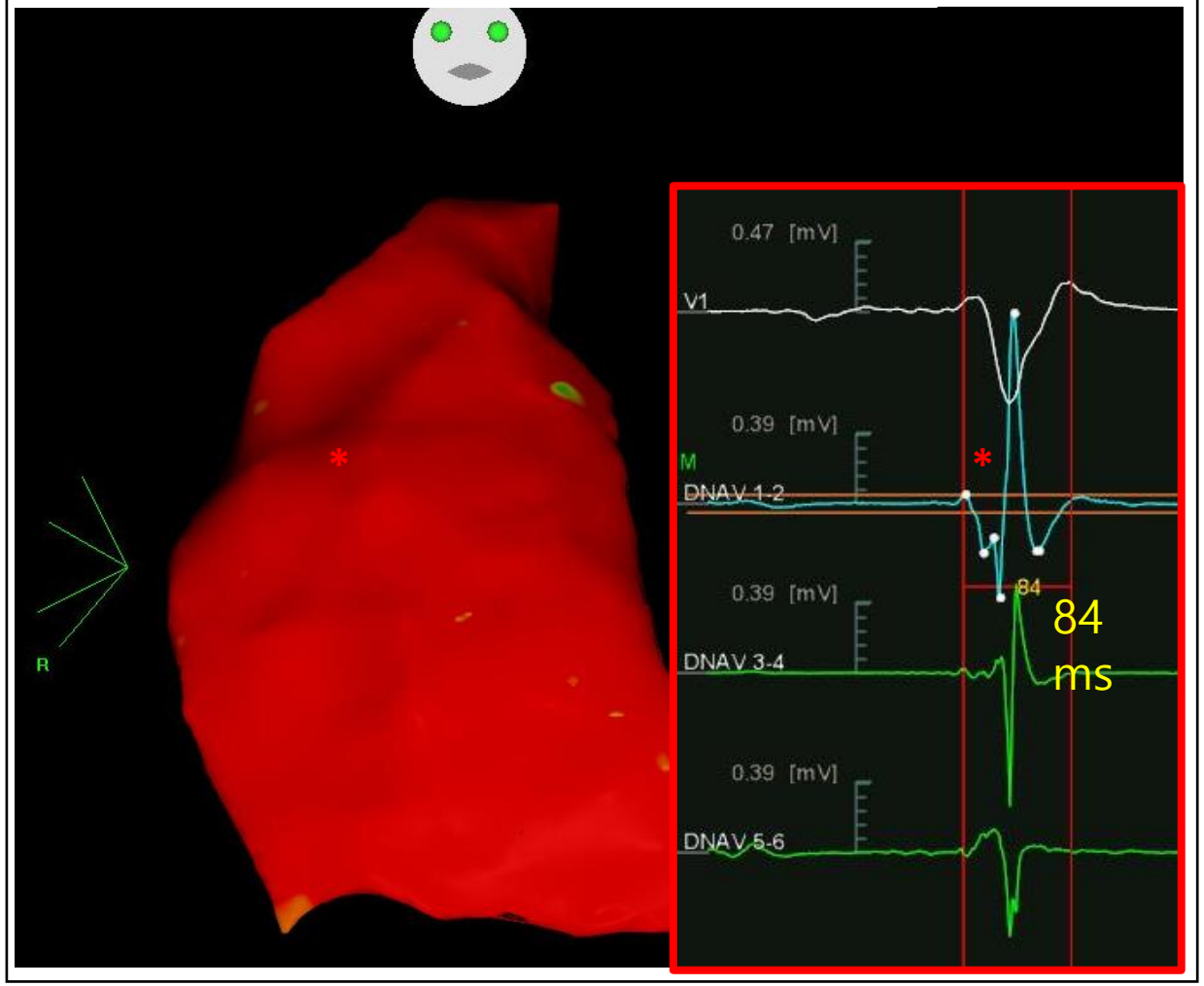


PHENOTYPE ABLATION (Late Potentials Abolition)

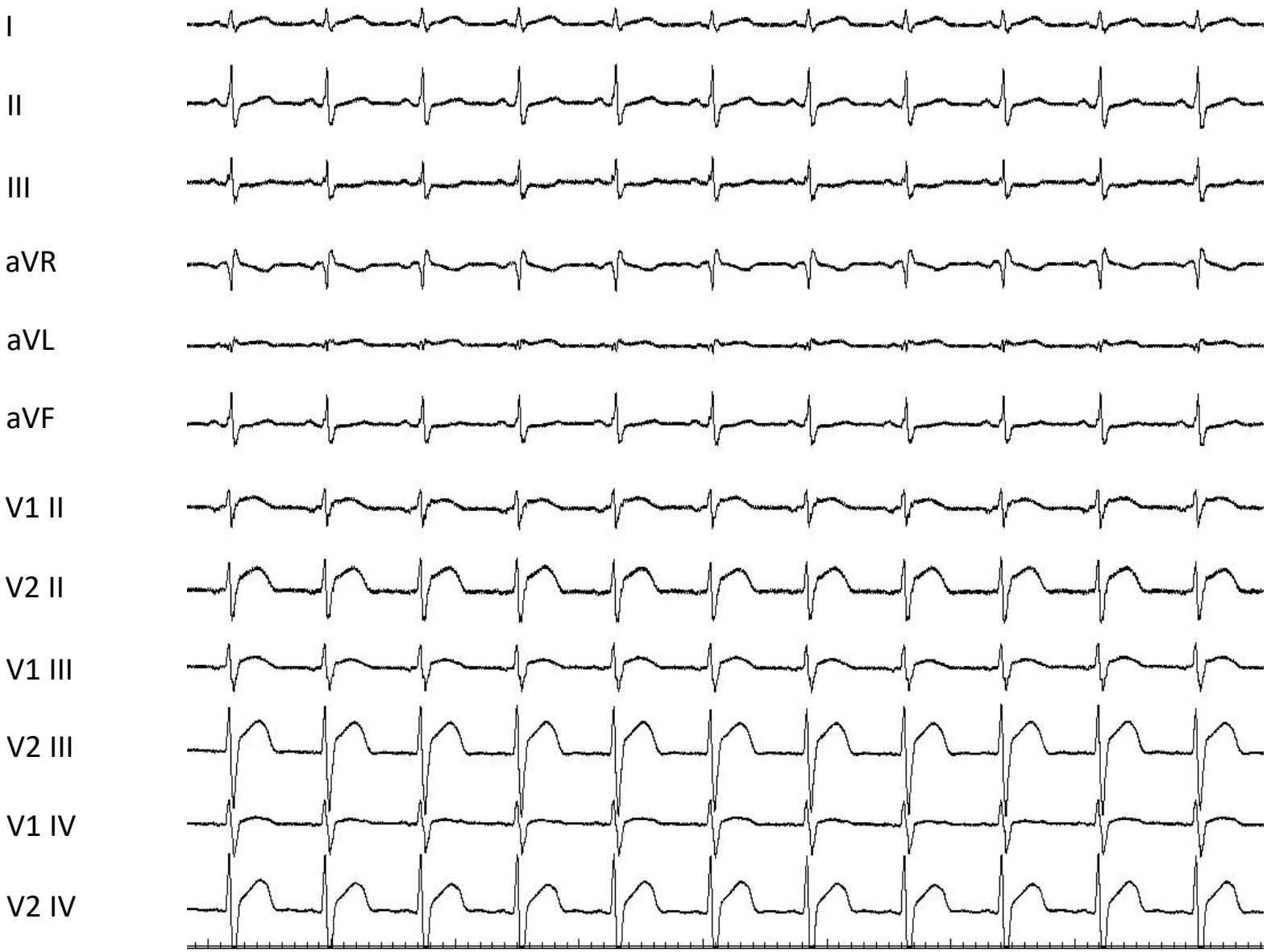
Before Ablation



After Ablation



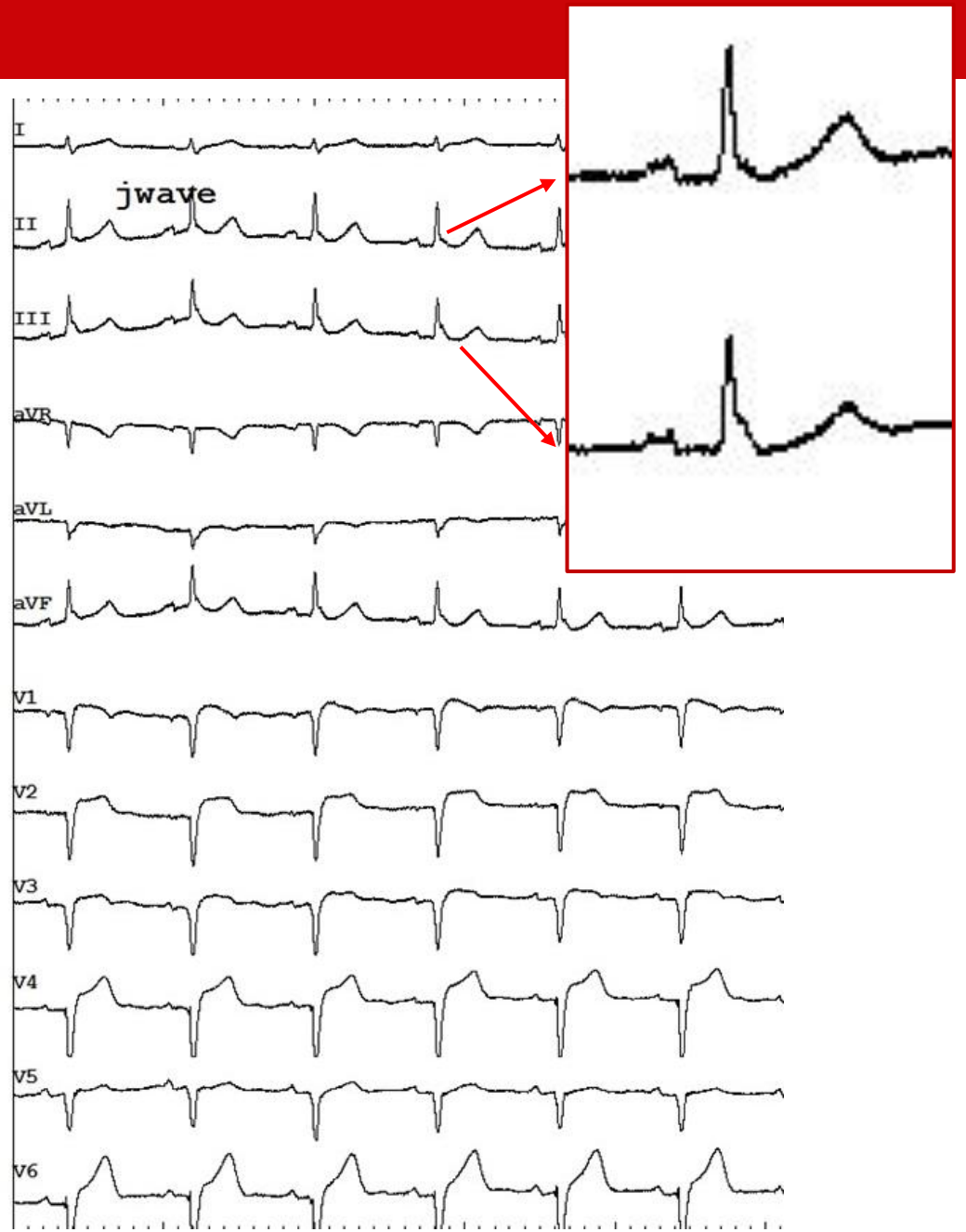
ECG AFTER ABLATION



Brugada and J-Wave Syndrome

SUMMARY

- Male 28 YO
- No family history of SCD
- 2019: positive Ajmaline test and ICD implantation
- 2020: VF episode with appropriate ICD shock
- 2021: epicardial mapping (admission reason)

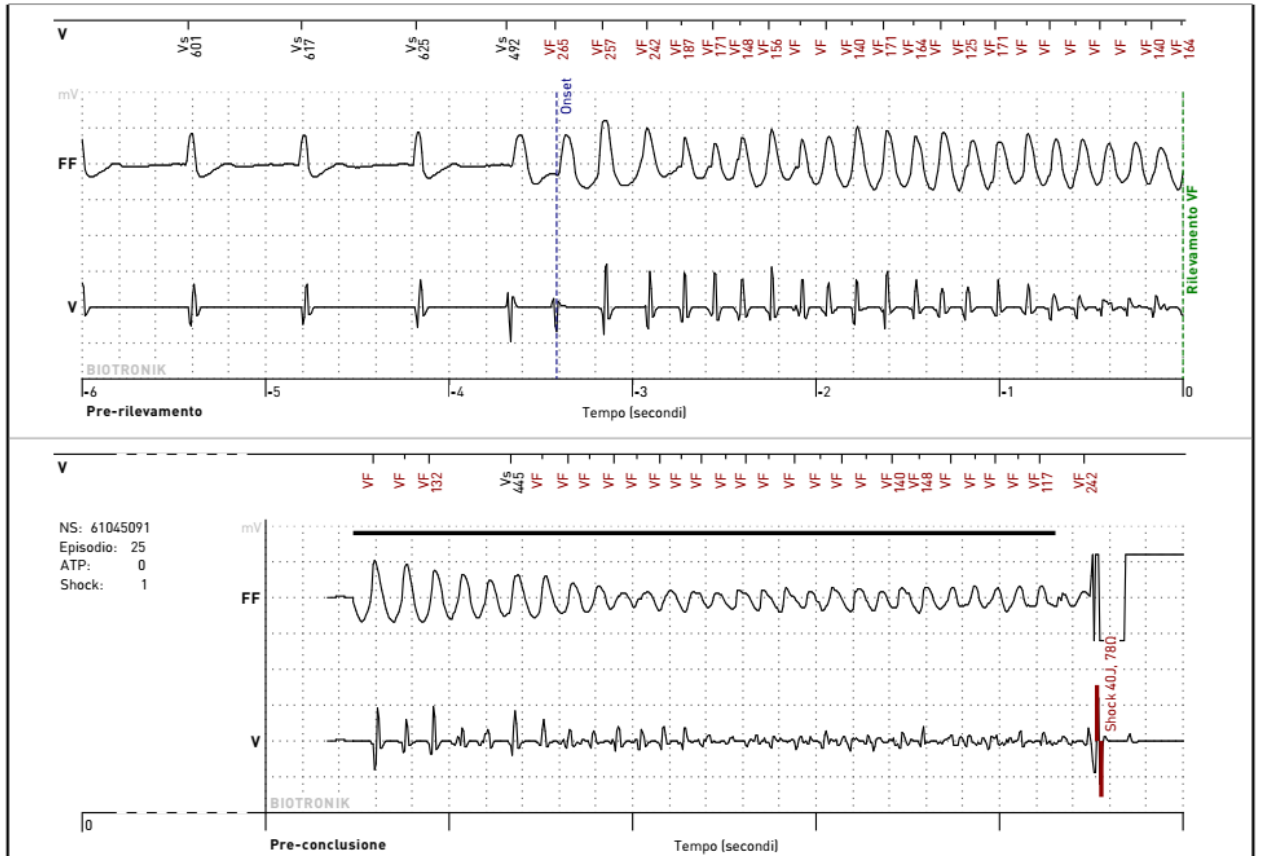


VF EPISODE (ICD registration, 2020)

Registrazioni - Episodio 25:

Generale	
Numero	25
Tipo	VF
Rilevamento	21-dic-2020 22.20.39
Term.	21-dic-2020 22.20.58
Durata	19s
Programmazione disp. impiantabile n.	3
Rilevamento	
RR medio al rilevamento iniziale [ms]	144
Onset [%]	59, soddisfatto
Stabilità [ms]	23
Criterio contatore morfologico [%]	OFF
Nuovo rilevamento	---

Terapia	
ATP erogati (totale)	0
ATP One Shot erogate	NO
Shock erogati	1
Shock abortiti	0
Energia massima [J]	40
Term.	
RR medio alla conclusione [ms]	752
Nota	
nessuno	



