



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

Ventricular Arrhythmias in Adult Congenital Heart Disease

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Objectives



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- 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

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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.

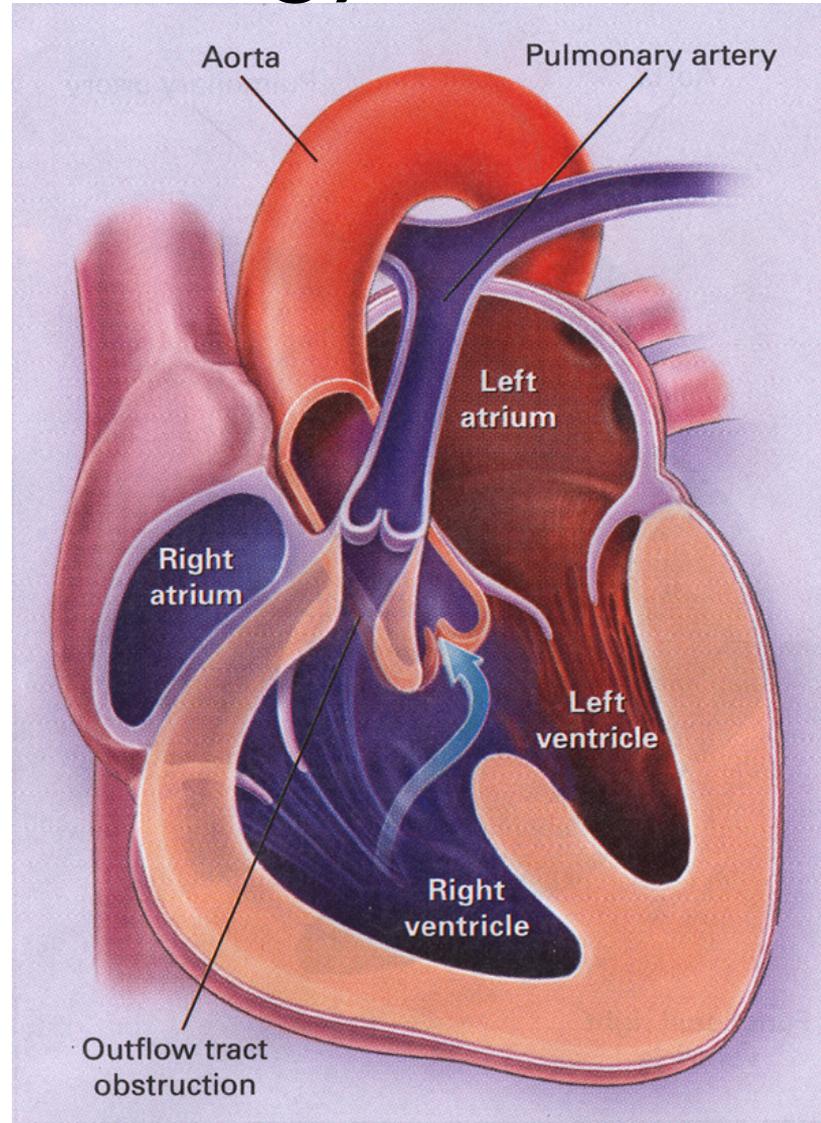


Major features (contd.)

- 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 ≥ 180 msec on ECG, or a QRS duration that increases at a rate of > 3 msec/year
- Tricuspid regurgitation

Value of Programmed Ventricular Stimulation



- 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:

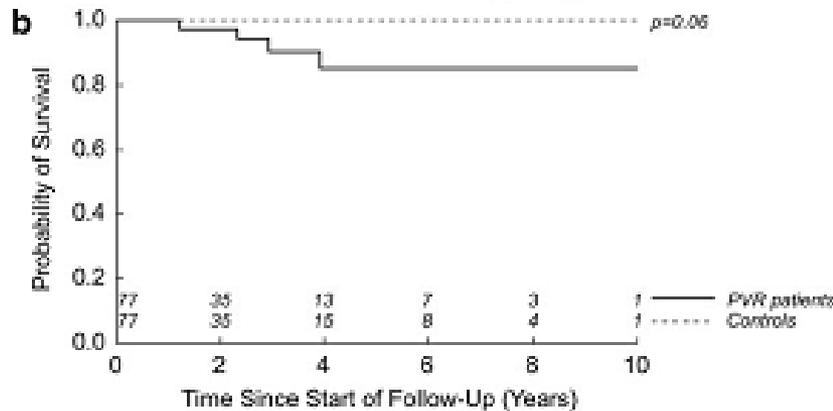
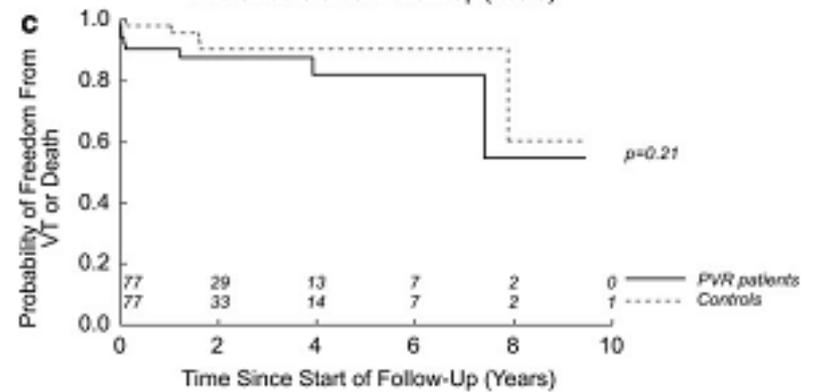
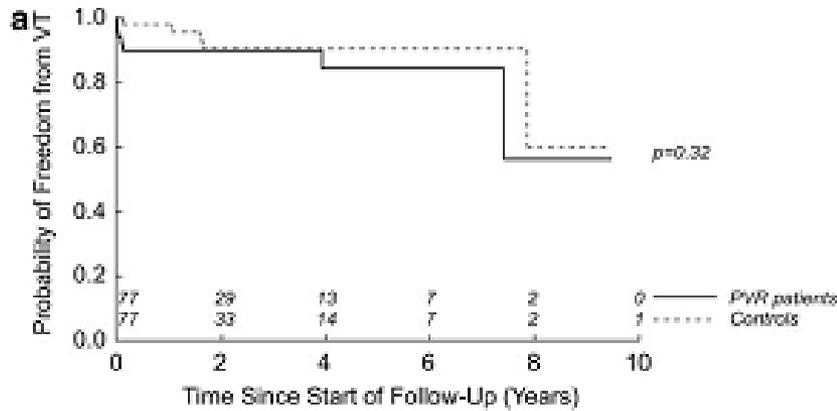
Moderate-to-severe RV dysfunction

RV enlargement (e.g., right ventricular end-diastolic volume >170 ml/m²)

