

Gestione del WPW in età pediatrica e adolescenziale: terapia, stratificazione del rischio e indicazioni all'ablazione

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Accessory Pathways : *NATURAL HISTORY*

- Prevalence of AP is estimated to be 1-3 in 1000 individuals
- In the first year of life the accessory pathway loses anterograde conduction in as many as 40% of patients
- Approximately 65% of adolescents with a AP pattern on a resting ECG are asymptomatic
 - Up to 46% of asymptomatic patients may not have retrograde conduction and may never develop tachycardias
 - Up to 30% of asymptomatic are thought to develop arrhythmias during a 10-year follow-up

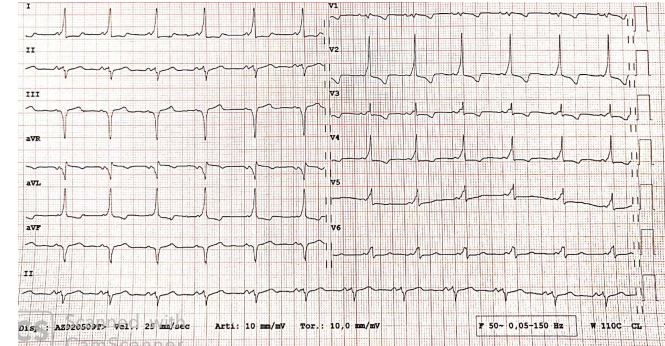
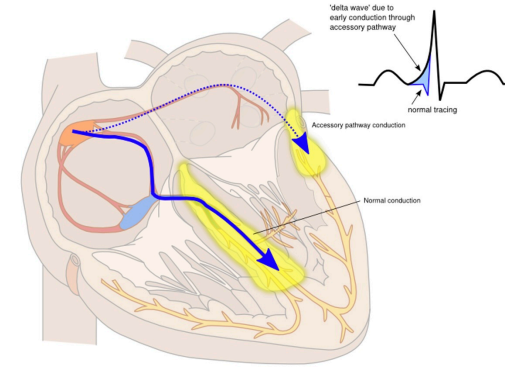


Accessory Pathways : *NATURAL HISTORY*

In 80% of cases it is associated with a **normal heart**

In 20% it is **associated** with cardiomyopathies or congenital heart disease:

- Ebstein
- Tricuspid atresia
- Congenitally corrected transposition of the great vessels,
- Hypertrophic cardiomyopathy
- Rhabdomyoma
- Tuberous sclerosis
- Storage diseases (Pompe and Danon disease)
- Mitochondrial diseases



The association with cardiomyopathy **increases the risk of AP**





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European Society
of Cardiology

European Heart Journal (2021) 42, 17–96

doi:10.1093/eurheartj/ehaa605

ESC GUIDELINES

2020 ESC Guidelines on sports cardiology and exercise in patients with cardiovascular disease

The Task Force on sports cardiology and exercise in patients with cardiovascular disease of the European Society of Cardiology (ESC)

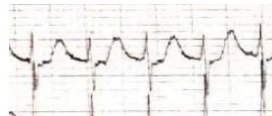
In pediatric age compared to adults the risk of induction of AF following a ARVT is anecdotal in healthy and small hearts despite very fast ARVT

The risk increases if we are faced with hearts with heart disease or genetic predispositions

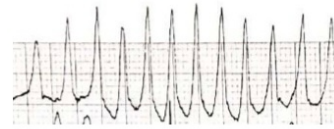
MECHANISM



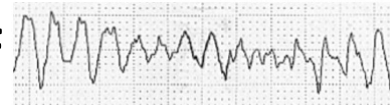
AVRT



AF

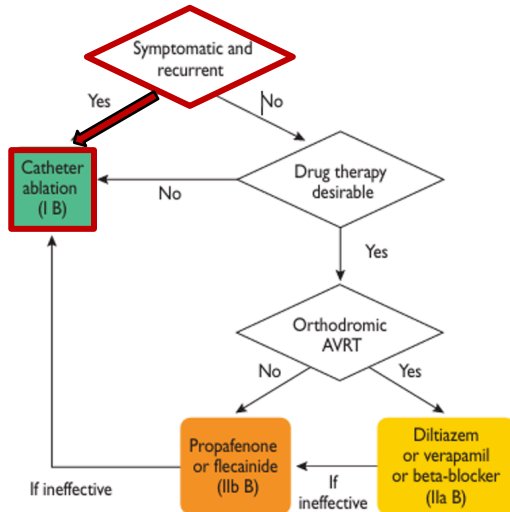


VF





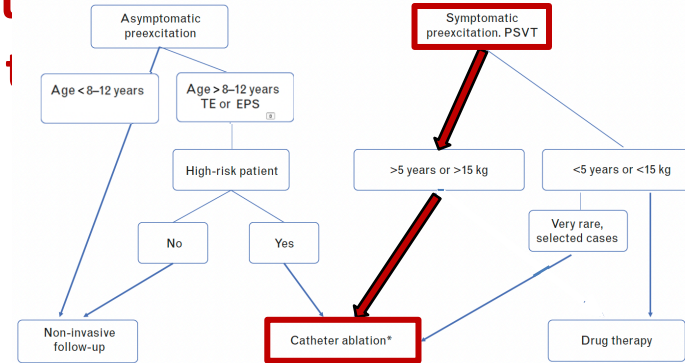
2019 ESC Guidelines for the management of patients with supraventricular tachycardia



with symptomatic pre-excitation the choice of what treatment

Diagnosis and treatment of fetal and pediatric age patients (0–12 years) with Wolff–Parkinson–White syndrome and atrioventricular accessory pathways

