KHRS 2019

Pediatric and Congenital

Idiopathic Ventricular Tachycardia in Children and Adolescents

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COI

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The author has no financial conflicts of interest to disclose concerning the presentation.

PACES/HRS Expert Consensus Statement on the Evaluation and Management of Ventricular Arrhythmias in the Child With a Structurally Normal Heart

Crosson JE, Callans DJ, Bradley DJ, Dubin A, Epstein M, Etheridge S, Papez A, Phillips JR, Rhodes LA, Saul P, Stephenson E, Stevenson W, Zimmerman F.

Heart Rhythm. 2014;11(9):e55-78. doi: 10.1016/j.hrthm.2014.05.010.

Treatment of children with ventricular arrhythmias and a structurally normal heart

Class 1

1. Asymptomatic infants and children with normal ventricular function and frequent but isolated ventricular ectopy or accelerated ventricular rhythm should be observed, with no medical or ablative therapy (Level of evidence: B).
2. Infants and children with well-tolerated idiopathic outflow tract tachycardia that is infrequent, slow, and self- terminating should be monitored, with no medical or ablative therapy (Level of evidence: B).
3. Children with VT or frequent ventricular ectopy thought to be causative of documented ventricular dysfunction should be treated either medically or with catheter ablation* (Level of evidence: C).
4. Children who experience hemodynamic compromise due to presumed idiopathic outflow tract tachycardia should be treated either medically or with catheter ablation* (Level of evidence: C).
5. Symptomatic children and infants older than 1 year with presumed intrafascicular verapamil-sensitive reentrant tachycardia should have initial medical management with a calcium-channel blocking agent or catheter ablation* (Level of evidence: B).
6. Infants and children with an acute presentation of polymorphic VT should have prompt correction of treatable causes such as electrolyte abnormalities or drug toxicity (Level of evidence: C).

Treatment of children with ventricular arrhythmias and a structurally normal heart

Class 2a

1. In asymptomatic infants and children with frequent complex or multiform ventricular ectopy, β-blocker therapy can be useful. If this does not control the arrhythmia, suppressive therapy with calcium-channel blockers can also be useful. If this arrhythmia is very well tolerated and infrequent, only observation can be useful (Level of evidence: C).
2. In symptomatic children with presumed idiopathic outflow tract tachycardia, or with rhythm correlated symptoms due to ventricular ectopy or accelerated idioventricular rhythm, suppressive therapy with a β-blocker or catheter ablation can be useful (Level of evidence: C).
3. In infants younger than 1 year with presumed intrafascicular verapamil-sensitive reentrant tachycardia, medical therapy with β-blocker therapy can be useful (Level of evidence: C).

Treatment of children with ventricular arrhythmias and a structurally normal heart

Class 2b

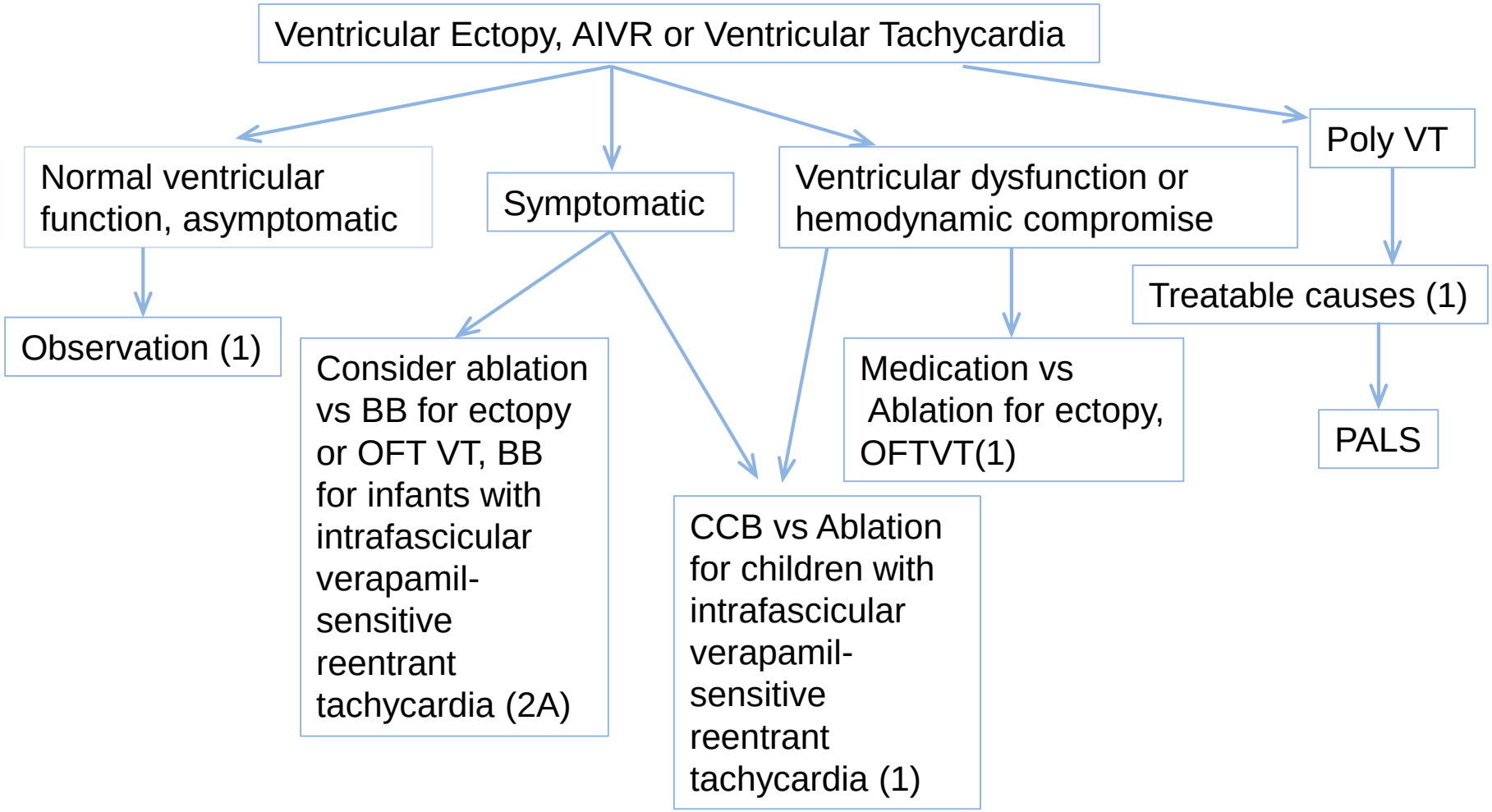
1. In infants and children with frequent complex or multiform ventricular ectopy, treatment with other agents (class I or III) after failure of β-blockers and/or calcium-channel blockers may be reasonable (Level of evidence: C).
2. Catheter ablation* may be reasonable in children with complex ventricular arrhythmias where one morphology dominates or when there is a suspected trigger that can be targeted (Level of evidence: C).
3. ICD implantation may be reasonable in children or older infants with polymorphic VT when the arrhythmia persists after acute treatable causes have been ruled out if sudden death risk persists (Level of evidence: C).

Treatment of children with ventricular arrhythmias and a structurally normal heart

Class 3

1. Catheter ablation in infants and toddlers is not recommended, except in the case of VT that cannot be adequately controlled medically and is not tolerated hemodynamically (Level of evidence: C).
2. Exercise restrictions are not recommended in children with normal ventricular function, no or minimal symptoms, and well-tolerated and/or well-controlled monomorphic ventricular arrhythmias (Level of evidence: C).
3. ICD implantation is not recommended in patients with IVT, regardless of symptoms, unless the tachycardia cannot be adequately controlled with medication and/or catheter ablation and in the judgment of the specialist the patient has a risk of sudden death higher than expected in this population (Level of evidence: C).

Treatment algorithm



Numbers in parentheses refer to the level of recommendation. AIVR = accelerated idio ventricular rhythm; BB = β-blocker; CCB = calcium-channel blocker; OFTVT = outflow tract tachycardia; PALS = Pediatric Advanced Life Support; VT = ventricular tachycardia.

Crosson JE, et al. Heart Rhythm. 2014;11:e55-78.

Indications for catheter ablation in children with idiopathic ventricular arrhythmias*

Class 1 Catheter ablation is recommended in children with:

1. Ventricular dysfunction or hemodynamic compromise presumed to be due to ventricular ectopy or tachycardia, either as primary therapy or in patients not controlled medically (Level of evidence: C).
2. Intrafascicular verapamil-sensitive reentrant tachycardia, either as primary therapy or if not controlled by calcium- channel blockers (Level of evidence: C).

Class 2a Catheter ablation can be useful in:

1. Symptomatic children with presumed idiopathic outflow tract tachycardia(Level of evidence: C).
2. Symptomatic children with rhythm-correlated symptoms due to frequent ventricular ectopy or accelerated idioventricular rhythm (Level of evidence: C).

