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**Winter Arrhythmia**  
School  
*Annual Cardiac Arrhythmia Meeting*  
*Division of Cardiology, University of Toronto*

# Ventricular Arrhythmias in Adult Congenital Heart Disease

Krish Nair



# Objectives

- To discuss ventricular tachycardia in Adult Congenital Heart Disease (ACHD) focusing on Tetralogy of Fallot (TOF)
- To discuss the role of programmed ventricular stimulation
- To discuss intra-operative mapping and cryoablation
- To discuss the techniques of Catheter ablation

# Ventricular Arrhythmias in Congenital Heart Disease

- Rare in the first and second decade of life  
Atrial arrhythmias are far more common
- TOF  
TGA  
cCTGA  
Single ventricle physiology (Fontan)  
Ebstein anomaly  
Eisenmenger syndrome  
Aortic coarctation



# Major features

- Much of the current literature surrounds ventricular arrhythmias in TOF
- Significantly higher incidence reported in patients following atrial switch repair.
- Monomorphic VT with a left bundle branch morphology is most commonly described in TOF, as well as Ebstein's anomaly
- Polymorphic VT and VF is best described in patients following atrial switch repair.



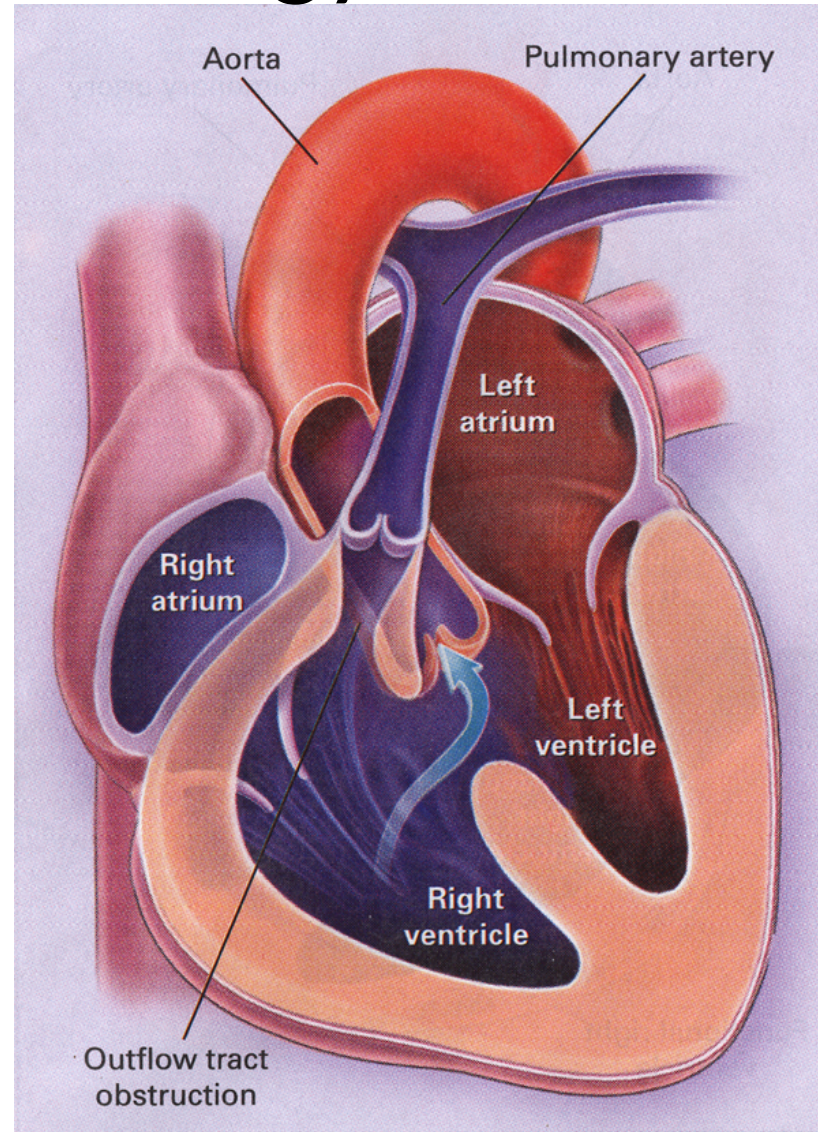


# Major features (contd.)

- Lack of data on ventricular arrhythmias in Fontan patients currently
- Atrial arrhythmias may be predictors of ventricular arrhythmias and sudden cardiac death in TGA, Ebstein's anomaly and Fontan
- Sudden death is recognised in Eisenmenger syndrome and in aortic coarctation



# Tetralogy of Fallot





# Tetralogy of Fallot

- Incidence of clinical sustained VT and SCD, 11.9% and 8.3% 35 years after repair\*
- Timing of initial repair
  - Late i.e. after first decade
- Type of repair
  - Evolution from ventriculotomy to transatrial/transpulmonary approach
  - VSD patch

\*

*Gatzoulis MA et al. Lancet. 2000,356: 975-81.*



# Risk stratification

- Transannular patch
- Severe PR
- Right ventricular dilatation
- QRS duration  $\geq 180$  msec on ECG, or a QRS duration that increases at a rate of  $> 3$  msec/year
- Tricuspid regurgitation

# Value of Programmed Ventricular Stimulation



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- Predicted subsequent cardiac arrest
  - » Alexander et al JCE 1999
- Negative Induction in all who died
  - » Chandar et al Am J Cardiol 1990
- False positive induction
  - » Lucron et al Am J Cardiol 1999
- Multicenter study of 252 patients
  - » Khairy P et al. Circulation 2004; 109:1994–2000



# Role of EP Study

- Yields insufficiently high positive predictive values so cannot be recommended for routine screening test in asymptomatic patients.
- TOF patients of intermediate risk will likely have the greatest benefit from an EP study.
- Needs robust stimulation protocols
- Inducible monomorphic or polymorphic sustained VT was predictive of subsequent clinical sustained VT or sudden death.

*Khairy P et al. Circulation 2004; 109:1994- 2000*





# Management Options

- Pharmacologic
- ICD
- PVR with and without intra-operative cryoablation
- Catheter Ablation



# Pharmacological therapy

- Amiodarone
- Beta-blockers and sotalol are commonly prescribed
- Avoid class IA or IC antiarrhythmic agents if either ventricle is dysfunctional.



## Class I: ICD therapy is indicated in adults with CHD

- 1. who are survivors of cardiac arrest due to ventricular fibrillation or hemodynamically unstable ventricular tachycardia
- 2. and spontaneous sustained ventricular tachycardia who have undergone hemodynamic and electrophysiologic evaluation
- 3. and a systemic left ventricular ejection fraction  $\leq 35\%$ , biventricular physiology, and New York Heart Association(NYHA) class II or III symptoms



# Issues with ICDs

- Lack of venous access to the ventricles or abnormal systemic veins
- Right sided AV valve disease
- Subcutaneous ICD is an option: however cannot pace

# Pulmonary valve replacement

- Free pulmonary regurgitation is common following transannular patch repair.
- Pulmonary regurgitation is the main underlying haemodynamic lesion for patients with ventricular tachycardia and sudden death.

Gatzoulis MA. *Lancet* 2000;356: 975-81.

- Indexed end systolic and end diastolic volumes are significantly higher in patients with inducible sustained VT. Marie PY. 1992;69(8):785-789.