Loira Leoni^a, Gabriele Bronzetti^b, Diego Colonna^c, Giulio Porcedda^d, Alessandro Rimini^e and Massimo Stefano Silvetti^f,
Area Pediatrica, Associazione Italiana di Aritmologia e Cardioritmologia (AIAC)
J Cardiovasc Med 2023, 24:1–13

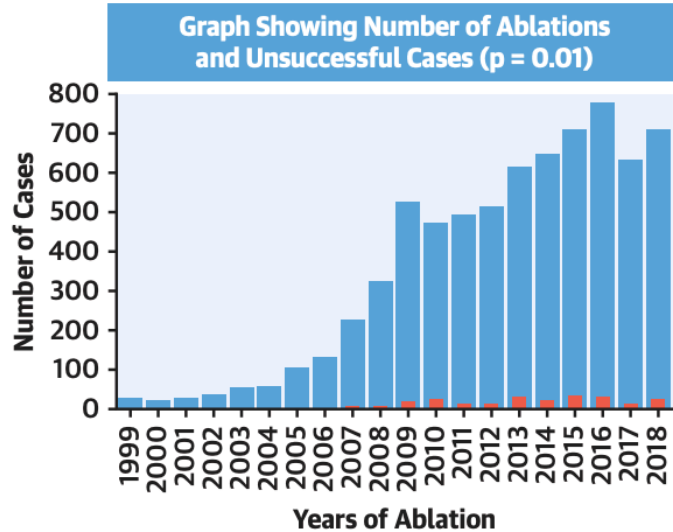


In the pediatric population, catheter ablation must be performed at HIGHLY EXPERIENCED CENTERS and in according TO AGE OR WEIGHT



Outcomes From Pediatric Ablation

A Review of 20 Years of National Data



CONCLUSIONS Overall **SUCCESS RATES FROM PEDIATRIC ABLATIONS**

ARE EXCELLENT and compare favorably to other registries.

Introduction of newer technologies have likely made **procedures safer** and **reduced radiation exposure**

7,069 Pediatric Ablations
Increasing success rates over time

2,445 AVNRT

92% Success Rate

4,187 Accessory Pathways

97% Success Rate

385 Atrial Tachycardia

89% Success Rate

Study used NICOR (National Institute of Cardiovascular Outcomes Research)



Cardiology

Cardiac Surgery: Systematic Review

Cardiology 2025;150:320–338
DOI: 10.1159/000541178

Received: April 17, 2024
Accepted: August 12, 2024
Published online: September 5, 2024

Conclusion: RFCA appears to be a highly effective and safe treatment option for AVRT and AVNRT in children, with high success rates and relatively low recurrence and complication rates.

Meta-Analysis of the Efficacy and Safety of Radiofrequency Catheter Ablation for Pediatric Paroxysmal Supraventricular Tachycardia

HIGHLY EFFECTIVE AND SAFE FOR AVRT AND AVNRT IN CHILDREN

 **ESC**
European Society
of Cardiology
Europace (2021) 23, 431–440
doi:10.1093/europace/eaab325

CLINICAL RESEARCH
Clinical electrophysiology and ablation

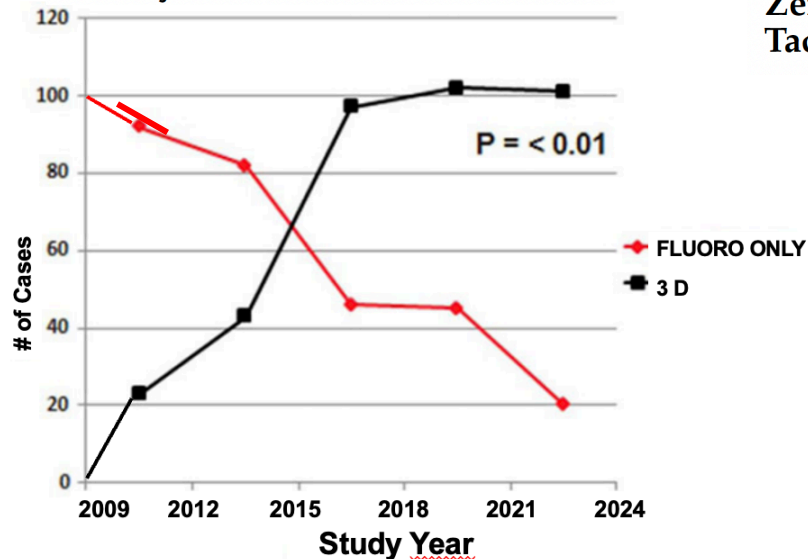
Pediatric catheter ablation at the beginning of the 21st century: results from the European Multicenter Pediatric Catheter Ablation Registry ‘EUROPA’

The present study proves over all **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.



