Arrhythmias in the adult with repaired congenital heart disease

Raja Selvaraj, JIPMER

Introduction

Adults with repaired CHD

- CHD - 1% of population
- Many undergo correction
- With improved care, many of these patients grow up to be adults
- Arrhythmias are an important morbidity
- Complex interplay between arrhythmias and hemodynamics

Mechanisms

- Underlying heart disease
- Chamber enlargement / pressure overload
- Fibrosis - sutures / patches
- Trauma to specialised conduction system

Topics

- Bradyarrhythmias
- Tachyarrhythmias
- Clinical evaluation
- Evaluation - Imaging / ECG
- Pacemaker for brady-arrhythmias
- Medical management of tachyarrhythmias
- Ablation for tachyarrhythmias
- Common arrhythmias (AVNRT, AP)
- Risk stratification

Cover

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Management

Hemodynamics correction

- Residual shunts
- Tricuspid regurgitation
- Pulmonary regurgitation
- Outflow tract obstruction

Drugs

- Class Ic (Flecainide, Propafenone)
- Class III (Amio, Dofetilide, Sotalol)
- Proarrhythmia / systemic problems

Ablation

- Relatively less efficacious
- But still generally safe, can be very beneficial

Atrial Tachyarrhythmias

- Macroreentrant Atrial tachycardia
  - Typical flutter most common
  - Atriotomy related
  - septal rare
  - may be combination
- Focal atrial tachycardia

Flutter Circuits

OBJECTIVES The purpose of this study was to compare atrial tachycardia circuits after a range of cardiac operations.

BACKGROUND Knowledge of circuits occurring in a given postsurgical substrate should help to ablate these challenging tachycardias and develop potential preventive strategies.

METHODS We analyzed tachycardia circuits in 83 consecutive patients (60 males; median age 47 years, range 9–73) after atrial incisions undergoing ablation of atrial tachycardias. A combined strategy of electroanatomic (CARTO) and entrainment mapping was used. Fifty-two patients (63%) underwent operation for congenital and 31 (37%) for acquired heart disease. Patients were divided into subgroups based on the intervention performed in the atria: right lateral atriotomy (39 patients), left atrial (11) and superior transseptal (10) approach to the mitral valve, biatrial heart transplantation (8), Mustard (8) and Fontan (4) procedure, and other interventions (3).

RESULTS Most of the 119 tachycardias mapped were isthmus-dependent atrial flutter (66) and incisional tachycardia (30). Isthmus-dependent atrial flutter was the most frequent arrhythmia in all subgroups except for Fontan patients, in whom incisional tachycardia was most frequent. The distribution of tachycardia circuits did not differ significantly among groups.

CONCLUSIONS The observed circuits did not differ among the postsurgical substrates. Isthmus-dependent atrial flutter should be the first circuit considered in patients after atrial incisions.

KEYWORDS Atrial flutter; Atrial tachycardia; Catheter ablation; Reentry

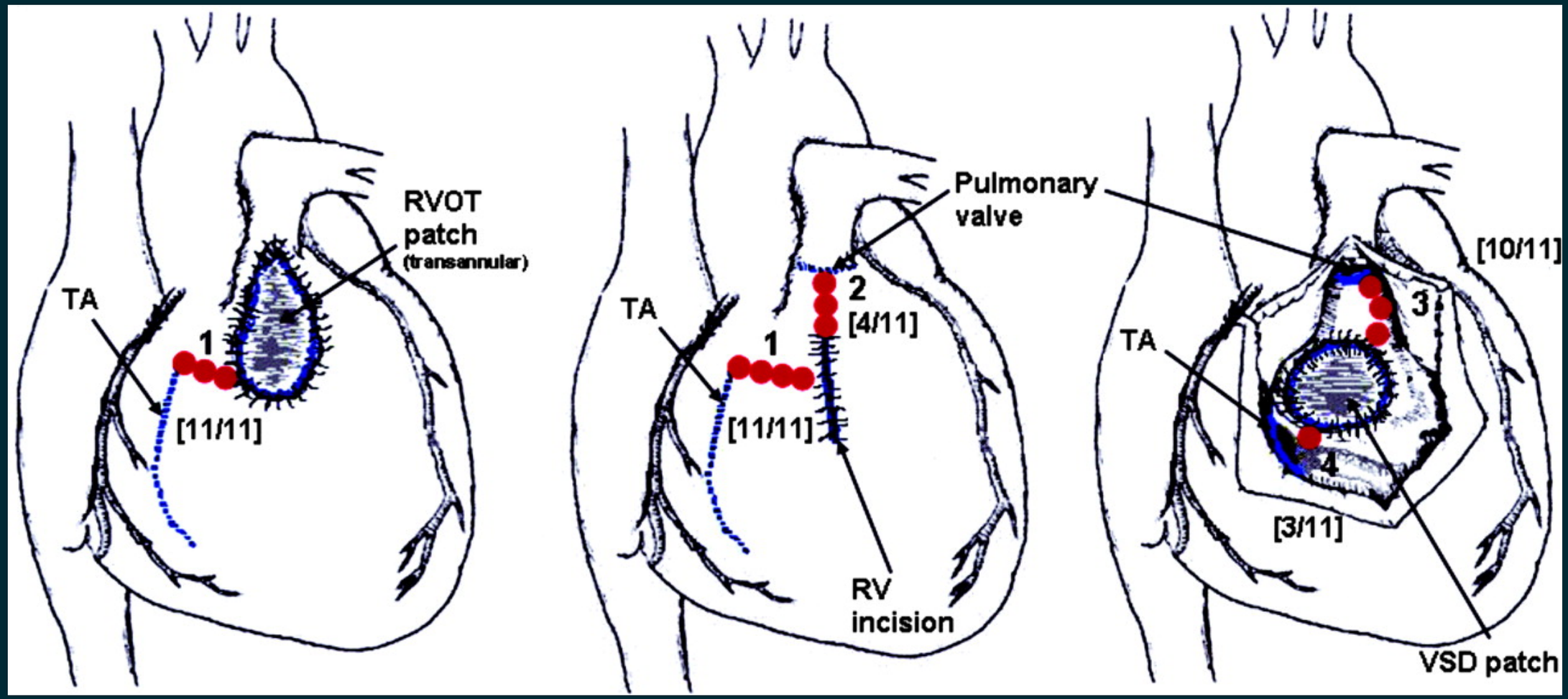
(Heart Rhythm 2005;2:64–72)

Peter Lukac et al. Ablation of atrial tachycardia after surgery for congenital and acquired heart disease using an electroanatomic mapping system: Which circuits to expect in which substrate?. Heart Rhythm 2005;2(1):64-72

Ventricular arrhythmias

- Monomorphic Ventricular tachycardia
  - Macroreentry around ventriculotomy / patches
- Polymorphic Ventricular tachycardia
  - LV outflow tract obstruction
  - d TGA with atrial switch (systemic RV)
  - TOF, poor RV function

Macroreentrant VT in repaired TOF



Zeppenfeld K, Schalij MJ, Bartelings MM, Tedrow UB, Koplan BA, Soejima K, Stevenson WG. Catheter ablation of ventricular tachycardia after repair of congenital heart disease: electroanatomic identification of the critical right ventricular isthmus. Circulation. 2007 Nov 13;116(20):2241-52

Substrate

- ASD
- DORV / TOF
- Ebsteins anomaly
- Mustard / Senning
- Fontan
  - almost 50% of classic fontan have arrhythmias
  - modern approaches (lateral tunnel / extracardiac) - less arrhythmias, more difficult access

Ablation

- Why
- When
- Who should do
- How

Ablation - Why ?