## Indications for PVR

**Severe pulmonary regurgitation associated with:**

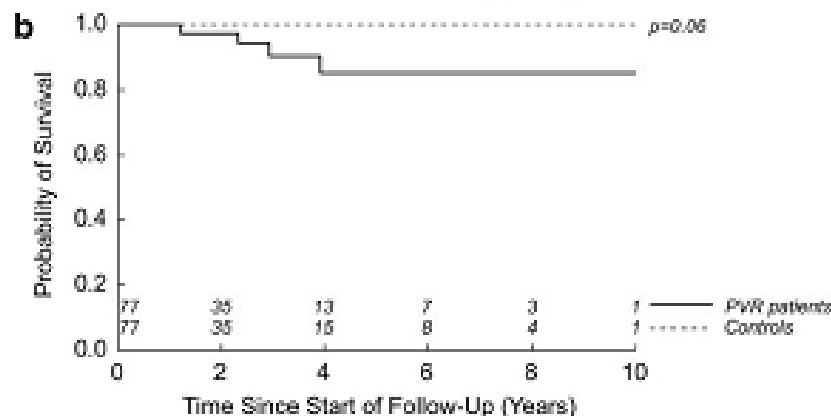
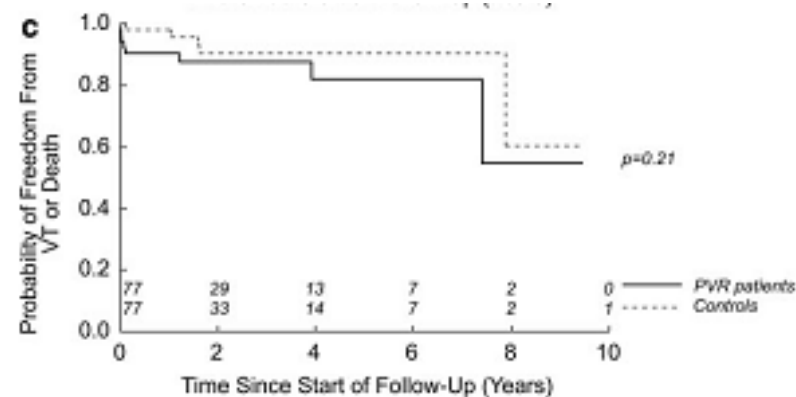
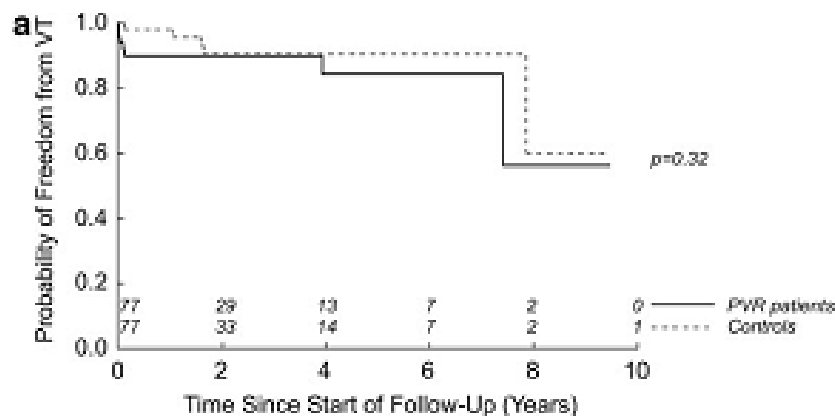
Moderate-to-severe RV dysfunction

RV enlargement (e.g., right ventricular end-diastolic volume  $>170$  ml/m<sup>2</sup>)

moderate-to-severe TR

and/or symptomatic or sustained atrial or ventricular arrhythmias .

# Pulmonary Valve Replacement: Impact on Survival and Ventricular Tachycardia



No significant differences were seen in VT, death, or combined VT and/or death

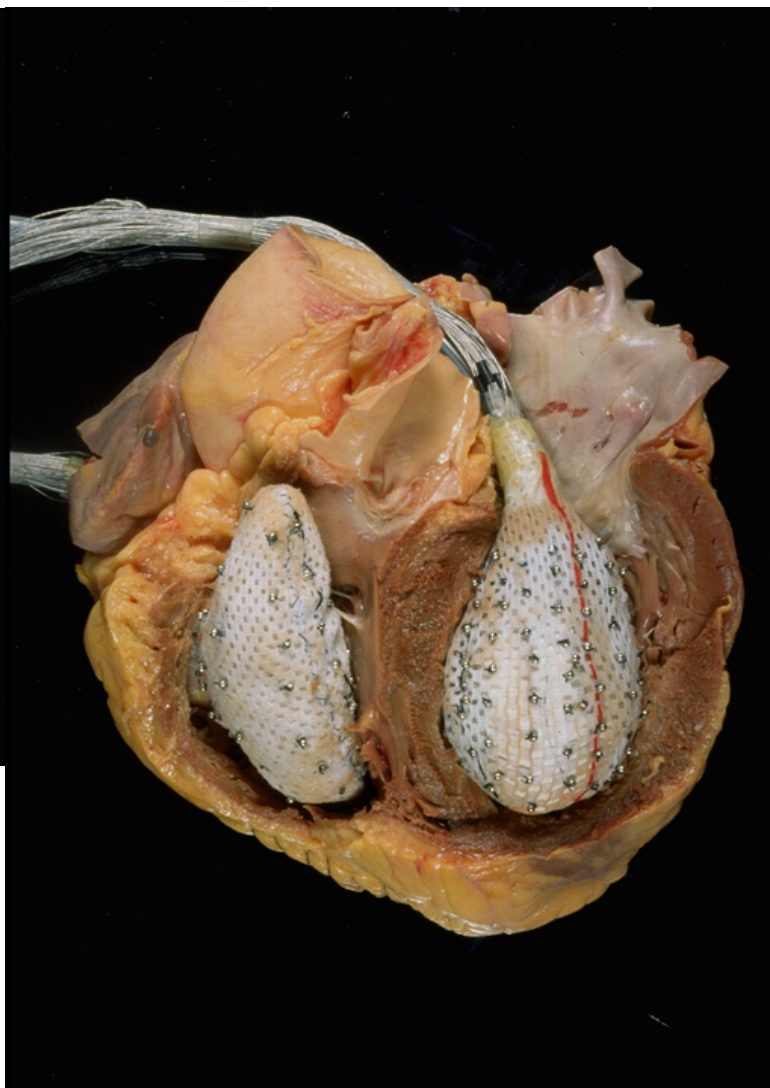
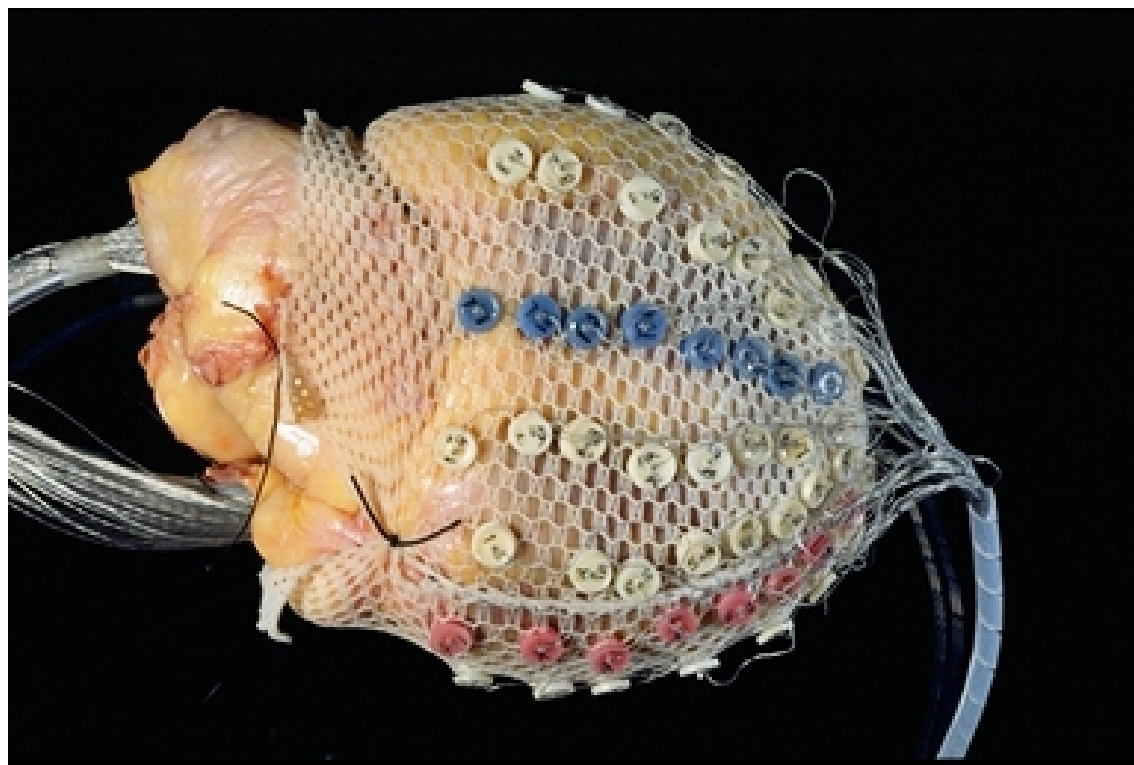
*Circulation* . 2009;119:445-451.

