9th 'Utrecht Sessions' on Atrioventricular Septum Defects
1st - 3rd February 2024

Utrecht Sessions

European Reference Network
for rare or low prevalence complex diseases

Network
Heart Diseases
(ERN GUARD-HEART)

Member
Motol University
Hospital — Czechia

Treating arrhythmias in AVSD
From brady to tachycardia

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FN MOTOL

Complexity of CHD	Type of CHD	Prevalence (in CHD population)	Atrial Arrhythmia			Ventricular Arrhythmia	Other Pacing Needs		
			AT	AF	Other		SND	AV block	Dyssynchrony, heart failure
Simple	Patent ductus arteriosus	6-8%							
	Pulmonary stenosis	6-8%							
	Ventricular septal defect	30-32%							
	Secundum atrial septal defect	8-10%							
Moderate	Aortic coarctation	5-7%							
	Anomalous pulmonary venous return	0.5-2.5%							
	Atrioventricular septal defect	3-5%							
	Aortic stenosis	3-5%							
	Ebstein anomaly	0.5-1.5%							
	Tetralogy of Fallot	8-10%							
	Primum atrial septal defect	2-3%							
Severe	Truncus arteriosus	1.5-2%							
	Pulmonary atresia	2-2.5%							
	Double outlet right ventricle	1.5-2%							
	D-transposition of the great arteries	6-7%							
	L-transposition of the great arteries	1-2%							
	Hypoplastic left heart syndrome	3-4%							
	Other (heterotaxy, other single								

Khairy et al PACES/HRS Expert Consensus Statement on Arrhythmias in Adult Congenital Heart Disease

IECD Guidelines, statements, position papers

Circulation
JOURNAL OF THE AMERICAN HEART ASSOCIATION

2012

American Heart Association

2012 ACCF/AHA/HRS Focused Update Incorporated Into the ACCF/AHA/HRS 2008 Guidelines for Device-Based Therapy of Cardiac Rhythm Abnormalities : A Report of the American College of Cardiology Foundation/American Heart Association Task Force on Practice Guidelines and the Heart Rhythm Society

Andrew E. Epstein, John P. DiMarco, Kenneth A. Ellenbogen, N.A. Mark Estes III, Roger A.

2013 ESC Guidelines on cardiac pacing and cardiac resynchronization therapy

2013

The Task Force on cardiac pacing and resynchronization therapy of the European Society of Cardiology (ESC). Developed in collaboration with the European Heart Rhythm Association (EHRA).

Authors/Task Force Members: Michele Brignole (Chairperson) (Italy)*, Angelo Auricchio (Switzerland), Gonzalo Baron-Esquivias (Spain), Pierre Bordachar

Pharmacological and non-pharmacological therapy for arrhythmias in the pediatric population: EHRA and AEPC-Arrhythmia Working Group joint consensus statement

2013

PACES/HRS Expert Consensus Statement on the Recognition and Management of Arrhythmias in Adult Congenital Heart Disease

2014

Developed in partnership between the Pediatric and Congenital Electrophysiology Society (PACES) and the Heart Rhythm Society (HRS). Endorsed by the governing bodies of PACES, HRS, the American College of Cardiology (ACC), the American Heart Association (AHA), the European Heart Rhythm Association (EHRA), the Canadian Heart Rhythm Society (CHRS), and the International Society for Adult Congenital Heart Disease (ISACHD).

2015 ESC Guidelines for the management of patients with ventricular arrhythmias and the prevention of sudden cardiac death

2015

ESC European Society of Cardiology

Europace (2018) 0, 1–35
doi:10.1093/europace/eux380

2018

EHRA POSITION PAPER

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

2021 PACES Expert Consensus Statement on the Indications and Management of Cardiovascular Implantable Electronic Devices in Pediatric Patients

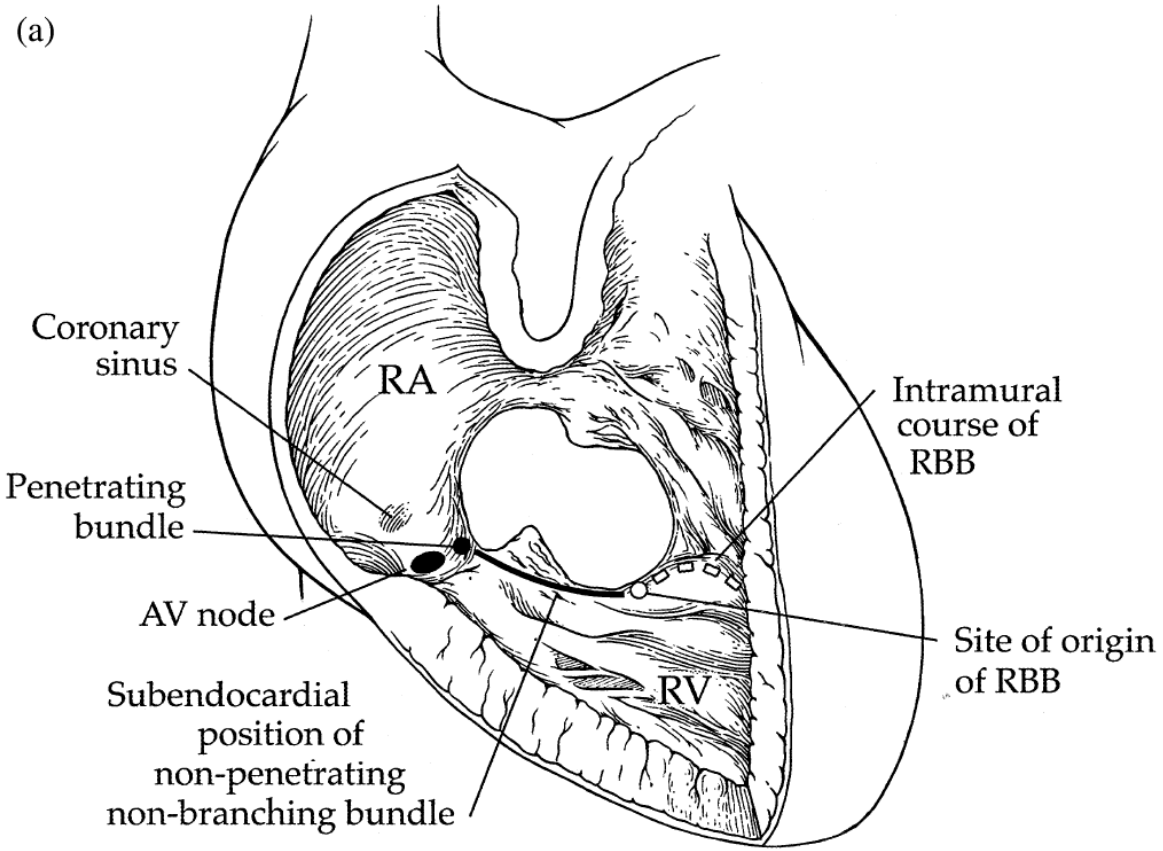
Developed in collaboration with and endorsed by the Heart Rhythm Society (HRS), the American College of Cardiology (ACC), the American Heart Association (AHA), and the Association for European Paediatric and Congenital Cardiology (AEPC). Endorsed by the Asia Pacific Heart Rhythm Society (APHRS), the Indian Heart Rhythm Society (IHRS), and the Latin American Heart Rhythm Society (LAHRS).

