Preexcitace

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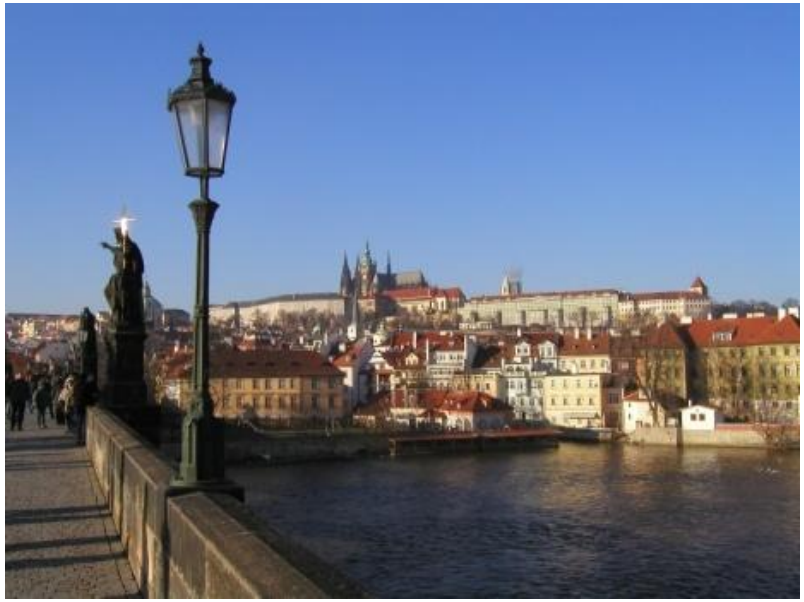
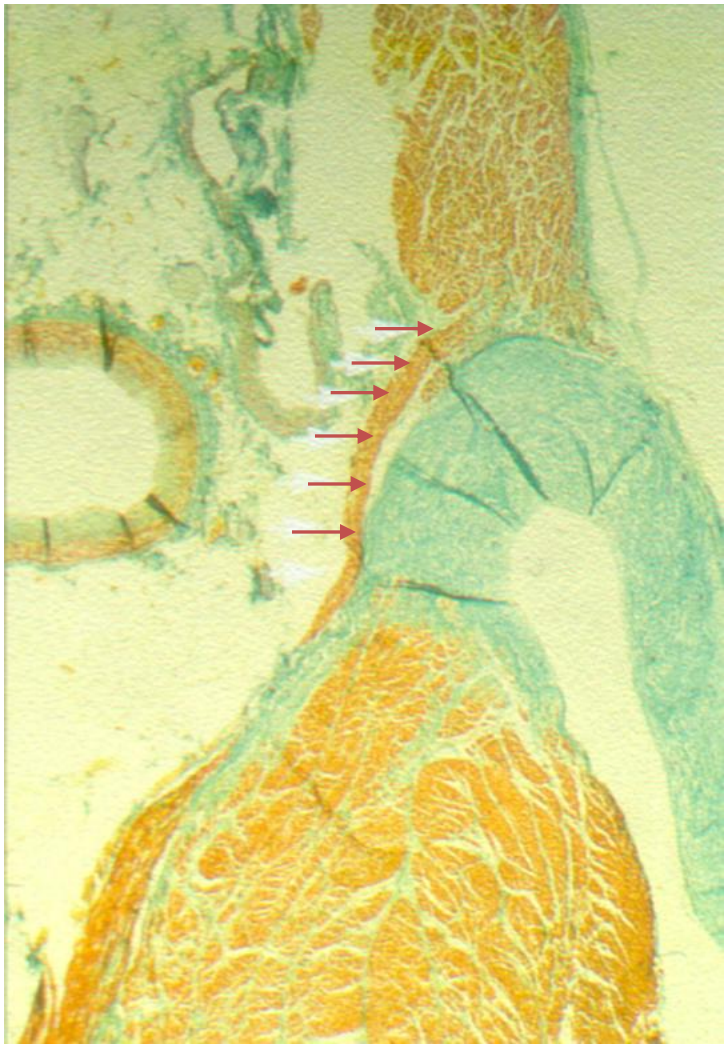
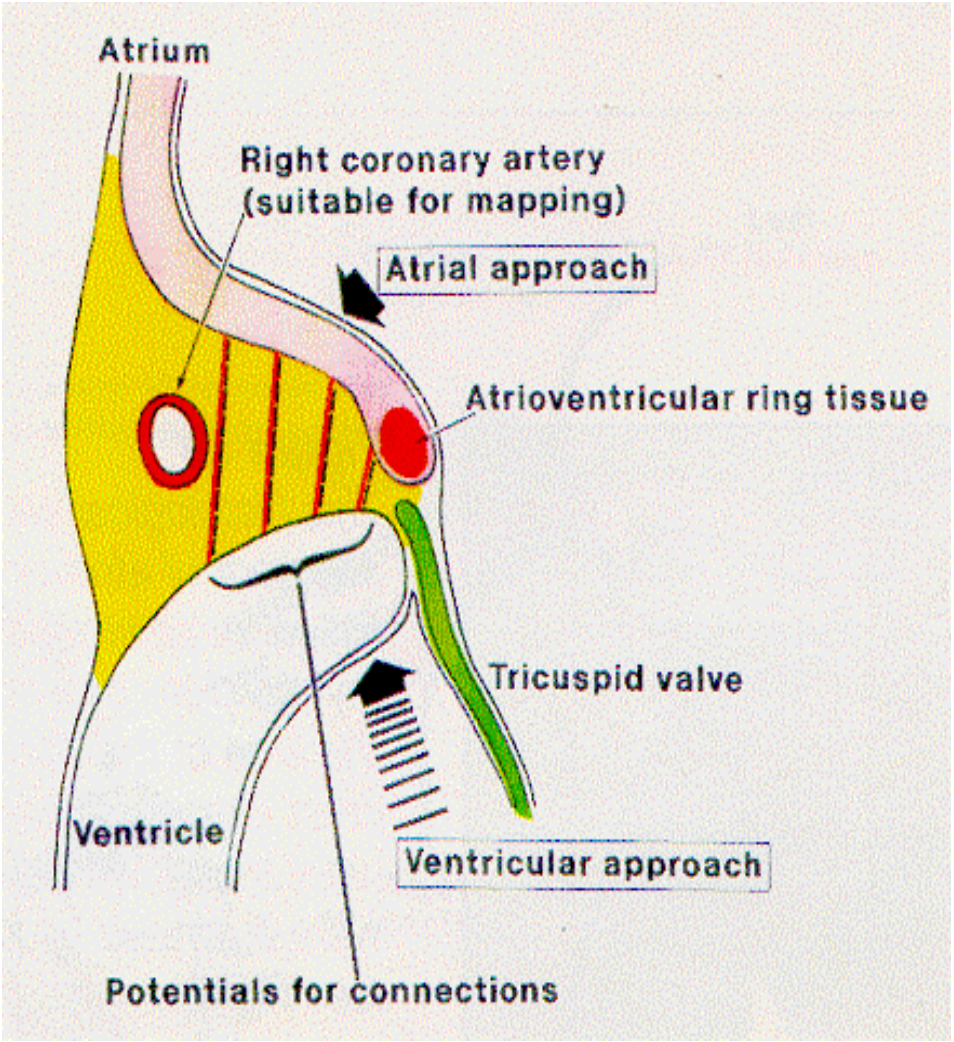


Anatomy of accessory atrioventricular pathways





Courtesy Paul T, Göttingen, Germany

Circulation
JOURNAL OF THE AMERICAN HEART ASSOCIATION

American Heart Association®

Accessory Atrioventricular Myocardial Connections in the Developing Human Heart: Relevance for Perinatal Supraventricular Tachycardias

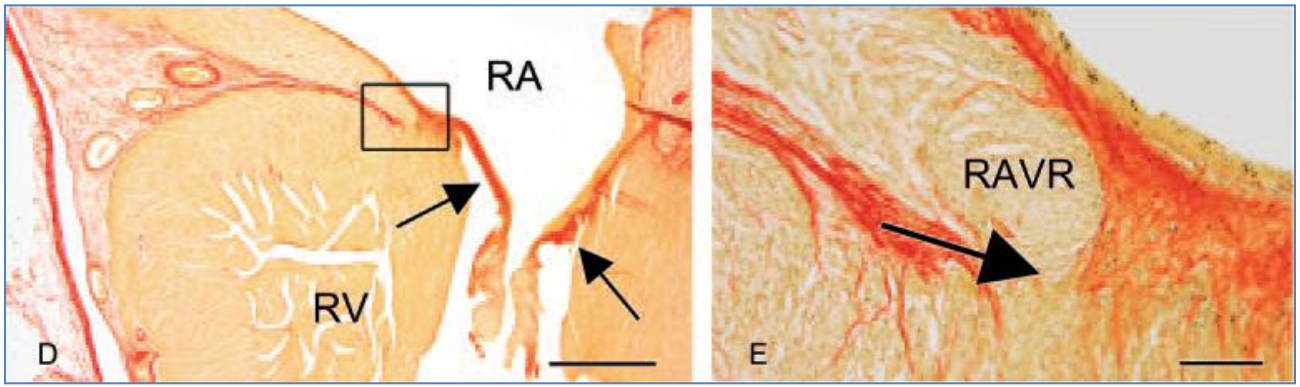
Nathan D. Hahurij, Adriana C. Gittenberger-De Groot, Denise P. Kolditz, Regina Bökenkamp, Martin J. Schalij, Robert E. Poelmann and Nico A. Blom

