

PLATFORM OF LABORATORIES FOR ADVANCES IN CARDIAC EXPERIENCE

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ARITMOLOGIA CLINICA E CARDIOLOGIA INTERVENTISTICA IN ETÀ
PEDIATRICA

# DIAGNOSTICA DELL'ESPRESSIONE ARITMICA NEI PAZIENTI PEDIATRICI: BEST PRACTICE

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Responsabile Unità Funzionale Aritmologia Ospedale San Raffaele Milano







# Epidemiology





## Epidemiology and pathophysiology of arrhythmias in structurally normal heart

- The prevalence of supraventricular arrhythmia (SVA) is 2,25/1000 people with an annual incidence in children < 19 years of age of 13/100.000
- In infancy, SVT results predominantly from accessory pathways, followed by a small number of cases of ectopic atrial tachycardia
- In teenage life, there is a significant increase in the prevalence of atrioventricular nodal reentry tachycardia (AVNRT), particularly in females
- Ventricular tachycardia (VT) is rare in children, representing only 1.8% of undergoing EPS. It is typically associated with a structurally normal heart, although children presenting with VT require careful evaluation for early manifestations of underlying cardiac disease



Europace (2013) **15**, 1337–138 doi:10.1093/europace/eut082

**EHRA/AEPC CONSENSUS STATEMENT** 

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

Josep Brugada<sup>1\*</sup>, Nico Blom<sup>2</sup>, Georgia Sarquella-Brugada<sup>3</sup>, Carina Blomstrom-Lundqvist<sup>4</sup>, John Deanfield<sup>5</sup>, Jan Janousek<sup>6</sup>, Dominic Abrams<sup>7</sup>, Urs Bauersfeld<sup>8†</sup>, Ramon Brugada<sup>9</sup>, Fabrizio Drago<sup>10</sup>, Natasja de Groot<sup>11</sup>, Juha-Matti Happonen<sup>12</sup>, Joachim Hebe<sup>13</sup>, Siew Yen Ho<sup>14</sup>, Eloi Marijon<sup>15</sup>, Thomas Paul<sup>16</sup>, Jean-Pierre Pfammatter<sup>17</sup>, and Eric Rosenthal<sup>18</sup>



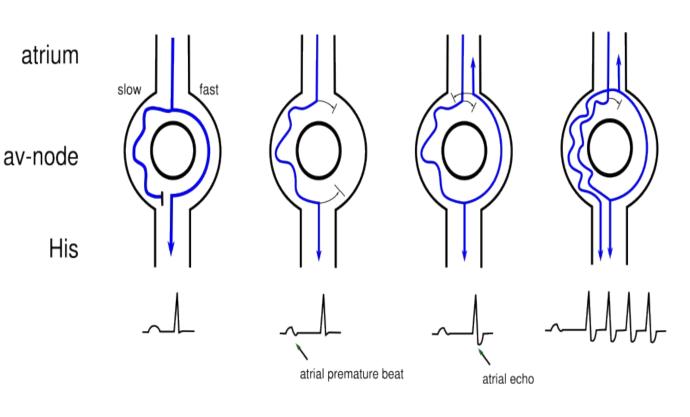


# Supraventricular arrhythmias



# Atrioventricular nodal re-entrant tachycardia (AVNRT)

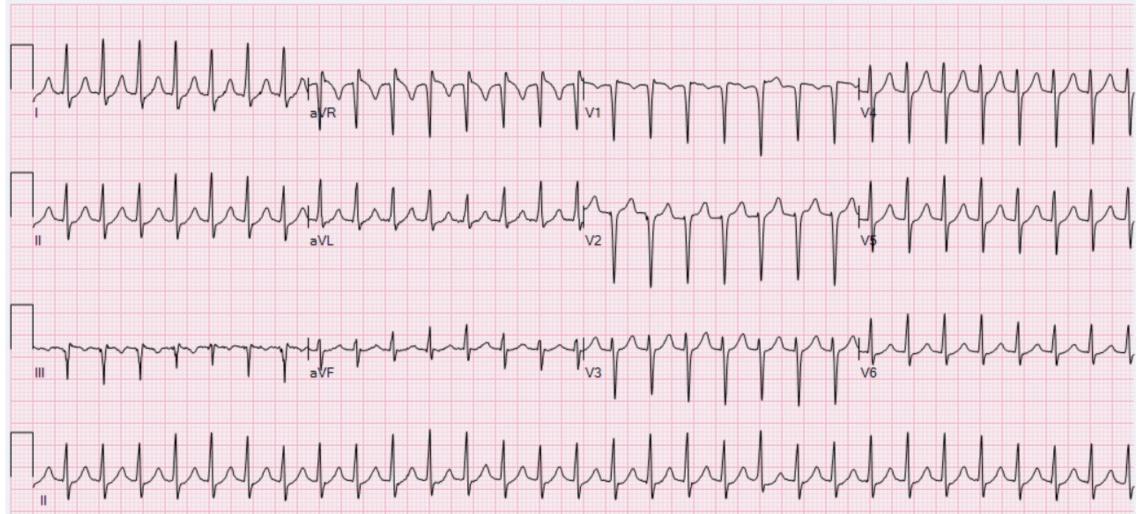
- The most common mechanism of AVNRT in children is anterograde slow pathway activation followed by retrograde activation of the atria via the fast pathway (slow-fast) and anterograde ventricular activation occurring shortly after via the His-Purkinje system.
- In the less common atypical AVNRT (fast-slow) the circuit is reversed, and earliest atrial activation seen in the low right atrium, producing a long RP tachycardia on the surface ECG with an inverted P-wave shortly before the following QRS complex





### 12 leads ECG of an AVNRT







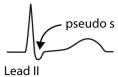
## 12 leads ECG findings in AVNRT



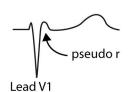
#### A) Typical AVNRT (slow-fast): 90% of all cases



In most cases the P-wave is hidden in the QRS complex.



The P-wave is sometimes seen after the QRS complex. It will present itself as "pseudo s" in lead II and "pseudo r" in lead V1.



#### B) Atypical AVNRT (fast-slow): 10% of all cases



P-wave before QRS complex

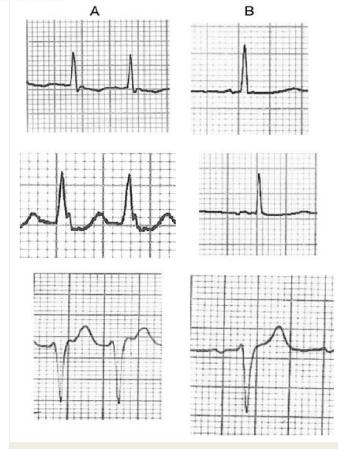
#### C) Very atypical AVNRT (slow-slow): <1% of all cases



P-wave on the ST-T segment

# Utility of the aVL lead in the electrocardiographic diagnosis of atrioventricular node re-entrant tachycardia

Darío Di Toro<sup>1,2\*</sup>, Claudio Hadid<sup>1,2</sup>, Carlos López<sup>1</sup>, Juan Fuselli<sup>2</sup>, Vidal Luis<sup>1</sup>, and Carlos Labadet<sup>1,2</sup>



**Figure 1** Three types of aVL notch (A) during atrioventricular node re-entrant (B) sinus rhythm.







Classification of AVNRT Types							
		AH/HA	VA (His)	Usual ERAA			
Typical AVNRT	Slow-fast	>1	<60ms	RHis, CS os, LHis			
Atypical AVNRT	Fast-slow	<1	>60ms	CS os, LRAS, dCS			
	Slow-slow	>1	>60ms	CS os, dCS			

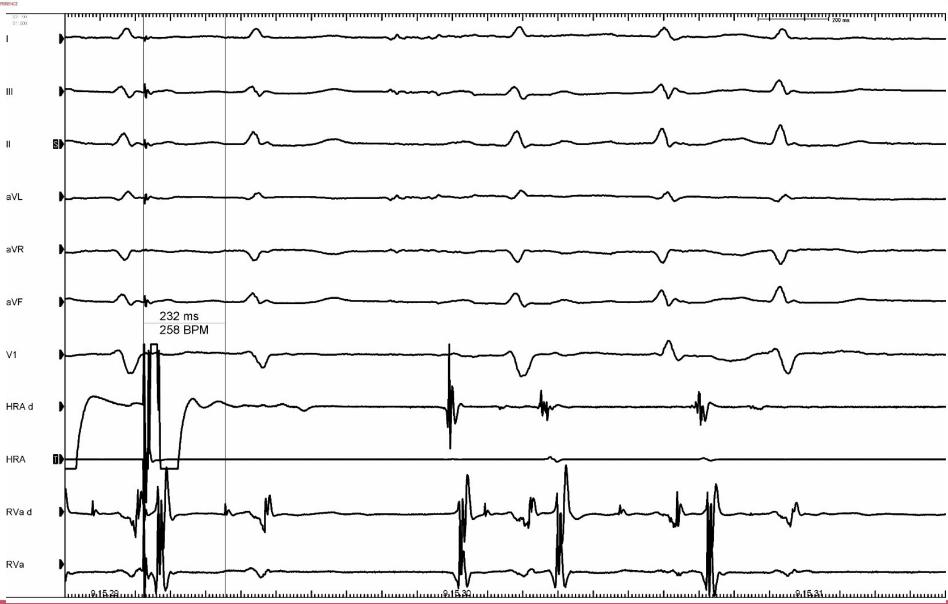
VA indicates interval measured from the onset of ventricular activation on surface ECG to the earliest deflection of the atrial activation in the His bundle electrogram; ERAA, earliest retrograde atrial activation; RHis, His bundle electrogram recorded from the right septum; LHis, His bundle electrogram recorded from the left septum; LRAS, low right atrial septum; CS os, ostium of the coronary sinus; and dCS, distal coronary sinus







### **Decremental**



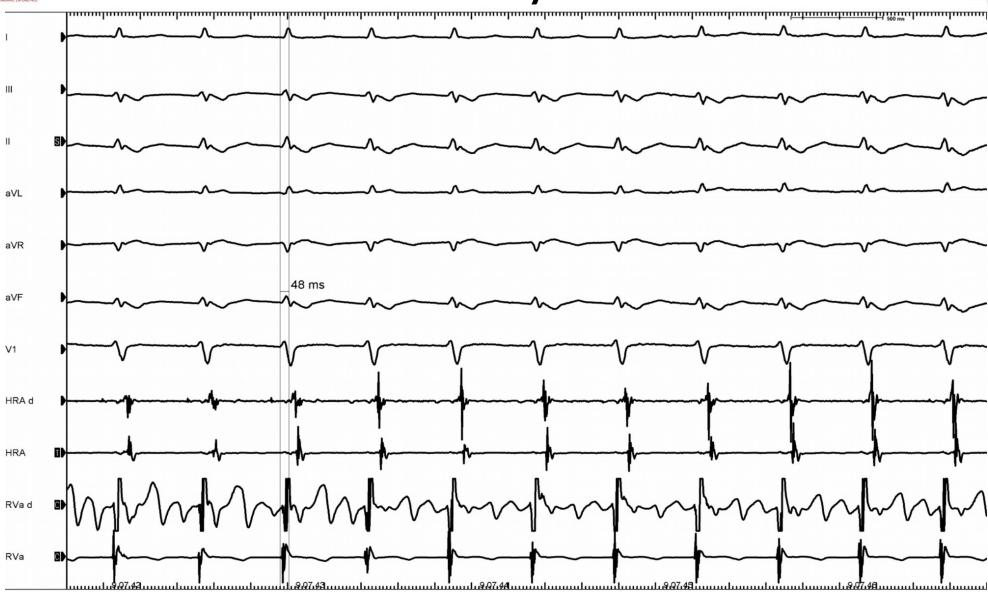


# Jump



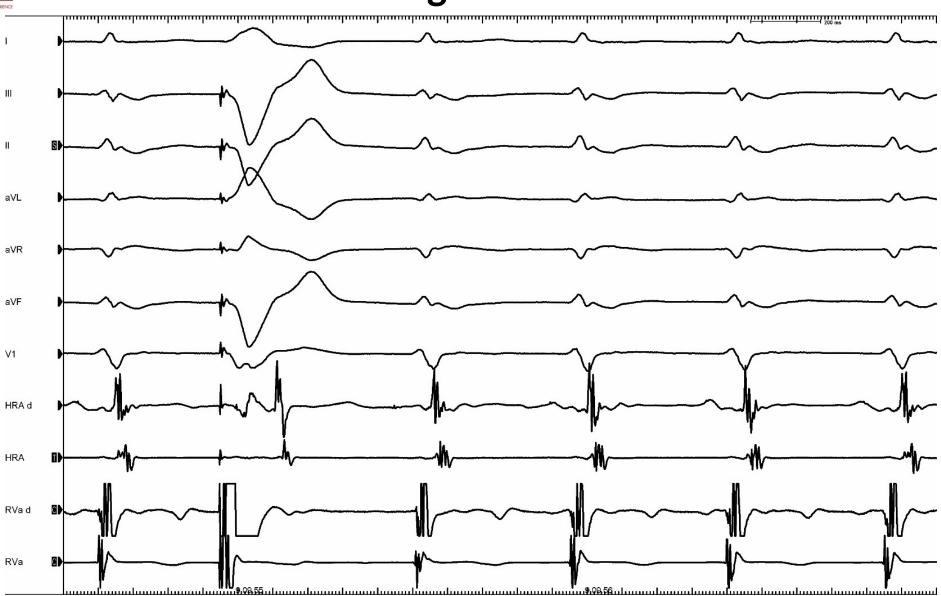


# **Tachy**



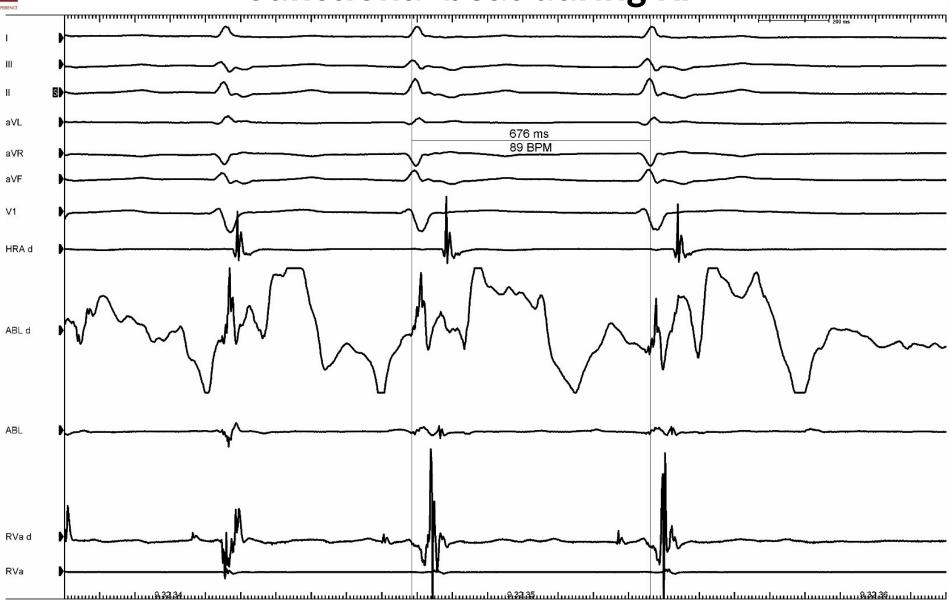


# **AVNRT: Pacing ad his refrattario**





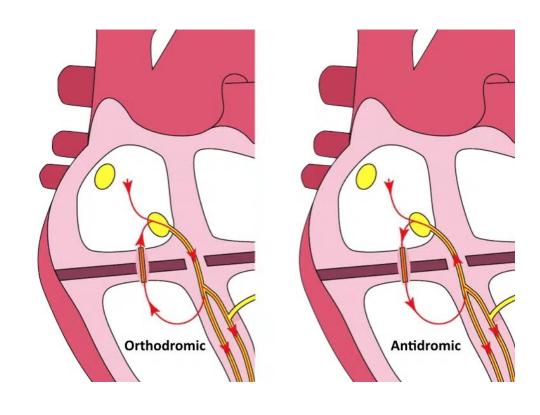
# **Junctional beat during RF**





# Atrioventricular reentry tachycardia (AVRT)

- Atrioventricular reentry tachycardia (AVRT) is induced by a muscular accessory pathway spanning the fibrous AV junction and providing continuity between atrial and ventricular myocardium, at a site electrophysiologically distinct from the AV node and proximal His-Purkinje system.
- Such connections have been described in the developing human heart, normally regressing by 20 weeks gestation. However, spontaneous regression of pathway function in infancy is well documented, although in what proportion of patients symptoms redevelop later in life remains uncertain.

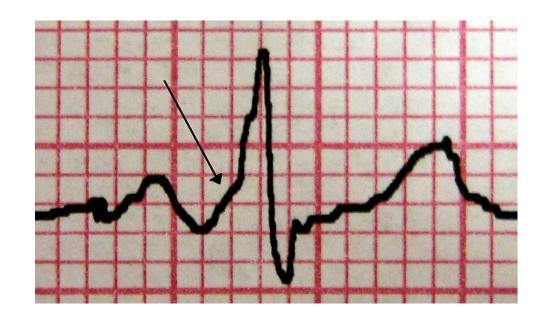






## 12 leads ECG manifest accessory pathway features

- When equipped with antegrade conduction, the accessory pathways manifest themselves on the surface ECG with particular signs:
  - Short PR interval (< 120 msec)</li>
  - Initial kneading of the QRS complex (delta wave)
  - Enlargement of the QRS complex (> 120 msec)
- The occult accessory pathways have only the retroconduction property.
- The non-manifest accessory pathways are characterized by electrophysiological properties of anteroconduction inferior to those of the normal Hissian conduction system, such that in basal conditions they are not visible.