- Arrhythmias often poorly tolerated
- Rapid VR during flutter because of young age / slower flutters
- Poor response to drugs
- Drug therapy not preferred
  - Long term side effects
  - Proarrhythmia
  - Negative inotropy

Ablation - When

Recommendations	Consensus statement	References
In patients with CHD and AT/AF rhythm control is recommended as the preferred initial strategy.		Expert consensus
Catheter ablation is recommended as first line therapy and preferred to long-term pharmacological treatment, in case of amenable, circumscribed substrates.		Expert consensus

ESC Scientific Document Group; Arrhythmias in congenital heart disease: a position paper of the European Heart Rhythm Association (EHRA), Association for European Paediatric and Congenital Cardiology (AEPC), and the European Society of Cardiology (ESC) Working Group on Grown-up Congenital heart disease, endorsed by HRS, PACES, APHRS, and SOLAECE, EP Europace, Mar 2018, https://doi.org/10.1093/europace/eux380

Ablation - Who

- Pediatric Electrophysiologist
- ACHD Electrophysiologist
- Electrophysiologist with support of pediatric Cardiologist

Ablation - How

- Understand anatomy
  - CHD
  - Repair
- Identify conduction barriers
- Identify circuits
- Linear ablation (barriers)
- Confirming block

Procedure planning

- Surgery notes
- Imaging
- Residual shunts

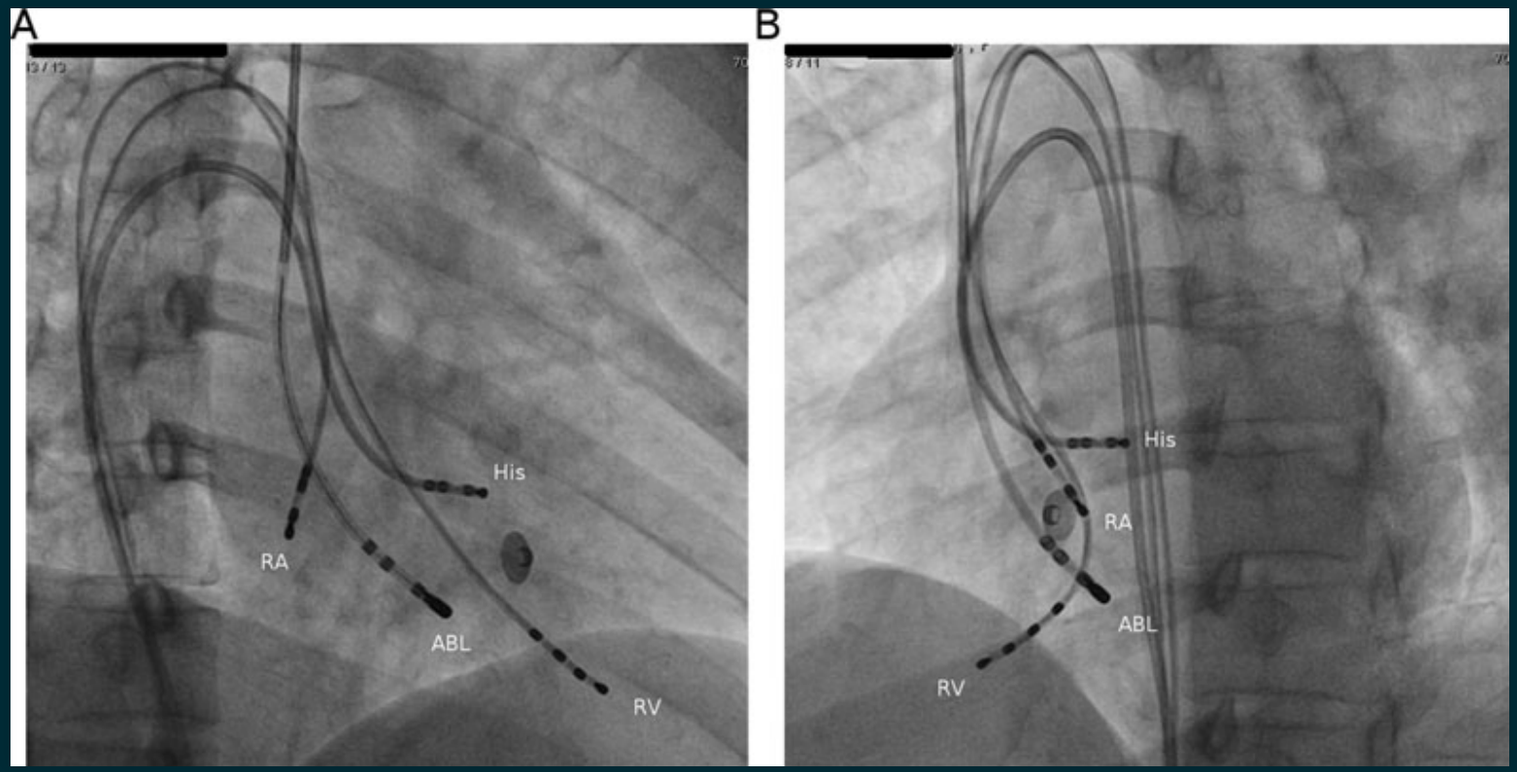
Access issues

- Difficult transseptal
  - CHD surgery
  - ASD device
- Baffle puncture in Mustard / Senning for CTI line
- Magnetic navigation may allow retrograde route

Access - Venous access

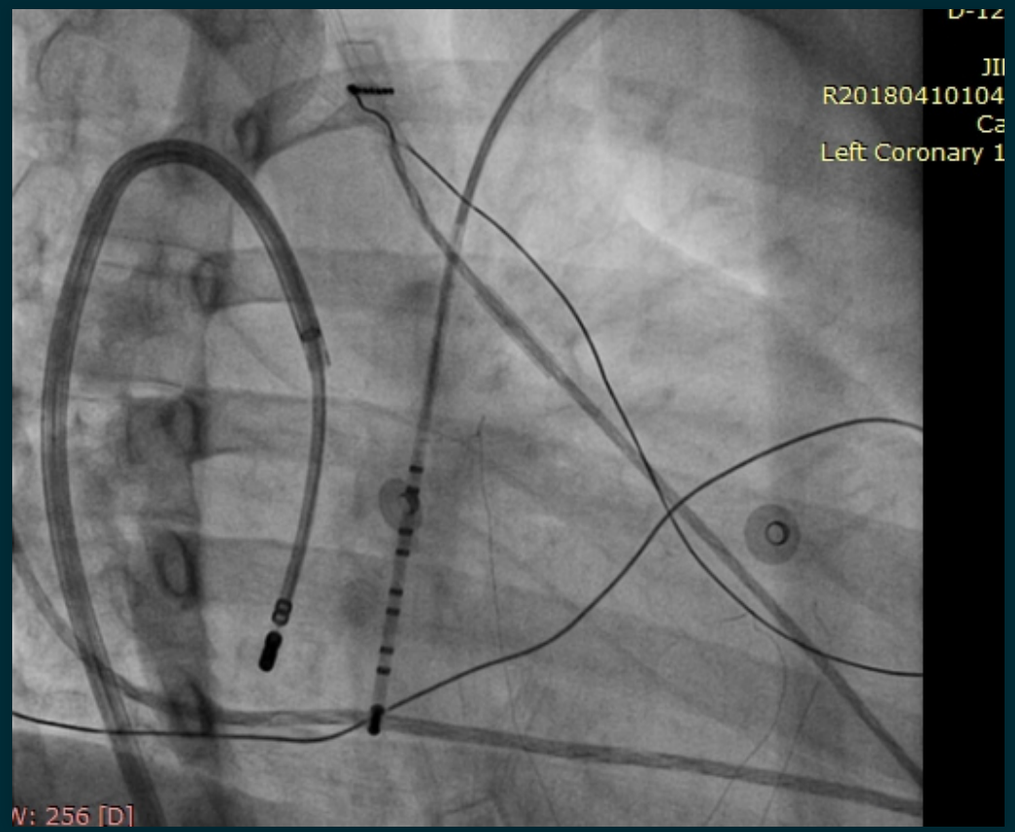
- Iliofemoral occlusions
  - Pediatric percutaneous procedures
- IVC interruption
- LSVC

Azygous continuation of IVC



RB Vidhyakar .. Raja Selvaraj. Ablation of atrioventricular nodal reentrant tachycardia using the superior approach in a patient with IVC interruption. Journal of Cardiovascular Electrophysiology 2012;23(12):1393-4

Azygous continuation of IVC



Mapping - Technology

- Use of 3D mapping
- Image integration
- Use of array
- Multielectrode mapping
- Use of magnetic navigation (1)

Ueda A, Suman-Horduna I, Mantziari L, Gujic M, Marchese P, Ho SY, Babu-Narayan SV, Ernst S. Contemporary outcomes of supraventricular tachycardia ablation in congenital heart disease: a single-center experience in 116 patients. Circ Arrhythm Electrophysiol. 2013 Jun;6(3):606-13. doi: 10.1161/CIRCEP.113.000415.