Circulation. 2008;117:2850-2858; originally published online May 27, 2008;

N = 45 human embryonic, fetal and neonatal hearts

Immunohistochemical examination of accessory myocardial AV connections

- 4 - 6 WOG A and V myocardium continuous at the primitive AV canal
- 6 - 10 WOG numerous accessory myocardial AV connections
  - left (45%), right (35%), septal (20%)
- 10 - 20 WOG accessory AV connections decreased in number
- >20 WOG no accessory myocardial AV connections observed

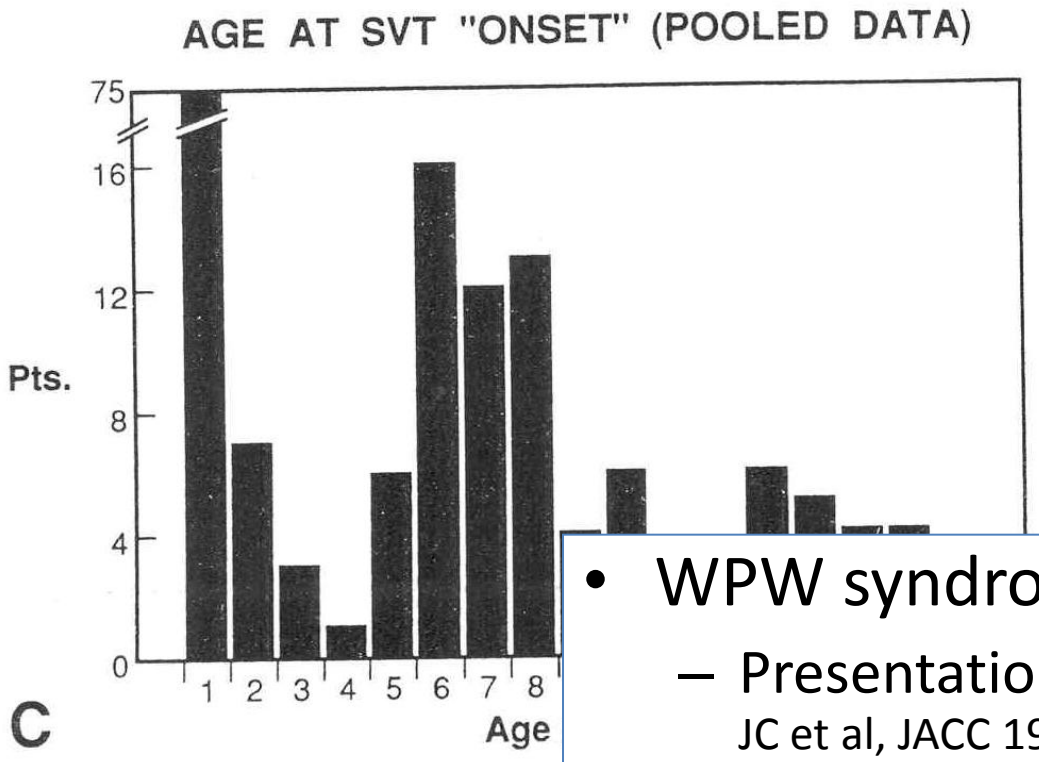


Conclusions from fetal studies

- AV ring isolation is a gradual process
  - May continue postnatally

- AV reentrant tachycardias often spontaneously obliterate before 1 year of age

Natural history in childhood



- WPW syndrome (prevalence 0.15-0.3 %)
  - Presentation with SVT <2 mo of age (Perry JC et al, JACC 1990)
    - 93 % disappear by 1 year
    - 31 % reappear at average age of 8 yrs
    - If symptomatic at >5 yrs, 78 % long-term persistance

Population-based studies

- Prevalence of supraventricular arrhythmia

2.25/1000 persons

- Annual incidence in children <19 years of age

13/100 000 persons

- In 10 583 children (www.paces.org) undergoing EPS/RFA

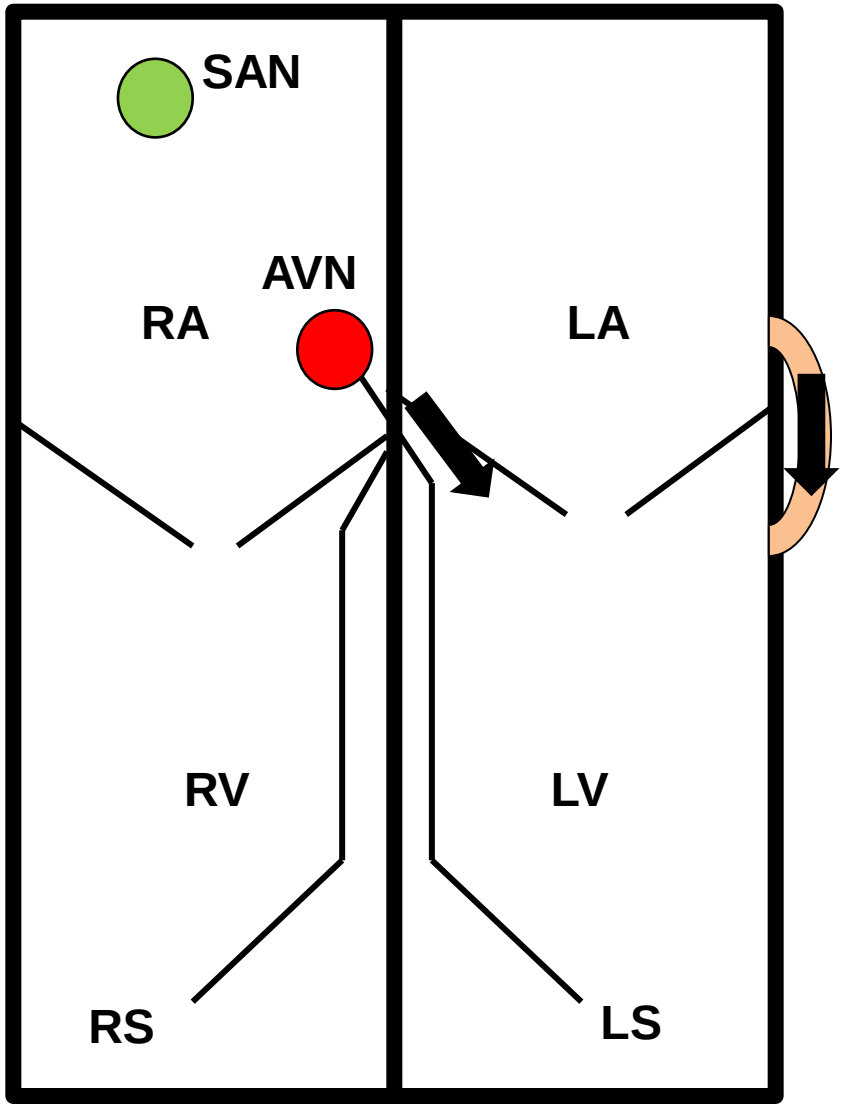
AVRT (accessory pathways) in 67%

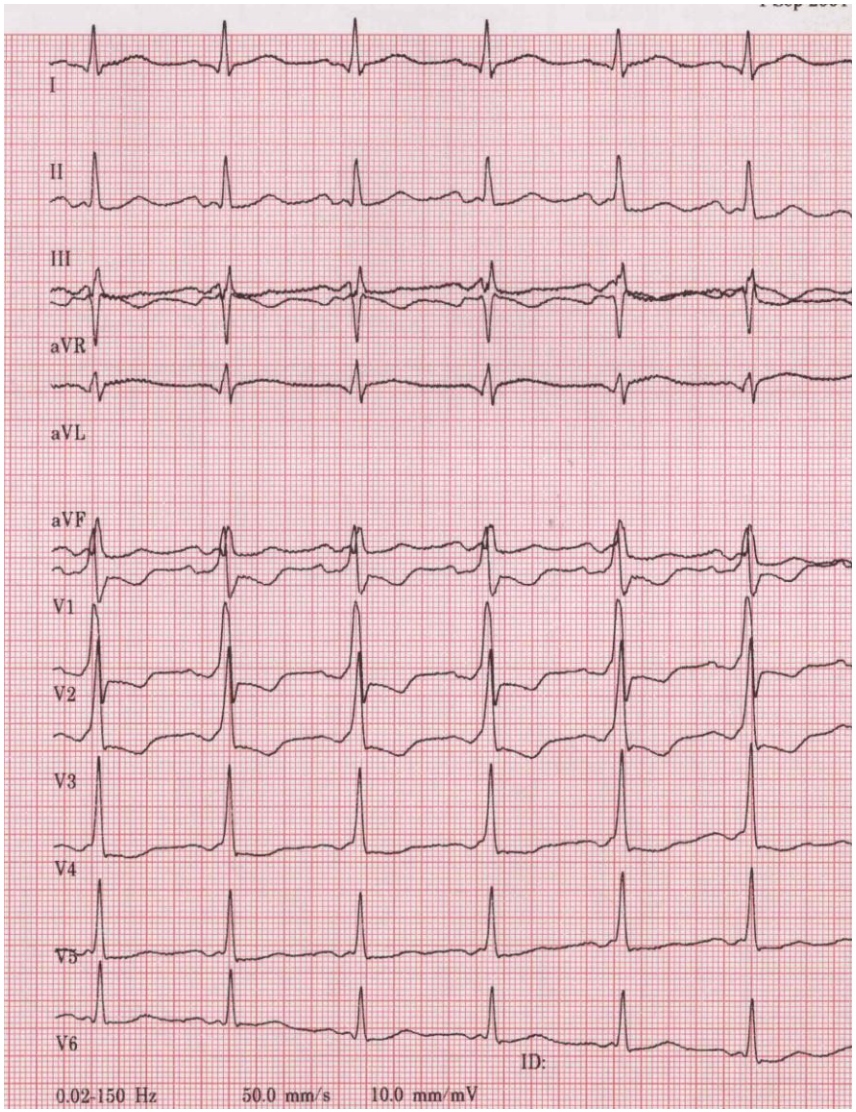
Kugler JD et al. JCE 2002
Van Hare GF et al. JCE 2004