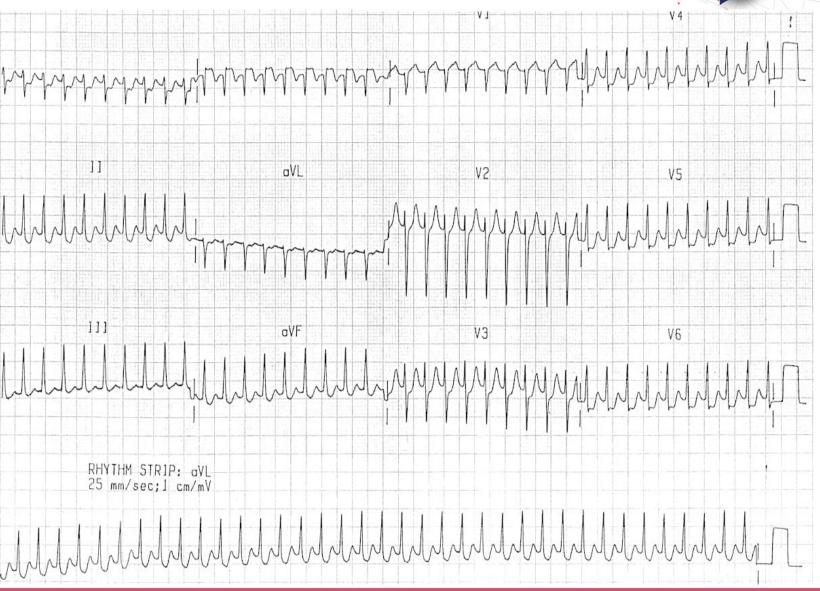




### 12 leads ECG in orthodromic AVRT



- Regular rythm
- Narrow QRS complexes
- Inverted P waves with an RP interval usually <50% of the tachycardia RR interval
- Constant RP interval regardless of the tachycardia cycle length

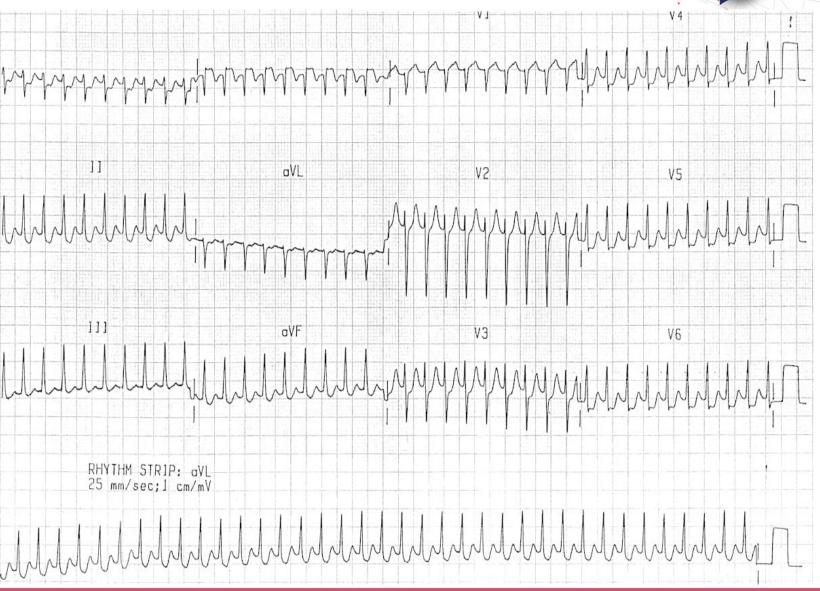




### 12 leads ECG in antidromic AVRT



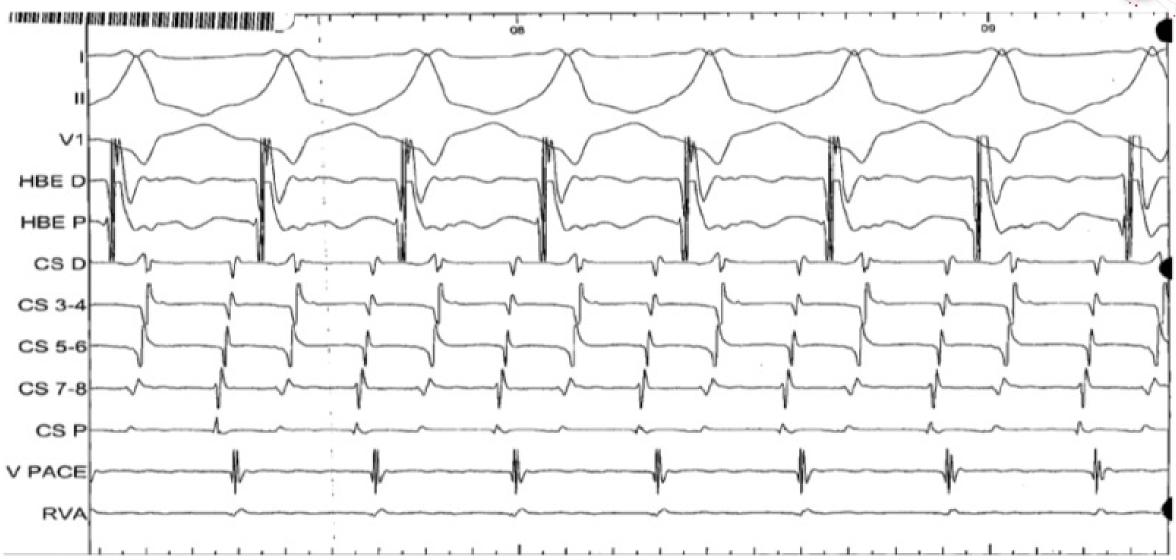
- Regular rythm
- Wide QRS complexes
- Inverted P waves with an RP interval usually >50% of the tachycardia RR interval
- Constant RP interval regardless of the tachycardia cycle length





# **Intracardiac ECG during EP study**

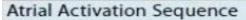






### **Exclusion of ANVRT**





Eccentric atrial activation sequence during the SVT excludes AVNRT (with rare exceptions of left-sided insertion of fast or slow AVN pathways).

#### Effects of BBB

If VA interval or tachycardia CL prolongs on development of BBB, AVNRT is excluded.

Spontaneous Oscillations of Tachycardia CL

Spontaneous changes in tachycardia CL accompanied by constant VA interval make AVNRT unlikely.

#### VES Delivered During SVT

VES delivered during SVT when the HB is refractory that resets or terminates the SVT excludes AVNRT.

VES during SVT that captures the atrium at the same coupling interval as that of the VES (exact capture phenomenon) excludes AVNRT.

VES delivered during SVT that captures the atrium at a shorter coupling interval than that of the VES (paradoxic capture phenomenon) excludes AVNRT.

#### Entrainment of SVT by Atrial Pacing

AH<sub>atrial pacing</sub> - AH<sub>SVT</sub> < 20 msec excludes AVNRT.

#### Entrainment of SVT by Ventricular Pacing

VA<sub>ventricular pacing</sub> – VA<sub>SVT</sub> < 85 msec excludes AVNRT.

PPI - tachycardia CL < 115 msec excludes AVNRT.

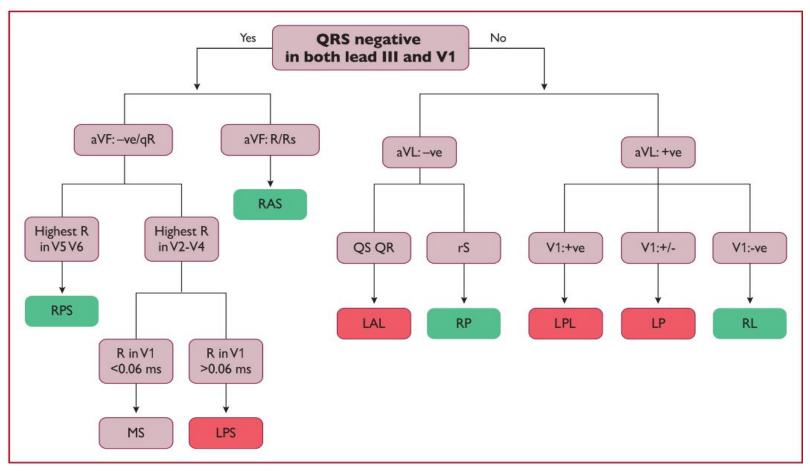
Ventricular fusion during entrainment indicates AVRT and excludes AVNRT.







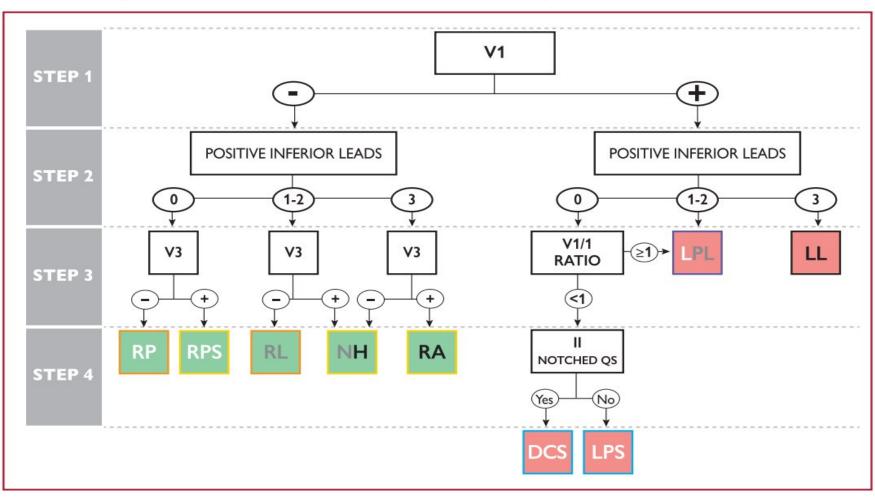
## Localization of accessory pathways in AVRT



The St George's algorithm for the localization of accessory pathways



# Localization of accessory pathways in the presence of maximum (spontaneous or evoked) pre-excitation





# This article originally appeared in The American Heart Journal in volume 5, issue 6, August 1930, pgs. 685–704



**Figure 1.** (Case I) Right bundle-branch block. The P-R interval is 0.1 second. The rate is 72. Time intervals for this and succeeding figures = 0.2 second. Horizontal lines cut off intervals of  $10^{-4}$  volt.



**Figure 6.** (Case III) Right bundle-branch block. The P-R interval is well under 0.1 second. The rate varies between 60 and 70.

### The American Heart Journal

Vol. V August, 1930

No. 6

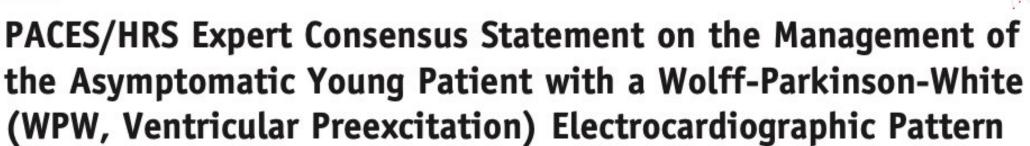
#### Original Communications

BUNDLE-BRANCH BLOCK WITH SHORT P-R INTERVAL IN HEALTHY YOUNG PEOPLE PRONE TO PAROXYSMAL TACHYCARDIA

LOUIS WOLFF, M.D., BOSTON, MASS., JOHN PARKINSON, M.D., LONDON, Eng., and Paul D. White, M.D., Boston, Mass.







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 Foundation (ACCF), the American Heart Association (AHA), the American Academy of Pediatrics (AAP), and the Canadian Heart Rhythm Society (CHRS)

- Prevalence of asymptomatic WPW: 0.1% 0.3 %;
- In pediatric patients with asymptomatic WPW, the incidence of SCD is reported to be between 0.0012 and 0.6%/year
- Most studies report a very low incidence of SCD, which seems to be lower in childhood than in adulthood





# Aborted Sudden Death in the Wolff-Parkinson-White Syndrome

Carl Timmermans, MD, Joep L.R.M. Smeets, MD, Luz-Maria Rodriguez, MD, Georgios Vrouchos, MD, Adri van den Dool, BS, and Hein J.J. Wellens, MD

In a population of 690 patients with Wolff-Parkinson-White (WPW) syndrome referred to our hospital from January 1979 to February 1995, 15 patients (2.2%) had an aborted sudden death out of the hospital. This retrospective study examines their clinical and electrophysiologic characteristics. Gender, accessory pathway localization, and presence of multiple accessory pathways were compared between patients with and without spontaneous ventricular fibrillation (VF). Whereas gender and the presence of multiple accessory pathways did not significantly differ between both groups, septally located pathways occurred significantly more

often in the VF group. In patients with aborted sudden death, spontaneous VF was found significantly more often in men (13 of 15). VF was the first manifestation of the WPW syndrome in 8 patients. The remaining 7 patients had documented episodes of atrial fibrillation, circus movement tachycardia, or both (n = 2). Ten of the 15 patients were exercising or under emotional stress at the time of aborted sudden death. Only 1 patient had 2 accessory pathways. The location of the accessory pathway was septal (midseptal or posteroseptal) in 11 patients, left lateral in 4, and right lateral in 1. (Am J Cardiol 1995;76:492-494)

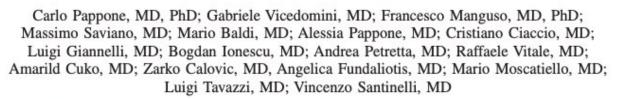
In a large retrospective series of 690 WPW patients, 15 (2,2%) had aborted sudden death, and in 8 of them (0,50%) VF was the first clinical manifestation of the syndrome

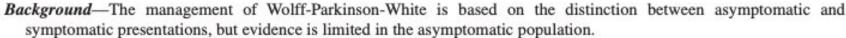
THE AMERICAN JOURNAL OF CARDIOLOGY® VOL. 76 SEPTEMBER 1, 1995



#### Wolff-Parkinson-White Syndrome in the Era of Catheter Ablation

**Insights From a Registry Study of 2169 Patients** 





Methods and Results—The Wolff-Parkinson-White registry was an 8-year prospective study of either symptomatic or asymptomatic Wolff-Parkinson-White patients referred to our Arrhythmology Department for evaluation or ablation. Inclusion criteria were a baseline electrophysiological testing with or without radiofrequency catheter ablation (RFA). Primary end points were the percentage of patients who experienced ventricular fibrillation (VF) or potentially malignant arrhythmias and risk factors. Among 2169 enrolled patients, 1001 (550 asymptomatic) did not undergo RFA (no-RFA group) and 1168 (206 asymptomatic) underwent ablation (RFA group). There were no differences in clinical and electrophysiological characteristics between the 2 groups except for symptoms. In the no-RFA group, VF occurred in 1.5% of patients, virtually exclusively (13 of 15) in children (median age, 11 years), and was associated with a short accessory pathway antegrade refractory period (P<0.001) and atrioventricular reentrant tachycardia initiating atrial fibrillation (P<0.001) but not symptoms. In the RFA group, ablation was successful in 98.5%, and after RFA, no patients developed malignant arrhythmias or VF over the 8-year follow-up. Untreated patients were more likely to experience malignant arrhythmias and VF (log-rank P<0.001). Time-dependent receiver-operating characteristic curves for predicting VF identified an optimal anterograde effective refractory period of the accessory pathway cutoff of 240 milliseconds.

Conclusions—The prognosis of the Wolff-Parkinson-White syndrome essentially depends on intrinsic electrophysiological properties of AP rather than on symptoms. RFA performed during the same procedure after electrophysiological testing is of benefit in improving the long-term outcomes. (Circulation. 2014;130:811-819.)

In an 8-year prospective registry of 550 initially asymptomatic subjects, ventricular fibrillation developed in 2.4% and malignant arrhythmias in 8.7%, primarily in patients with accessory pathway anterograde refractory period, 240 ms and AV re-entrant tachycardia initiating AF

DOI: 10.1161/CIRCULATIONAHA.114.011154

Circulation is available at http://circ.ahajournals.org







# Invasive risk stratification in asymptomatic patients

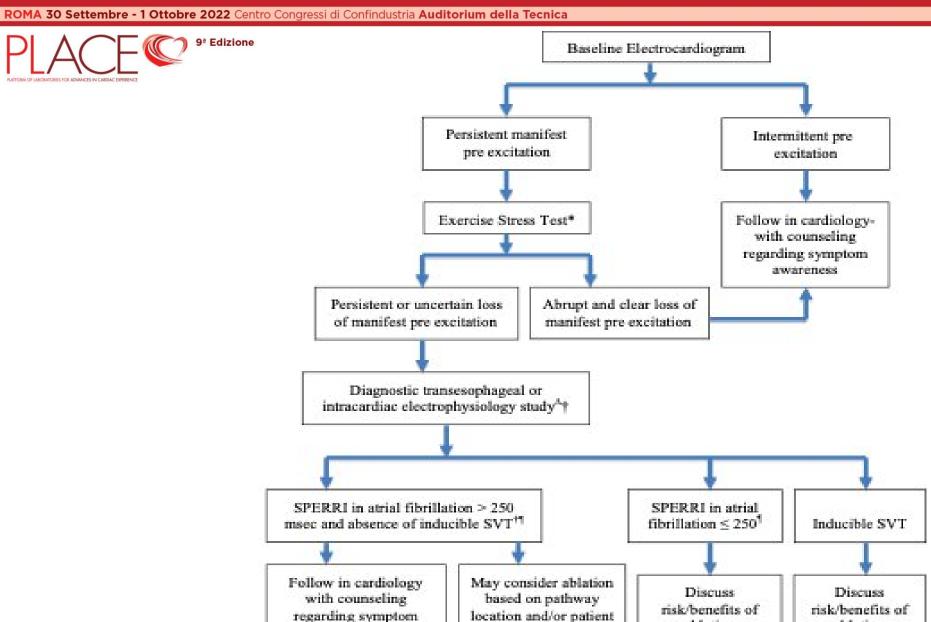
- The measurement of SPERRI during AF induction
- Anterograde and retrograde AV characteristics
- The anterograde effective refractory period of the AP (APERP)
  - The number and location of AP

A SPERRI IN AF ≤ 250ms OR AND APERP OF ≤ 250ms ARE CRITICAL VALUES



# Recommendations for young asymptomatic patients (8-21 years) with WPW ECG pattern

- Utilization of invasive risk stratification (transesophageal or intracardiac) to assess SPERRI (IIA B/C);
- Patients at higher risk of SCD (SPERRI ≤ 250 msec) consider catheter ablation (IIA B/C);
- Patients at lower risk (SPERRI > 250) msec consider to defer CA if the ablation may incur an increased risk of adverse events (II B/C);
- If the patients develop cardiovascular symptoms may be eligible to CA



characteristics

(Class IIB)

ablation.