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

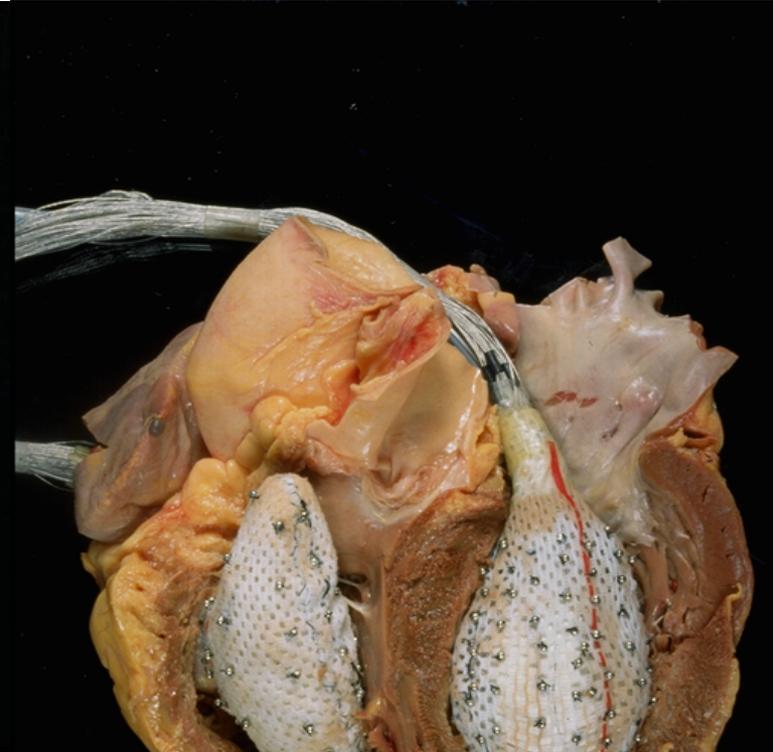
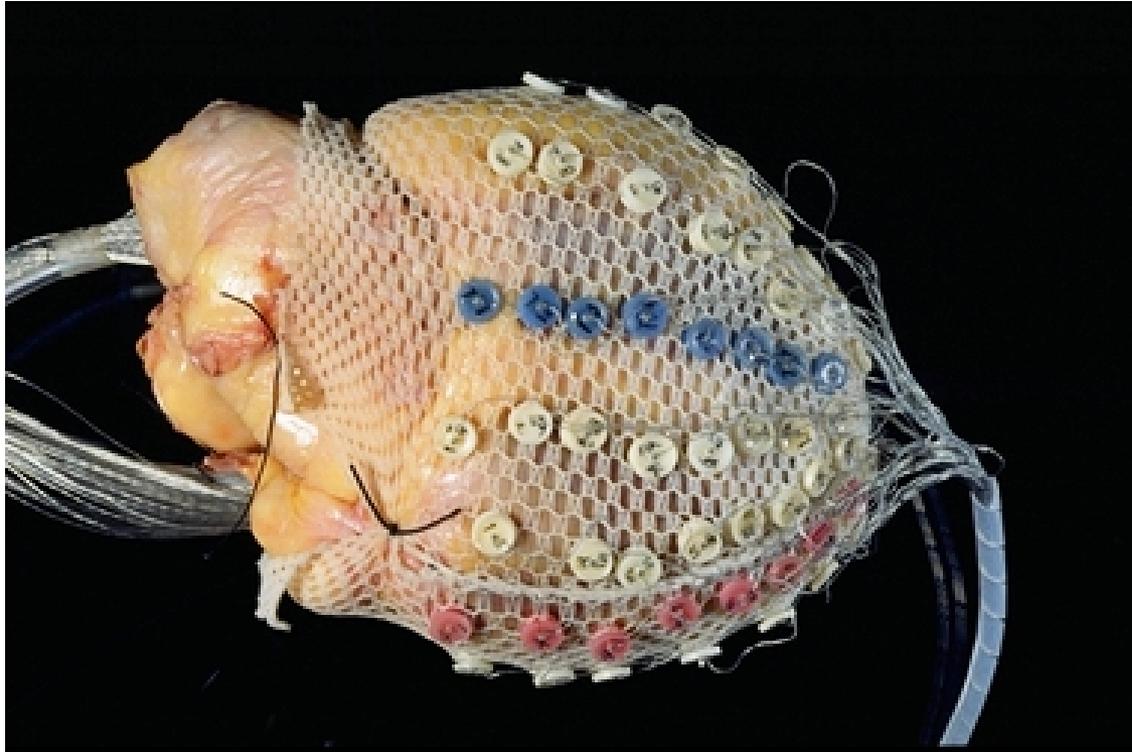
Circulation . 2009;119:445-451.

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

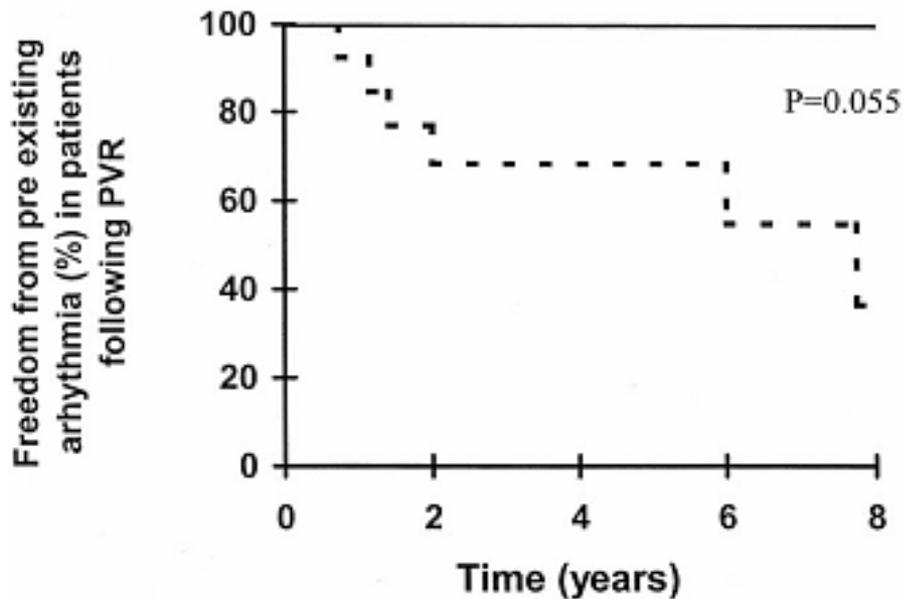


Intraoperative Ablation



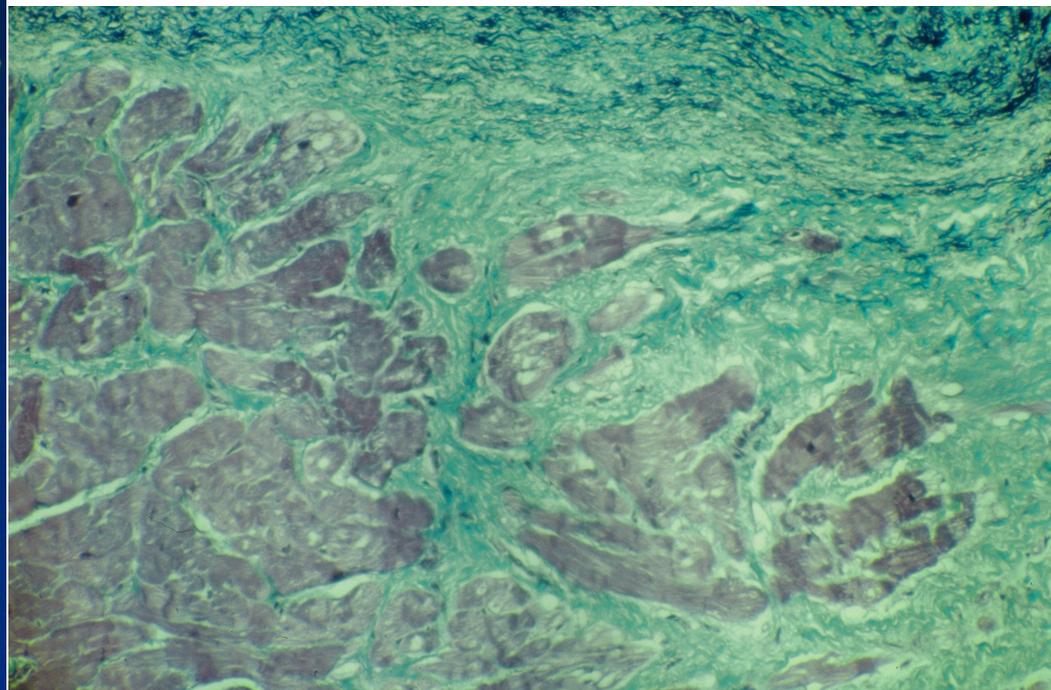
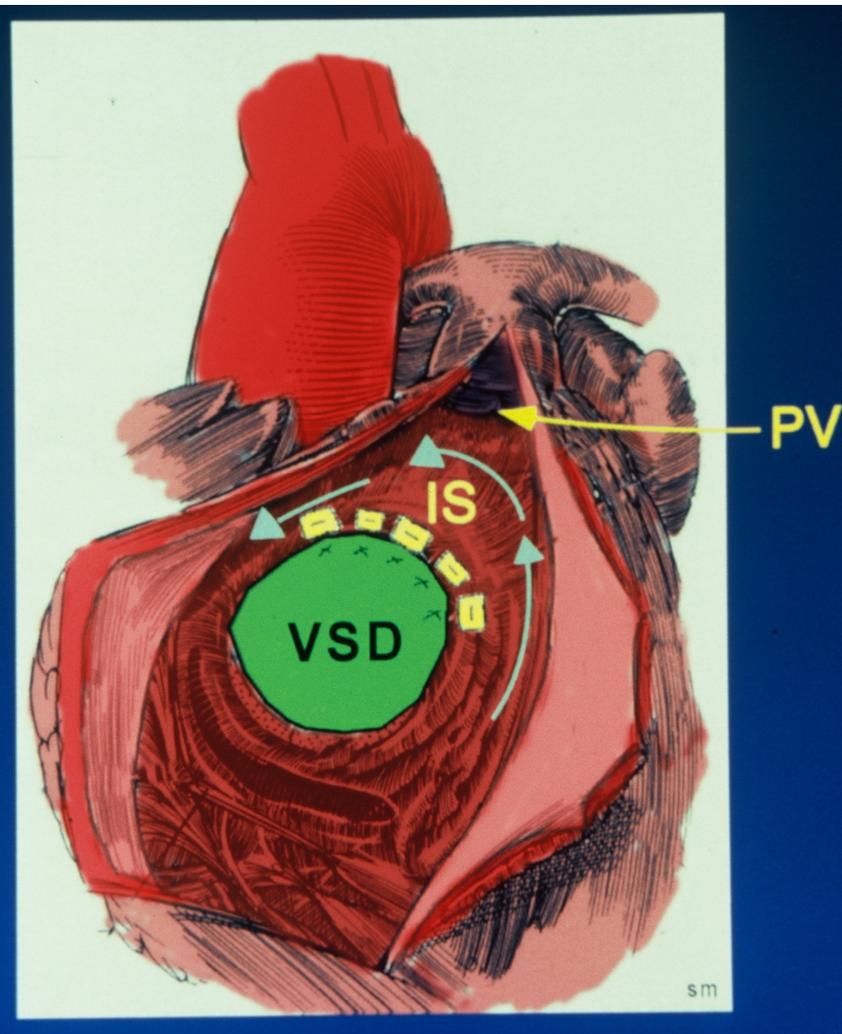