Terminology

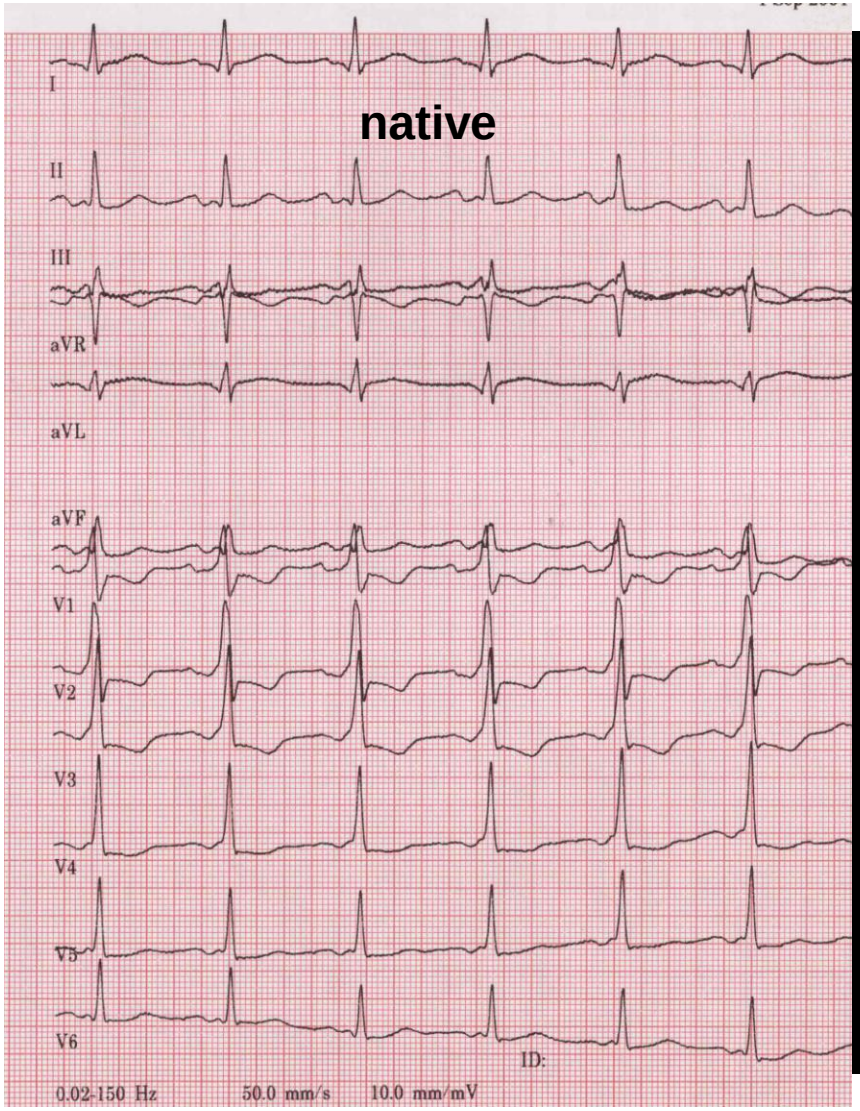
- Wolff-Parkinson-White Syndrome
  - Preexcitation in sinus rhythm
  - Supraventricular tachycardia
- Wolff-Parkinson-White Pattern
  - Asymptomatic preexcitation in sinus rhythm
  - No symptoms
- Manifest pathway
  - Antegrade +/- retrograde conduction
- Concealed pathway
  - Retrograde conduction only

