Arrhythmia recognition and treatment

South Wales and South West Congenital Heart Disease Network

Contents

- Bradyarrhythmias
- Tachyarrhythmias
- Channelopathies/Cardiomyopathies

Bradyarrhythmias

- Neonates
- Infants/Adolescents
- Pacing

Neonatal bradyarrhythmias

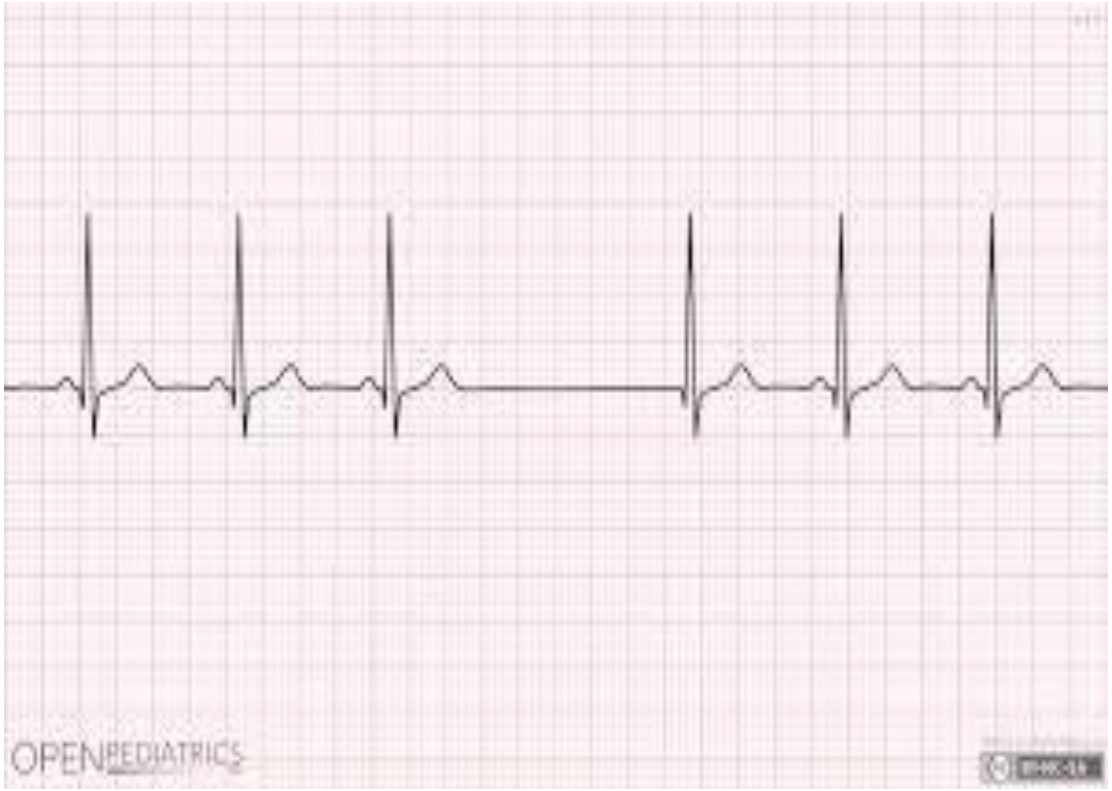
Age	Heart rate 2nd to 98th percentile in bpm (mean)
0-1 days	93-154 (123)
1-3 days	91-159 (123)
3-7 days	90-166 (129)
7-30 days	107-182 (140)
1-3 months	121-179 (150)

Neonatal bradyarrhythmias

Sinus bradycardia:

- Hypoxia
- Acidosis
- Infection / sepsis
- Electrolyte abnormalities
- Neonatal hypothyroidism
- Increased intracranial pressure
- Hypervagal states- e.g. high position of NG tube, Gastro oesophageal reflux disease
- Obstructive jaundice

Neonatal bradyarrhythmias



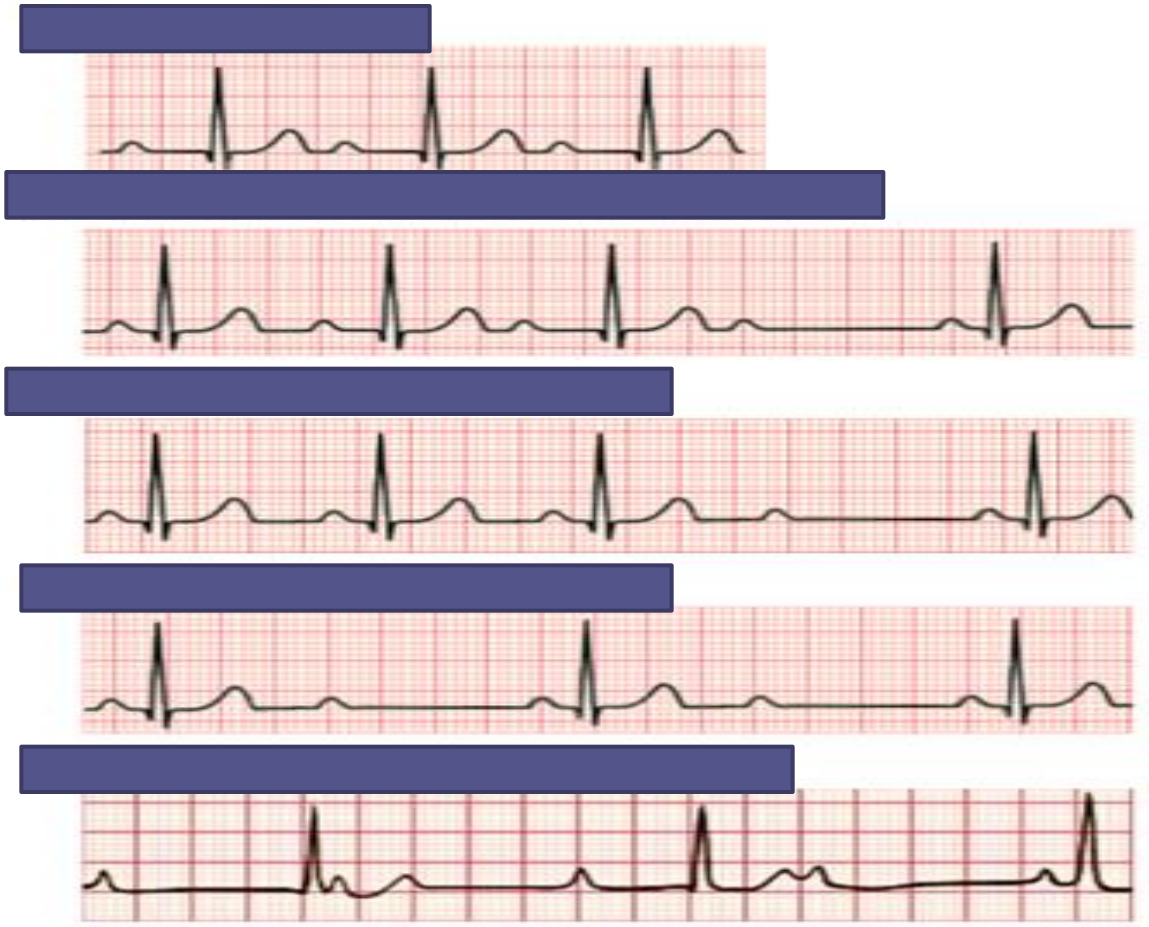
Sinus node dysfunction

- Common in ccTGA
- Left atrial isomersim
- Post surgical/intervention
- Long QT
- Drug overdose

Treatment

- Isoprenaline
- Atropine
- Usually self reverting
- Not as significant as AV block

Neonatal bradyarrhythmias



AV block

- Congenital
- Maternal CTD
- Congenital syphilis
- Post surgical
- Long QT syndrome
- Drug overdose

Treatment

- Atropine/isoprenaline
- Pacing if needed

Infant/Adolescent bradyarrhythmias

- Cardiomyopathies/channelopthies become more a problem
- Patients more likely to have multiple congenital interventions

Pacing

Disorders of atrioventricular conduction
Complete congenital atrioventricular block
Class I
(1) Complete congenital atrioventricular block in a newborn or an infant with a ventricular rate ,55 b.p.m. or with CHD and a ventricular rate ,70 b.p.m. (C)
(2) Complete congenital atrioventricular block with a wide complex escape rhythm, complex ventricular ectopy, or ventricular dysfunction. (B)
(3) Complete congenital atrioventricular block beyond first year of life with an average heart rate ,50 bpm, abrupt pauses in ventricular rate 2–3× basic cycle length, or associated with symptoms of chronotropic incompetence. (B)
Class II
(1) Complete congenital atrioventricular block in asymptomatic children and adolescents with an acceptable rate, a narrow QRS complex and normal ventricular function. (C)
Other non-surgical atrioventricular block
Class I
(1) Advanced second- or third-degree AV block associated with symptomatic bradycardia, ventricular dysfunction, or low cardiac output. (C)
Post-operative atrioventricular block
Class I
1. Post-operative advanced second- or third-degree AV block not expected to resolve or persisting at least 7 days after cardiac surgery. (B)
Class IIb
1. Transient post-operative third-degree AV block with residual bifascicular block. (C)