(Class IIA)

ablation

(Class IIB)

regarding symptom

awareness

(Class IIA)

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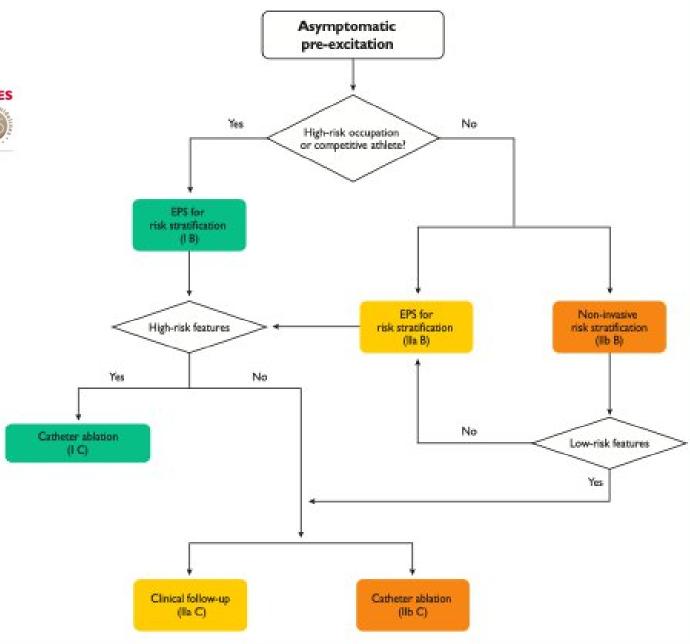


European Heart Journal (2020) 41, 655—720 European Society doi:10.1093/eurheartj/ehz467



# 2019 ESC Guidelines for the management of patients with supraventricular tachycardia

- One in five pts will develop an arrhythmia
  - AVRT 80 %;
  - AF 20-30%;
- The risk of cardiac arrest/FV has been estimated at 2.4 per 1000 person years







# Clinical ad electrophysiological features associated with risk of SCD

- Younger age;
- Inducibility of AV-reciprocating tachycardia during EPS;
- Multiple APs;
- SPERRI ≤ 250 msec;
- ERP of AP ≤ 250 msec

IN THE BASELINE STATE OR DURING ISOPROTERENOL INFUSION

Pediatric Cardiology (2019) 40:892–900 https://doi.org/10.1007/s00246-019-02110-6

#### **REVIEW ARTICLE**



# Management of Asymptomatic Wolff–Parkinson–White Pattern in Young Patients: Has Anything Changed?

Daniela Raposo<sup>1</sup> · Natália António<sup>1,2</sup> · Helena Andrade<sup>3</sup> · Pedro Sousa<sup>2</sup> · António Pires<sup>1,3</sup> · Lino Gonçalves<sup>1,2</sup>

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#### Abstract

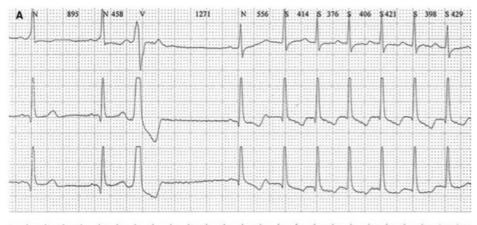
The approach to pediatric asymptomatic Wolff–Parkinson–White (WPW) patients is controversial. The objective of this review is to update the last consensus of specialists of the Pediatric and Congenital Electrophysiology Society/Heart Rhythm Society on this subject in order to summarize the most recent evidence on the management of young patients with asymptomatic WPW pattern. A systematic review of the literature published between 2008 and 2018 was performed taking into account the protocol of the Preferred Reporting Items for Systematic Reviews and Meta-Analyses (PRISMA) in PubMed (including Cochrane), Embase, and Web of Science. Observational, experimental, and multicentric studies were included. Out of a total of 37 articles selected, 4 were considered eligible. Most studies considered a cutoff age of 8 or greater as recommended in the 2012 consensus. The identification of a shortest pre-excitatory RR interval (SPERRI) ≤ 250 ms seems to be the best predictor for risk stratification. The importance of routine isoprenaline use to improve the sensitivity of the electrophysiological study to identify patients at high risk of sudden death was consensual. Prophylactic ablative therapy has been indicated in asymptomatic children with an accessory pathway (AP) who have a low SPERRI and/or a low effective anterograde period of the AP and/or multiple APs. Despite the evidence found in the most recent studies, more studies are warranted in this setting.

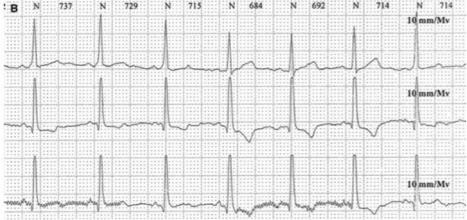






### Intermittent loss of preexcitation:





- has been associated with longer ERPs;
  - One fifth of pts have AP ERPs ≤ 250 ms;

# Development of Rapid Preexcited Ventricular Response to Atrial Fibrillation in a Patient with Intermittent Preexcitation

LEE W. GEMMA, M.D., LEONARD A. STEINBERG, M.D., ERIC N. PRYSTOWSKY, M.D., and BENZY J. PADANILAM, M.D.

From the St. Vincent Medical Group, Indianapolis, Indiana, USA

Intermittent preexcitation during sinus rhythm is indicative of an accessory pathway at a very low risk for sudden death. We present the case of a 49-year-old man with intermittent preexcitation who subsequently developed rapid atrial fibrillation with a shortest preexcited R-R interval of 230 milliseconds. Electrophysiology study showed intermittent preexcitation at baseline and 1:1 anterograde accessory pathway conduction to 220 milliseconds in the presence of 1 mcg/min isoproterenol infusion. The pathway was successfully ablated at the lateral mitral annulus. Accessory pathways highly sensitive to catecholamines may show intermittent preexcitation at baseline with potential for rapid conduction during atrial fibrillation and sudden death. (J Cardiovasc Electrophysiol, Vol. 24, pp. 347-350, March 2013)



Catheter ablation of Aps in the anteroseptal or midseptal region is associated with small risk of AV Block

#### **EP CASE EXPRESS**

doi:10.1093/europace/euu157 Online publish-ahead-of-print 25 June 2014

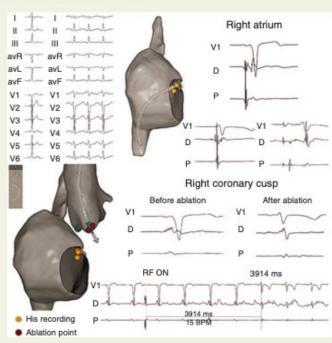
### Successful radiofrequency ablation of an anteroseptal accessory pathway from the right coronary cusp

#### Teresa Oloriz, Simone Gulletta, and Paolo Della Bella\*

Arrhythmia Unit and Electrophysiology Laboratories, Department of Cardiology and Cardiothoracic Surgery, Ospedale San Raffaele, Via Olgettina 60, Milan, Italy.

\* Corresponding author. Tel: +39 02 26436247; fax: +39 02 26437326. E-mail address: dellabella.paolo@hsr.it

A 13-year-old presented recurrent narrow QRS tachycardia and pre-excitation pattern consistent with a right-sided anteroseptal accessory pathway (AP) was referred for ablation. The ablation catheter (a 3.5 mm ThermoCool SmartTouch Biosense Webster) was placed in the anteroseptal region at the site of earliest ventricular activation. Mechanical bumping of the AP disclosed, during nodal atrioventricular (AV) conduction, the presence of the His bundle potential at that site, suggesting close proximity of the AV conduction to the AP. A complex electrogram with atrial activity followed by an early and fragmented ventricular electrogram preceded by a pre-potential was recorded at the right coronary cusp (RCC). The application of radiofrequency (10-20 W) (contact force 15-30 g) successfully abolished the conduction through the AP in <4 s (Figure). The measured distance between the ablation site and the His recording on the right atrium was 16 mm. As the conduction system penetrates to the left, it becomes located at the base of the interleaflet triangle between the non-coronary and RCC. Ablation on the RCC requires RF energy to be very cautiously and limitedly delivered. Electroanatomical mapping allowed stable contact



and the contact force provided an effective ablation modality of RF.

The full-length version of this report can be viewed at: http://www.escardio.org/communities/EHRA/publications/ep-case-reports/ Documents/Successful-radiofrequency-ablation.pdf.

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ORIGINAL PAPER



#### Catheter ablation in ASymptomatic PEDiatric patients with ventricular preexcitation: results from the multicenter "CASPED" study

Marta Telishevska<sup>1</sup> · J. Hebe<sup>2</sup> · T. Paul<sup>3</sup> · J. H. Nürnberg<sup>2</sup> · U. Krause<sup>3</sup> · R. Gebauer<sup>4</sup> · M. Gass<sup>5</sup> · C. Balmer<sup>6</sup> · F. Berger<sup>6</sup> · S. Molatta<sup>7</sup> · M. Emmel<sup>8</sup> · W. Lawrenz<sup>9</sup> · T. Kriebel<sup>10</sup> · G. Hessling<sup>1</sup>

Received: 5 September 2018 / Accepted: 26 November 2018 / Published online: 5 December 2018 © Springer-Verlag GmbH Germany, part of Springer Nature 2018

#### Abstract

**Background** As there are limited data about the clinical practice of catheter ablation in asymptomatic children and adolescents with ventricular preexcitation on ECG, we performed the multicenter "CASPED" (Catheter ablation in ASymptomatic PEDiatric patients with Ventricular Preexcitation) study.

Methods and results In 182 consecutive children and adolescents aged between 8 and 18 years (mean age  $12.9 \pm 2.6$  years; 65% male) with asymptomatic ventricular preexcitation, a total of 196 accessory pathways (APs) were targeted. APs were right sided (62%) or left sided (38%). The most common right-sided AP location was the posteroseptal region (38%). Ablation was performed using radiofrequency (RF) energy (93%), cryoablation (4%) or both (3%). Mean procedure time was  $137.6 \pm 62.0$  min with a mean fluoroscopy time of  $15.6 \pm 13.8$  min. A 3D mapping or catheter localization system was used in 32% of patients. Catheter ablation was acutely successful in 166/182 patients (91.2%). Mortality was 0% and there were no major periprocedural complications. AP recurrence was observed in 14/166 patients (8.4%) during a mean follow-up time of  $19.7 \pm 8.5$  months. A second ablation attempt was performed in 20 patients and was successful in 16/20 patients (80%). Overall, long-term success rate was 92.3%.

Conclusion In this retrospective multicenter study, the outcome of catheter ablation for asymptomatic preexcitation in children and adolescents irrespective of antegrade AP conduction properties is summarized. The complication rate was low and success rate was high, the latter mainly depending on pathway location. The promising results of the study may have future impact on the ongoing risk-benefit discussion regarding catheter ablation in the setting of asymptomatic preexcitation in children and adolescents.

In the CASPED retrospective multicenter study, involving 182 children and adolescents with asymptomatic pre-excitation, catheter ablation achieved a 91% success rate without significant complication





Recommendations for the management of patients with asymptomatic pre-excitati als with a

Catheter ablation is associated with high cure (>95%) and low risk of major complication (<0,5%)

Recommendation	Classa	Level <sup>b</sup>
Performance of an EPS, with the use of isopre- naline, is recommended to risk stratify individ- uals with asymptomatic pre-excitation who have high-risk occupations/hobbies, <sup>c</sup> and those who participate in competitive athletics. <sup>439,450–452,454–460</sup>	1	В
Catheter ablation is recommended in asymptomatic patients in whom electrophysiology testing with the use of isoprenaline identifies high-risk properties, such as SPERRI ≤250 ms, AP ERP ≤250 ms, multiple APs, and an inducible AP-mediated tachycardia.	1	В
Catheter ablation is recommended in high-risk patients with asymptomatic pre-excitation after discussing the risks, especially of heart block associated with ablation of anteroseptal or MS APs, and benefits of the procedure. 439,440,473—476	1	С

1611		
Performance of an EPS to risk stratify individuals with asymptomatic pre-excitation should be considered. 439,450–452,454–460	lla	В
Non-invasive evaluation of the conducting properties of the AP in individuals with asymptomatic pre-excitation may be considered. 459,461–463,465–469	Шь	В
Invasive risk stratification with an EPS is recommended in patients without 'low-risk' characteristics at non-invasive risk stratification. 462,463,465–469,477	1	С
Clinical follow-up should be considered in a patient with asymptomatic pre-excitation and a low-risk AP at invasive risk stratification. 450,452,456,463,477	lla	c
Catheter ablation may be considered in a patient with asymptomatic pre-excitation, and a low-risk AP at invasive or non-invasive risk stratification. 405,450,452,456,463,477	Шь	с
Catheter ablation should be considered in patients with asymptomatic pre-excitation and LV dysfunction due to electrical dyssynchrony. 478–481	lla	С
Catheter ablation may be considered in patients with low-risk asymptomatic pre-excitation in appropriately experienced centres according to patient preferences. 203,439,450,453,454,471,474,482	Шь	c



# Criteria of succesful ablation sites



Stable catheter position, as confirmed fluoroscopically and by observing a stable electrogram (<10% change in amplitude in atrial and ventricular electrograms over 5-10 beats).

Atrial electrogram amplitude > 0.4 mV, or A/V ratio > 0. Both atrial and ventricular electrogram components should be recorded from the ablation (tip) electrode. When ablating from the atrial aspect of the annulus, the atrial electrogram is usually equal to or larger than the ventricular electrogram. Sometimes, the )o can merge and it may be difficult to determine whether both components are present. Rapid atrial or ventricular pacing resulting in block in the BT can help eliminate ventricular or atrial electrogram (respectively) so that the exact morphology of the other component can be visualized.

Local AV interval on the ablation catheter is usually short (25-50 msec, except for previously damaged, slowly conducting, oblique, or epicardial BTs).<sup>51</sup>

The local ventricular electrogram on the ablation catheter should precede the onset of the delta wave on the ECG by a mean of 0-10 msec for left-sided BTs and 10-30 msec for right-sided BTs (the local ventricular electrogram is measured from the peak of the bipolar electrogram or the maximal dV/dT in the unipolar electrogram).

QS (or, less preferably, rS) morphology of the unipolar electrogram. Right-sided BTs usually have unipolar recordings that show more pronounced (rapid and deeper) QS configuration than left-sided BTs. 46,51

Continuous electrical activity (defined as isoelectric interval of <5 msec be)een ventricular and atrial electrograms).

Presence of BT potential.

during anterograde activation mapping

Local VA interval during retrograde activation of the BT is short (25-50 msec, except for previously damaged, slowly-conducting, oblique, or epicardial BTs), usually resulting in inscription of the atrial electrogram on the ascending portion of the terminal ventricular electrogram. The "pseudodisappearance" of the atrial electrogram within the terminal portion of the ventricular electrogram (forming a W sign) during orthodromic AVRT is a manifestation of an extremely short local VA interval, which correlates with successful ablation sites.

Surface QRS to local atrial electrogram interval ≤ 70 msec (during orthodromic AVRT).

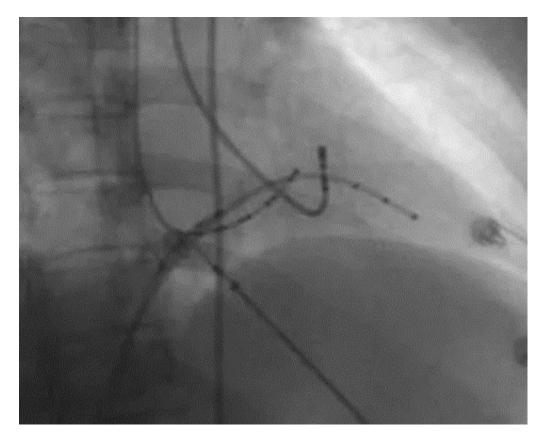
The local VA interval remains constant regardless of which direction the ventricular wavefront engaging the BT is traveling (i.e., despite pacing from different ventricular sites). If one uses the ventricular approach to ablate a concealed BT, the ventricular insertion site can be identified as one that maintains a constant local VA interval, despite differences in direction of activation to the ventricular site.

and additional criteriaduring retrograde activation mapping

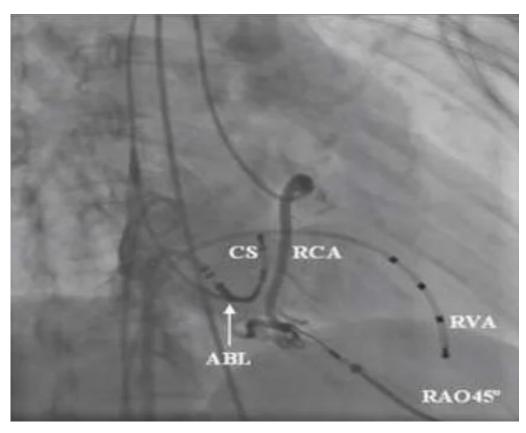


#### **NVRT:** catheter ablation





Retrograde ablation of left side AP, RAO



Ablation of right side AP



Clinical electrophysiology and ablation



#### Pediatric catheter ablation at the beginning of the 21st century: results from the European Multicenter Pediatric Catheter Ablation Registry 'EUROPA'

Ulrich Krause<sup>1\*</sup>, Thomas Paul<sup>1</sup>, Paolo Della Bella<sup>2</sup>, Simone Gulletta<sup>2</sup>, Roman A. Gebauer<sup>3</sup>, Christian Paech<sup>3</sup>, Peter Kubus<sup>4</sup>, Jan Janousek<sup>4</sup>, Paola Ferrari<sup>5</sup>, and Paolo De Filippo<sup>5</sup>

#### Aims

Contemporary data from prospective multicentre registries on catheter ablation in pediatric patients are sparse. Aim of the European Pediatric Catheter Ablation Registry EUROPA was to contribute data to fill this gap of knowledge.

#### Methods and results

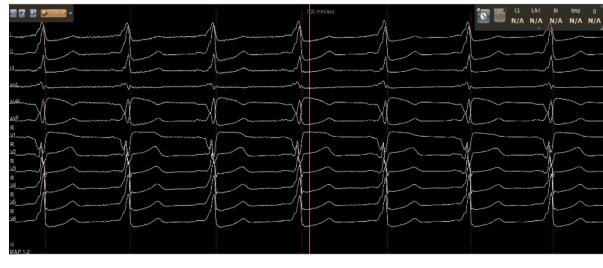
From July 2012 to June 2017, data on catheter ablation in pediatric patients ( $\leq$ 18 years of age) including a 1-year follow-up from five European pediatric EP centres were collected prospectively. A total of 683 patients (mean age 12.4 $\pm$ 3.9 years, mean body weight 50.2 $\pm$ 19 kg) were enrolled. Target tachycardia was WPW/atrioventricular-nodal re-entrant tachycardia (AVRT) in 380 (55.7%) patients, AVNRT in 230 (33.8%) patients, ventricular tachycardia (VT) in 24 (3.5) patients, focal atrial tachycardia (FAT) in 20 (2.9%) patients, IART in 14 (2%) patients, and junctional ectopic tachycardia in 3 (0.45) patients. Overall procedural success was 95.6%. Compared with all other substrates, success was significantly lower in FAT patients (80%, n = 16, P = 0.001). Mean procedure duration was 136 $\pm$ 67 min and mean fluoroscopy time was 4.9 $\pm$ 6.8 min. Major complications occurred in 0.7% of the patients. No persisting AV block requiring permanent pacing was reported. At 1-year follow-up (605/683 patients, 95%), tachycardia recurrence was reported in 7.8% of patients. Recurrence after VT ablation (33%) was significantly higher (P = 0.001) than after ablation of all other substrates.