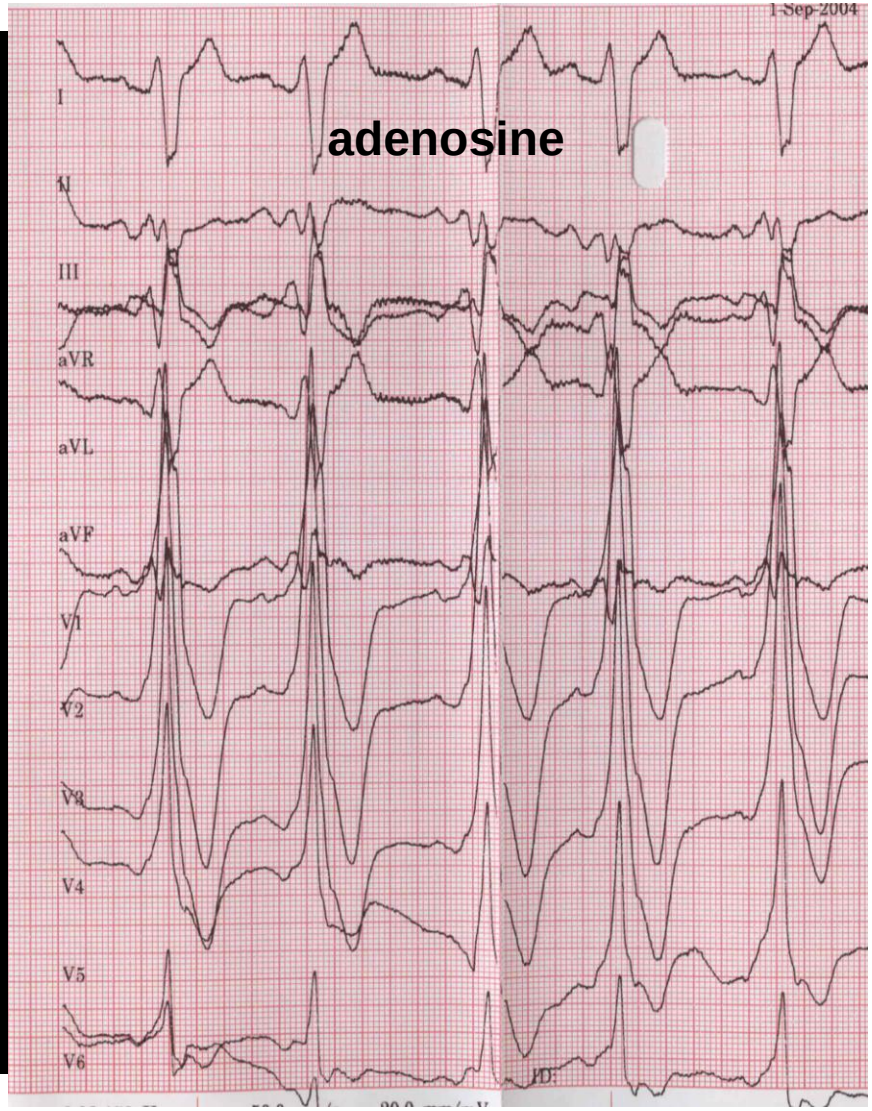
WPW electrophysiology



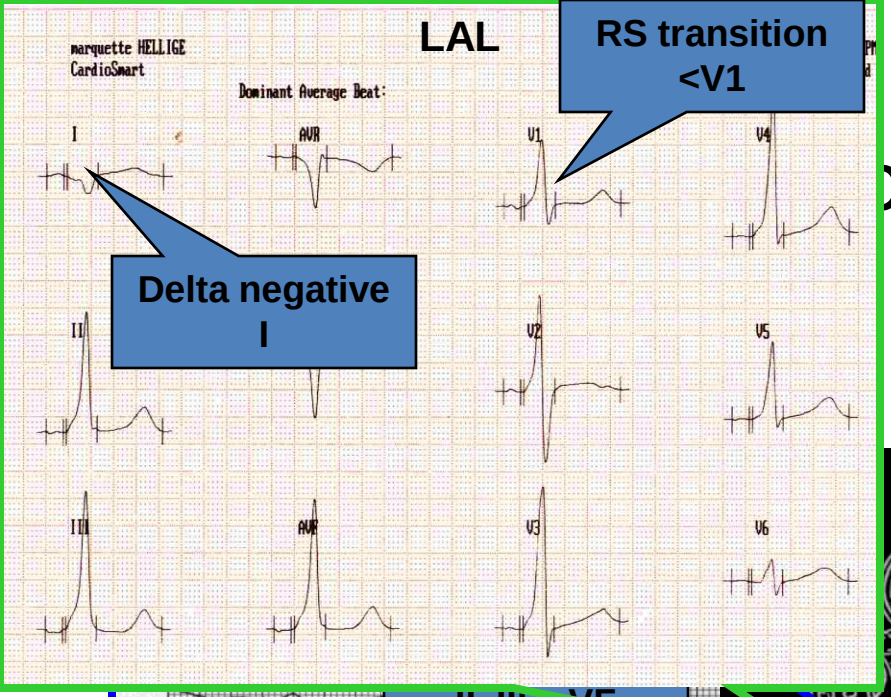


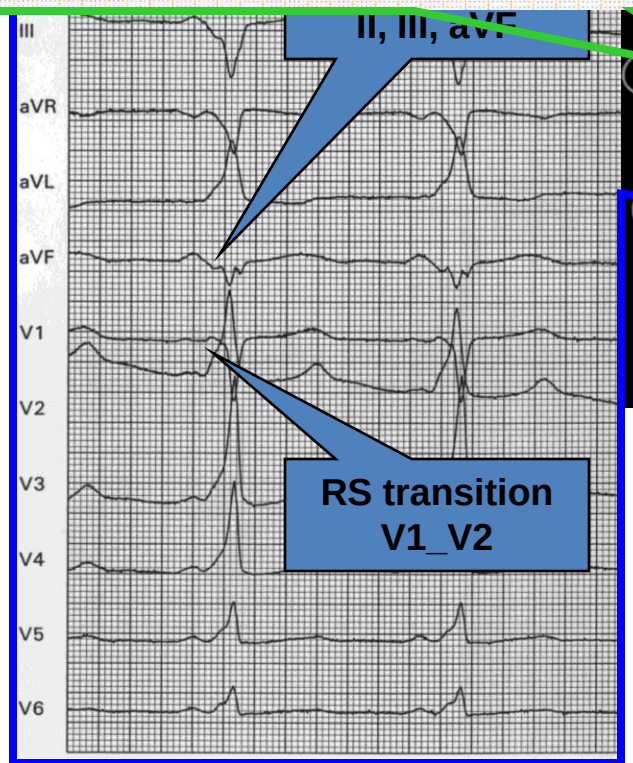
WPW electrophysiology

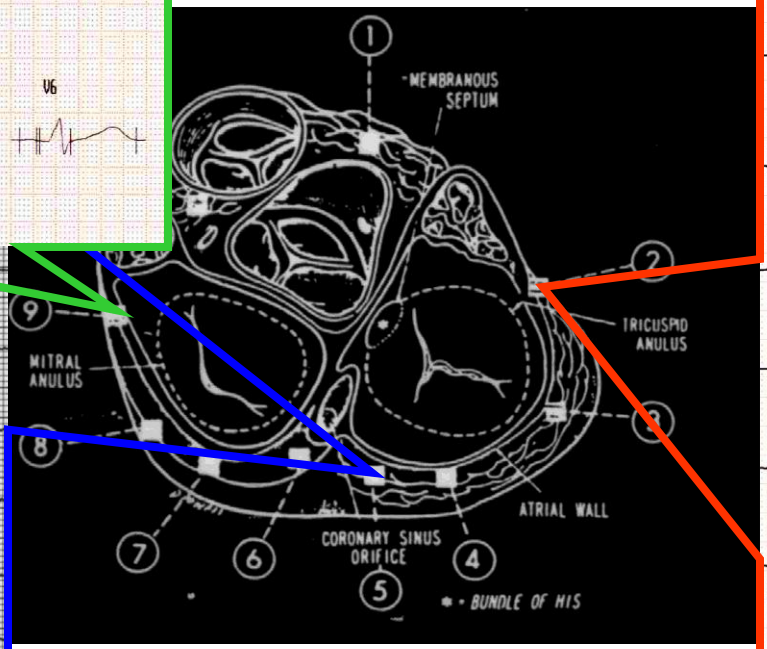