Sinus node dysfunction
Class I
(1) Sinus node dysfunction with correlation of symptoms during age-inappropriate bradycardia. (B)
Class IIa
1. Asymptomatic sinus bradycardia in children and CHD with resting rate ,40 b.p.m. or pauses in ventricular rate .3 s. (C)
2. Sinus node dysfunction with intra-atrial reentrant tachycardia with the need for antiarrhythmics when other therapeutic options, such as catheter ablation, are not possible. (C)
3. Congenital heart disease and impaired haemodynamics due to sinus bradycardia or loss of AV synchrony. (C)
Class IIb
(1) Asymptomatic sinus bradycardia in the adolescent with CHD with resting rate ,40 bpm or pauses in ventricular rate .3 s. (C)
Other indications. Neuromuscular disease associated with AV conduction disease [e.g. myotonic muscular dystrophy, Kearns–Sayre syndrome, Erb dystrophy (limb girdle), peroneal muscular atrophy etc].
Class I
(1) Third-degree or advanced second-degree AV block with or without symptoms. (B)
Class IIb
(1) Any degree of AV block, because the progression of the conduction disease may be unpredictable. (B)
Neurocardiogenic syncope
Class IIb
(1) Significantly symptomatic patients in who prolonged asystole can be demonstrated spontaneously or at tilt-table testing. (C)

Pacing

Table 5 The consensus panel recommendations on preferred pacemaker implantation access, pacing modes, and ventricular lead placement in pediatric patients with AV block, systemic LV, and absence of intracardiac shunts

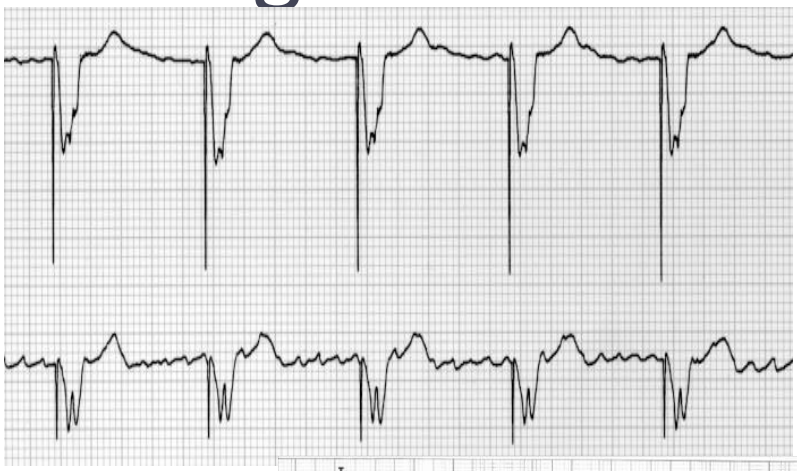
Patient size (kg)	Access	Pacing mode	Ventricular lead placement
<10	Epicardial Endocardial—in specific situations (failed epicardial, centre preference)	VVIR DDD(R)—in case of a specific haemodynamic indication	LV apex RV septum
10–20	Epicardial Endocardial	VVIR DDD(R) – in case of a specific haemodynamic indication	LV apex RV septum
>20	Endocardial Epicardial—in specific situations (e.g. concomitant with other cardiac surgery)	DDD(R) VVIR	RV septum LV apex or free wall—based on surgical feasibility

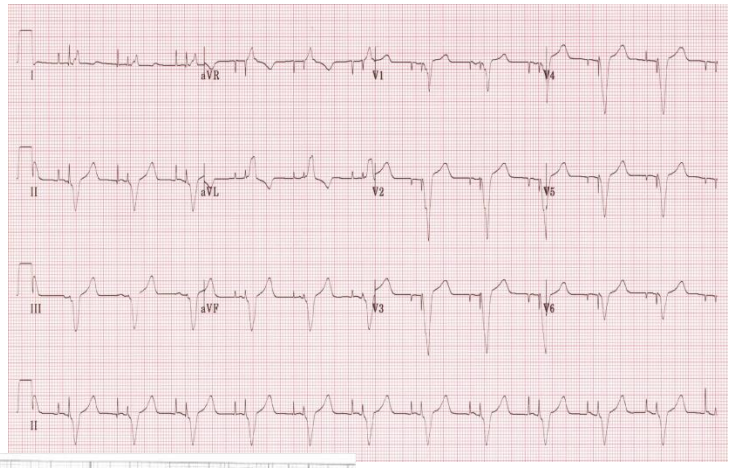
AV, atrioventricular; LV, left ventricle; RV, right ventricle

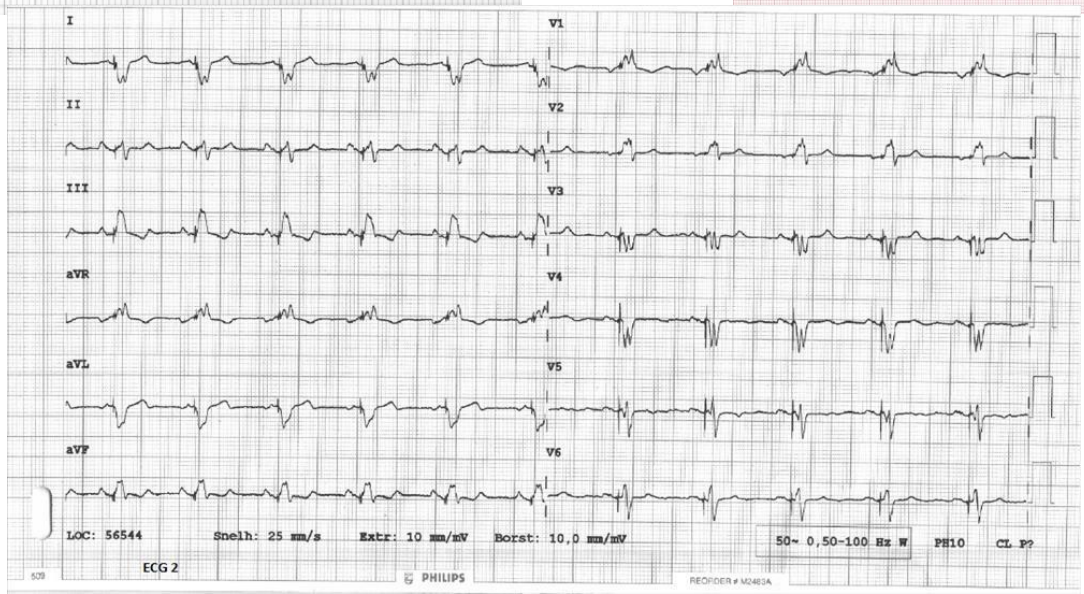
Pacing

- Recognising a problem
- Need to know what system/to which structure
- Remember the pacemaker- is a computer with a massive storage and diagnostic system