Brugada + J wave – L.L. 2021

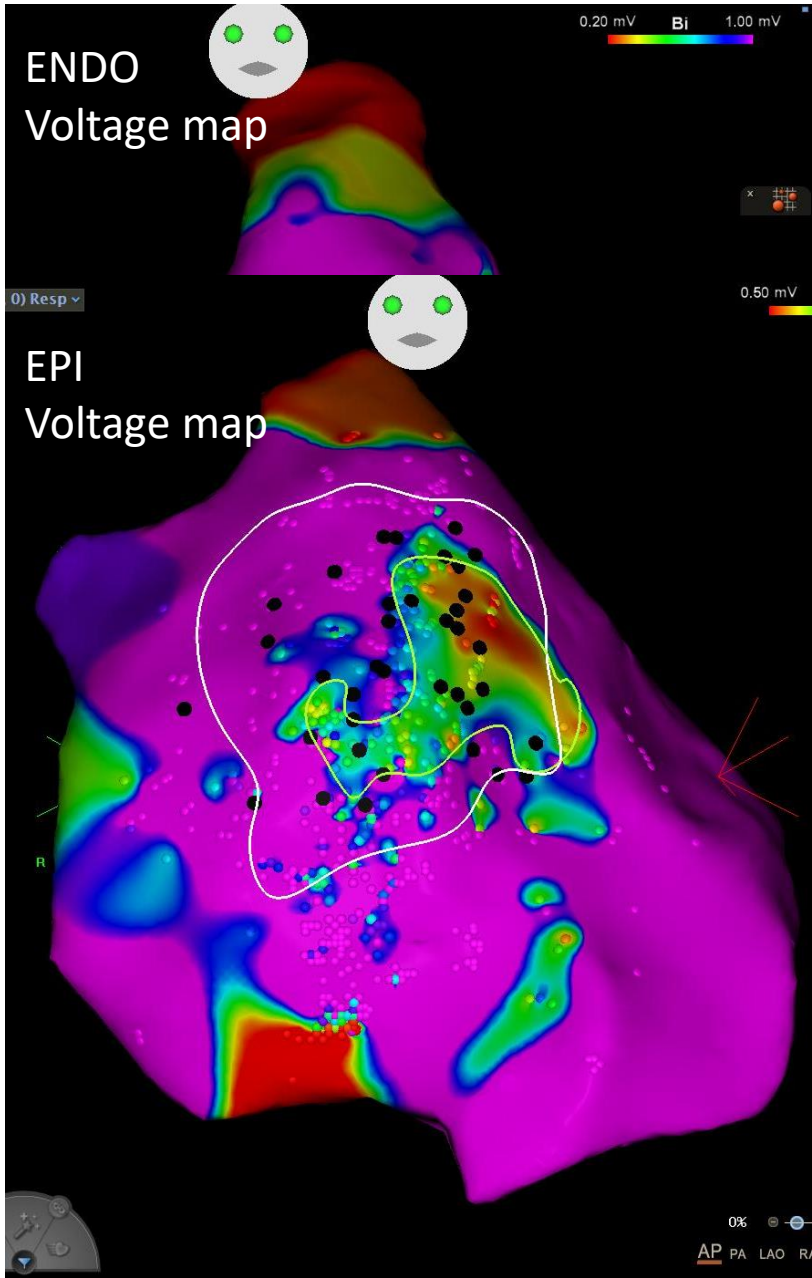
MAPPING DETAILS

Ablation date 05-05-2021

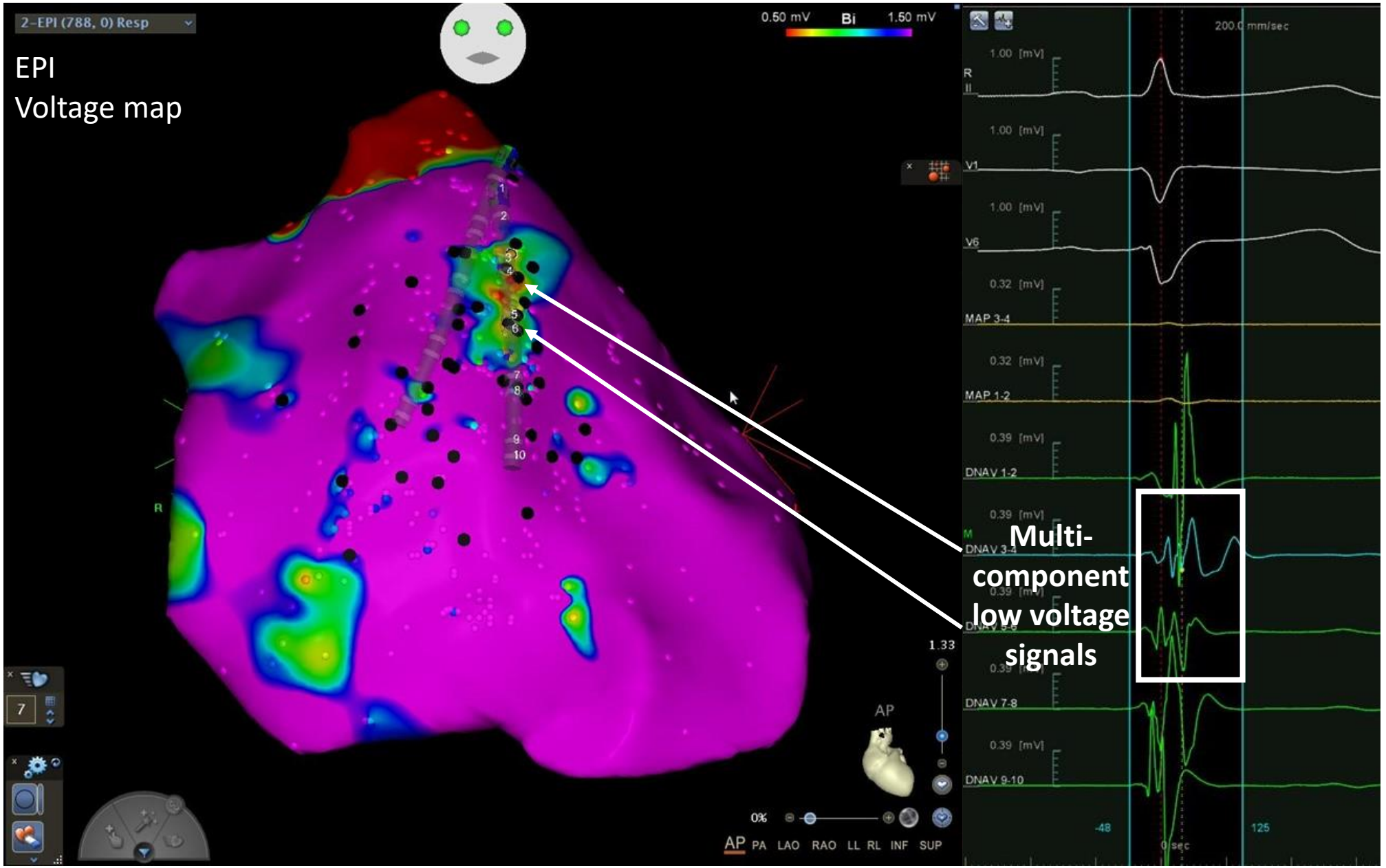
DecaNAV substrate mapping catheter

Endocardial mapping - Healthy RV

Epicardial mapping – Low voltage area and prolonged signals on anteroseptal wall

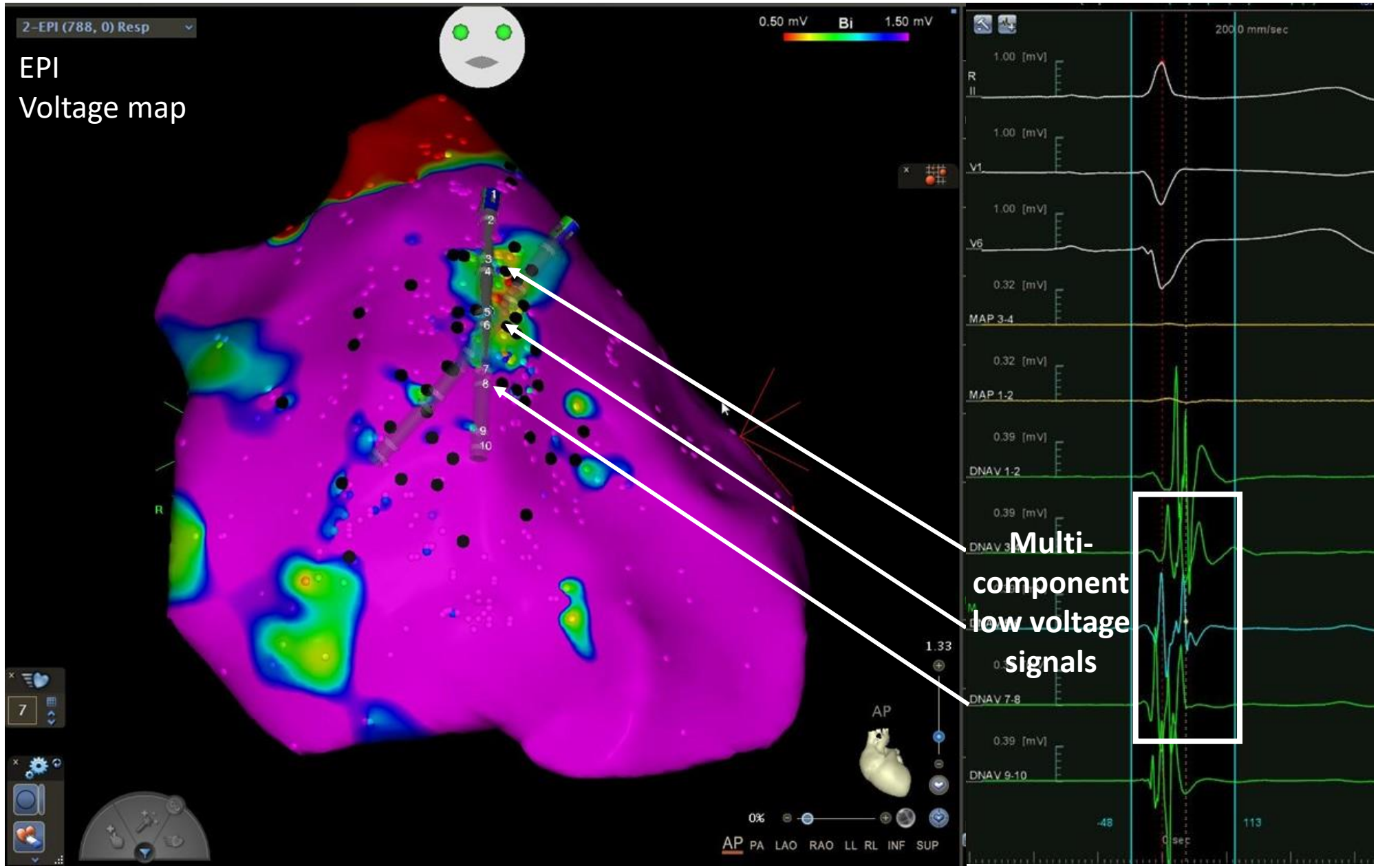


SIGNAL ANALYSIS – Target area



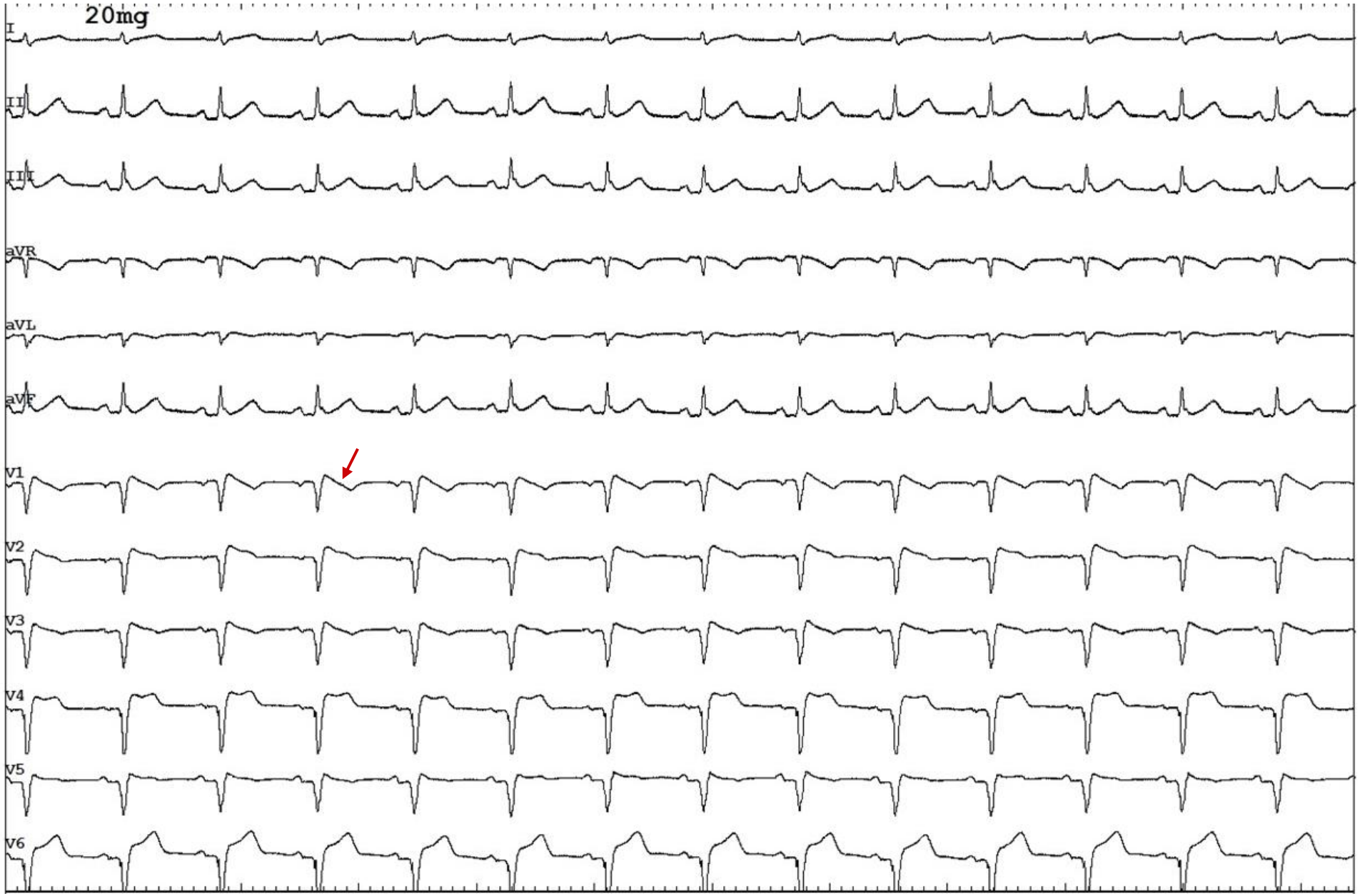
Brugada + J wave – L.L. 2021

SIGNAL ANALYSIS – Target area



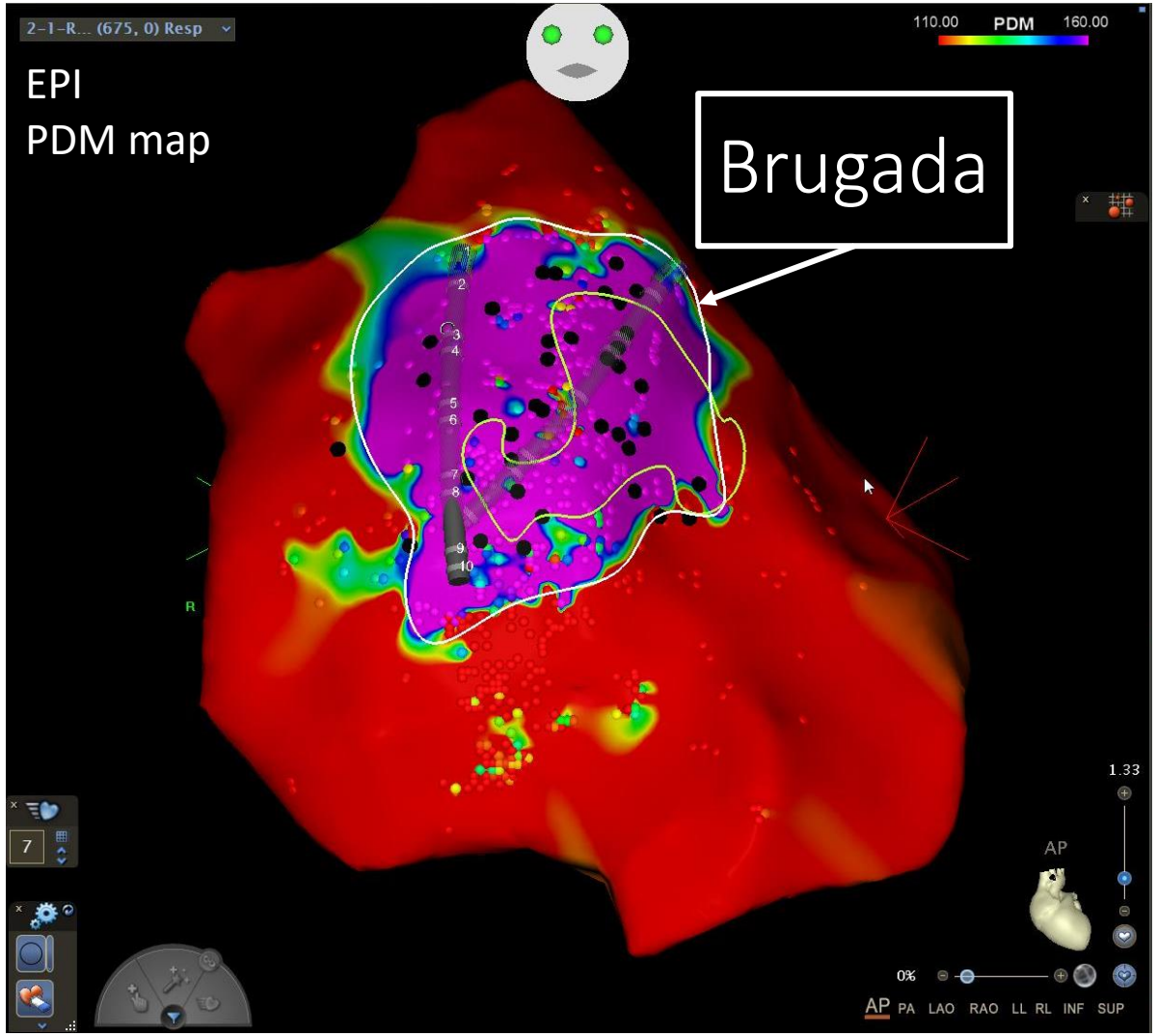
Brugada + J wave – L.L. 2021

ECG post 20mg ajmaline infusion (Brugada pattern manifestation)

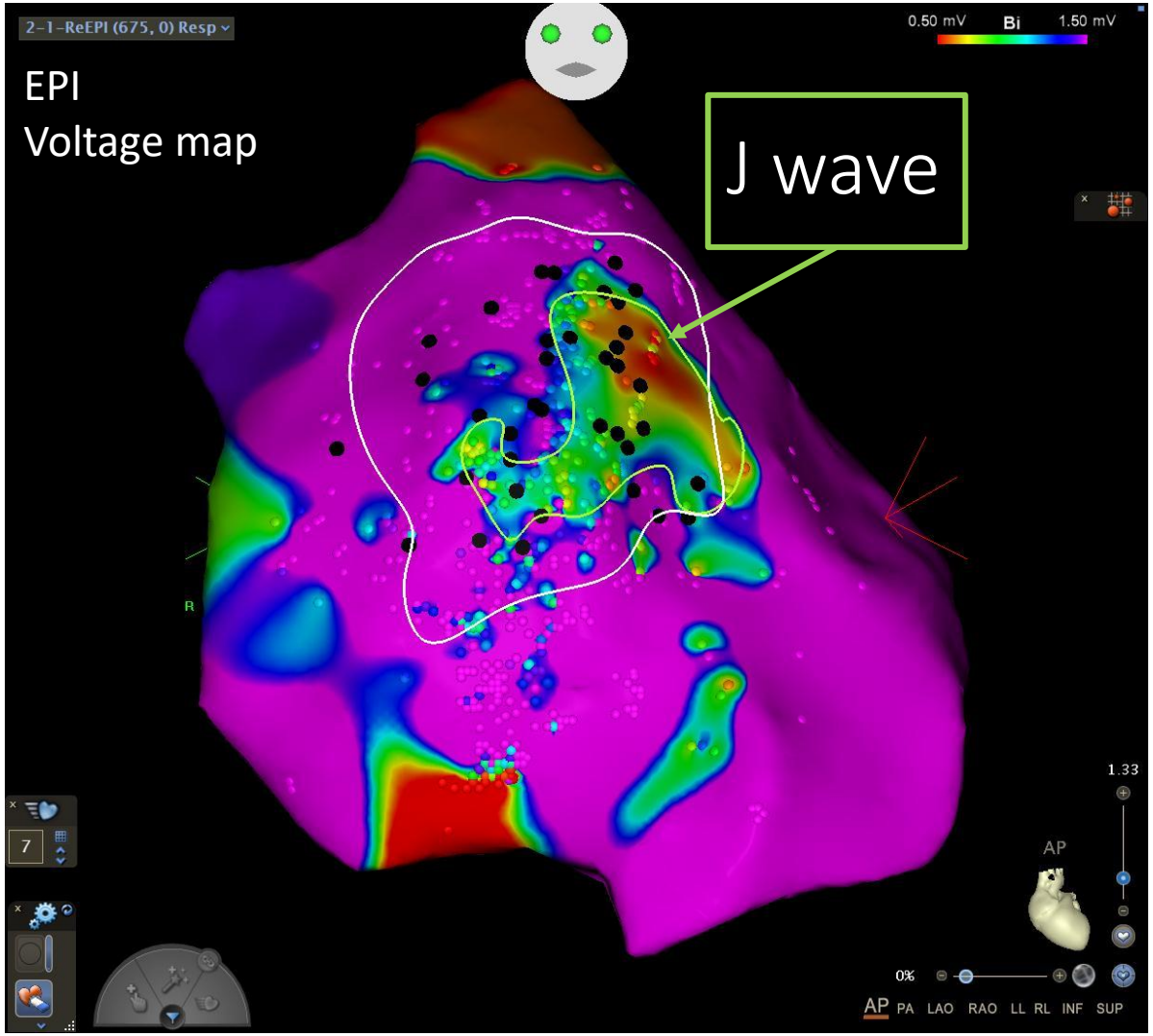


Brugada + J wave – L.L. 2021

AREA OF INTEREST: Brugada and Jwave areas



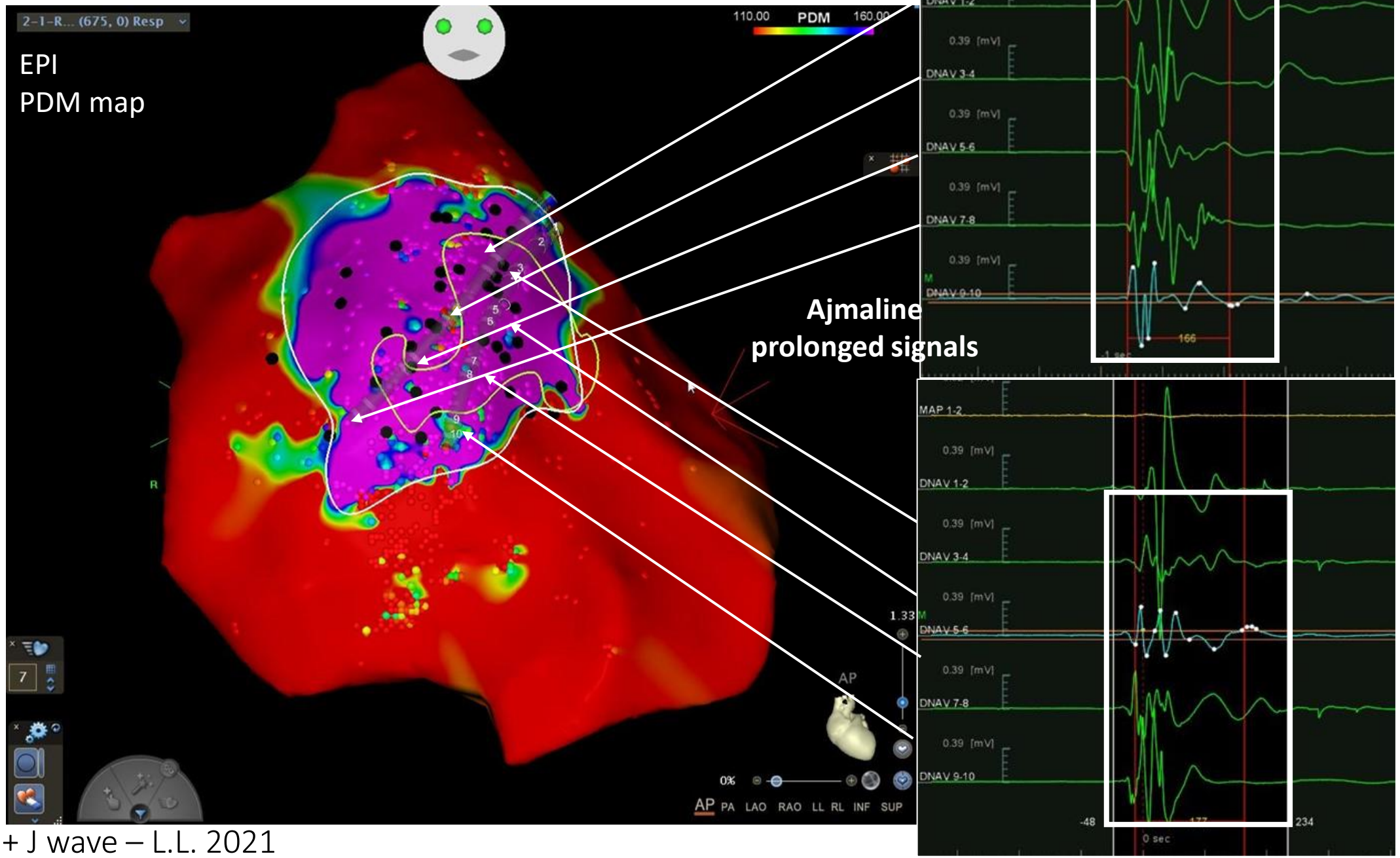
Ajmaline prolonged signals



Low voltage signals

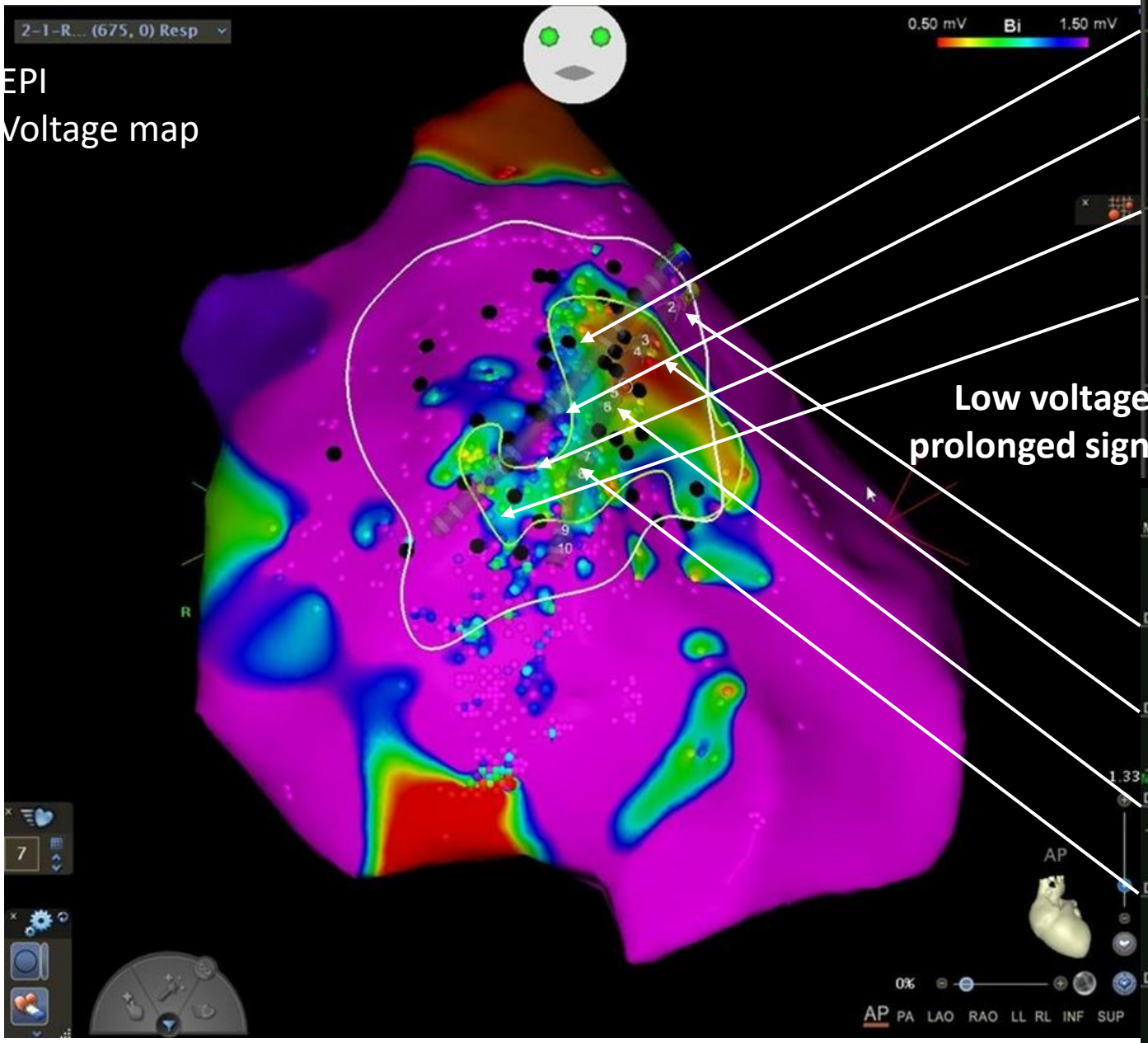
Brugada + J wave – L.L. 2021

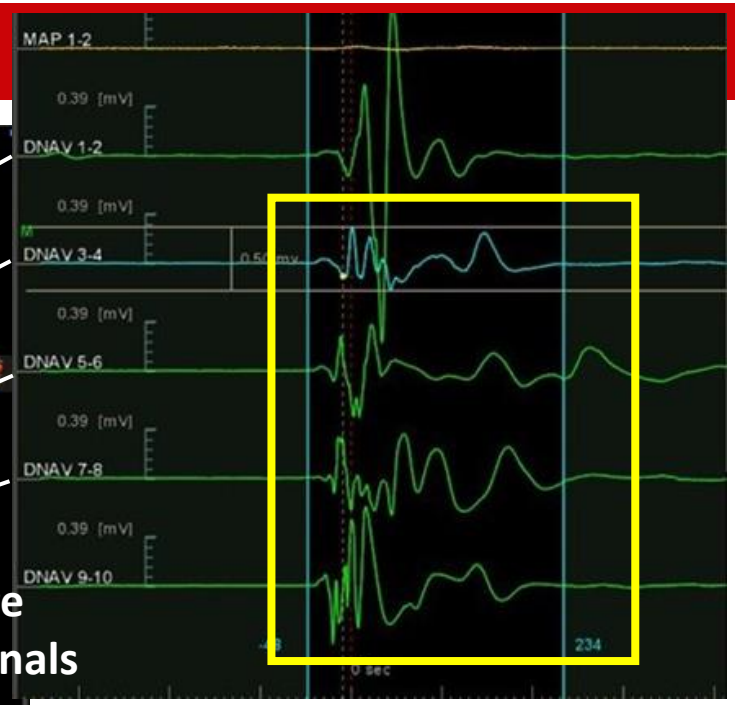
SIGNAL ANALYSIS – Brugada area



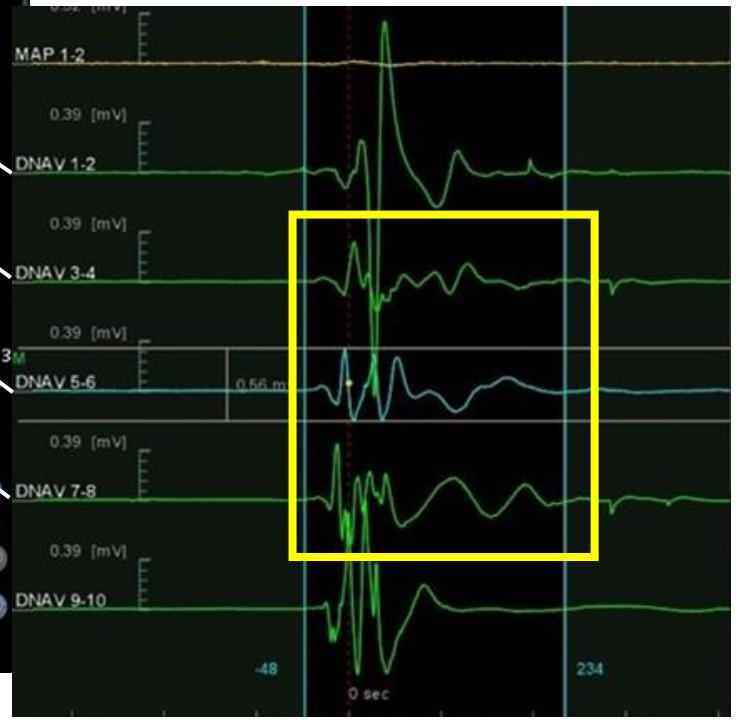
Brugada + J wave – L.L. 2021

SIGNAL ANALYSIS – J wave area





Low voltage prolonged signals



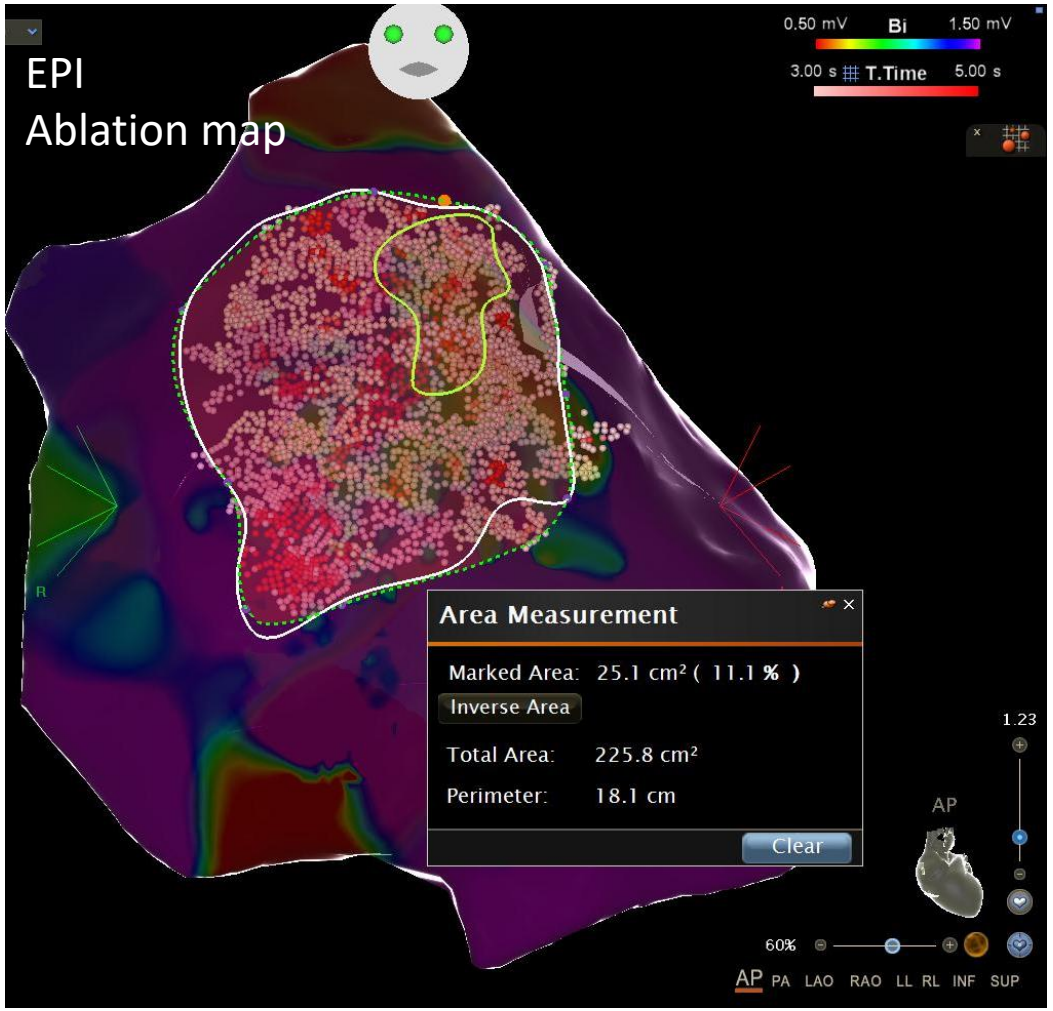
Brugada + J wave – L.L. 2021

ABLATION STRATEGY

Homogenization of both areas (fragmented low voltage potentials area and ajmaline prolonged signals area) with SmartTouch SF catheter