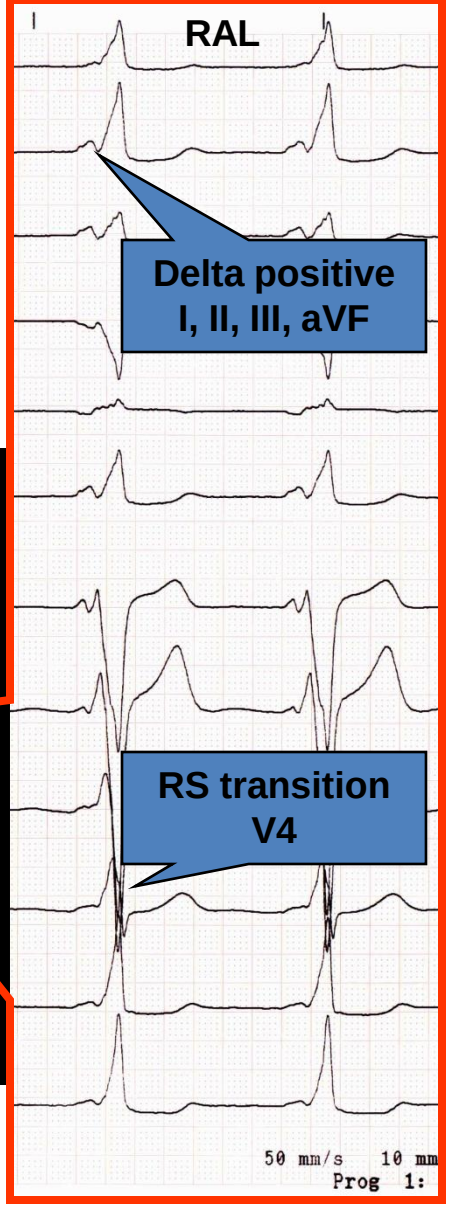


on during sinus

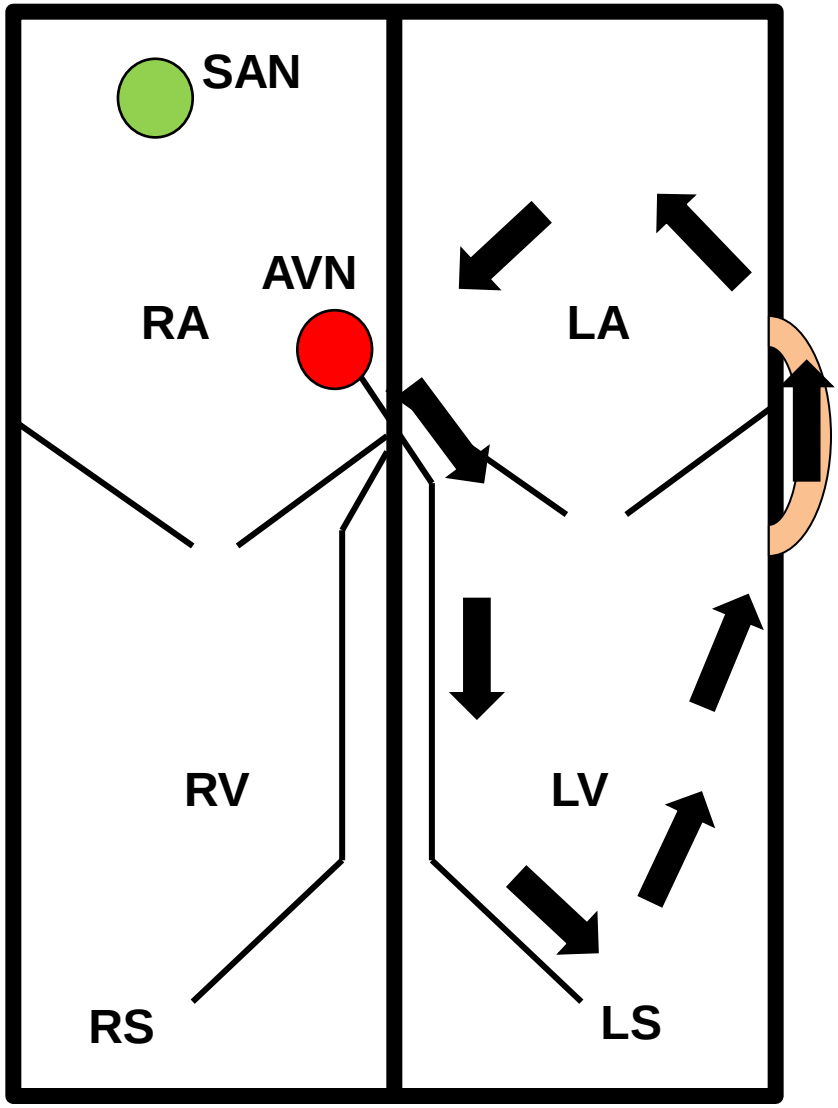


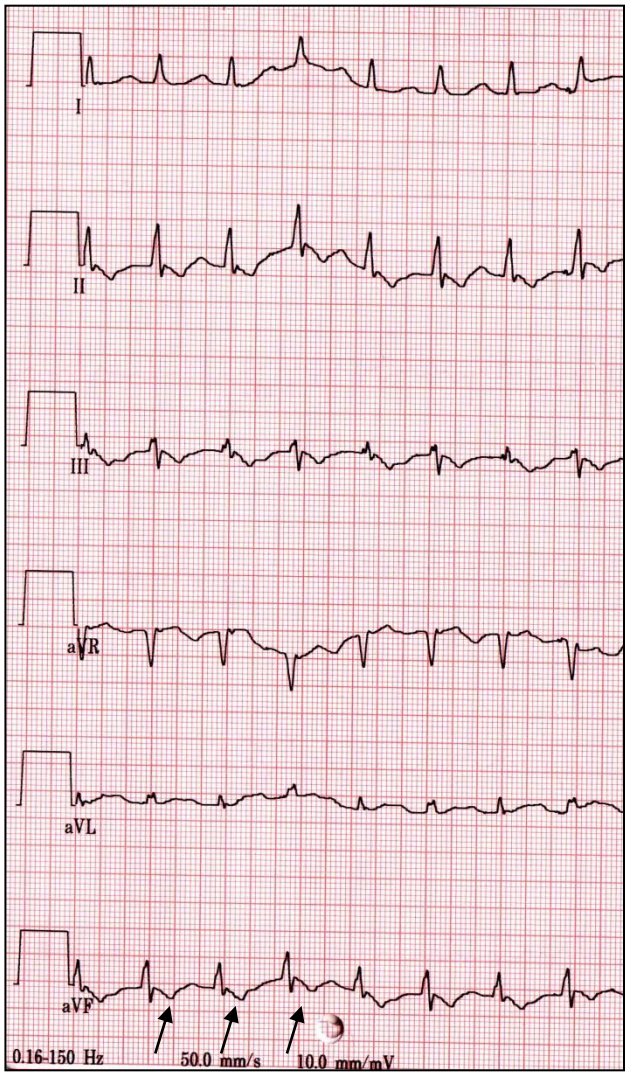




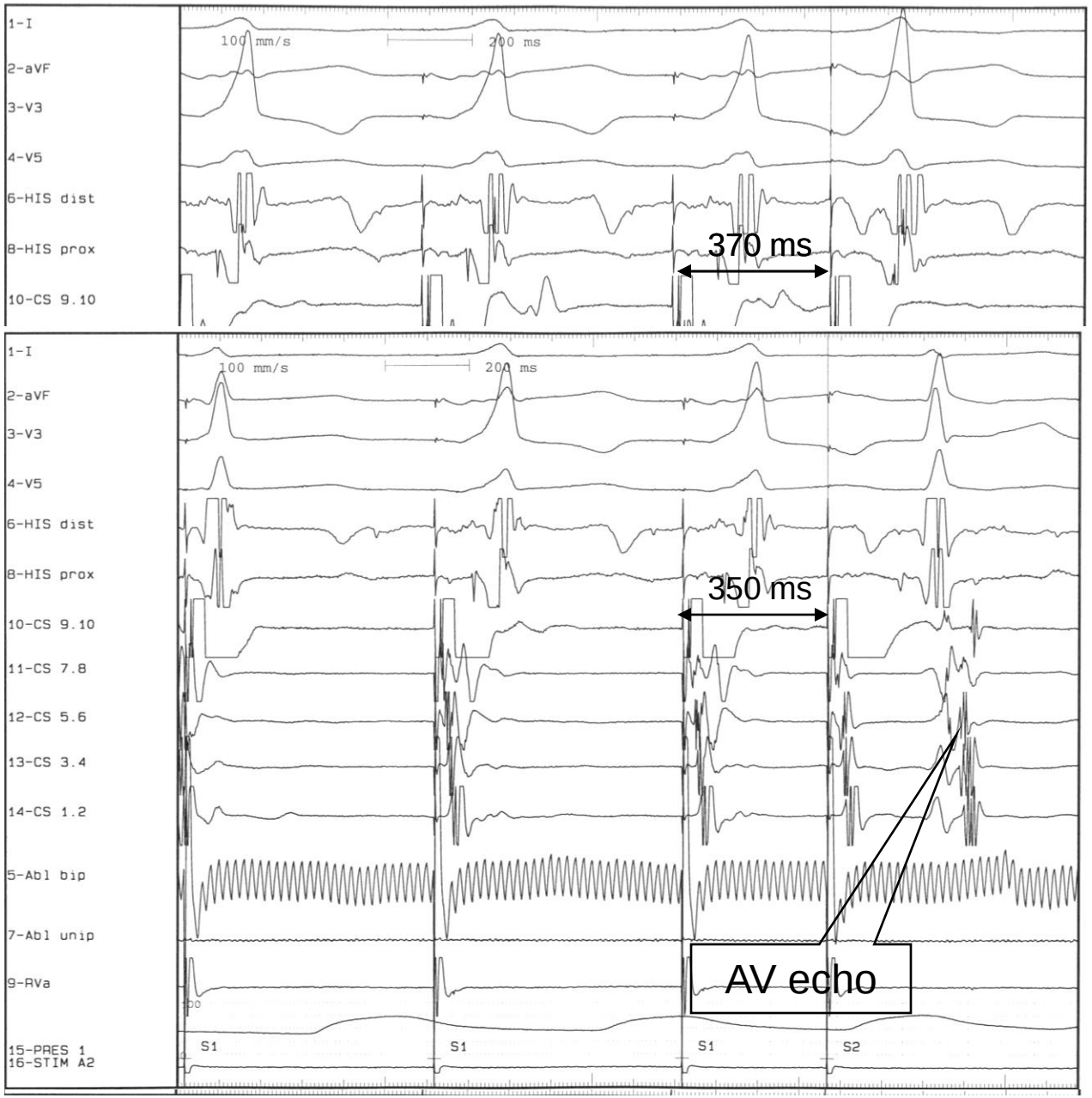


Ortodromic AV-reentrant tachycardia (AVRT)