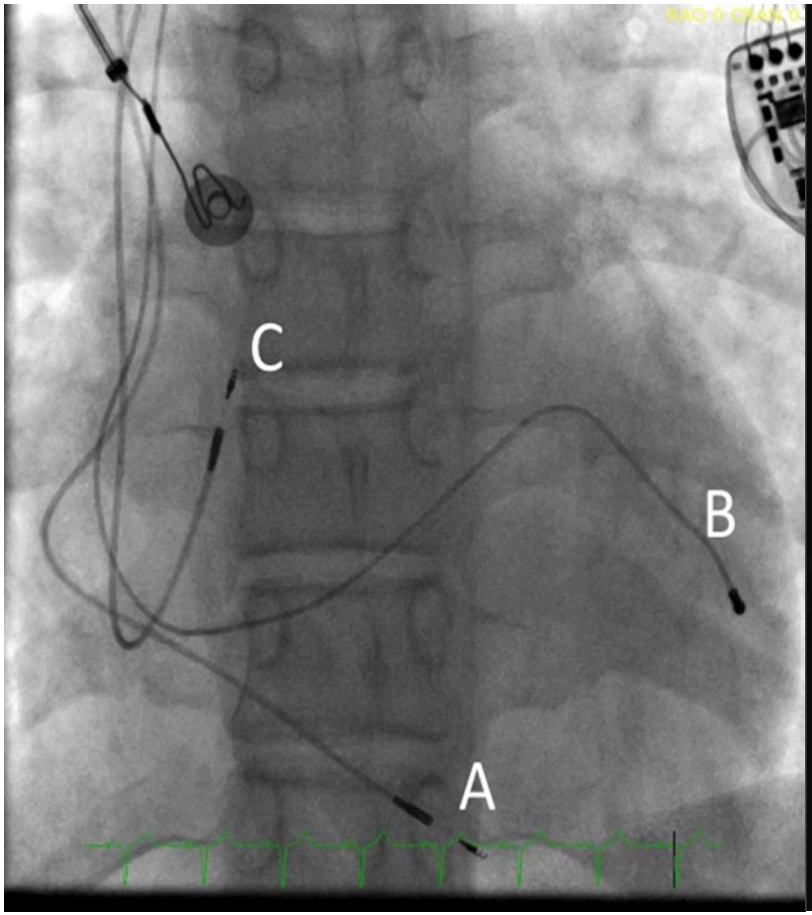
Pacing

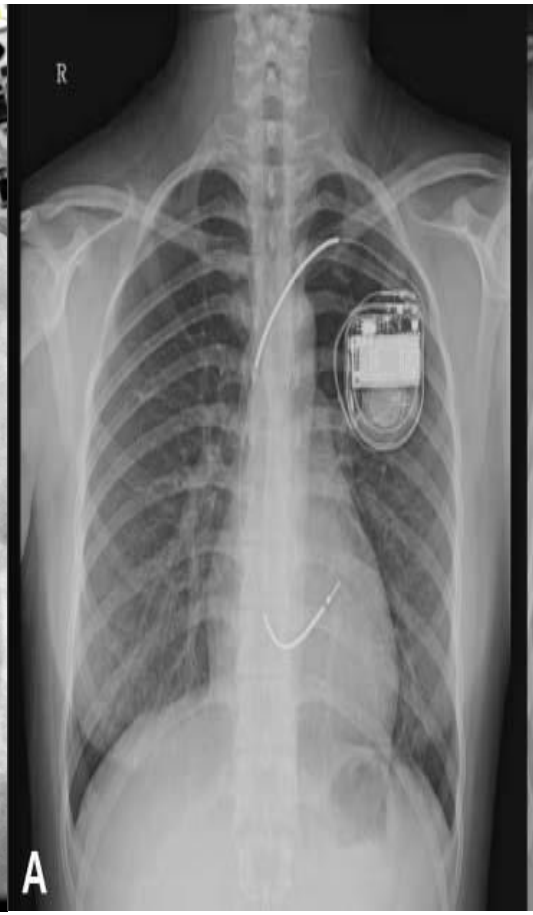


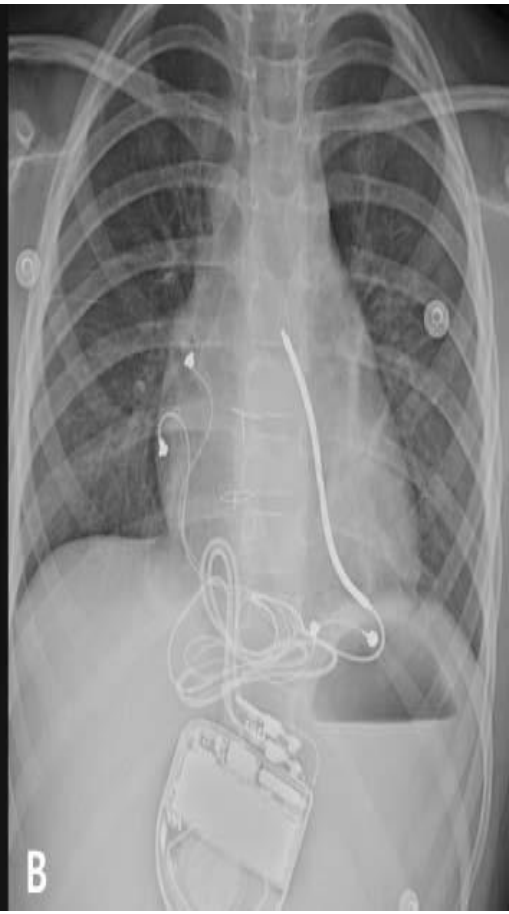

I
aVR
V1
V4
II
aVL
V2
V5
III
aVF
V3
V6
II


I
II
III
aVR
aVL
aVF
V1
V2
V3
V4
V5
V6
LOC: 56544
Snelh: 25 mm/s
Extr: 10 mm/mV
Borst: 10,0 mm/mV
50~ 0,50-100 Hz W
PH10
CL P?
ECG 2
PHILIPS

Pacing

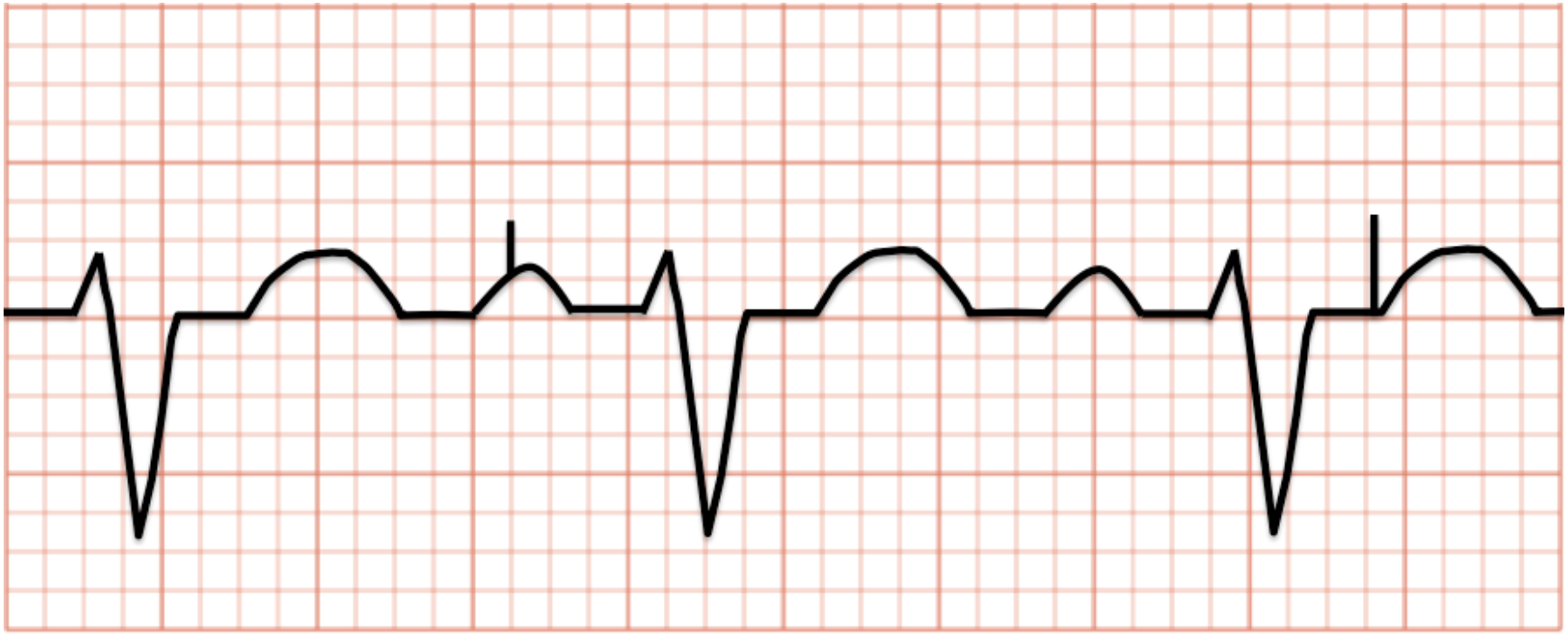




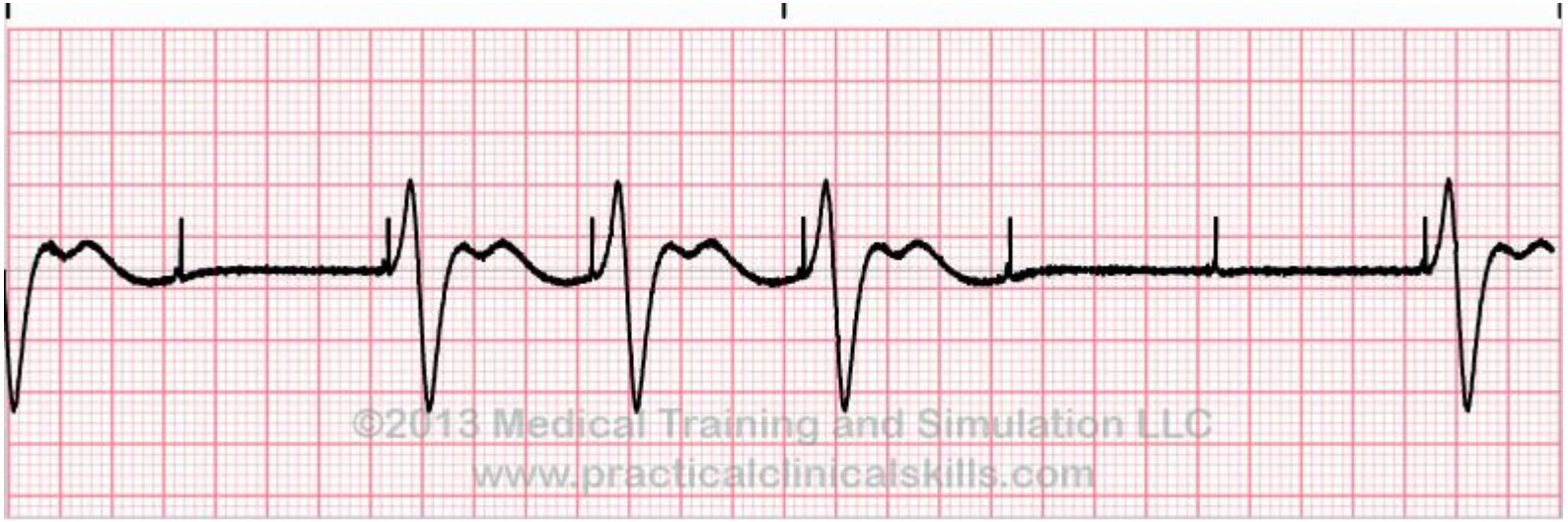


Pacing

Failure to sense in the A
Can reduce the sensitivity- help the PPM see the P waves
Lead may have displaced slightly