Catheter

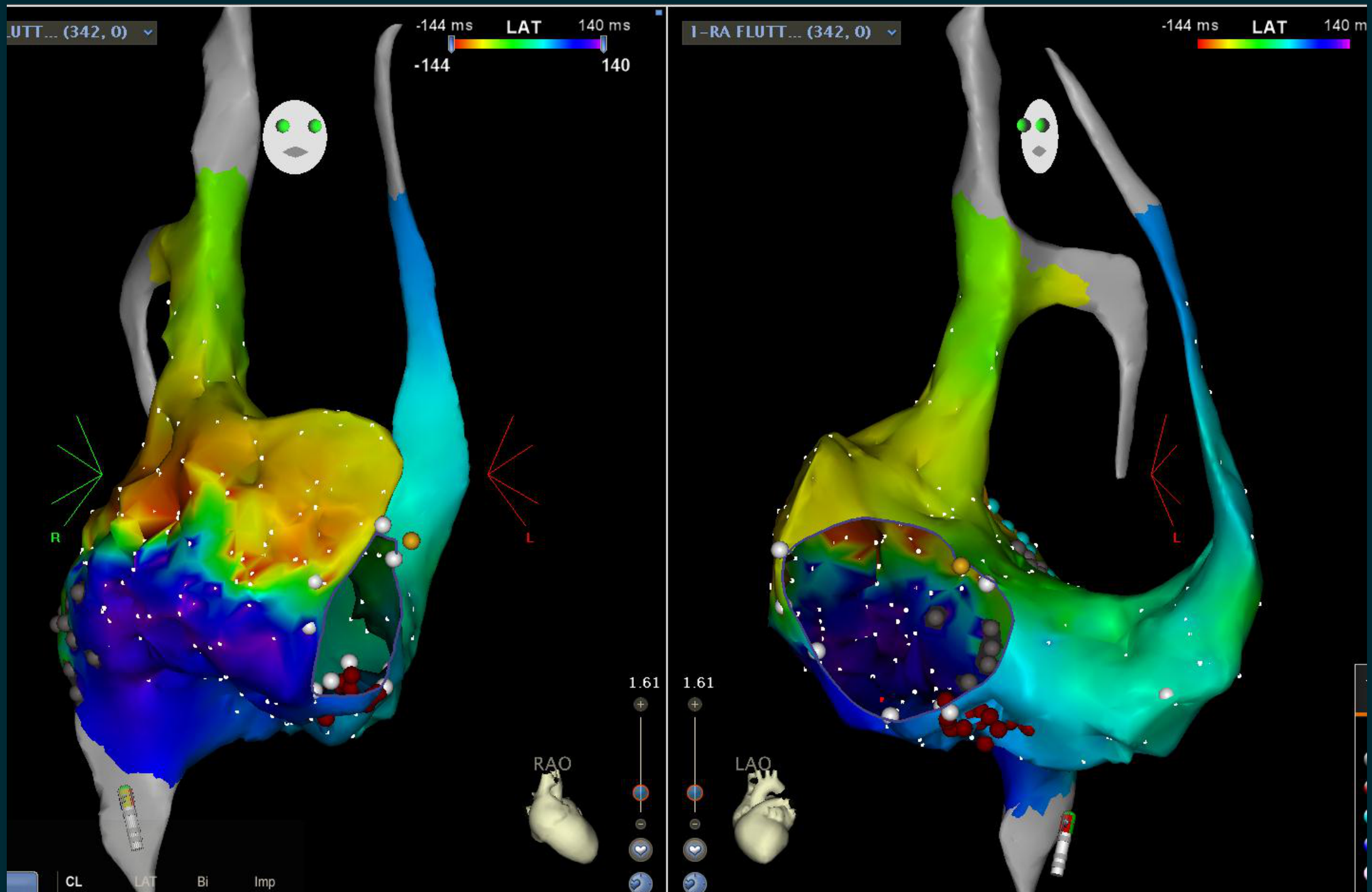
- Large tips
- Cooled tips
- Equally effective in producing transmural atrial lesions (1)

McGreevy KS, Hummel JP, Jiangang Z, et al. “Comparison of a saline irrigated cooled-tip catheter to large electrode catheters with single and multiple temperature sensors for creation of large radiofrequency lesions.” J Interv Card Electrophysiol. 2005; 14(3): 139-45

Substrate

- Conduction blocks - Atriotomy
  - Line of low voltage
  - Line of double potentials
- Scarring from pressure overload
  - Low voltage regions
  - Late potentials