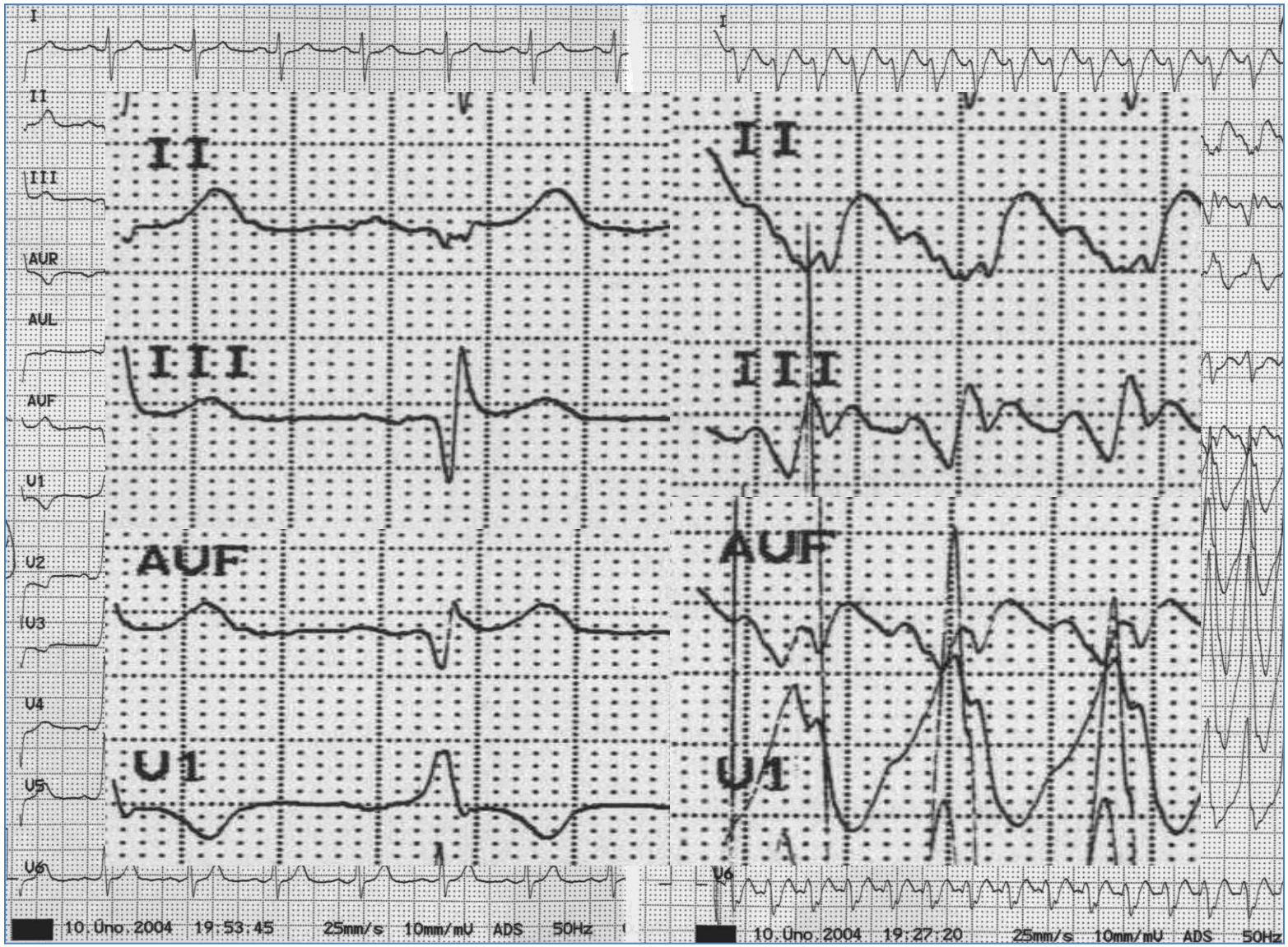




Response to premature atrial stimuli

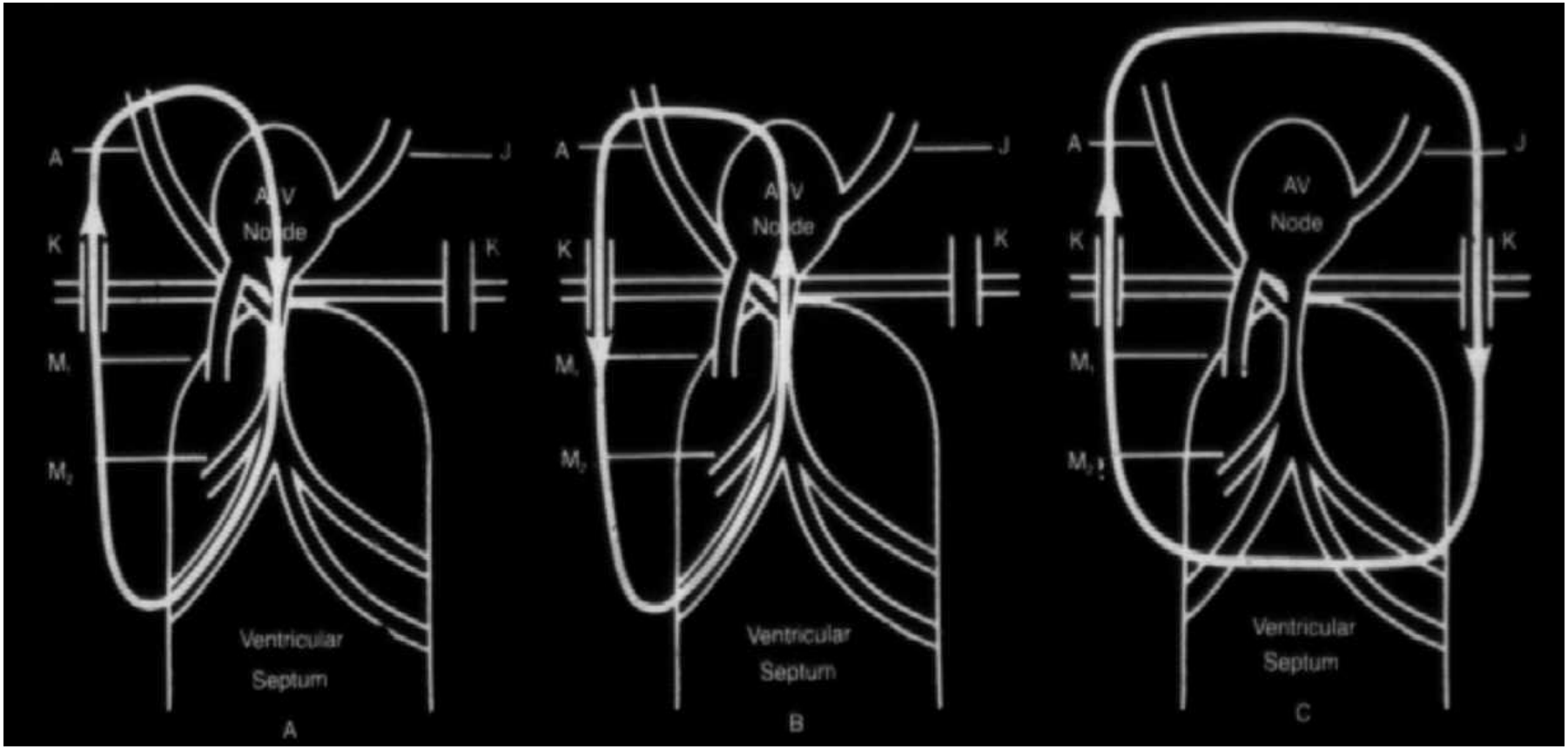


Antidromic AV-reentrant tachycardia (AVRT)



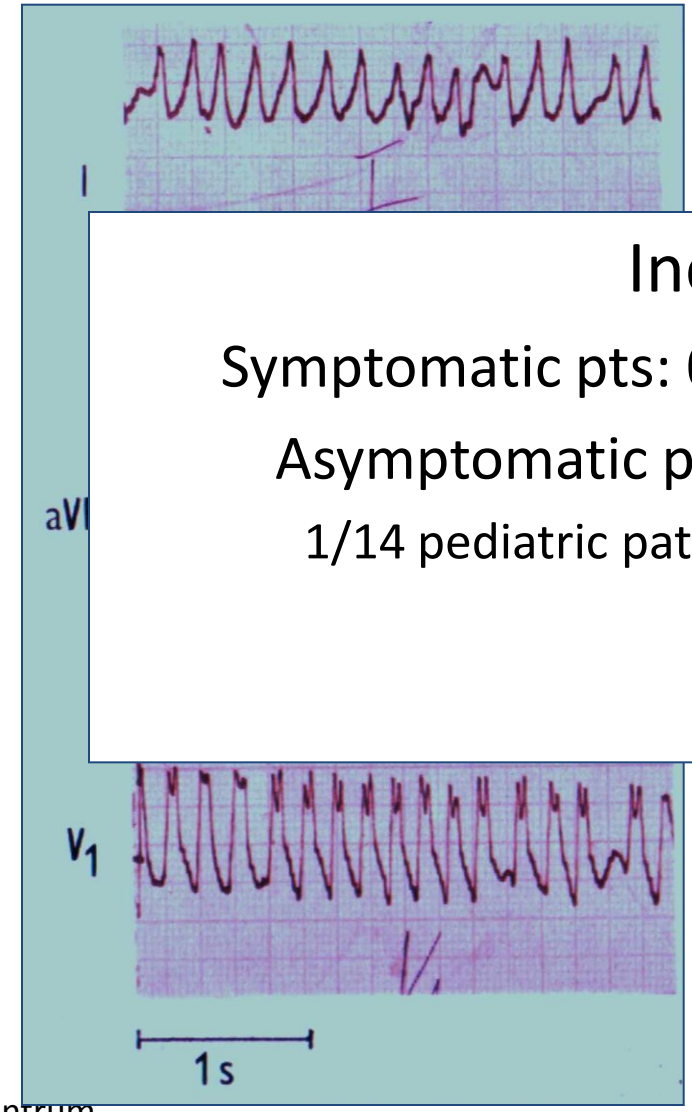
Mechanisms of pathway-mediated SVT

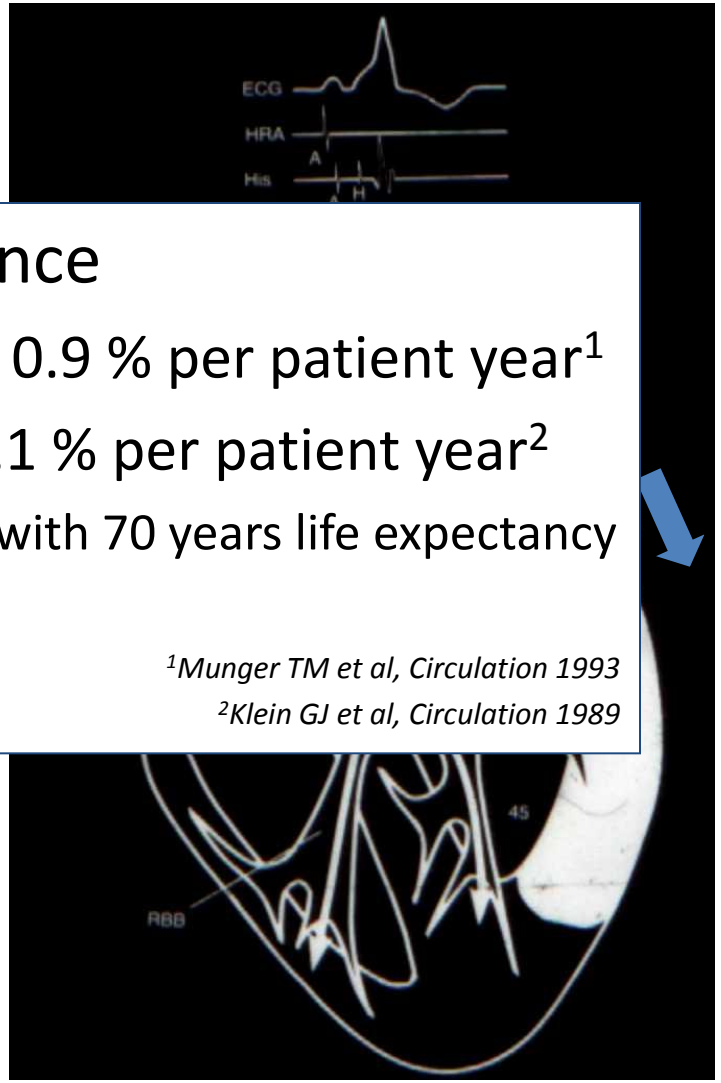
Ortodromic | Antidromic | Multiple pathways



The asymptomatic WPW pattern

Cardiac arrest in the WPW syndrome and atrial fibrillation





Incidence

Symptomatic pts: 0.3 – 0.9 % per patient year[1]

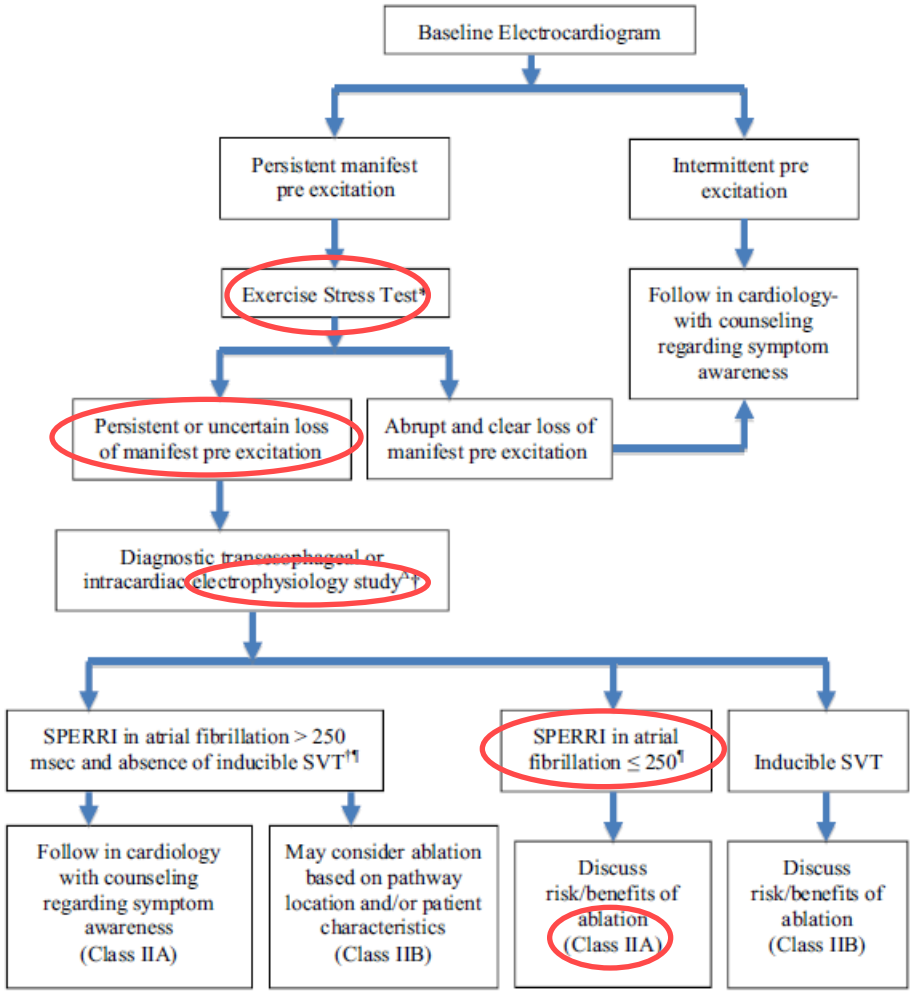
Asymptomatic pts: 0.1 % per patient year[2]