Benefits include a reduction in right ventricular volumes, along with a transient decrease in QRS duration

The notion that pulmonary valve replacement alone affords sufficient protection against sudden death has been refuted by recent studies



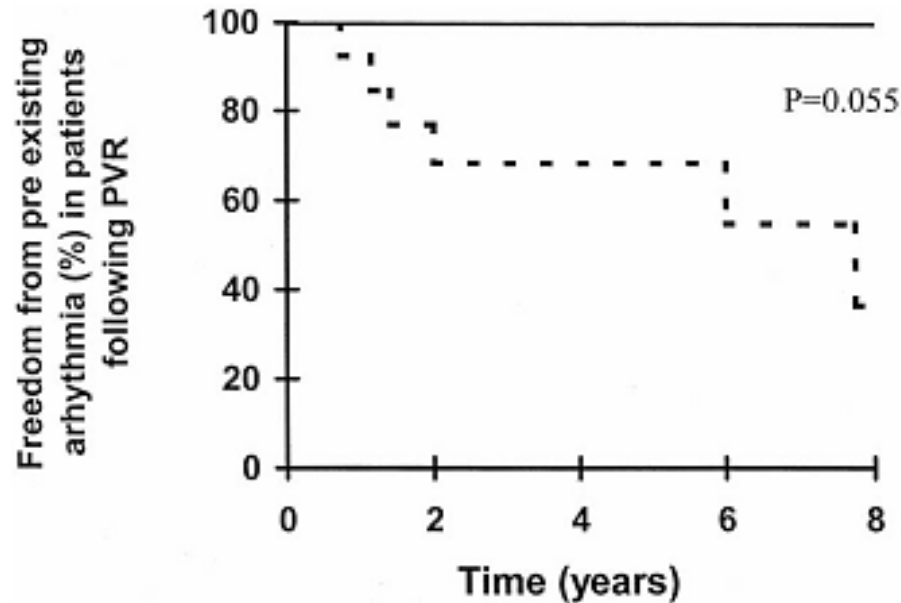
# Intraoperative Ablation



# Pulmonary Valve Replacement with concomitant intraoperative ablation therapy



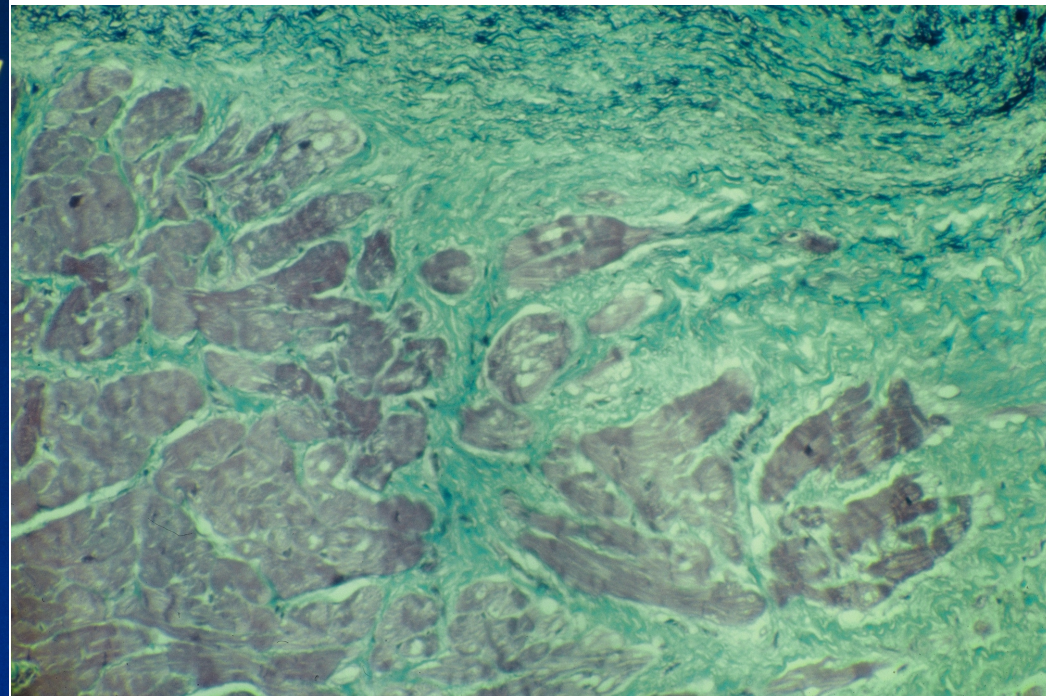
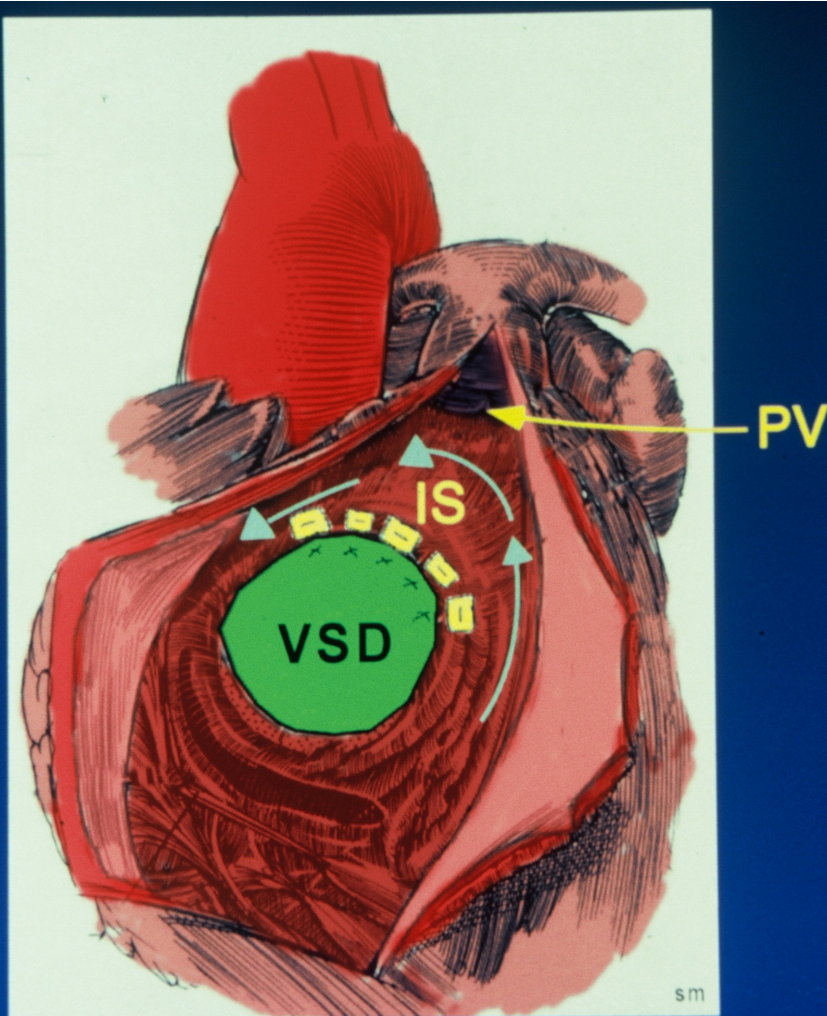
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Intraoperative ablation reduces the incidence of arrhythmias  
Optimal approach and its impact on sudden death remains to be determined