Pulmonary Valve Replacement with concomitant intraoperative ablation therapy



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

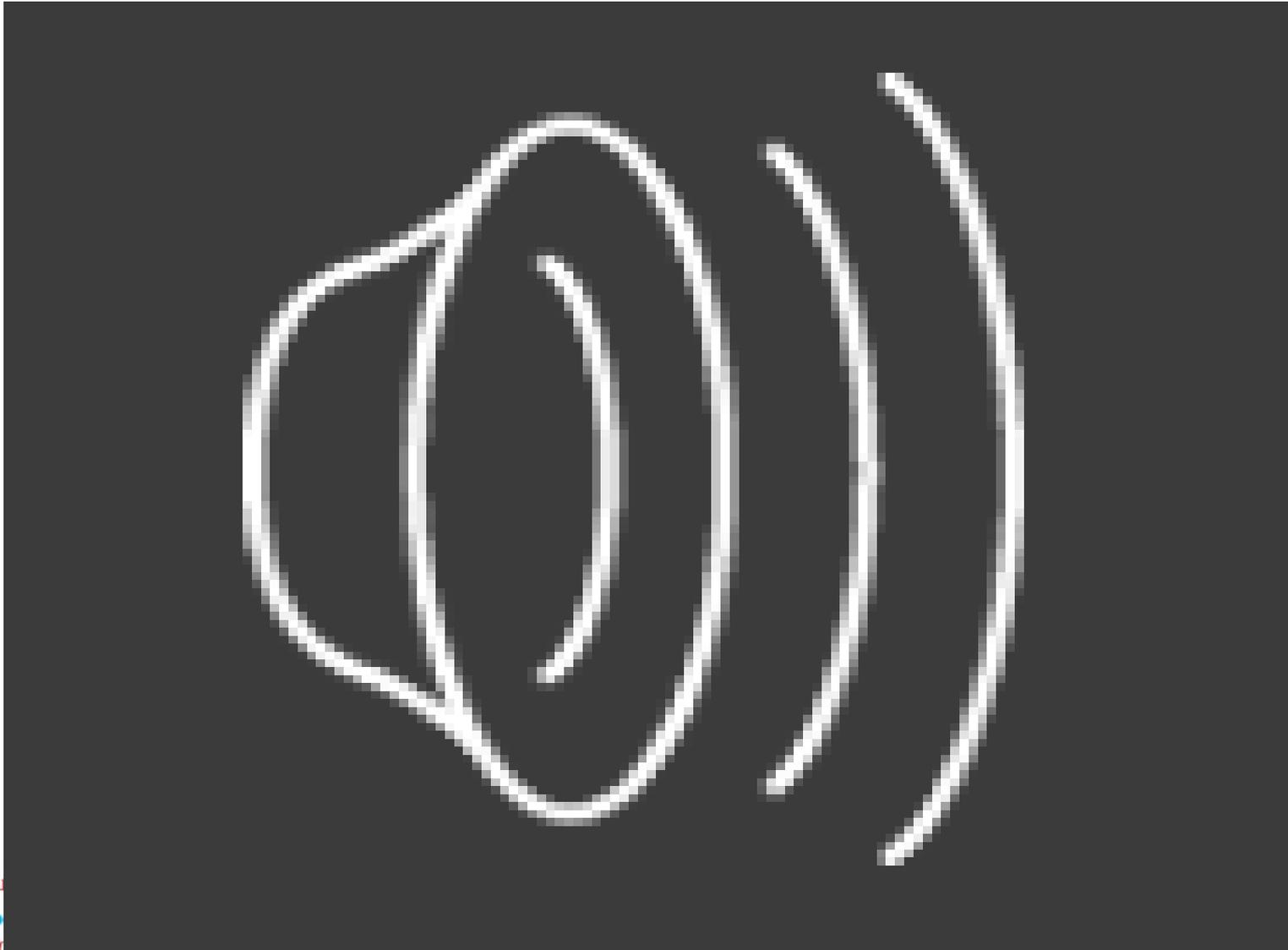
Circulation . 2001;103:2489-2494



1. FLASHING LIGHT DISPLAY



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2. ELECTROGRAM ANALYSIS



500 ms

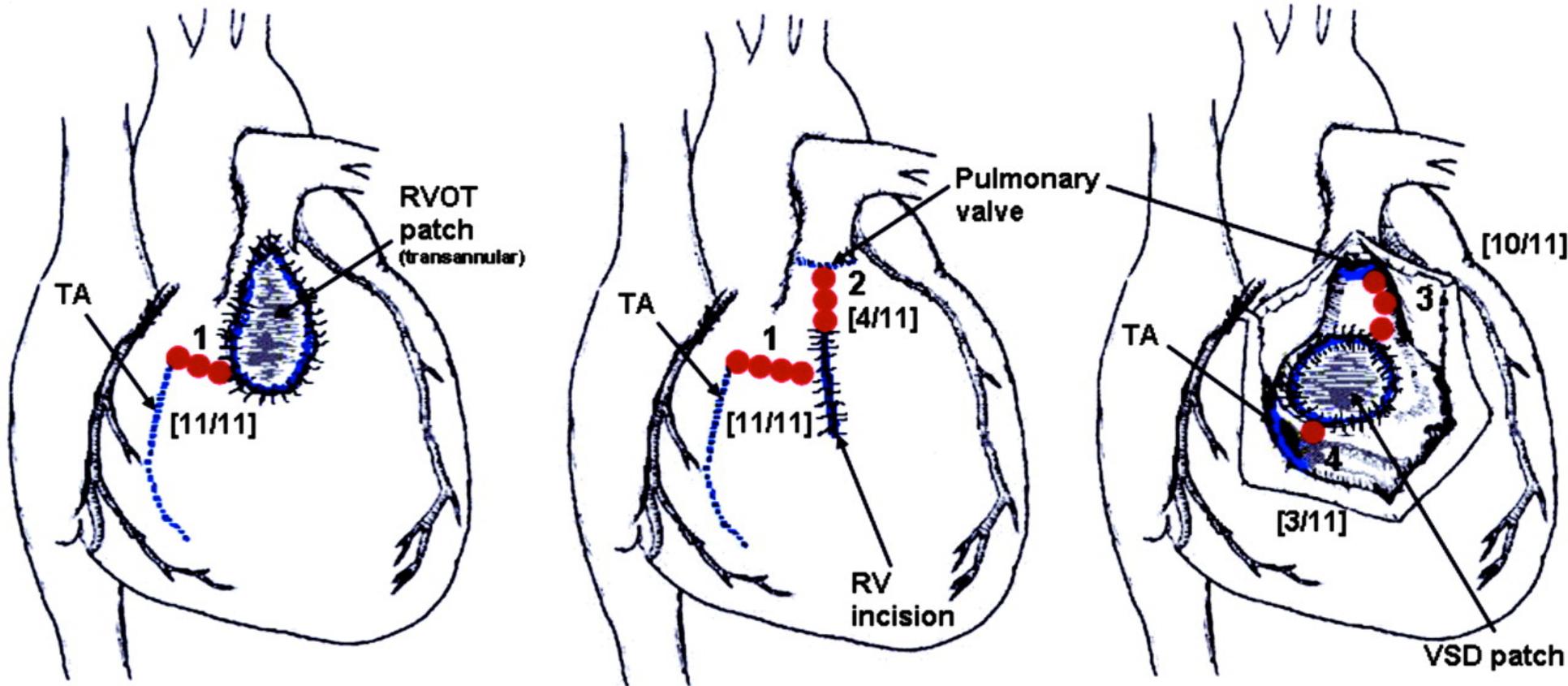


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CATHETER ABLATION

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4 ANATOMIC ISTHMUSES



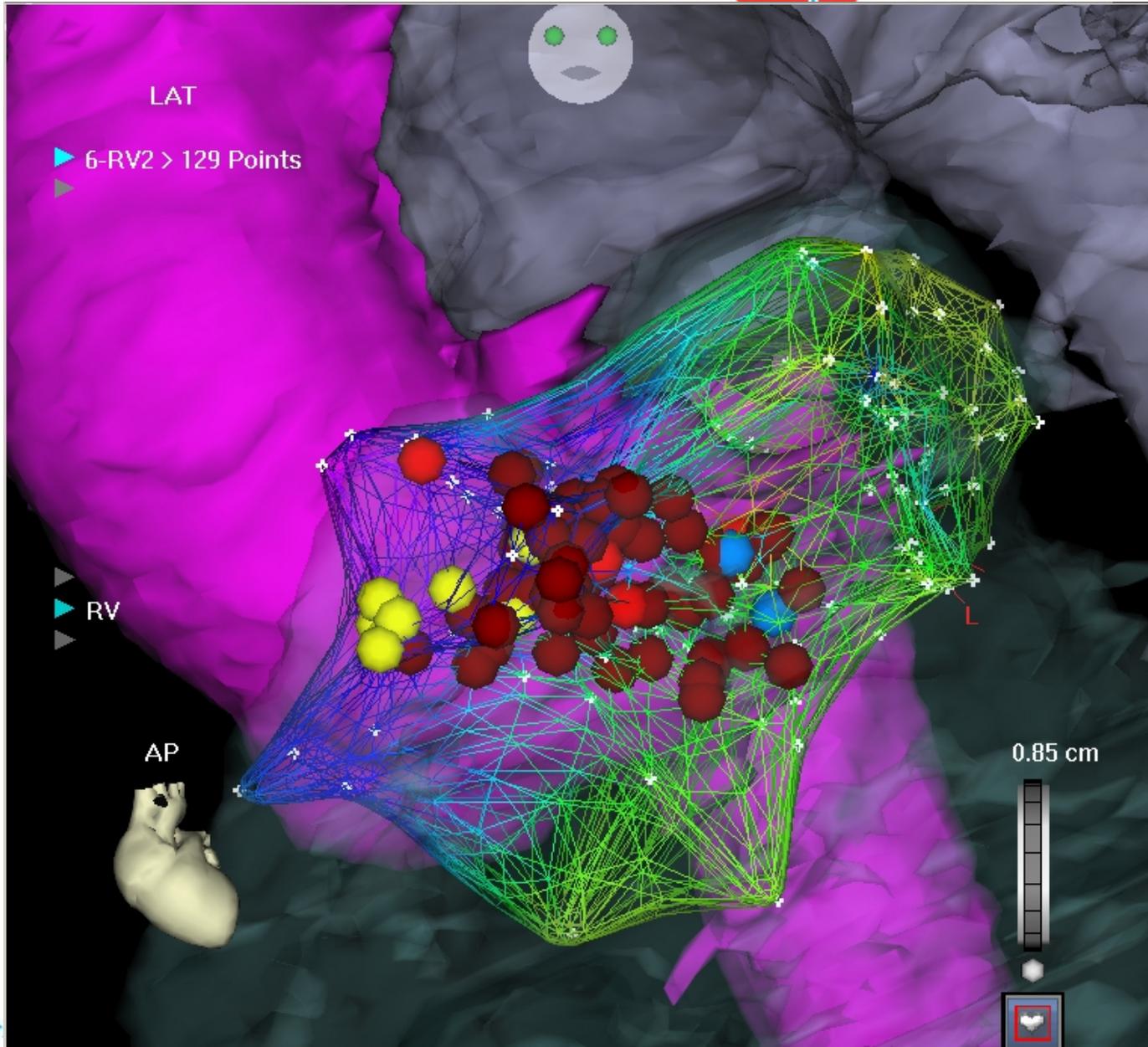
Schematic of the localization of anatomic boundaries (blue lines) for VT after repair of CHD and the resulting anatomic isthmuses (red lines); frequency of the distinct isthmuses in brackets.