The conduction system and arrhythmias in common atrioventricular canal

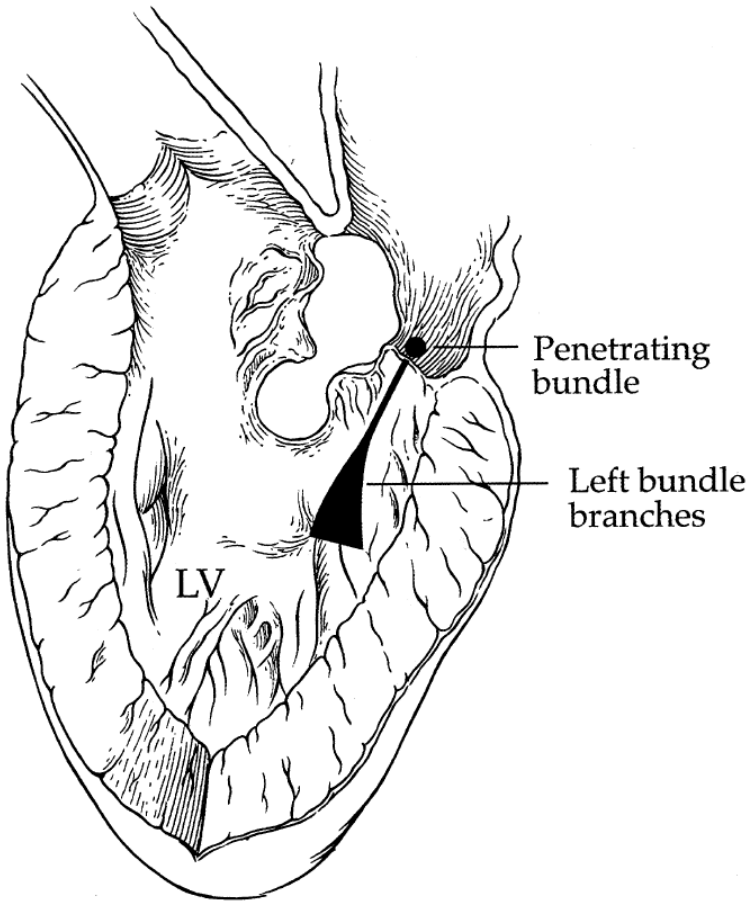
Progress in Pediatric Cardiology 10 (1999) 153–159

Naomi J. Kertesz*

(a)

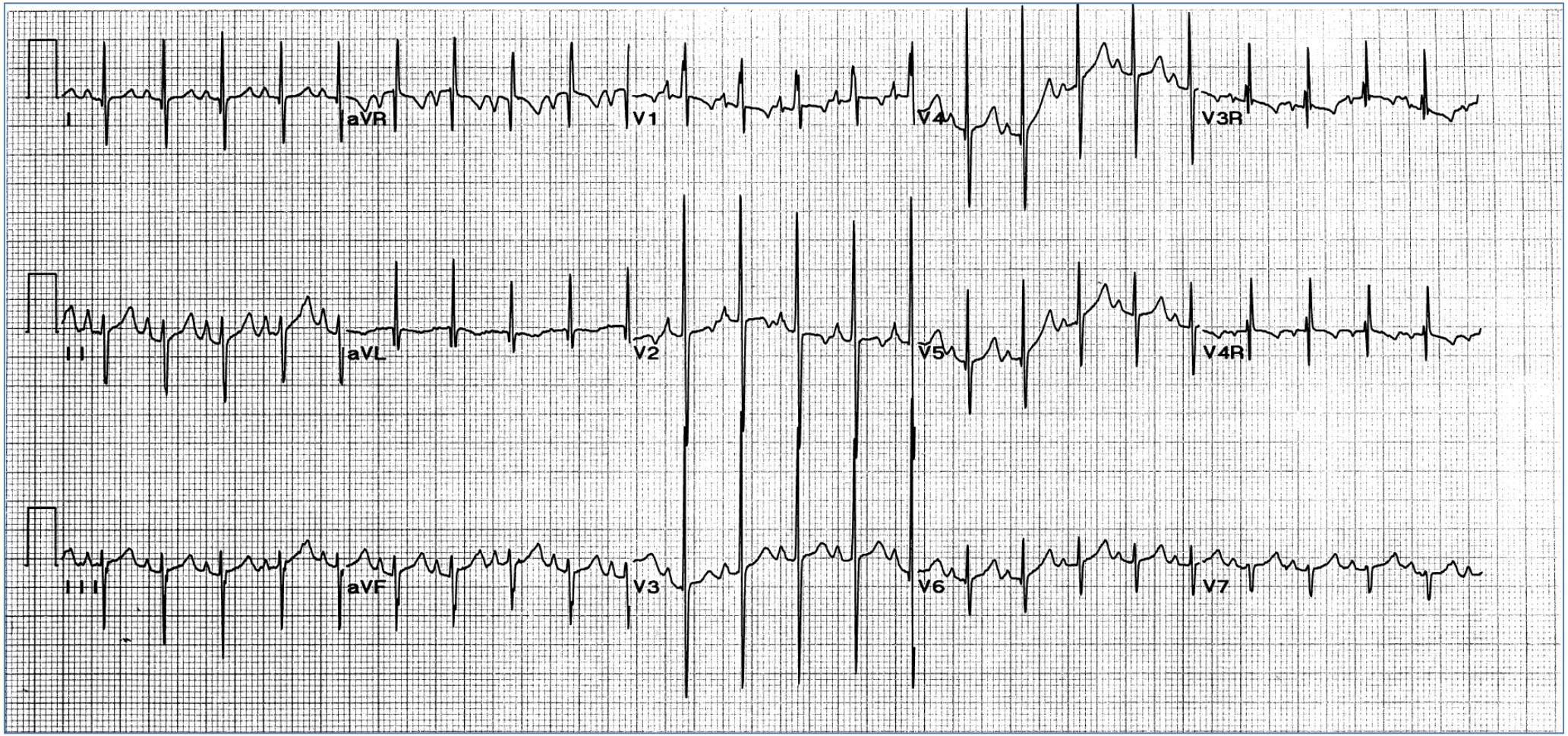


(b)