META-ANALYSIS Heart Rhythm 2026

Zero- or minimal-fluoroscopy vs conventional fluoroscopy in catheter ablation of supraventricular tachycardias: An updated systematic review and meta-analysis



Article

Zero-Fluoroscopy Catheter Ablation of Supraventricular Tachycardias in the Pediatric Population

CONCLUSION ZF/MF ablation provides **equivalent acute efficacy, marginal improvement in long-term success, and markedly reduces radiation exposure without increasing complications.**

These findings support ZF/MF as a safe and effective alternative to CF, with particular relevance for **minimizing radiation exposure in vulnerable populations.**

EQUIVALENT EFFICACY AND SAFETY + MINIMIZING RADIATION EXPOSURE IN VULNERABLE POPULATIONS



What to do in asymptomatic patients?

What is their risk of sudden death?

Is it better to just observe or to perform ablation on everyone?



Ventricular pre-excitation: symptomatic and asymptomatic children have the same potential risk of sudden cardiac death

Table 1 Transoesophageal electrophysiological study results

	WPW	Asymptomatic VPE	P-value
Patients (n)	54	73	N/A

The **accessory pathway in asymptomatic** patients can be just as "dangerous" as in symptomatic patients because **asymptomatic status is not predictive of the conduction speed of the accessory pathway.**

30% of asymptomatic are thought to develop arrhythmias during a 10-year follow-up

Every asymptomatic is a potential symptomatic with the same potential risk of SCD !

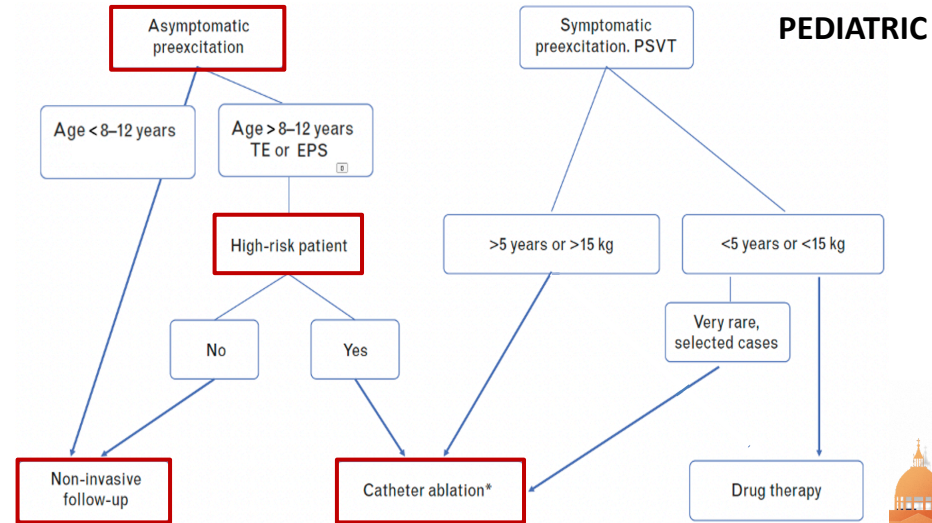
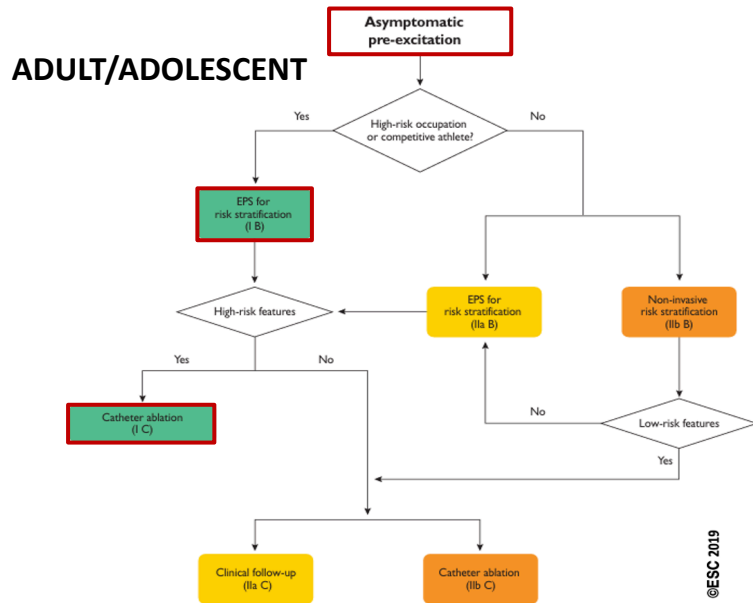
shortest pre-excited R–R interval; TEEPS: transoesophageal electrophysiological study; VPE, ventricular pre-excitation; WPW, Wolff–Parkinson–White syndrome.