**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 ± 29.3 months of follow-up, 91% of patients remained VT free.

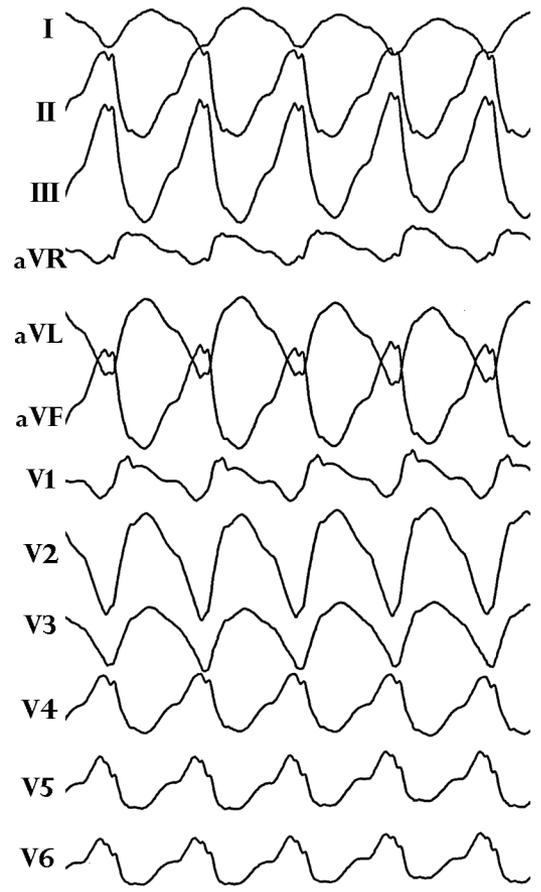
3. PACE MAPPING STRATEGY