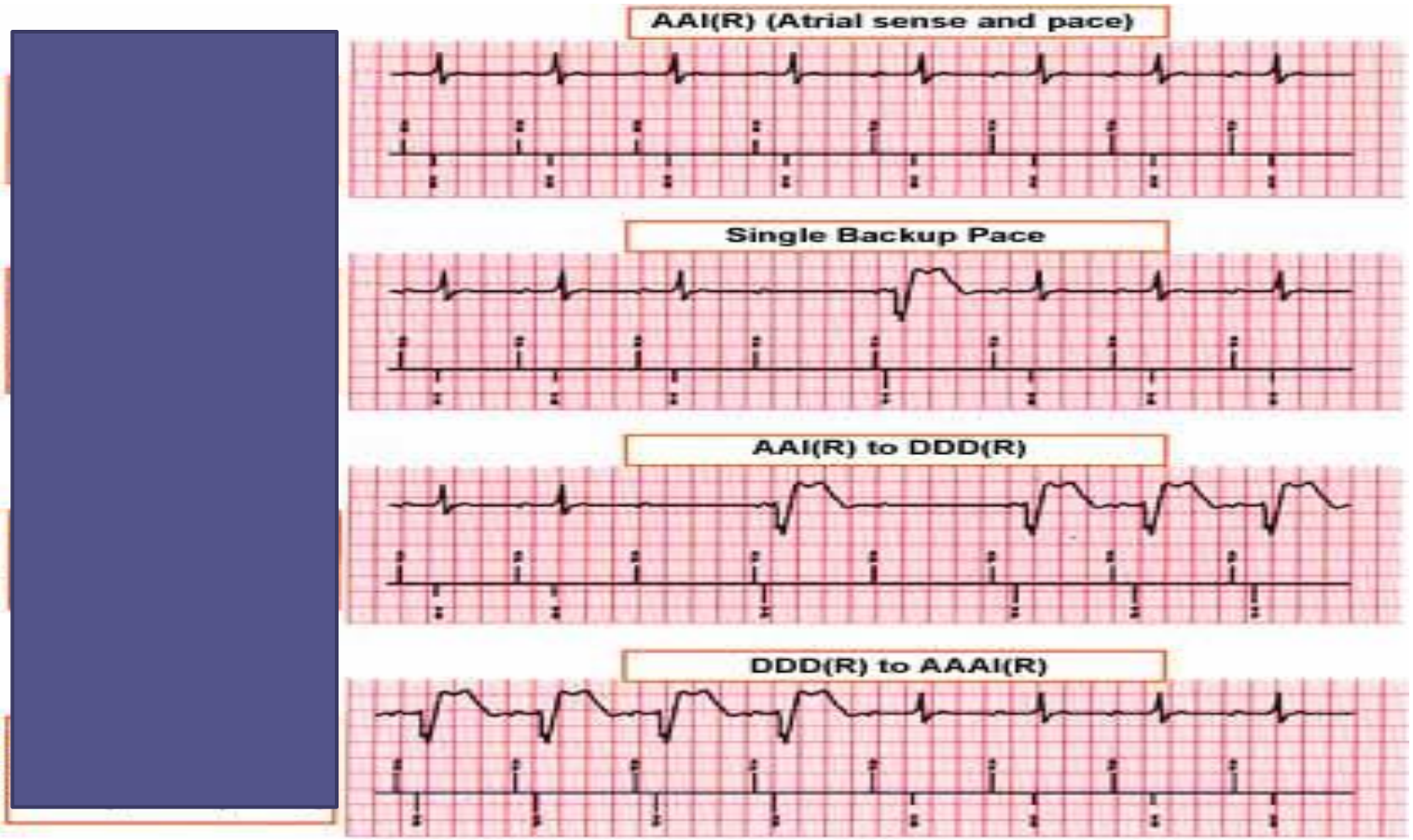


Pacing



Failure to capture
Can increase output
If no success will need new lead/repositioning

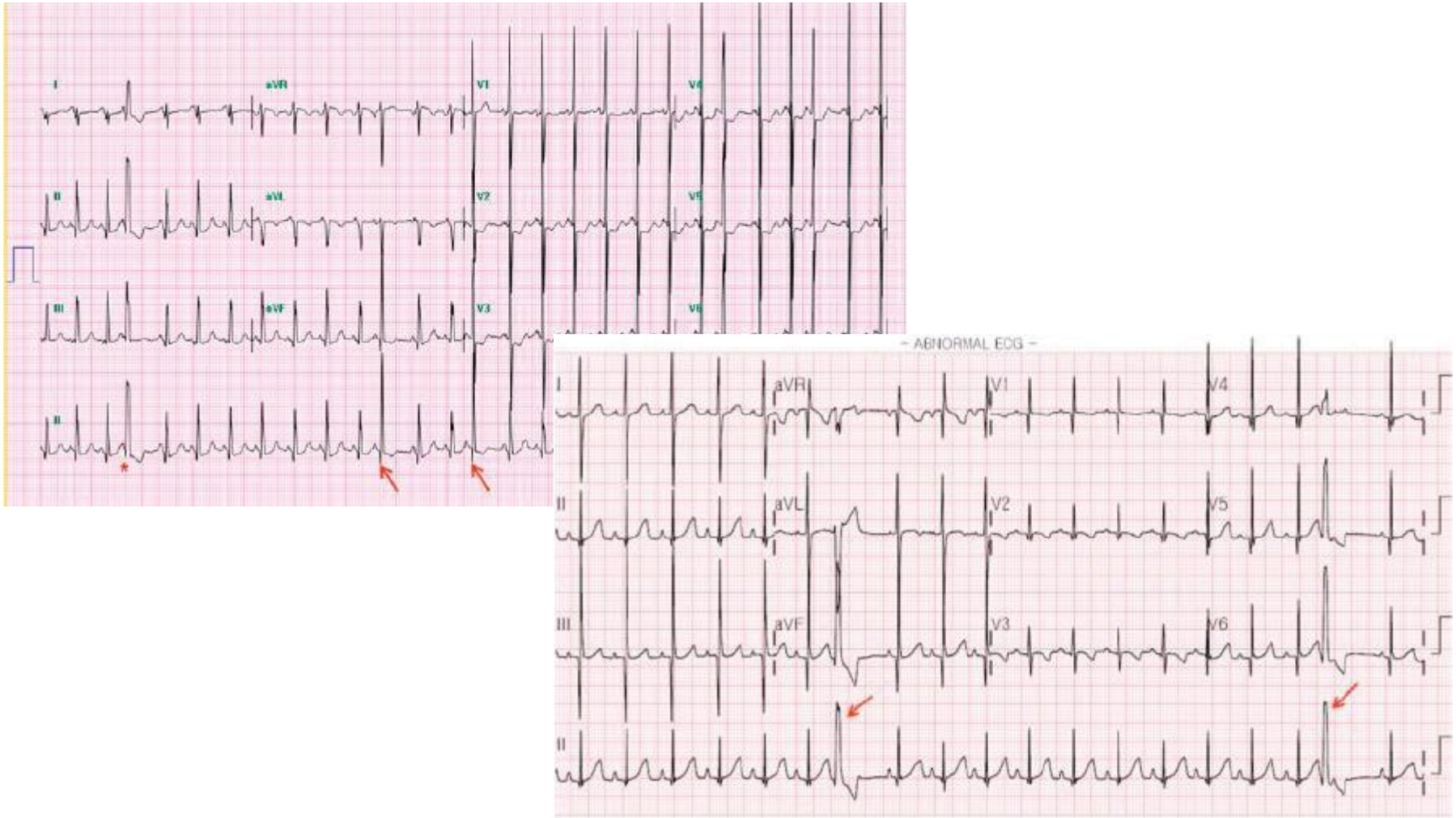
Pacing


AAI(R) (Atrial sense and pace)
Single Backup Pace
AAI(R) to DDD(R)
DDD(R) to AAAI(R)