1/14 pediatric patients with 70 years life expectancy

[1]*Munger TM et al, Circulation 1993*

[2]*Klein GJ et al, Circulation 1989*

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



* patients unable to perform an exercise stress test should undergo risk-stratification with an EP study
Δ prior to invasive testing, patients and the parents/guardians should be counseled to discuss the risks and benefits of proceeding with invasive studies, risks of observation only, and risks of medication strategy.
† patients participating at moderate-high level competitive sports should be counseled with regards to risk-benefit of ablation (Class IIA) and follow the 36th Bethesda Conference Guidelines[6]
¶ in the absence of inducible atrial fibrillation, the shortest pre-excited RR interval determined by rapid atrial pacing is a reasonable surrogate

Cohen MI et al. HeartRhythm 2012

Shortest preexcited RR evaluation

Acute treatment

Hemodynamically stable regular narrow QRS tachycardia

Table I Recommendations for acute treatment of haemodynamically stable regular narrow QRS tachycardia in infants and children

Drug/intervention	Dosage (iv)	Class	Level
Vagal manoeuvres	Ice immersion, gastric tube insertion in infants,	I	B
Transoesophageal atrial overdrive pacing[a]	Valsalva, and head stand in older children	I	B
Adenosine → **WPW – atrial fibrillation!**	Rapid bolus starting dosages:		B
	For infants: 0.15 mg/kg.	I	
	For >1 year of age: 0.1 mg/kg Increasing dosage up to 0.3 mg/kg.	I	
Verapamil[b,c] **Contraindicated <1 year!!!**	0.1 mg/kg slowly over 2 min	I	B
Flecainide[b]	1.5–2 mg/kg over 5 min	IIa	B
Propafenone[b]	Loading: 2 mg/kg over 2 h Maintenance: 4–7 μg/kg/min	IIa	B
Amiodarone	Loading: 5–10 mg/kg over 60 min. Maintenance infusion:5–15 μg/kg/min	IIb	B

iv, intravenously; Class, recommendation class; Level, level of evidence.
[a]Most effective if AV reentrant tachycardias or atrial flutter.
[b]Myocardial depressant effect.
[c]Contraindicated in infants <1 year of age.

If unstable and critically ill – cardioversion!

Brugada J et al. EHRA/AEPC consensus statement. Europace 2013

Oral prophylactic treatment

Prophylactic antiarrhythmic drugs for SVT during the first 6–12 months of life[1]
Postpone ablation to >5 yrs[2]
In-hospital initiation except beta-blockers and amiodarone

Table 3 Sug…mic drugs for SVT and VT in infants and children

Drug	Total daily dosage per body weight divided in × doses	Main contraindications and precautions	Features prompting lower dose or discontinuation	AV nodal slowing
Digoxine		**Not for WPW syndrome!**	Bradycardia	Moderate
Propranolol	1–3 mg/kg in 3× daily	Asthma bronchiale	Bradycardia	Moderate
Atenolol	0.3–1.3 mg/kg in 1× daily	Asthma bronchiale	Bradycardia	Moderate
Verapamil	4–8 mg/kg in 3× daily	Myocardial depressant effect **Contraindicated <1 year!!!**	Bradycardia	Marked
Flecainide	2–7 mg/kg in 2× daily	Contraindicated if creatinine clearance <50 mg/mL or …F. Caution if conduction system disease. **Not in structural heart disease!**	QRS duration increase >25% above baseline	None
Propafenone	200–600 mg/m² or 10–15 mg/kg in 3× daily	Contraindicated if reduced LVEF. Caution if conduction system disease and renal impairment.	QRS duration increase >25% above baseline	Slight
Sotalol	2–8 mg/kg in 2× daily **3 x daily in infants!**	Contraindicated if significant LV hypertrophy, systolic HF, pre-existing QT prolongation, hypokalaemia, creatinine clearance <50 mg/mL and asthma bronchiale. Moderate renal dysfunction requires careful adaptation of dose	QT interval >500 ms	Similar to high-dose beta-blockers
Amiodarone	Loading: 10 mg/kg for 10 days. Maintenance: 5 mg/kg in 1× daily	Caution when using concomitant therapy with QT-prolonging drugs, HF. Dose of vitamin K antagonists and of digitoxin/digoxin should be reduced.	QT interval >500 ms	Slight

LVEF, left ventricular ejection fraction; HF, heart failure.

[1]*Lemler MS et al. AHJ 1997*
[2]*Friedman RA, PACE 2002*
Brugada J et al. EHRA/AEPC consensus statement. Europace 2013

Catheter ablation

AVNRT

Chronic Therapy

Catheter ablation for slow pathway modification is recommended in symptomatic patients or in patients with an ICD. 243–247

AVRT

Chronic therapy

Catheter ablation of the accessory pathway is recommended in patients with symptomatic AVRT and/or pre-excited AF.[a] 320–322

Catheter ablation of concealed accessory pathways may be considered in symptomatic patients with frequent episodes of AVRT 244–247

AFl

Chronic therapy

One-time or repeated cardioversion associated with AAD are recommended as a long-term alternative for patients with infrequent AFL recurrences or refusing ablation. 152,153

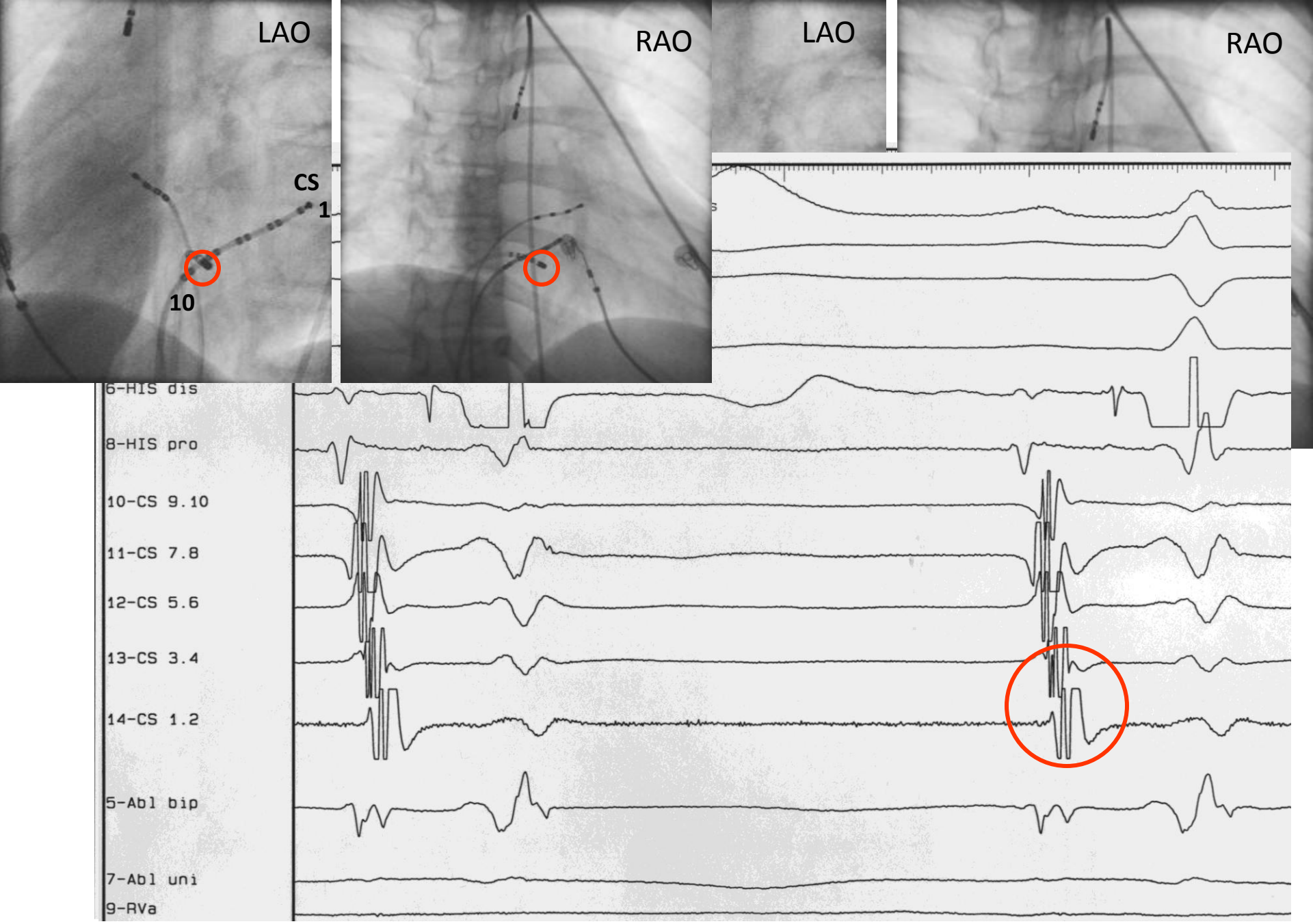
In patients with recurrent or poorly tolerated typical AFL, CTI ablation is recommended for preventing recurrences with a low incidence of complications. 153,154