#### Conclusion

The present study proves overall high efficacy and safety of catheter ablation of various tachycardia substrates in pediatric patients. Of note, complication rate was exceptionally low. Long-term success was high except for patients after VT ablation.



### Electrophysiology study of atrioventricular arrhythmias







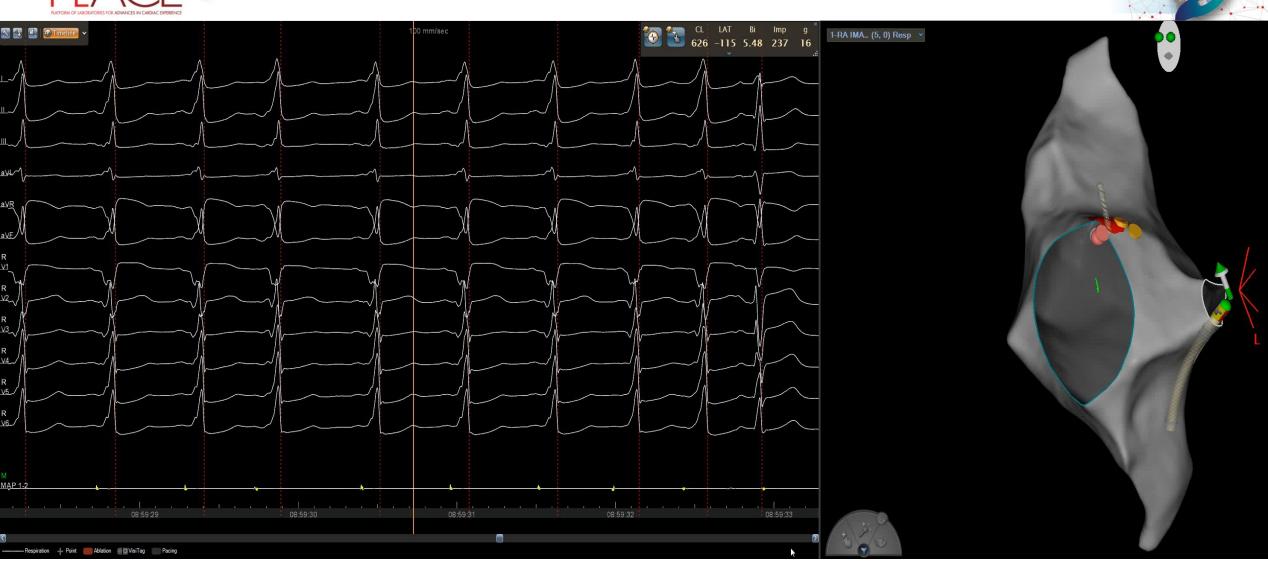


## **Baseline** ecg

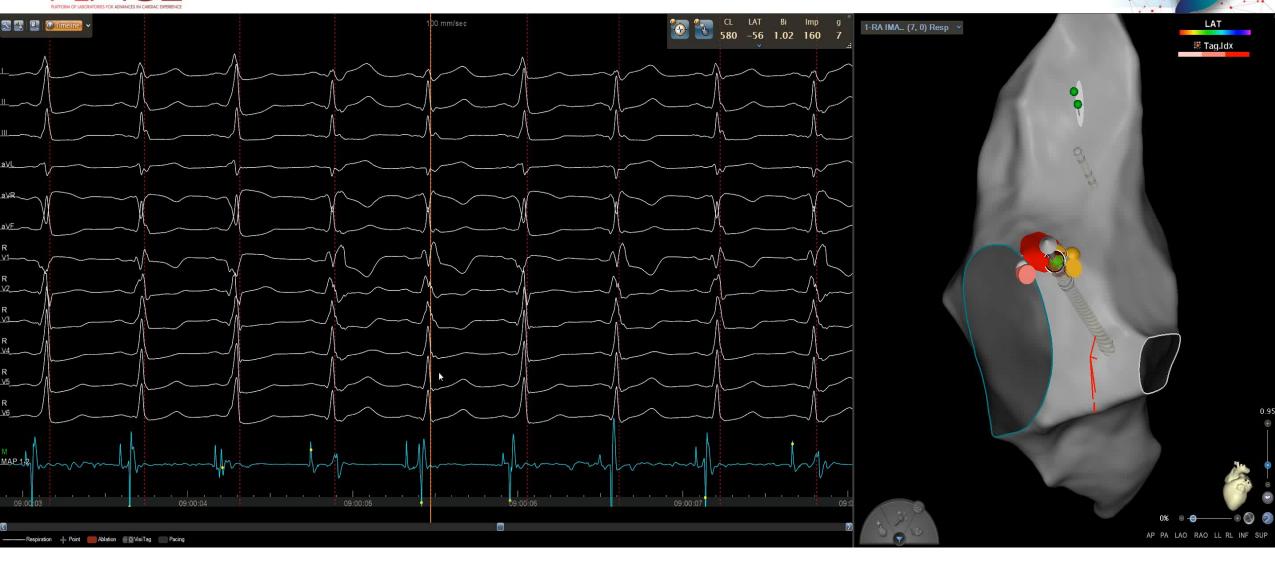






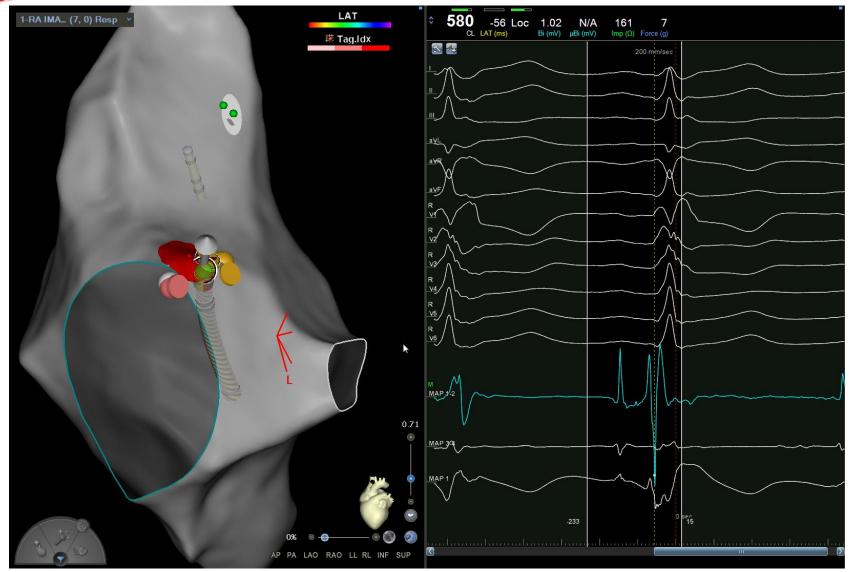






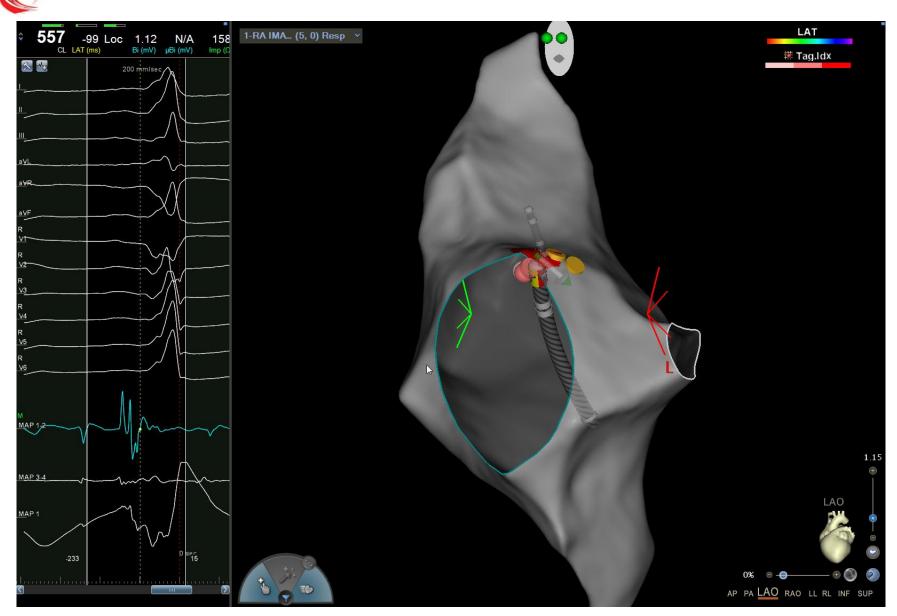
PLACE

9ª Edizione



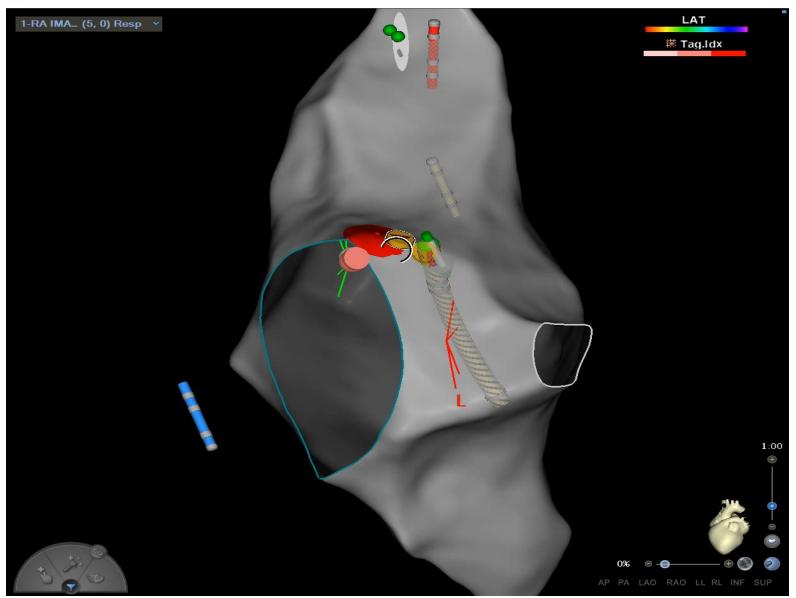
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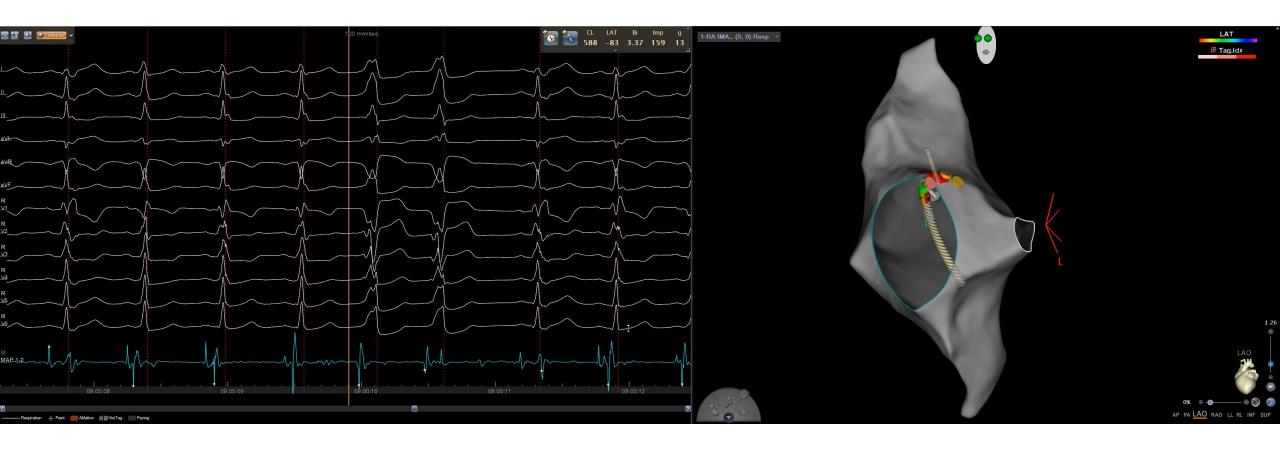
















# Insertable Cardiac Monitors in children





# 2021 PACES Expert Consensus Statement on the Indications and Management of Cardiovascular Implantable Electronic Devices in Pediatric Patients (9)

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).

#### Recommendations

COR	Insertable Cardiac Monitors			
I	Noninvasive cardiac rhythm monitoring is indicated in all patients prior to placement of an ICM.			
I	ICM is indicated in syncopal patients with high-risk criteria when comprehensive evaluation does not define a cause of syncope or lead to a specific treatment, and who do not have conventional indications for a pacemaker or ICD.	B-NR		
IIa	ICM is reasonable in the evaluation of patients with recurrent syncope of uncertain origin but not a high risk of SCD.	B-NR		
IIa	ICM is reasonable in patients with infrequent symptoms (>30-day intervals) suspected to be due to an arrhythmia, when the initial noninvasive evaluation is nondiagnostic.	C-LD		
IIa	ICM implantation is reasonable for guiding the management of patients with cardiac channelopathies or structural heart diseases associated with significant rhythm abnormalities.	C-LD		
IIb	ICM may be considered in patients with suspected reflex syncope presenting with frequent or severe syncopal episodes.	C-LD		
IIb	ICM may be considered in carefully selected patients with suspected epilepsy in whom anticonvulsive treatment has proven ineffective.	C-LD		
IIb	ICM may be considered in patients with severe but infrequent palpitations when other monitoring methods have failed to document an underlying cause.	C-LD		
IIb	ICM implantation may be considered for detecting subclinical arrhythmias in patients with cardiac channelopathies or other diseases associated with significant rhythm abnormalities.	C-EO		







### Recommendation-specific supportive text

Several observational studies have demonstrated a benefit of ICM in establishing a diagnosis for recurrent symptoms of unclear etiology when other monitoring methods have failed to document an underlying cause.

Syncope: Cardiac or undefined syncope may be present in up to 8% of syncopal events in children and adolescents. In adults, monitoring with an ICM has been shown to be more cost-effective for establishing a diagnosis than other methods of rhythm monitoring and should be the method of choice when arrhythmogenic syncope is suspected but not proven. 223,226

Palpitations: ICM implantation should be considered on an individual basis, taking into account each patient's underlying cardiac condition, the severity of symptoms, and age-and development-related monitoring limitations. 233,234

Bradyarrhythmias: ICM may be useful in the monitoring of bradyarrhythmias and their correlation with clinical symptoms. ICM may also be useful for patients at risk for intermittent or progressive AV block including patients with neuromuscular diseases, progressive cardiac conduction diseases, and Kearns-Sayre syndrome. 91,98

Other conditions: ICM may be useful for occult arrhythmia detection in asymptomatic children with potentially lethal cardiac diseases (e.g., inherited primary arrhythmia syndromes, cardiomyopathies) and may identify events that warrant changes in patient management. 226,230-232 Furthermore, monitoring with an ICM may provide psychological reassurance for parents of children at risk for malignant arrhythmias. 233





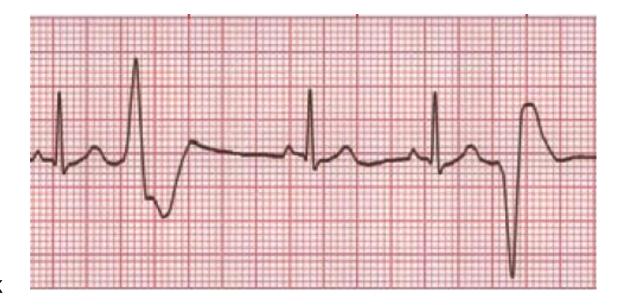
## Ventricular arrhythmias





## Premature Ventricular Complexes (PVCs)

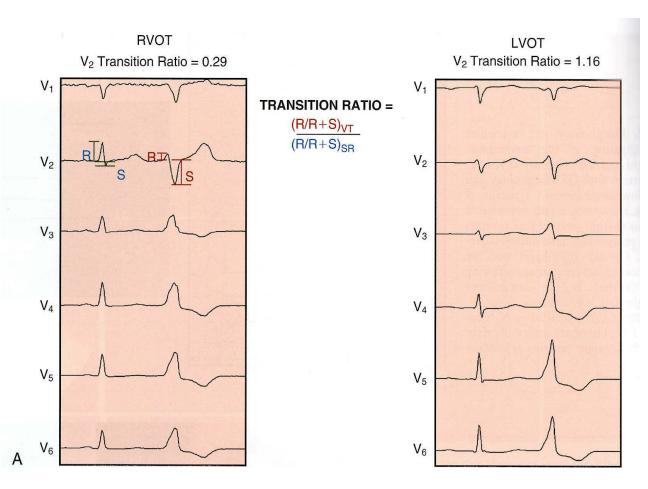
- PVCs are a common finding in pediatric patients of all ages and may be seen on a Holter monitor in about 40 percent of healthy children. When PVCs are rare, isolated and monomorphic, they usually do not require extensive evaluation in a healthy child and have an excellent prognosis.
- However, when the PVC burden is more frequent or more complex it necessitates further work-up and possibly longitudinal follow-up.
- With frequent ectopy (generally defined as ≥ 10 percent of beats in a 24-hour period), there is a risk of developing ventricular dysfunction even in a normal heart: in most studies, 20 to 30 percent ectopy is needed to increase the risk of ventricular dysfunction over time.