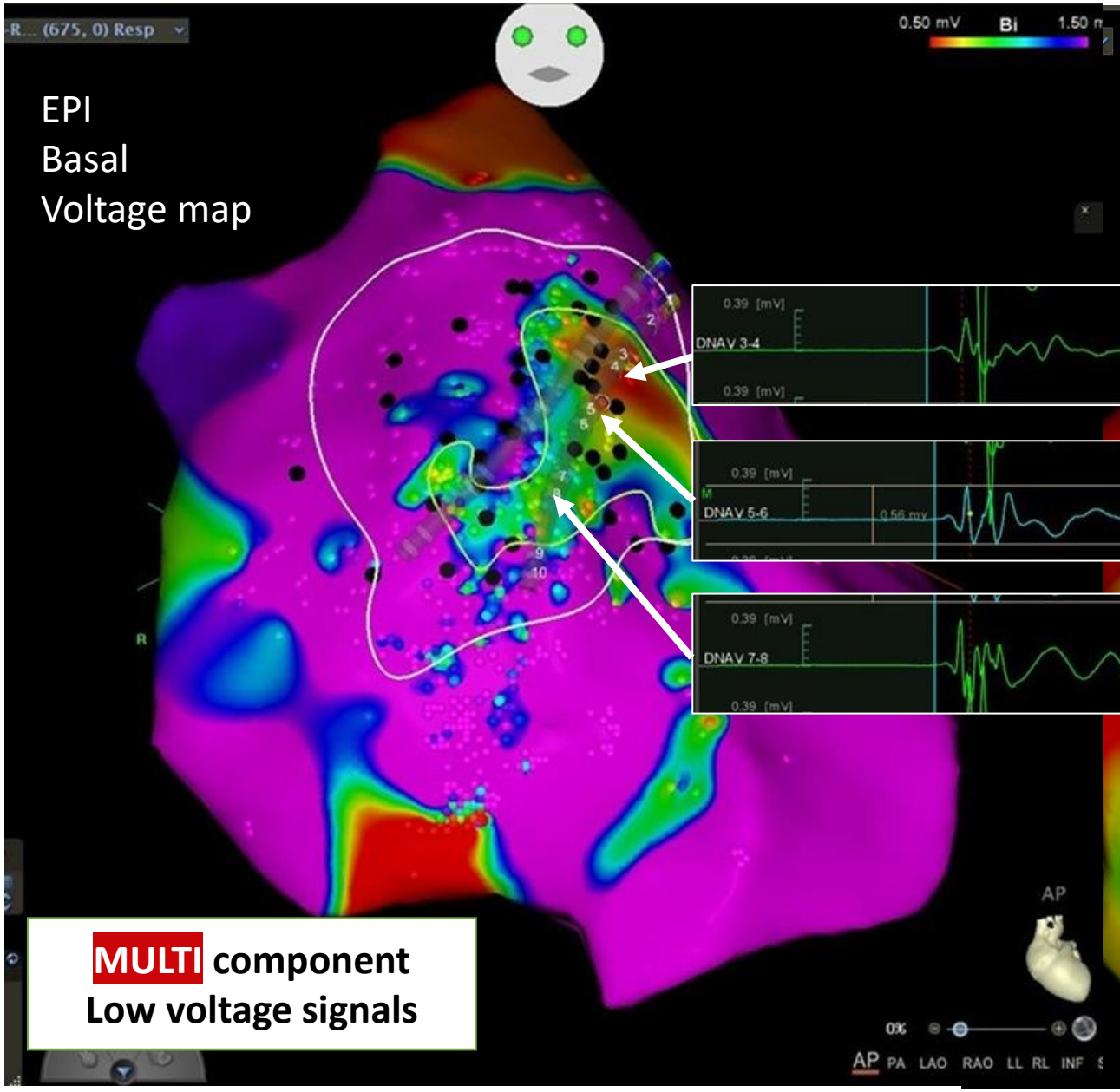
40-45 watts with fast dragging technique, to reach superficial lesion

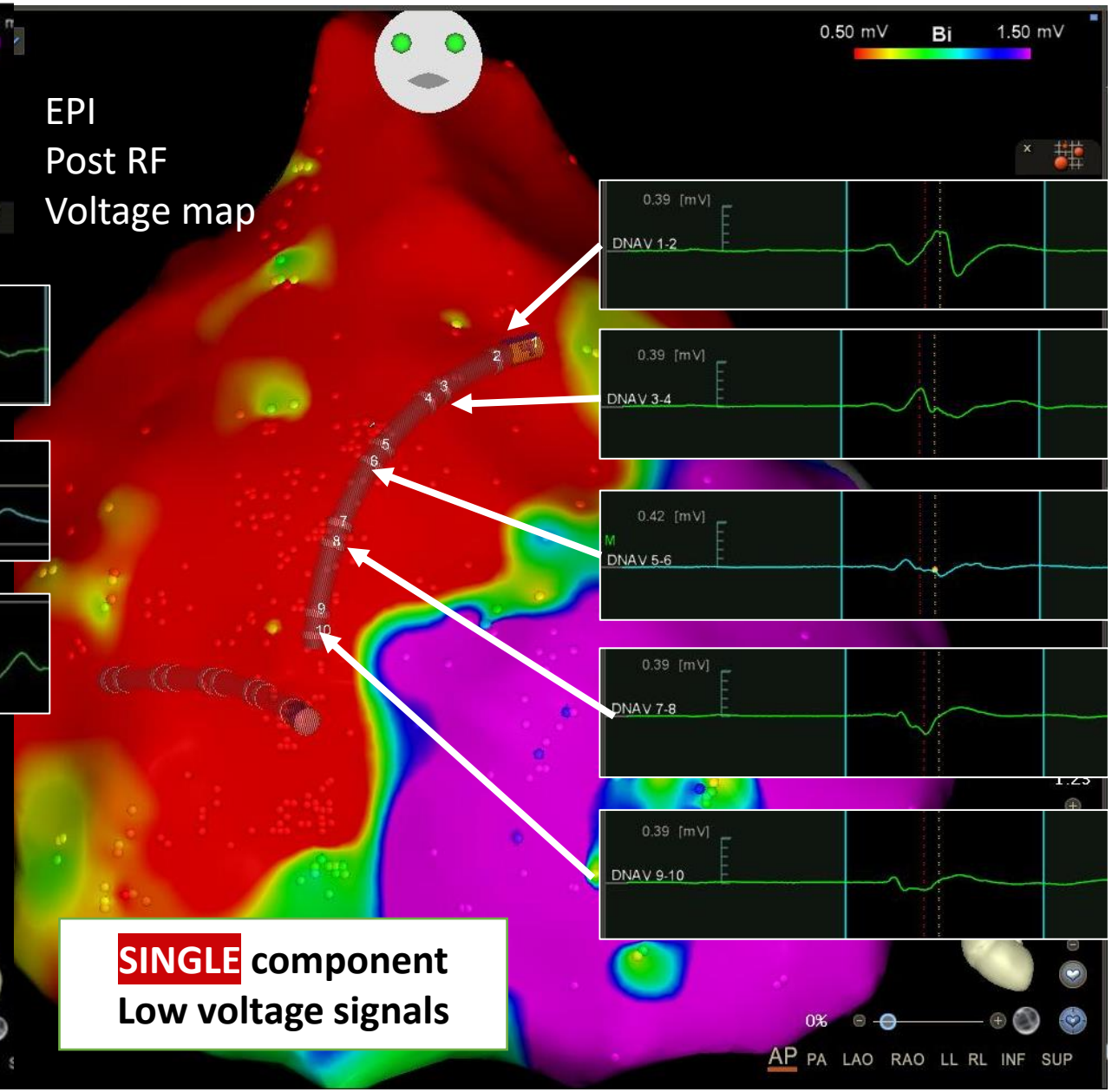
42 minutes of total ablation for a 25.1 cm^2 area



Brugada + J wave – L.L. 2021

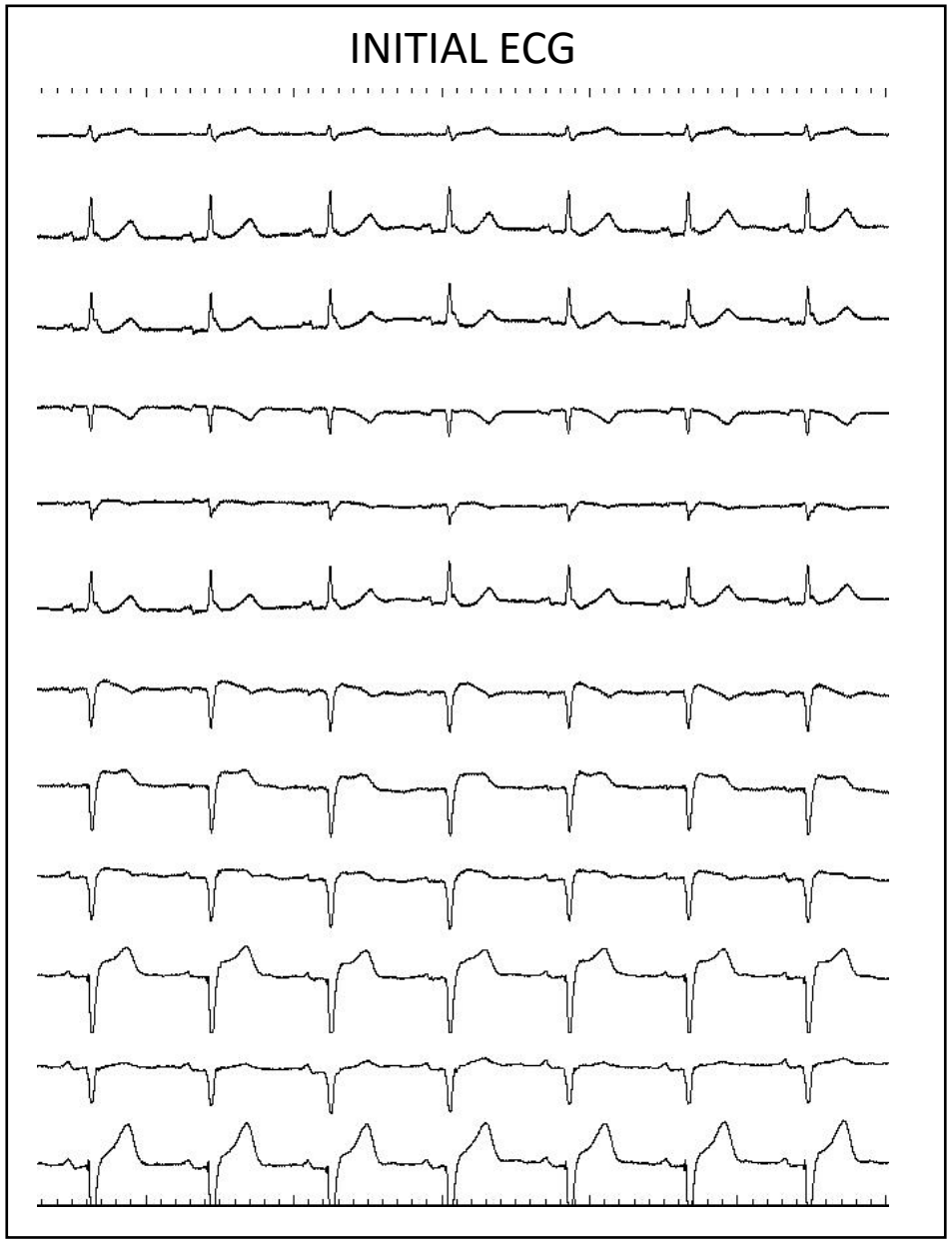
SIGNAL COMPARISON in the target area: PRE AND POST ABLATION

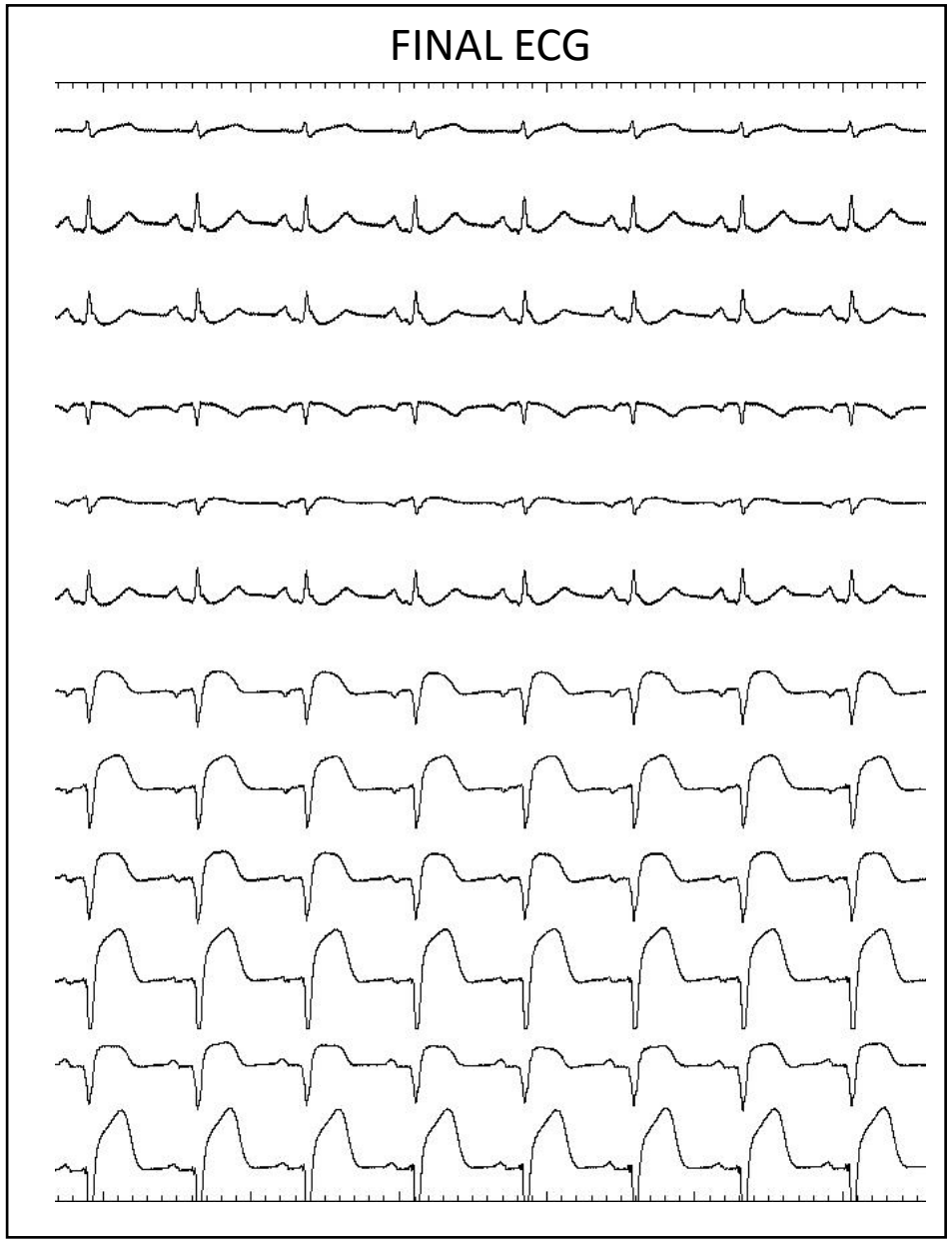




Brugada + J wave – L.L. 2021

ECG MODIFICATION POST RF: no J wave



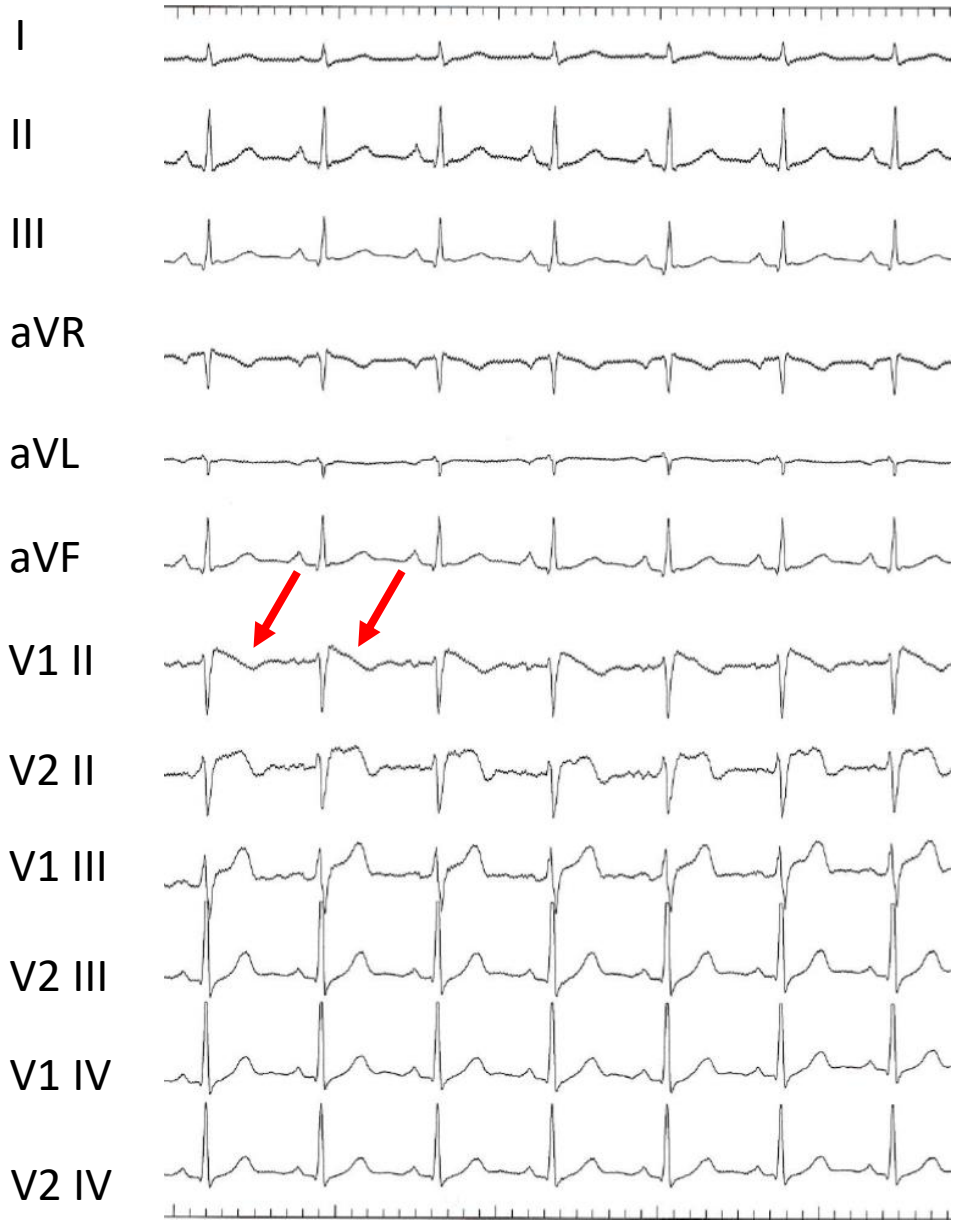


Brugada + J wave – L.L. 2021

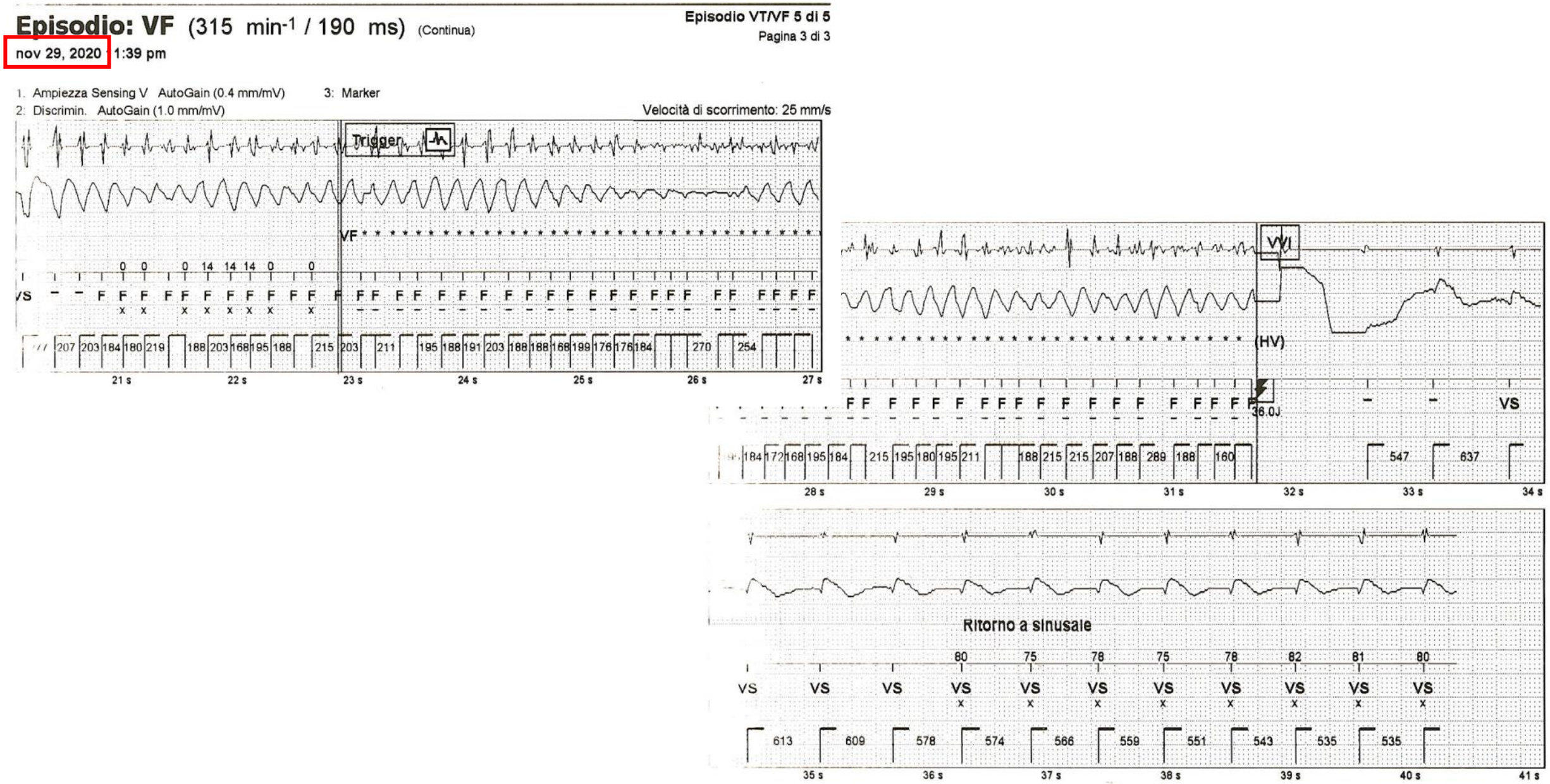
Brugada Syndrome and ARVD

SUMMARY

- Male 48 YO
- 2011: CA due to VF, Brugada syndrome type I and ARVD diagnosis, ICD implantation
- 2013: 2 ICD therapies, positive CACNB2 mutation test
- 2019: 1 ICD therapy
- November 2020: 1 ICD therapy, after less than ten hours one more FV recurrence and Brugada ablation