*Circulation . 2001;103:2489-2494*







# 1. FLASHING LIGHT DISPLAY



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## 2. ELECTROGRAM ANALYSIS



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14<sup>th</sup>

Collingwood  
February

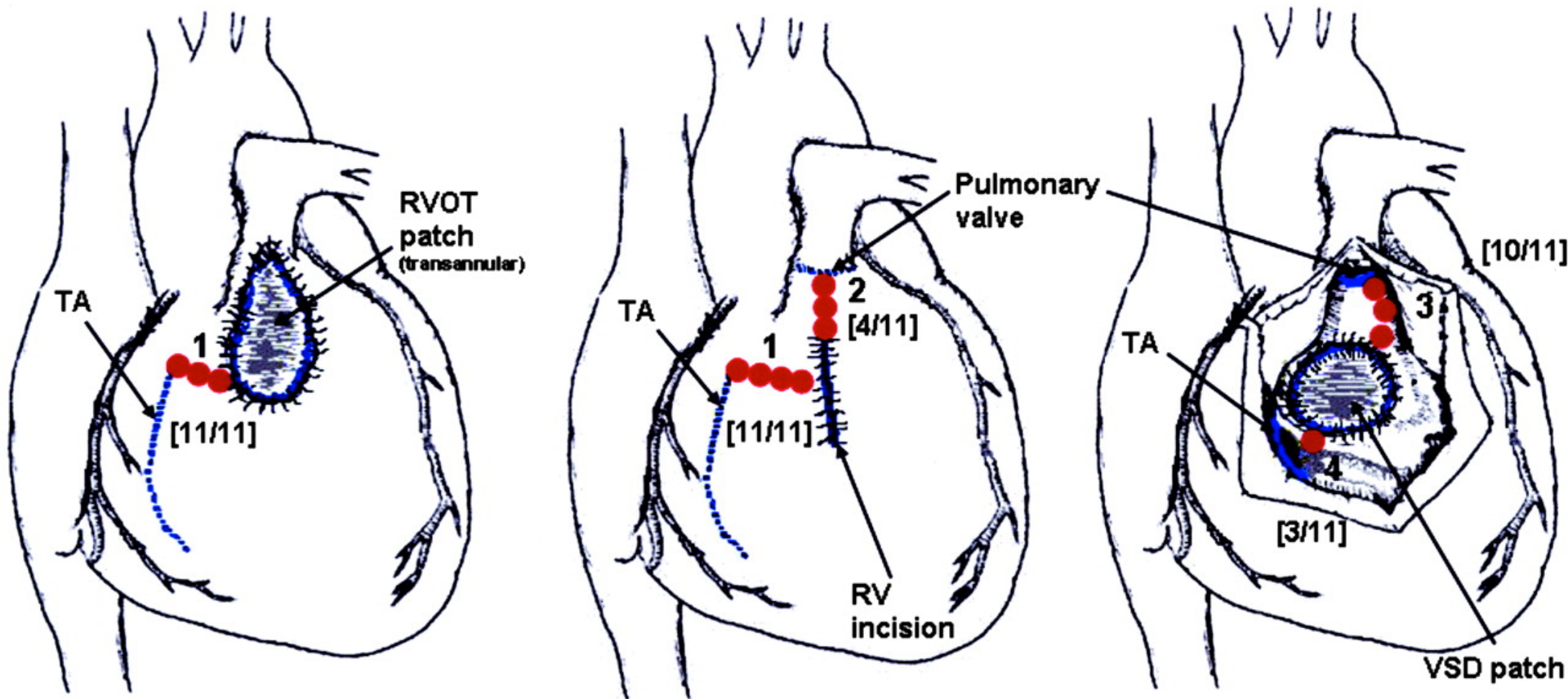


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# CATHETER ABLATION

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## 4 ANATOMIC ISTHMUSES



Schematic of the localization of anatomic boundaries (blue lines) for VT after repair of CHD and the resulting anatomic isthmuses (red lines); frequency of the distinct isthmuses in brackets.

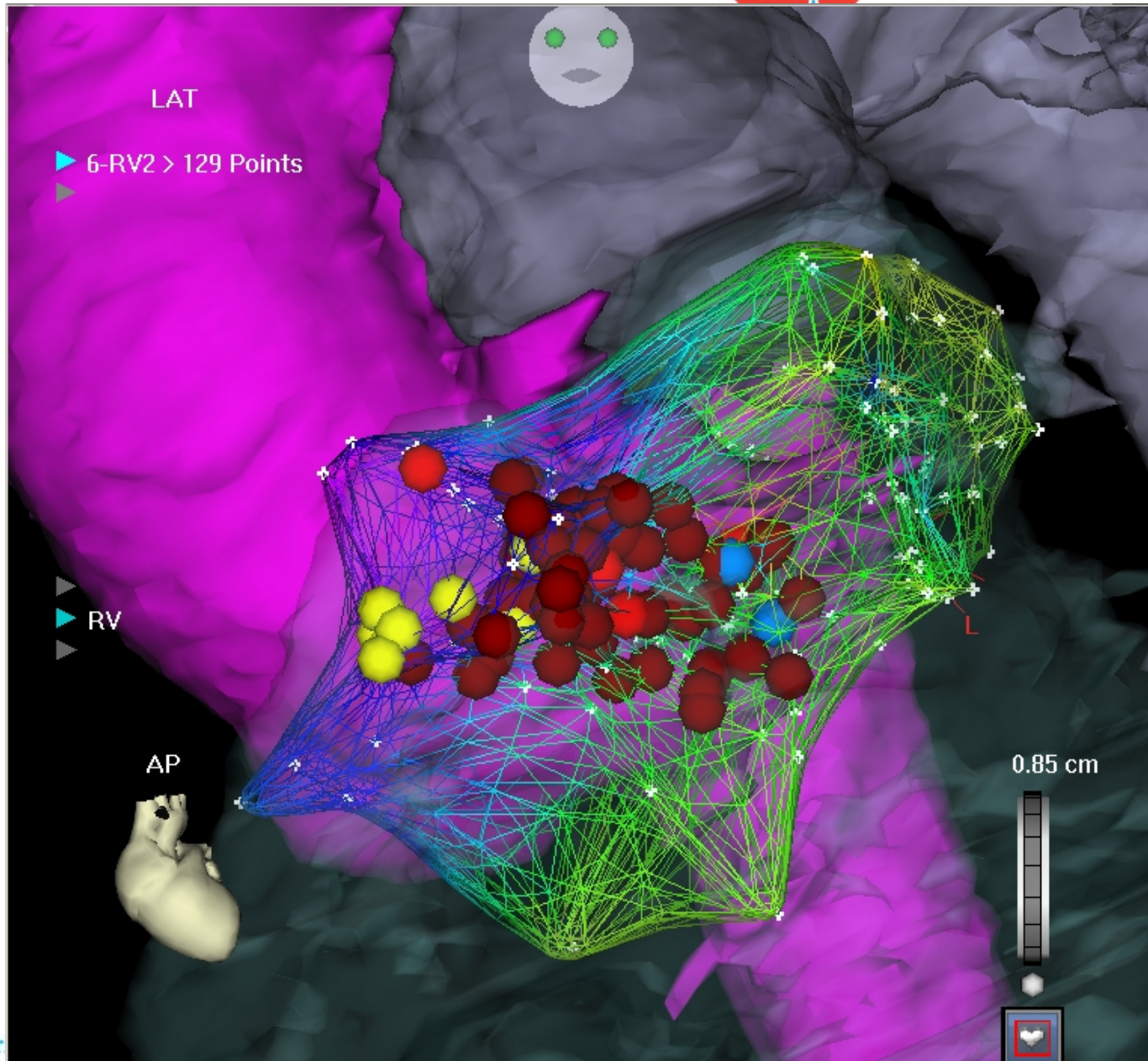
**Zeppenfeld, K et al. Circulation 2007 ;  
116 :2241-2252**



- The reentry circuit isthmuses of all induced 15 VTs were identified.
- 11 of 15 VTs in anatomic isthmus 1
- Transecting the anatomic isthmuses by ablation lesions abolished all VTs.
- At  $30.4 \pm 29.3$  months of follow-up, 91% of patients remained VT free.



### 3. PACE MAPPING STRATEGY

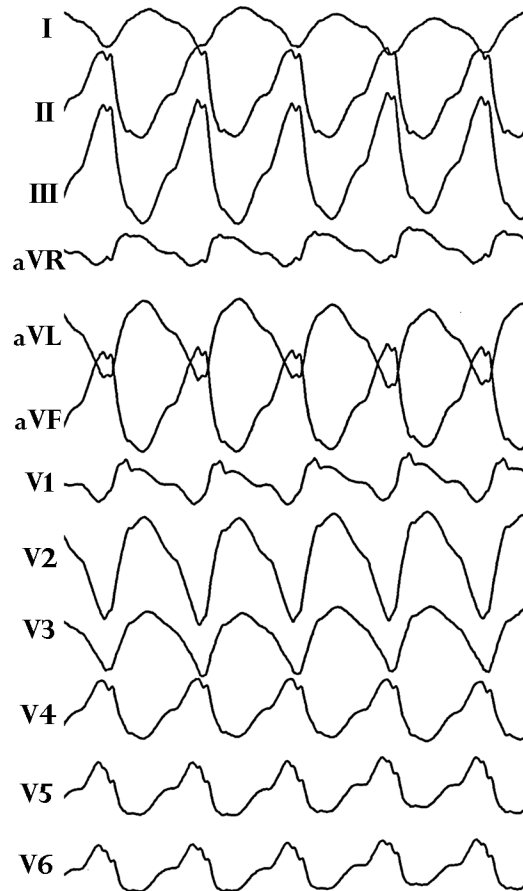


# -PACE MAPPING STRATEGY



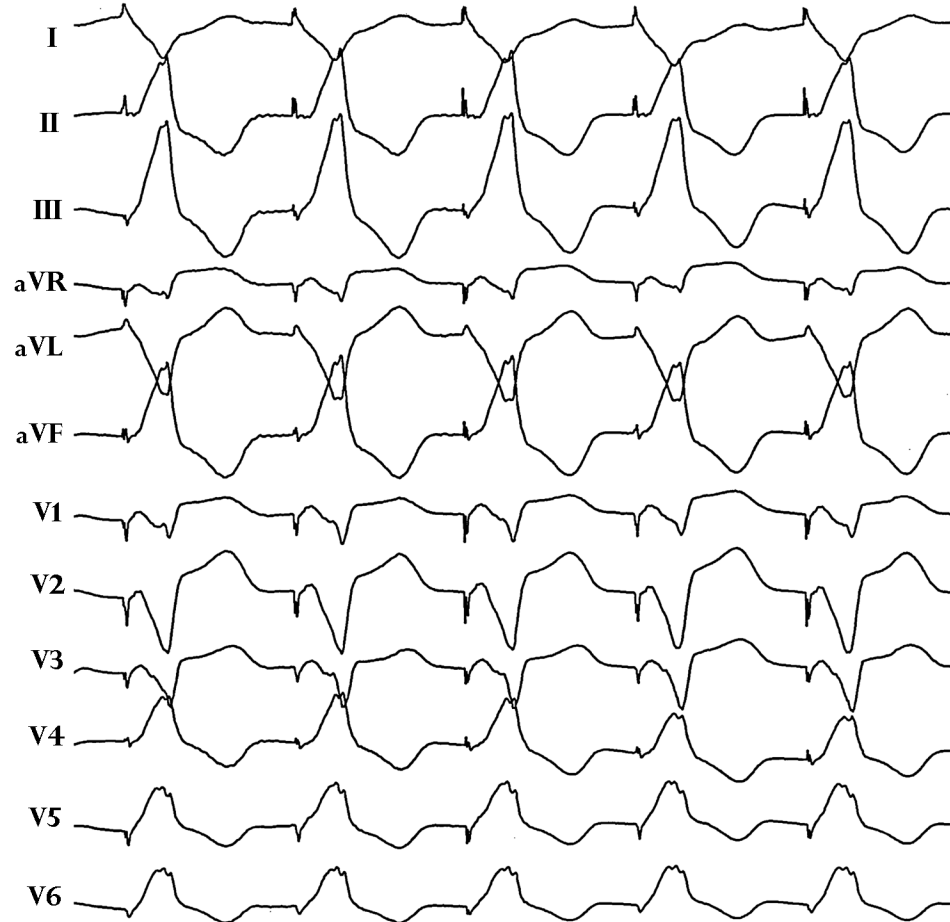
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**CLINICAL VT**