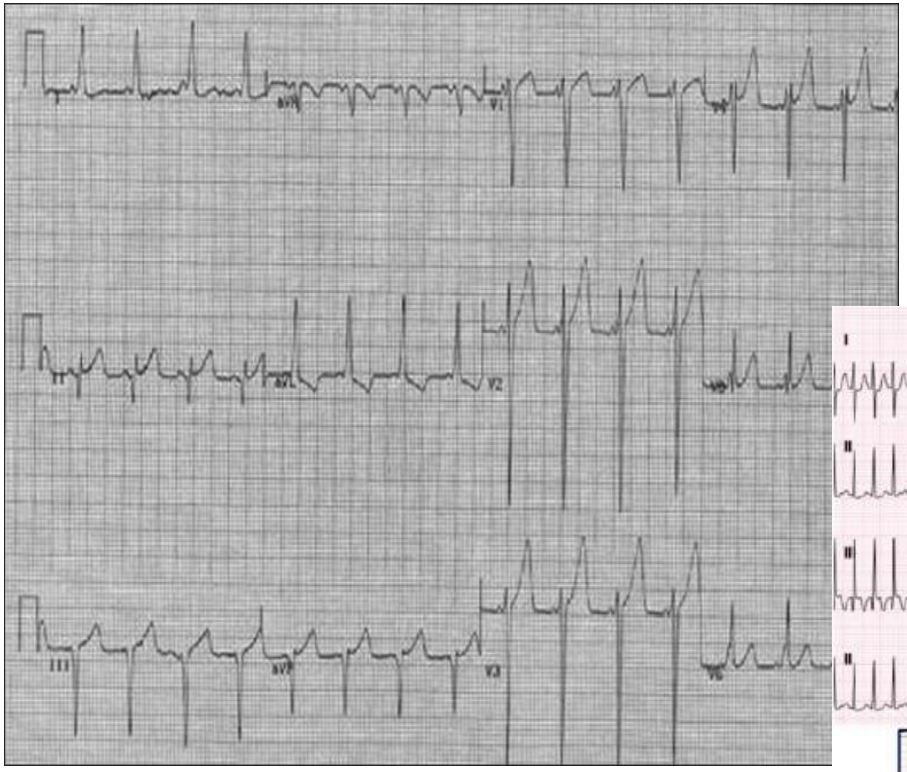
Tachyarrhythmias

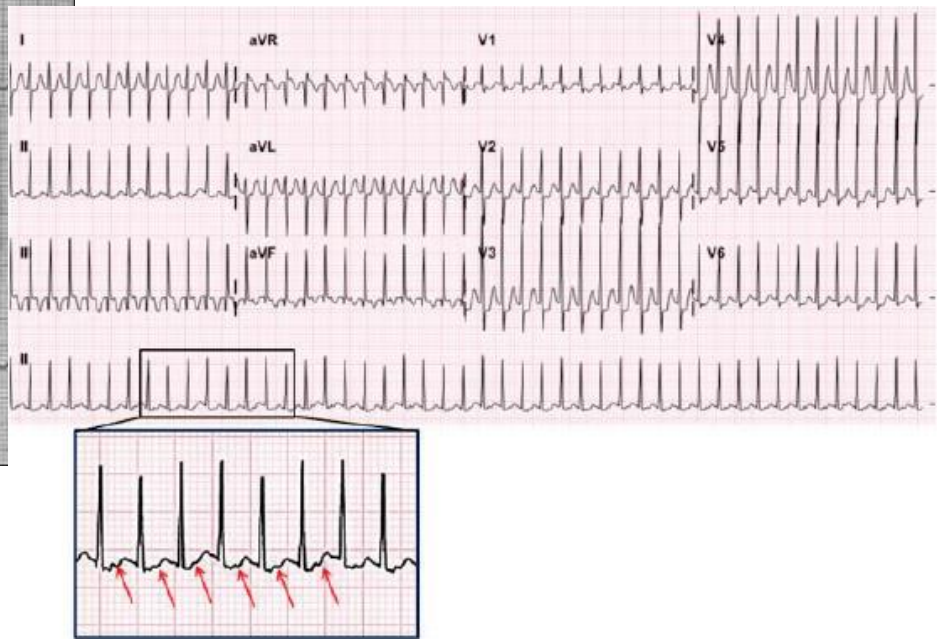
- AVRT
- PJRT
- JET
- Flutter
- Atrial tachycardia
- AVNRT
- VT/ectopy

Tachyarrhythmias



Tachyarrhythmias

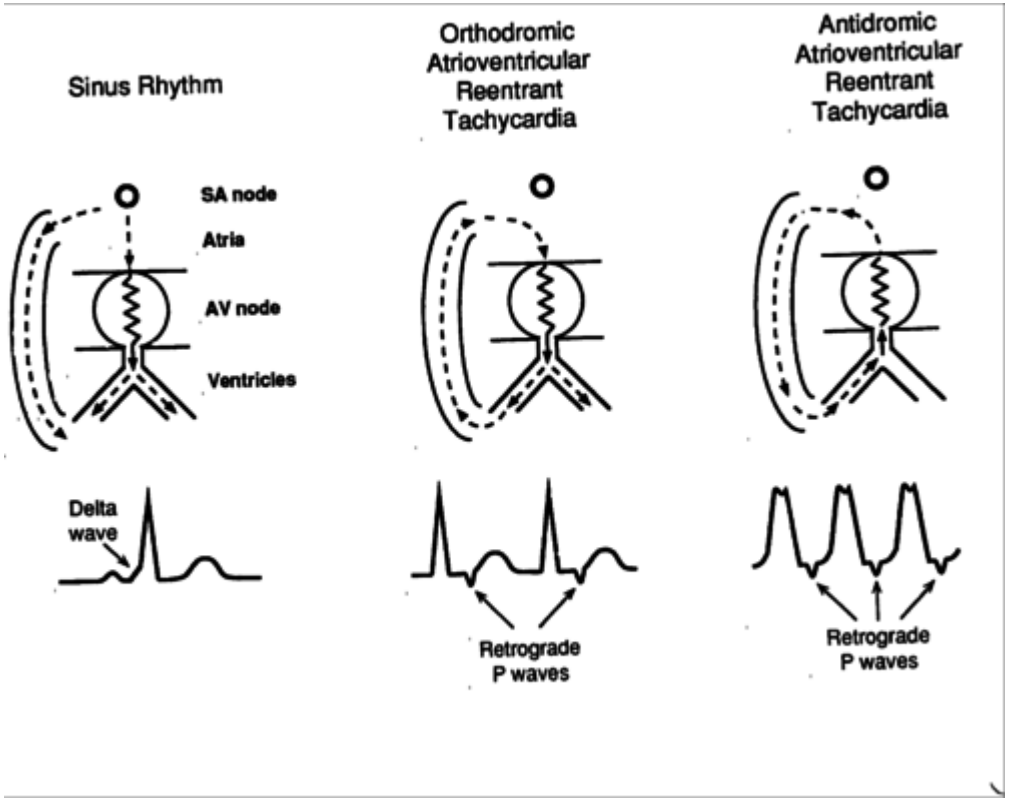




Tachyarrhythmias

- AVRT
- Most common source of sustained SVT in neonatal period
- 60% of cases taken for EPS according to EHRA survey
- Degree of pre-excitation dependent on the node/pathway properties

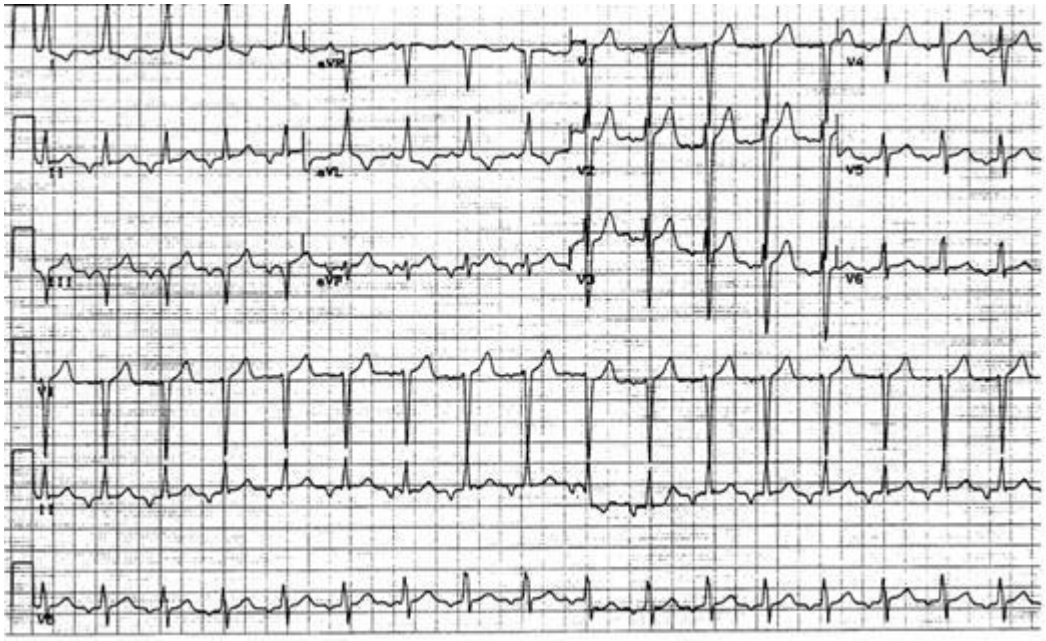
Tachyarrhythmias



Tachyarrhythmias

- Normal management-ABC etc.
- Adenosine reasonable starting point in AVRT- 100mcg/kg as a fast bolus with flush- may need to repeat up to 500mcg/kg
- Longterm medical management options include propanolol/flecainide
- Should all be referred to EP for invasive EP study at some point- small risk of sudden death with rapidly conducting pathway-ERP less than 250ms