Typical flutter

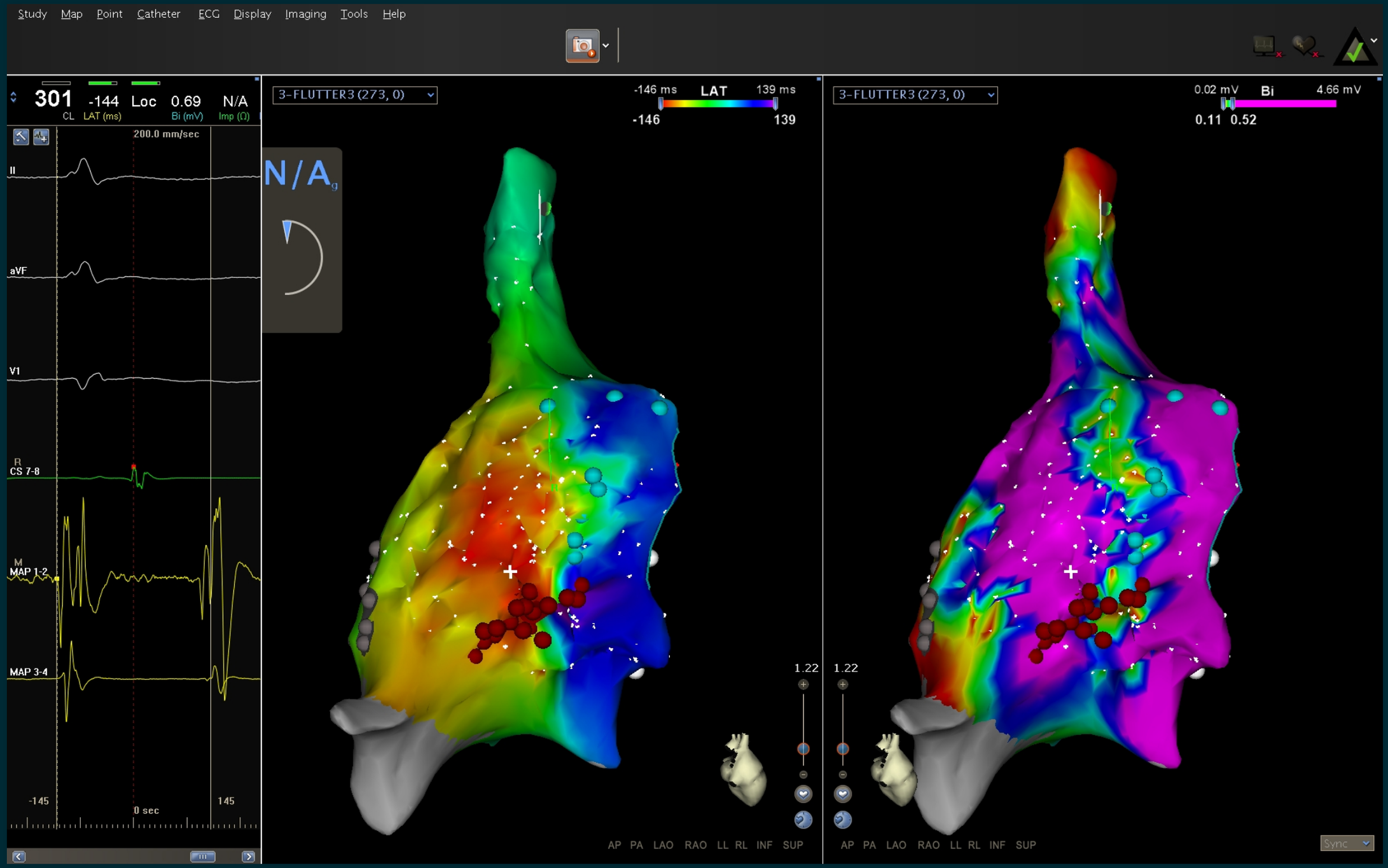


AP PA LAO RAO LL RL INF SUP

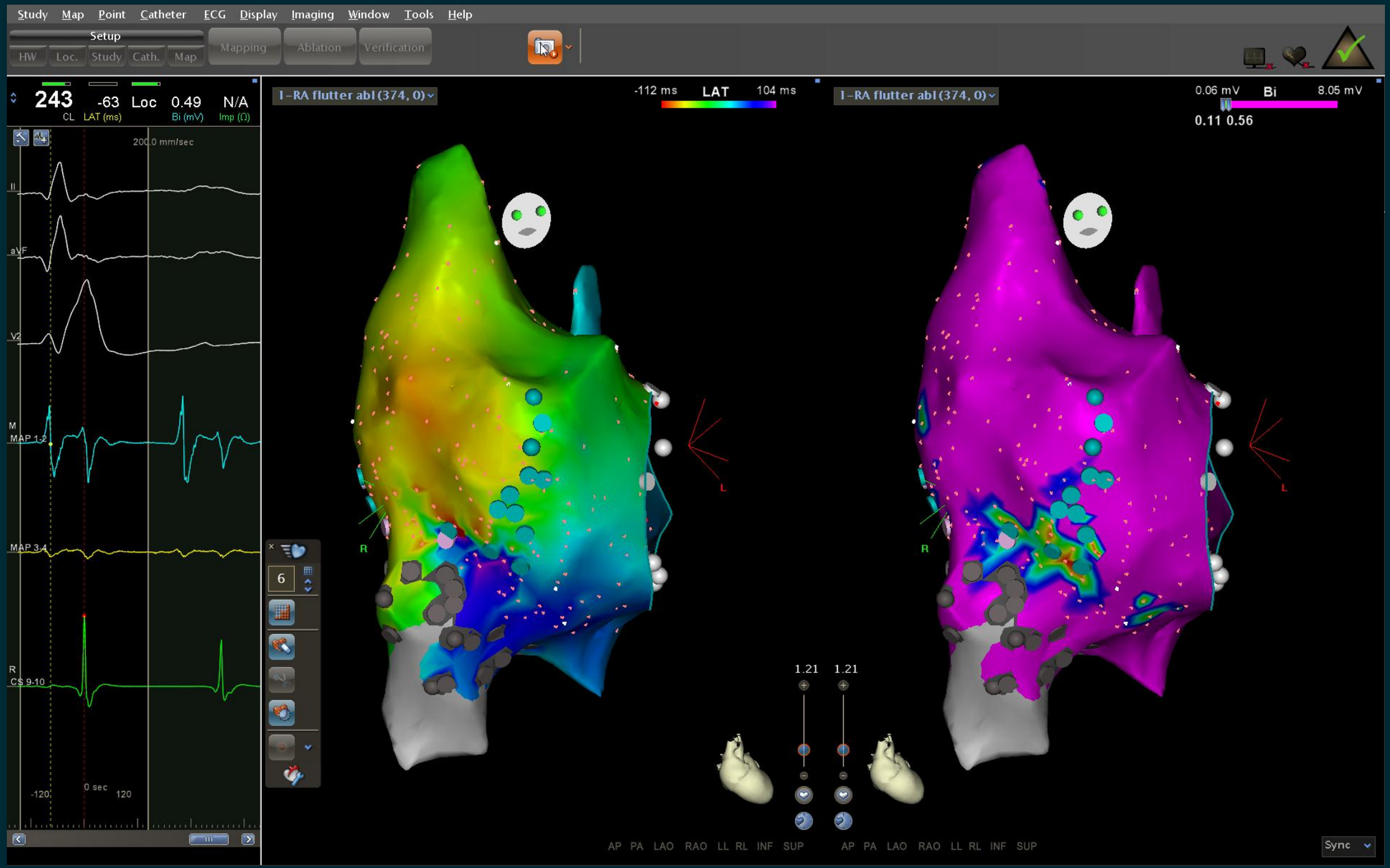
AP PA LAO RAO LL RL INF SUP

29 . 1

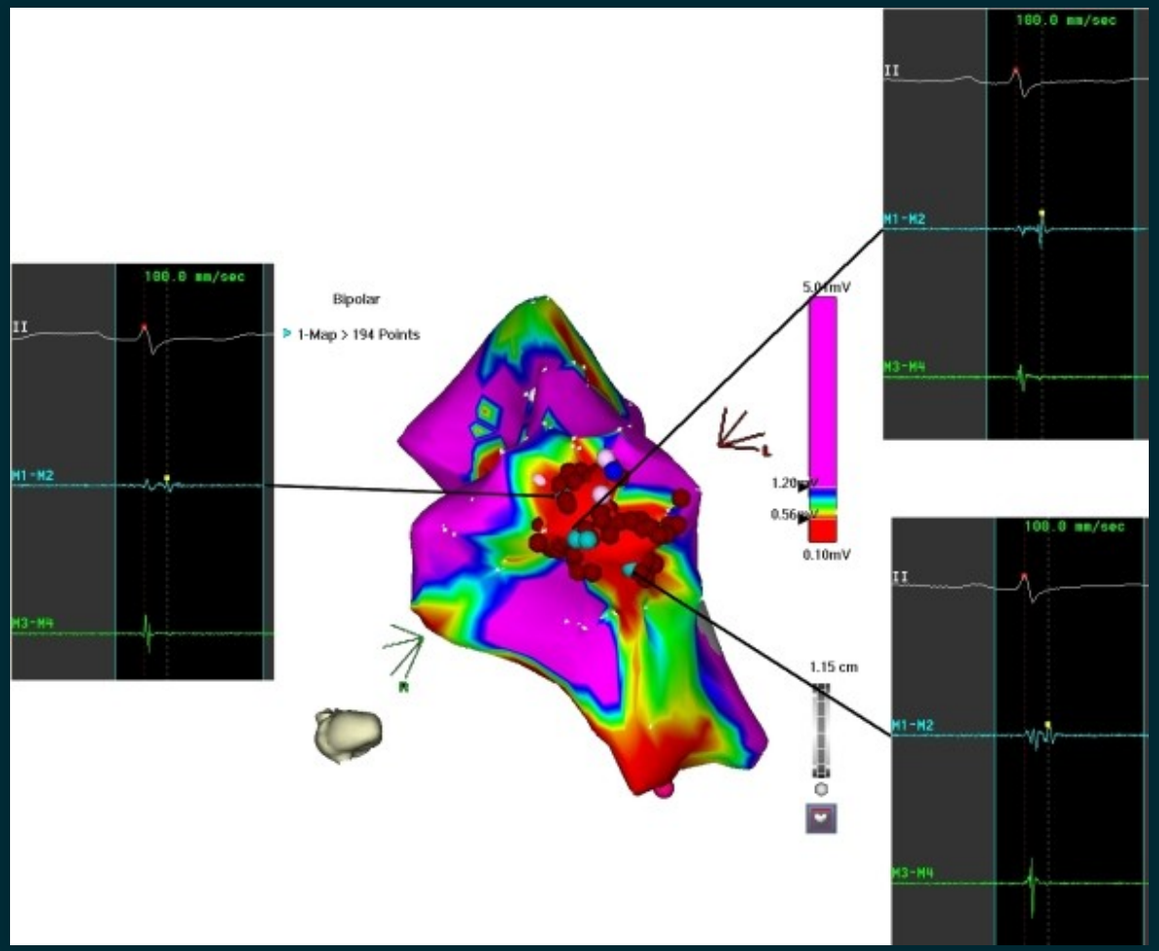
Substrate - Atriotomy



Substrate - Atriotomy

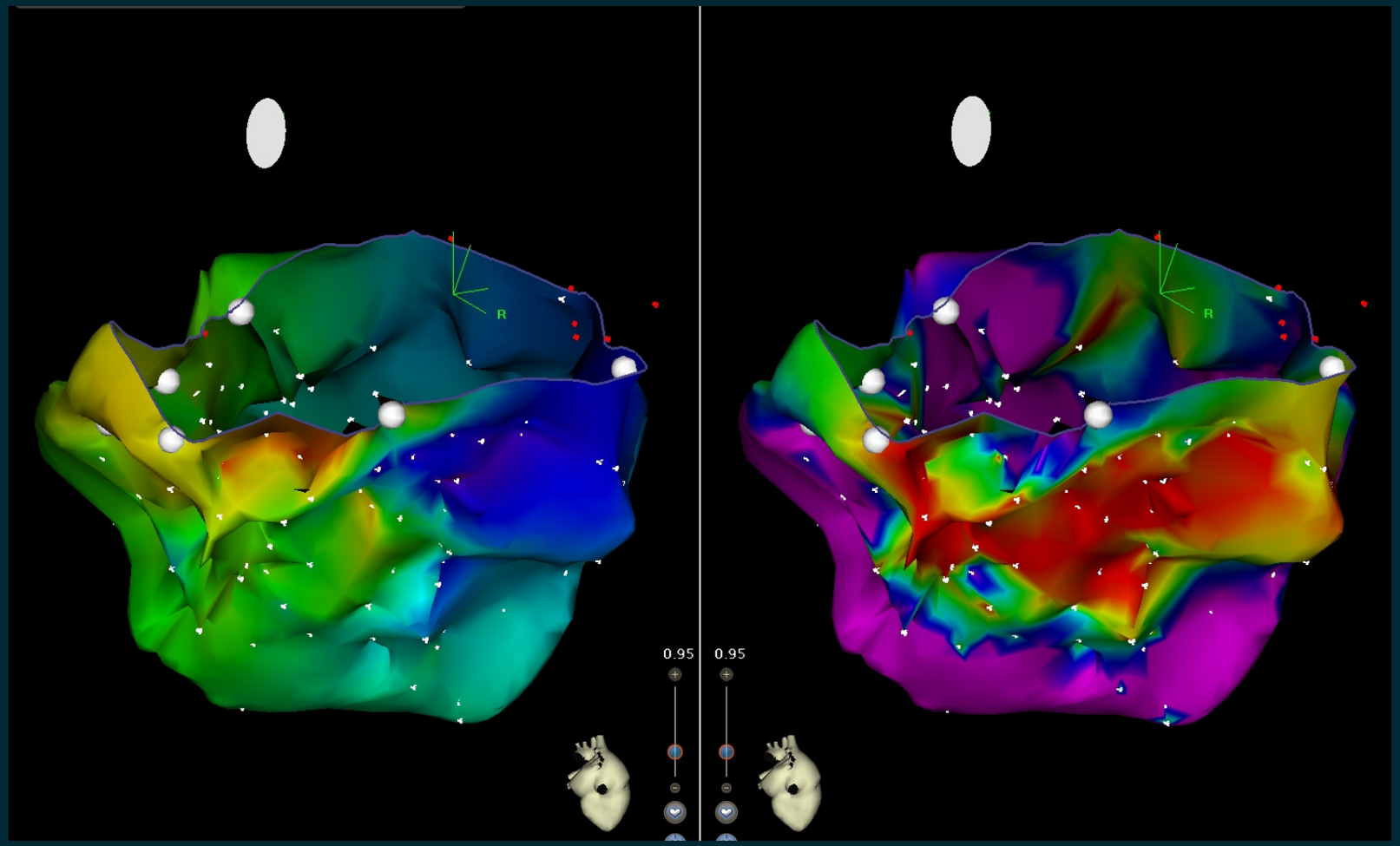


Substrate - Fibrosis / slow conduction