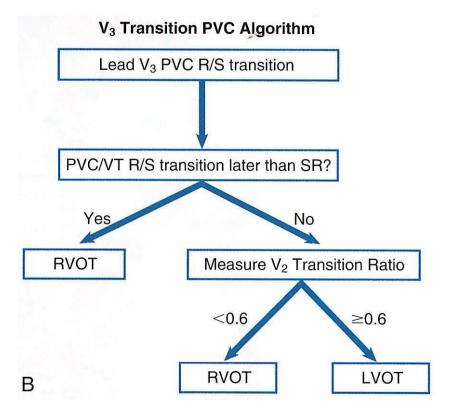




# Ventricular arrhythmias in the absence of structural heart Outflow tract ventrices tachycardia









# Ventricular arrhythmias in the absence of structural heart Outflow tract ventrice are tachycardia

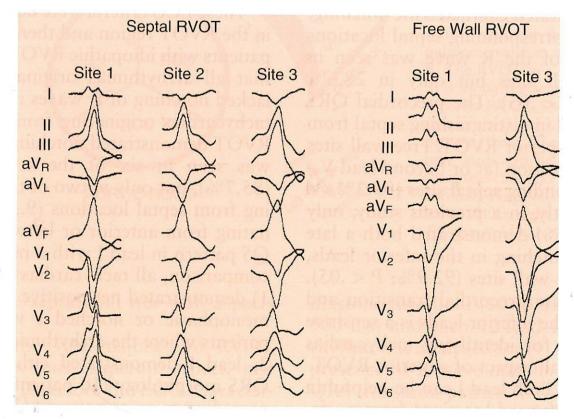
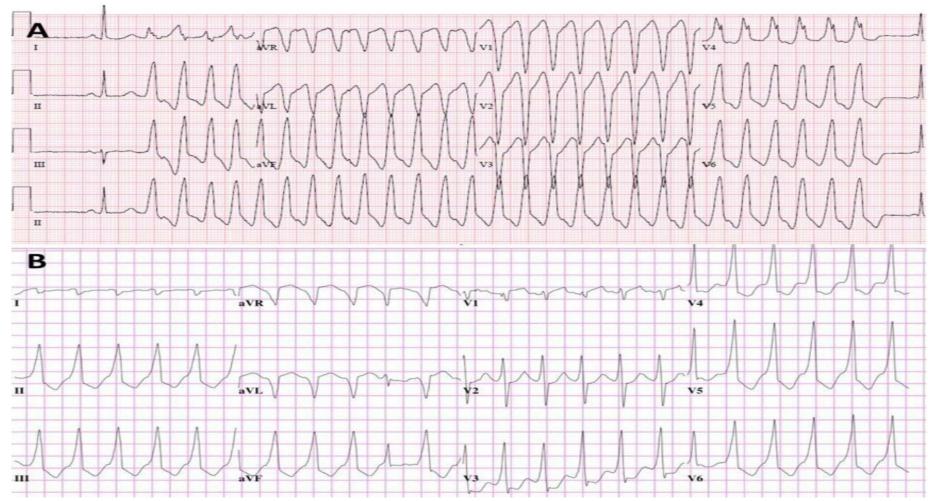


Figure 28-6. Electrocardiogram (ECG) morphologies of tachycardias that were successfully ablated in the superior right ventricular outflow tract (RVOT). The left-hand side of the figure displays the 12-lead ECG morphology of characteristic septal RVOT tachycardias from sites 1 through 3, whereas the right-hand side of the figure displays the ECG morphology of characteristic free-wall RVOT tachycardias from sites 1 and 3. Tachycardias originating from free wall show notching in the inferior leads and late precordial transition (≥V₄). In comparison, tachycardias originating from septal RVOT have earlier precordial transition and lack notching of the R wave in the inferior leads. For both septal and free-wall tachycardias, lead I helps distinguish anterior (leftward) from posterior (rightward) location. See text for details.



# Ventricular arrhythmias in the absence of structural heart Outflow tract ventrices tachycardia







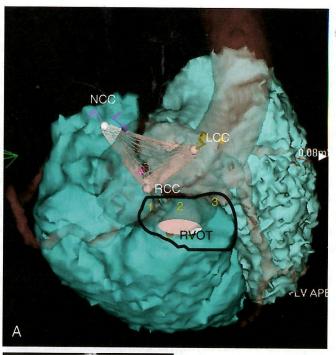


# Ventricular arrhythmias in the absence of structural heart Outflow tract ventrices tachycardia

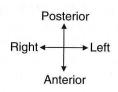


TABLE 28-1 Diagnostic Criteria				
RVOT source  • Septal  • Free wall  • Anterior sites  • Posterior sites	<ul> <li>LBBB pattern; R-wave transition after V<sub>3</sub>; tall R in leads 2, 3, aV<sub>F</sub></li> <li>Absence of notching in leads 2, 3, aV<sub>F</sub>; precordial transition &lt; V<sub>4</sub></li> <li>Presence of notching in leads 2, 3, aV<sub>F</sub>; precordial transition ≥ V<sub>4</sub></li> <li>Negative lead 1</li> <li>Positive lead 1</li> </ul>			
Coronary cusps  • Left coronary cusp  • Right coronary cusp  • Left-right junction	<ul> <li>Strongly inferior axis; precordial transition earlier than in NSR</li> <li>m or w pattern in V<sub>1</sub></li> <li>QS or rS pattern in V<sub>1</sub></li> <li>QS pattern, with notching on downstroke, in V<sub>1</sub></li> </ul>			
Aortomitral continuity	RBBB, inferior axis, qR pattern in V <sub>1</sub>			
LV summit	Inferior axis, delayed upstroke in lead 2, QS in lead 1, R/S ratio in $V_2 < 1$			

LBBB, Left bundle branch block pattern; LV, left ventricle; NSR, normal sinus rhythm; RBBB, right bundle branch block pattern; RVOT, right ventricular outflow tract.

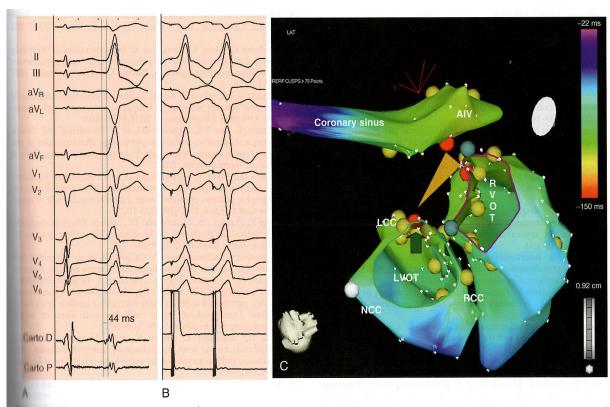








#### PVCs from the mitral anulus: identification and electrophysiological aspects



**28-18.** A, Local activation from the left coronary cusp (LCC; pre-QRS by 44 ms) during the clinical arrhythmia (frequent monomorphic with left bundle branch block morphology, inferior axis, and early precordial transition by lead V<sub>3</sub>). B, Pace map from this location, which excellent match of the PVC. C, Electroanatomic maps of the right ventricular outflow tract (RVOT), left ventricular outflow tract (LVOT), the coronary sinus/anterior interventricular vein (AIV) that were acquired during the PVCs. Relative to the RVOT, coronary sinus, AIV, right cusp (RCC), and noncoronary cusp (NCC), the LCC demonstrates the earliest activation (green arrow). Arrhythmias arising from the motive the anterior septal superior RVOT, LCC, and distal coronary sinus-AIV junction (triangle between three red dots) can manifest activation/best pace maps from one or more surrounding locations and so can be frequently challenging to localize accurately.

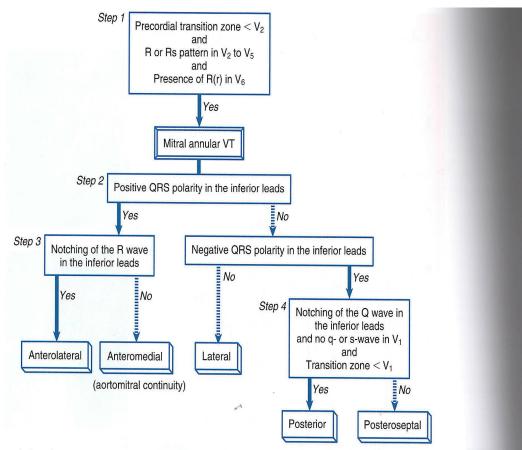


Figure 29-3. Proposed algorithm to predict the precise focus of a ventricular tachycardia/premature ventricular contractions originating final mitral annulus based on the QRS wave configuration in 12-lead electrocardiogram recordings. VT, Ventricular tachycardia. (From Tada II. Naito S, et al. Idiopathic ventricular arrhythmia arising from the mitral annulus: a distinct subgroup of idiopathic ventricular arrhythmia arising from the mitral annulus: a distinct subgroup of idiopathic ventricular arrhythmia. Coll Cardiol. 2005;45:877-886. With permission)



### Ventricular arrhythmias in the absence of structural heart Outflow tract ventrices tachycardia

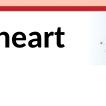
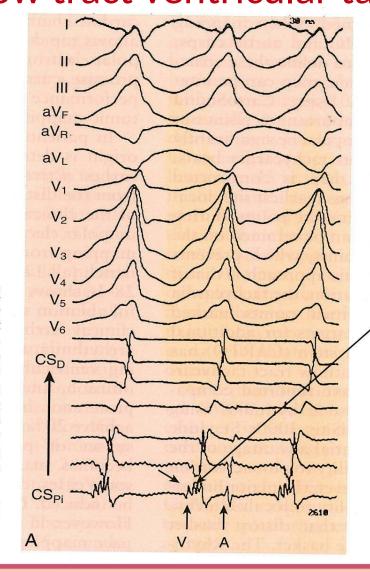
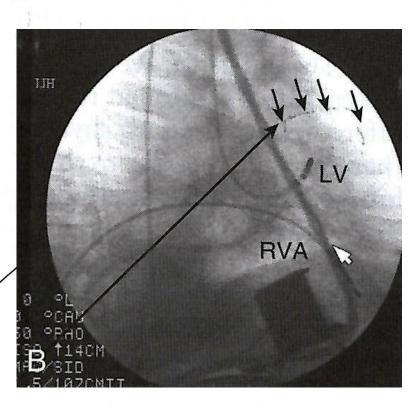


Figure 28-15. Twelve-lead electrocardiogram and intracardiac electrograms from a patient with ventricular tachycardia of a left ventricle (LV) summit origin (left). Intracardiac recordings of the epicardial surface are achieved using a 2-F multielectrode mapping catheter advanced through the coronary sinus (CS) into the anterior interventricular vein (right, black arrows). The earliest activation during tachycardia is recorded at the proximal electrode positioned in the proximal anterior interventricular vein (left, arrow). This site also resulted in excellent pace map. The fluoroscopic image on the right was obtained in an right anterior oblique projection. Catheters at the right ventricular apex (RVA) and base of the LV endocardium under the aortic valve are also seen. A, Atrium; D, distal; Pi, proximal; V, ventricle. (Courtesy Dr. Enrique Rodriguez.)

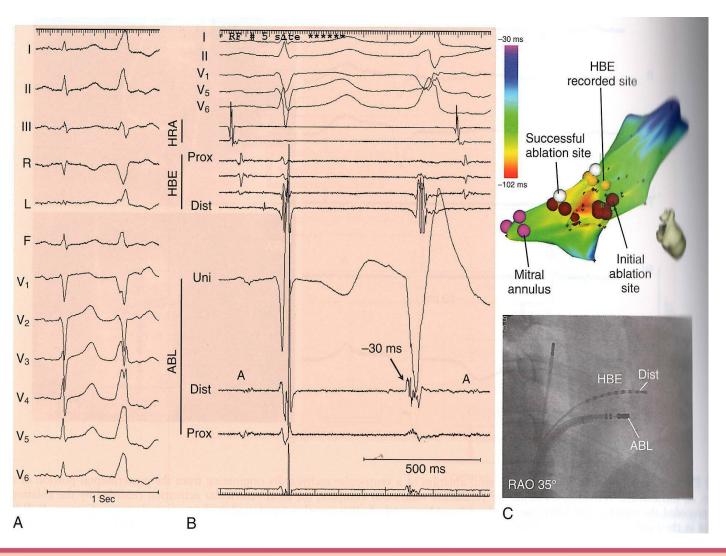


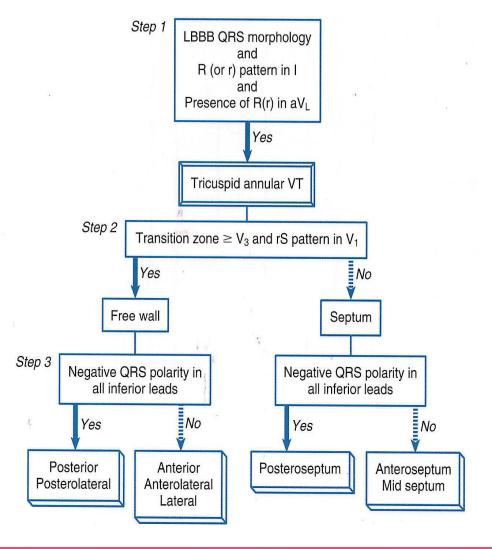




# PVCs from the tricuspid anulus: identification and electrophysiological aspects



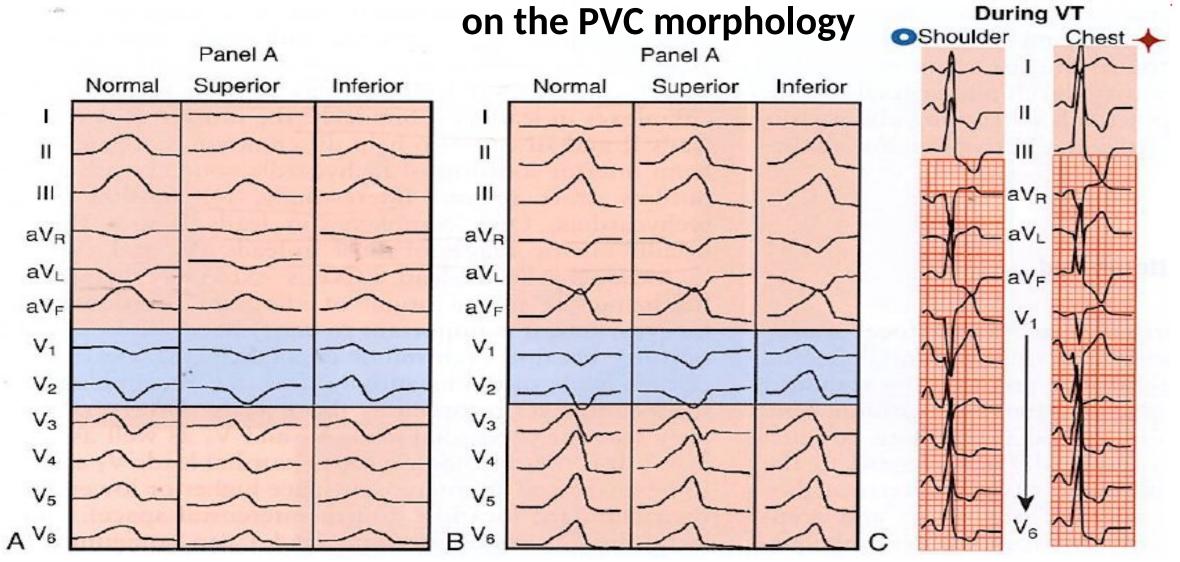


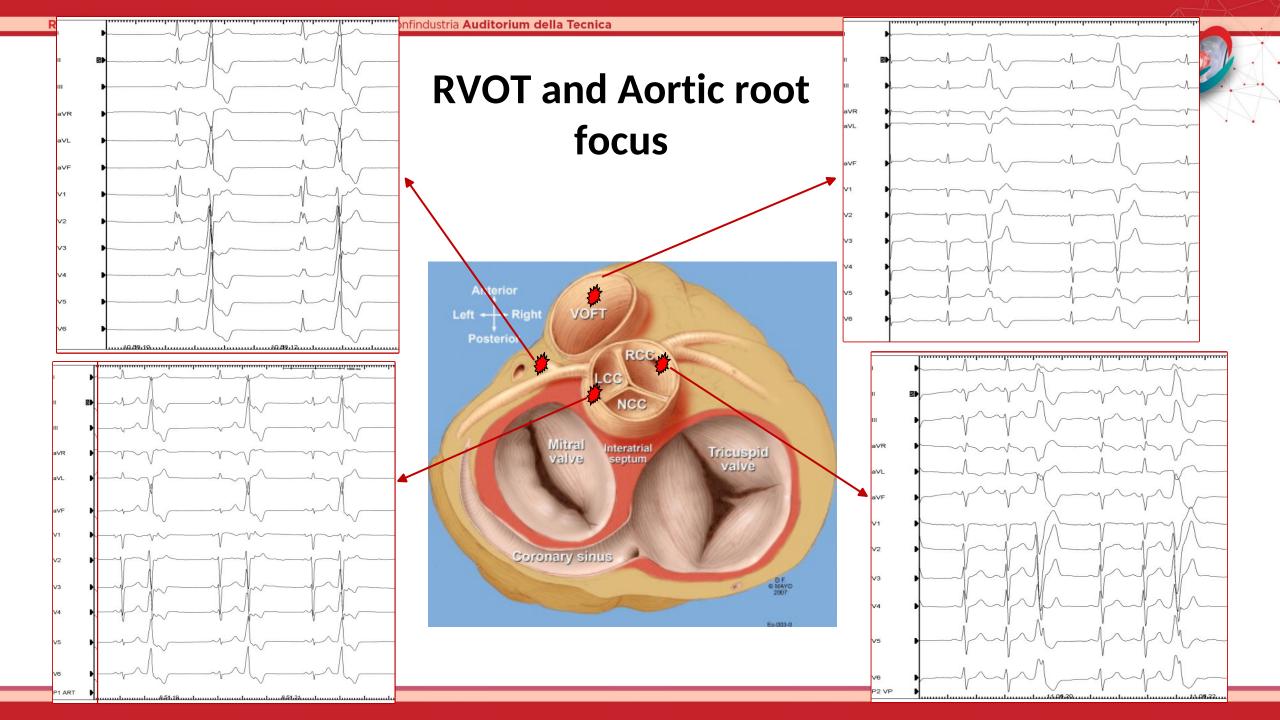


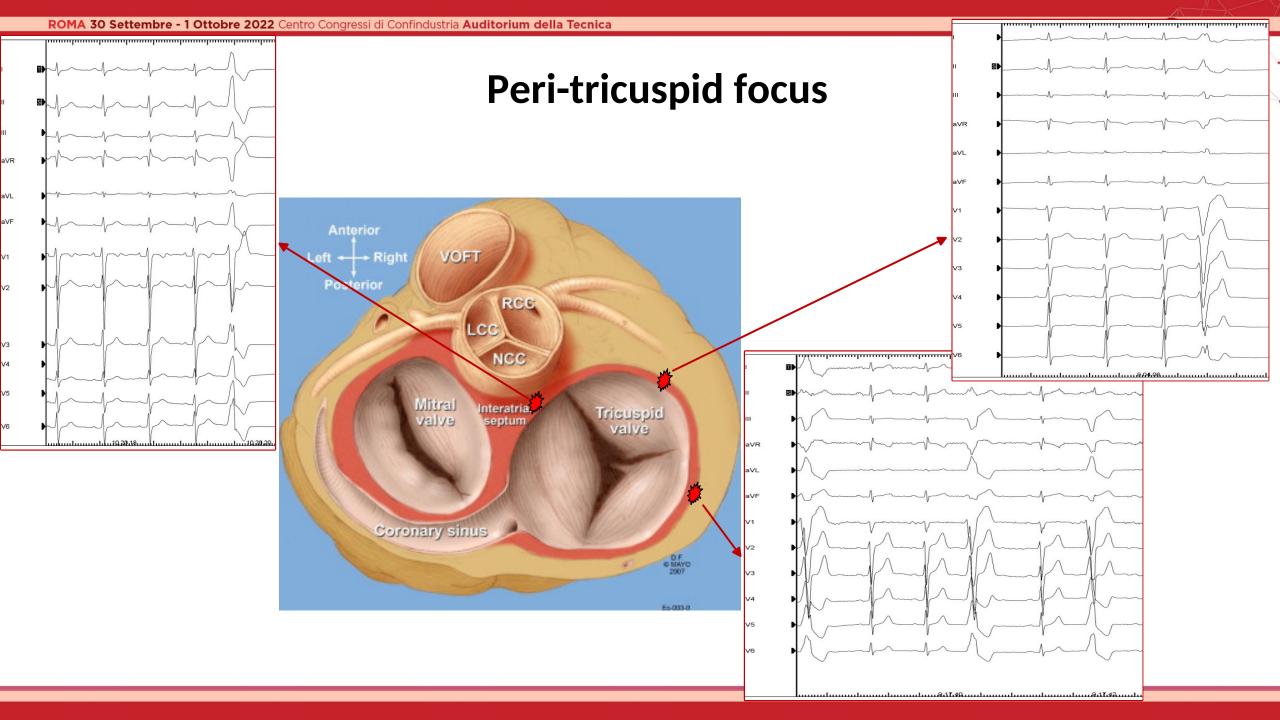


# ROMA 30 Settembre - 1 Ottobre 2022 Centro Congressi di Confindustria Auditorium della Tecnica Normalia 9º Edizione The effect of vertical displacement of V<sub>1</sub> and