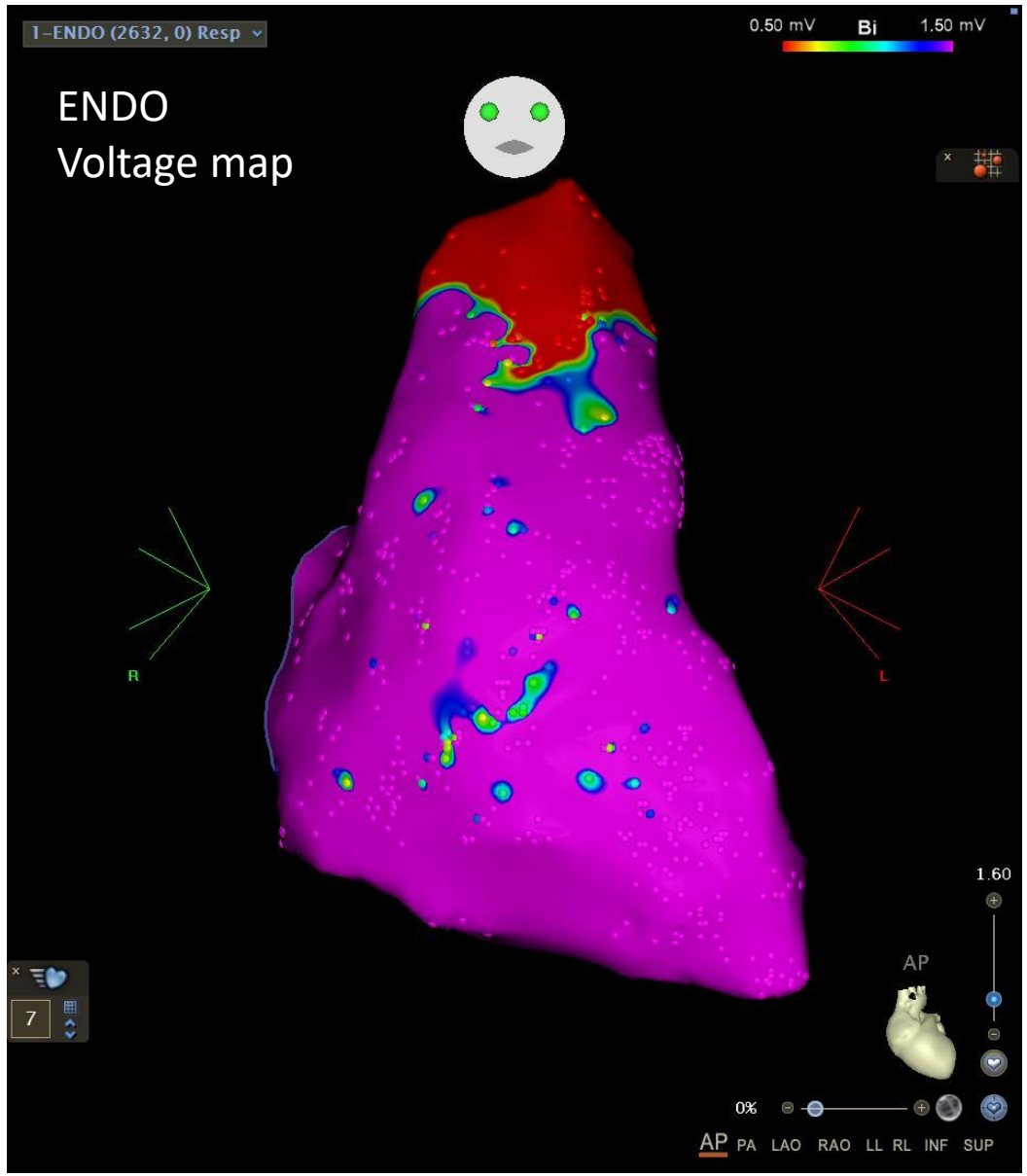


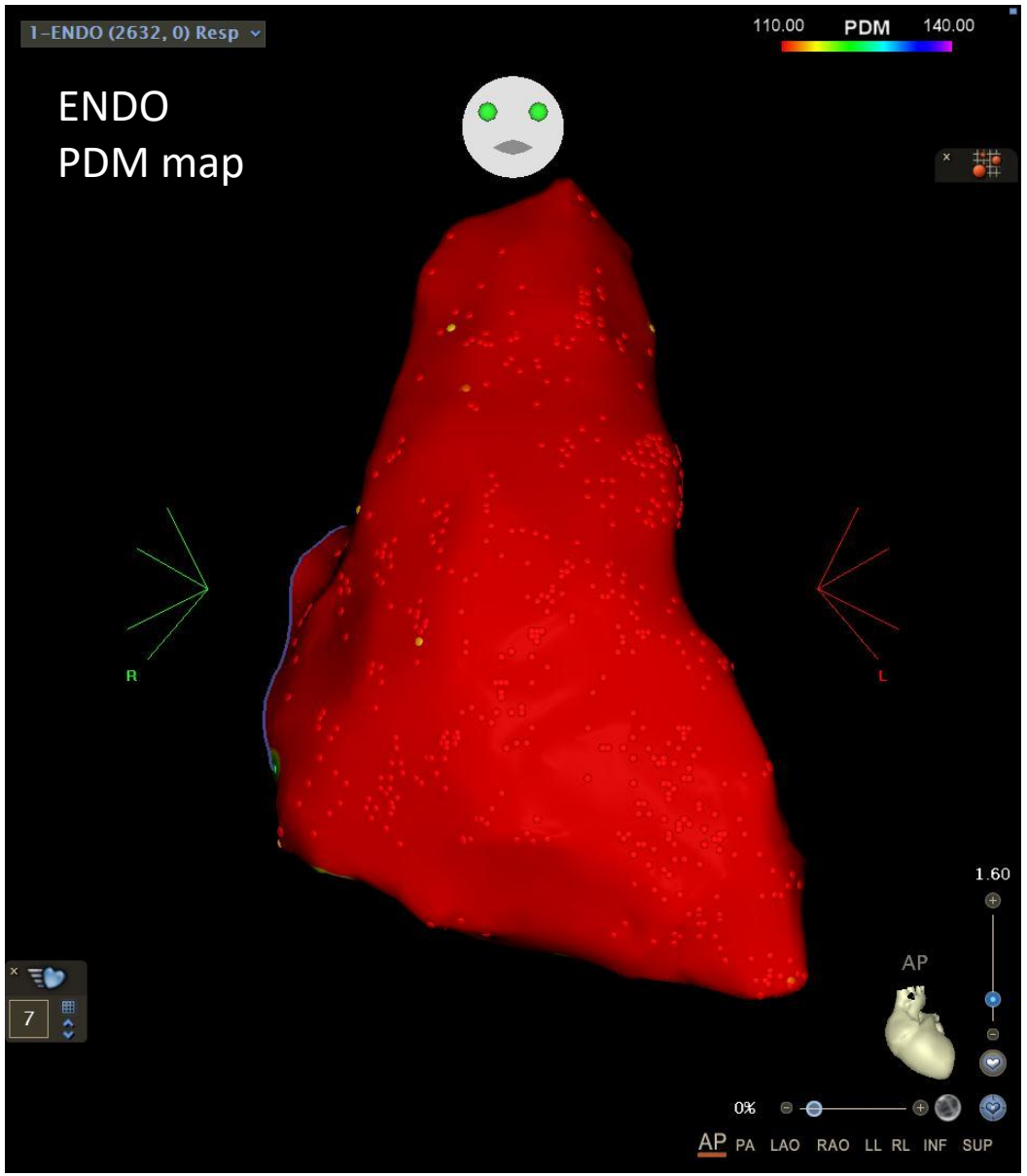
VF EPISODE (ICD registration 29/11/2020)



Brugada + ARVD – M.A. 2020

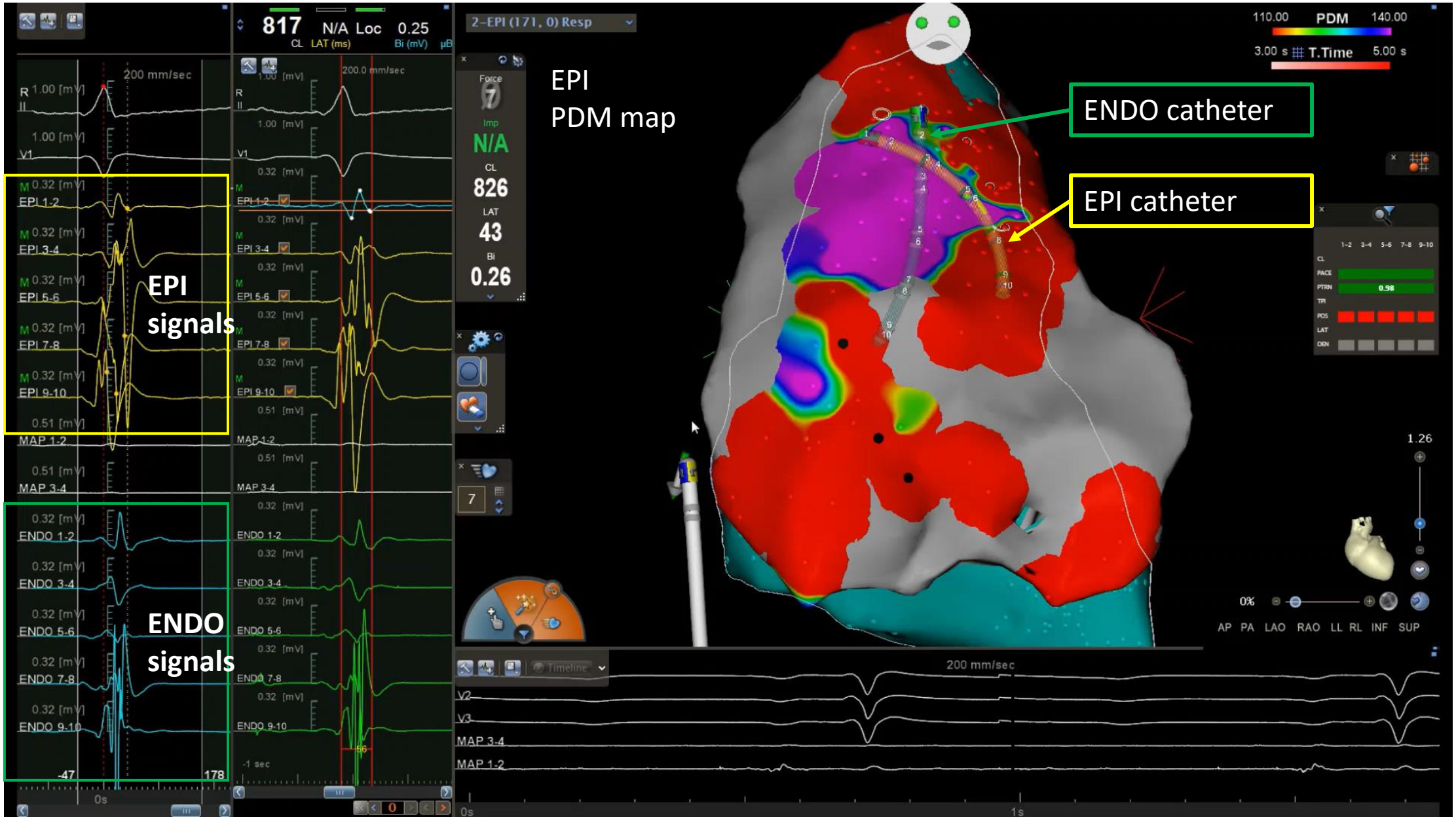
ENDOCARDIAL MAP: healthy RV





Brugada + ARVD – M.A. 2020

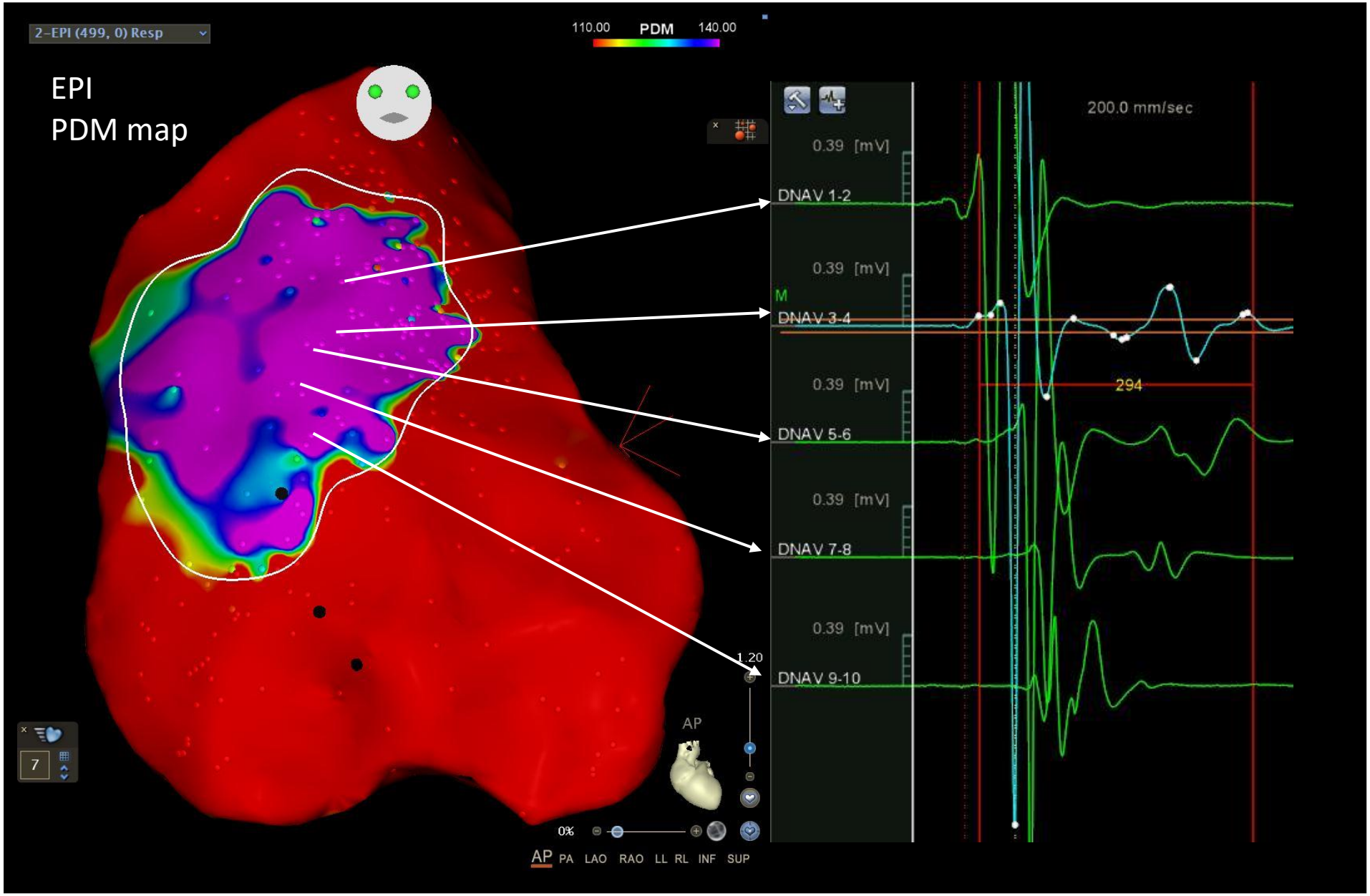
BASELINE EPICARDIAL MAPPING



BASELINE EPICARDIAL MAPPING: ENDO vs EPI SIGNALS

Brugada + ARVD – M.A. 2020

TARGET AREA (area with prolonged signals)



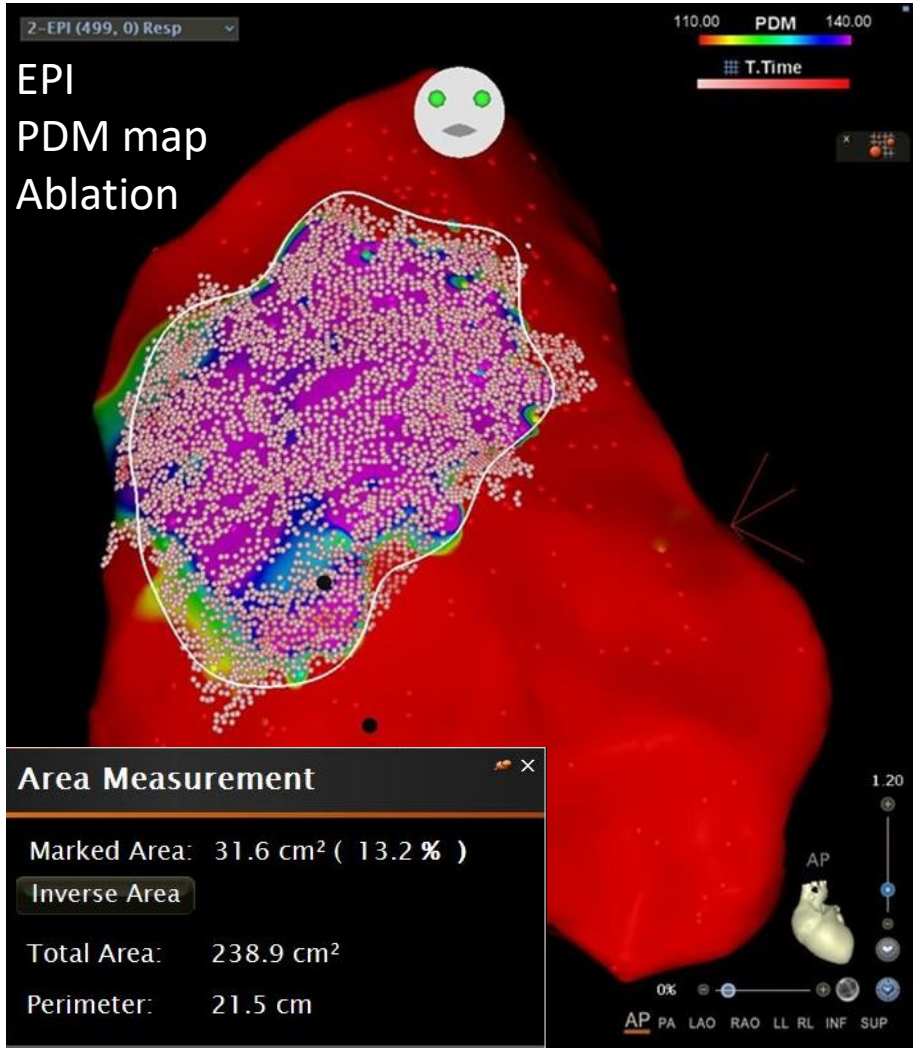
Brugada + ARVD – M.A. 2020

ABLATION STRATEGY

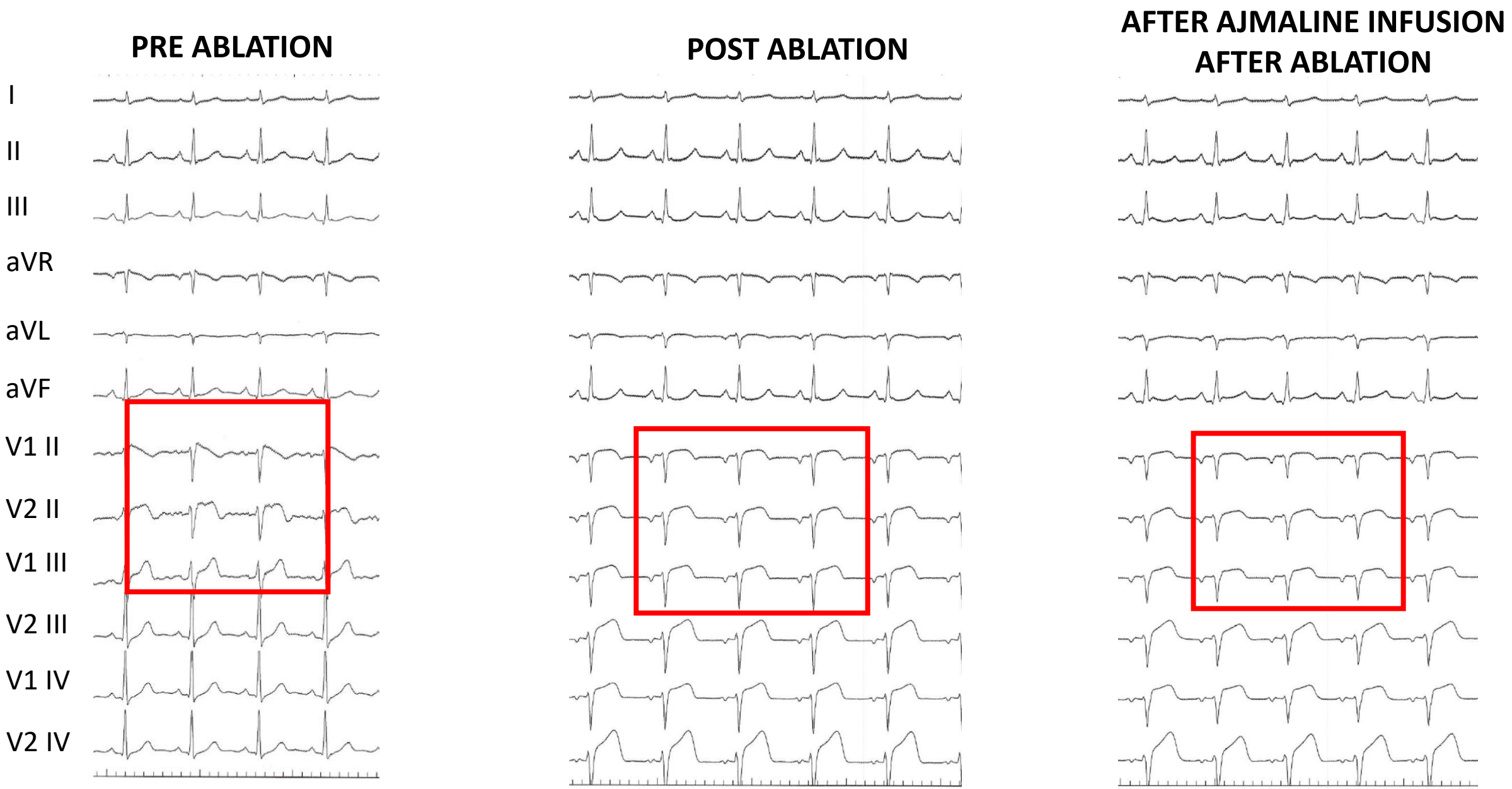
Homogenization of fragmented low voltage potentials area with SmartTouch SF catheter

40-45 watts with fast dragging technique, to reach superficial lesion

40 minutes of total ablation on a 31.6 cm^2 area



ECG MODIFICATION AFTER ABLATION

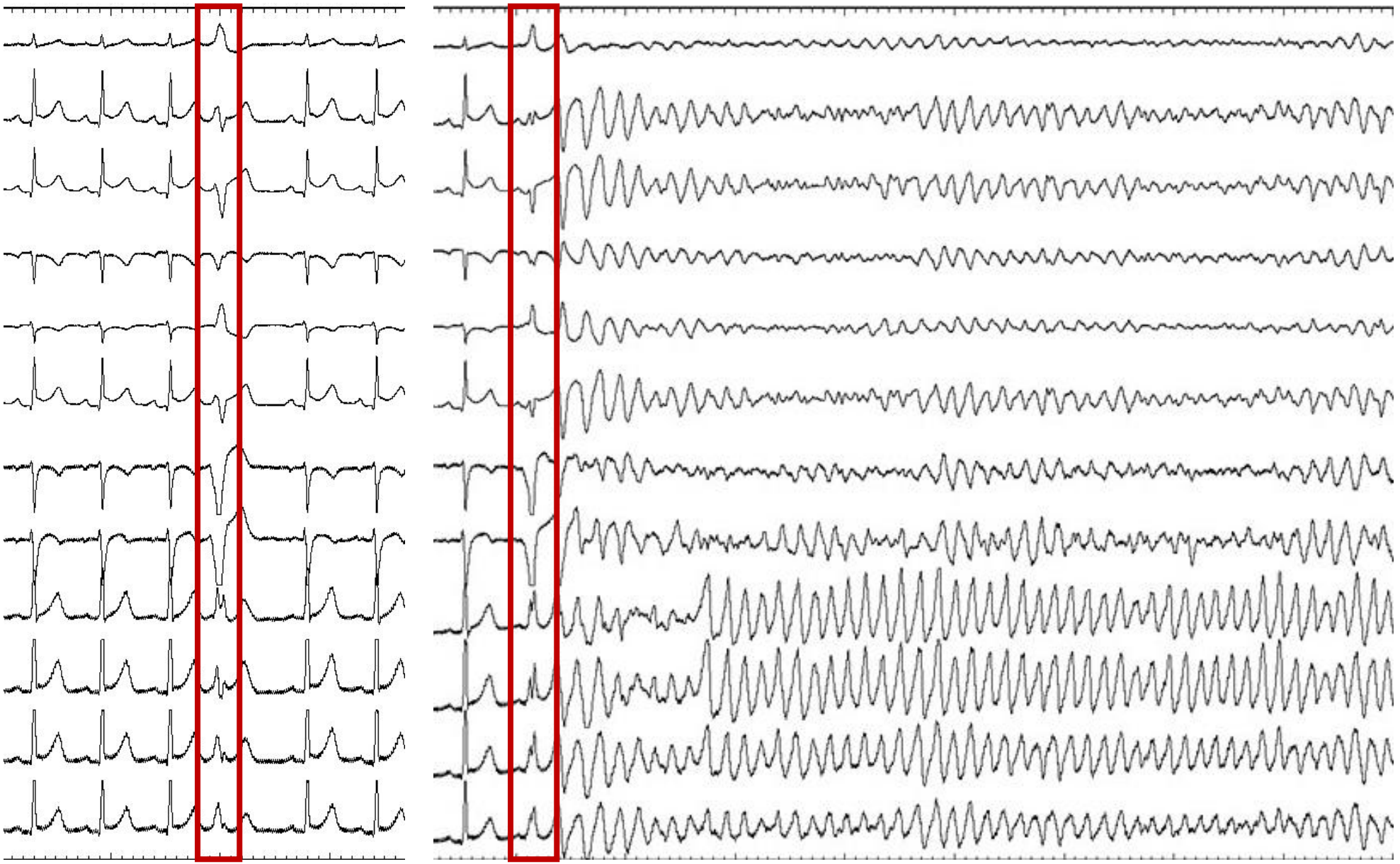


Brugada + ARVD – M.A. 2020

INITIAL ECG: frequent PVCs

1-Month Later….