Class 2b Catheter ablation may be reasonable to consider in children with polymorphic ventricular arrhythmia where one morphology dominates or when there is a suspected trigger that can be targeted(Level of evidence: C).

Class 3 Catheter ablation is not recommended in:

1. Infants and toddlers, except in the case of VT that cannot be adequately controlled medically and is not tolerated hemodynamically (Level of evidence: C).
2. Asymptomatic ventricular ectopy or tachycardia that is not suspected of causing ventricular dysfunction(Level of evidence: C).
3. Ventricular arrhythmias due to transient reversible causes, such as acute myocarditis or drug toxicity(Level of evidence: C).

*Catheter ablation for ventricular arrhythmias in children should be performed only by centers and physicians with expertise in ablation therapy in pediatric patients

Crosson JE, et al. Heart Rhythm. 2014;11:e55-78.

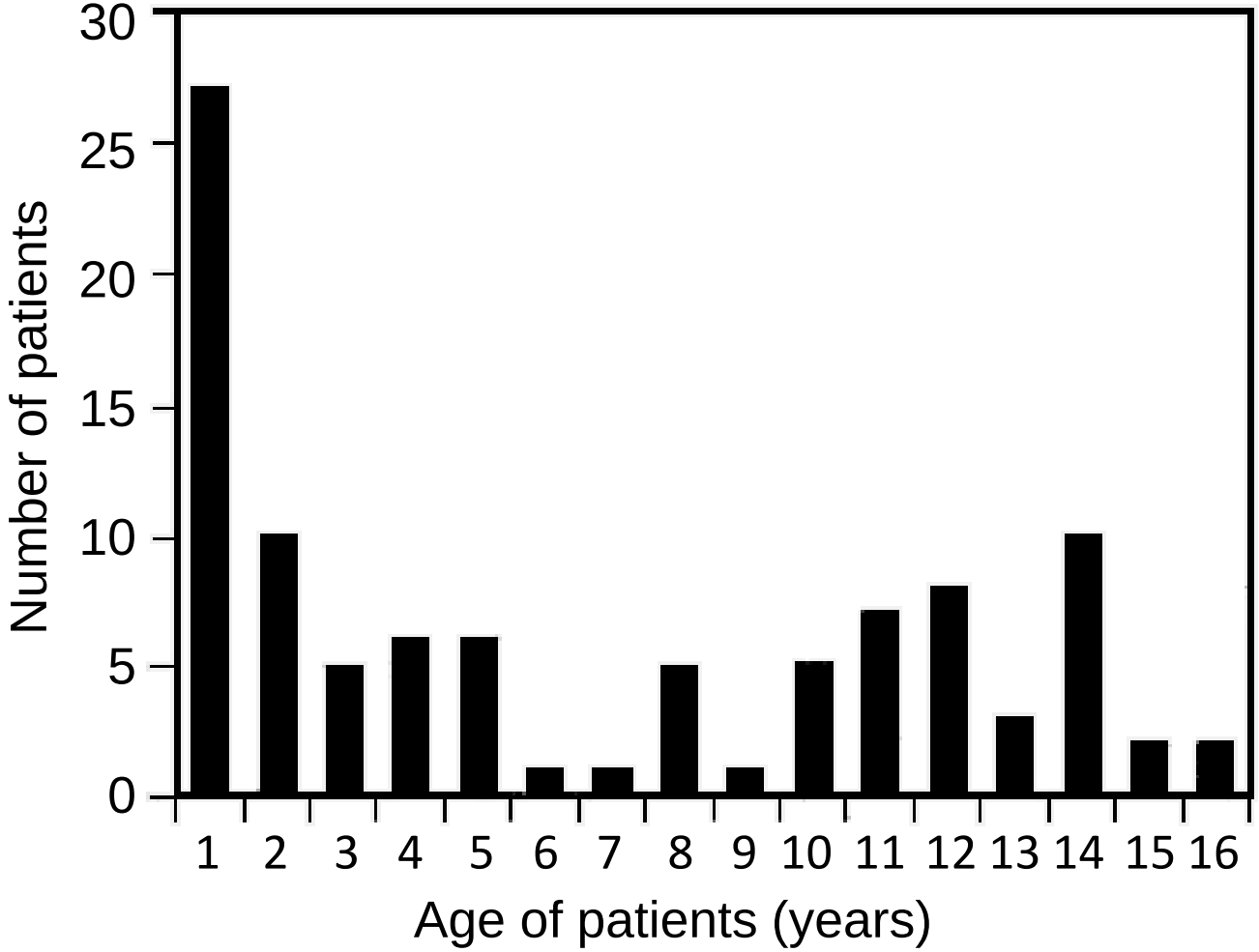
Comparisons of the idiopathic ventricular tachycardia (VT) at different age group

Parameters	<1 year n = 5	1–10 years n = 18	≧10 years n = 34	p value
Male (%)	4 (80%)	5 (27.8%)	23 (65.7%)	**0.019**
Symptoms				
Palpitation	2 (40%)	14 (77.8%)	21 (61.8%)	0.106
Heart failure or shock	1 (20%)	2 (11.1%)	9 (26.4%)	
Syncope/near syncope	0	1 (5.6%)	3 (8.8%)	
Associated dilated cardiomyopathy	0	2 (11.1%)	6 (17.6%)	0.519
VT spontaneous resolution	4 (80%)	1 (5.6%)	3 (8.8%)	**<0.001**
LBBB vs. RBBB	4/1	8/10	4/30	**0.001**
Tachycardia cycle length (ms)	268 ± 62	341 ± 84	380 ± 91	**0.014**
VT QRS duration (ms)	91 ± 13	126 ± 13	125 ± 17	**0.01**

p value less than 0.05 was presented as bold italics.
Abbreviation: LBBB- left bundle branch block, RBBB- right bundle branch block, VT ventricular tachycardia.

Chiu SN. et al. Arch Dis Child. 2017;102:1137-1142.

Age distribution of the patients with idiopathic VT in children



- 98 children from 12 European centers
- M:F=53:45
- Mean age 5.4 years (range 0.1–15.1)
- 27 patients (27%) <1 year (17 newborn)

Pfammatter JP, et al. JACC 1999; 33: 2067–72

Comparison of Clinical Characteristics of Patients Depending on the QRS Morphology During VT

	RBBB (Presumed Left VT)	LBBB (Presumed Right VT)	P Value
Infants	4/27 (14%)	23/71 (32%)	<0.05
Symptoms	18/27 (67%)	18/71 (25%)	<0.01
Max. heart rate in VT	182 ± 38 beats/min	175 ± 37 beats/min	NS
VT resolution	10/27 (37%)	54/71 (76%)	<0.01
Ablation	4/27 (14%)	5/71 (7%)	NS

LBBB: left bundle branch block morphology; RBBB: right bundle branch block morphology; VT: ventricular tachycardia.

Pfammatter JP, et al. JACC 1999; 33: 2067–72

Comparison of Clinical Characteristics of VT Depending on the Age at Initial Manifestation