In patients with depressed LV systolic function, ablation may be considered to revert dysfunction due to tachycardiomyopathy, and prevent recurrences. 155,156

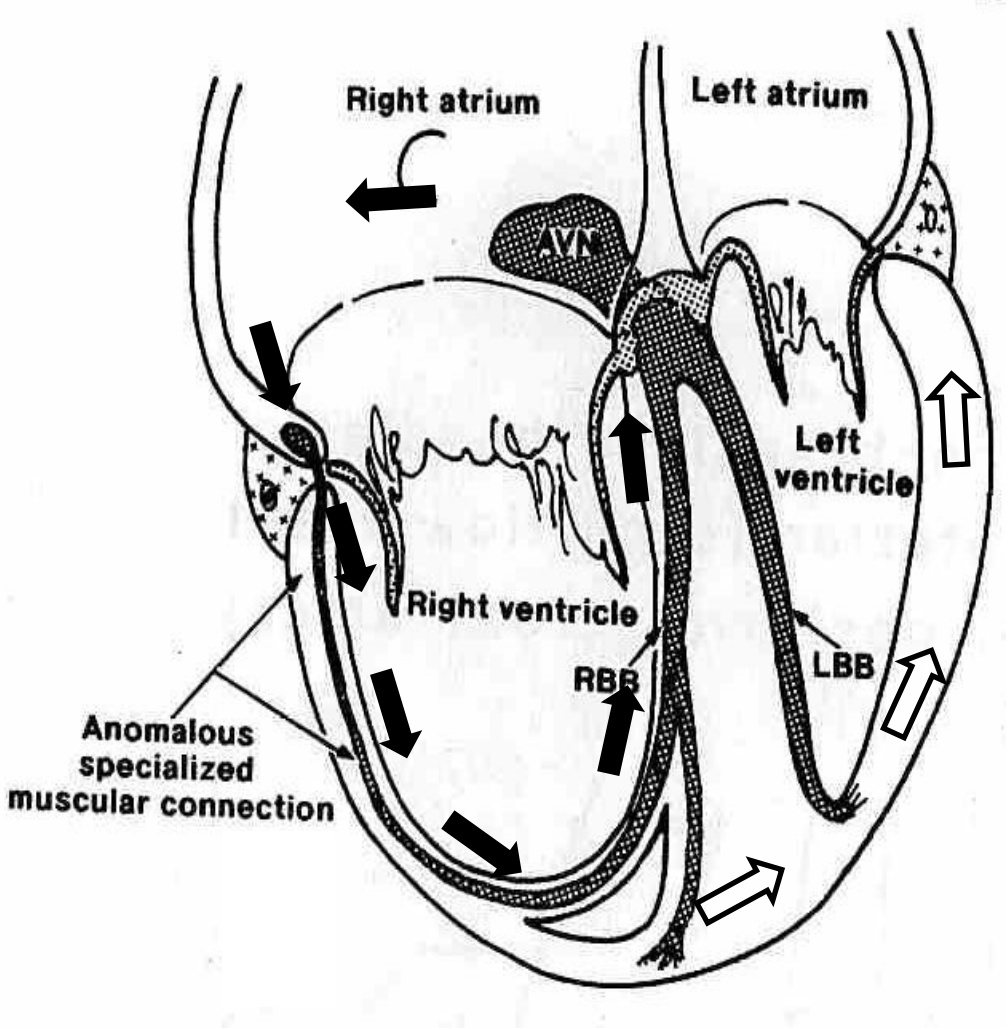
Asymptomatická preexcitace

Table 17 Management of asymptomatic pre-excitation

Recommendation	Reference
Electrophysiologic testing may be considered for risk stratification in subjects with asymptomatic ventricular pre-excitation.	322,339–345
Catheter ablation of accessory pathways may be considered in asymptomatic patients with accessory pathways with antegrade refractory period <240 ms, inducible atrioventricular reentrant tachycardia triggering pre-excited atrial fibrillation, and multiple accessory pathways.[a]	322,343,345
Observation without treatment may be reasonable in asymptomatic WPW patients who are considered to be at low risk following electrophysiology study or due to intermittent pre-excitation.	322,341
Screening programmes may be considered for risk stratification of asymptomatic subjects with pre-excited ECG.	322,341

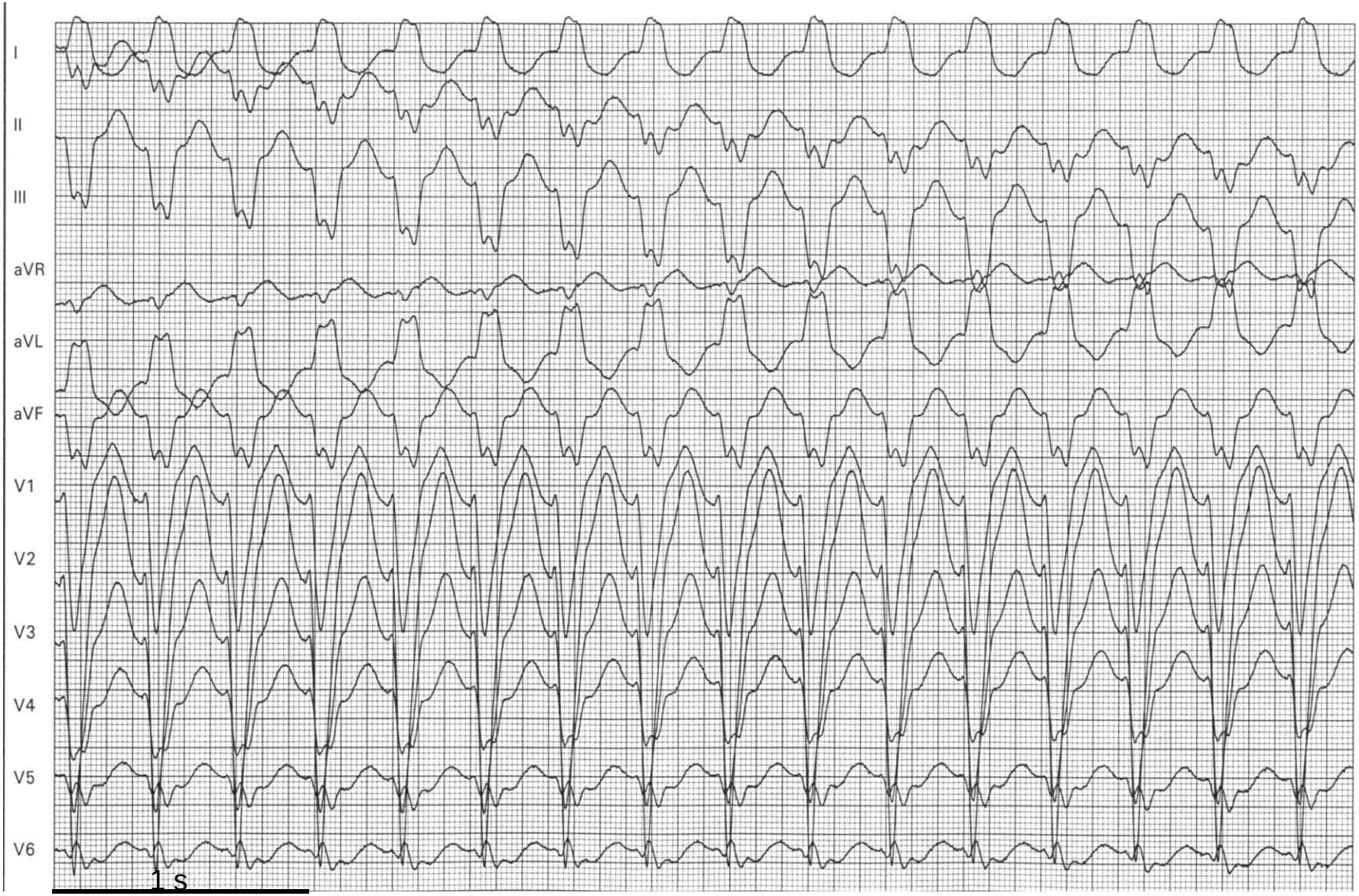

LAO
RAO
LAO
RAO
CS
1
10
6-HIS dis
8-HIS pro
10-CS 9.10
11-CS 7.8
12-CS 5.6
13-CS 3.4
14-CS 1.2
5-Abl bip
7-Abl uni
9-RVa

Atriofascicular connection (“Mahaim”)



- AV node and His bundle like structure
- Lateral tricuspid annulus
- Connected to distal right bundle
- Only antegrade conduction
- Decremental properties

Mahaim-mediated SVT



Nodo-/fasciculoventricular pathways