^a% of total patients.

^b% of patients studied during isoproterenol infusion or stress testing.

^c% of patients with AF.

In asymptomatic patients, *non invasive and invasive risk stratification* may identify a subgroup of patients with essentially no risk of SCD, which may assist in decision making for certain individuals



RISK STRATIFICATION IN WPW



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of Cardiology

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

ESC GUIDELINES



2019 ESC Guidelines for the management of patients with supraventricular tachycardia

Asymptomatic patients with **intermittent** pre-excitation (at rest
or during exercise) or **abrupt disappearance** of pre-excitation
during stress testing ***may be considered to be at low risk***
Exercise stress test



Fall of the second paradigm

Loss of ventricular preexcitation during noninvasive testing does not exclude high-risk accessory pathways: A multicenter study of WPW in children

Escudero et al Heart Rhythm, Vol 17, No 10, October 2020

Table 2 Clinical and EPS findings in patients with nonpersistent preexcitation and SCA or RC-AF

Patient no.	Age at event (y)	Testing defining nonpersistent preexcitation	Event type	Clinical SPERRI (ms)	Symptoms before the event	HD	AP location	Anesthesia at EPS	EPS data (ms)
1	12.9	Holter monitoring	RC-AF	230	SVT	No	Left anterolateral	Sedation	APERP 225 SPERRI 200
2	13.9	Unknown	RC-AF	200	Palpitations	No	Left lateral	GA	APERP 310 SPCL 260
3	16.9	EST (loss at HR 140 beats/min)	RC-AF	200	Syncope, SVT	Ebstein anomaly	Right posteroseptal	GA	None tested
4	9.4	ECG/Holter monitoring	R-SCA		SVT	No	Right posterior	GA	No preexcitation during atrial fibrillation
5	16.3	ECG/Holter monitoring	R-SCA		SVT	No	Left posterolateral	GA	APERP 280
6	18.7	ECG/Holter monitoring	R-SCA		SVT	No	Right anterosseptal	GA	APERP 350 SPERRI 370

AP = accessory pathway; APERP = accessory pathway effective refractory period; CHD = congenital heart disease; ECG = electrocardiogram; EPS = electrophysiology study; EST = exercise stress test; GA = general anesthesia; HR = heart rate; RC-AF = rapidly conducted preexcited atrial fibrillation; R-SCA = resuscitated sudden cardiac arrest; SPERRI = shortest preexcited R-R interval in atrial fibrillation at electrophysiology study; SPCL = shortest preexcited paced cycle length; SVT = supraventricular tachycardia.