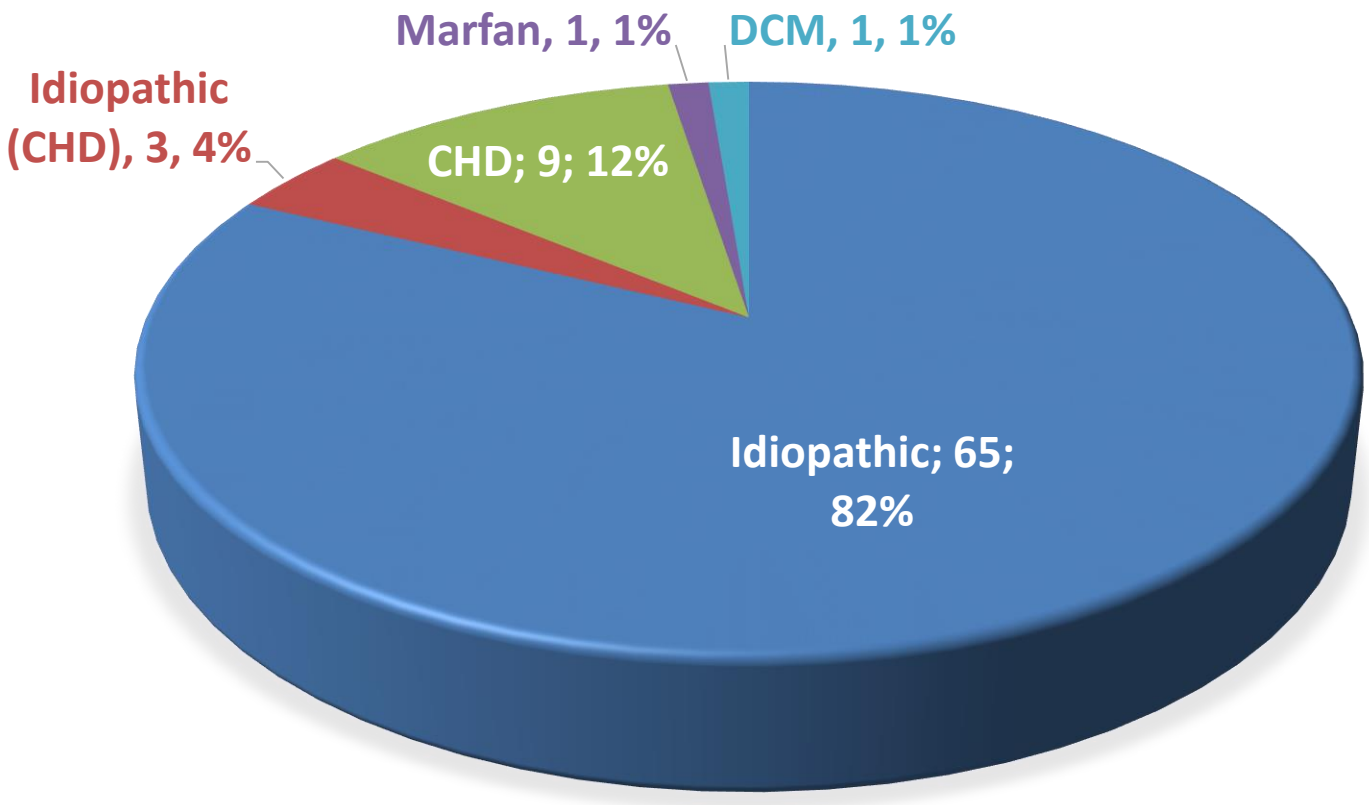
	Infants	Children	p Value
Number of patients	27	71	
Symptoms initially	6 (22%)	30 (38%)	<0.05
Sustained VT	12 (44%)	24 (34%)	NS
RBBB	4 (14%)	23 (32%)	<0.05
VT resolution	24 (89%)	40 (56%)	<0.01
Never treated	9 (33%)	16 (23%)	NS
Still treated	2 (7%)	21 (30%)	<0.05
Follow-up	48 months	46 months	NS

RBBB; right bundle branch block morphology

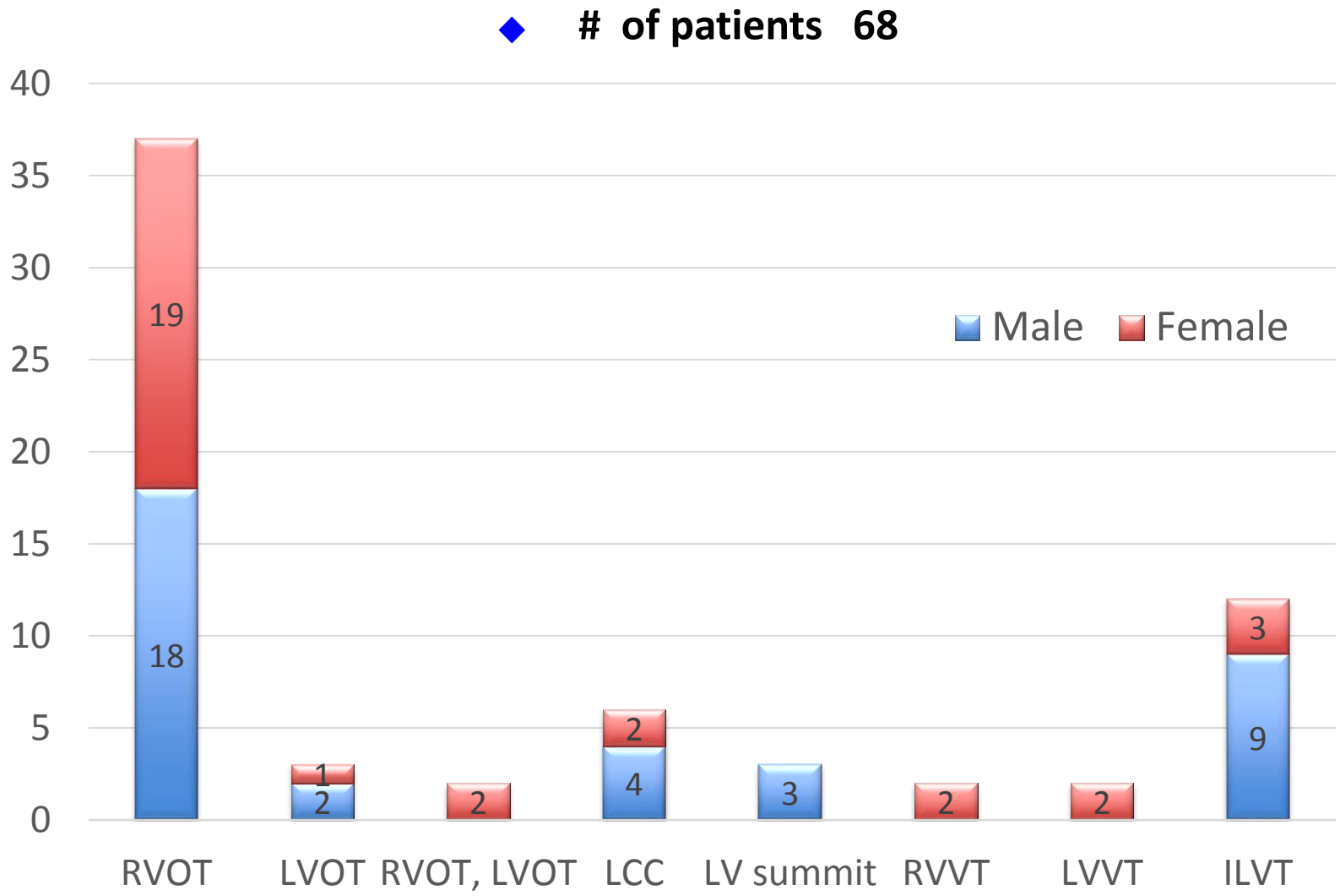
Pfammatter JP, et al. JACC 1999; 33: 2067–72

Cause of VT & PVC in RFA patients

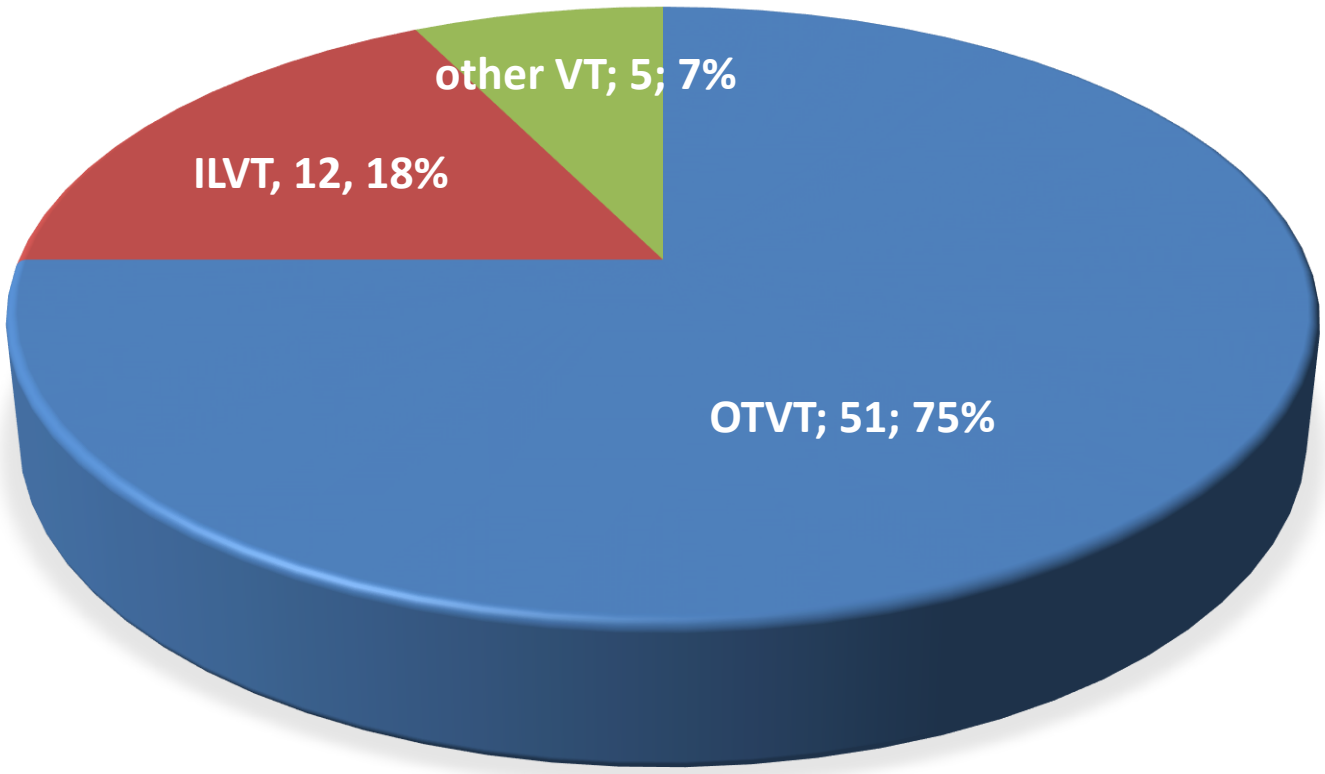
- # of patients 79
- Age (years) 12.5 ± 6.1