### Ventricular arrhythmias (Vas)

- Ventricular tachycardia (VT) is rare in children representing only 1.8% of children undergoing EPS
- Ventricular tachycardia is well described in infancy, and although symptoms are less common compared to older children (22 vs. 34%), and may be incessant leading to ventricular dysfunction
- Few data are available regarding the invasive VA treatment with catheter ablation





### Most common Vas in children

- Ventricular outflow tract ventricular tachycardia (RVOT, LVOT)
  - Fascicular ventricular tachycardia
- Ventricular tachycardia in the setting of inherited channelopathies (QT long, Brugada ...)

#### **CONSENSUS STATEMENT**



# 2019 HRS/EHRA/APHRS/LAHRS expert consensus statement on catheter ablation of ventricular arrhythmias

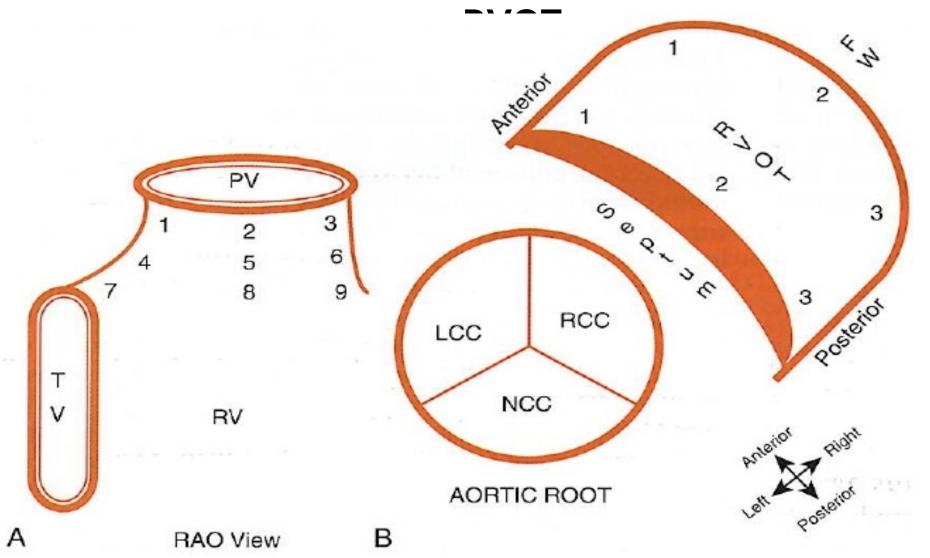
Reco	mmendations	for catheter	ablation of bu	ndle branch	reentrant VT	and for	catheter	ablation of fa	ascicular <b>V</b>	<b>/T</b>

COR	LOE	Recommendations	References
1	B-NR	In patients with bundle branch reentrant VT, catheter ablation is useful for reducing the risk of recurrent VT.	S4.6.1–S4.6.9
1	B-NR	<ol> <li>In patients with idiopathic left fascicular reentrant VT for whom medications are ineffective, not tolerated, or not the patient's preference, catheter ablation is useful.</li> </ol>	S4.6.10–S4.6.22
1	B-NR	3. In larger pediatric patients (≥15 kg) with idiopathic left fascicular reentrant VT in whom medical treatment is ineffective or not tolerated, catheter ablation is useful.	S4.6.23-S4.6.26
1	B-NR	<ol> <li>In patients with focal fascicular VT with or without SHD, catheter ablation is useful.</li> </ol>	S4.6.11,S4.6.27-S4.6.29
1	B-NR	5. In patients with postinfarction reentrant Purkinje fiber-mediated VT, catheter ablation is useful.	S4.6.30–S4.6.32



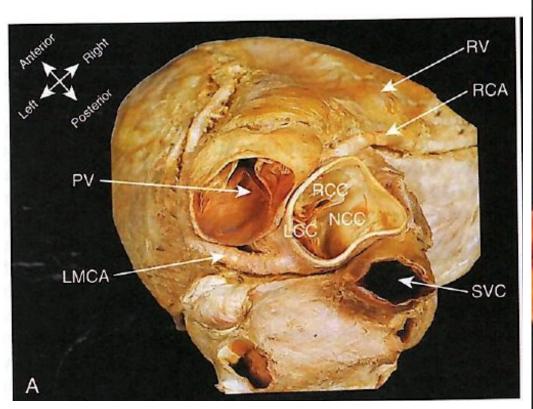


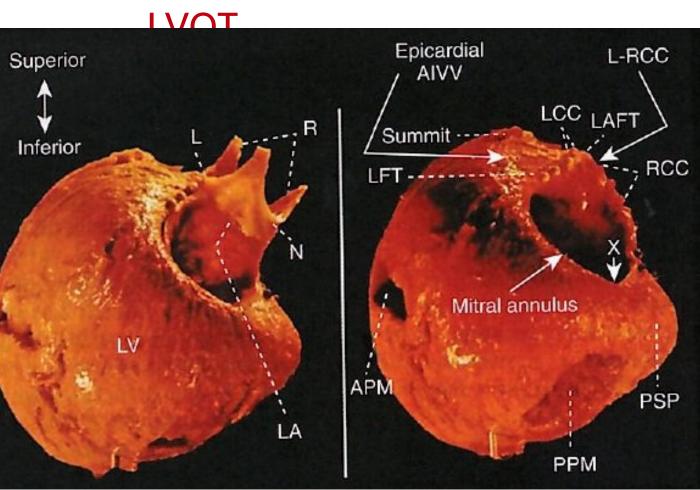




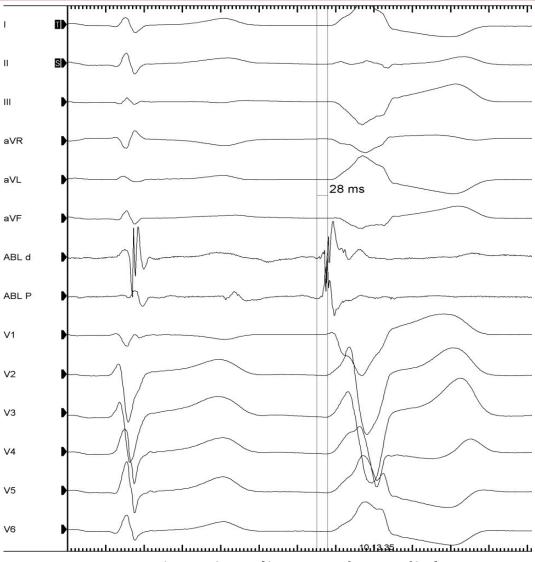


# Ventricular arrhythmias in the absence of structural heart Anatomical relationship between RVOT and

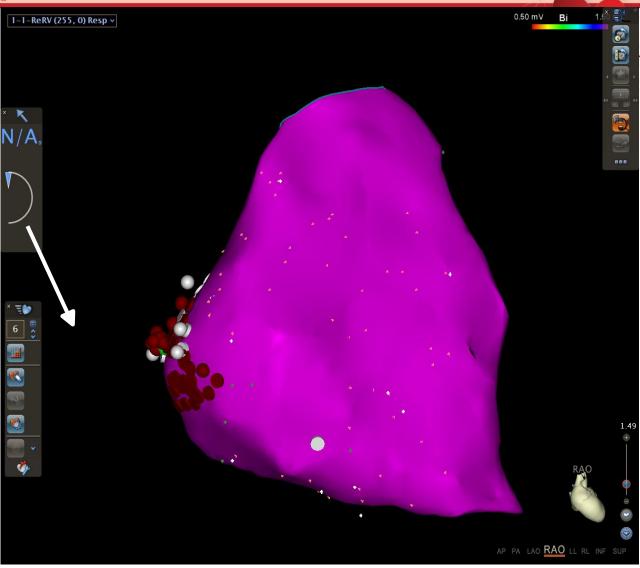




Basal condition ECG: LBB-upper axis extrasystole; likely peritricuspid focus. There are no ECG signs of ARVD in RS complexes.



Site of earliest endocardial activation (RV basal lateral inferior free wall)



RV bipolar voltage map



#### **Final result**





ORIGINAL ARTICLES WILEY

#### Etiology is a predictor of recurrence after catheter ablation of ventricular arrhythmias in pediatric patients



#### Abstract

Background: Ventricular arrhythmias (VAs) are rare in pediatric patients, especially in absence of structural heart disease (SHD). Few data are available regarding the invasive VAs treatment with catheter ablation (CA) in pediatric patients and predictors of outcomes have not been fully investigated.

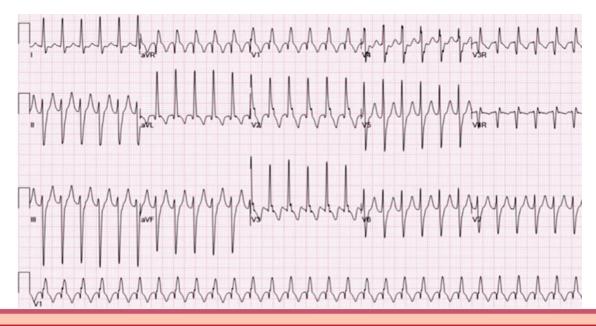
Objective: To describe the clinical presentation, procedural characteristics, and outcomes in pediatric patients undergoing CA for VAs.

**Methods:** Eighty-one consecutive pediatric patients (58 male [72%], 15.5 ± 2.2 years) treated by CA for ventricular tachycardia (VT) or premature ventricular beats (PVBs) were retrospectively evaluated. Study endpoints were VAs recurrence and mortality for any cause.

**Results:** Ninety-five procedures were performed in 81 patients, 52 (55%) PVBs and 43 (45%) VT ablations. During a follow-up of 35.0 months (interquartile range = 13.0-71.0), 14 patients (14.7%) had a VA recurrence: 11 (33.3%) patients treated with CA for VT and 3 (6.2%) patients treated for PVBs (p < .001). One patient (1%) died 26 months after the procedure during an electrical storm. Patients with SHD had higher VAs recurrence rate, as compared with idiopathic VAs (pairwise log-rank p < .001). Patients treated with CA for VT had higher VA recurrence rate, as compared with PVB patients (pairwise log-rank p = .002). At Cox multivariate analysis only SHD was an independent predictor of VAs recurrence (hazard ratio = 5.56, 95% confidence interval = 2.68-11.54, p < .001).

Conclusion: CA of VAs is effective and safe in a pediatric population. CA of idiopathic and fascicular VAs are associated with lower recurrence rate, than VAs in the setting of SHD.









#### patients Study type

- Single tertiary referral center (San Raffaele Hospital, Milan, Italy)
- Experimental Prospective study
- Enrollement period: July 2021 January 2021

#### Aim of the study

Describe the clinical presentation, procedural characteristics and outcomes in pediatric patients undergoing CA for VAs.











#### **STUDY POPULATION:**

Patients undergoing catheter ablation for Ventricular Tachycardia (VT) and Premature Ventricular Beats (PVBs) between July 2010 and January 2019 were enrolled and prospectively followed-up.

#### **INCLUSION CRITERIA:**

- Age less than 18 years at diagnosis
- ECG documentation of at least 1 VA episode
- At least 1 catheter ablation of VA in our Hospital





**Results: Population** 

#### **STUDY POPULATION:**

Ninety-five procedures were performed in **81 patients**, 52 (55%) PVBs and 43 (45%) VT ablations. During a follow-up of 35.0 months (IQR: 13.0-71.0), 14 patients (14.7%) had a VA recurrence: 11 (33.3%) patients treated with CA for VT and 3 (6.2%) patient treated for PVBs (p<0.001).

One patient (1%) died 26 months after the procedure during an electrical storm.





### Results: Patients' baseline characteristics

	Idiopathic (N=55)	Fascicular (N=12)	SHD (N=14)	Total (N=81)	p value						
<b>Age (years)</b> Mean (SD)	15.3 (2.3)	15.9 (1.7)	16.0 (2.3)	15.5 (2.2)	0.42						
<b>Gender (M)</b> n (%)	40 (72.7%)	7 (58.3%)	11 (78.6%)	58 (71.6%)	0.49	LVEDV (ml) Mean	60.8 (13.2)	64.2 (8.8)	67.7 (21.1)	62.5 (14.4)	0
SCD family history	0 (0.0%)	0 (0.0%)	2 (14.3%)	2 (2.5%)	0.007	(SD)	27515.5	29200.0	32201.0	28558.0	0
Age at presentation	12.9 (3.3)	13.2 (3.2)	13.1 (2.8)	13.0 (3.1)	0.93	PVBs number/24h Mean (SD)	27313.3	29200.0	32201.0	20330.0	0
(years) Mean (SD)						Mean (3D)	(15963.9)	(17326.3)	(23617.3)	(17604.1)	
Presentation with:											
-Near syncope n (%)	7 (12.7%)	2 (16.7%)	4 (28.6%)	13 (16.0%)	0.35	Beta blockers n (%)	29 (52.7%)	6 (50.0%)	9 (64.3%)	44 (54.3%)	0.
-Palpitations n (%)	32 (58.2%)	12 (100.0%)	14(100.0%)	58 (71.6%)	< 0.001	Class I AADs n (%)	6 (10.9%)	3 (25.0%)	3 (21.4%)	12 (14.8%)	0.
-Heart failure n (%)	0 (0.0%)	1 (8.3%)	2 (14.3%)	3 (3.7%)	0.03	<b>CCBs</b> n (%)	0 (0.0%)	1 (8.3%)	0 (0.0%)	1 (1.2%)	0.
-ICD Shock n (%)	1 (1.8%)	0 (0.0%)	7 (50.0%)	8 (9.9%)	< 0.001	Amiodarone n (%)	5 (9.1%)	3 (25.0%)	4 (28.6%)	12 (14.8%)	0.
-Syncope n (%)	2 (3.6%)	4 (33.3%)	6 (42.9%)	12 (14.8%)	< 0.001			24			
ICD n (%)	1 (1.8%)	1 (8.3%)	8 (57.1%)	10 (12.3%)	< 0.001						
LVEF (%) Mean (SD)	58.4 (6.2)	59.8 (3.5)	54.3 (7.1)	57.9 (6.2)	0.06						





### Results: Procedural charateristics

	No			
	recurrence	<b>VA</b> recurrence	Total	р
	(N=81)	(N=14)	(N=95)	value
Induction:				w
-Ventricular stimulation n (%)	14 (17%)	7 (50%)	21 (22%)	<0.001
- Isoproterenol n (%)	12 (15%)	1 (7%)	13 (14%)	0.44
- Spontaneous n (%)	12 (15%)	2 (14%)	14 (15%)	0.95
PVBs ablation n (%)	49 (60%)	3 (21%)	52 (55%)	< 0.001
VT ablation n (%)	32 (40%)	11 (79%)	43 (45%)	< 0.001
Endocardial ablation n (%)	80 (99%)	13 (93%)	93 (98%)	0.15
<b>Epicardial ablation</b> n (%)	4 (5%)	4 (29%)	8 (8%)	< 0.001
Previous PES n (%)	18 (22%)	4 (29%)	22 (23%)	0.60
<b>VT induced</b> n (%)	23 (28%)	7 (50%)	30 (32%)	0.11

Strategy:				
-Late potential target n (%)	11 (14%)	4 (29%)	15 (16%)	0.15
-Pacemapping n (%)	2 (2%)	0 (0%)	2 (2%)	0.55
-Activation mapping n (%)	74 (91%)	10 (71%)	84 (88%)	0.03
RF time (s) Mean (SD)	419.8 (232.3)	538.5 (256.7)	437.3(238.4)	0.04
<b>Procedure time (min)</b> (Mean (SD)	176.5 (80.2)	200.7 (97.8)	180.0 (82.9)	0.42
<b>Left ventricular ablation</b> n(%)	41 (51%)	7 (50%)	48 (51%)	0.96
Right ventricular ablation n(%)	46 (57%)	7 (50%)	53 (56%)	0.63