ECG changes

Left anterior hemiblock QRS morphology – superior axis



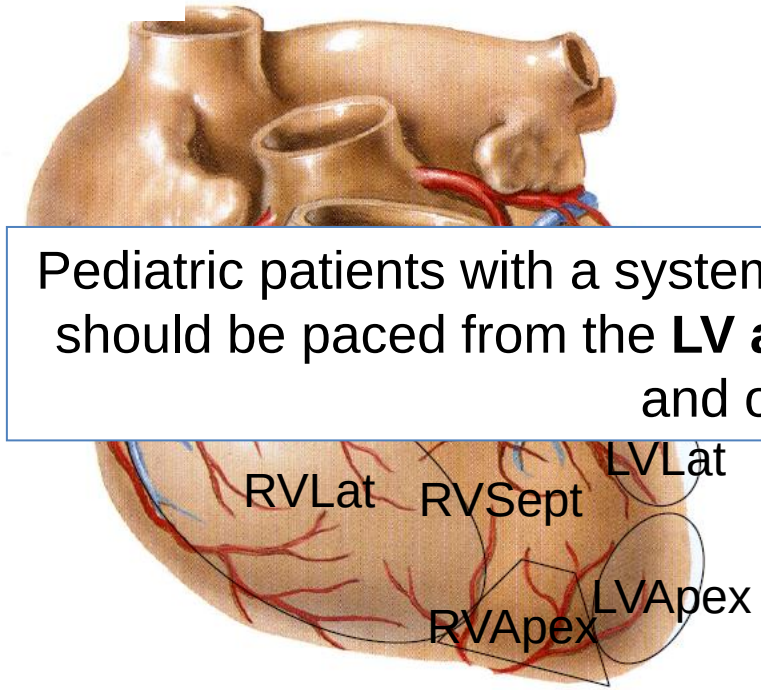
Pacing for AV block in AVSD

- Surgical AV block in the majority of AVSD requiring pacing
- Repair in infancy
  - Epicardial pacing
- Later on if pacemaker needed
  - Standard strategies of PM implantation including CRT if clinically indicated
    - CS ostium location if transvenous CRT lead planned?
  - Conduction system pacing not yet described
    - LBAP likely not simply feasible because of anatomy

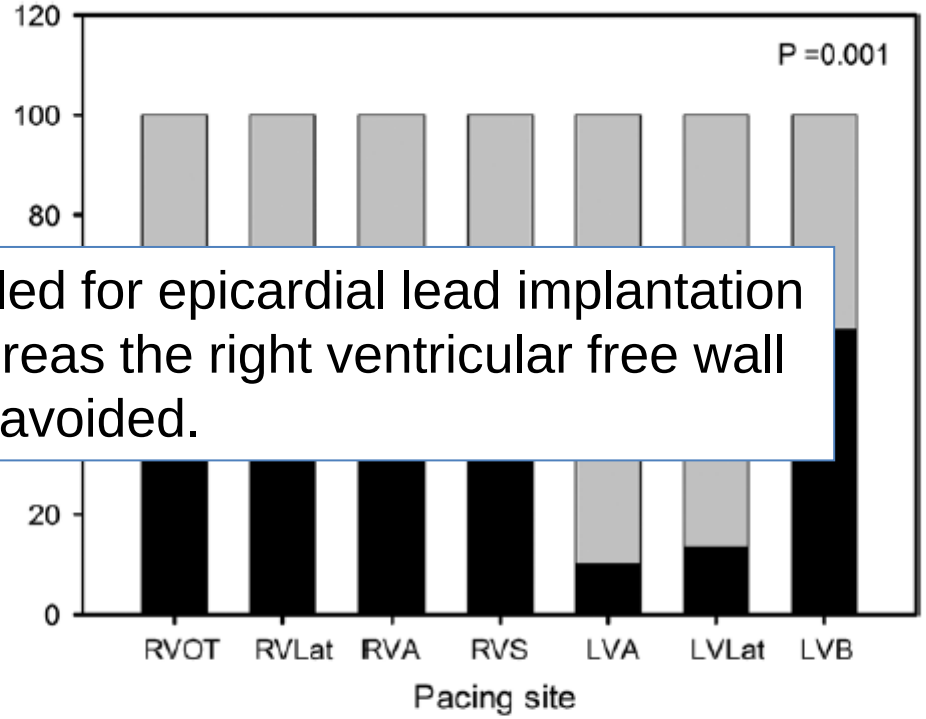
Permanent epicardial pacing in children

Preserving ventricular function by choosing the optimal ventricular pacing site

Cross-sectional study (N=178, 21 centers)



Proportion of pts with LVEF<55 %



Pediatric patients with a systemic LV who are scheduled for epicardial lead implantation should be paced from the **LV apex or free wall**, whereas the right ventricular free wall and outflow tract should be avoided.

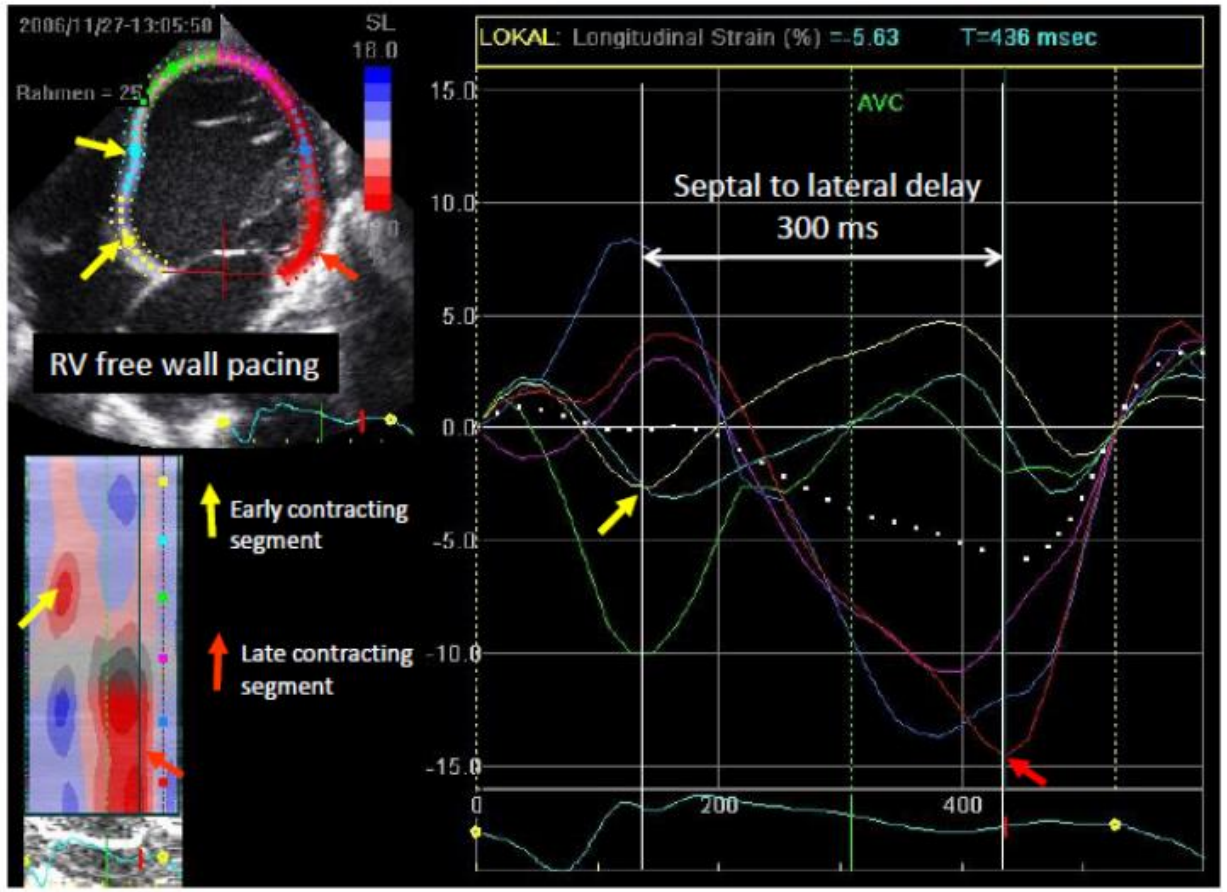
Preservation of LV function (LV EF ≥55 %):
LV apical/lateral pacing: OR 8.26, p=0.018