Superior axis V3 transition



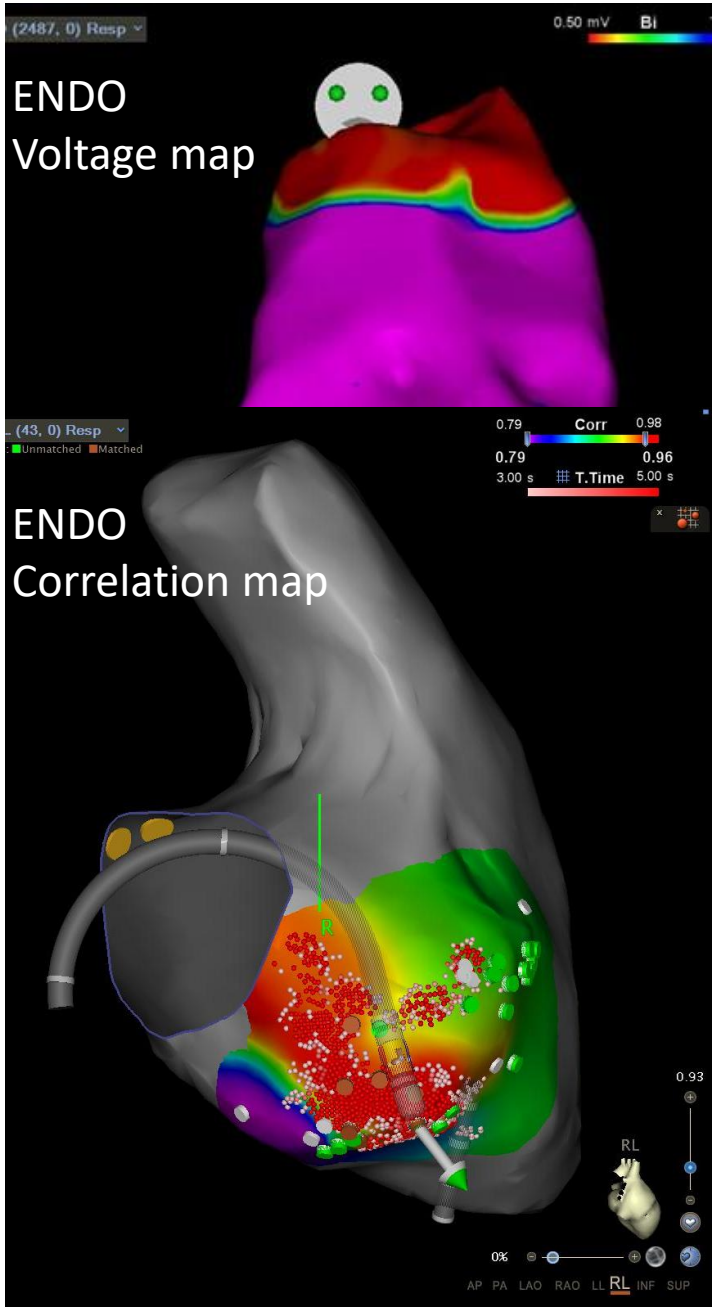
MAPPING DETAILS

Ablation date 22-12-2020

DecaNAV substrate mapping catheter

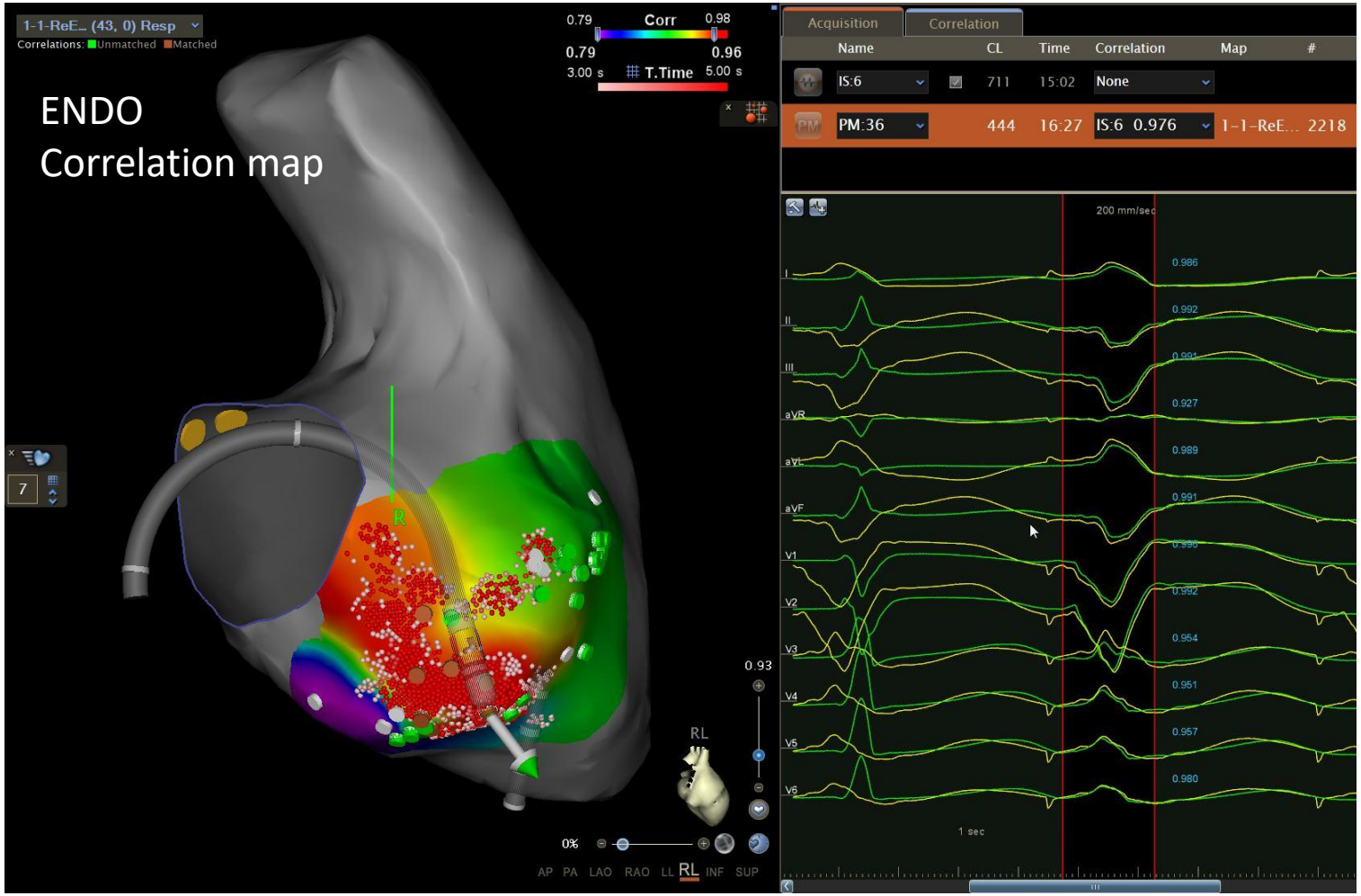
Endocardial mapping - Healthy RV

Endocardial mapping - Frequent PVC, origin identification with pace mapping



PVC mapping: PASO CORRELATION MAP

- Best correlation (98%) on lateral free wall, 1 cm from tricuspid valve
- Large area with good pace mapping and 25 to 35 msec endocardial signal precocity



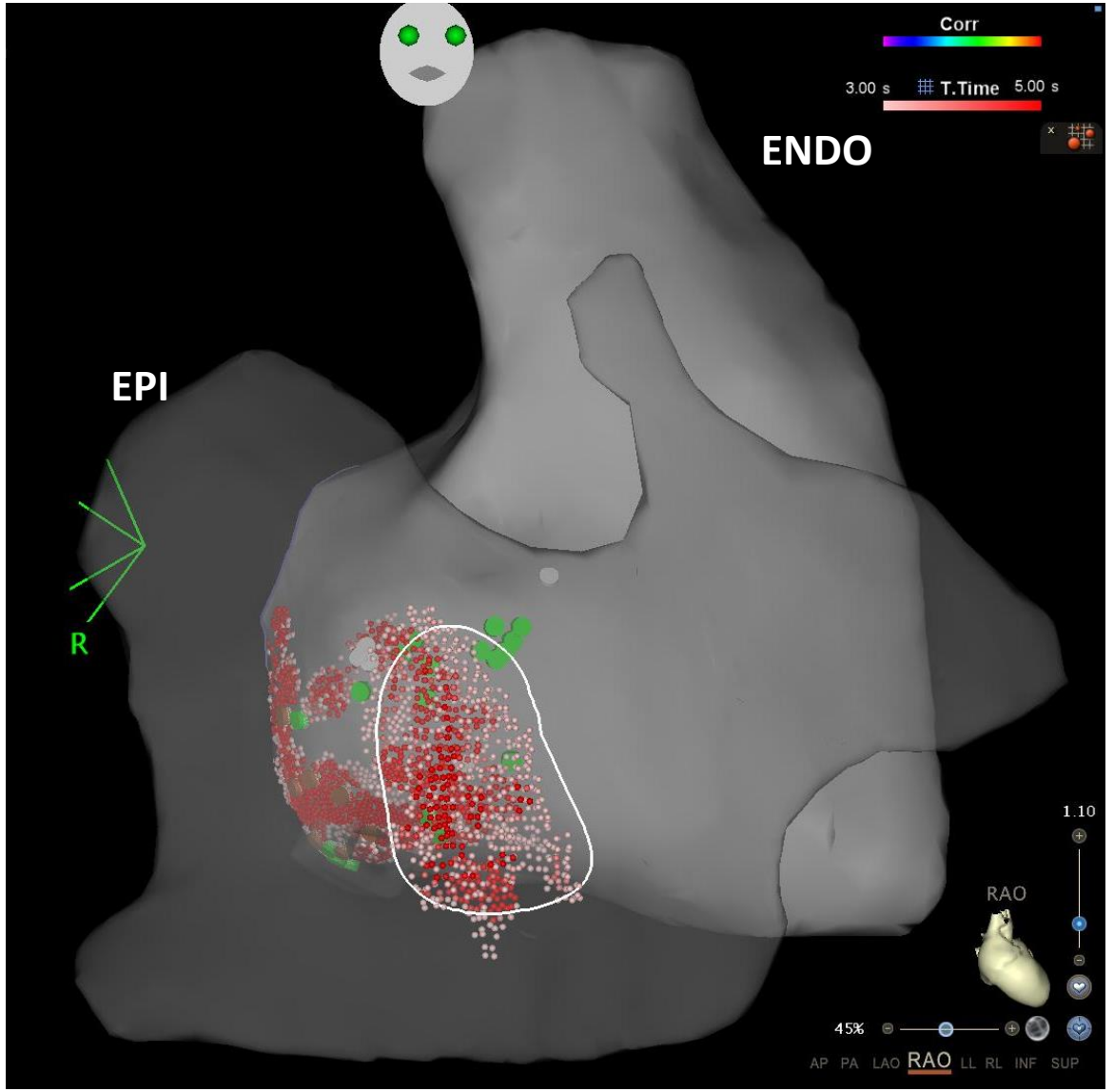
Brugada + ARVD – M.A. 2020

ABLATION STRATEGY

Ablation over the best correlation area with QDOT catheter

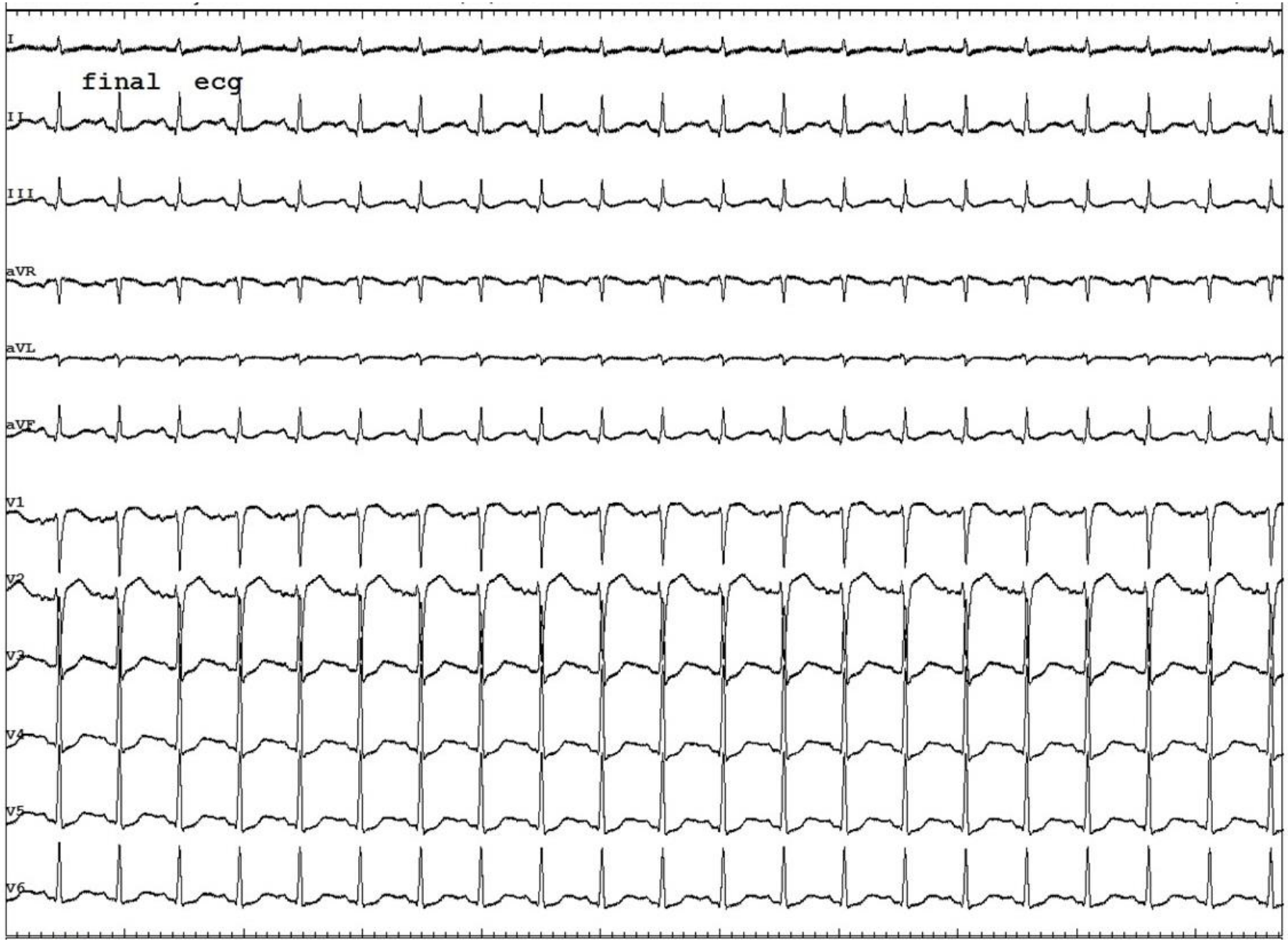
40-45 watts with fast dragging technique

Since the endocardial precocity area was large, an intramural origin of the PVC was targeted with epicardial anatomical “kissing” ablation



Brugada + ARVD – M.A. 2020

FINAL ECG: no more PVCs

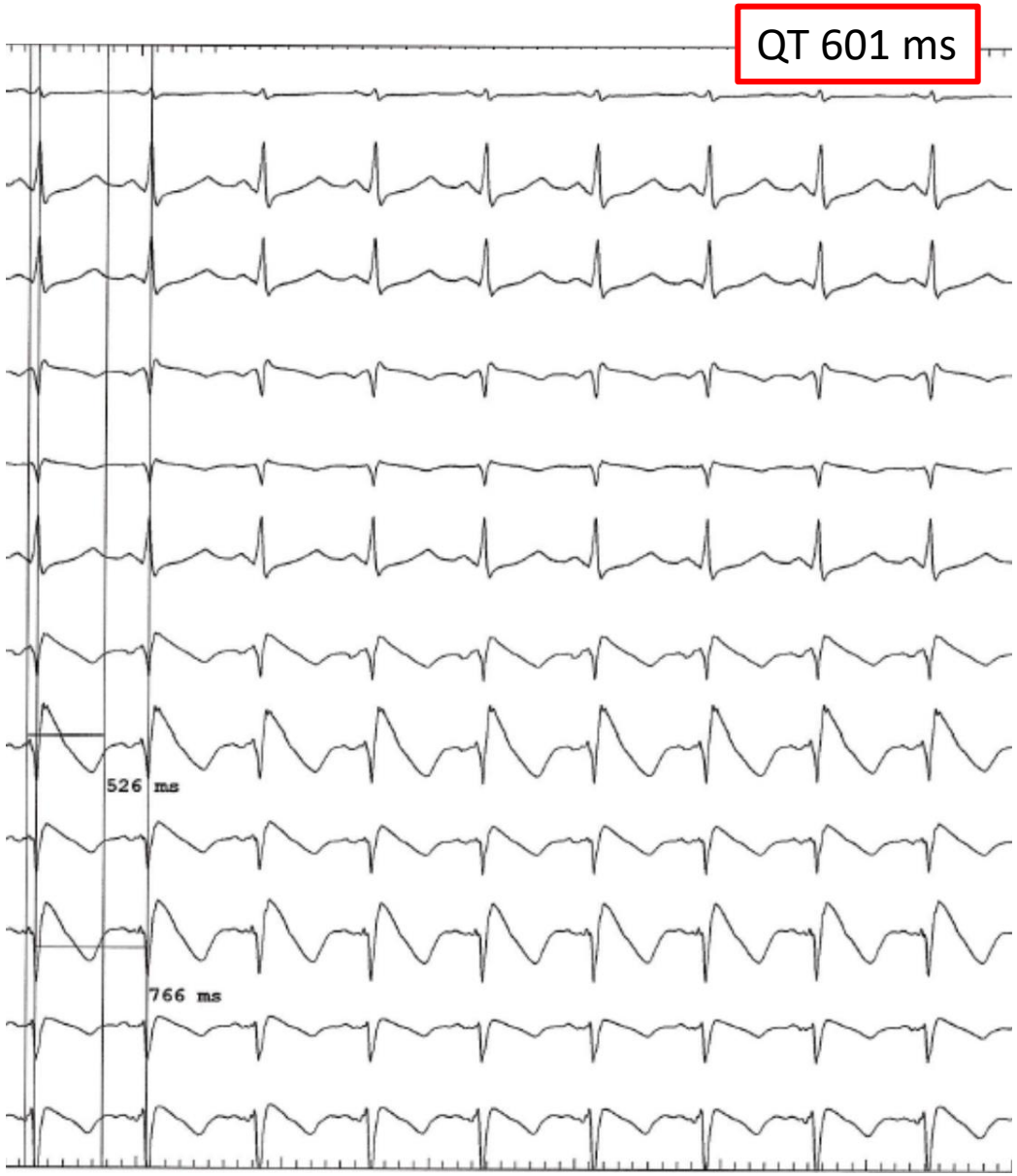


Brugada + ARVD – M.A. 2020

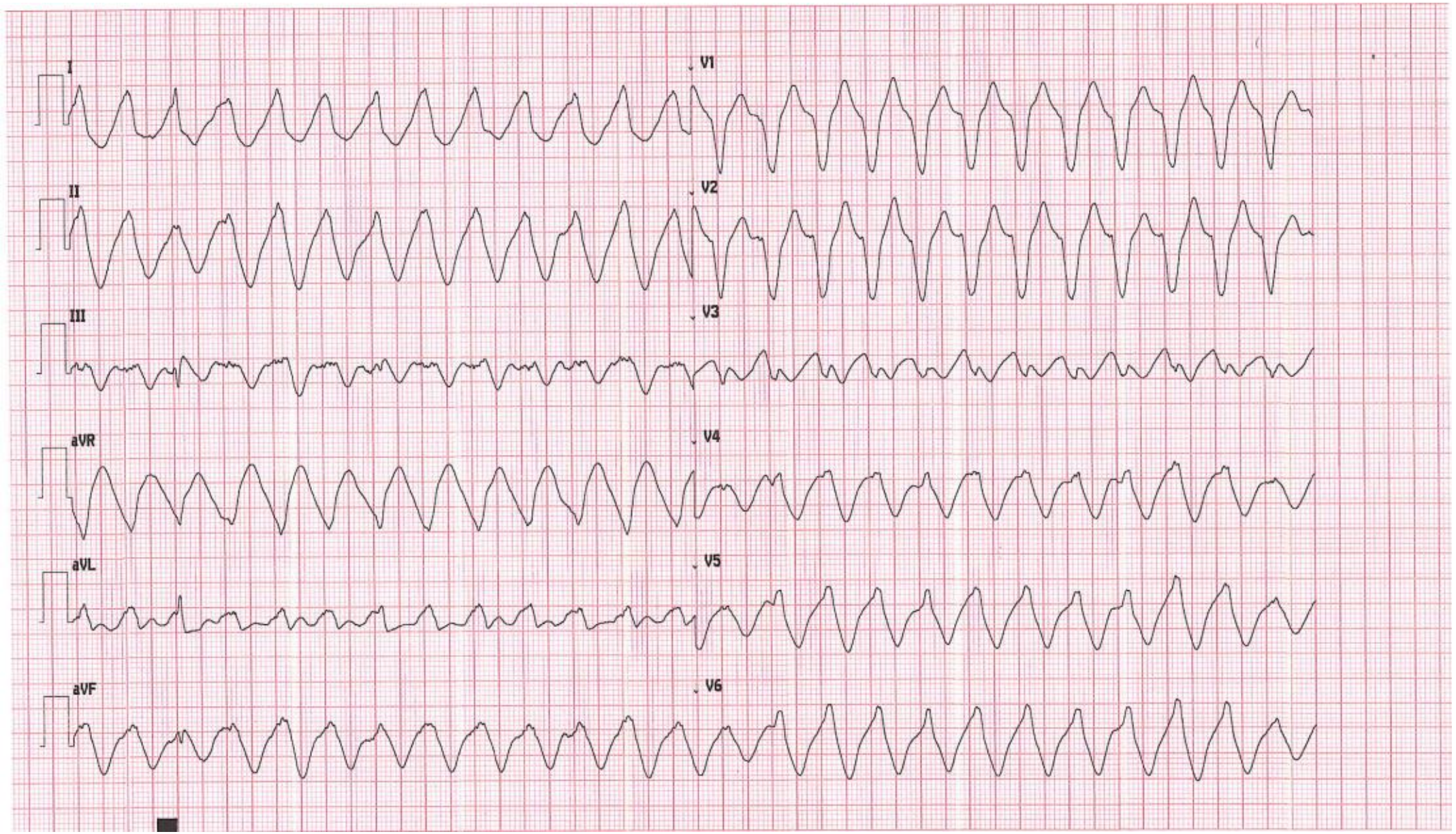
Brugada, ARVD and LQT Syndrome

SUMMARY

- Female 52 YO
- No family history of SCD
- 2010: Brugada type I diagnosis
- 2021: episode of VT and ICD implantation
- 2021: epicardial mapping (admission reason)



VT EPISODE (ER admission, 2021)