Results: Clinical characteristics of pate at and without ventricular arrhythmias recurrence

	No	VA		
	recurrence (N=67)	recurrence (N=14)	Total (N=81)	p value
Age (years) Mean (SD)	15.4 (2.3)	16.0 (1.9)	15.5 (2.2)	0.39
<b>Gender (M)</b> n (%)	47 (70%)	11 (79%)	58 (72%)	0.52
SCD family history	0 (0%)	2 (14%)	2 (2%)	< 0.001
Age at presentation (years) Mean (SD)	13.1 (3.1)	12.1 (3.0)	12.9 (3.1)	0.19
SHD	14 (21%)	12 (86%)	26 (32%)	<0.001
Clinical presentation with:				
- Palpitations n (%)	45 (67%)	13 (93%)	58 (72%)	0.05
- Heart failure n (%)	1 (1%)	2 (14%)	3 (4%)	0.02
- <b>Lipothymia</b> n (%)	9 (13%)	4 (29%)	13 (16%)	0.16
- Syncope n (%)	5 (7%)	7 (50%)	12 (15%)	<0.001
- ICD Shock n (%)	2 (3%)	6 (43%)	8 (10%)	< 0.001
ICD n (%)	3 (4%)	7 (50%)	10 (12%)	<0.001
LVEF (%) Mean (SD)	58.5 (5.8)	54.9 (7.4)	57.9 (6.2)	0.06

LVEF (%) Mean (SD)	58.5 (5.8)	54.9 (7.4)	57.9 (6.2)	0.06
LVEDV (ml) Mean (SD)	61.3 (13.3)	68.2 (18.7)	62.5 (14.4)	0.20
PVBs number/24h Mean	28366.8	29336.3	28558.0	0.86
(SD)	(16745.0)	(21448.6)	(17604.1)	
Beta blockers n (%)	35 (52%)	9 (64%)	44 (54%)	0.41
Class I AADs n (%)	8 (12%)	4 (29%)	12 (15%)	0.11
CCBs n (%)	1 (1%)	0 (0%)	1 (1%)	0.64
Amiodarone n (%)	6 (9%)	6 (43%)	12 (15%)	<0.001

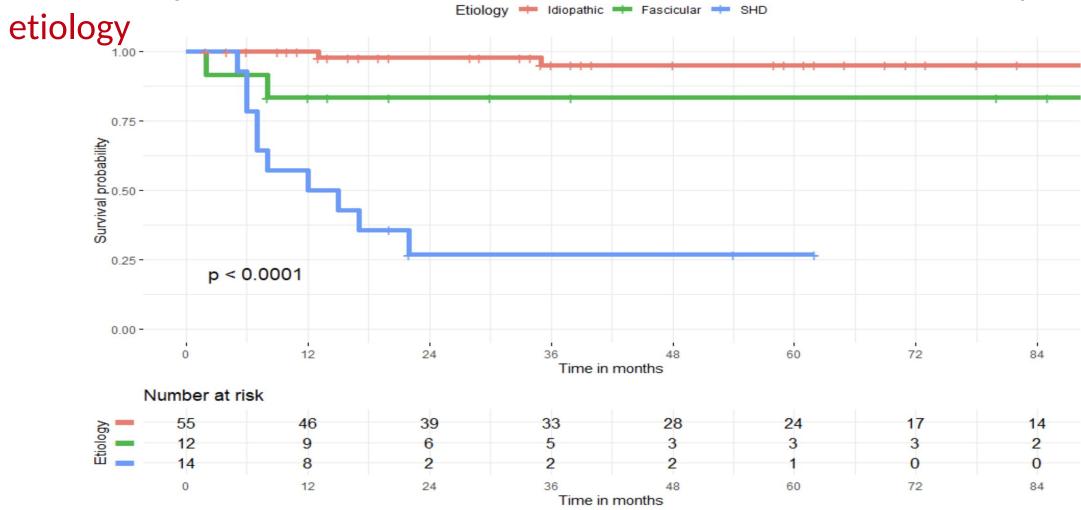


9ª Edizione

#### **Etiology as a predictor of recurrence after catheter** ablation of VAs in paediatric patients



Results: Kaplan-Meier curve of survival free from VAs recurrence by



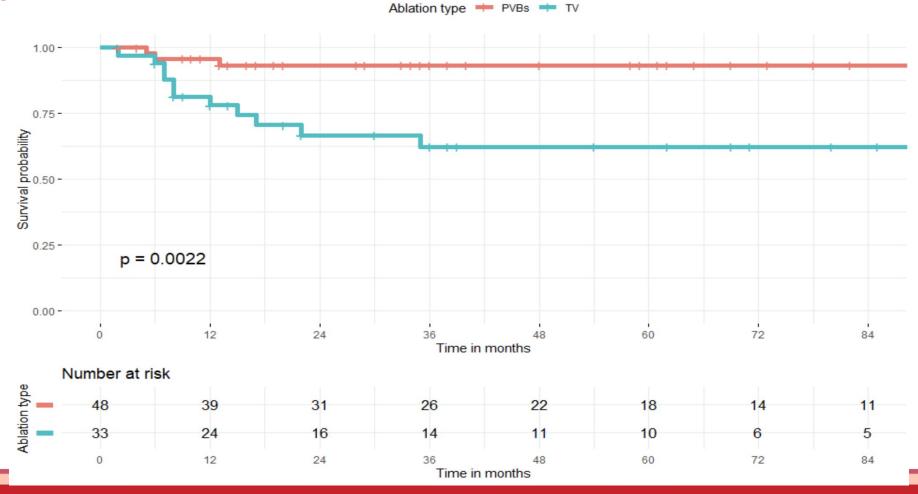


9ª Edizione

### Etiology as a predictor of recurrence after catheter ablation of VAs in paediatric patients



Results: Kaplan-Meier curve of survival free from VA recurrence in patients undergoing for premature ventricular beats and ventricular tachycardia ablation







Patients treated with **CA for VT had higher VA recurrence rate**, as compared with PVB patients (Pairwise Log-Rank p=0.002). At Cox multivariate analysis only SHD was an independent predictor of VAs recurrence (HR=5.56, CI 95% 2.68 -11.54, p<0.001).





### Main findings

- Catheter ablation in pediatric patients is an effective procedure, with low recurrence rate during the follow-up (mortality of 1% during fup)
- Patients treated with CA for VT had higher VA recurrence rate, as compared with PVB patients (Pairwise Log-Rank p=0.002)
- The SHD appeared as the main independent predictor of VA recurrence at the multivariate analysis (HR=5.56, CI 95% 2.68 -11.54, p<0.001)</li>

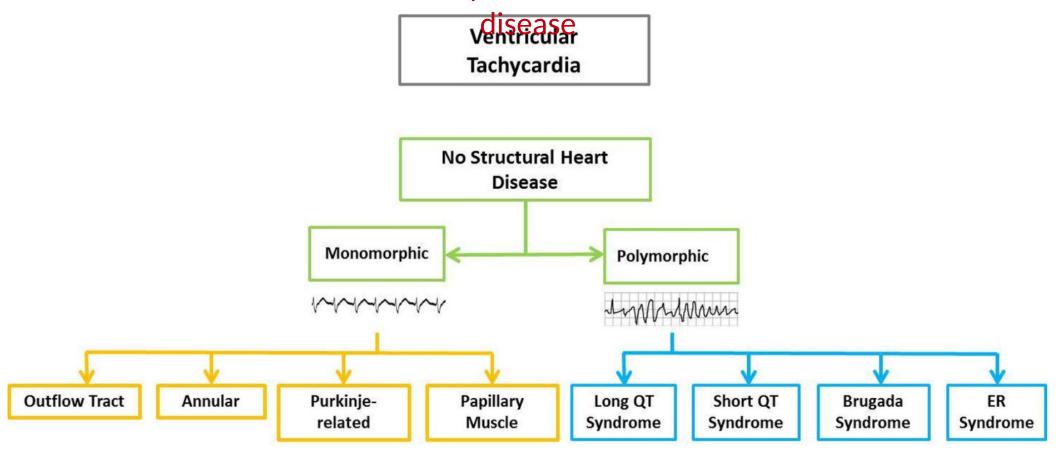




### Ventricular arrhythmias in the absence of structural heart disease



Classification of ventricular tachycardia in the absence of structural heart

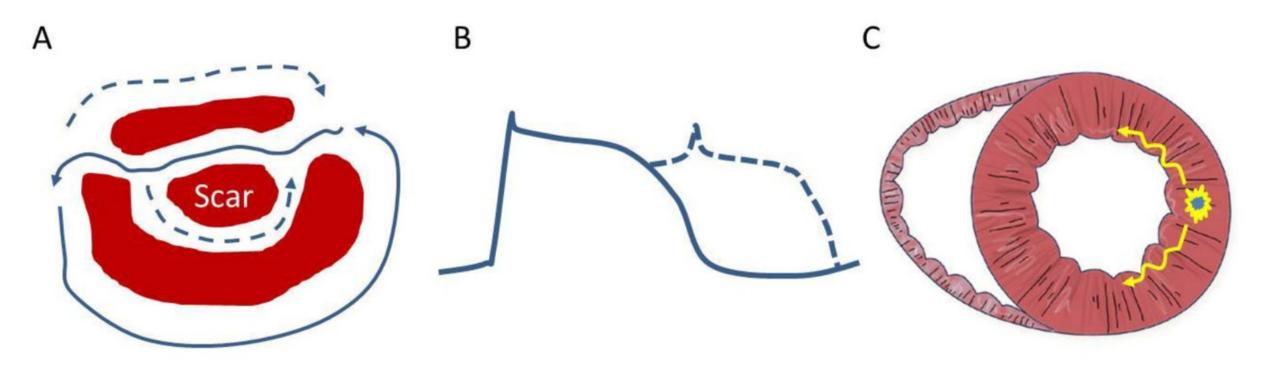




## Ventricular arrhythmias in the absence of structural heart disease



#### Mechanisms of ventricular arrhythmias

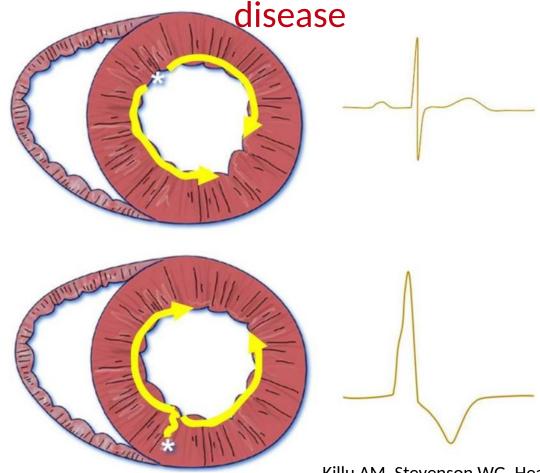




## Ventricular arrhythmias in the absence of structural heart disease



Classification of ventricular tachycardia in the absence of structural heart



AC, arrhythmogenic cardiomyopathy; EP, electrophysiology; RV, right ventricular; VT, ventricular tachycardia.



Isoproterenol required for initiation at EP study

### Ventricular arrhythmias in the absence of structural heart Outflow tract ventrices tachycardia



Table 1         Useful characteristics to assist differentiating idiopathic from scar-related monomorphic RV VT							
Characteristic	Idiopathic RV VT	Scar-related RV VT					
Any sinus rhythm ECG abnormality	Rare	++					
VT QRS morphology							
QRS transition after V4	Rare	++					
Notched down stroke in V1/2	Rare	++					
Delay in nadir of V1	Rare	++					
Multiple VT morphologies	Rare	++					

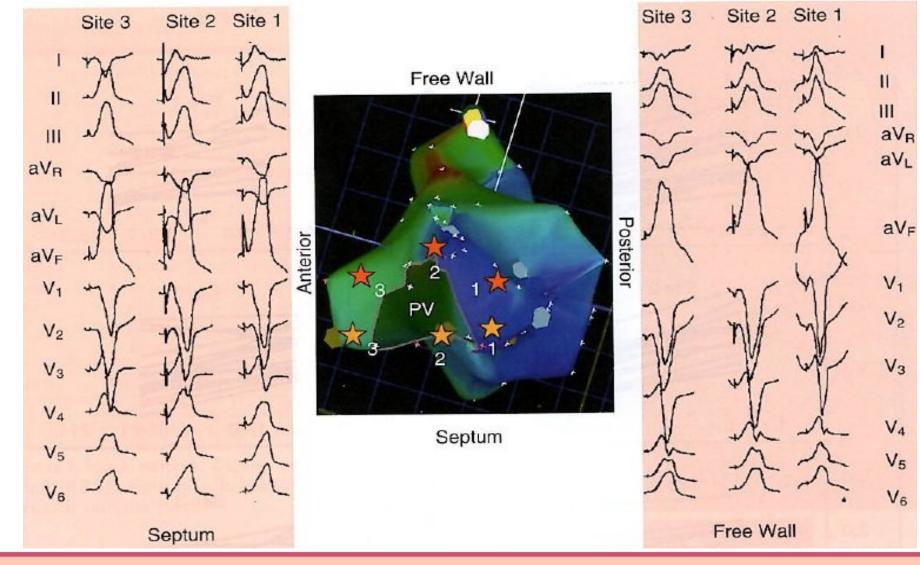
Uncommon (except AC)



9ª Edizione

### 12 EKG pace maps from sites 1 to 3 along the septum and free-wall aspects of the RVOT

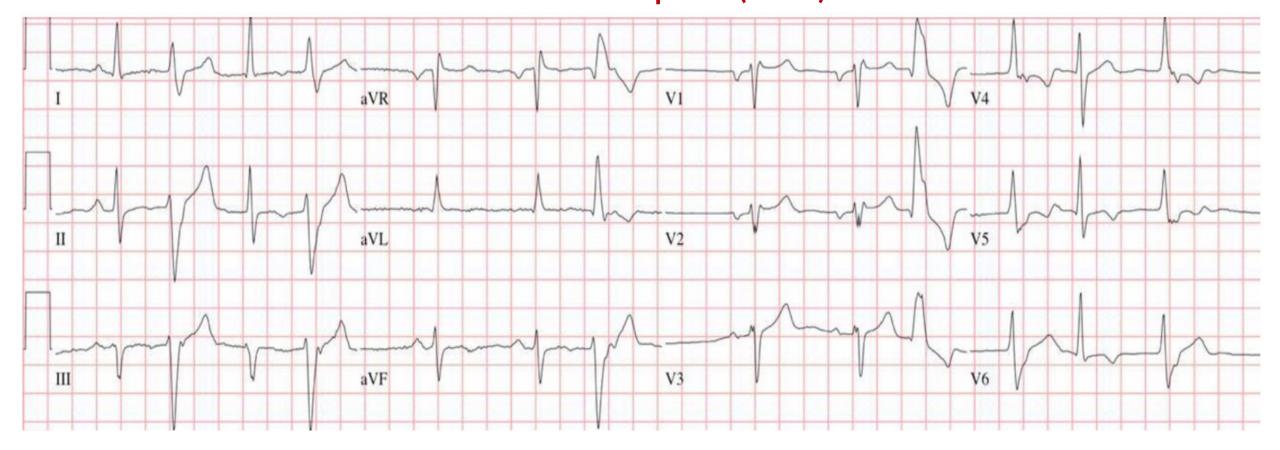






# Ventricular arrhythmias in the absence of structural heart Papillary muscle presentature ventricular complex (PVC)

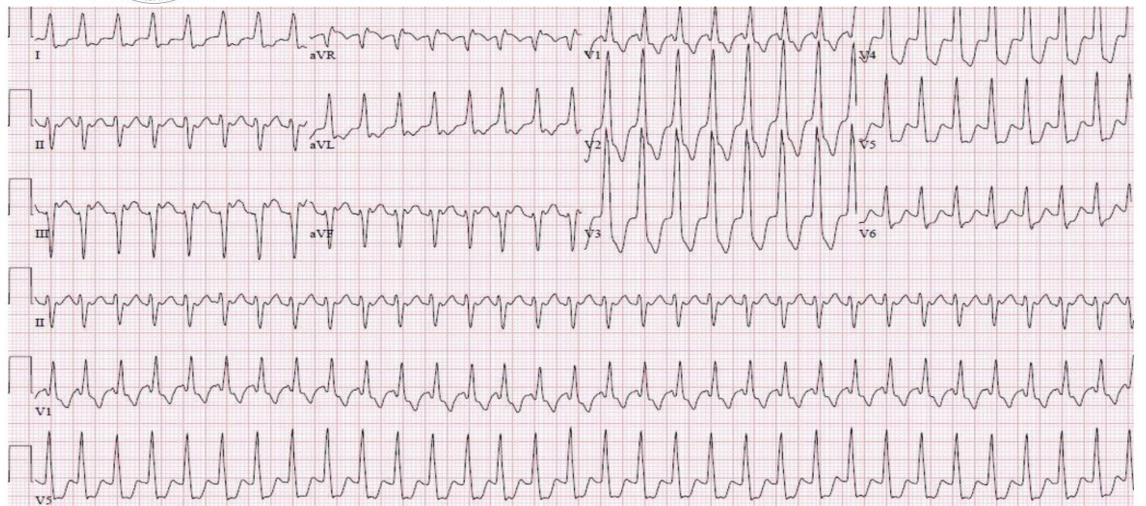






### Ventricular arrhythmias in the absence of structural heart





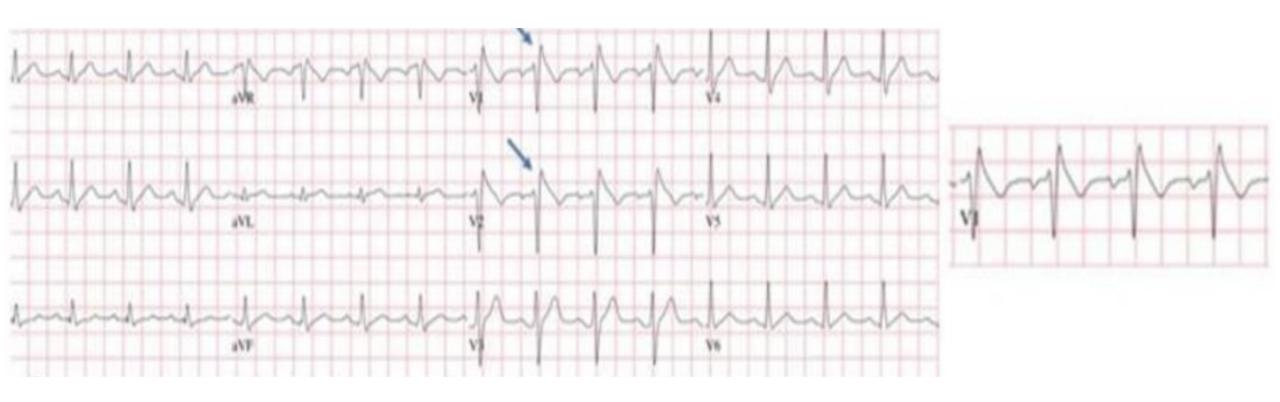


### Ventricular arrhythmias in the absence of structural heart ECG examples of inheined arrhythmia syndromes





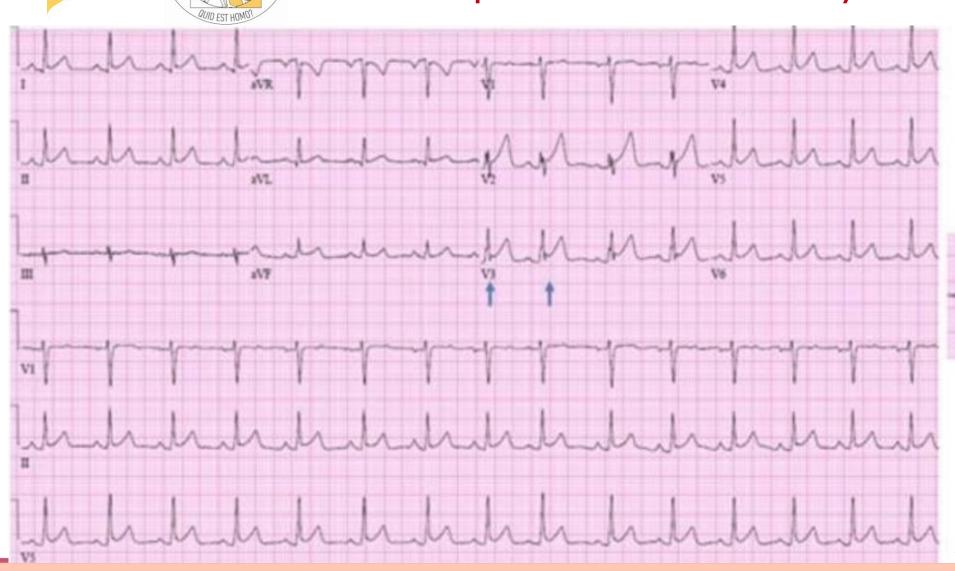
### Ventricular arrhythmias in the absence of structural heart ECG examples of inheined arrhythmia syndromes