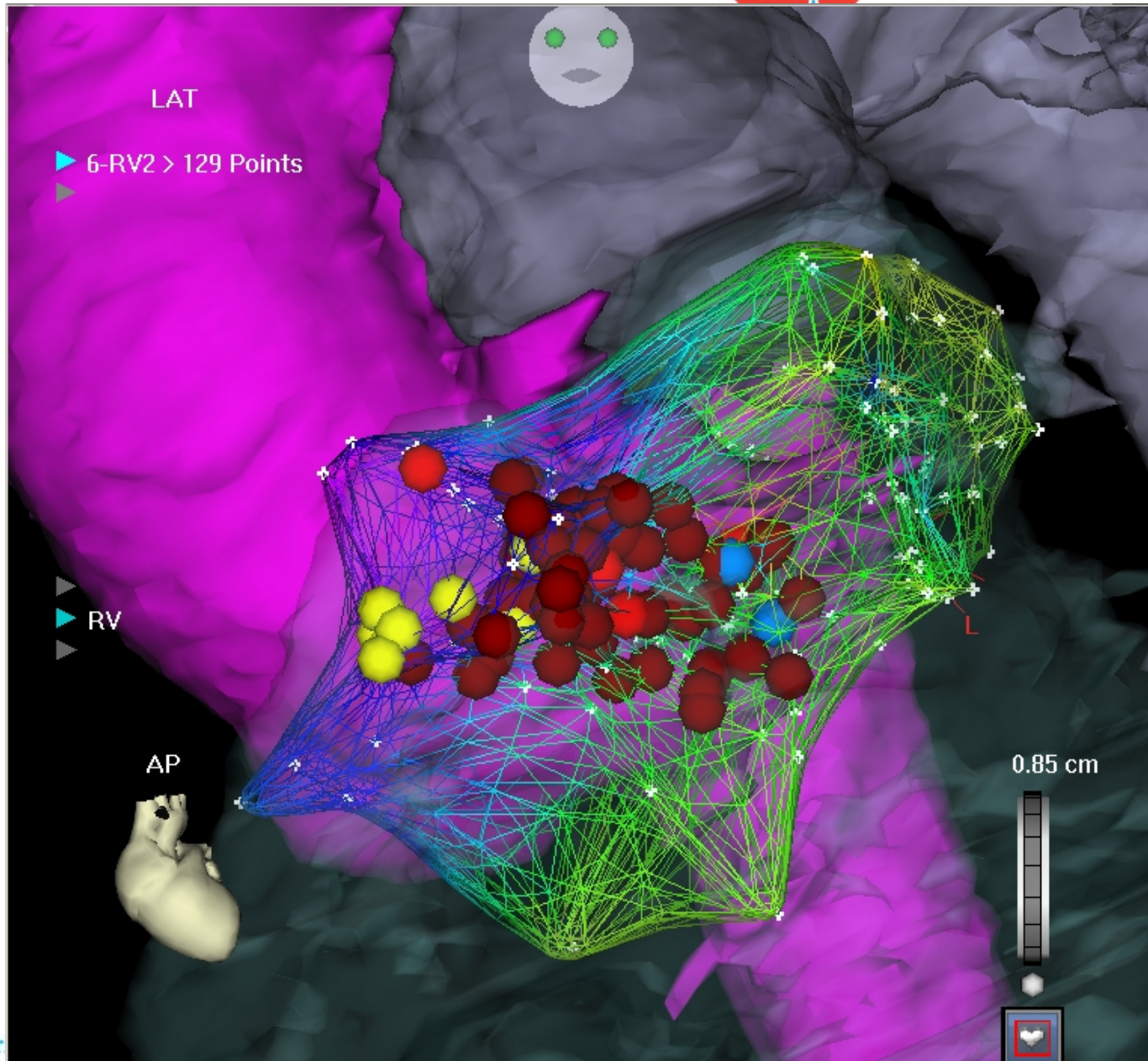
CL = 340 ms

**PACE-MAP (BLUE POINTS)**





### 3. Pace mapping strategy

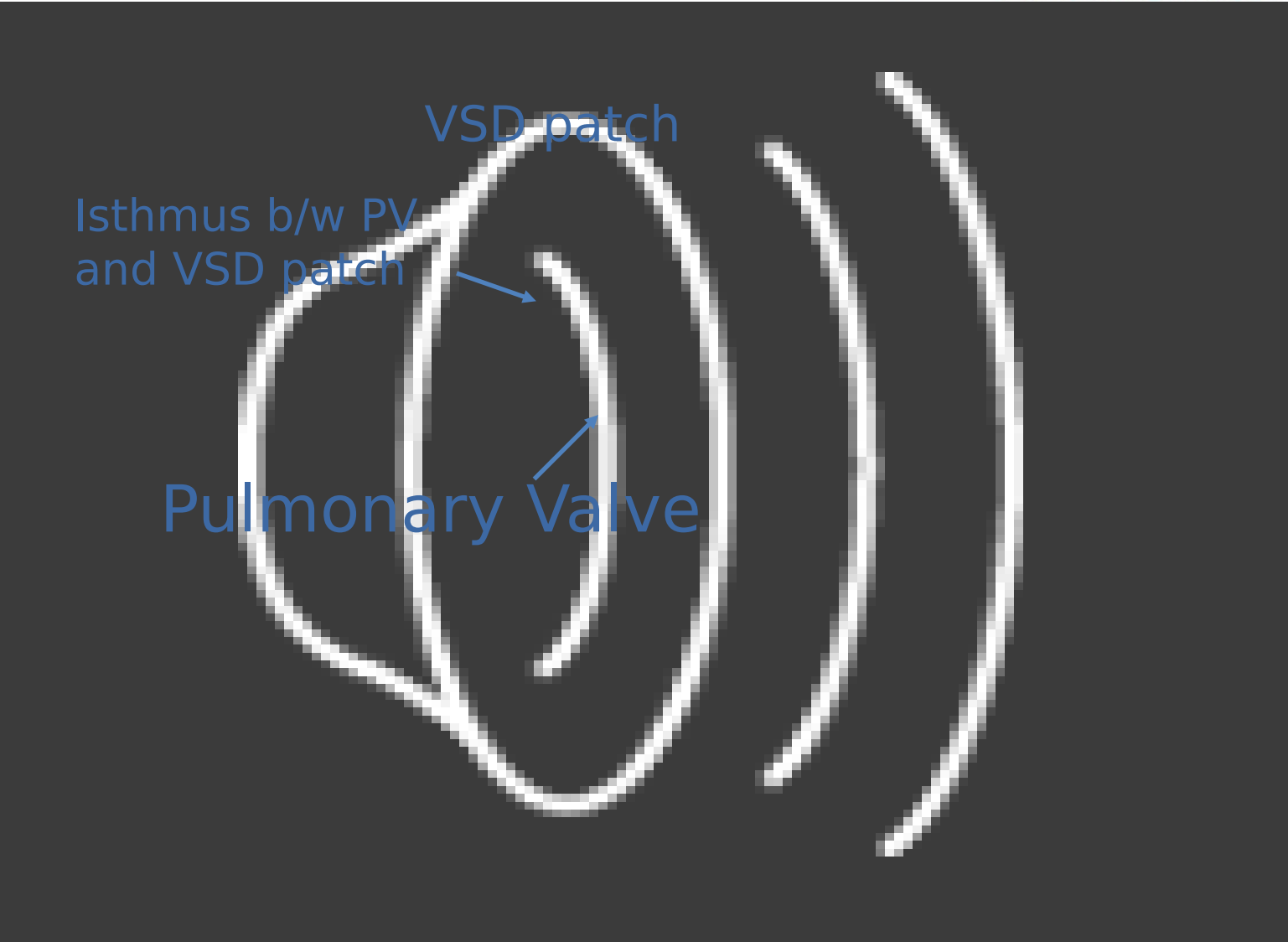




## 4-MERGING WITH CARTOSOUND



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VSD patch

Isthmus b/w PV  
and VSD patch

Pulmonary Valve

1-RV (0, 19)