Raja Selvaraj et al. Ventricular tachycardia in repaired double chambered right ventricle – Identification of the substrate and successful ablation. Indian Pacing and Electrophysiology Journal 2012; 12(1):27-31

Substrate - Isthmus for reentrant VT

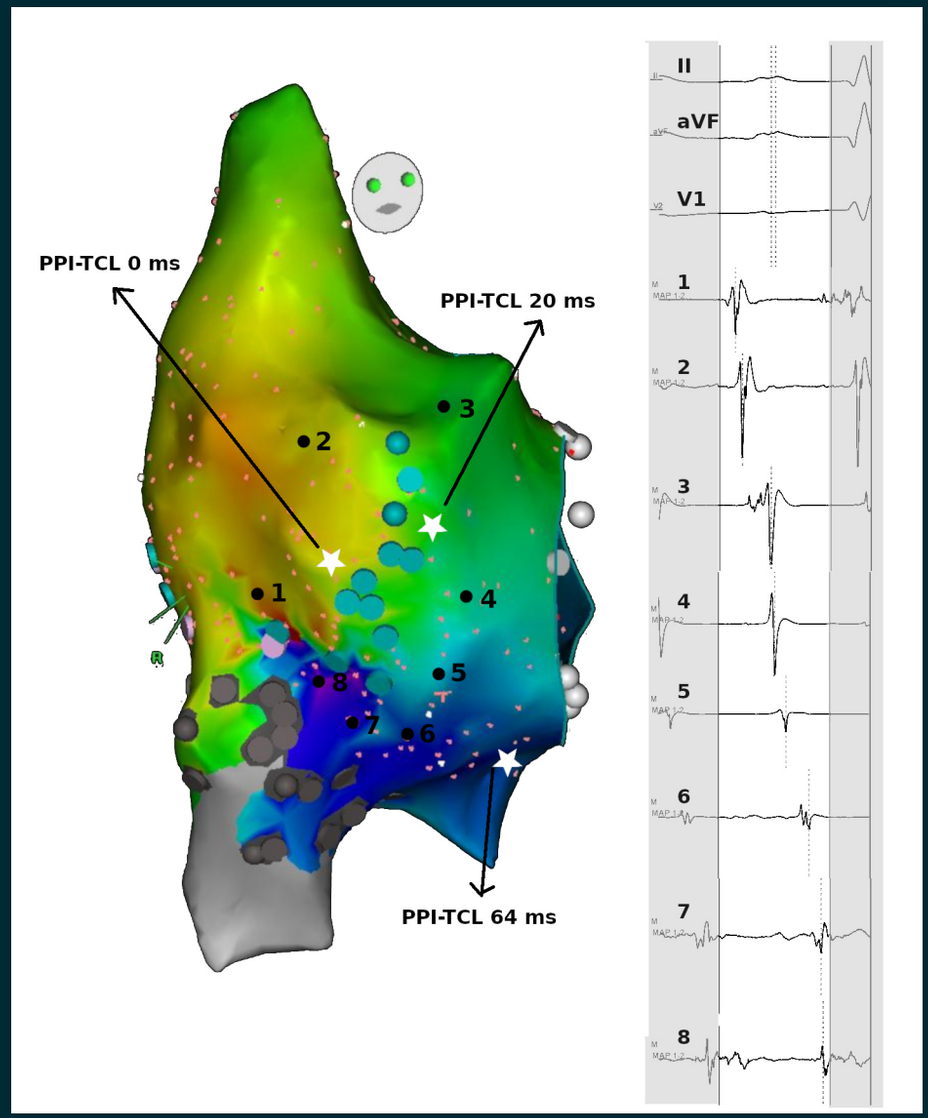


Senthil Kumar.. Raja J Selvaraj. Peritricuspid reentrant ventricular tachycardia in Ebstein's anomaly. Europace 2014;16(11):1633