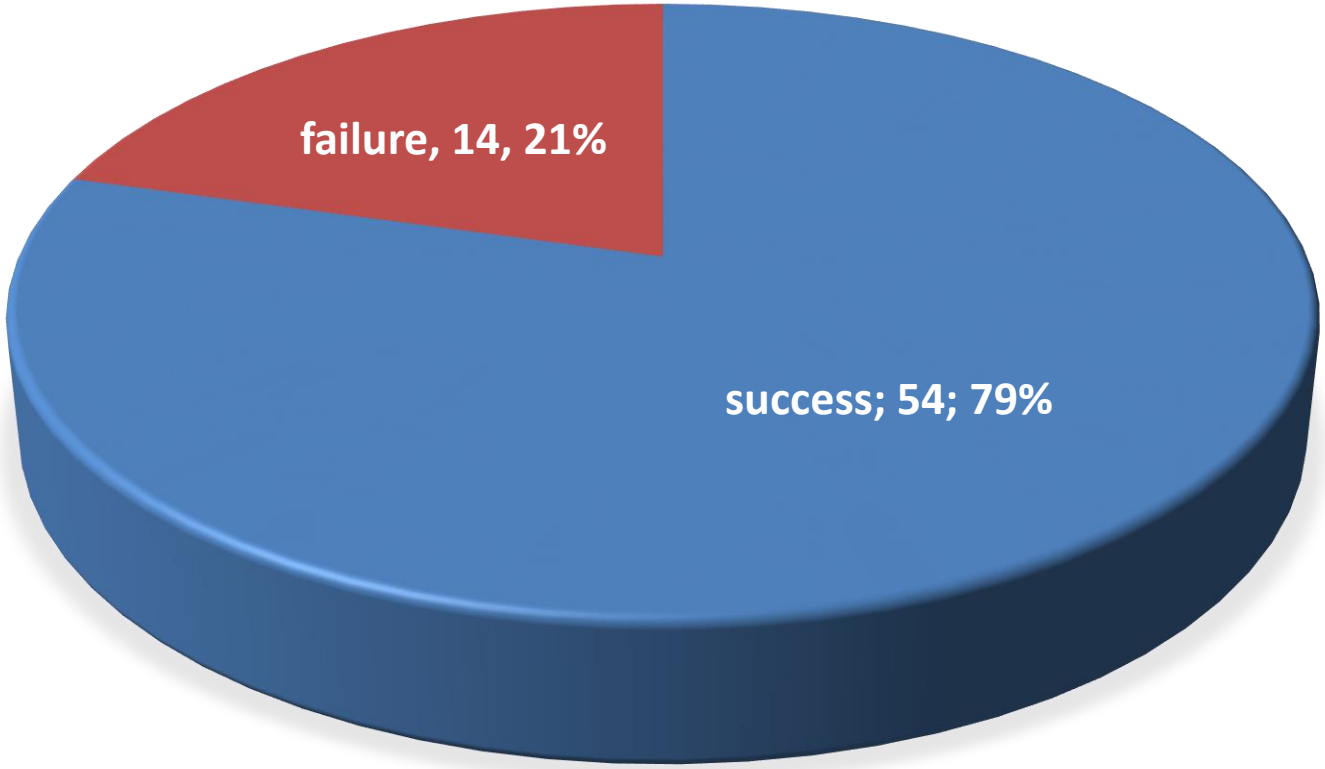
Origin of idiopathic VT & PVC



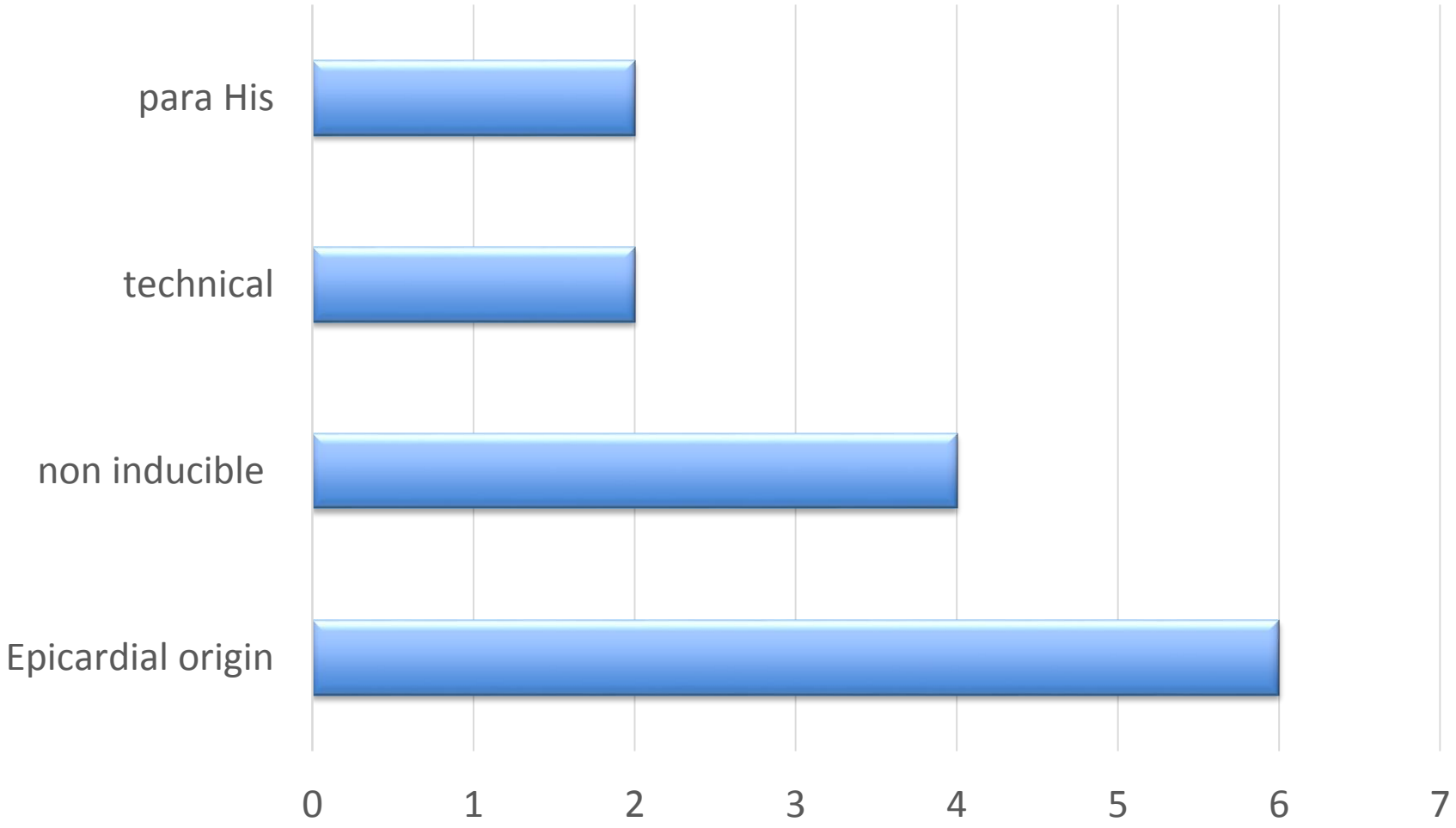
Types of idiopathic VT & PVC



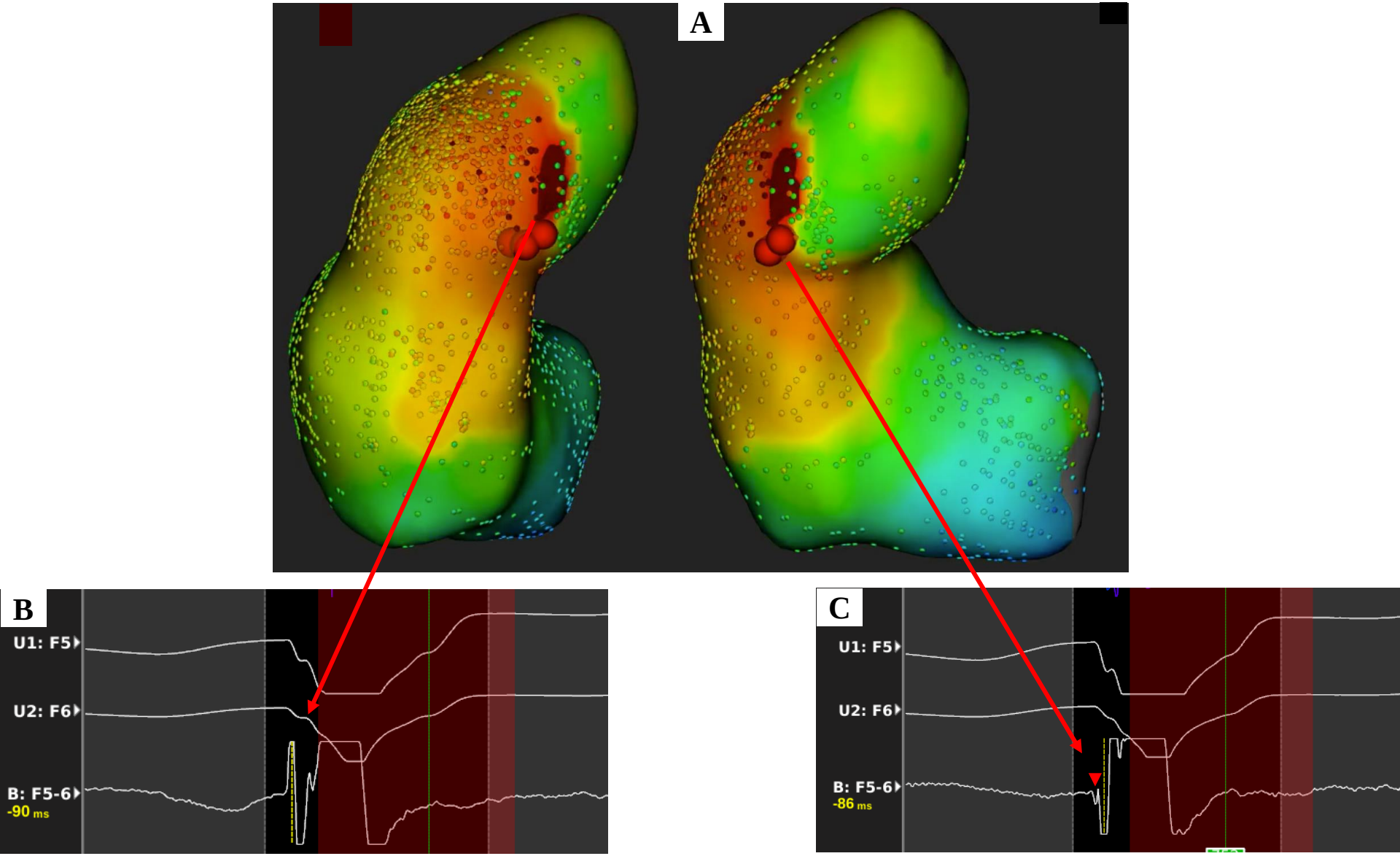