LAT



PA

Pulmonary valve

Isthmus b/w  
PV and VSD

LVOT

VSD patch

Right Ventricle



1.34

+

-

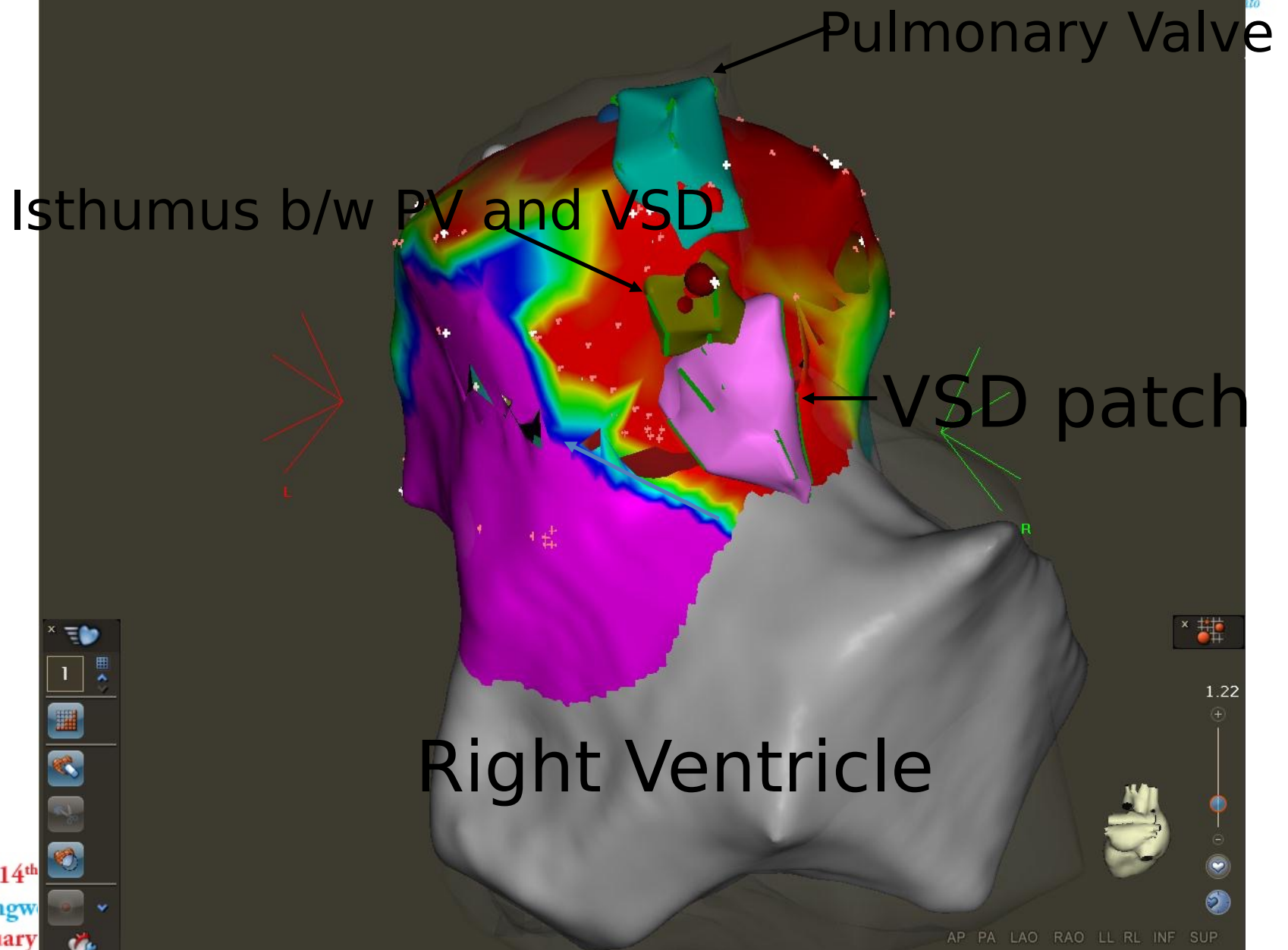
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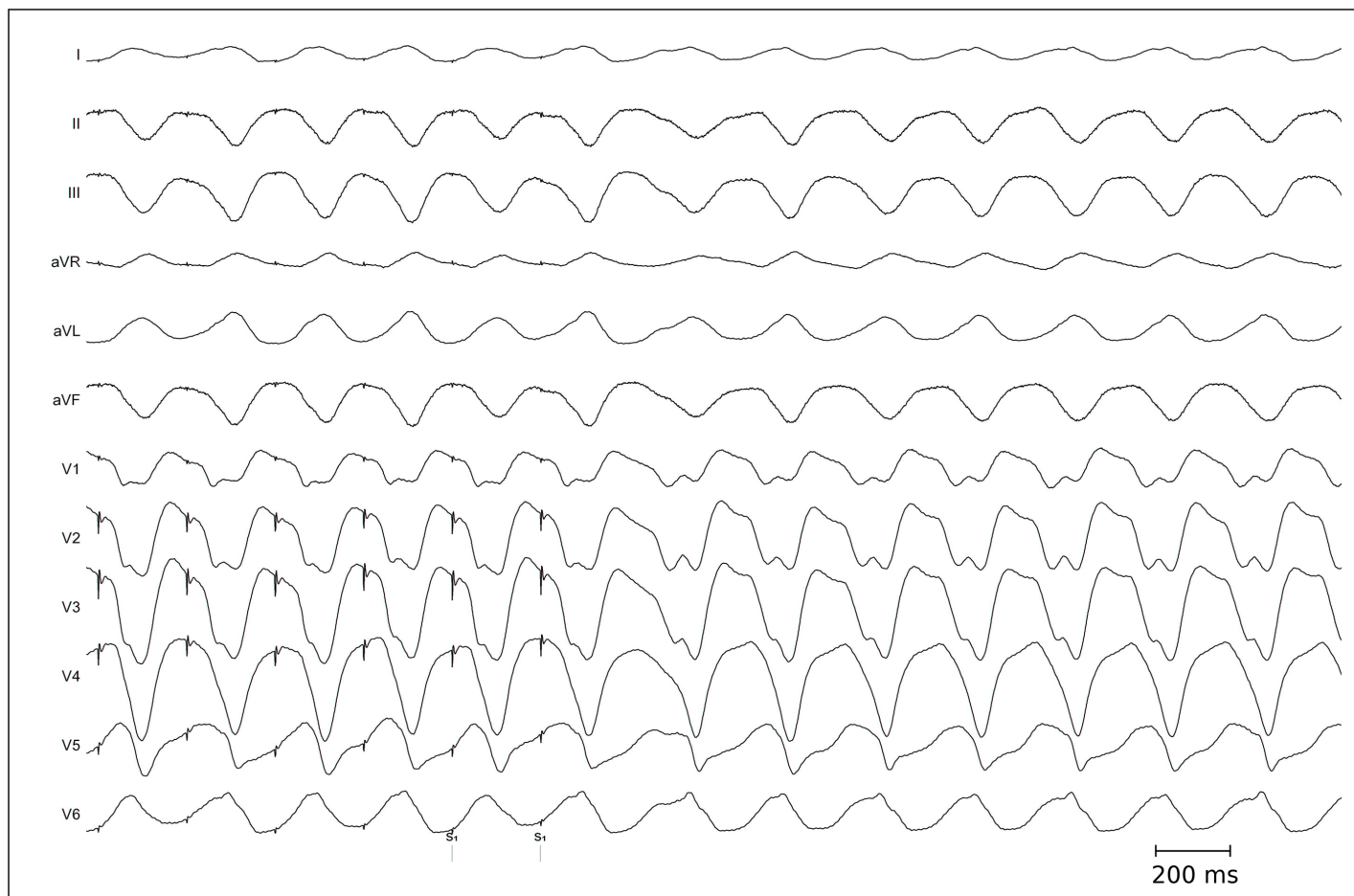
AP PA LAO RAO LL RL INF SUP