Janousek J et al. Circulation 2013;127:613-623

Results of LV apical pacing

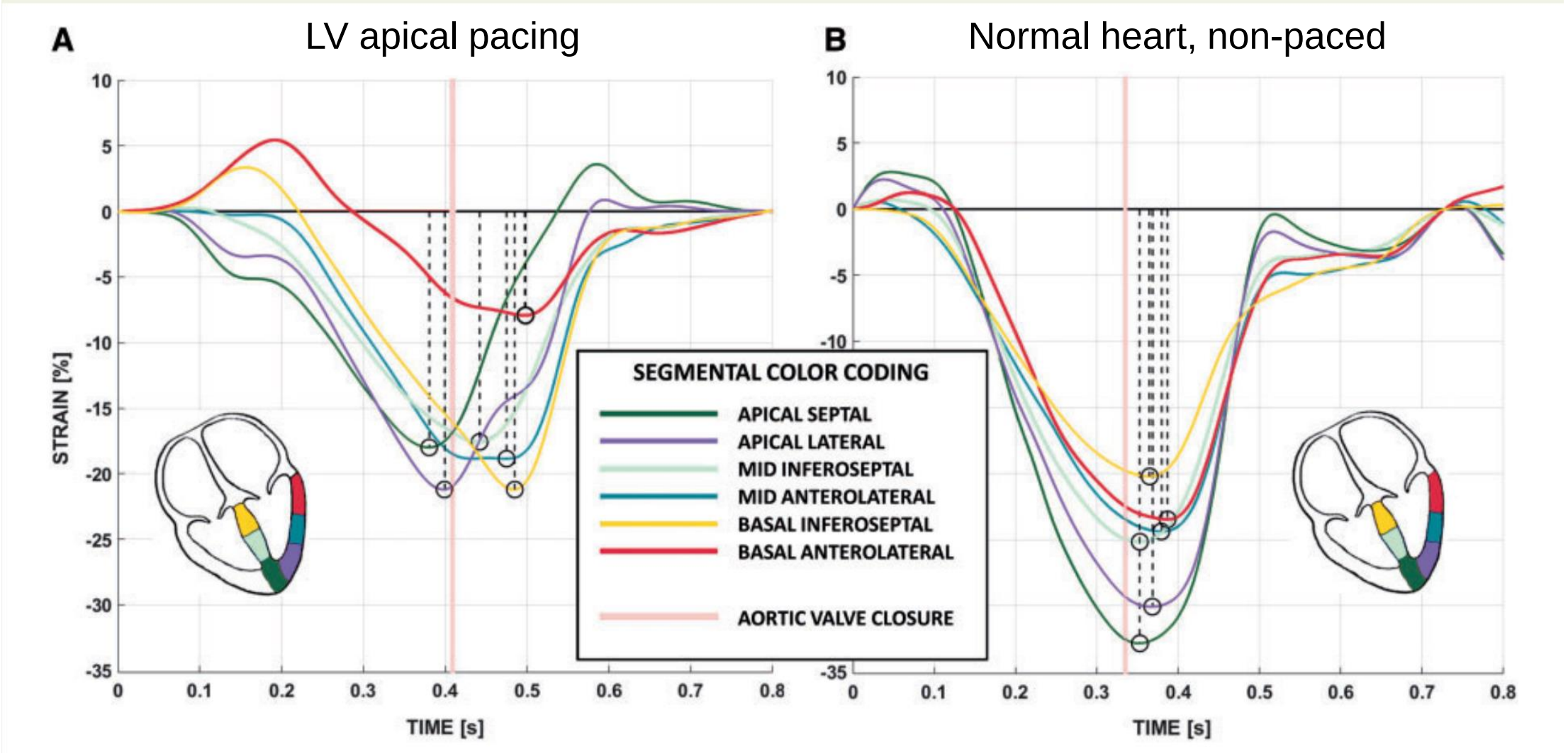
LV contraction pattern

RV free wall pacing



Kovanda J et al. Left ventricular apical pacing in children: feasibility and long-term effect on ventricular fucntion. Europace 2019

LV contraction pattern in epicardial pacing



Kovanda J et al. Left ventricular apical pacing in children: feasibility and long-term effect on ventricular fucntion. Europace 2019

Results of LV apical pacing

Structurally normal heart vs structural heart disease vs normal controls

AVSD in 5

Table I **Demographic and clinical data**

	Group A (*N* = 22)	Group B (*N* = 14)	Group C (*N* = 25)	*P*-value
	CAVB (structurally normal heart)	CAVB (structural heart disease)	Age-matched healthy controls	
Age at implantation (years)	1.04 (0.02–5.79)	1.74 (0.75–2.79)	–	NS
Pacing duration (years)	4.35 (2.68–6.18)	3.14 (1.73–8.79)	–	NS

Table 3 **Left ventricular metrics at cross-sectional evaluation**

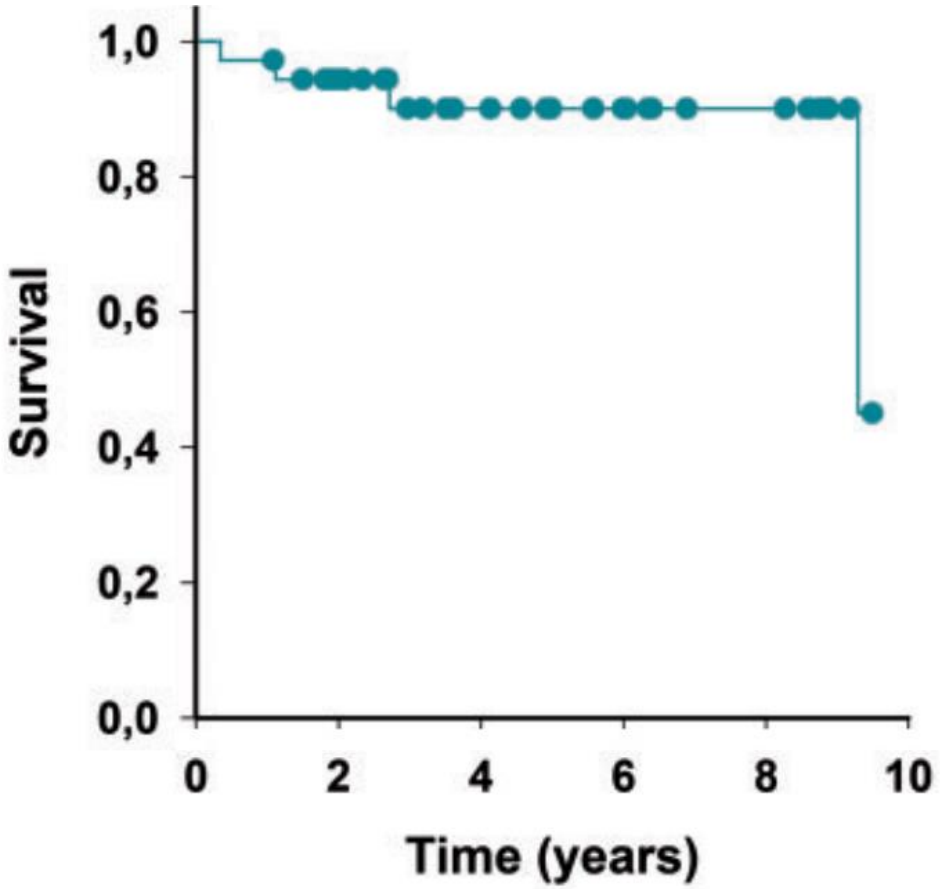
Group	A	B	C	*P*-value overall	*P*-value A vs. C	*P*-value B vs. C	*P*-value A vs. B
	CAVB (structurally normal heart)	CAVB (structural heart disease)	Age-matched healthy controls				
LVEDD (*Z* score)	1.47 (1.18)	0.86 (1.46)	−0.29 (1.28)	<0.001	<0.001	0.024	NS
LV SF (%)	40 (5.6)	39 (3.9)	36 (4.2)	NS	–	–	–
LV EF (Teichholz) (%)	69.8 (6.4)	69.8 (4.8)	66 (5.7)	NS	–	–	–