Brugada + ARVD + LQTs – P.G. 2021

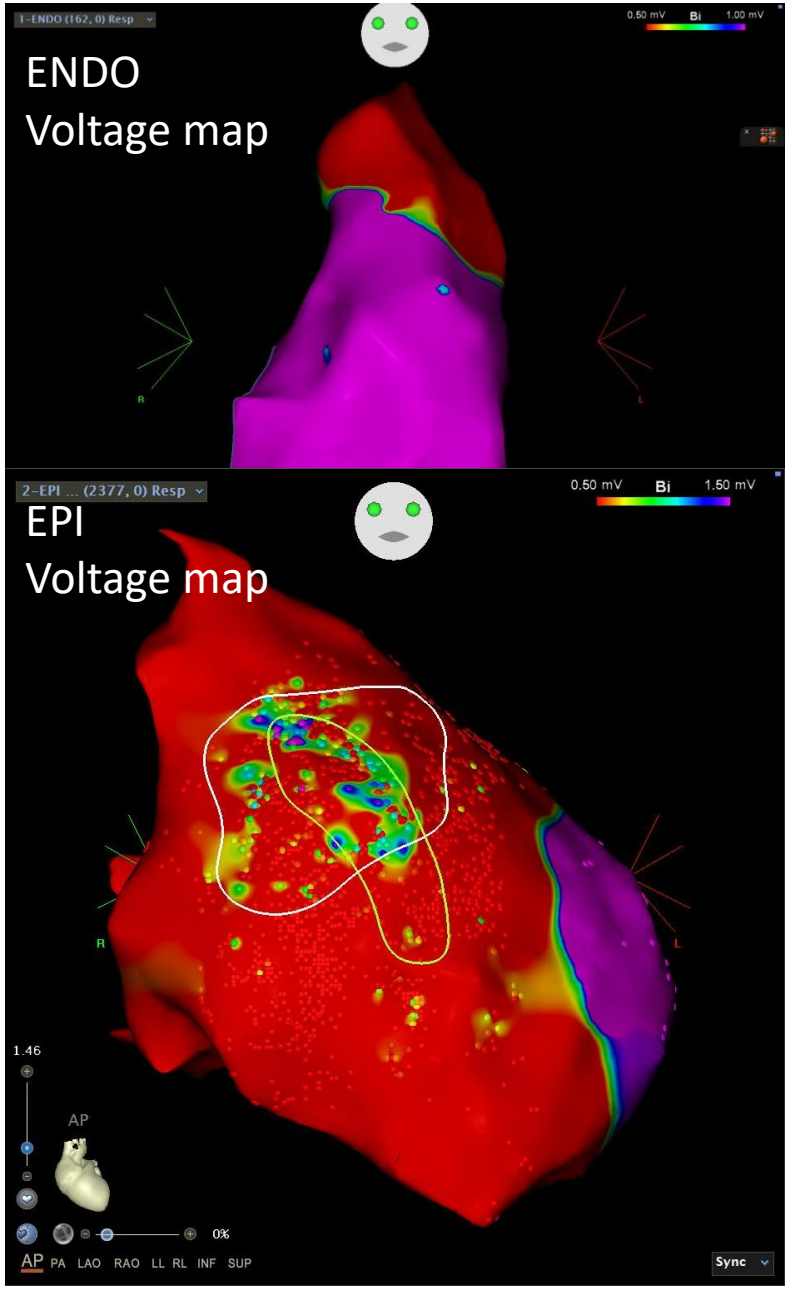
MAPPING DETAILS

Ablation date 07-09-2021

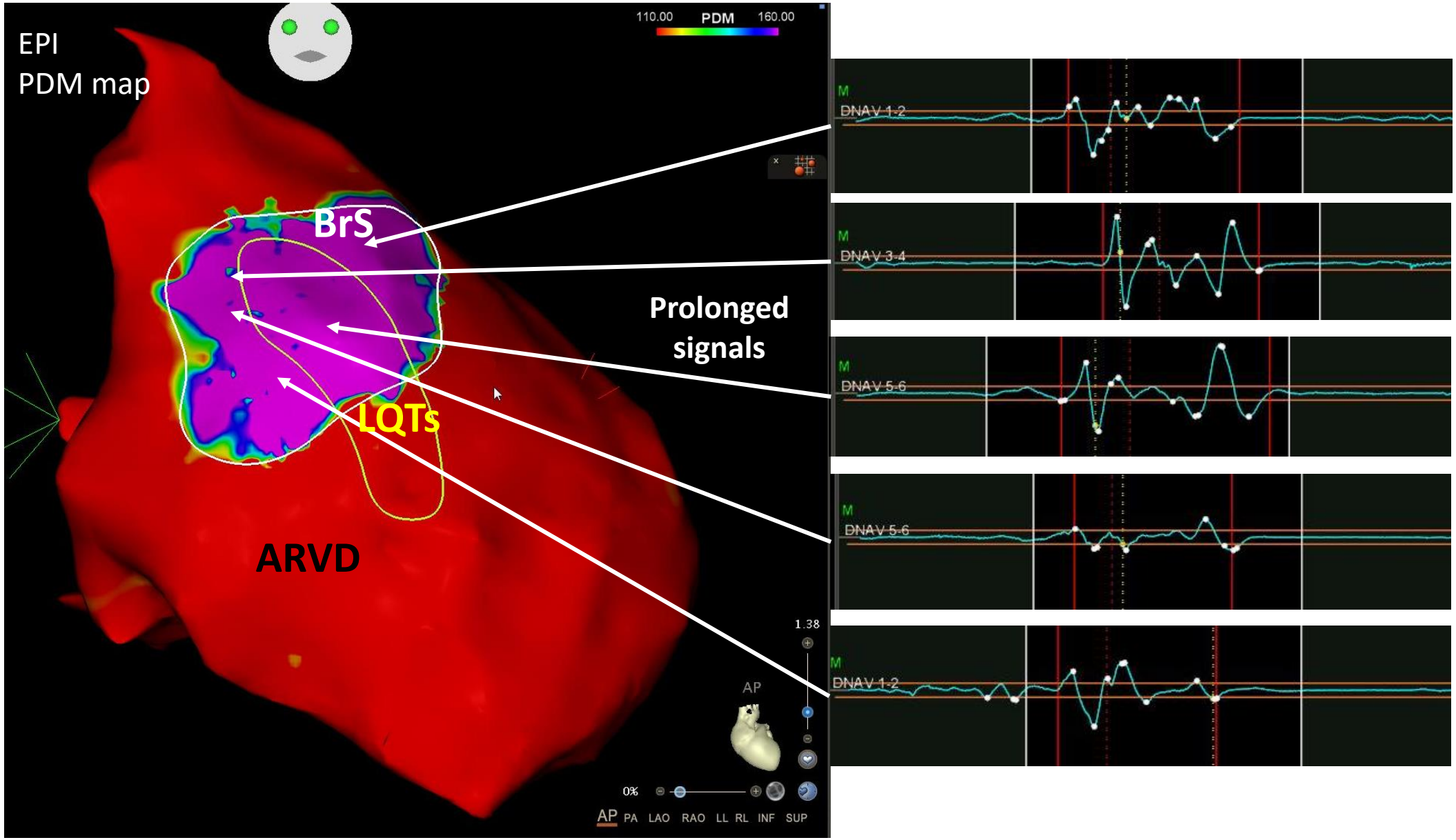
DecaNAV substrate mapping catheter

Endocardial mapping - Healthy RV

Epicardial mapping – Extended low voltage area related to ARVD

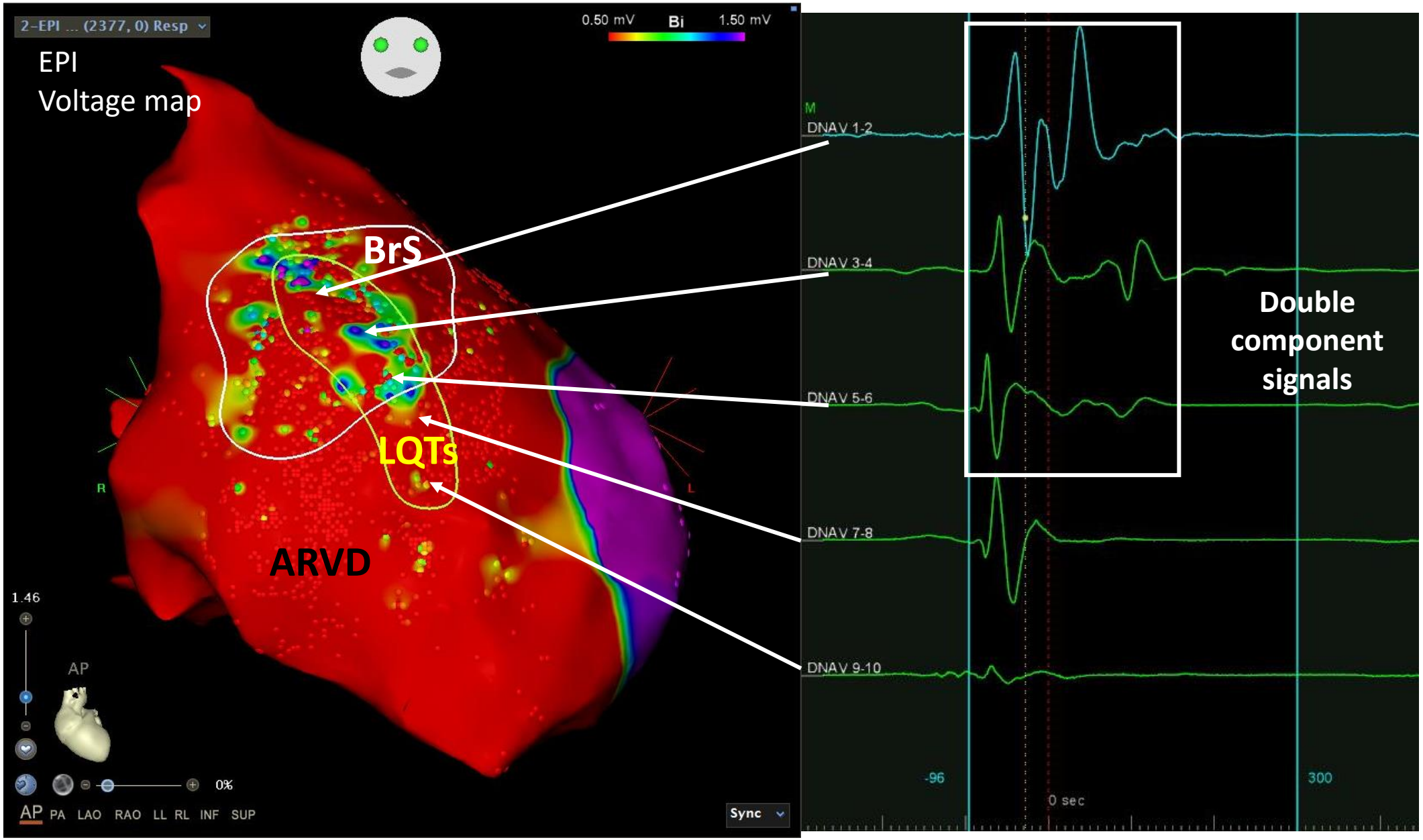


SIGNAL ANALYSIS – Brugada area



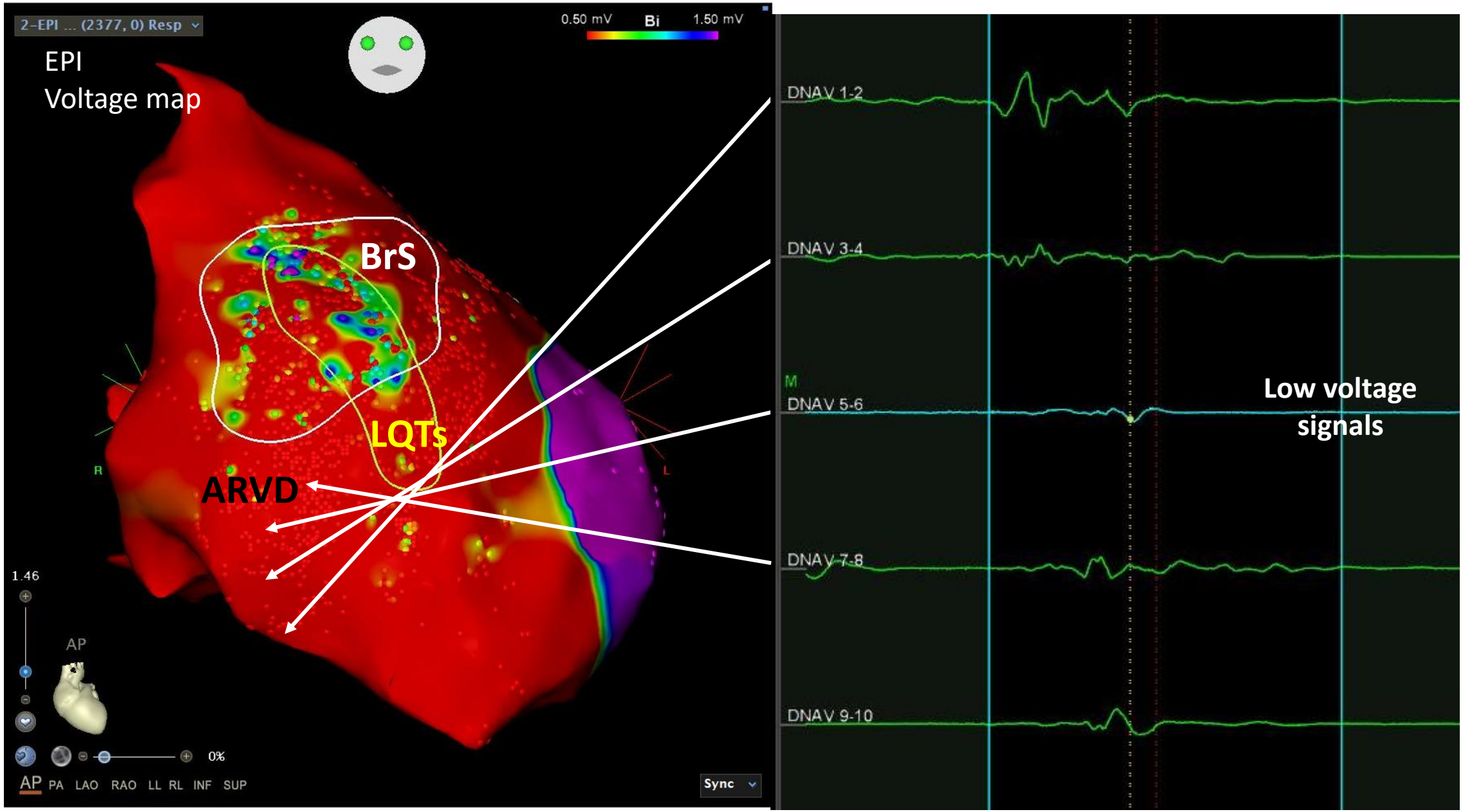
Brugada + ARVD + LQTs – P.G. 2021

SIGNAL ANALYSIS – LQTs area



Brugada + ARVD + LQTs – P.G. 2021

SIGNAL ANALYSIS – ARVD



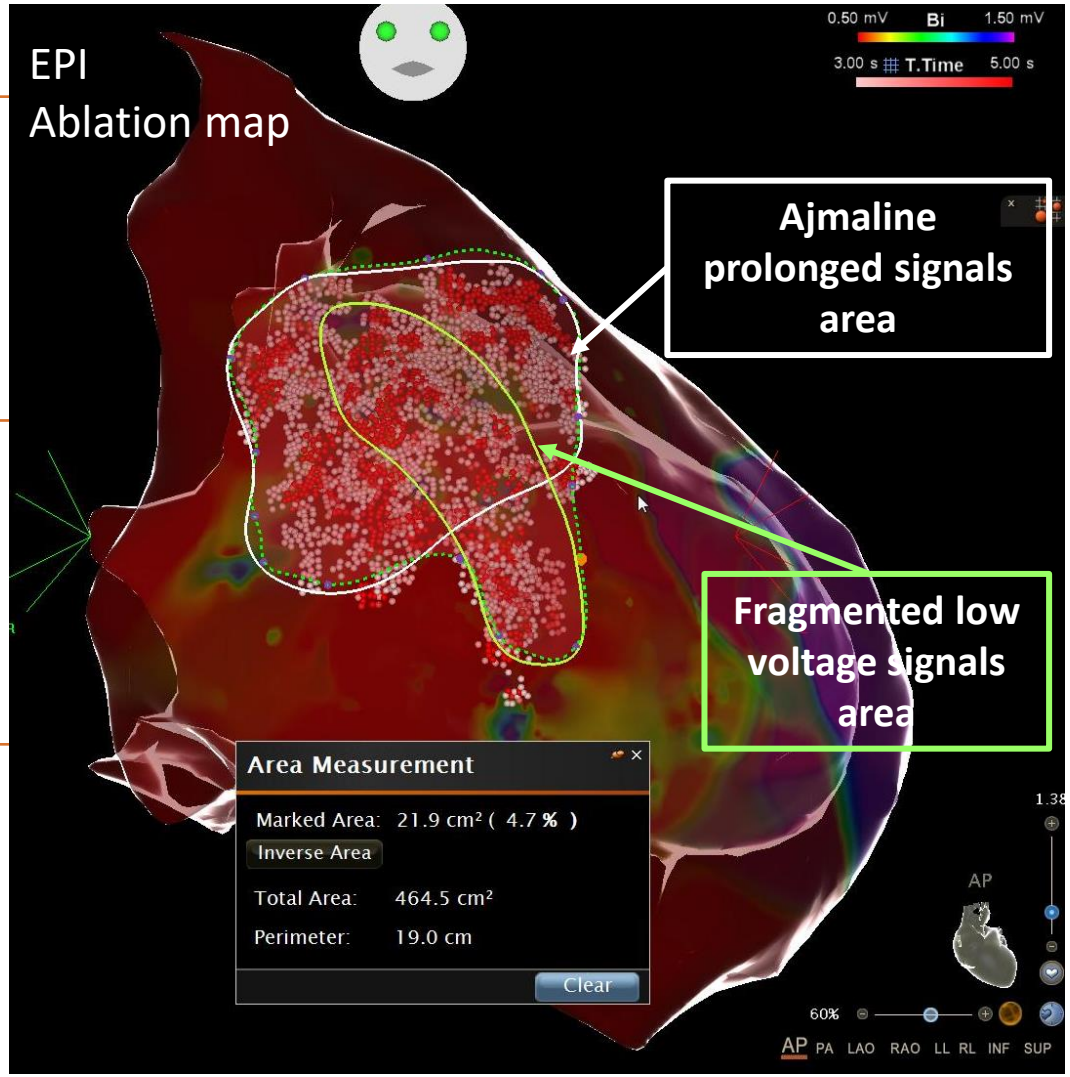
Brugada + ARVD + LQTs – P.G. 2021

ABLATION STRATEGY

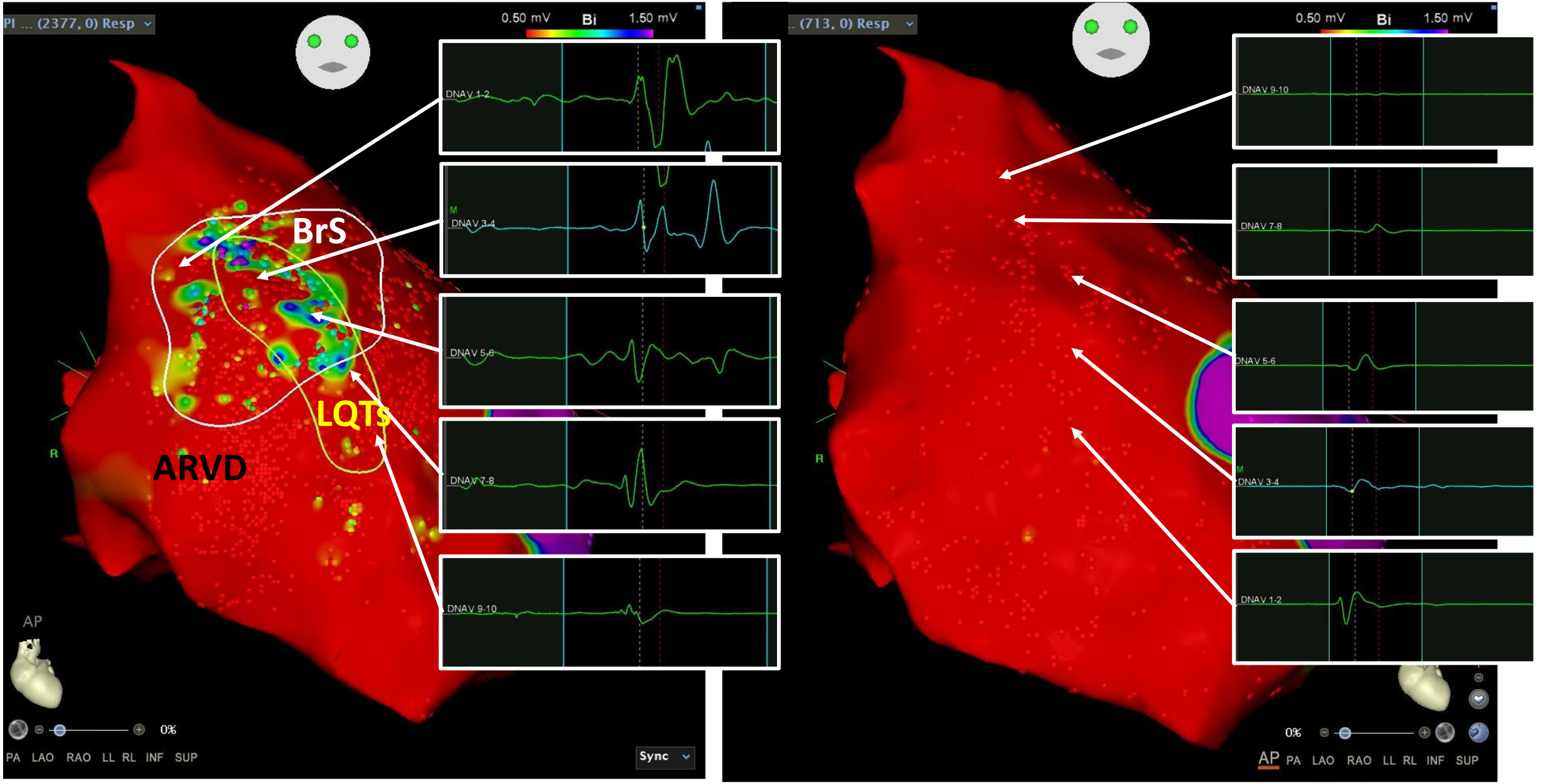
Homogenization of the area (fragmented low potentials and prolonged signals area) with SmartTouch SF catheter

40-45 watts with fast dragging technique, to reach superficial lesion

37 minutes of total ablation for a 21.9 cm^2 area



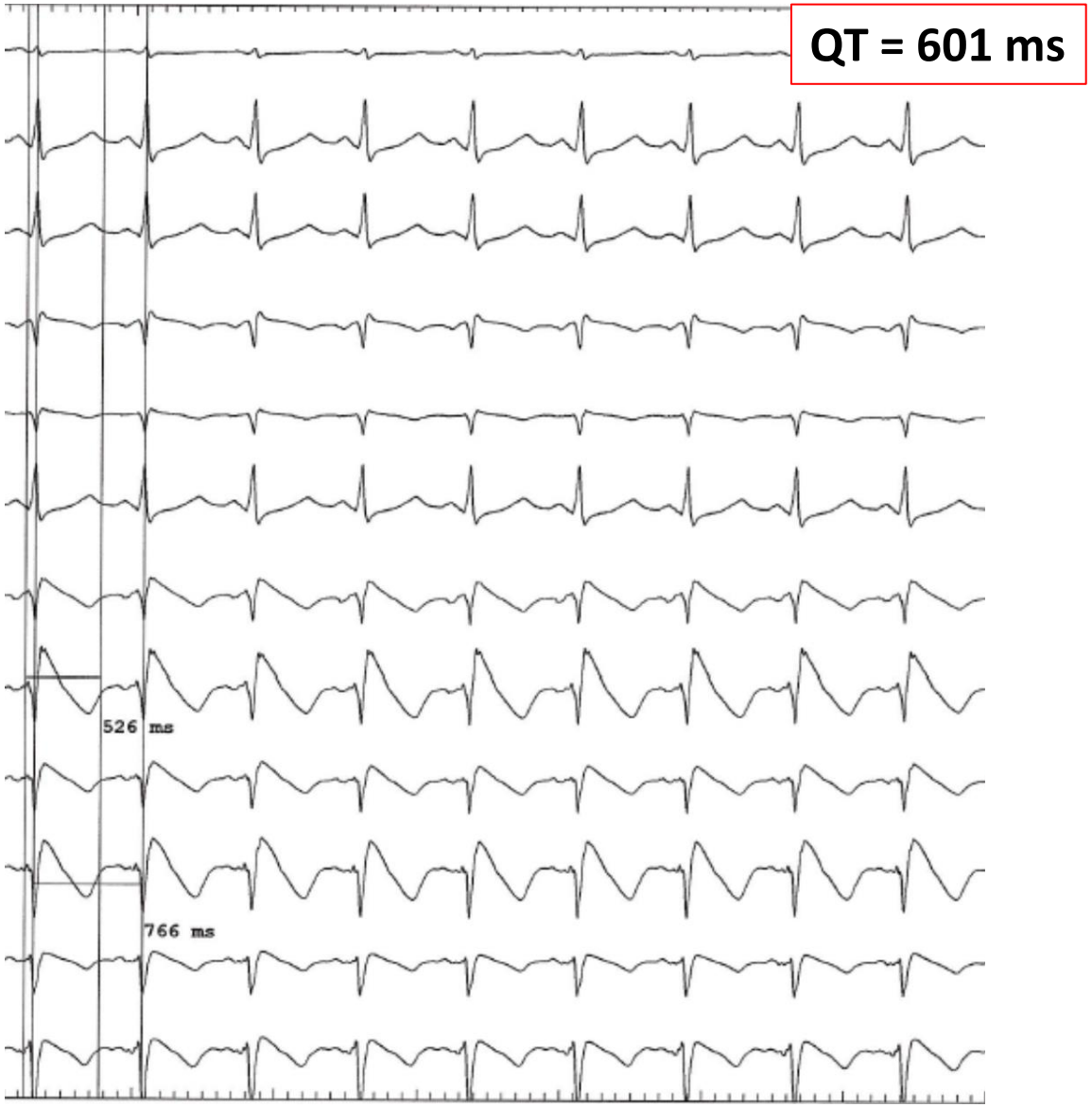
SIGNAL COMPARISON in the target area: PRE AND POST ABLATION