-PACE MAPPING STRATEGY

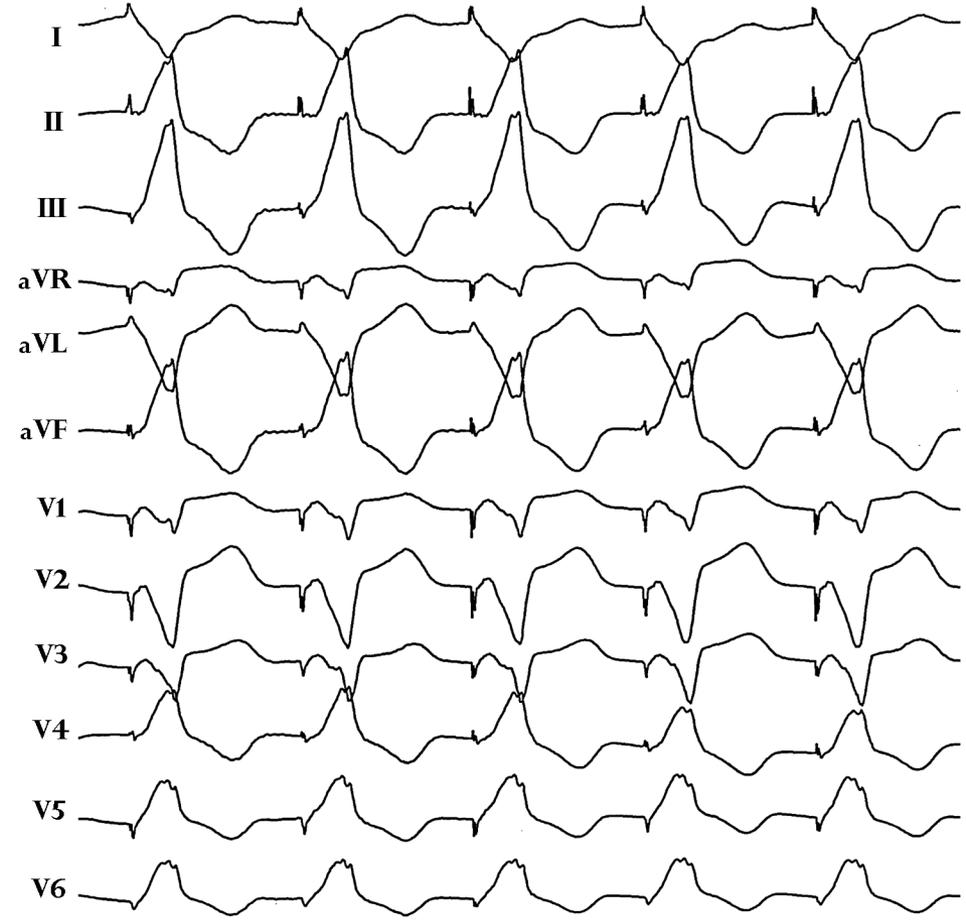


CLINICAL VT

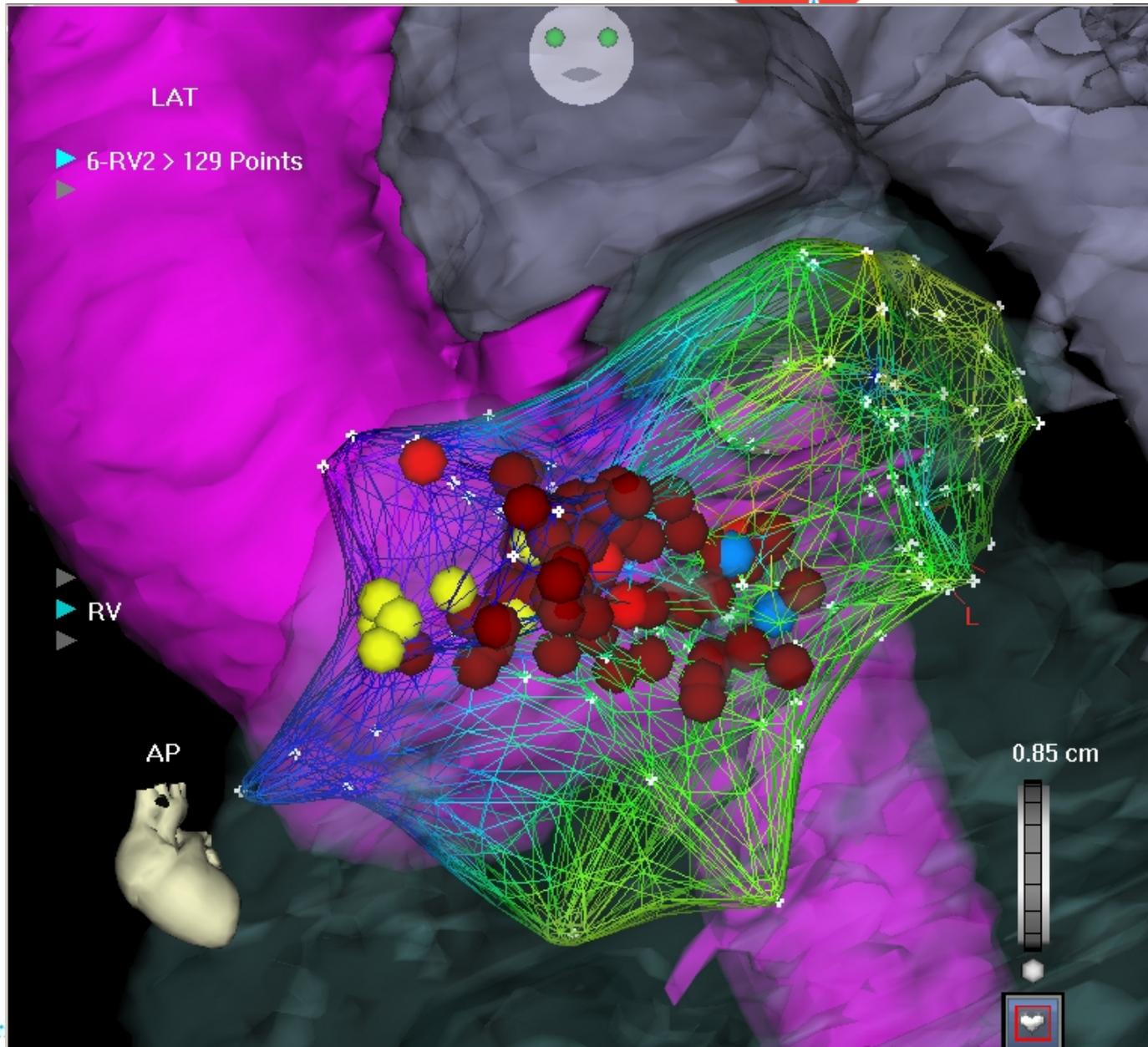


CL = 340 ms

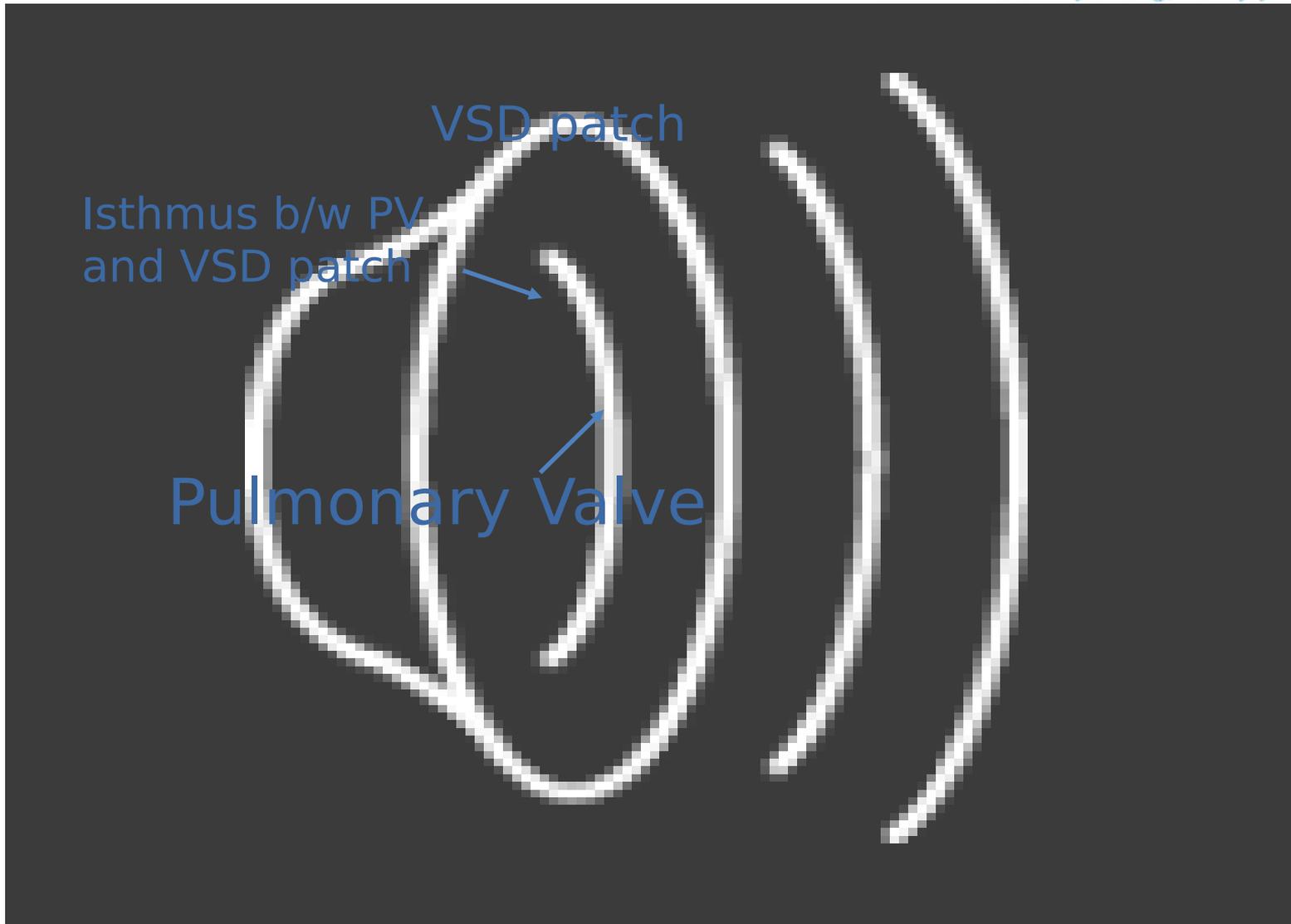
PACE-MAP (BLUE POINTS)