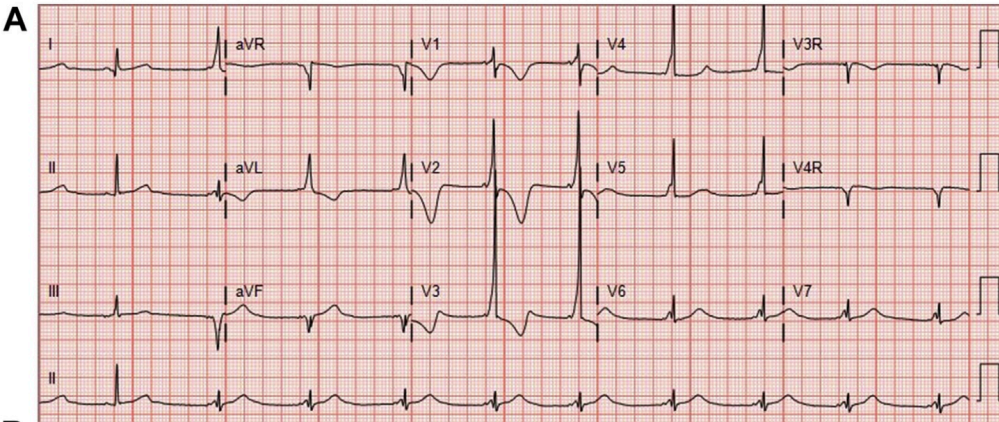
1589 patients

244 non persistent preexcitation

CONCLUSION

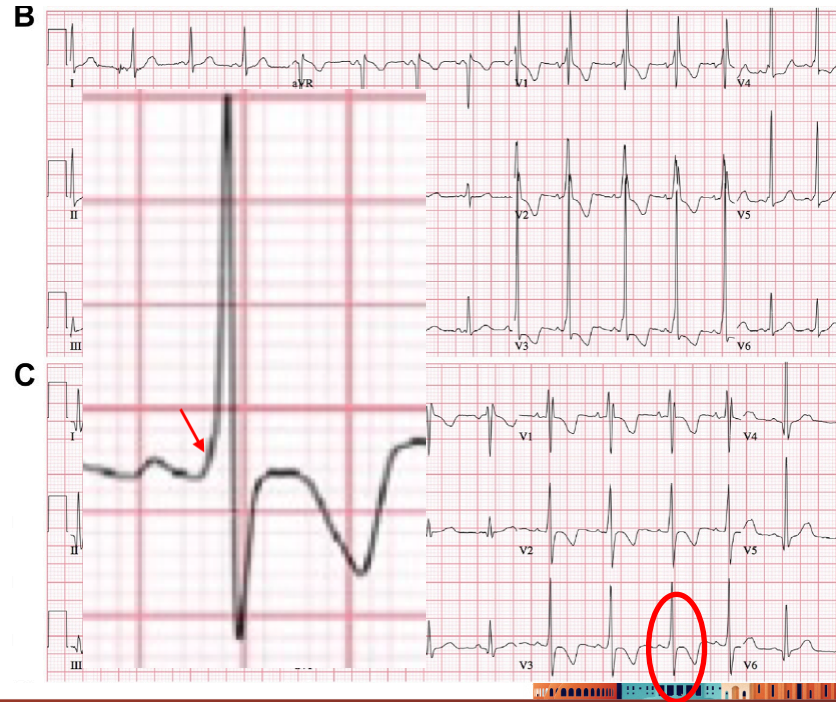
In this large, multicenter, international study of children with WPW, the overall likelihood of meeting **high-risk criteria at invasive EPS was lower in patients with nonpersistent preexcitation**, although **13% of these patients had high-risk APs at EPS**. In addition, there were **patients with nonpersistent preexcitation who experienced SCA or RC-AF, all of whom were previously symptomatic**. This study demonstrates that the presence of nonpersistent preexcitation in children with WPW may not guarantee the absence of a high-risk AP or the absence of the risk of SCA or RC-AF.

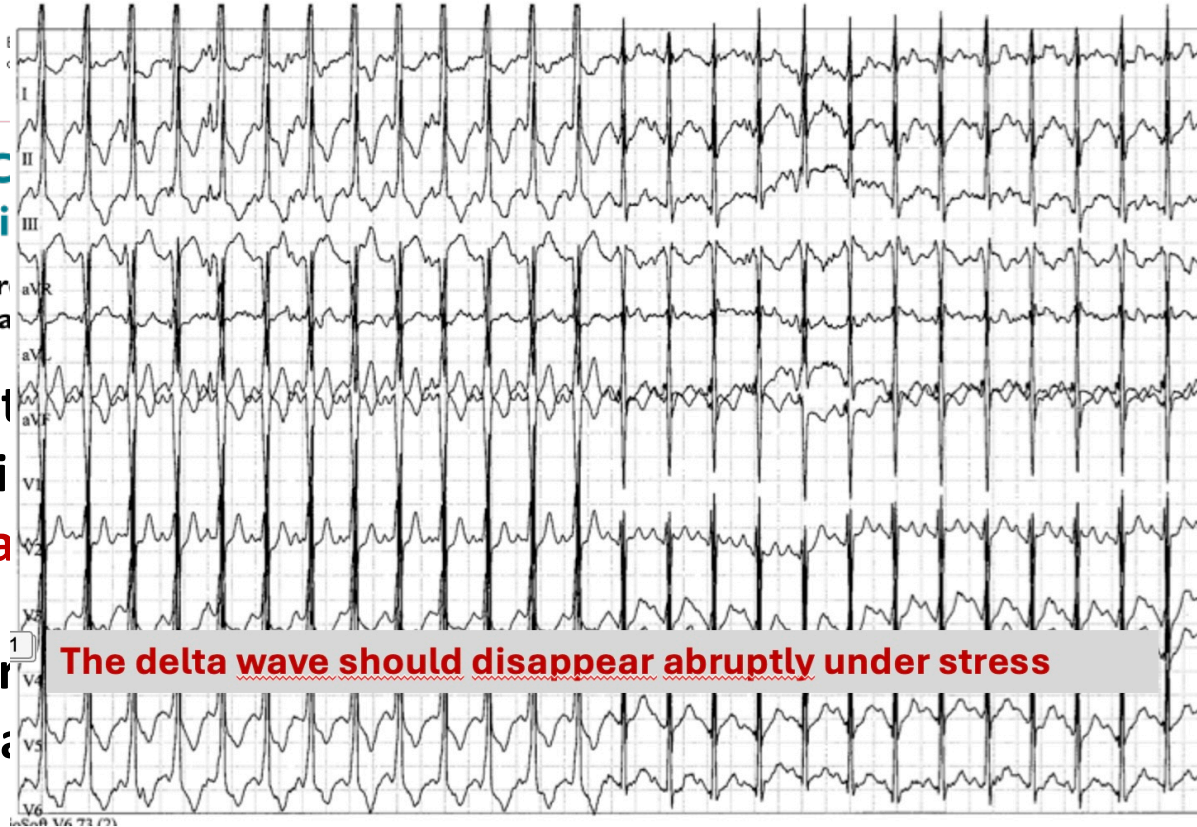




LATENT?

Electrocardiograms (ECGs) for patients 4 and 5 with nonpersistent preexcitation and sudden cardiac arrest (SCA).