Success rate of RFA in idiopathic VT & PVC



Reason of Failure of ablation in idiopathic VT & PVC

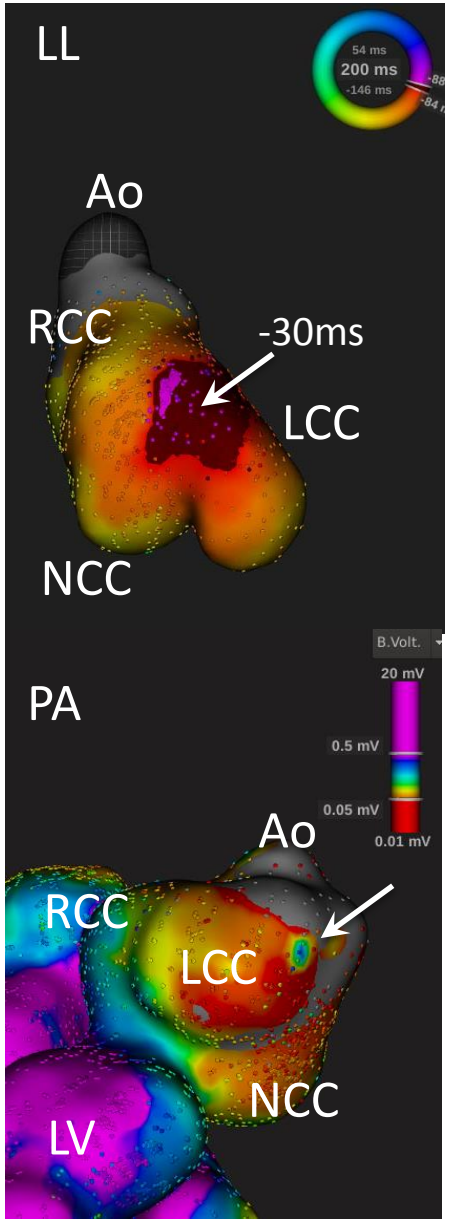


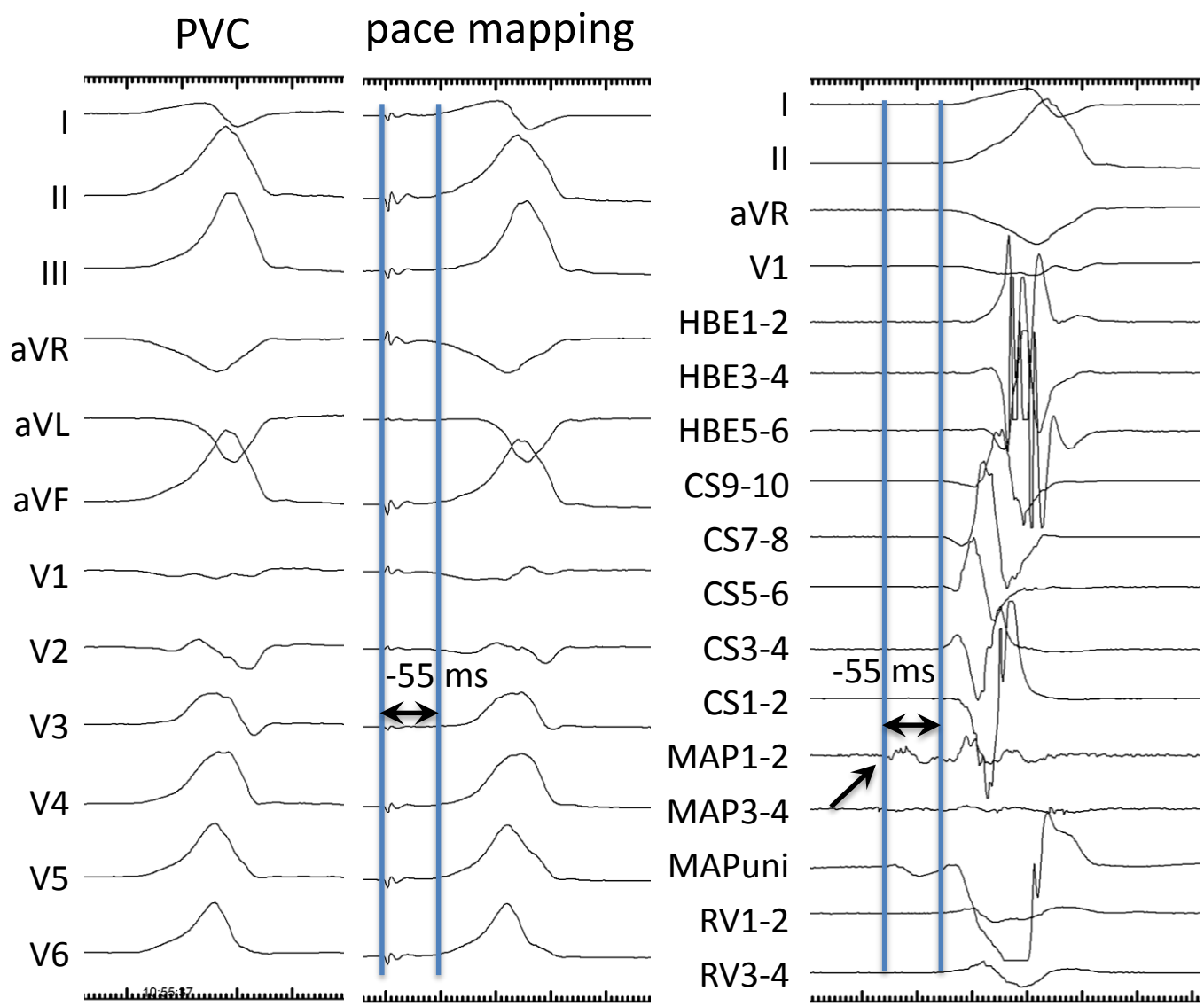
RVOT mapping of a PVC (8y Female)