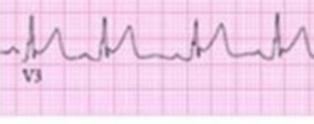




#### Ventricular arrhythmias in the absence of structural heart

### ECG examples of inheisted arrhythmia syndromes





Killu AM, Stevenson WG. Heart 2018;0:1-12. doi:10.1136/heartjnl-2017-

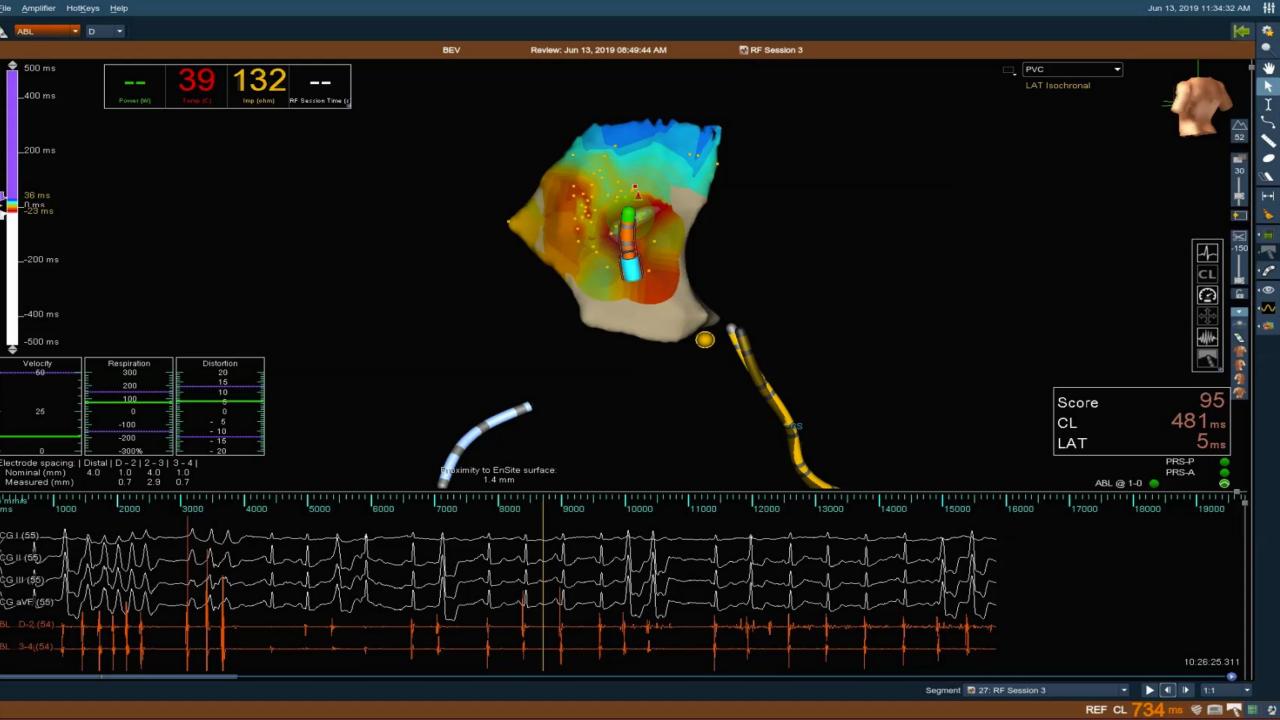


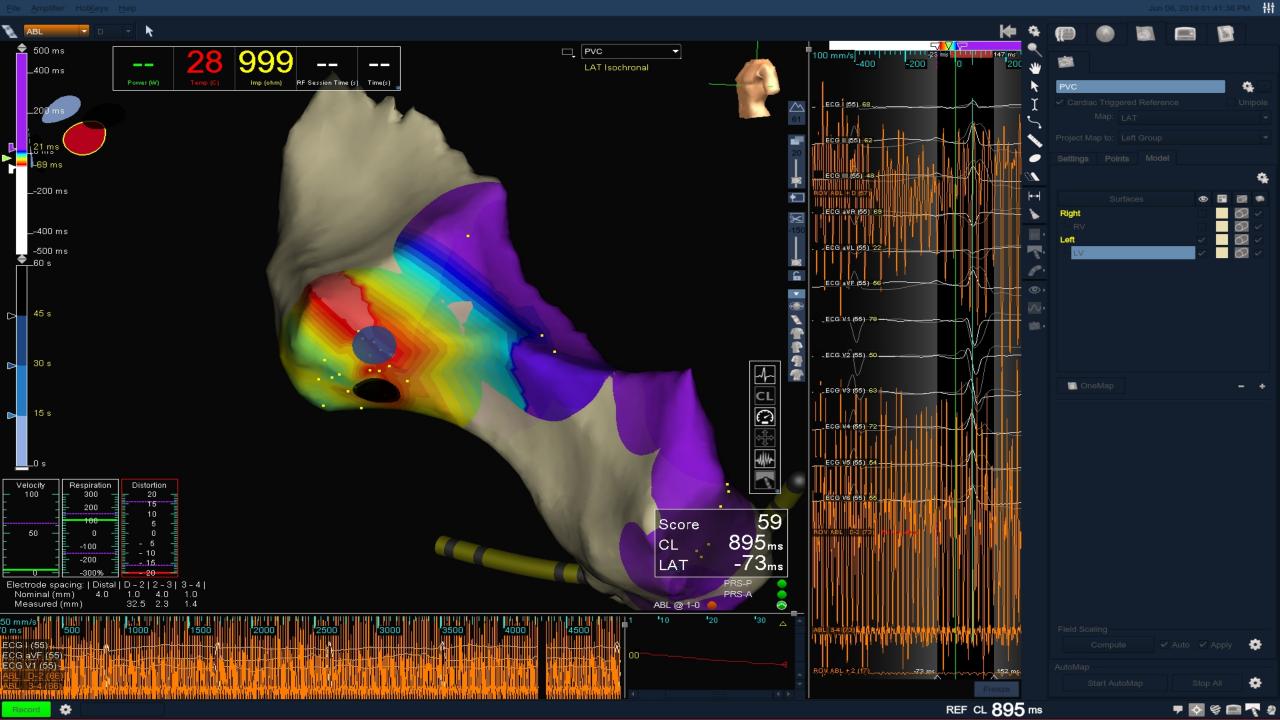
#### Ventricular arrhy

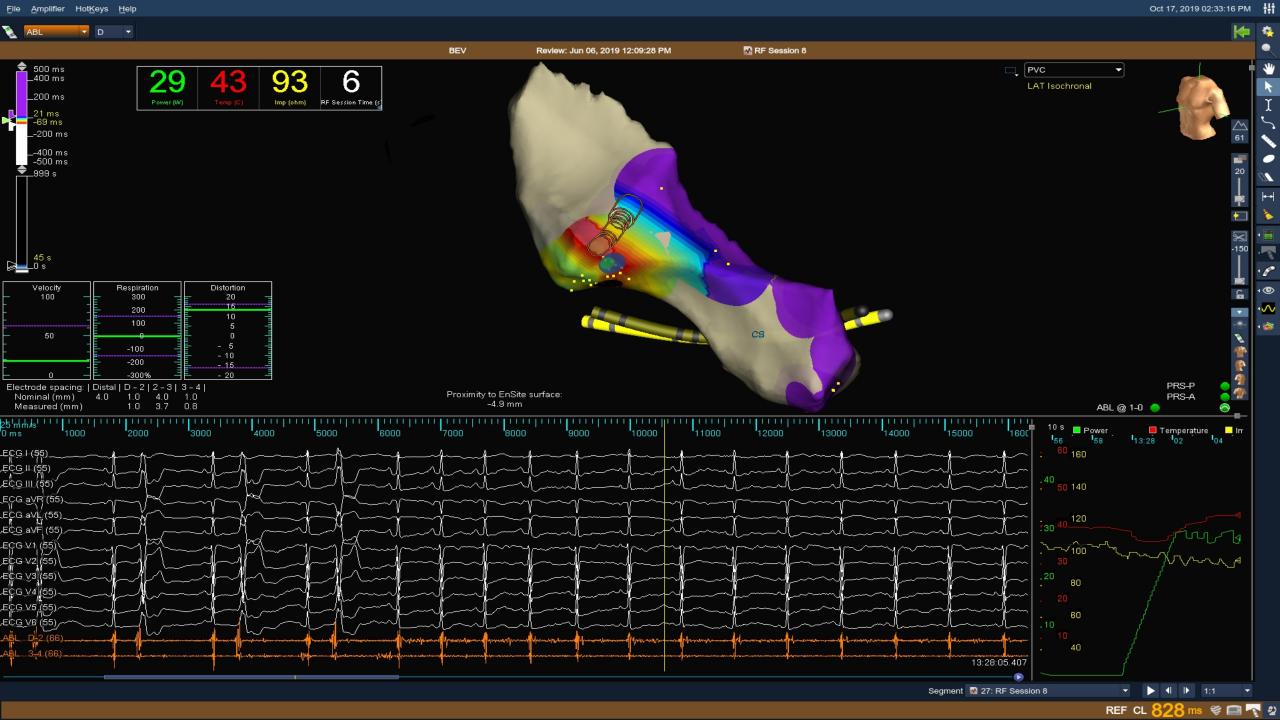
Comondicasco	ythm	ias i	n the	absend	ce of	structur	al	hear
Sumndisease	Su	mn	disea	se				

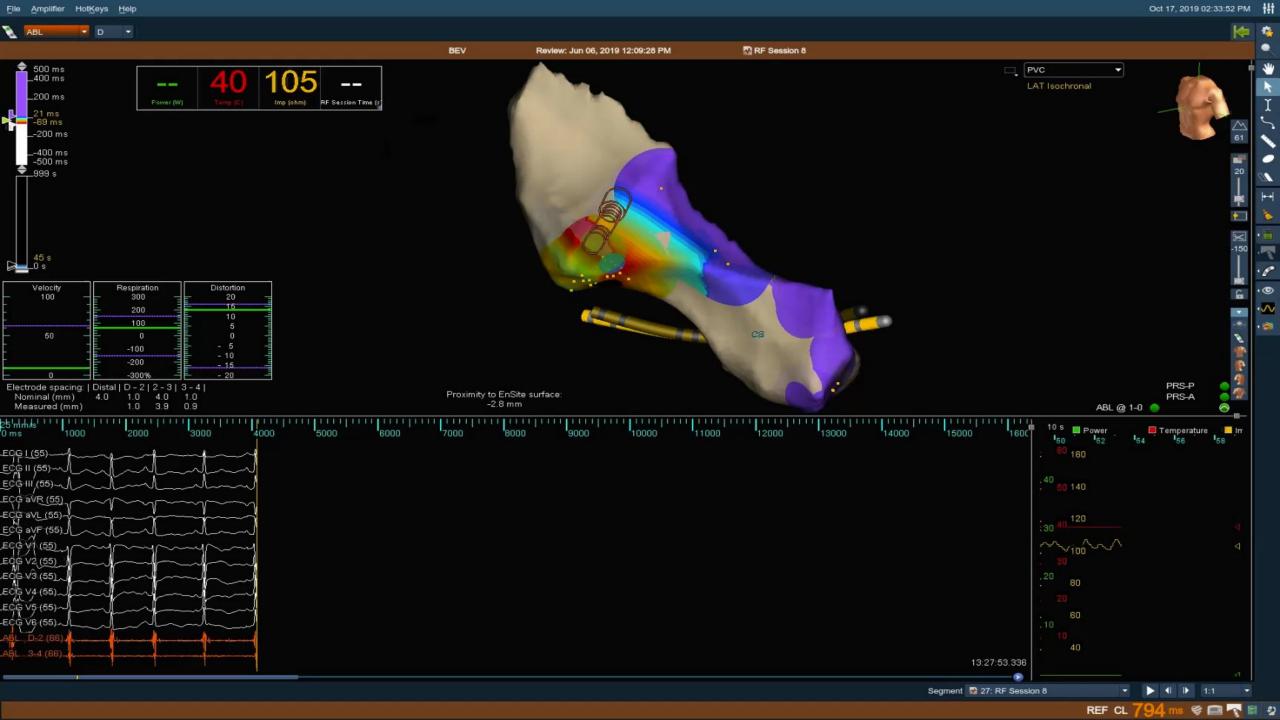
Table 2 Summary features of idiopathic monomorphic VTs and inherited arrhythmia syndromes							
Idiopathic ventricular arrhythmia type	Age group (years)	Gender	Likely mechanism	ECG characteristics	Effective medications	Success rate of ablation	Miscellaneous comments
Non-inherited VT in structura	lly normal hearts						
Outflow tract VT	30–50	F>M	DAD	LBBB more common than RBBB, inferior axis	BB, CCB, Class IC or III AAD	>80%	Atrial/ventricular burst pacing usually induce VTIsoproterenol or epinephrine facilitate induction
Interfascicular VT	20–40	M>F	Re-entry	RBBB/LS axis (LPF exit); RBBB/RI axis (LAF exit)	CCB (Verapamil), BB	80%	
Automatic fascicular VT (propranolol sensitive)	Variable	Variable	Abnormal automaticity or triggered activity	Typical or atypical LBBB or RBBB	BB (Propranolol)	Not established	
Mitral annular VT	50–70	M≥F	Abnormal automaticity of triggered activity	RBBB morphology, R>S in V1–V4	BB, class Ic or III AAD	>90%	
Tricuspid annular VT	50–70	M≥F	Abnormal automaticity of triggered activity	LBBB morphology, —aVR, +I. Other features vary depending on exact location	BB, class Ic or III AAD	90%	Success rate of ablation lower if arising from septal aspect of tricuspid annulus
Papillary muscle VT	40–70	M≥F	Abnormal automaticity of triggered activity	QRS>150 ms, RBBB morphology RS or rS in V3—V6Axis is superior if from PPM	BB, CCB, class Ic or III AAD	Limited data	
Inflammatory VT (eg, cardiac sarcoidosis, myocarditis)	Variable: depends on underlying condition	Variable: depends on underlying condition	Functional or scar-related re- entry most common depending on phase of disease	Variable, depending on site of inflammation	Supportive with immunosuppressionMechanical support and cardiac transplantation may be required in fulminant cases	Cardiac sarcoidosis: ~50% at 1–2 years <sup>44</sup> Myocarditis: 90% at 2 years <sup>45</sup>	Cardiac MRI, FDG-PET scan and endocardial biopsy important
Inherited VT in structurally no	ormal hearts						
LQTS	Variable	M = F	APD prolongation Re-entry	PMVT QTc usually 460 ms in sinus rhythm	BB, LCSD, pacing, ICD	N/A	Variable additional pharmacological options depending on subtype
SQTS	Variable. Consider in young patient with AF and short QT interval	M = F	APD shortening Re-entry	PMVT QTc<360 ms, short/absent ST segment, peaked T-wave, PR depression	ICD, Quinidine	N/A	Caused by shortening of repolarisation and APD. 2° to gain of function (K <sup>+</sup> channel) or loss of function (Ca <sup>2+</sup> channel) mutations
CPVT	<30	M = F	DAD	PMVT, bidirectional VT Normal sinus rhythm ECG at rest	BB (nadolol), CCB, flecainide, propafenone, LCSD, avoidance of competitive sport	N/A	Due to a malfunctioning ryanodine receptor → results in diastolic Ca <sup>2+</sup> overload with DAD
BrS	20–40	M >> F	Re-entry	PMVT ST elevation in V1–V3 in sinus rhythm (may require provocation)	ICD Quinidine, isoproterenol (for VT storm)	Limited data: reportedly >80%	
ERS	20–50	$M \ge F$	Re-entry	VF J-point elevation in sinus rhythm	ICD Quinidine	N/A	

















### TAKE HOME MESSAGE

Catheter ablation of ventricular arrhythmias is useful and effective in a pediatric population. Idiopathic and fascicular VAs have lower recurrence rates when compared with VA associated with cardiomyopathies