3. Pace mapping strategy



4-MERGING WITH CARTOSOUND



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PA

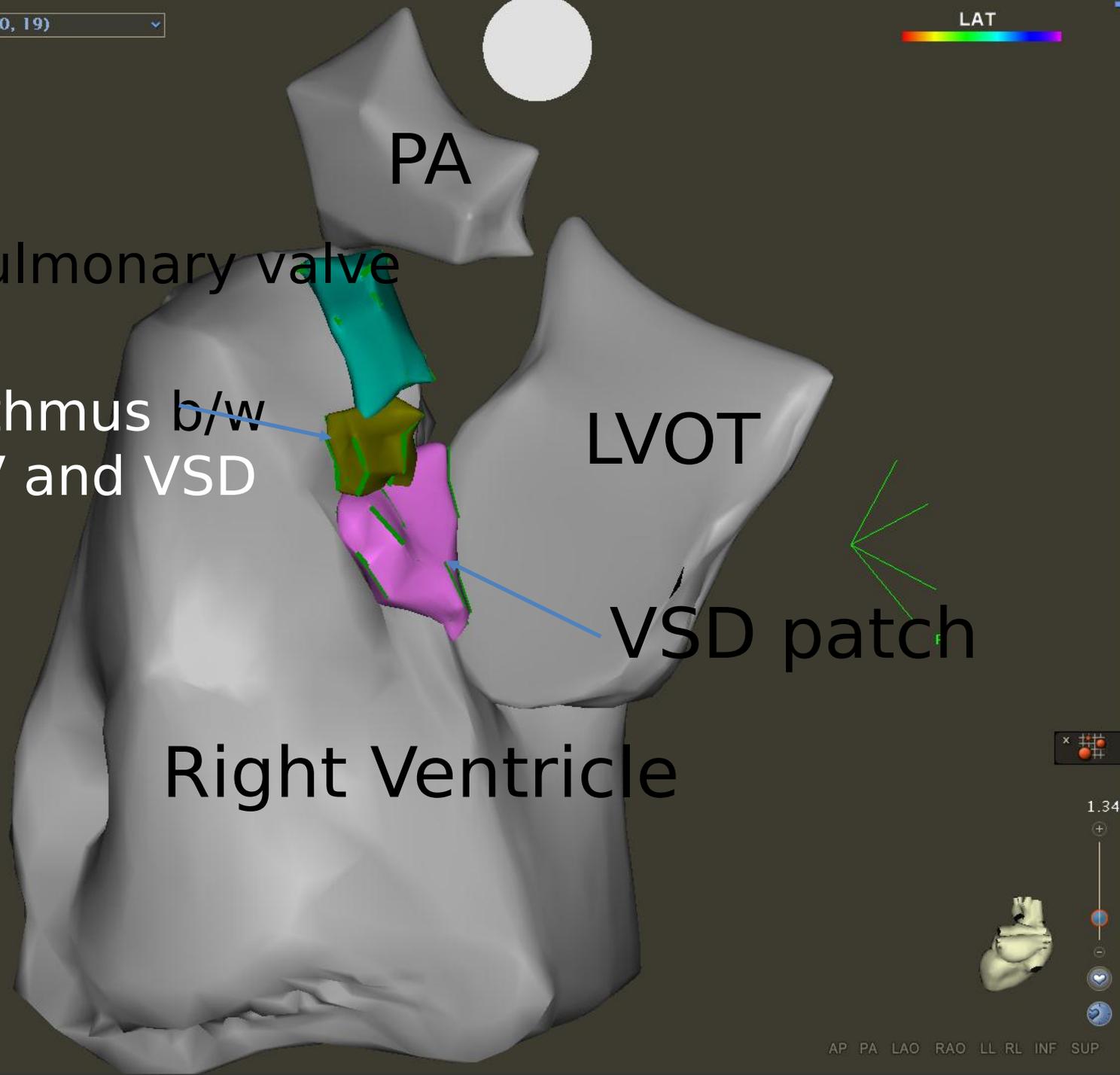
Pulmonary valve

Isthmus b/w
PV and VSD

LVOT

VSD patch

Right Ventricle



Pulmonary Valve
Isthmus b/w PV and VSD

VSD patch

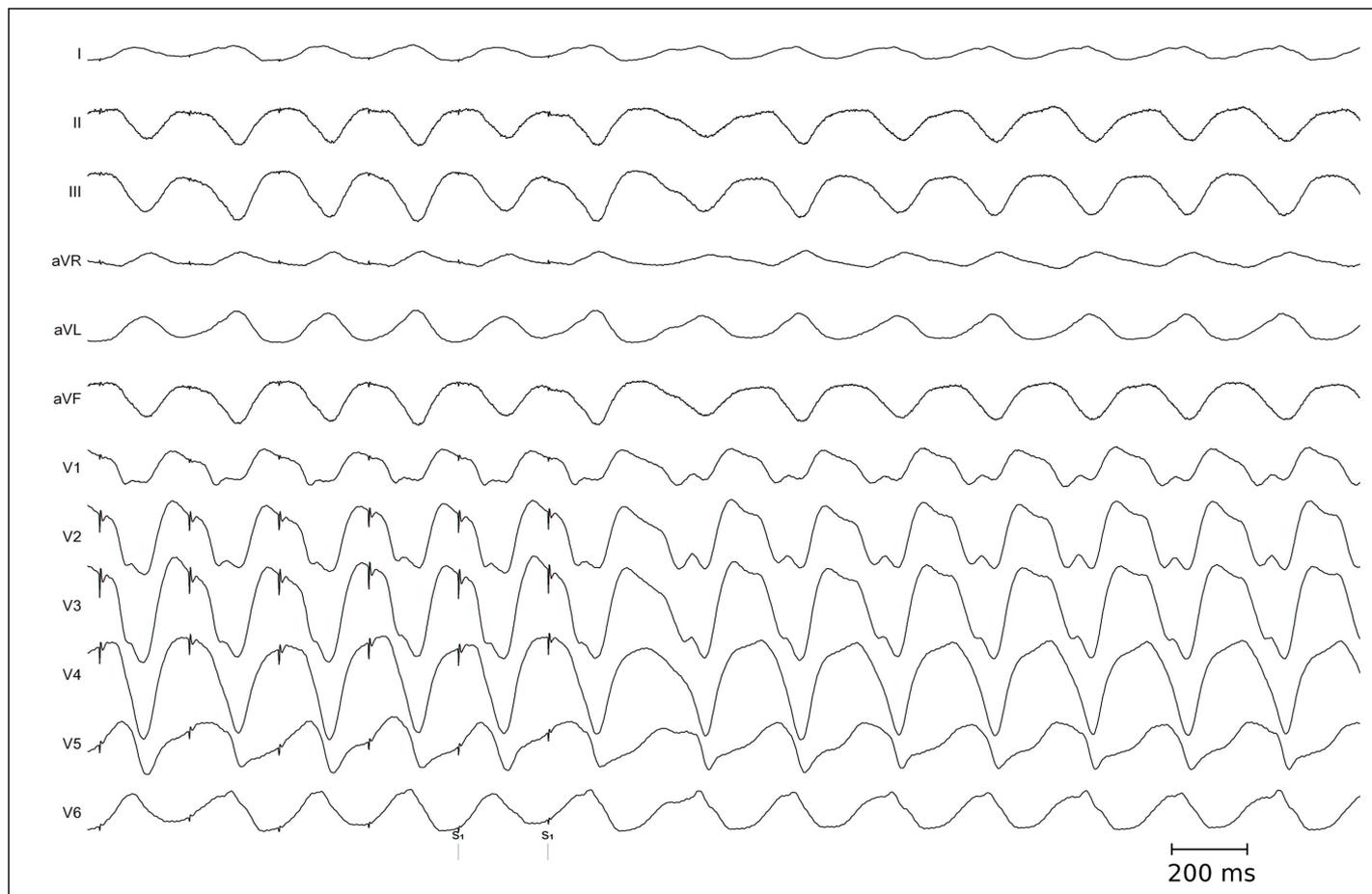
Right Ventricle



1.22



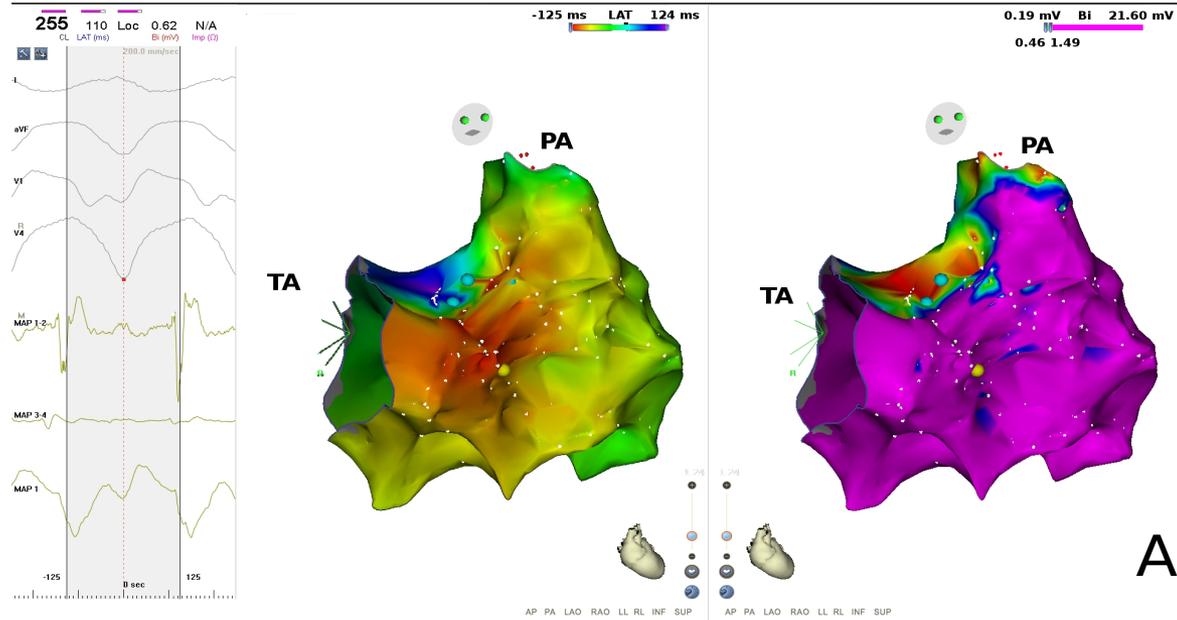
5. ACTIVATION MAPPING STRATEGY: CHALLENGE OF MAPPING A FAST TACHYCARDIA WITH A BROAD QRS