Kovanda J et al. Left ventricular apical pacing in children: feasibility and long-term effect on ventricular fucntion. Europace 2019

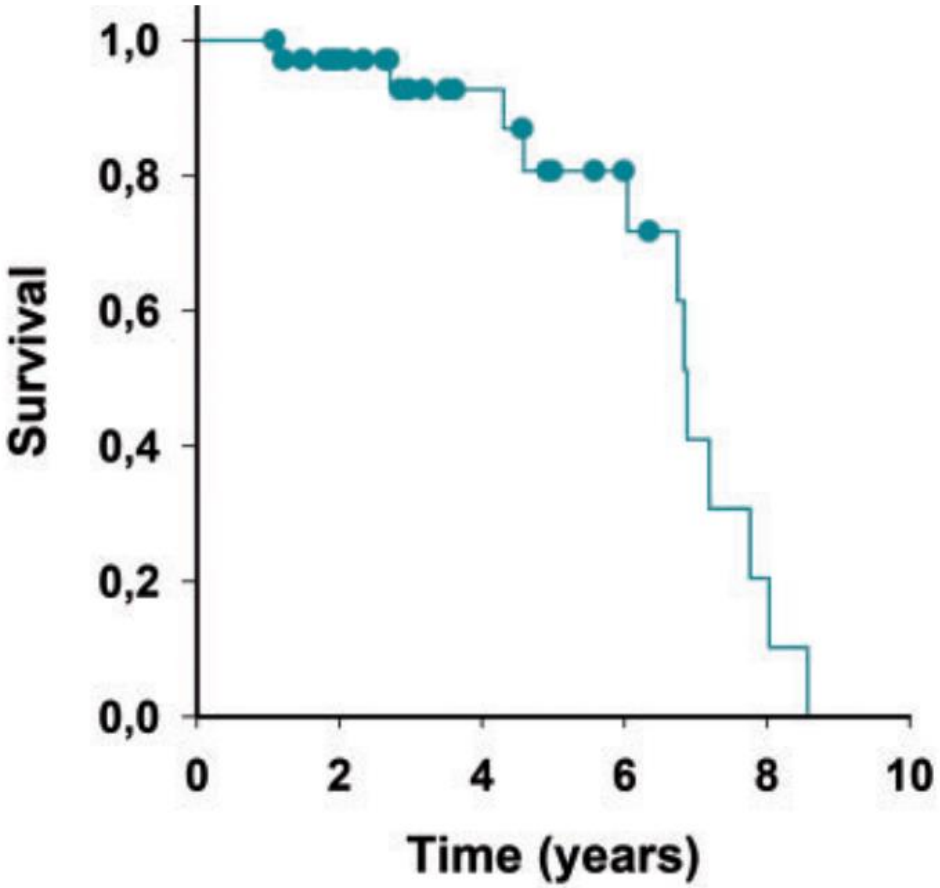
Results of LV apical pacing

Pacing system survival

Freedom from pacemaker-related surgical revision (elective generator replacement excluded).



Freedom from generator replacement for battery depletion



Kovanda J et al. Left ventricular apical pacing in children: feasibility and long-term effect on ventricular fucntion. Europace 2019

Tachyarrhythmias

- Mainly supraventricular
  - Congenital substrates for SVT
    - Accessory pathways
      - Midseptal pathways not present
      - Cave ablation in posteroseptal area: posterior AV node displacement
    - Atrioventricular nodal reentrant tachycardia
    - Twin AV nodes
  - Acquired substrates
    - Cavo-annular isthmus dependent IART
    - Atriotomy dependent IART
    - Complex biatrial IART
    - Focal atrial tachycardia
    - Atrial fibrillation

Atrioventricular Nodal Reentrant Tachycardia in Patients With Congenital Heart Disease

Outcome After Catheter Ablation

Circ Arrhythm Electrophysiol. 2017;10:e004869.

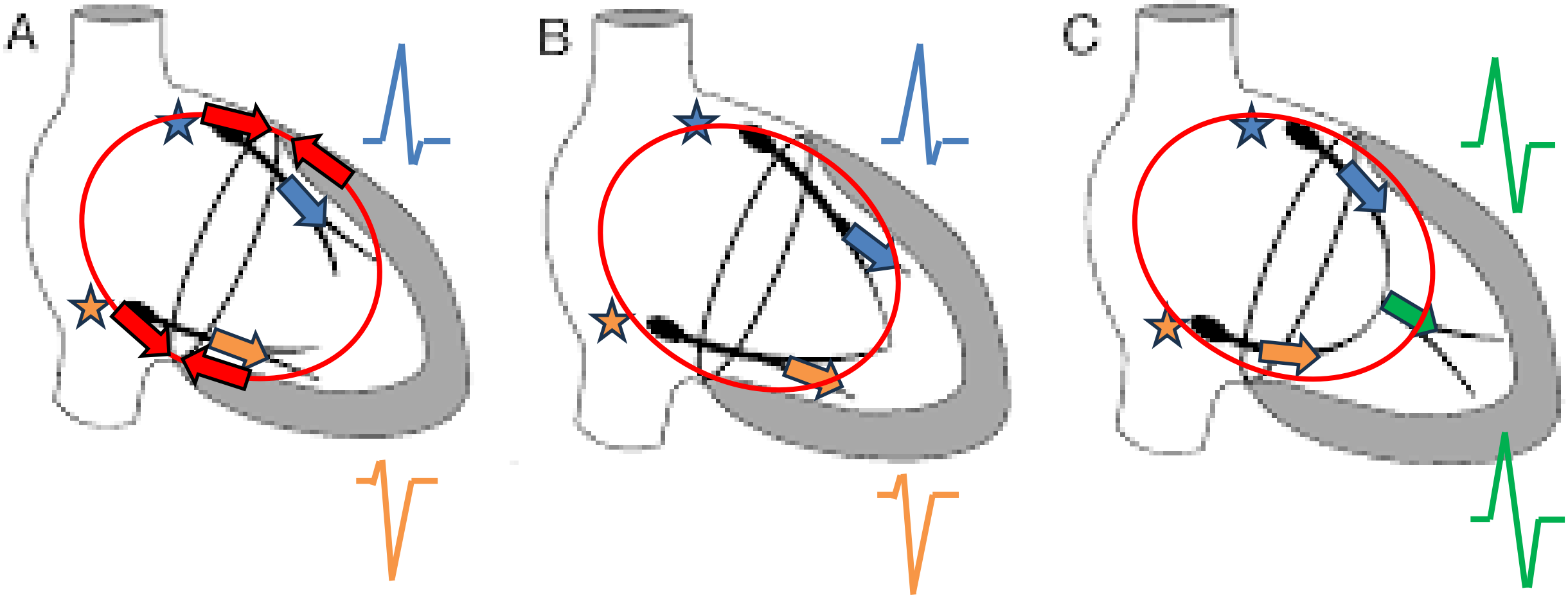
John Papagiannis, MD; Daniel Joseph Beissel, MD; Ulrich Krause, MD;