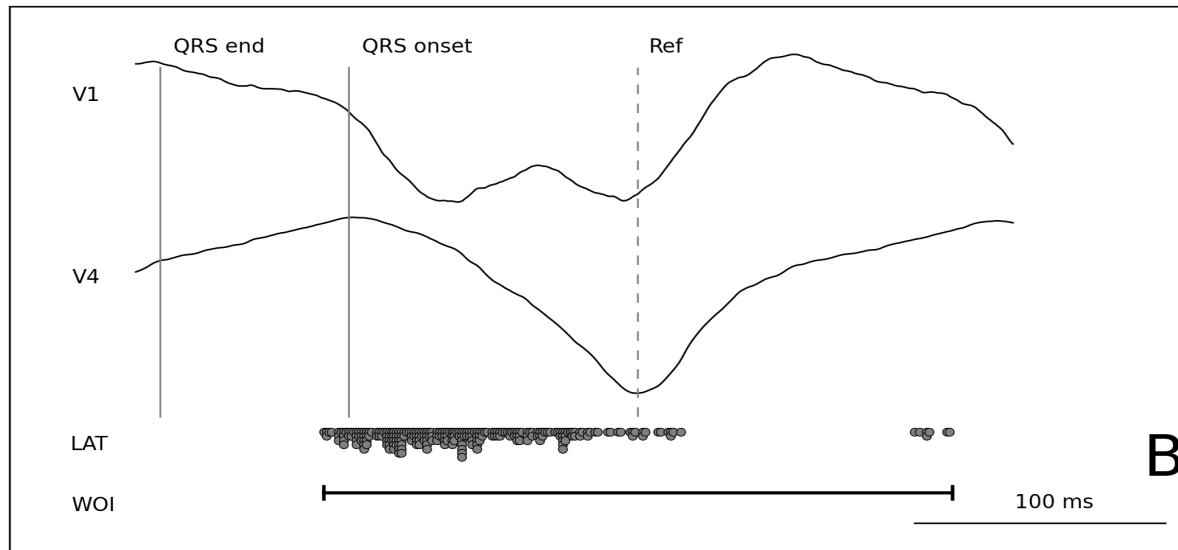
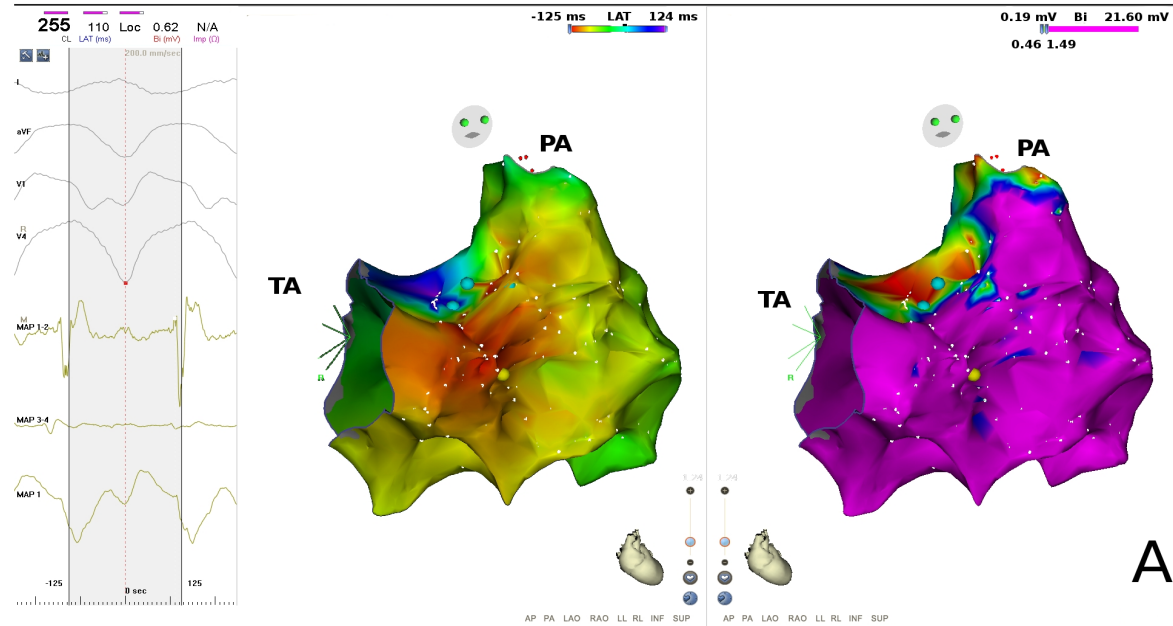
## 5. ACTIVATION MAPPING STRATEGY: CHALLENGE OF MAPPING A FAST TACHYCARDIA WITH A BROAD QRS



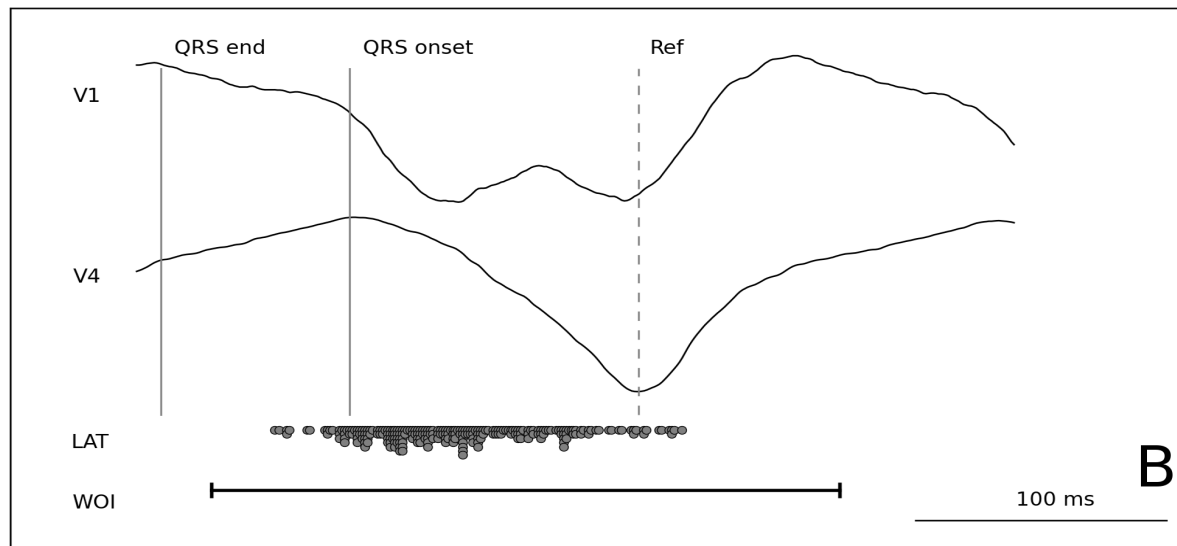
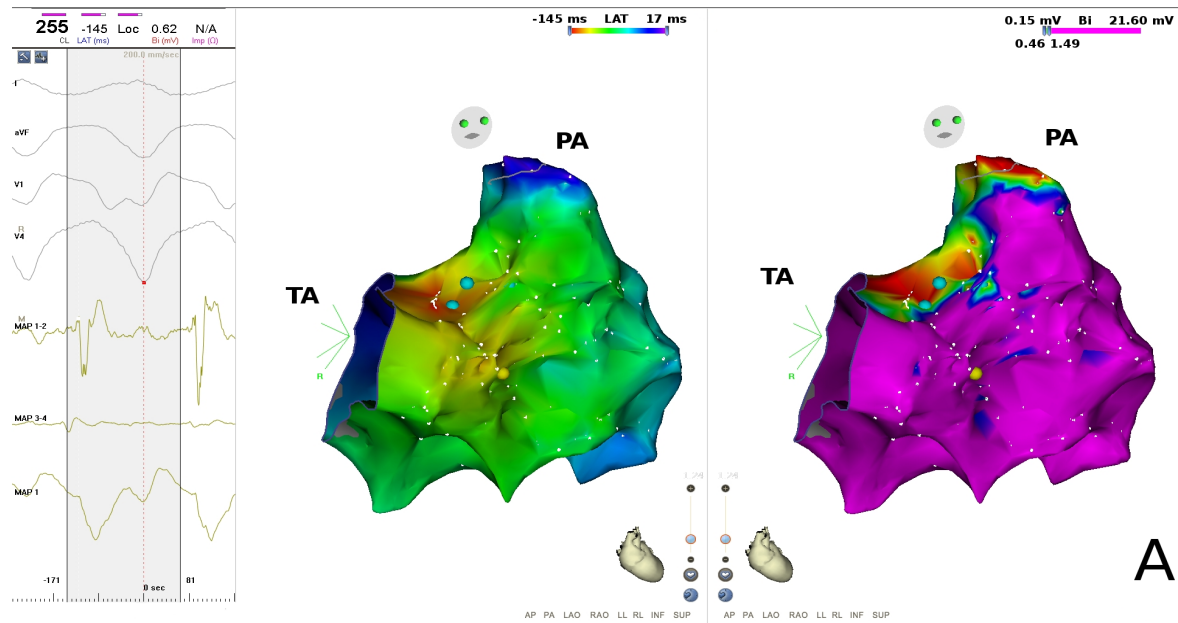
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- Selvaraj R, Krishnakumar Nair. Chasing 'Red' Herrings: Making Sense of the Colors while Mapping. 2014 Feb 6. **Circulation: Arrhythmia And Electrophysiology.**



- Selvaraj R, Krishnakumar Nair. Chasing 'Red' Herrings: Making Sense of the Colors while Mapping. 2014 Feb 6. Circulation: Arrhythmia And Electrophysiology.