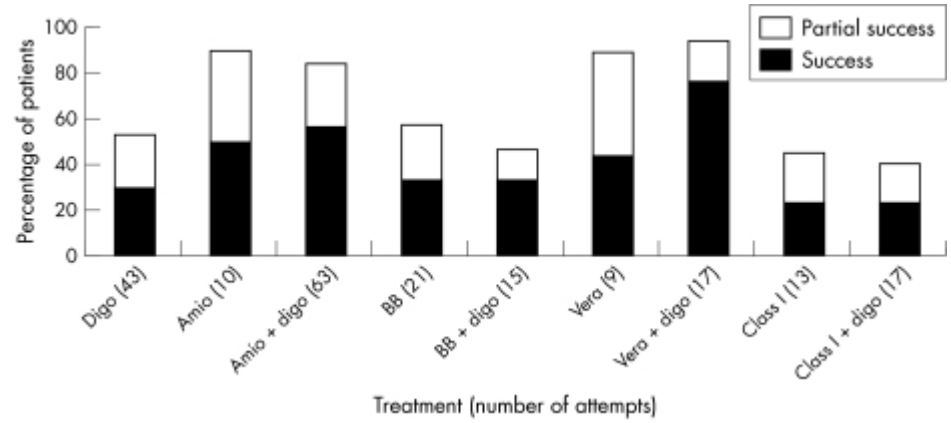
Tachyarrhythmias

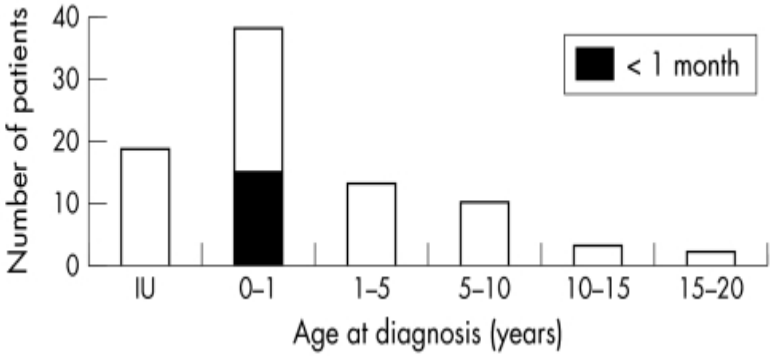


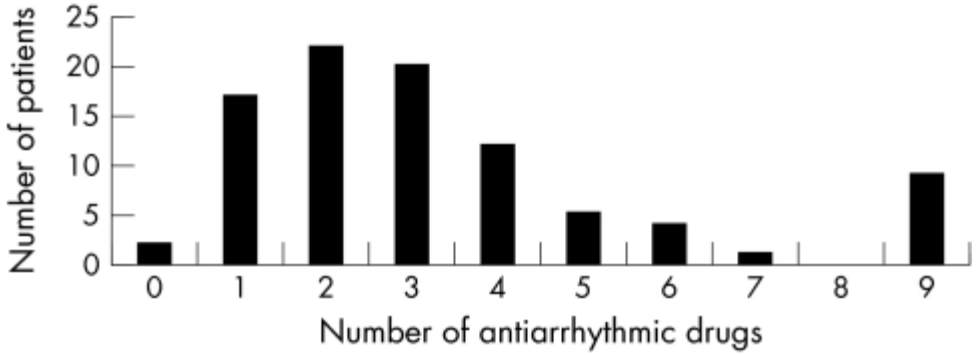
Diagnosis of PJRT

- incessant narrow QRS tachycardia
- negative P waves in inferior leads
- atrioventricular ratio of 1:1.

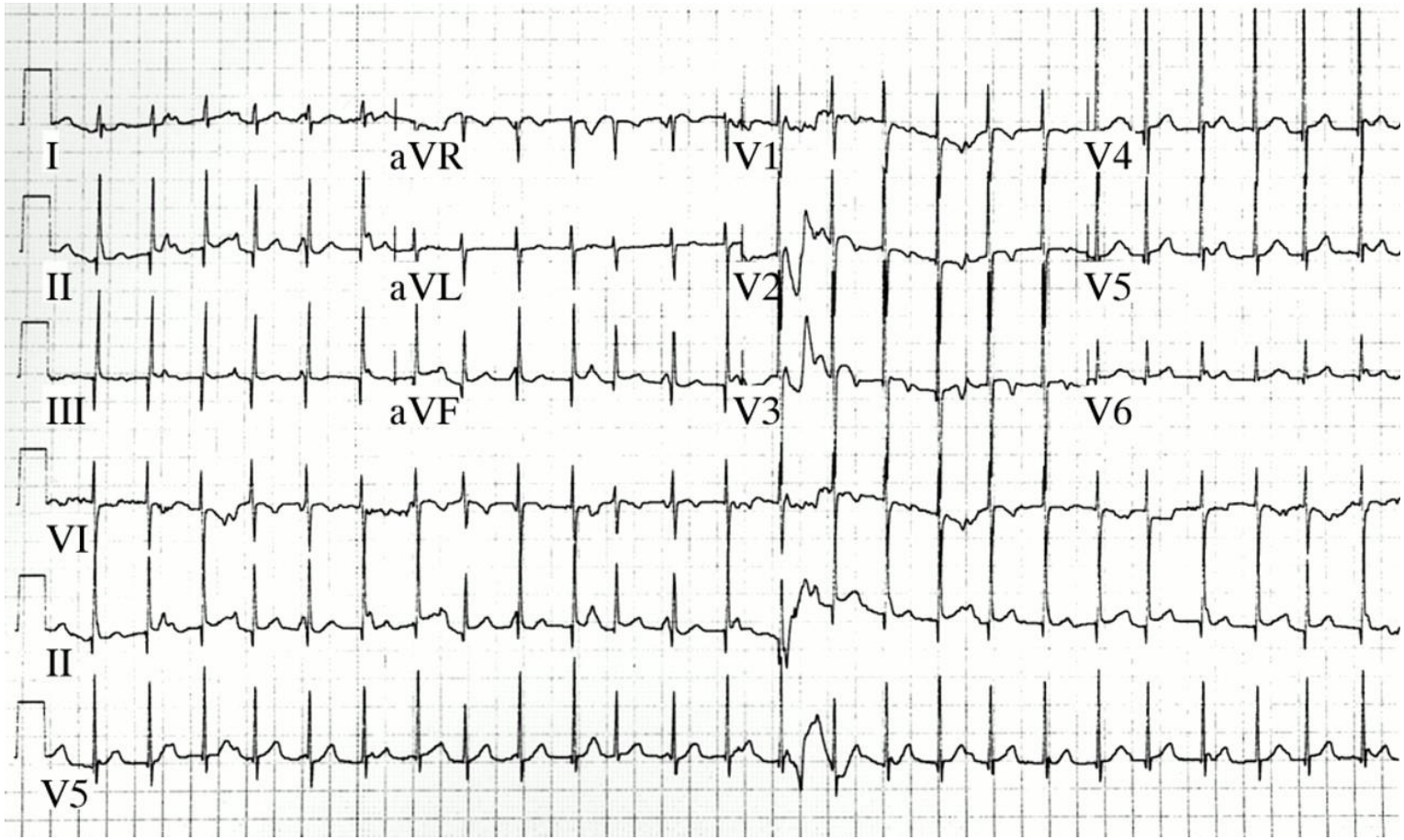
Tachyarrhythmias







Tachyarrhythmias



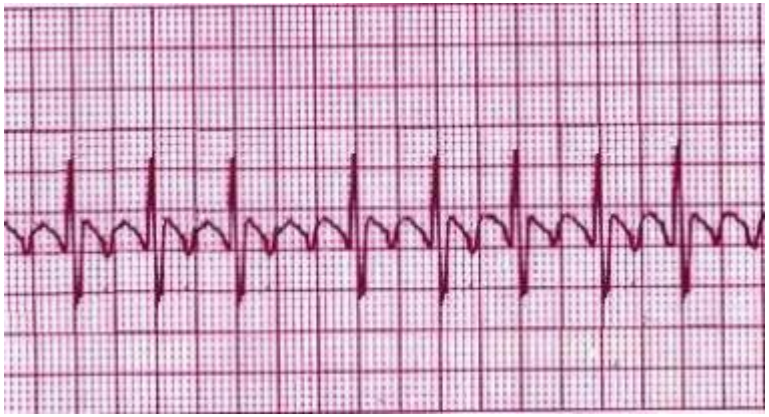
Tachyarrhythmias

- NCT- with AV dissociation
- Common under 6 months- almost always post surgical
- Oedema/trauma around the bundle of His
- Common cardiac lesions associated with the development of JET
  - - Tetralogy of Fallot (ToF)
  - - Ventricular septal defect (VSD)
  - - Atrioventricular septal defect (AVSD)
  - - Transposition of the great arteries (TGA)
  - - Total anomalous pulmonary venous drainage (TAPVD)
- Peri-operative risk factors in the development of JET
  - - Infant < 6 month old
  - - Long cardiopulmonary bypass and cross-clamp times
  - - Extensive myocardial ischaemia/injury (reflected in CK-MB)
  - - Transient AV block immediately post cardiopulmonary bypass
  - - Postoperative inotropic support, particularly dopamine
  - - Acidosis / electrolyte abnormalities, particularly hypomagnesaemia

Tachyarrhythmias

- Treatment- normally resolved by day 8 post op
- Supportive measures-reduce inotropes/cool/fluids and electrolytes
- Sequential pacing
- Amiodarone-Load: 5mg/kg over 1-4 hours and Maintenance infusion of 5-15mg/kg/min

Tachyarrhythmias

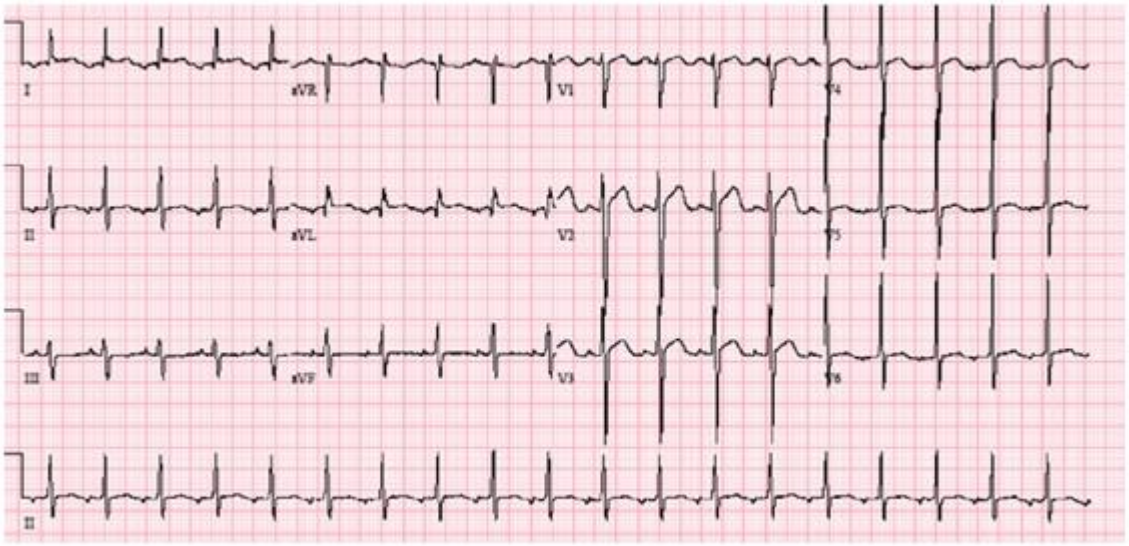


Atrial flutter

- Sawtooth P wave pattern in the inferior leads
- Can occur foetally/neontally
- Can be seen post surgically and in conditions with an enlarged right atrium