Group A: Complex CHD	N	Group B: Simple CHD	N
Tetralogy of Fallot	10	ASD 2	28
D-TGA after Mustard or Senning	9	VSD	8
Single ventricle physiology	6	Aortic valve disease	7
Ebstein anomaly	6	Pulmonary valve stenosis	5
Double outlet right ventricle	5	Mitral valve disease	3
Partial atrioventricular canal	4	ASD sinus venosus	2
CC-TGA	4	Partial anomalous pulmonary venous return	2
Common atrioventricular canal	3	Aortic aneurysm (Marfan syndrome)	1
Pulmonary atresia-intact ventricular septum	2	Subaortic stenosis VSD	1
Total anomalous pulmonary venous return	1	Supravalvar aortic stenosis (Williams syndrome)	1
Shone complex	1		
Total	51		58

- Outcome
  - All AVSD successfull
  - No permanent AV block
- Pecularities
  - An inverted relationship of the slow and fast pathways
  - Left-sided ablation of the posterior extension of AV node

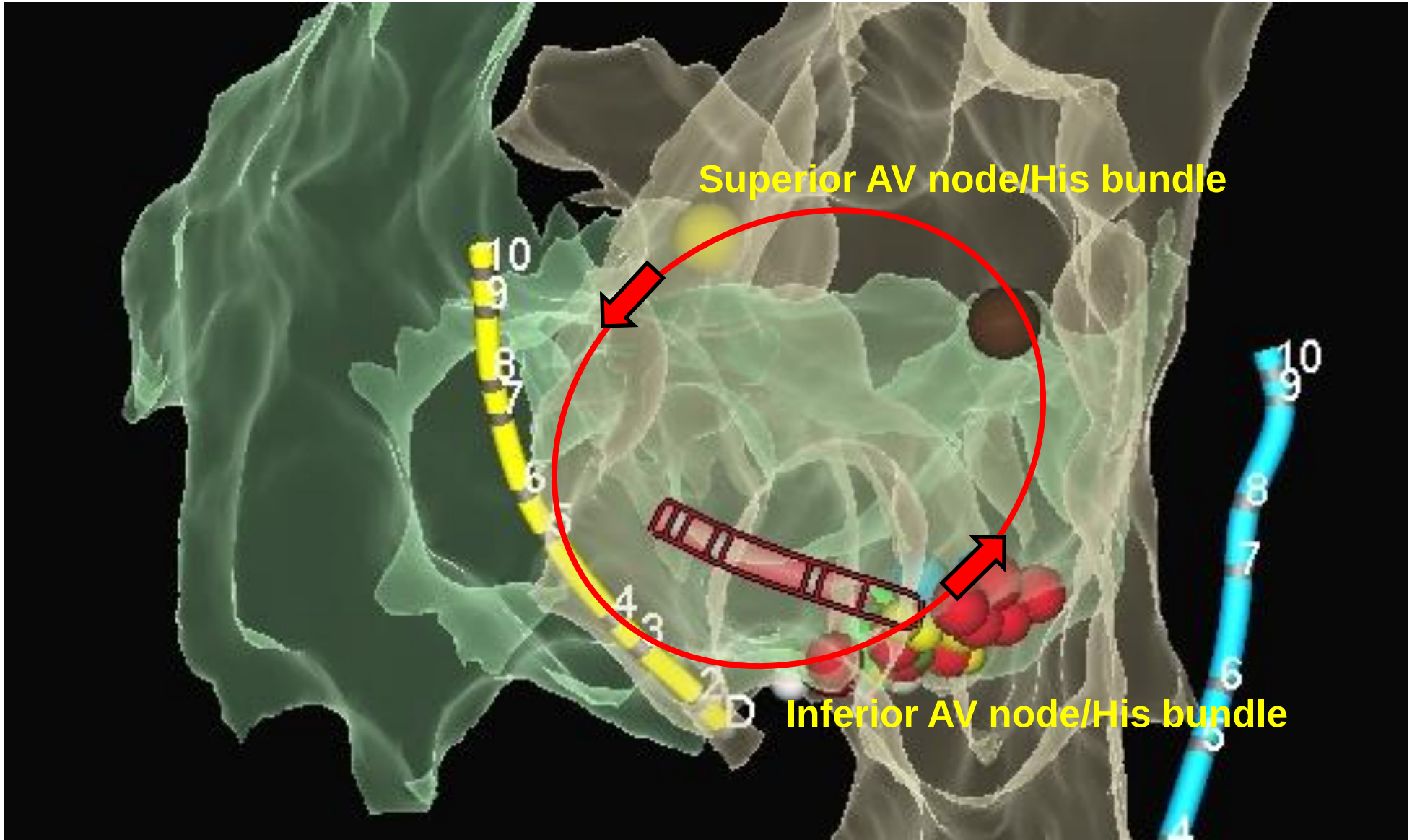
Twin (dual) AV nodes

Typically in unbalanced AVSD and dextroisomerism



Epstein MR et al. JCE 2003

Ablation of SVT due to twin AV nodes



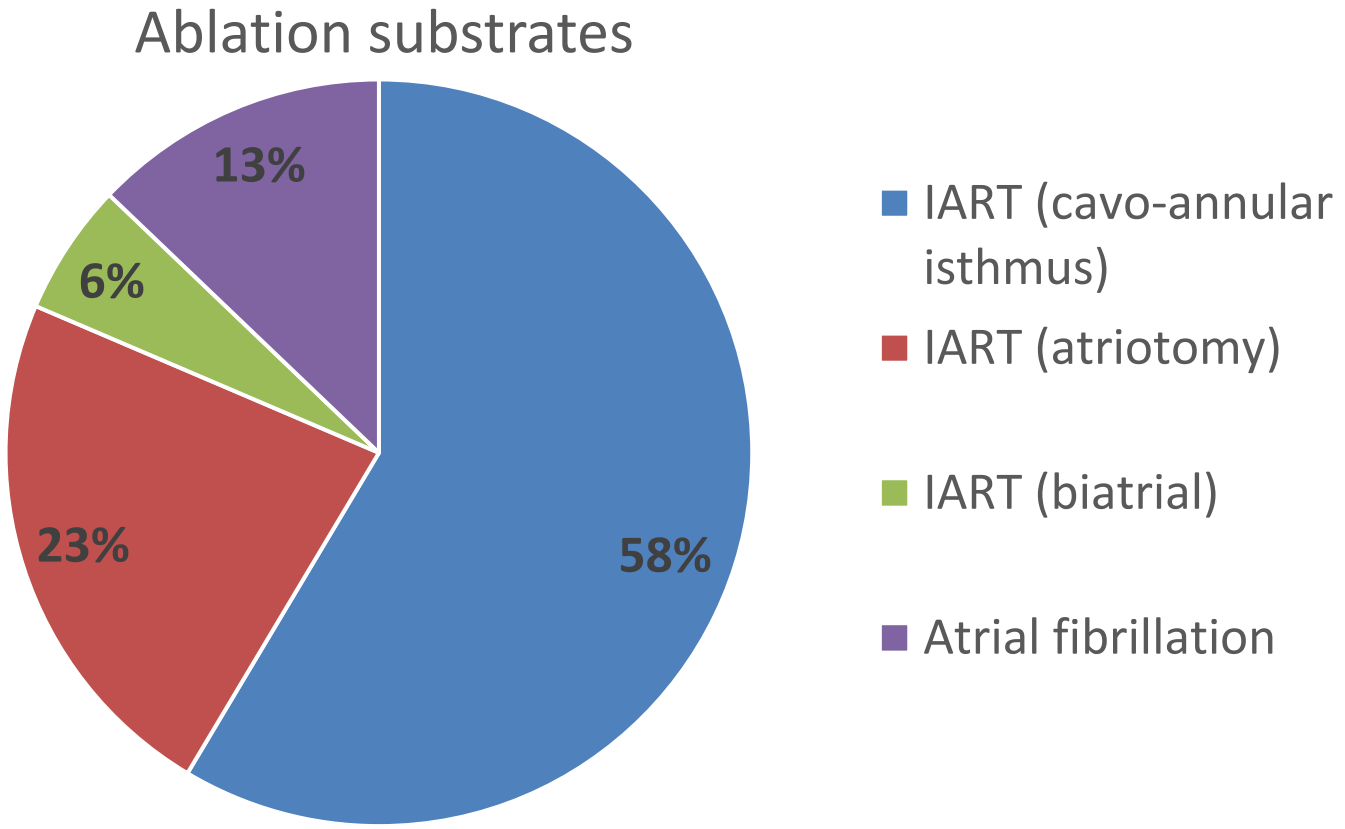
ESC European Society of Cardiology
Europace (2023) **25**, 1–8
https://doi.org/10.1093/europace/euad275

CLINICAL RESEARCH

Catheter ablation of atrial tachyarrhythmias in patients with atrioventricular septal defect

Victor Waldmann[1,2,3,4]*, Francis Bessière [5], Kevin Gardey [5],

- N=56
- Age 38.1±17.4 years
- Acute success = 96.4 %
  - 1.6 arrhythmia/patient
- Repeated ablation
  - 15 (26.8 %) of patients
  - Left-sided and biatrial mechanism



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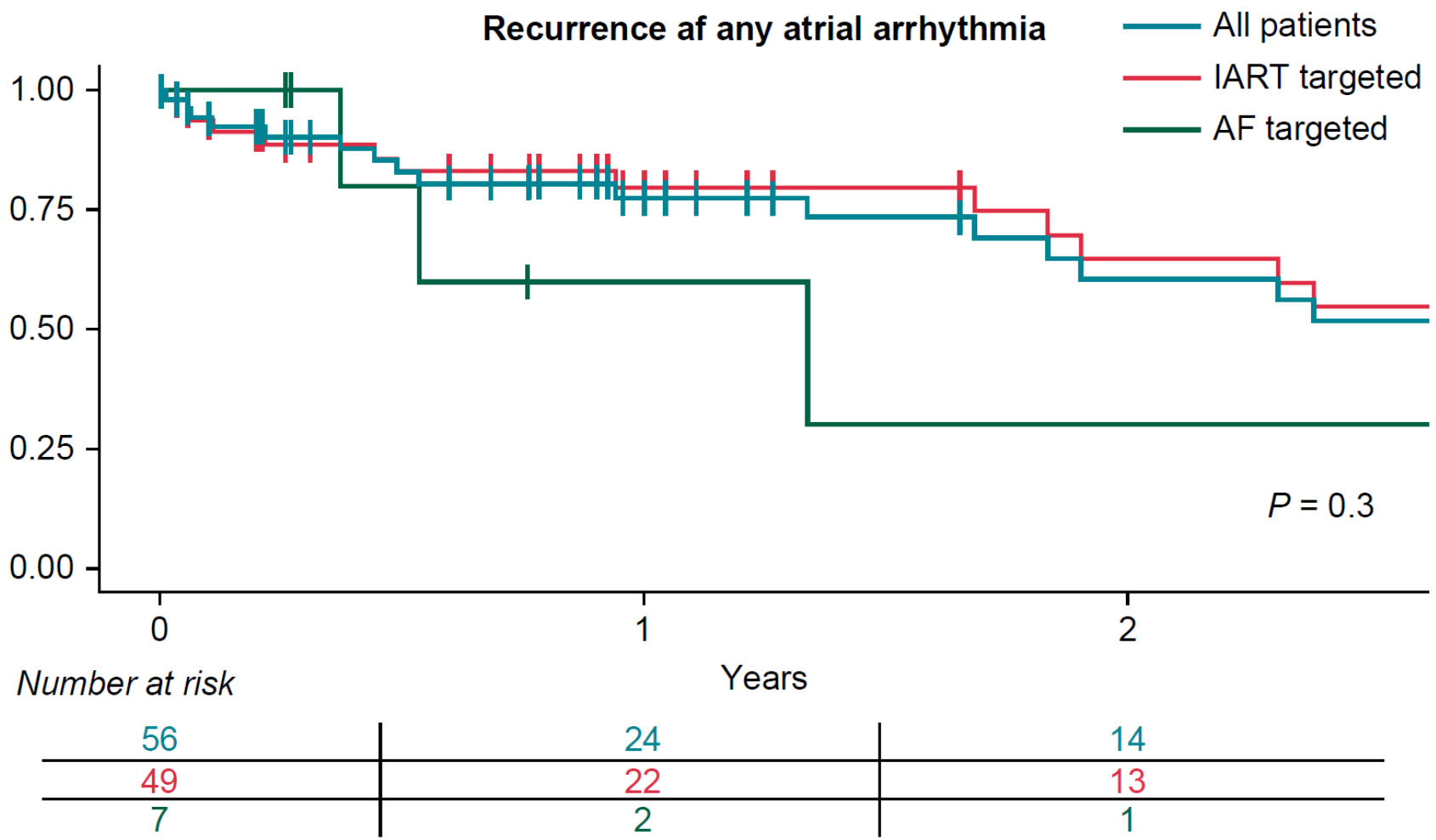