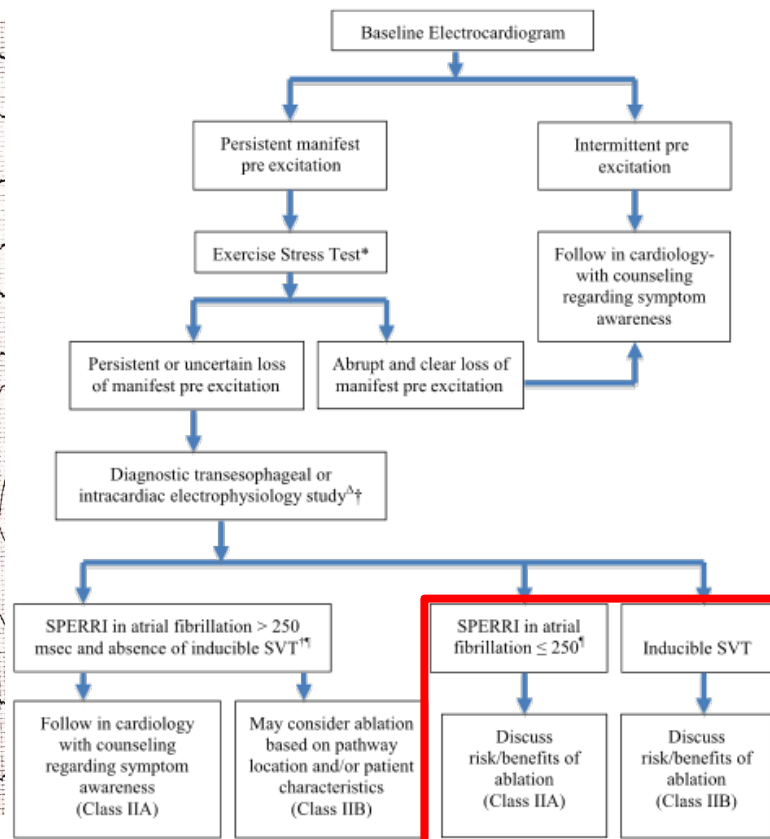
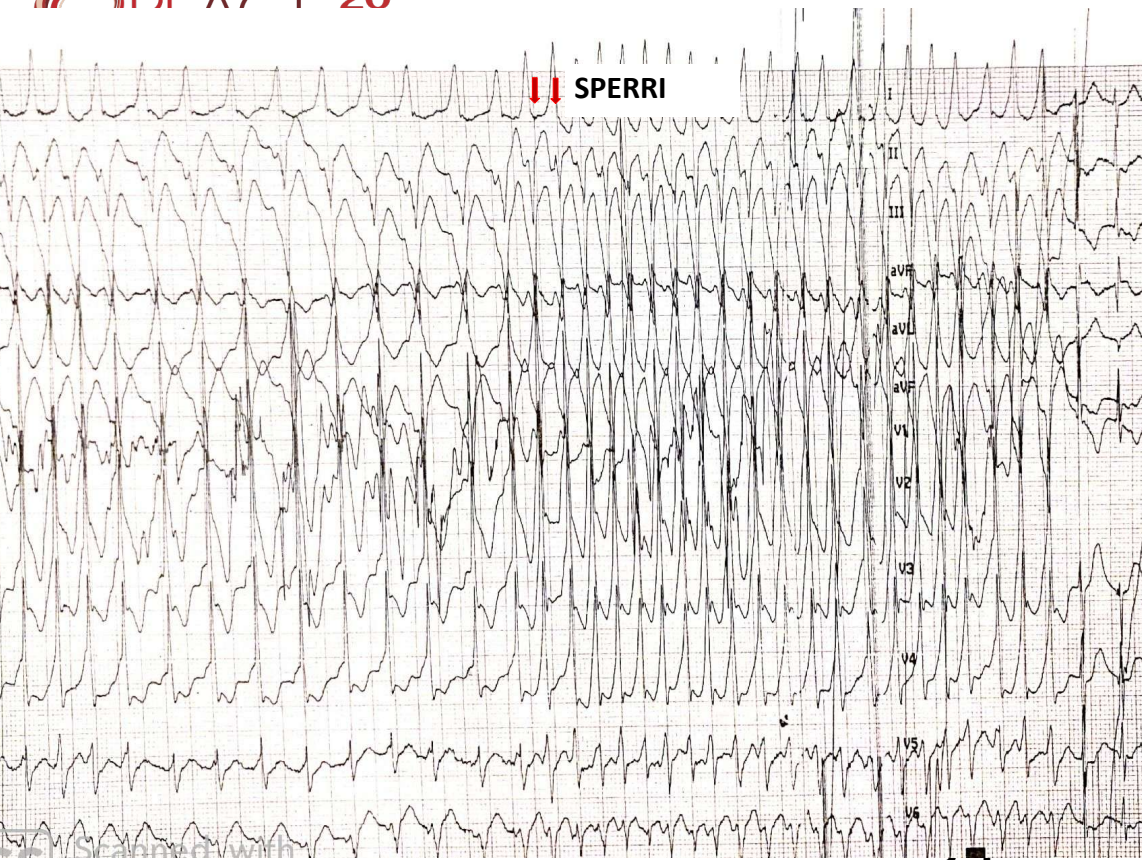




low risk
risks may be

the sudden

RISK STRATIFICATION IN WPW



REVIEW ARTICLE

Risk-Stratification Strategy for Sudden Cardiac Death in the Very Young Children with Asymptomatic Ventricular Preexcitation