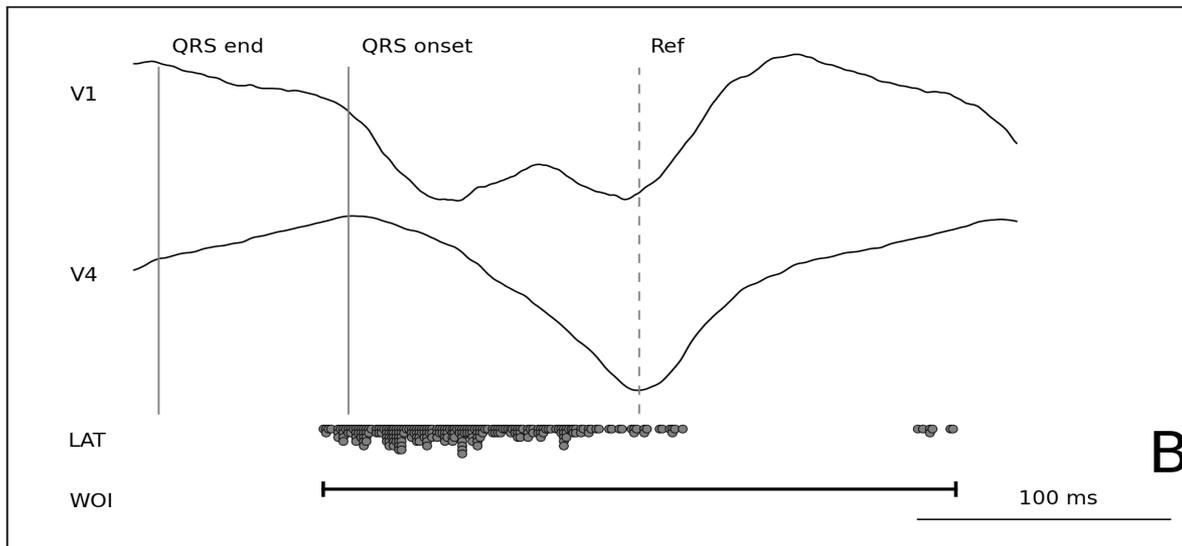
- Selvaraj R, Krishnakumar Nair. Chasing 'Red' Herrings: Making Sense of the Colors while Mapping. 2014 Feb 6. **Circulation: Arrhythmia And Electrophysiology.**

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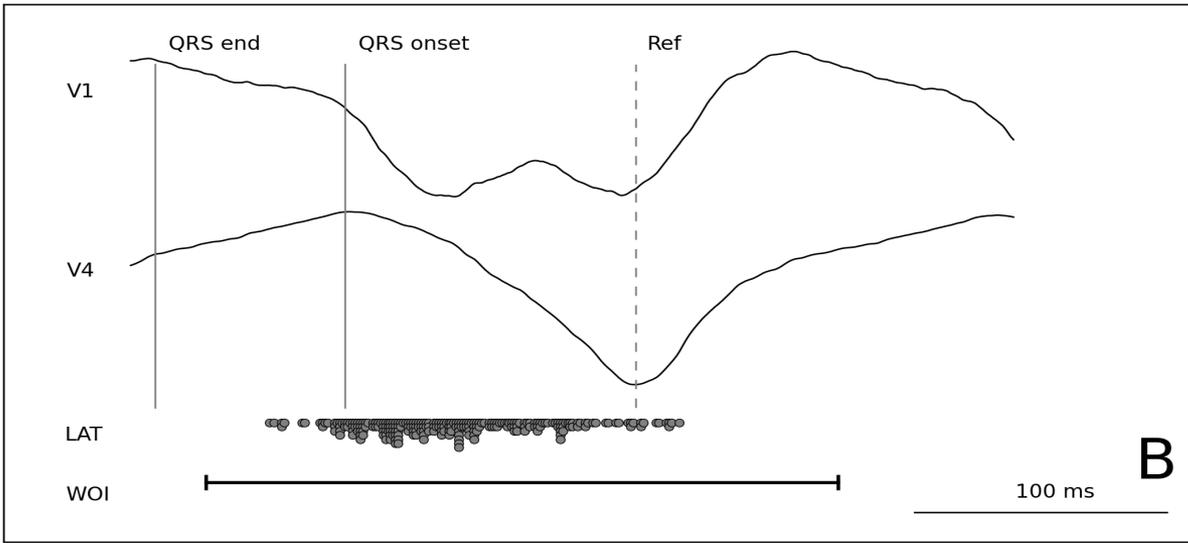
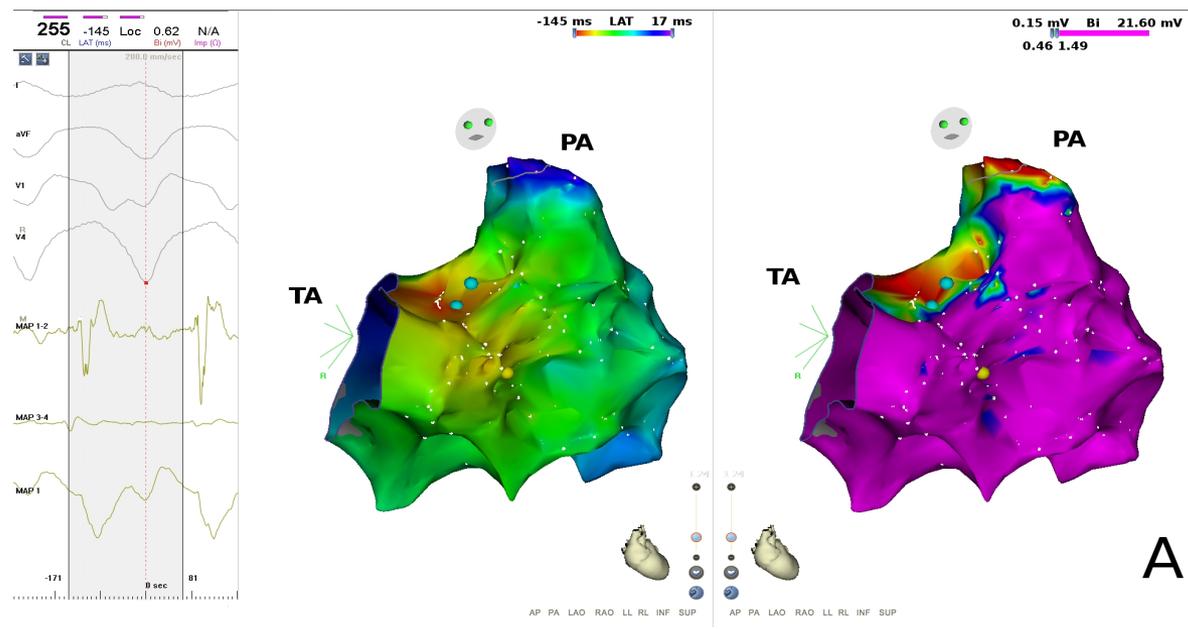


A



B

- Selvaraj R, Krishnakumar Nair. Chasing 'Red' Herrings: Making Sense of the Colors while Mapping. 2014 Feb 14. **6. Circulation: Arrhythmia And Electrophysiology.**