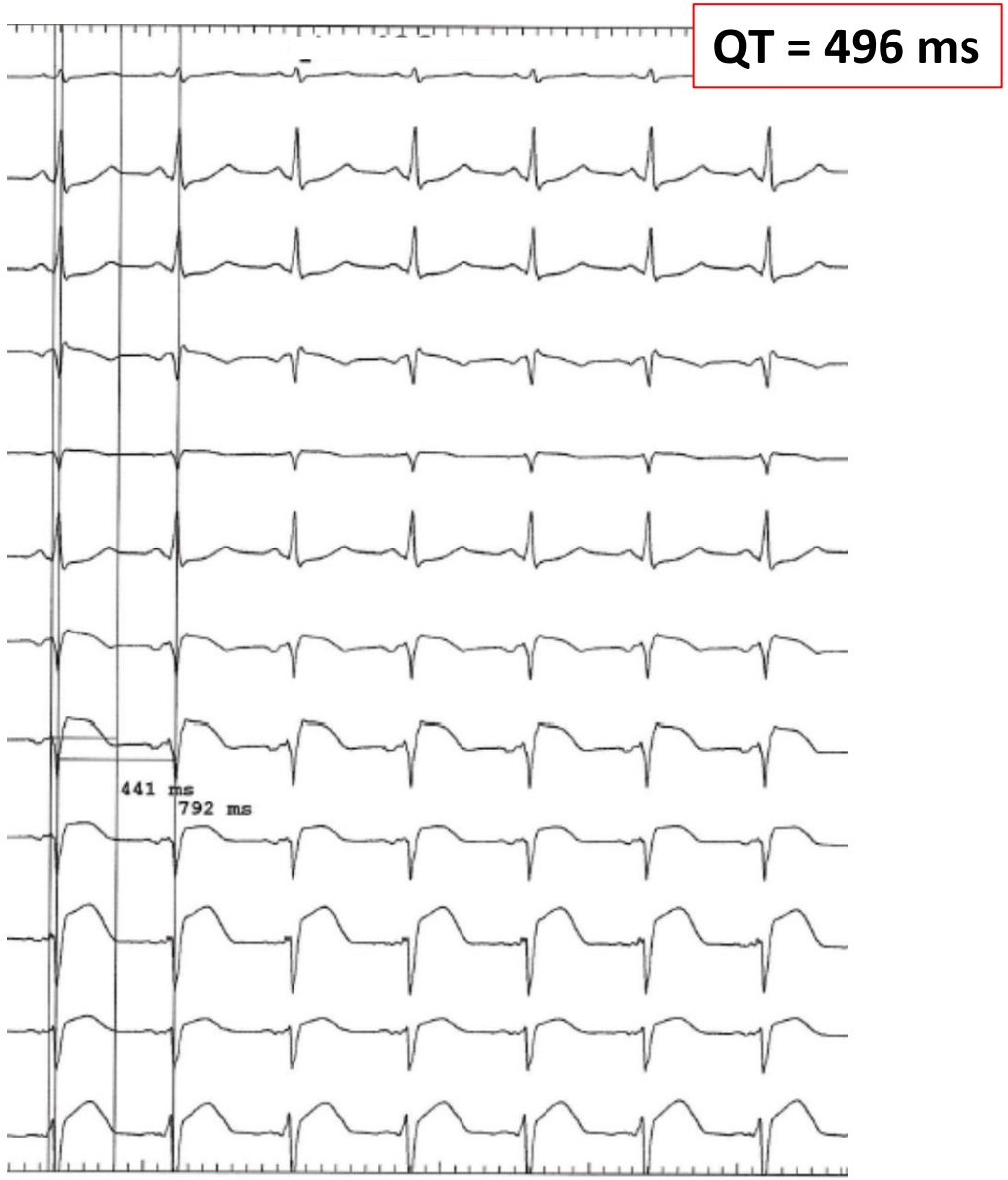
Brugada + ARVD + LQTs – P.G. 2021

FINAL ECG

BASELINE ECG



ECG AFTER ABLATION

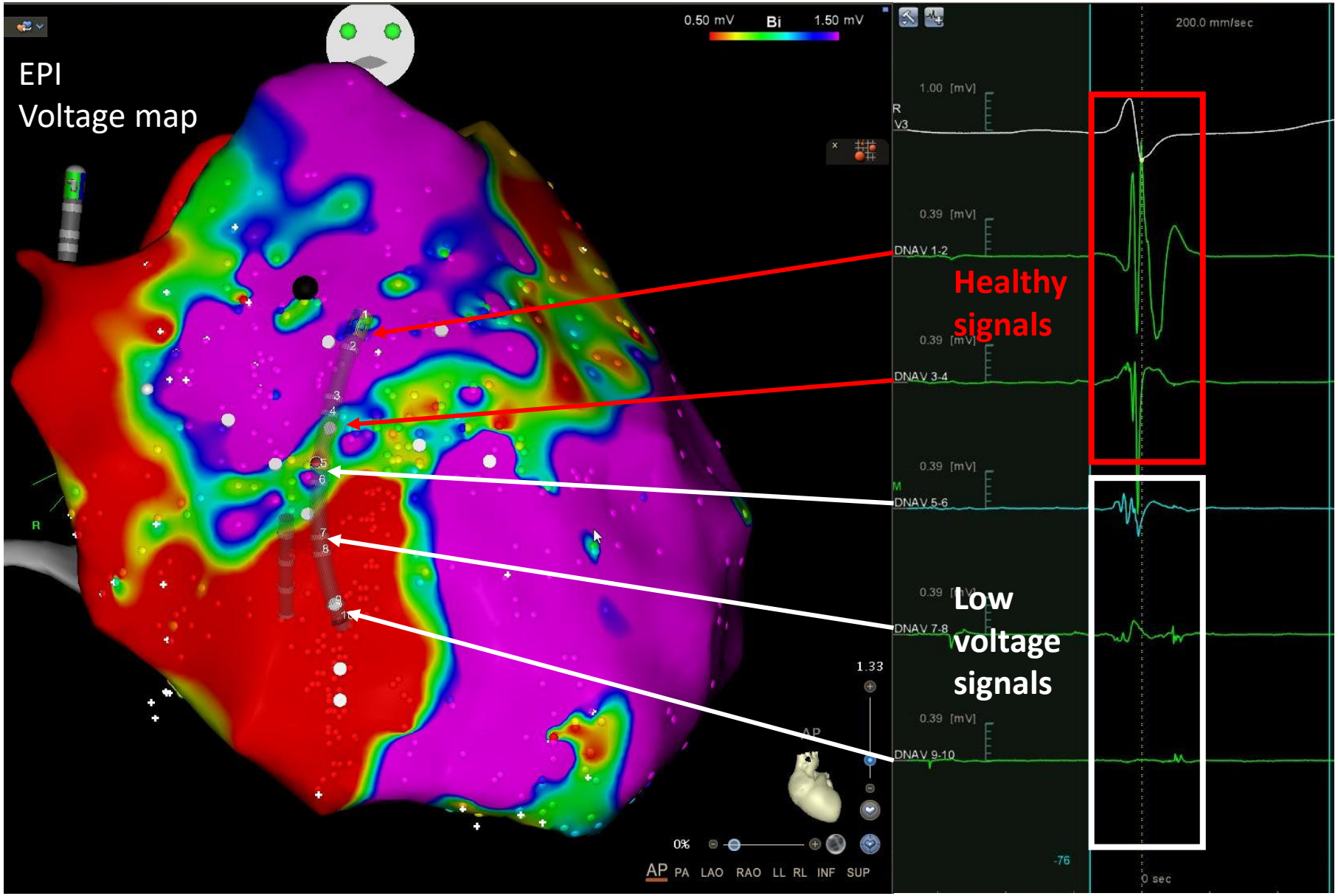


Arrhythmogenic RV Cardiomiopathy

SUMMARY

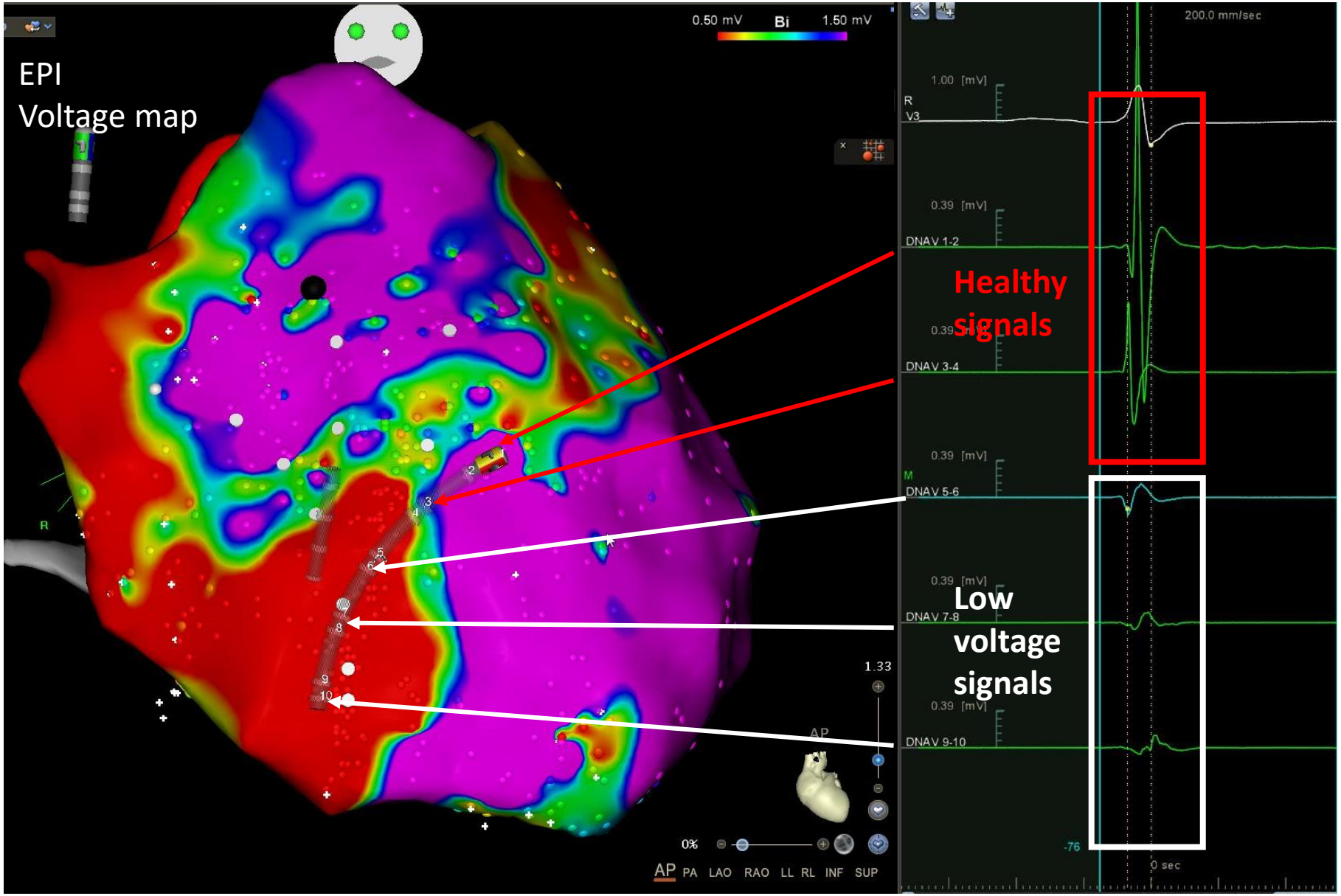
- Female 50 YO
- 2011: ARVD diagnosis and ICD implantation
- 2013: atrial flutter ablation
- 2017: two endo-epi VT ablations. After that, no more VT episodes
- 2021: VF episode (admission reason)
- Fast VT episodes shocked by ICD
- 15% burden of two PVC morphologies, both anteroapical RV
- Firstly, endocardial ablation with acute PVC elimination
- After few months new episodes and ER admission
- New ablation with epicardial approach in the same region

SIGNAL ANALYSIS – ARVD AREA



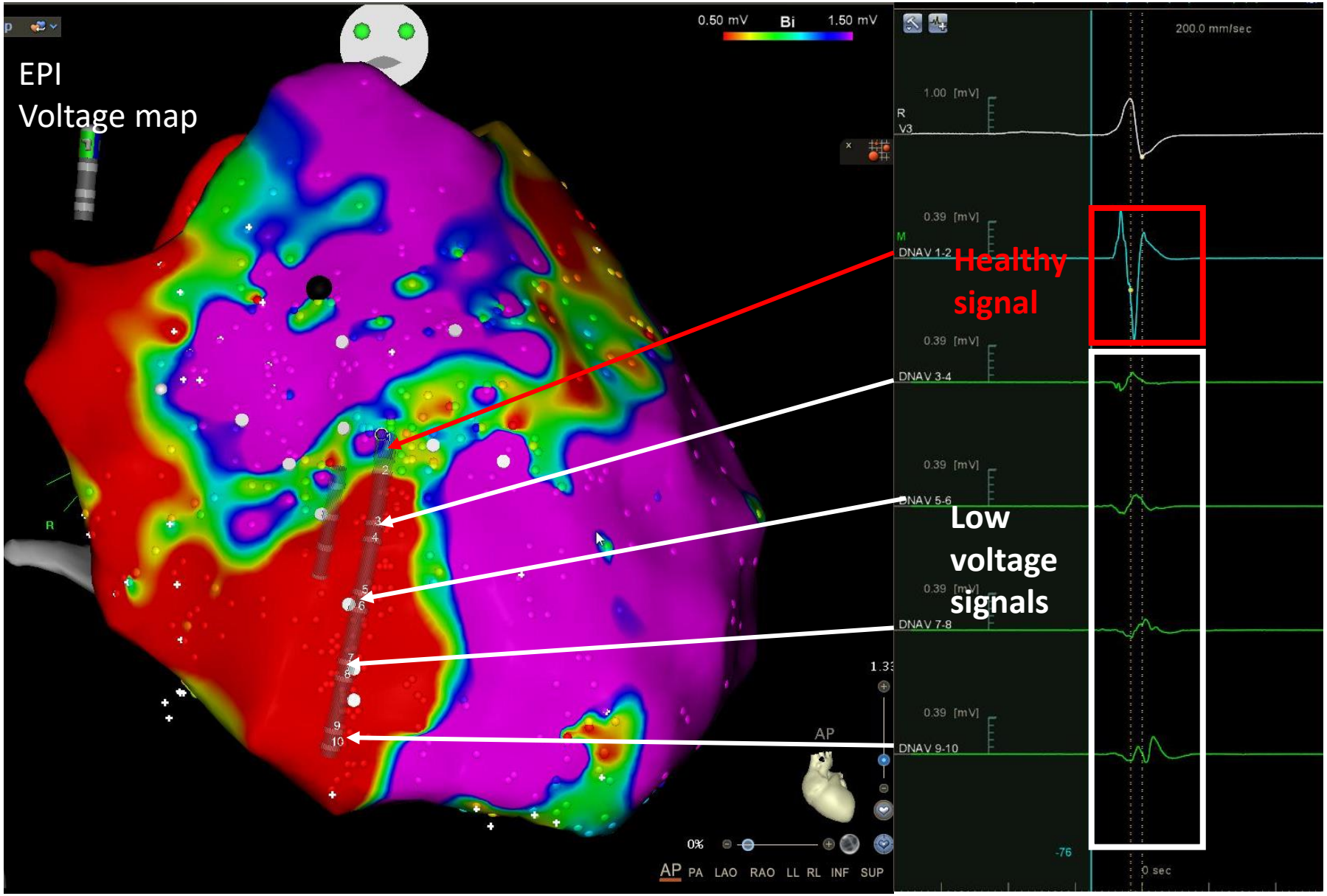
ARVD triggered – D'A.A. 2017

SIGNAL ANALYSIS – ARVD AREA



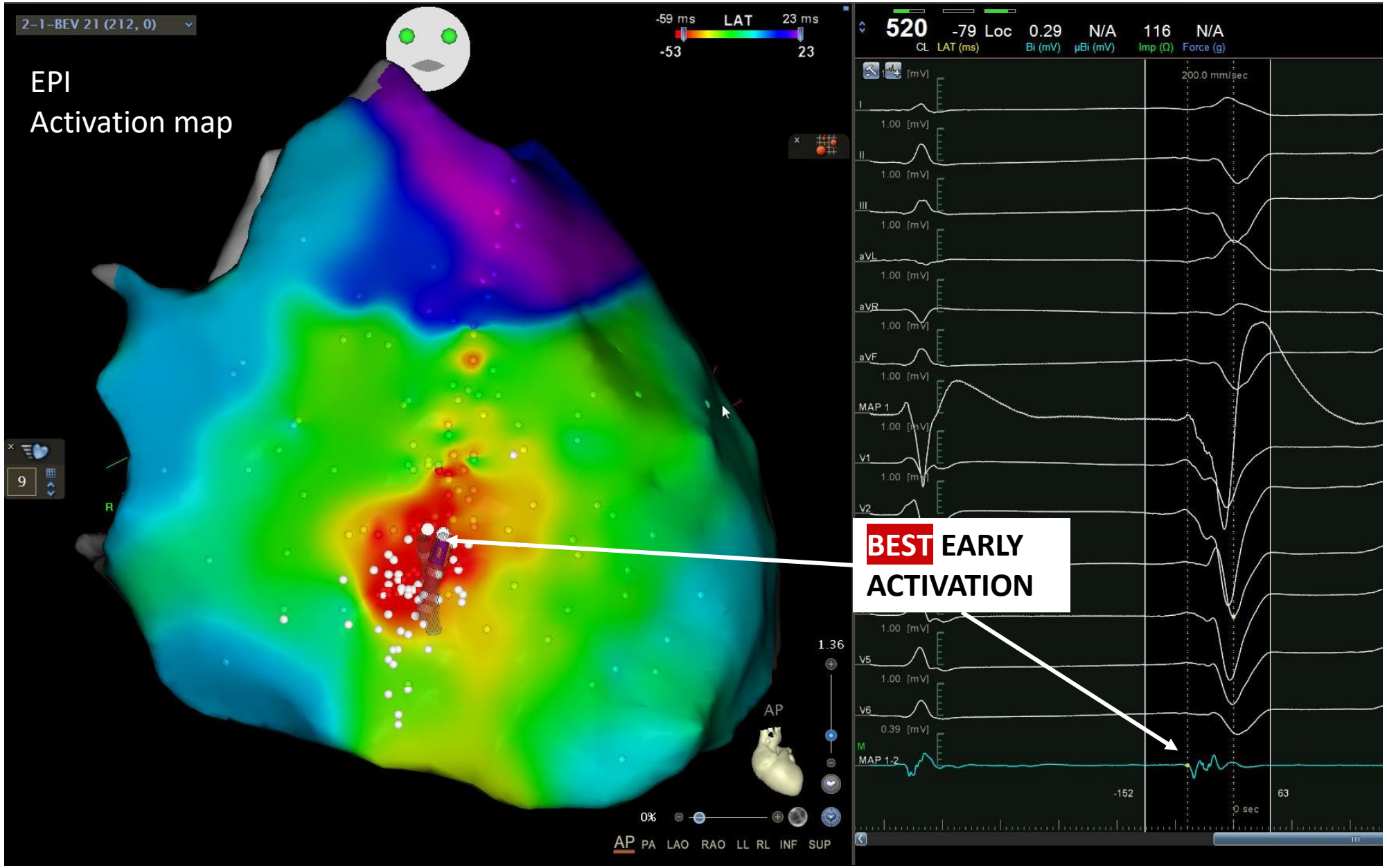
ARVD triggered – D'A.A. 2017

SIGNAL ANALYSIS – ARVD AREA



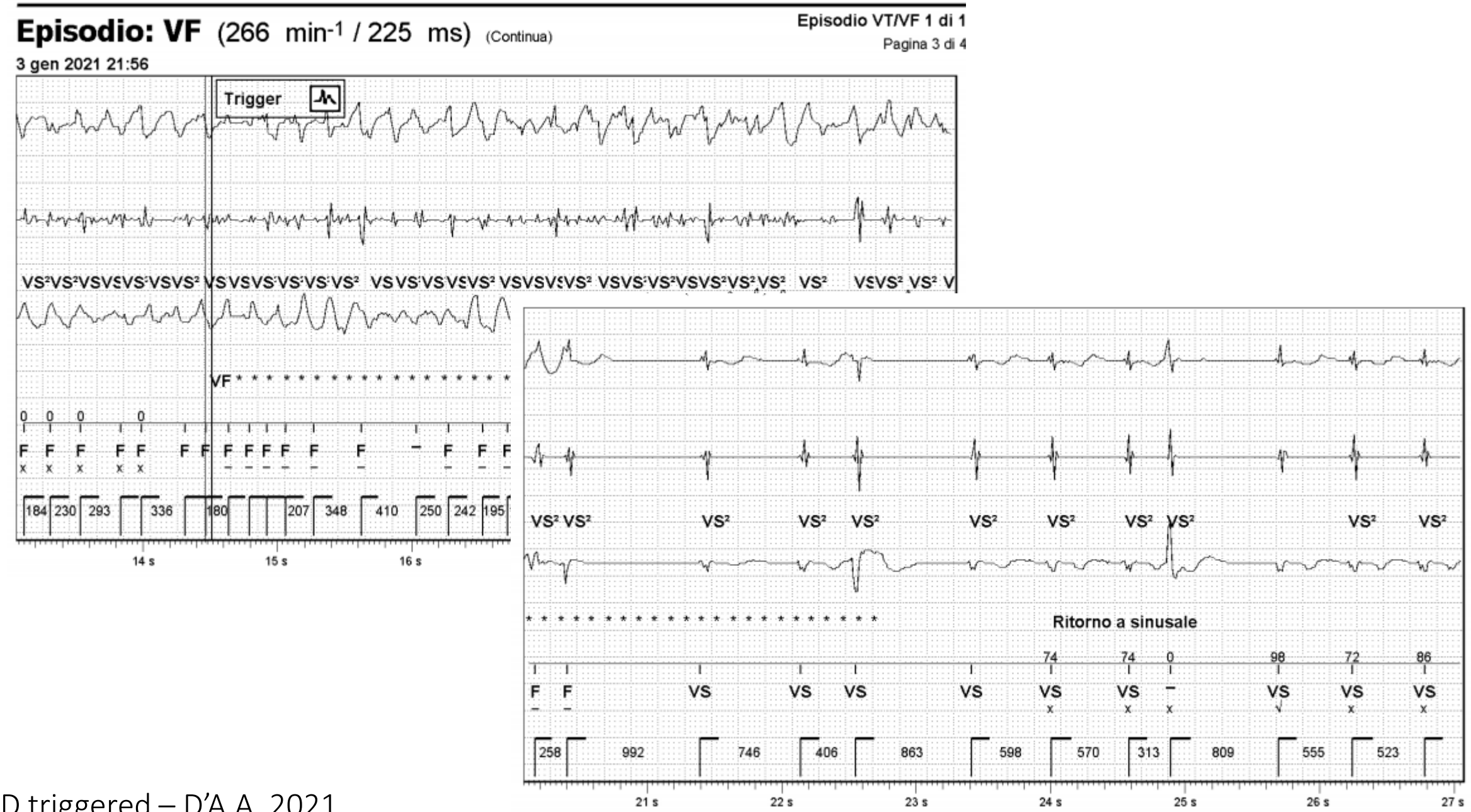
ARVD triggered – D'A.A. 2017

SIGNAL ANALYSIS – BEST PRECOCITY



ARVD triggered – D'A.A. 2017

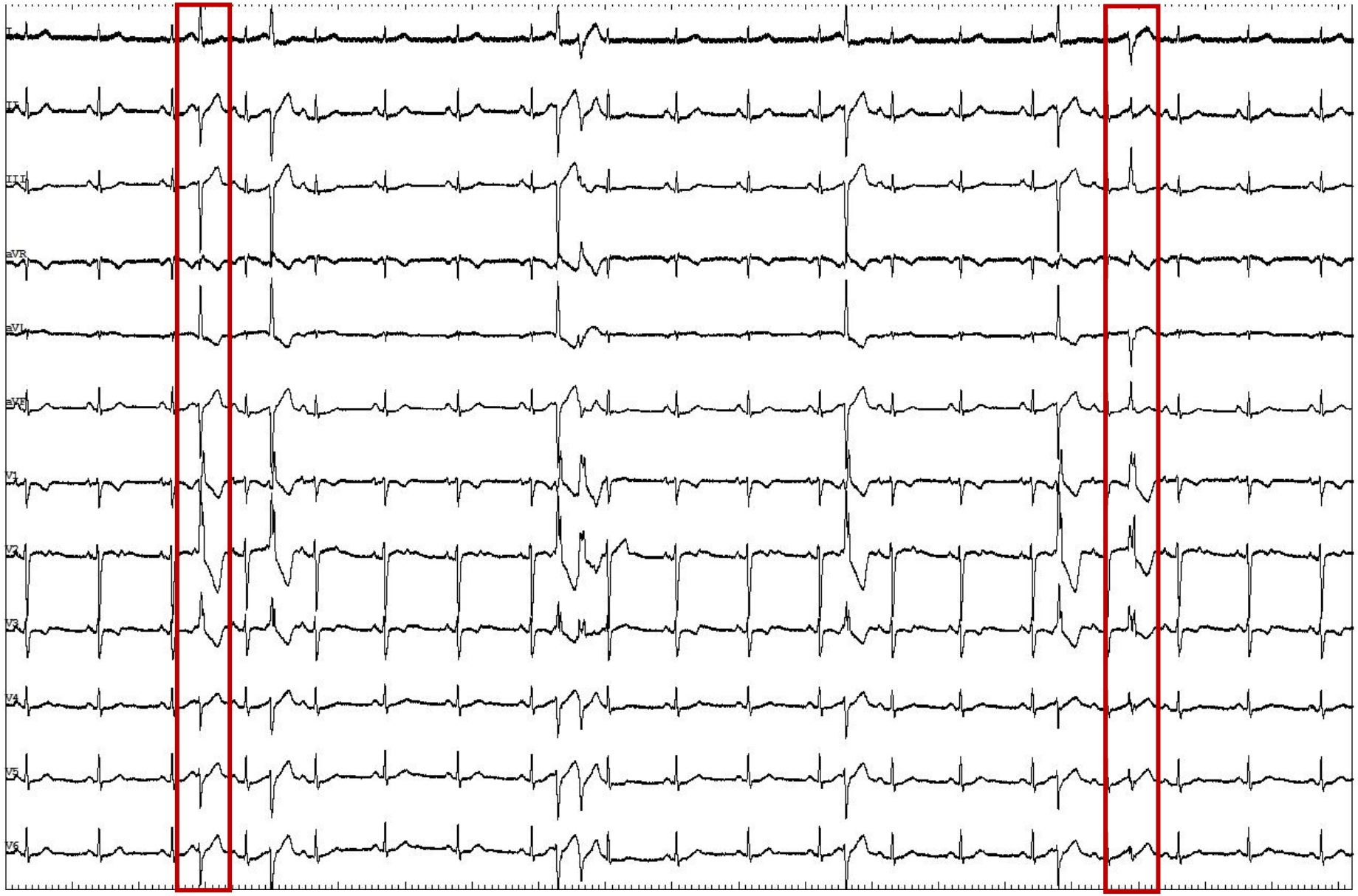
VF EPISODE (ICD registration, 2020)