Moises Rodriguez-Gonzalez^{1,2,*}, Ana Castellano-Martinez^{2,3} and Alvaro A. Perez-Reviriego^{1,2}

- | |
|--|
| 1. APERP cycle length < 250 ms at rest or < 220 ms during adrenergic stress, where the AP fails to conduct anterogradely on rapid atrial pacing. |
| 2. SPERRI in spontaneous or induced atrial fibrillation < 250 ms at rest or < 220 ms during adrenergic stress. |
| 3. Inducibility of an AVRT, with and without isoproterenol. |
| 4. The presence of multiple AP. |





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European Heart Journal (2022) **43**, 3997–4126

<https://doi.org/10.1093/eurheartj/ehac262>

ESC GUIDELINES

2022 ESC Guidelines for the management of patients with ventricular arrhythmias and the prevention of sudden cardiac death

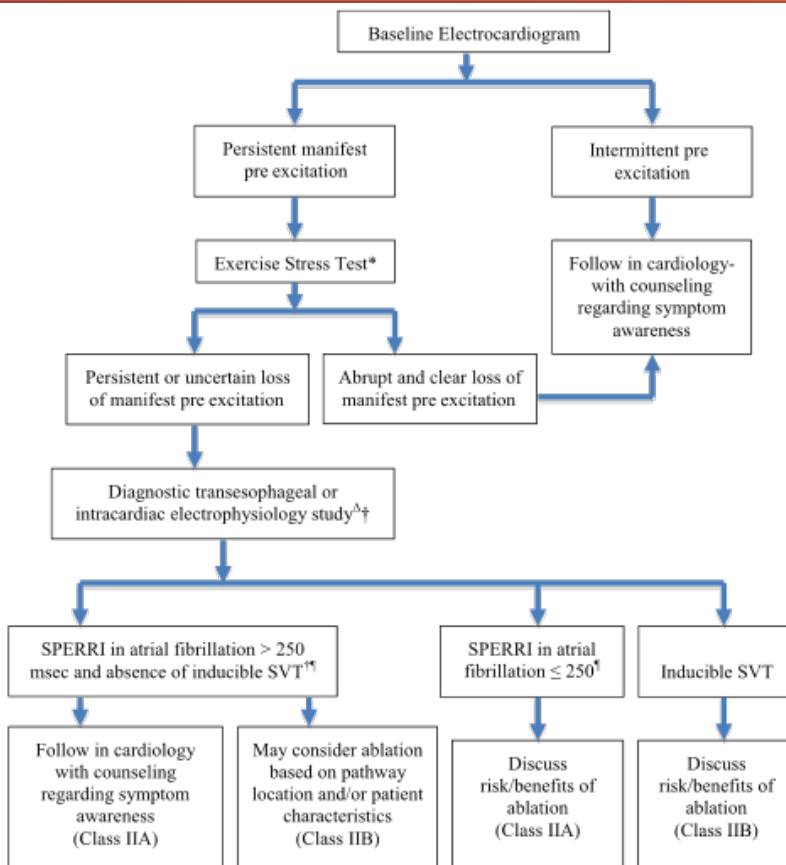
The current guidelines say that in children with asymptomatic accessory pathways, **risk stratification is not recommend before the age of 8 years.**



PACES/HRS Expert Consensus Statement on the Management of the Asymptomatic Young Patient with a Wolff-Parkinson-White (WPW, Ventricular Preexcitation) Electrocardiographic Pattern

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)

Children ages 5–8 years may be placed in the “younger-observe”



2020 ESC Guidelines on sports cardiology and exercise in patients with cardiovascular disease

The Task Force on sports cardiology and exercise in patients with cardiovascular disease of the European Society of Cardiology (ESC)

In recreational athletes with asymptomatic pre-excitation, risk assessment may first be pursued via non-invasive testing.

The sensitivity of non-invasive screening for AP is good, but its specificity is low.

Of note, *in children younger than 12 years, the risk of AF-induced VF and sudden death is very low. Generally, a conservative approach approach is recommended in this age group*

Recommendations for exercise and sports participation in individuals with paroxysmal supraventricular tachycardia and pre-excitation

Recommendations	Class ^a	Level ^b
In individuals with palpitations, a comprehensive assessment to exclude (latent) pre-excitation, structural heart disease, and VAs is recommended. ⁵⁰⁰	I	B
Participation in all sports activities is recommended in individuals PSVT without pre-excitation. ⁵⁰⁰	I	C
Ablation of the accessory pathway is recommended in competitive and recreational athletes with pre-excitation and documented arrhythmias. ⁵⁰⁰	I	C
In competitive/professional athletes with asymptomatic pre-excitation, an EP study is recommended to evaluate the risk for sudden death. ^{497,500}	I	B
In competitive athletes with PSVT but without pre-excitation, curative treatment by ablation should be considered.	IIa	C

EP = electrophysiological; PSVT = paroxysmal supraventricular tachycardia; VA = ventricular arrhythmia.