Activation mapping - Identify circuit

- Careful mapping with density
- Entrainment

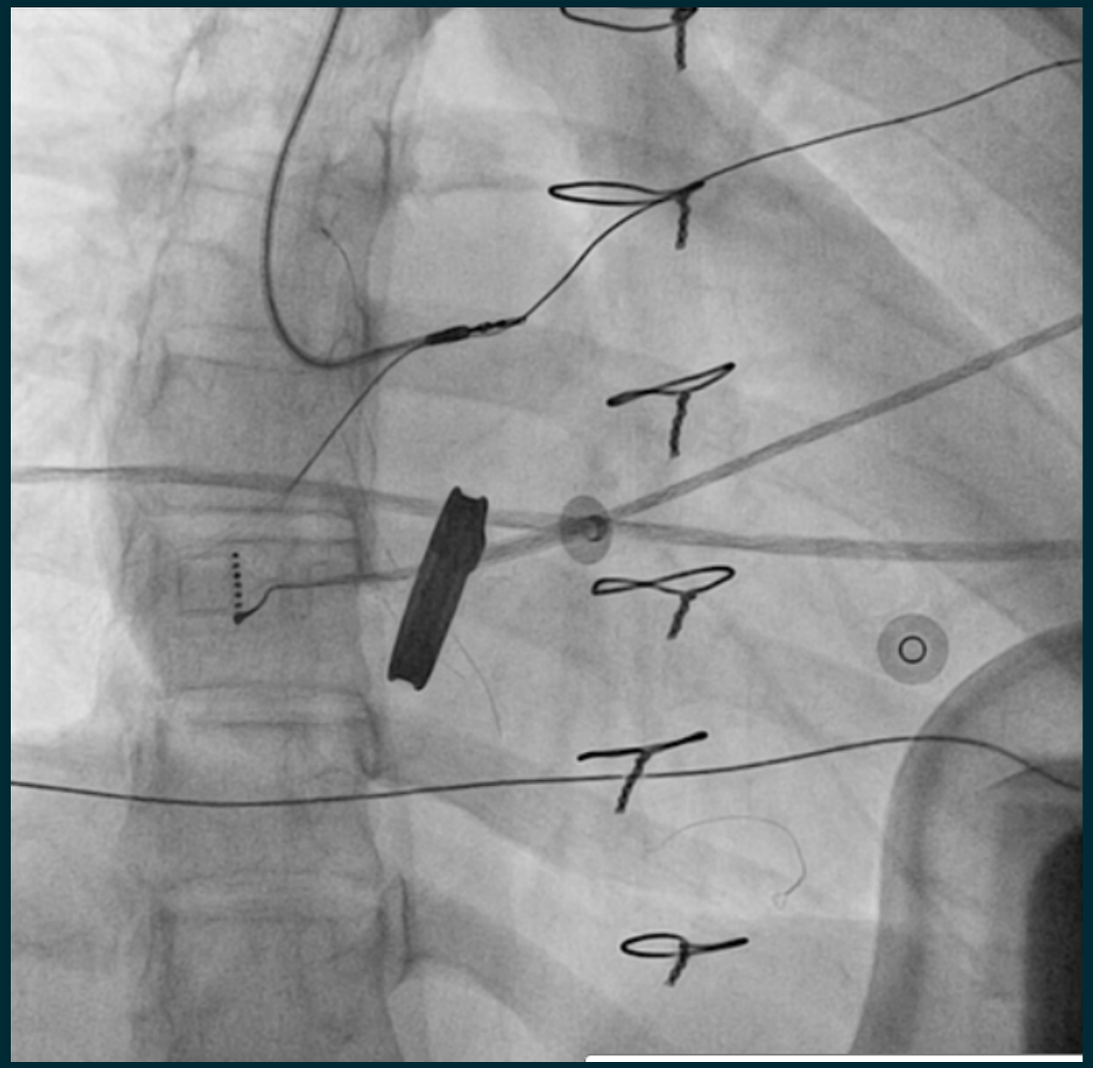
Activation and entrainment

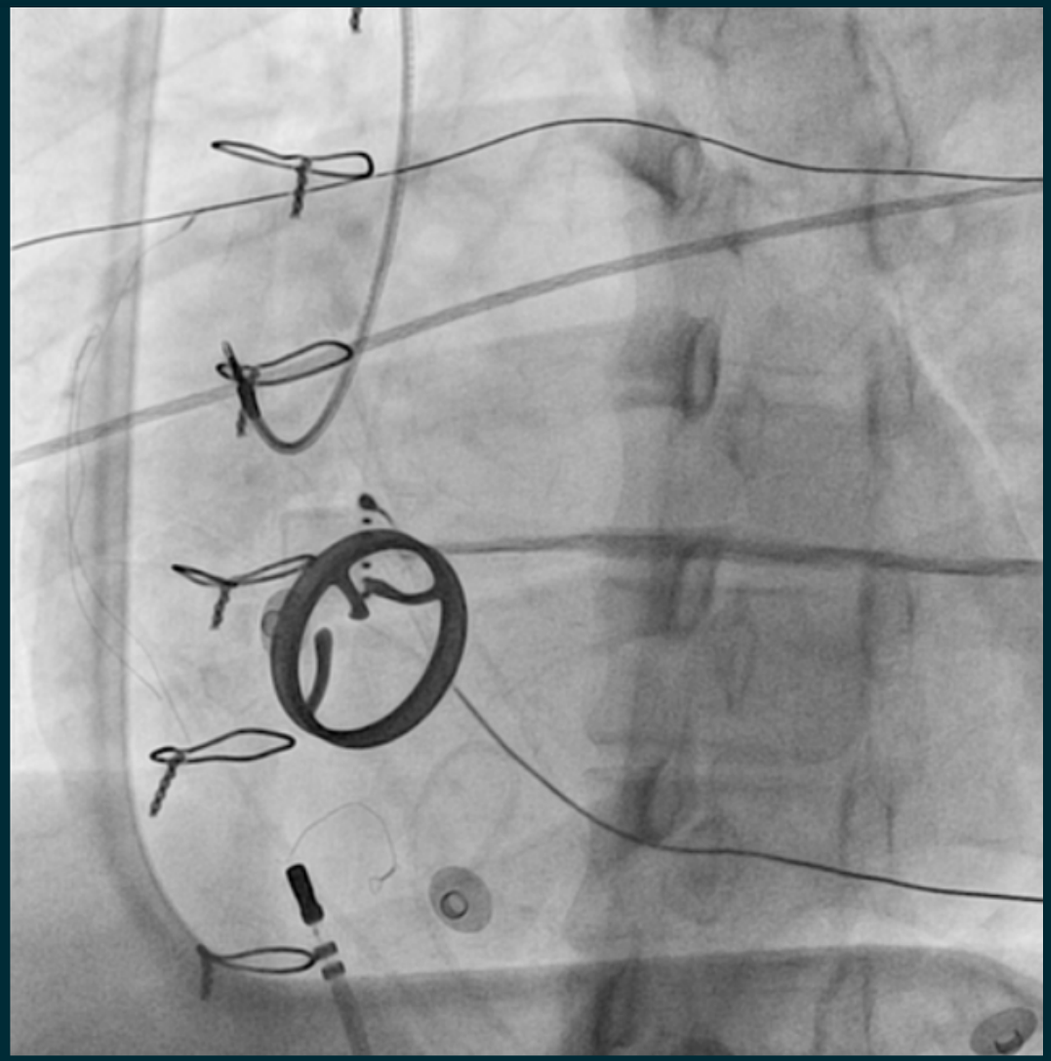


Activation mapping - Basics

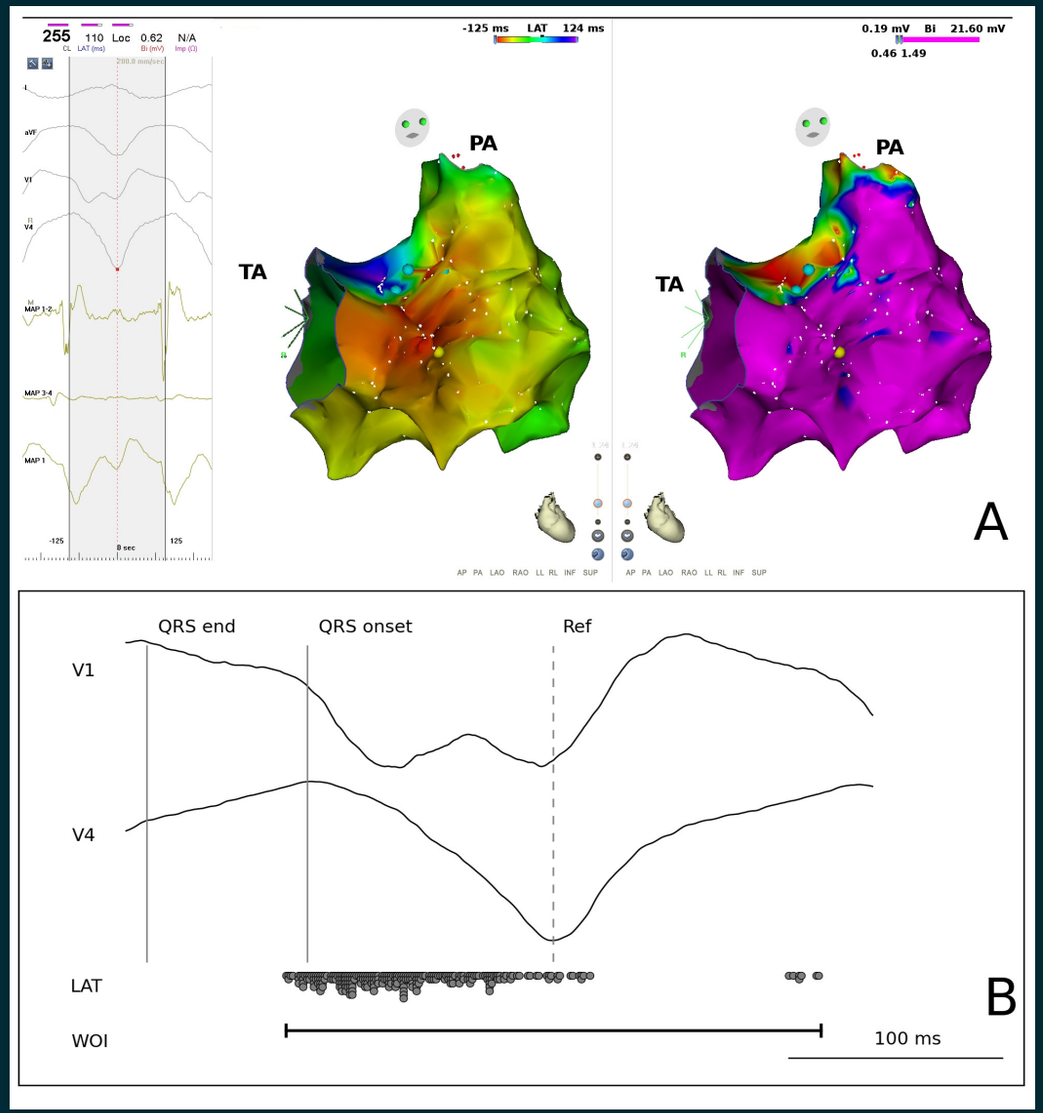
- Selecting a good reference electrogram - Stable and reliable
- Setting WOI
- Consistent activation time marking