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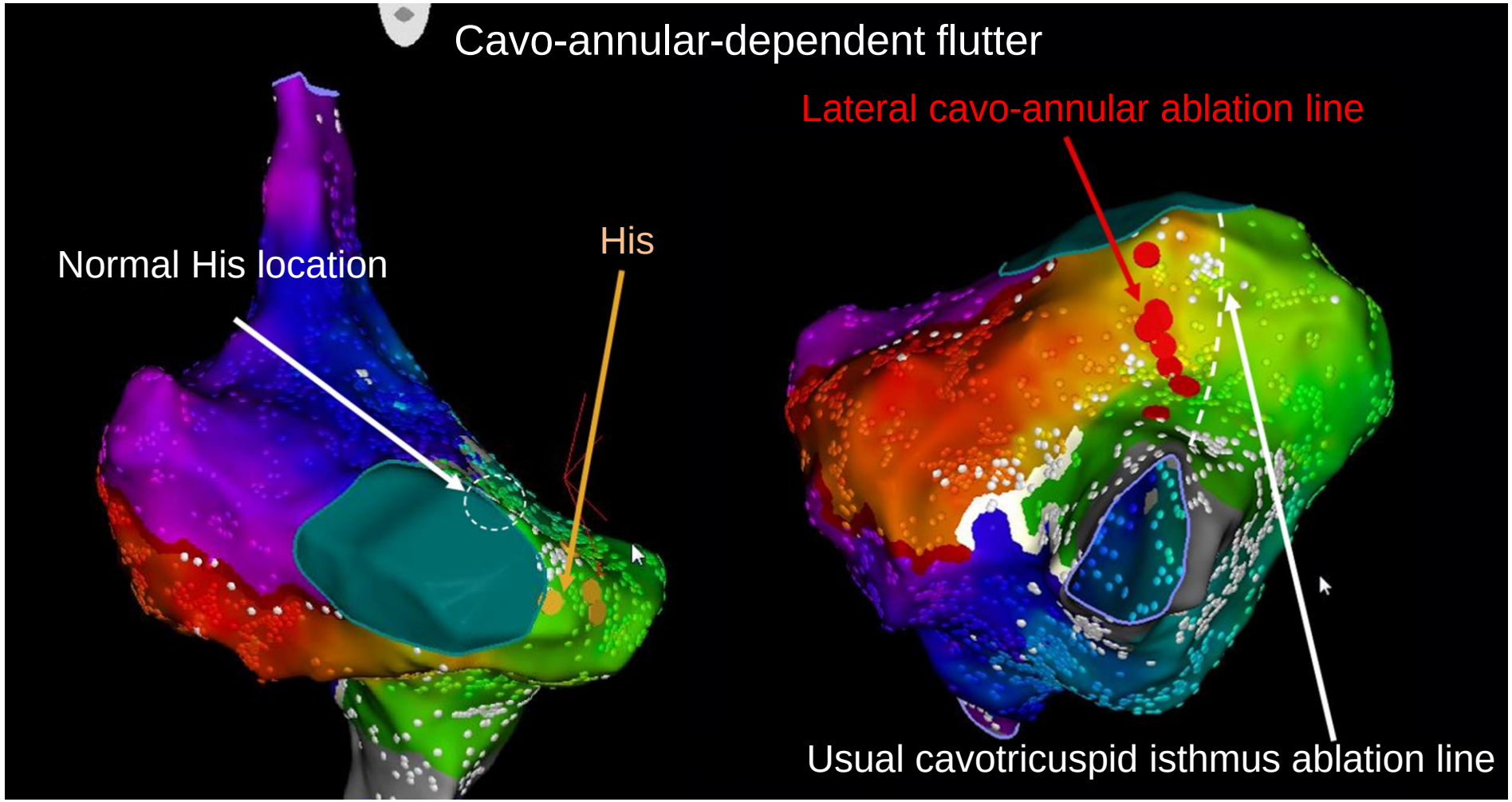

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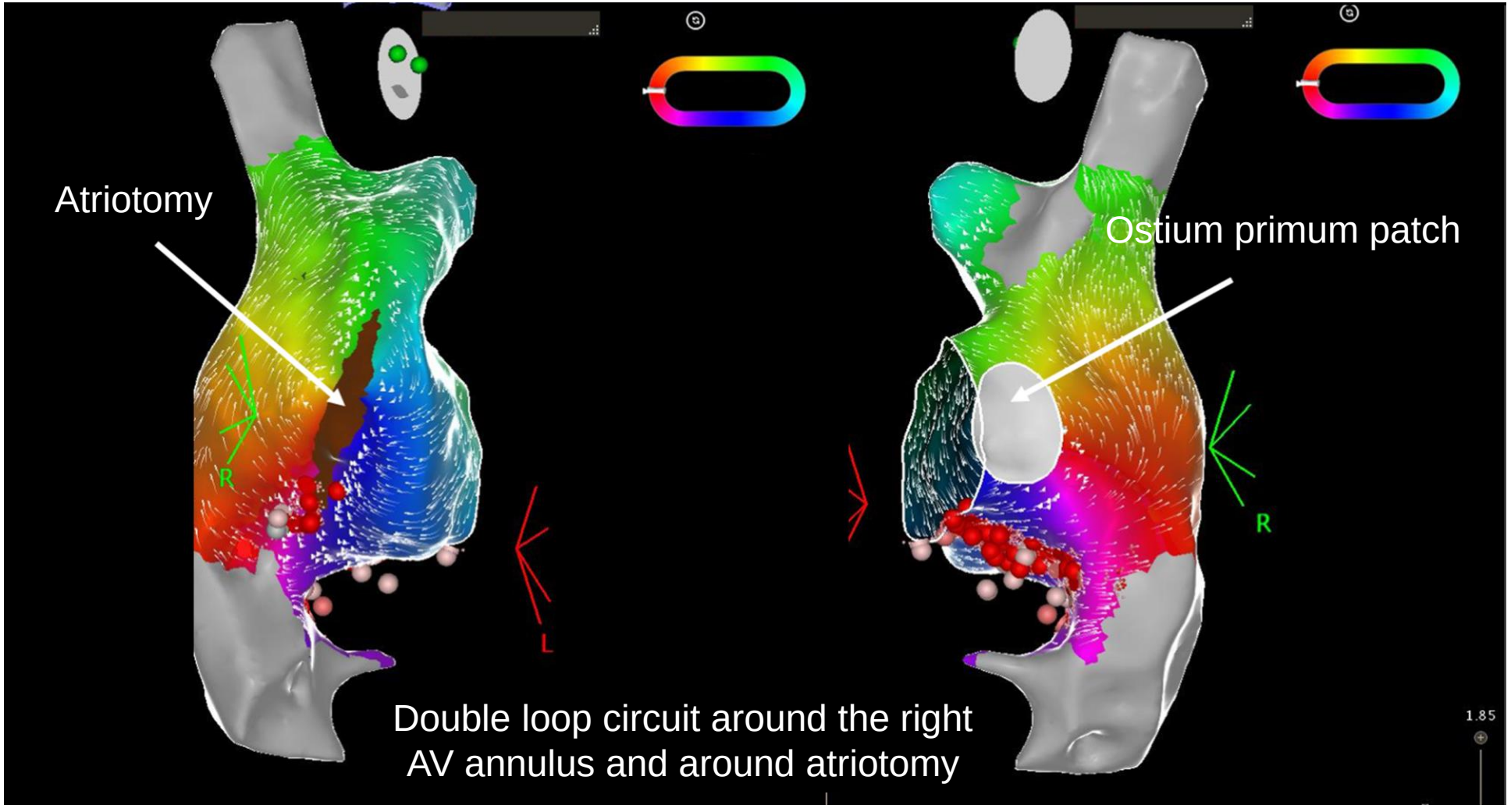

Summary

- Arrhythmias in AVSD not particularly different from other types of biventricular hearts
  - With the exception of functionally univentricular AVSD
- Same treatment principles apply
- LV apex should be the preferential pacing site early on
- AV conduction system anatomy needs to be acknowledged to prevent AV block during ablation
- Unusual and complex ablation substrates may be present