^aClass of recommendation.

^bLevel of evidence.

When to perform ablation in asymptomatic pediatric patients?

ABLATION:

In patients > 8 years
risk stratification

NO ABLATION

In children under
up to 8-12 years
procedural risk

In patients > 8 years
invasive risk stratification

Table 4 Indications for catheter ablation and oral prophylactic antiarrhythmic drugs for recurrent SVT or VT

Clinical situation	Recommendation	Class	Level	Reference
WPW syndrome and episode of aborted SCD	Catheter ablation	I	C	
WPW syndrome and syncope combined with preexcited RR interval during AF <250 ms or antegrade APERP during PES <250 ms	Catheter ablation	I	C	
Incessant or recurrent SVT associated with ventricular dysfunction	Catheter ablation	I	C	
Recurrent monomorphic VT with haemodynamic compromise and amenable to catheter ablation	Catheter ablation	I	C	
WPW syndrome and recurrent and/or symptomatic SVT and age > 5 years	Catheter ablation	I	C	
	Flecainide, propafenone	I		
	Sotalol	I		
	Amiodarone	IIb		
WPW syndrome and recurrent and/or symptomatic SVT and age < 5 years	Flecainide, propafenone	I	C	
	Sotalol	IIa		
	Catheter ablation	IIb		
	Amiodarone	IIb		
WPW syndrome and palpitations with inducible sustained SVT during EP test, age > 5 years	Catheter ablation	I	C	
	Flecainide, propafenone	I		
	Sotalol	I		
	Amiodarone	IIb		
Single or infrequent SVT (no pre-excitation), age > 5 years	None	I	C	
	Valsalva maneuver	I		
	'Pill-in-Pocket'	IIa		
	Flecainide (3 mg/kg), Diltiazem (120 mg) + Propranolol (80 mg)			
	Sotalol			
	Beta-blocking agents	I		
	Catheter ablation	IIb		
SVT, age > 5 years, chronic AA therapy has been effective in control of the arrhythmia	Catheter ablation	IIa	C	
SVT, age < 5 years (including infants), when AA medications, including Classes I and III are not effective or associated with intolerable side effects	Catheter ablation	IIa	C	
Asymptomatic preexcitation, age > 5 years, no recognized tachycardia, risks and benefits of procedure and arrhythmia clearly explained	Catheter ablation	III	C	
Asymptomatic preexcitation, age < 5 years	Any AA drug	III	C	
SVT controlled with conventional AA medications, age < 5 years	Catheter ablation	III	C	
Idiopathic monomorphic ventricular tachycardia	Propranolol	IIb	C	167,170
	Sotalol			
	Flecainide, propafenone	IIa	C	
	Verapamil			
	Procainamide			
	Amiodarone			

WPW, Wolff-Parkinson-White; SVT, supraventricular tachycardia; LV, left ventricular; SCD, sudden cardiac death; AF, atrial fibrillation; APERP, accessory pathway effective refractory period; PES, programmed electrical stimulation; EP, electrophysiological; AA, antiarrhythmic; level, level of Evidence; Class, recommendation classification.
*Patients should be free of significant LV dysfunction, sinus bradycardia, or pre-excitation.

asymptomatic and invasive

and in any case
low and the

non-invasive and



Thirty percent of asymptomatic patients become symptomatic over a 10-year follow-up period.

Asymptomatic patients are asymptomatic until they become symptomatic; the risk of a fast accessory pathway is not correlated with asymptomatic patients.

The risk of sudden death correlates with the risk of AF, so it increases in adults but is very low in pediatric patients.

Risk stratification is essential in pediatric patients.

Be careful not to confuse latent pathways with true intermittent pathways.

In asymptomatic children younger than 8-12 years of age, the assessment is limited to observation.

In children older than 8-12 years of age, risk stratification is necessary in asymptomatic children to guide the decision regarding ablation.

In asymptomatic adults, stratification is important, but the possibility of proceeding directly with ablation should be considered.



MASTER II LIVELLO

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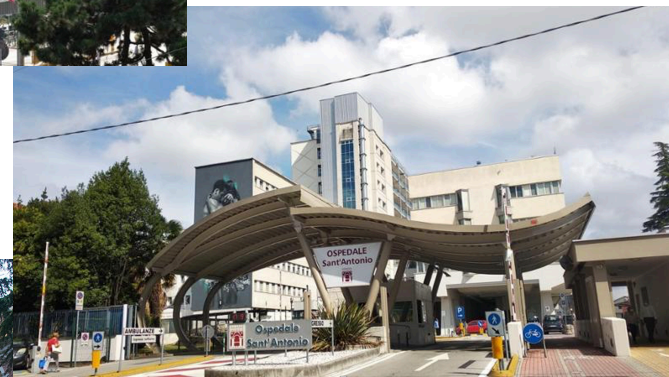


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