ARVD triggered – D'A.A. 2021

INITIAL ECG: frequent PVCs (2 morphologies)

Sharp QRS Suggesting an origin from the conduction system



ARVD triggered – D'A.A. 2021

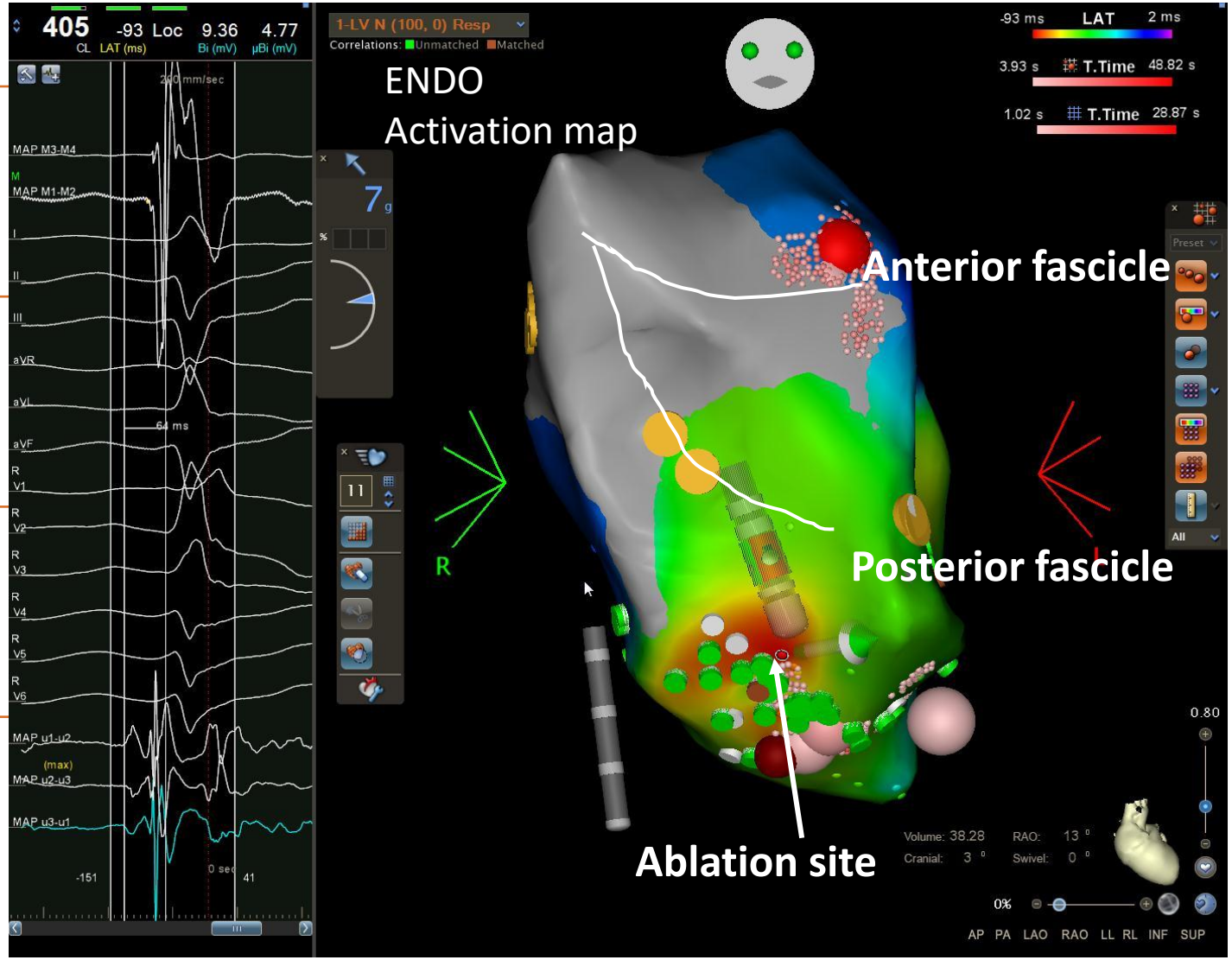
ABLATION STRATEGY (Postero-medial papillary muscle)

Firstly, posterior fascicle PVC was targeted

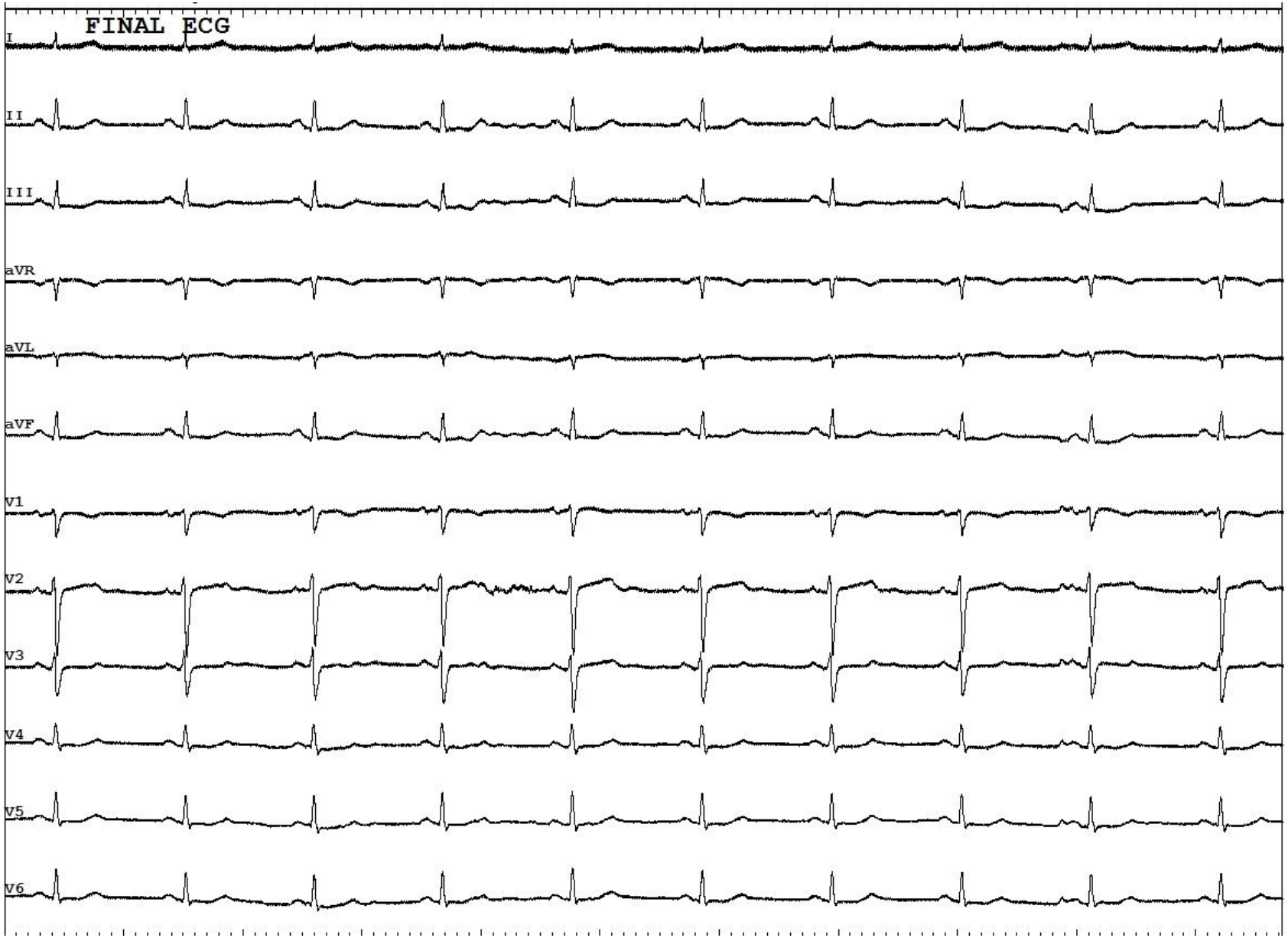
Identification of earliest activation signal during PVC in the area of postero-medial papillary muscle

5 minutes of RF application with different orientation around the insertion of papillary muscle

Elimination of first PVC morphology



FINAL ECG



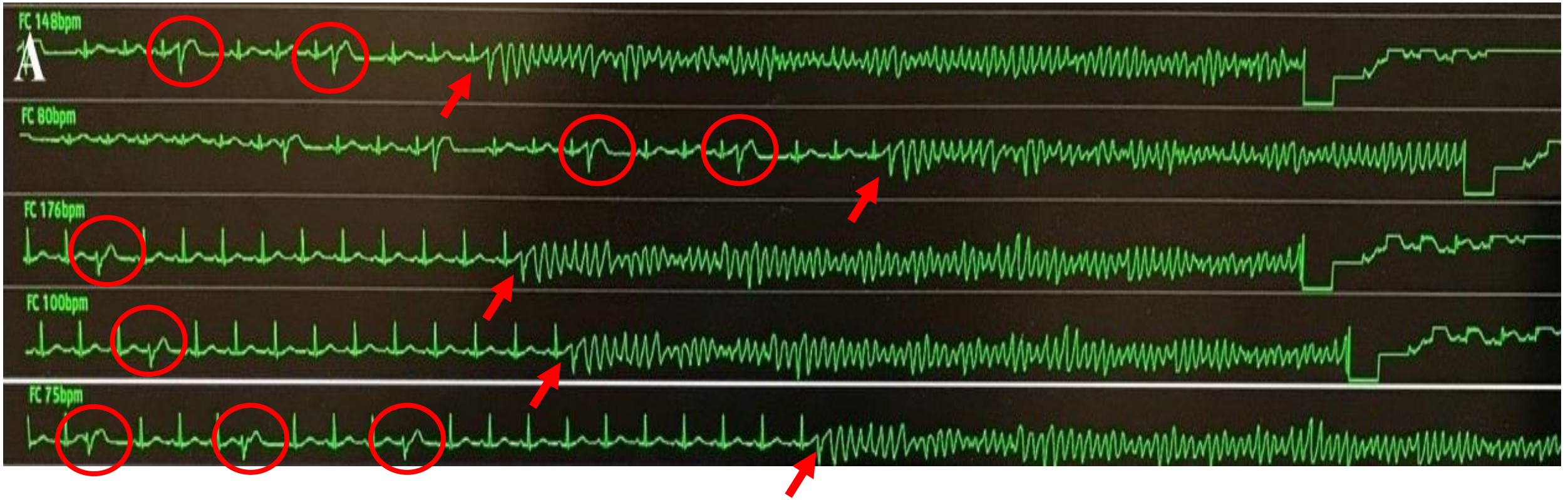
ARVD triggered – D'A.A. 2021

Peripartum cardiomyopathy

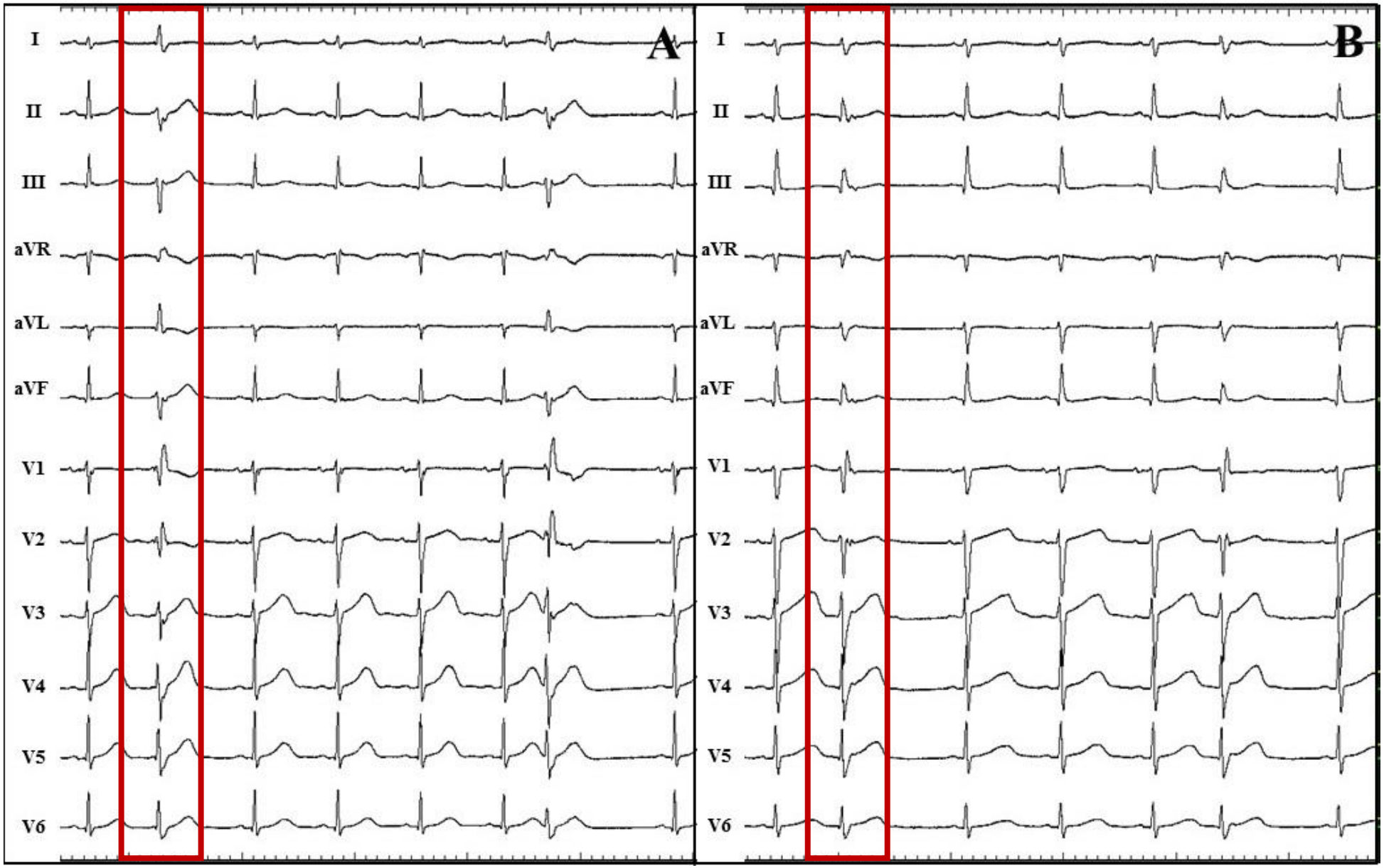
SUMMARY

- Female 30 YO
- No family histyory of SCD
- No signs of BrS, ER or LQTs
- Resuscitated cardiac arrest while resting
- 3 months after pregnancy
- No AADs before the event

VF EPISODES (ICU monitoring)



INITIAL ECG: 2 PVC morphologies



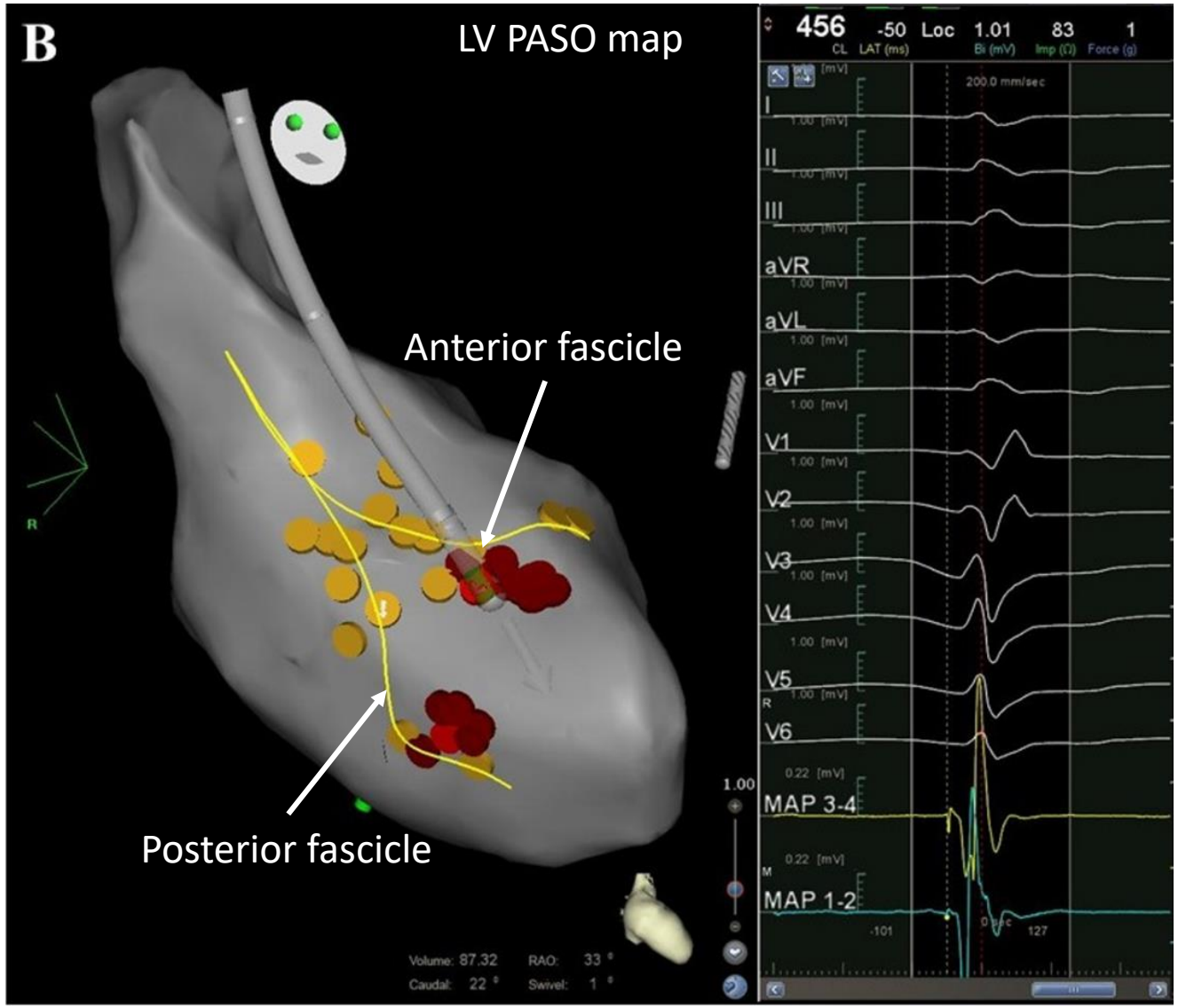
ABLATION STRATEGY (anterior fascicle PVC)

Secondly, anterior fascicle PVC was targeted

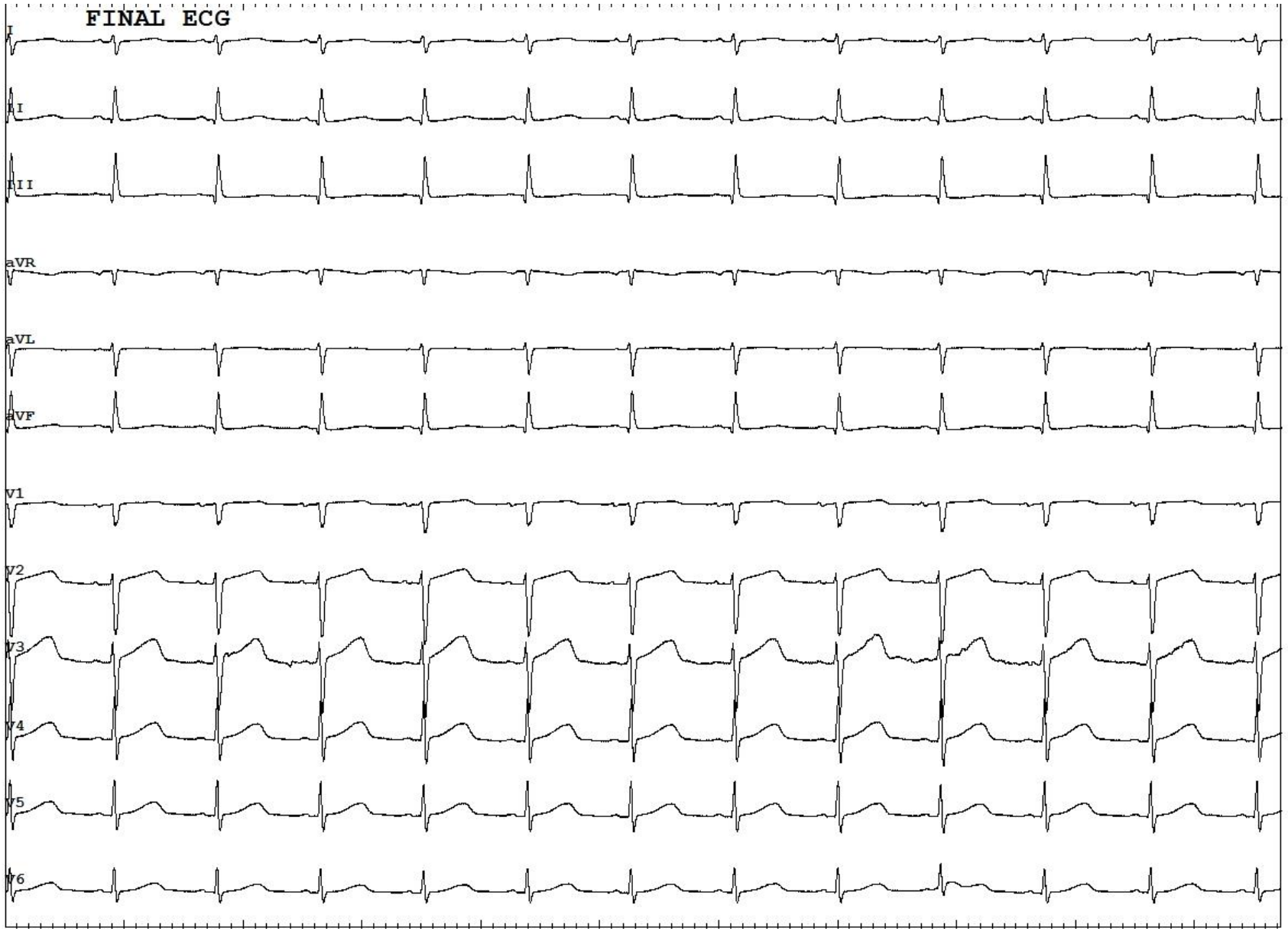
Identification of the best signal precocity during PVC in the area of the proximal anterior fascicle

4 minutes of RF application

Elimination of the second PVC morphology



FINAL ECG



PPCM – C.K. 2018

Katia, 50 arresti cardiaci in 6 giorni: la storia (a lieto fine) del suo cuore impazzito

di Ettore Mautone

Era impazzito all'improvviso, e senza una ragione, il cuore di Katia, una ragazza di 29 anni originaria di Napoli, residente a Roma, in vacanza a San Giorgio a Cremano a Pasqua. Prima soccorsa da sua madre a casa, (che per una coincidenza pochi giorni prima aveva seguito un corso di rianimazione), poi ricoverata all'ospedale Boscotrecase in pronto soccorso, quindi curata in rianimazione all'ospedale del mare. Infine trasferita al policlinico San Donato di Milano, dopo un volo organizzato nei minimi dettagli dalla rete dei soccorsi campani, per raggiungere uno dei pochi centri specializzati in Italia in elettrofisiologia. Ad attenderla un altro medico campano, Carlo Pappone, beneventano, tra i maggiori esperti al mondo nel fronteggiare il cuore matto di Katia.

Right Ventricle & Conduction System: the essence of life

- In this patient population, regions localized in the epicardium of the right ventricle harbor structural electrophysiological abnormalities.

- Catheter ablation of these areas resulted in successful suppression of malignant arrhythmias potentially leading to sudden cardiac death, thus supporting their arrhythmogenic role.

- These findings provide a novel approach for the management of cardiomyopathies patients requiring a potential definite and pathophysiological treatment.

«The *language* of signals and the shape of substrates are the expressions of the disease. Their abolition save the life of these patients»