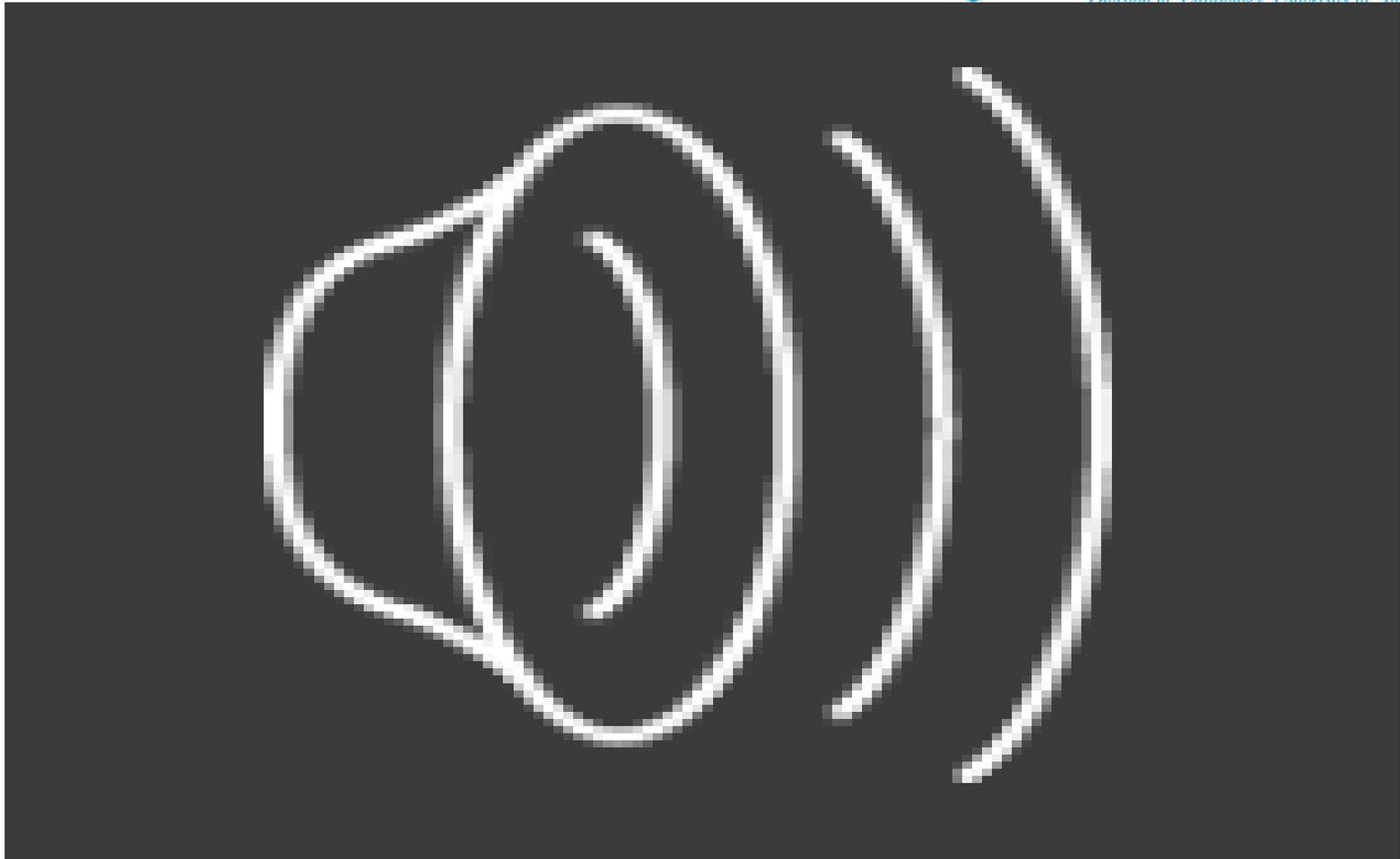


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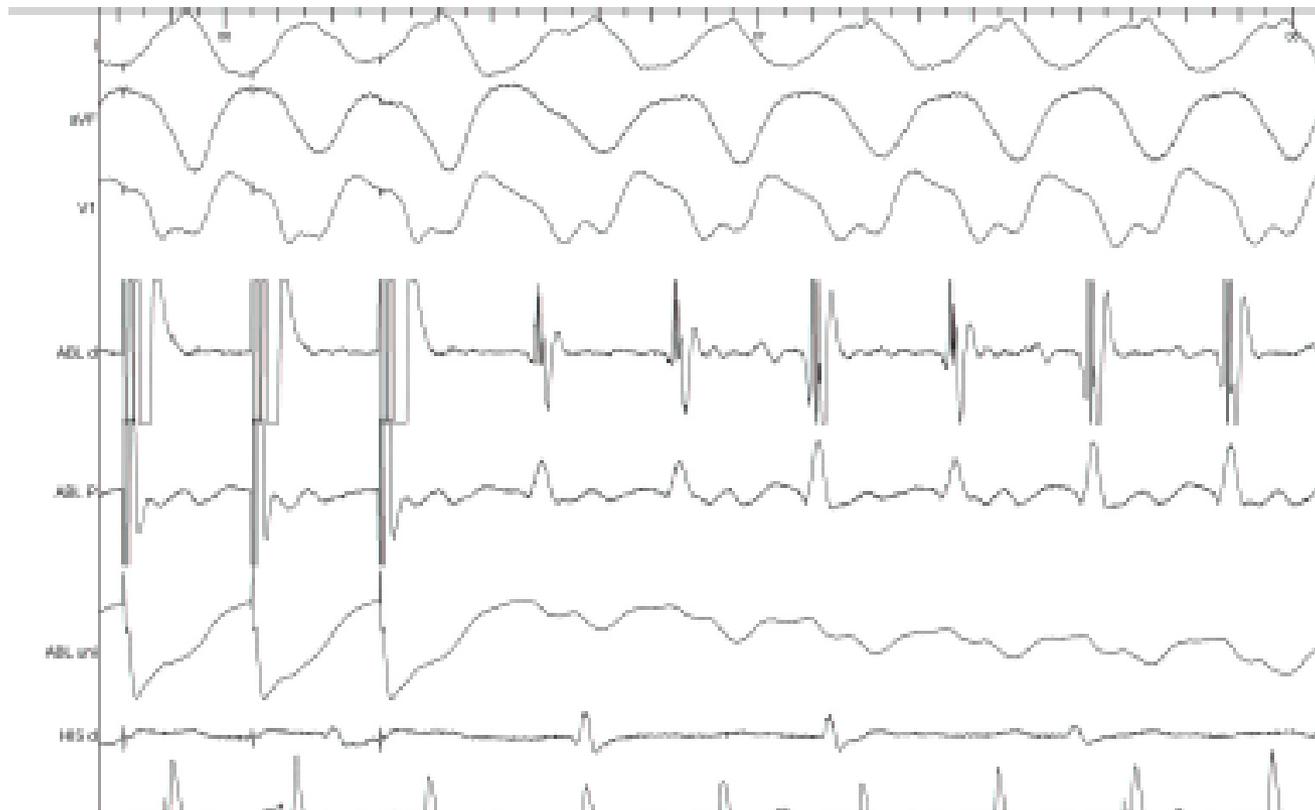
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6. CONCEALED ENTRAINMENT AND PPI-TCL=30

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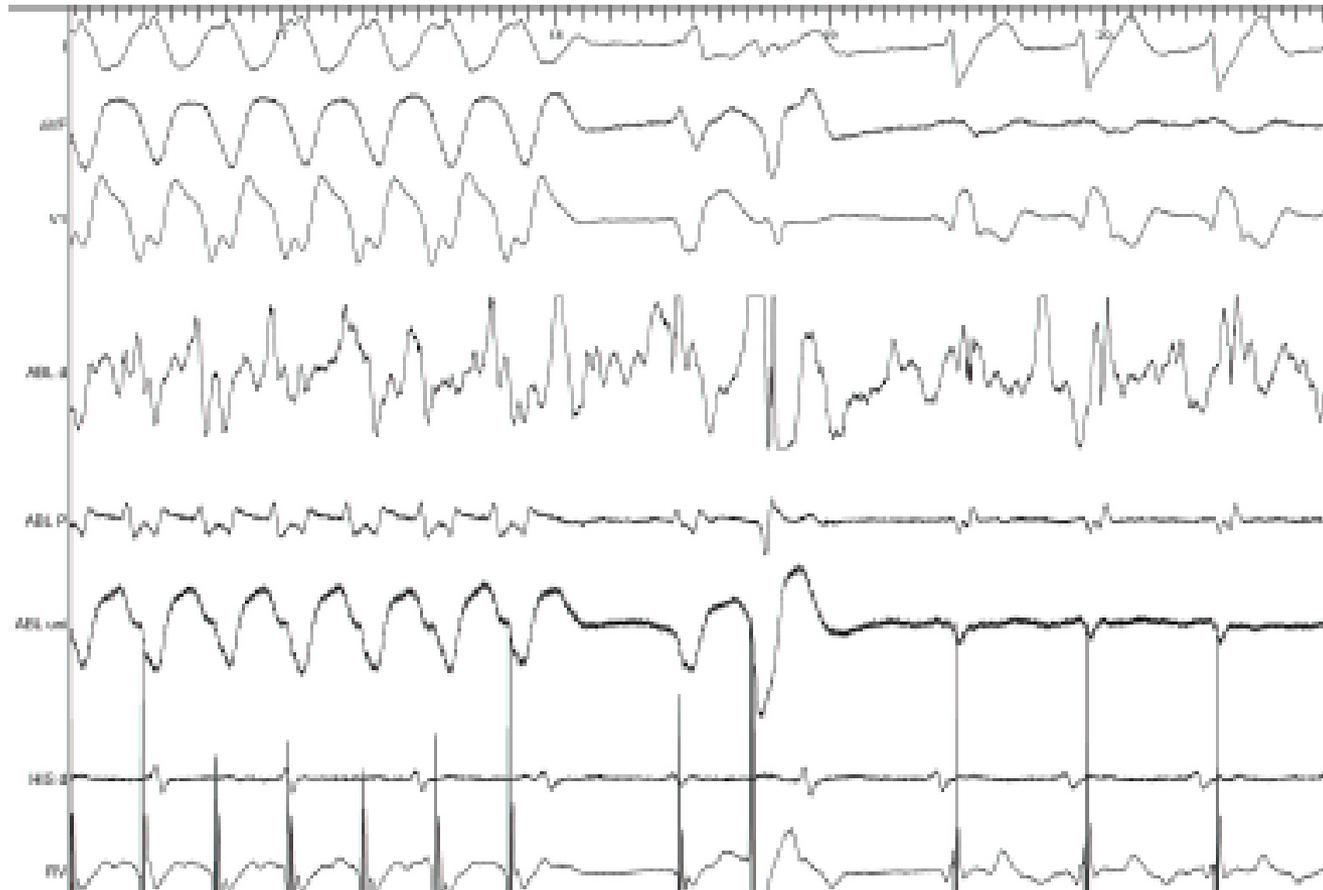
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VT Termination on RF#2



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Summary



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- ▶ 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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