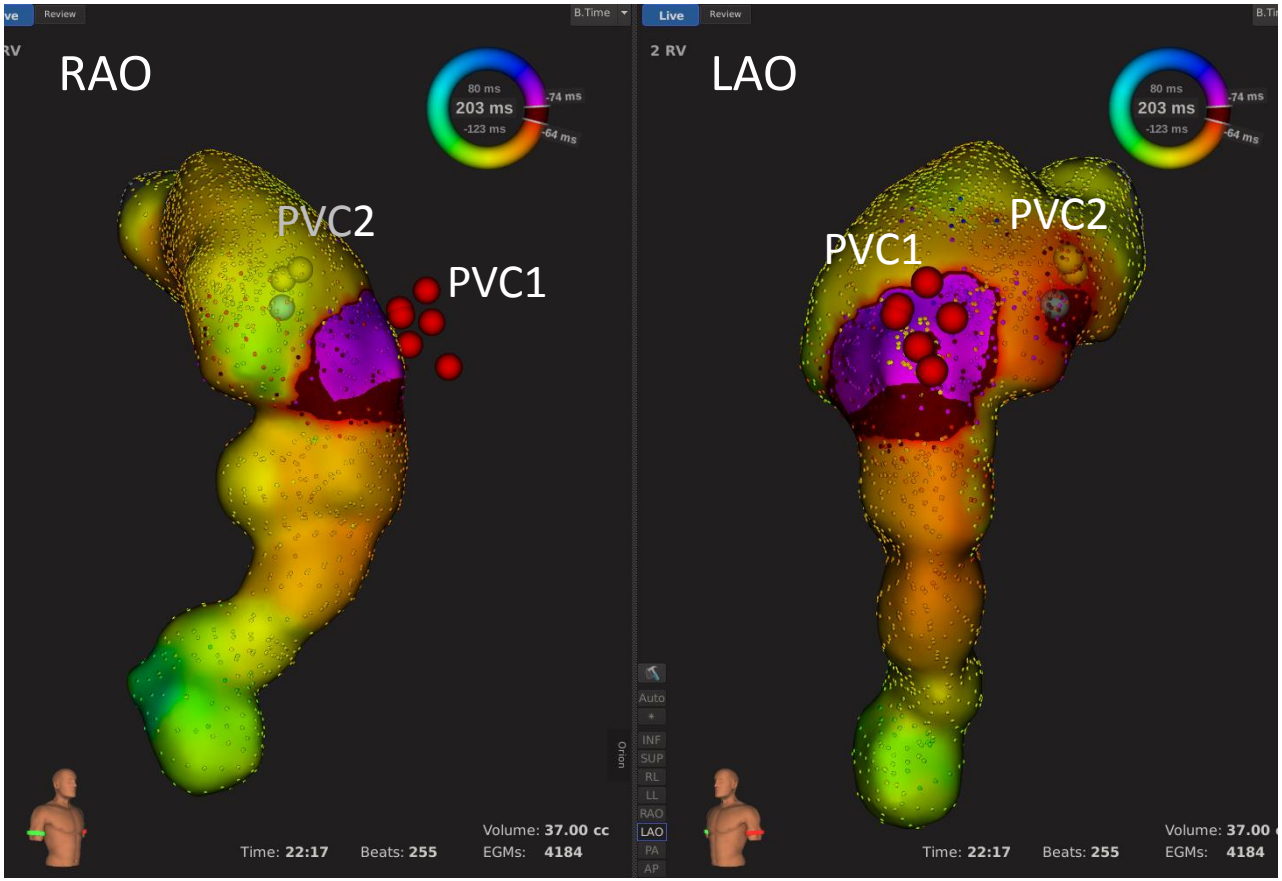
Mori H, Muraji S, Sumitomo N, et al. Heart Rhythm 2019 (in press)

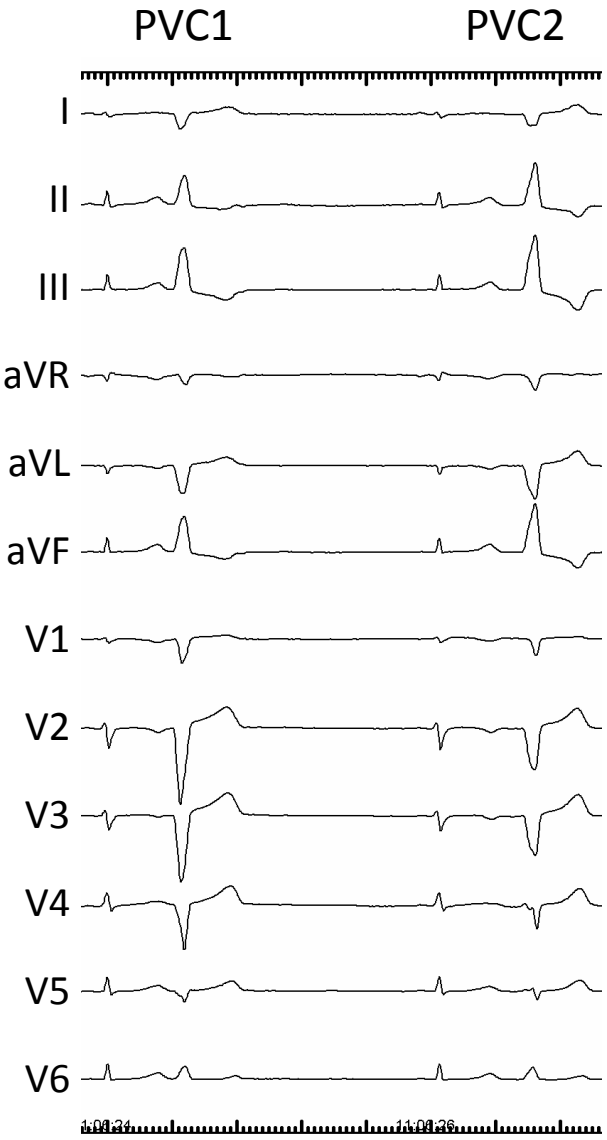
s/p Rastelli for TOF/PA, PVC from LCC (15y Male)



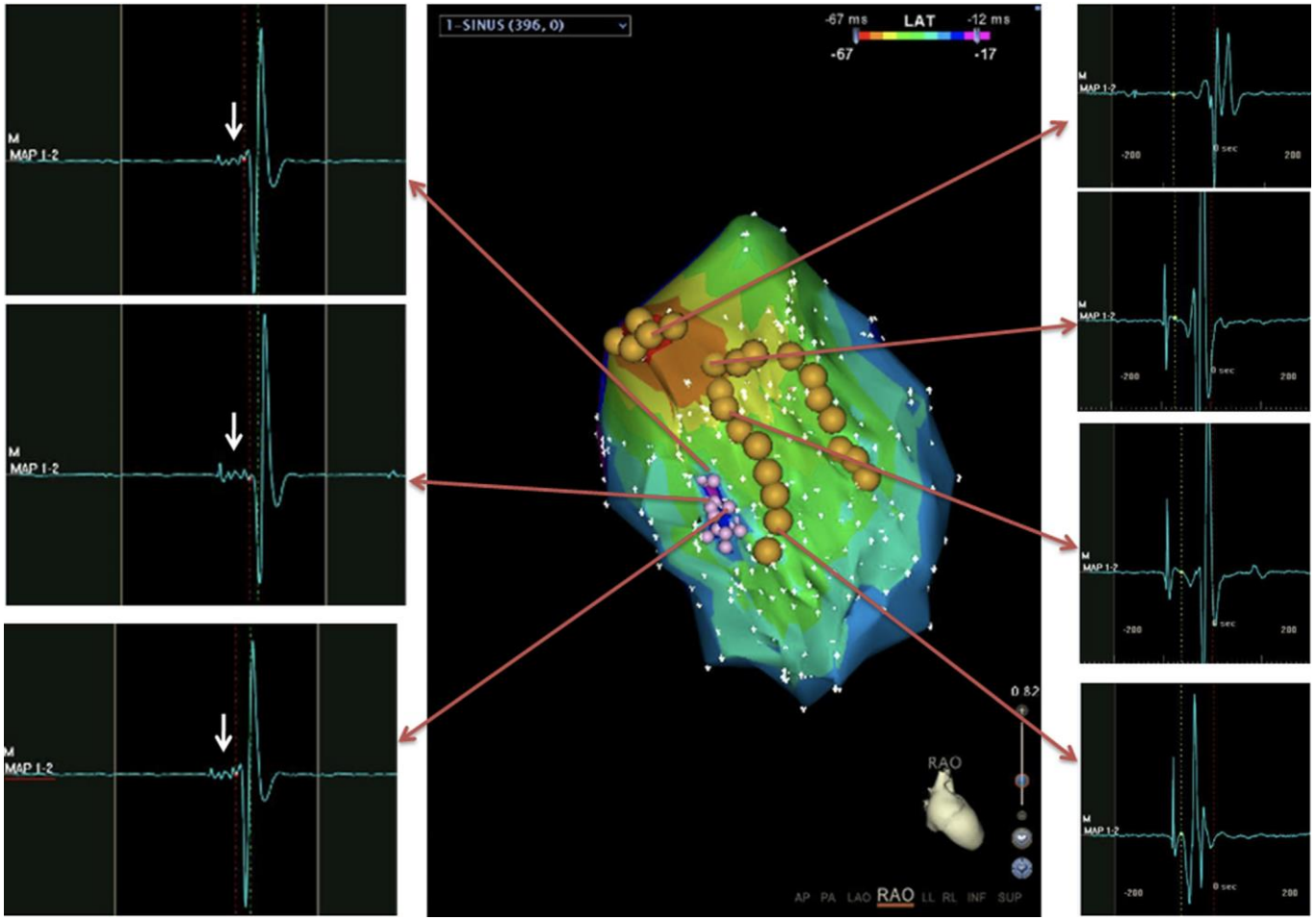


RVOT PVC (20y Female)