Atrial activation reference

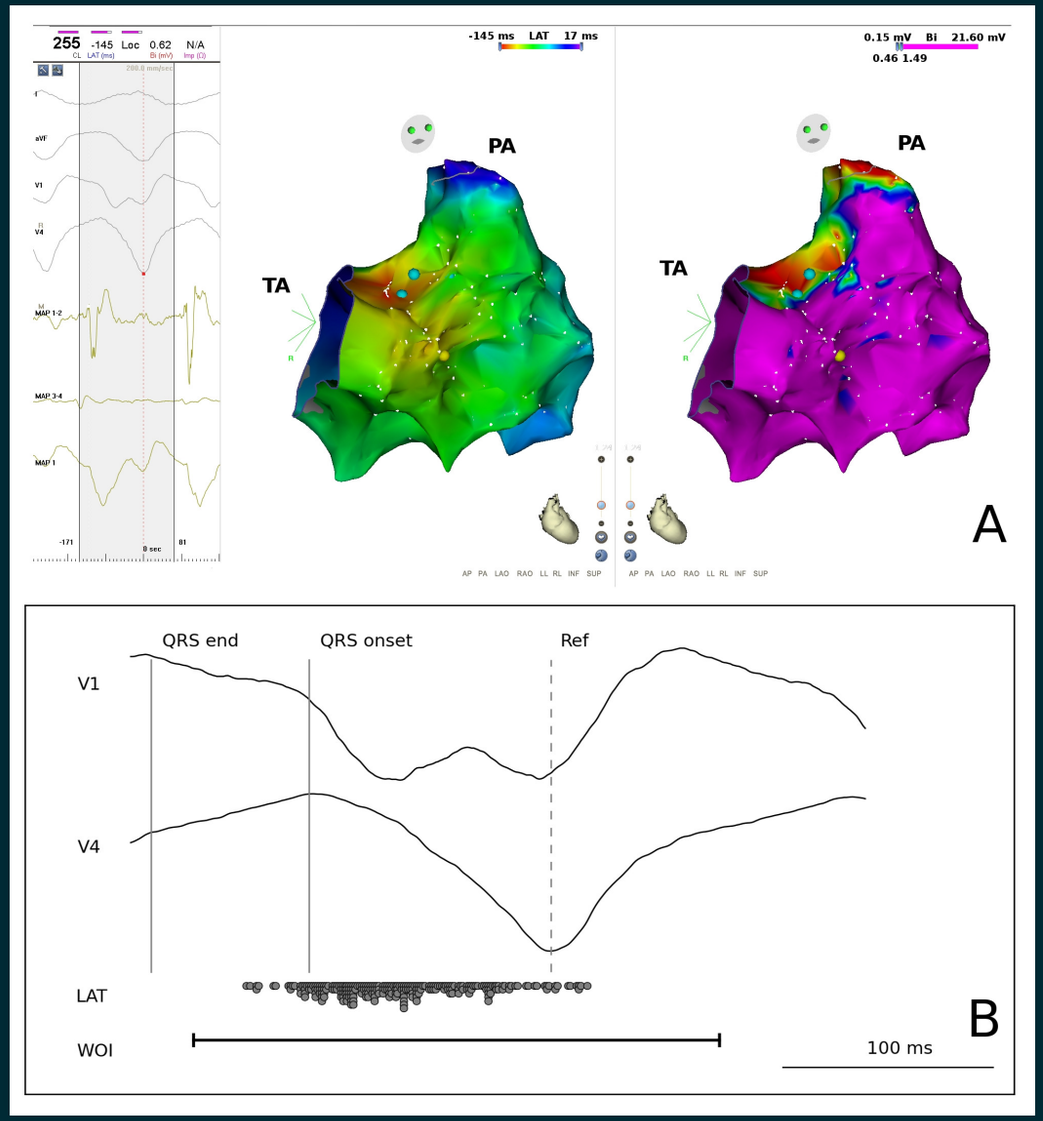




WOI - focal /reentry



WOI - focal /reentry

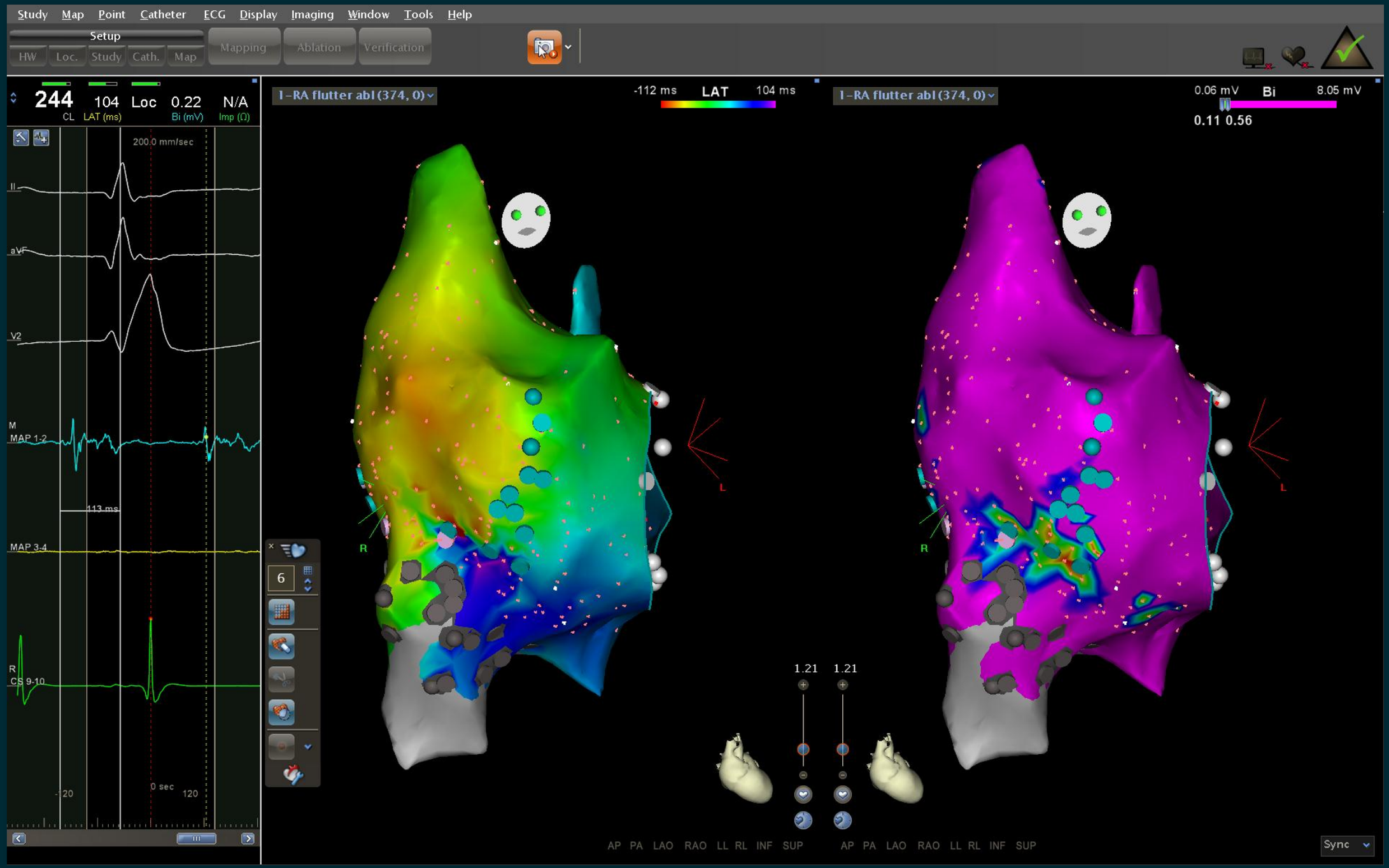


Raja J. Selvaraj et al. Circulation: Arrhythmia and Electrophysiology. 2014;7:553-556

Ablation - Identify isthmus

- Narrow
- Slow conduction
- Easy to ablate

Isthmus


Study Map Point Catheter ECG Display Imaging Window Tools Help
Setup
HW Loc. Study Cath. Map
Mapping
Ablation
Verification
244 CL
104 LAT (ms)
Loc
0.22 Bi (mV)
N/A Imp (Ω)
200.0 mm/sec
II
aVF
V2
M MAP 1-2
113 ms
MAP 3-4
R CS 9-10
-20
0 sec
120
1-RA flutter abl (374, 0)
-112 ms LAT 104 ms
1-RA flutter abl (374, 0)
0.06 mV Bi 8.05 mV
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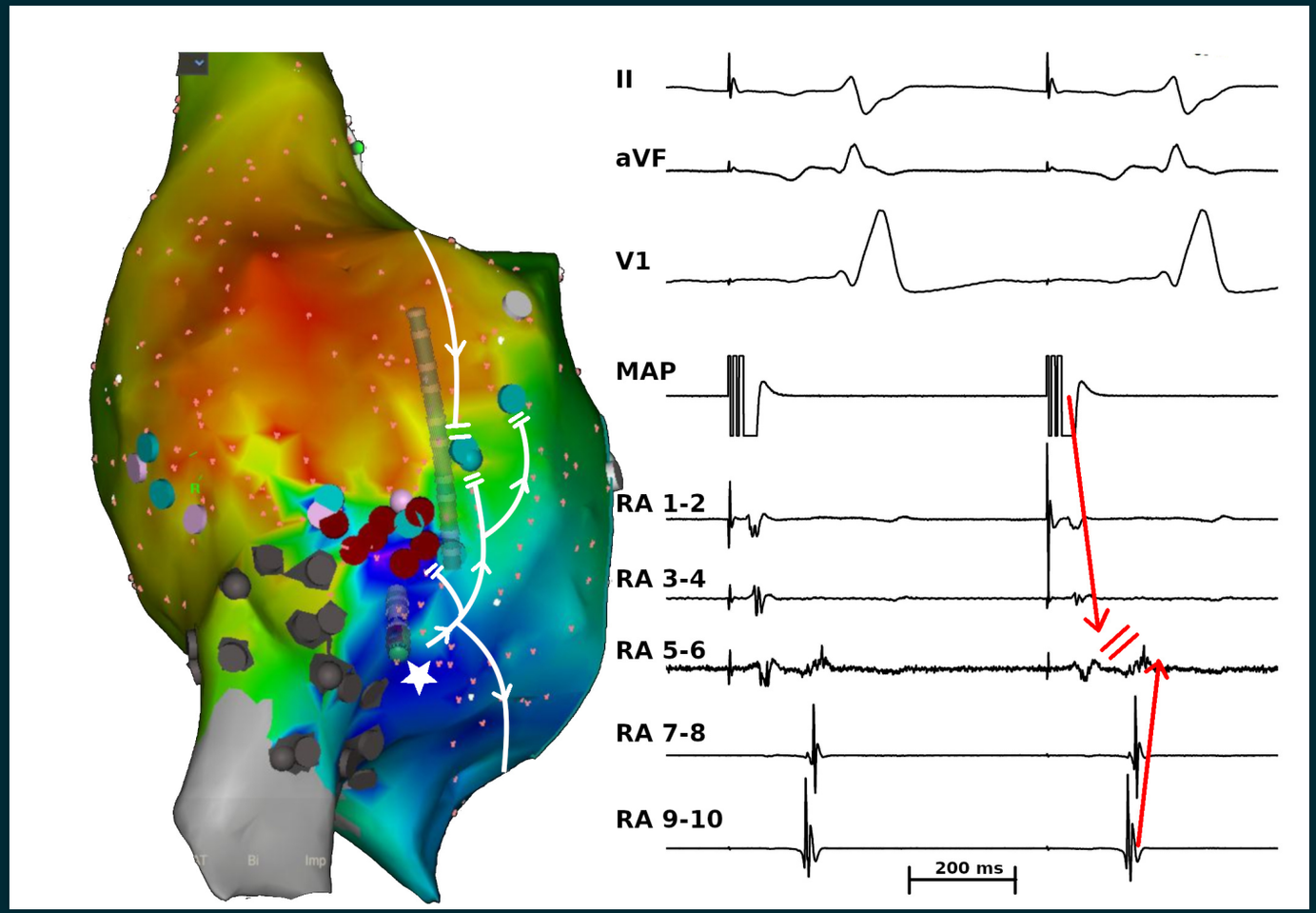
AP PA LAO RAO LL RL INF SUP
Sync

Ablation - Confirm block



Sudden death in CHD

- 25 to 100 times risk of sudden death compared to age matched controls (1)
- AS, CoA, TOF
- Arrhythmia / Circulatory - Embolic, aneurysm / Heart failure

Silka MJ, Hardy BG, Menashe VD, Morris CD. A population-based prospective evaluation of risk of sudden cardiac death after operation for common congenital heart defects. J Am Coll Cardiol. 1998 Jul;32(1):245-51. PubMed PMID: 9669277.

TOF - Ventricular arrhythmias and Sudden death

- Multicenter study - 793 patients
- Mean 20 yrs after repair
- Outcomes
  - Sustained VT - 33
  - Sudden death - 16