- Neonatally not likely to be longterm
- AV blocking agents amio/dig/propanolol will slow V rate till flutter terminates
- Cardiovert 1j/2j per kg
- Easy to ablate in adolescents

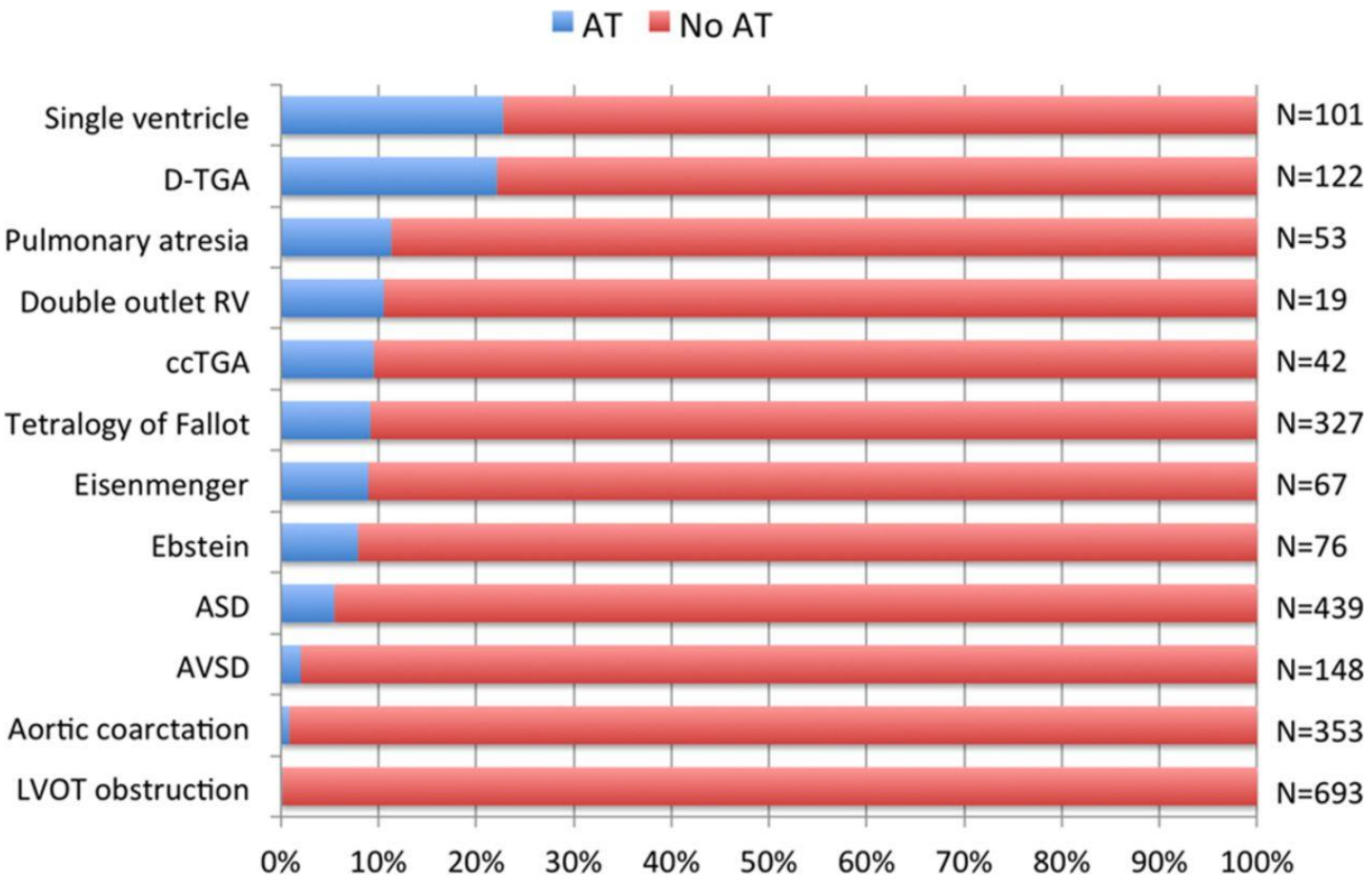
Tachyarrhythmias



Atrial tachycardia

- Can be focal or macro-re-entrant
- Key is comparison to the sinus p wave
- Rate may not be that fast
- Is particularly problematic in the Fontan group