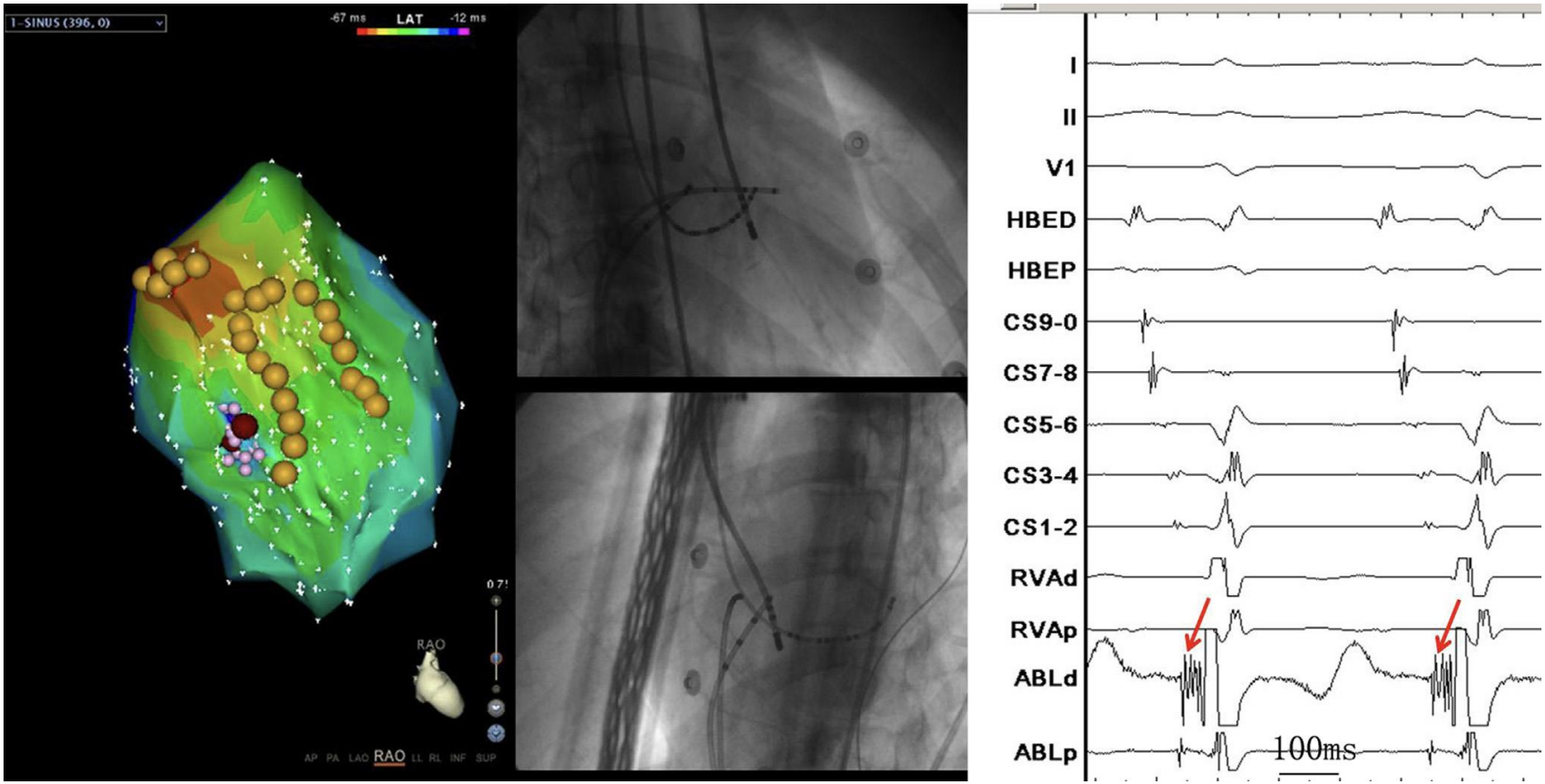
Identification of the left His-Purkinje system(HPS) during sinus rhythm(SR)



fragmented antegrade Purkinje potential (FAP)

Zhan XZ, et al. Heart Rhythm. 2016;13:1460-7.

Electroanatomic mapping, radiograph, and intracardiac electrogram target at the site of successful ablation during sinus rhythm (SR)



FAP; 23/24 ILVT: 5/15 Lt AP ($p<0.01$)
RFA success at FAP in 23/23 ILVT

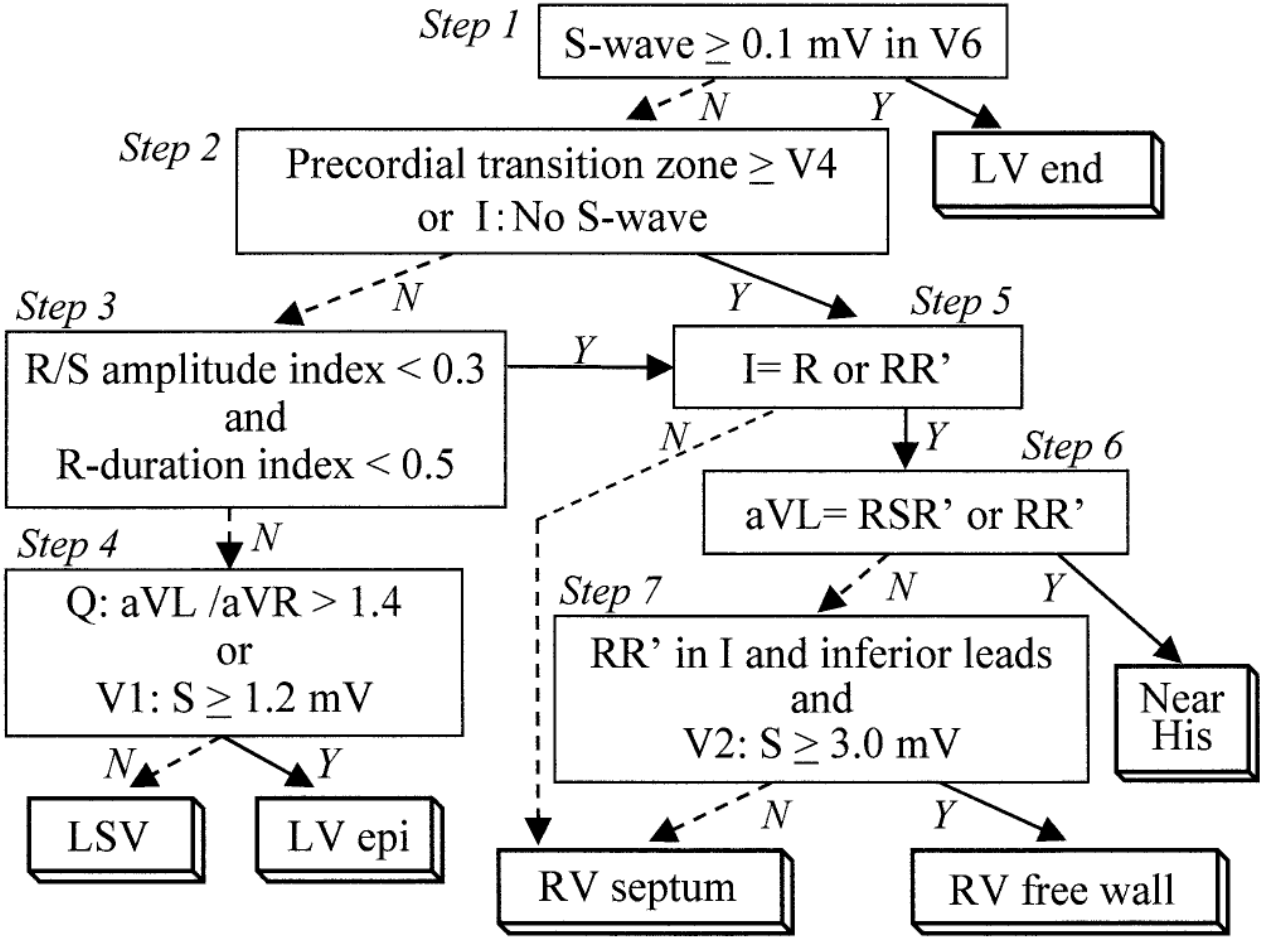
↙□ fragmented antegrade Purkinje potential (FA

Zhan XZ, et al. Heart Rhythm. 2016;13:1460-7.