- Selvaraj R, Krishnakumar Nair. Chasing 'Red' Herrings: Making Sense of the Colors while Mapping. 2014 Feb 14. Circulation: Arrhythmia And Electrophysiology.



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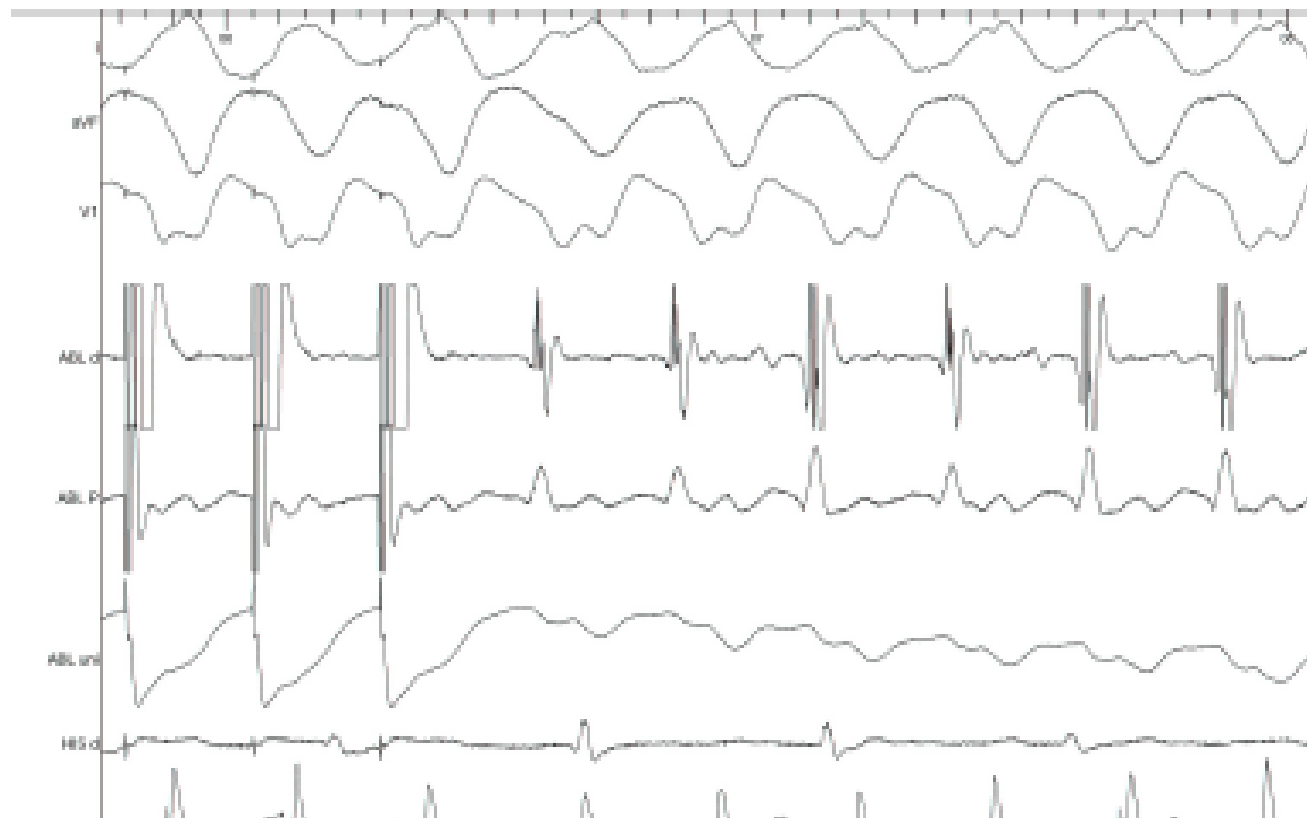
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## 6. CONCEALED ENTRAINMENT AND PPI-TCL=30

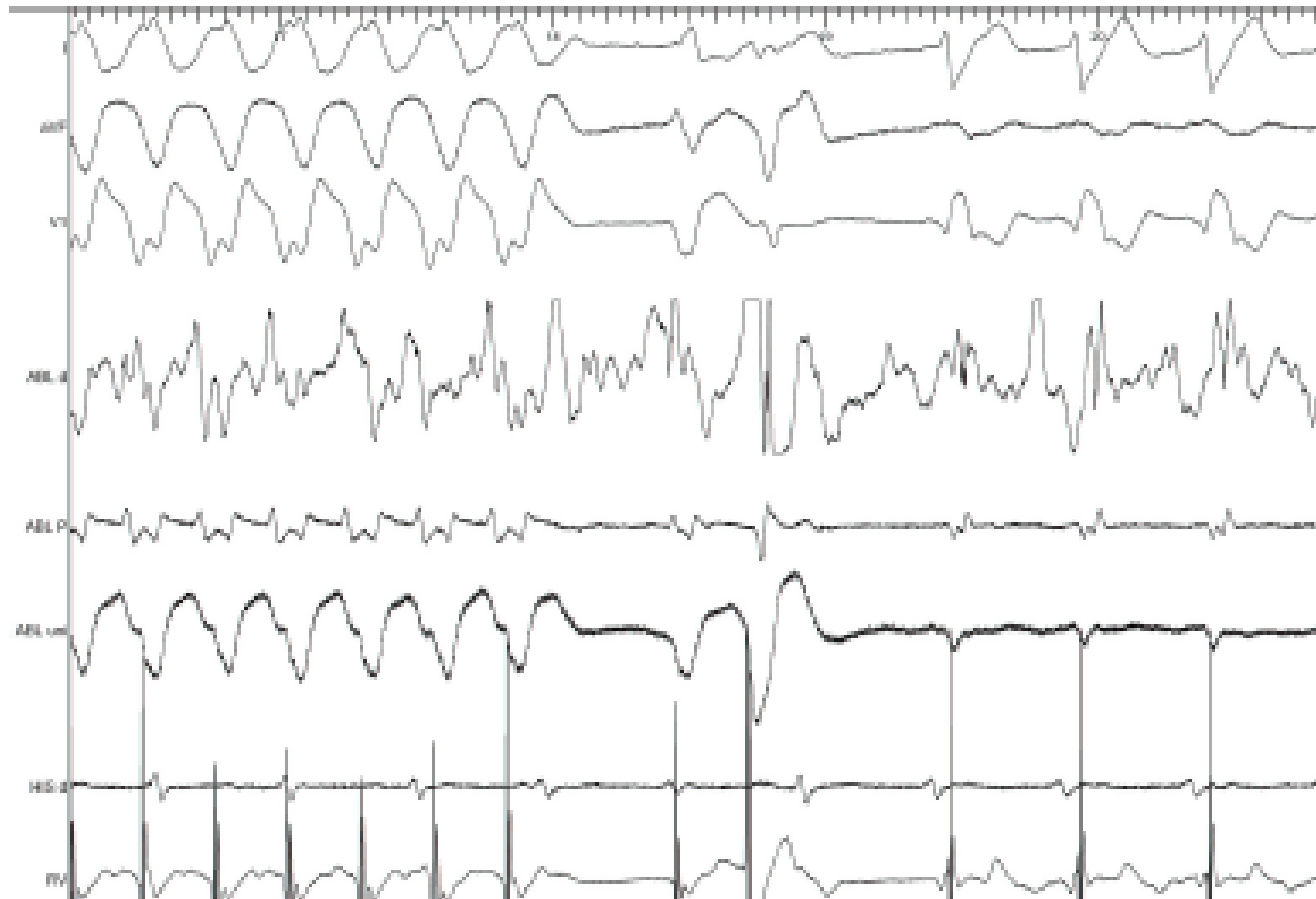
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# VT Termination on RF#2





# Summary

- ▶ Our understanding of ventricular arrhythmias in ACHD continues to grow and so too will our ability to better risk stratify these patients in the future.
- ▶ Critical isthmuses in VT ablation in patients with repaired TOF have been identified



# SUMMARY

Novel techniques and combination of techniques may be necessary to achieve success:

- Including activation mapping, pace mapping with Voltage mapping, and Entrainment mapping
- Studying EGM morphology and pattern on multielectrode catheters
- Re-annotation of signals and Re-setting windows of interest on CARTO
- Use of ICE and CARTO -SOUND



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# THANK YOU

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