  - Atrial arrhythmias - 29
- Same substrate for VT and sudden death
  - Pulmonary regurgitation
  - QRS widening

Gatzoulis MA, Balaji S, Webber SA, Siu SC, Hokanson JS, Poile C, Rosenthal M, Nakazawa M, Moller JH, Gillette PC, Webb GD, Redington AN. Risk factors for arrhythmia and sudden cardiac death late after repair of tetralogy of Fallot: a multicentre study. Lancet. 2000 Sep 16;356(9234):975-81. PubMed PMID: 11041398.

ICDs in ACHD

- Difficult anatomy
- Inappropriate shocks
- Need for multiple surgeries
- Poor quality of life

Czosek et al. Impact of Cardiac Devices on the Quality of Life in Pediatric Patients. Circulation: Arrhythmia and Electrophysiology. 2012;5:1064-1072

Recommendation

Recommendations	Consensus statement	References
ICD is recommended for patients with CHD who are survivors of an aborted cardiac arrest due to VF or haemodynamically unstable VT after evaluation to define the cause of the event and exclusion of any reversible causes.		57,58,61,295,305,306
ICD is recommended for patients with CHD with symptomatic sustained VT who have undergone haemodynamic and electrophysiological evaluation.		57,58,61,143,295,296
ICD is recommended in adults with CHD and a systemic LVEF ≤ 35%, biventricular physiology and NYHA functional Class II or III.		20,307,308

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Thank you for the attention !