Conclusion

- The incidence of idiopathic VT in children was 75% in outflow VT, and 18% in verapamil sensitive VT.
- For successful ablation of outflow VT, not only to find earliest activation site, and almost perfect pace mapping site, but to find prepotential is mandatory for difficult ablation patients.
- For successful ablation of verapamil sensitive VT, simultaneous recording of P1 and P2 potential, earliest activation of P2 potential, best pacemapping site, and the portion of PPI equal VTCL could be suitable ablation site. Fragmentation near the P2 potential might become suitable ablation site for verapamil sensitive VT.

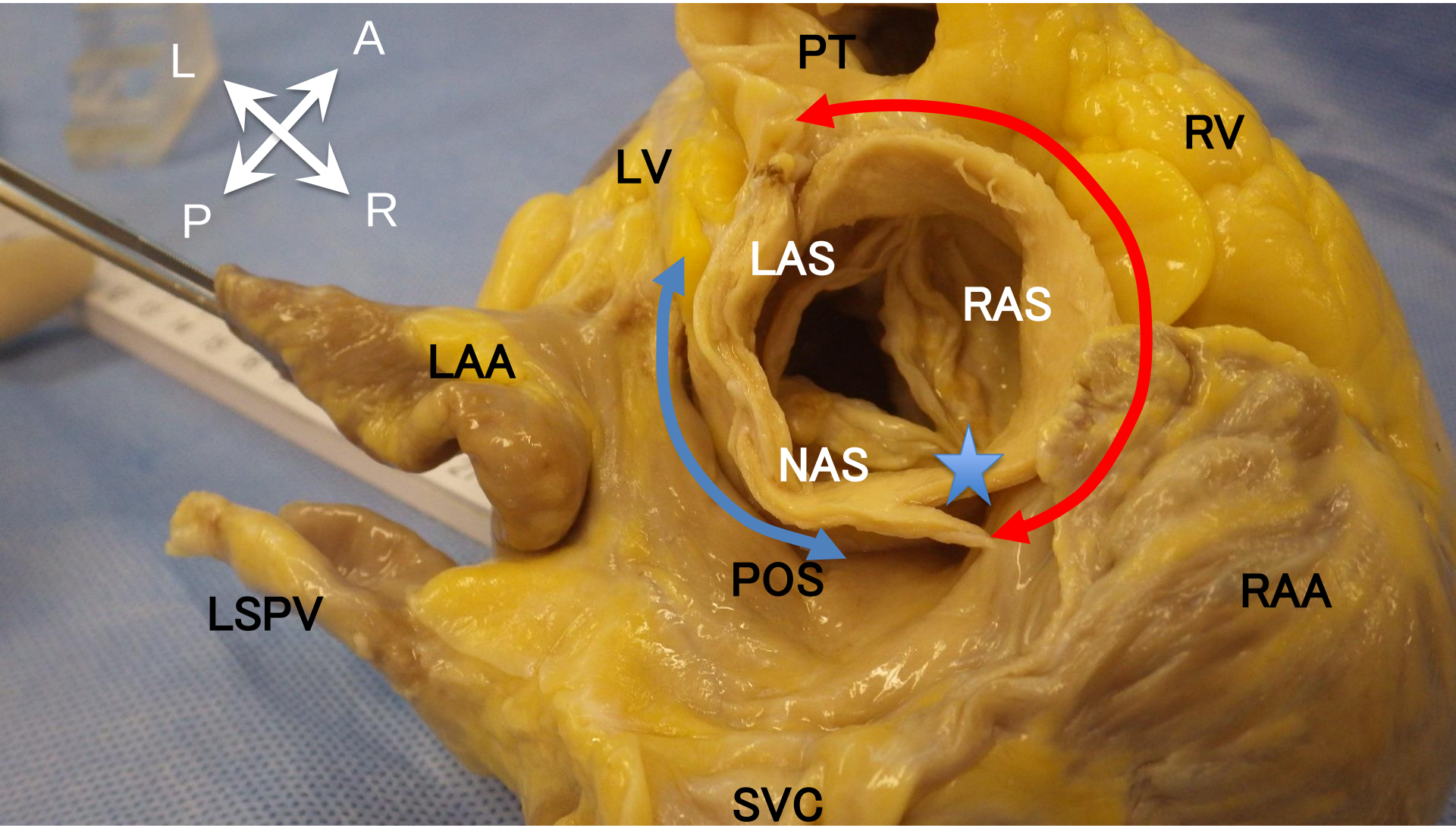
Stepwize ECG algorithm for determination of the location of the origin of OT-VT



R/S amplitude index: R/S in V1 or V2
R duration index: R/QRS duration V1, V2

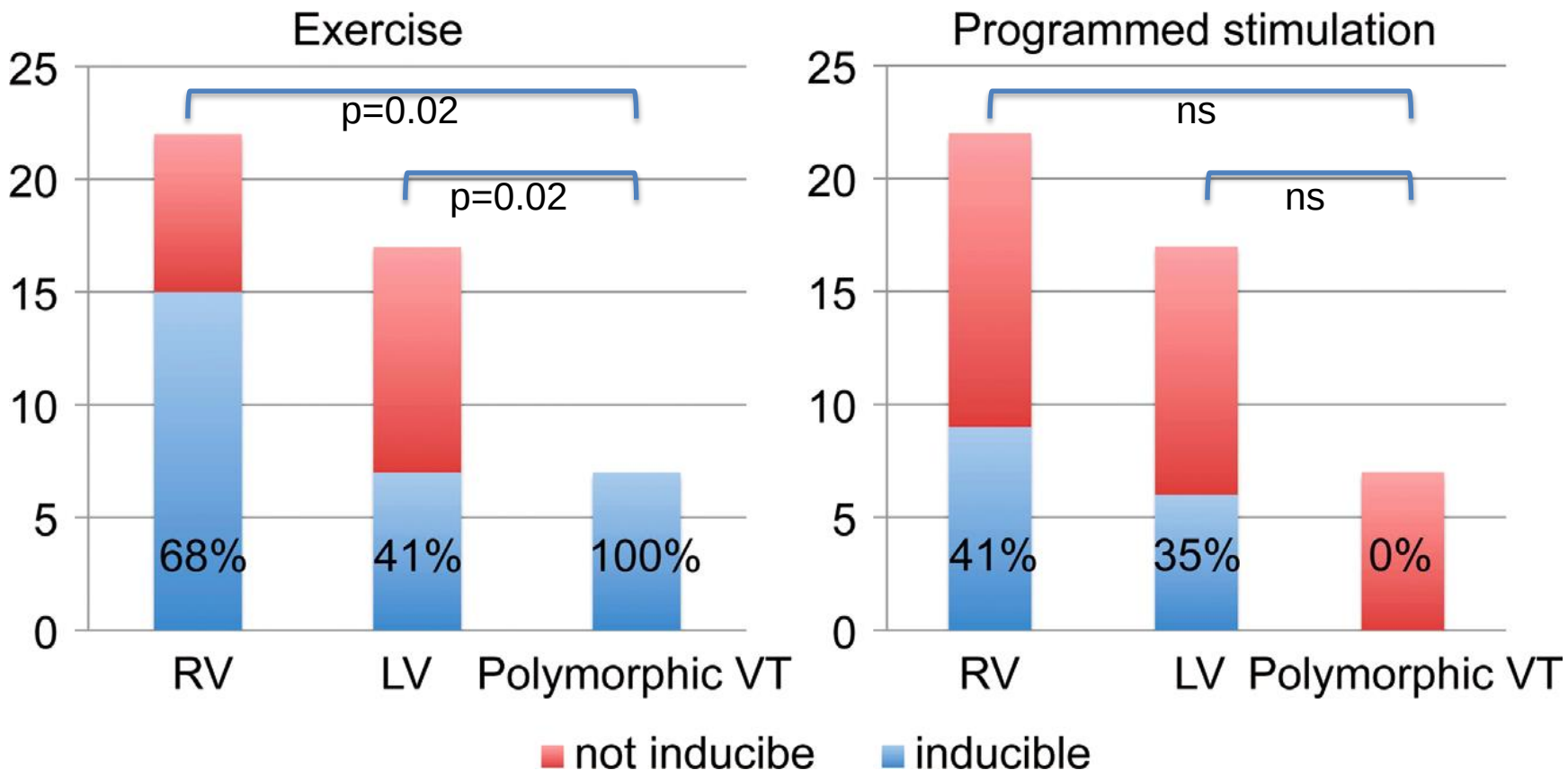
LSV: Left sinus of valsalva

Ito S, et al. JCE, 2013;14:1280-1286



Courtesy of Prof. Osamu Igawa

Induction of ventricular tachycardia (VT) by exercise and programmed stimulation



Fukuhara J, Sumitomo N, et al. Circ J. 2011;75:672-6.