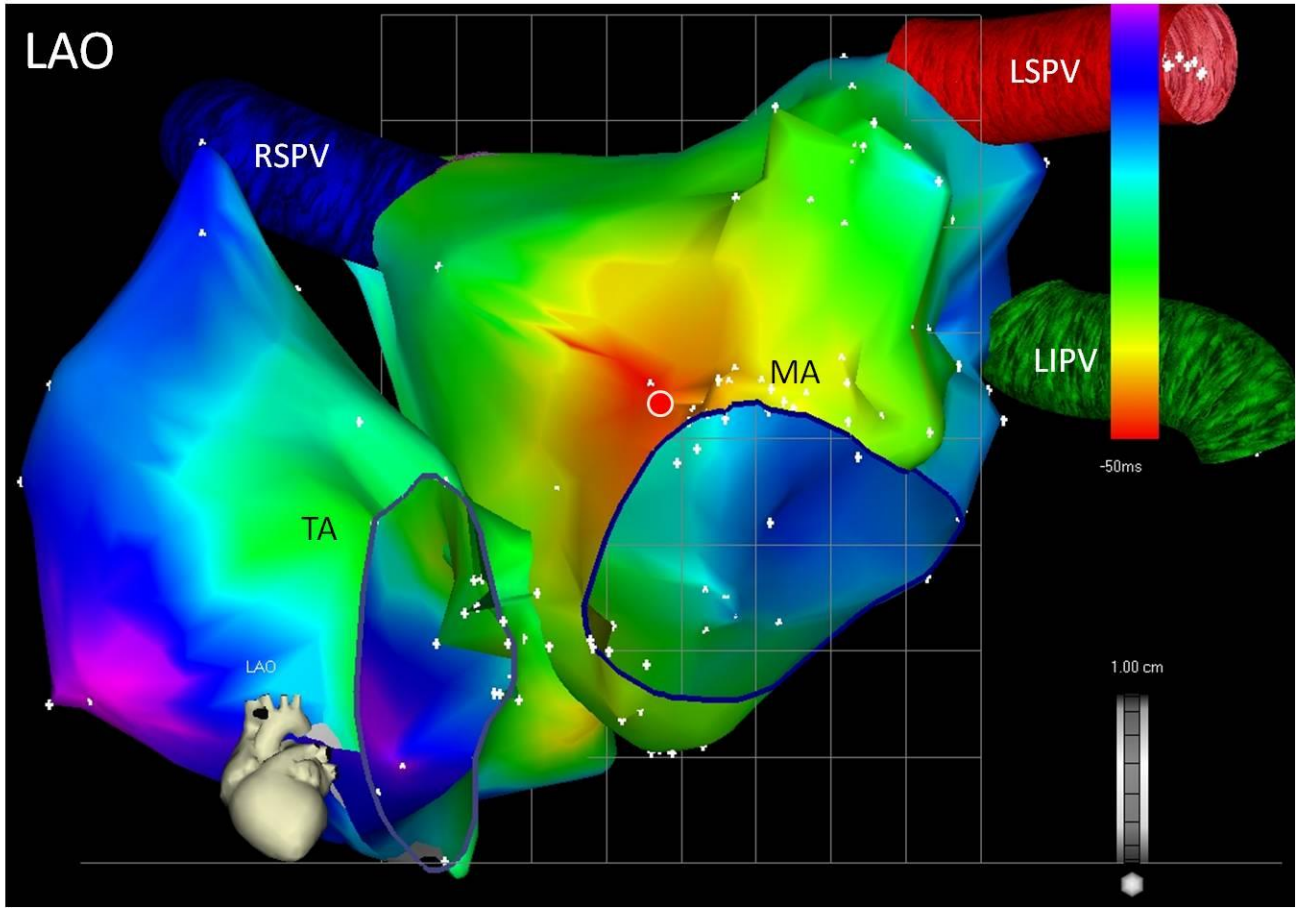
Tachyarrhythmias



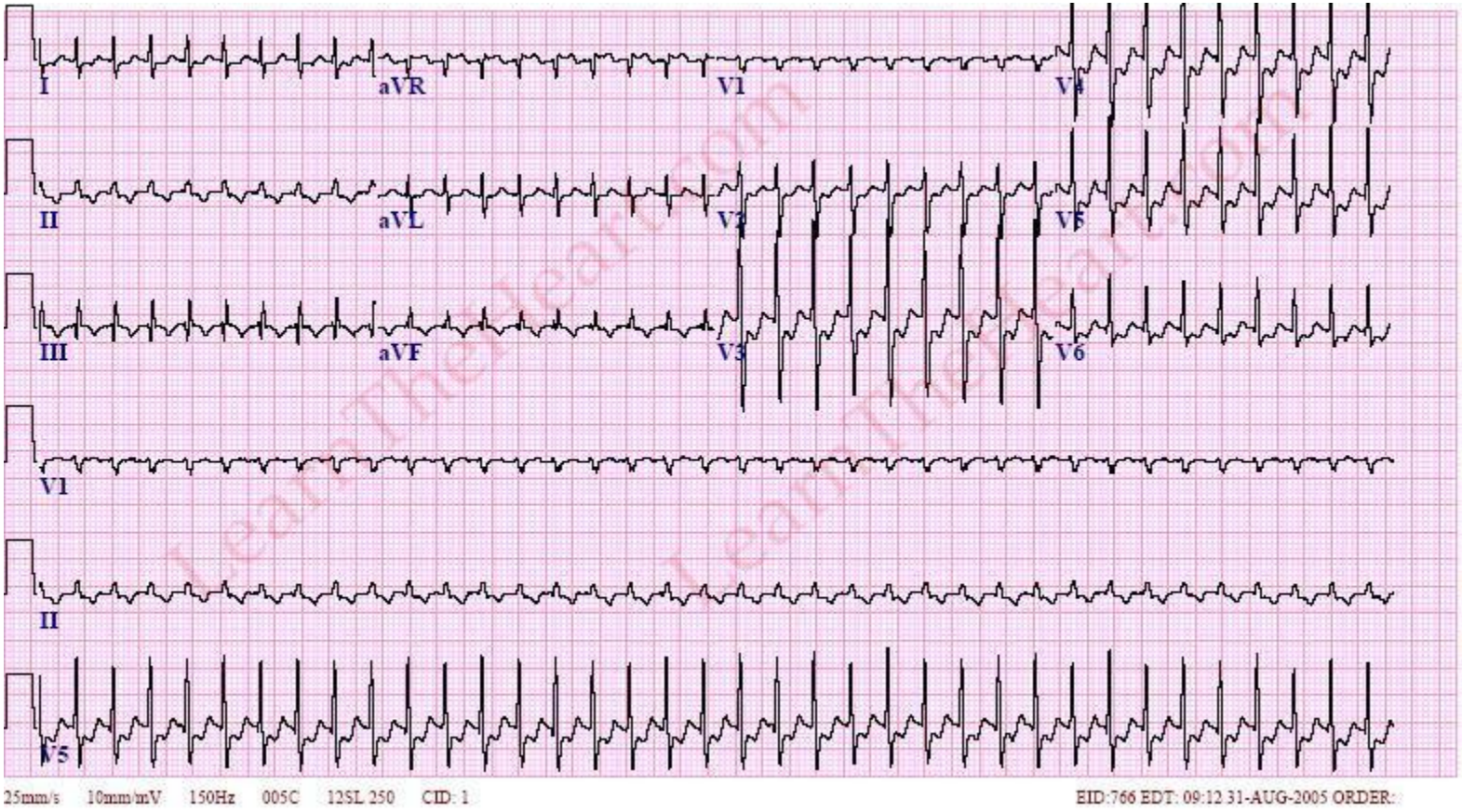
Tachyarrhythmias

- Treatment
  - Short lived- can do nothing
  - Can treat with beta-blockers/dig/amiodarone
  - Amenable to catheter ablation
  - In the single ventricle/Fontan group- does constitute an emergency and should be considered for immediate cardioversion

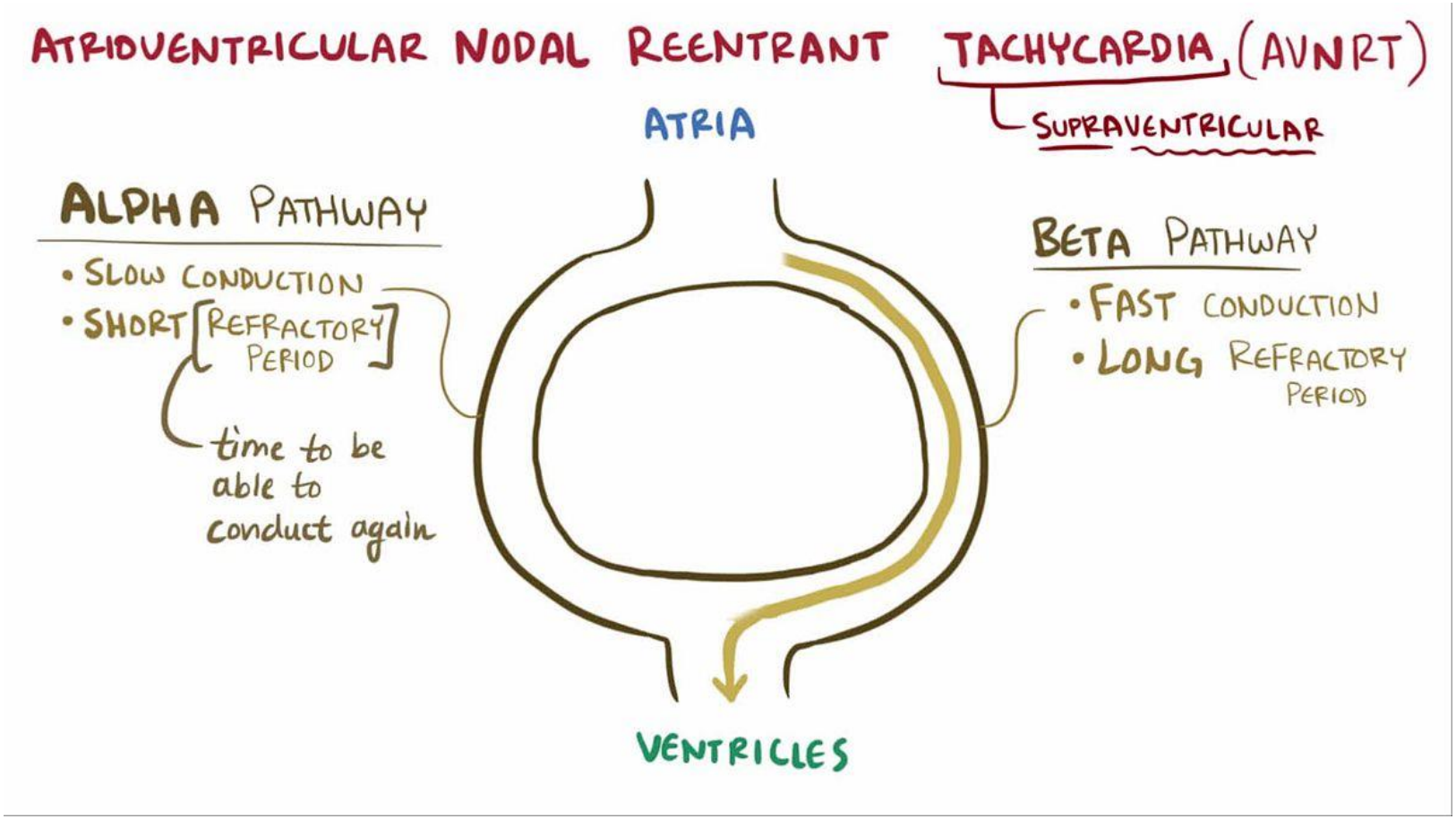
Tachyarrhythmias



Tachyarrhythmias



Tachyarrhythmias

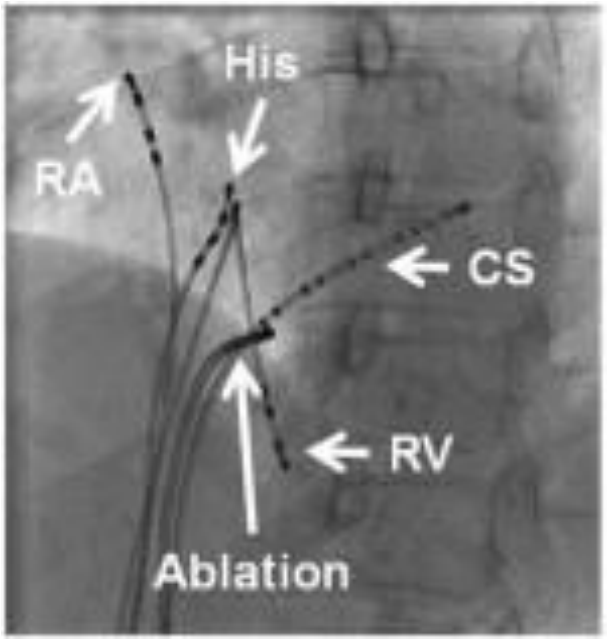


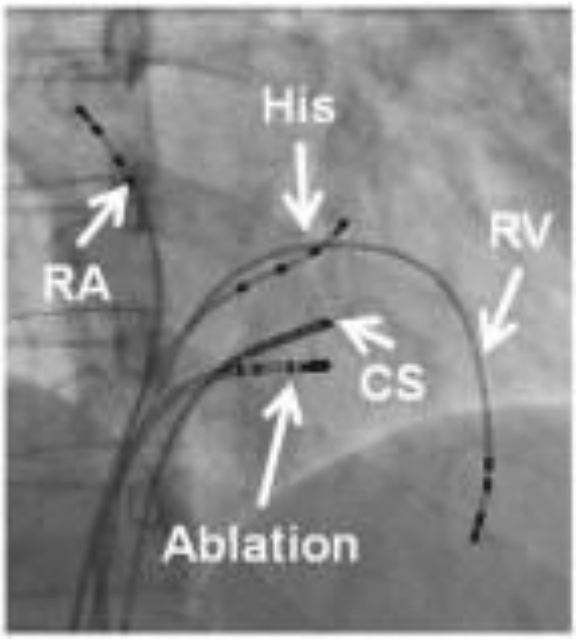
Tachyarrhythmias

- More and more common with age- pre-dominant SVT in adulthood
- Short RP
- Regular
- Often don’t see a P
- Vagal manoeuvres frequently work
- Easily terminated with adenosine
- Can be put on a beta-blocker

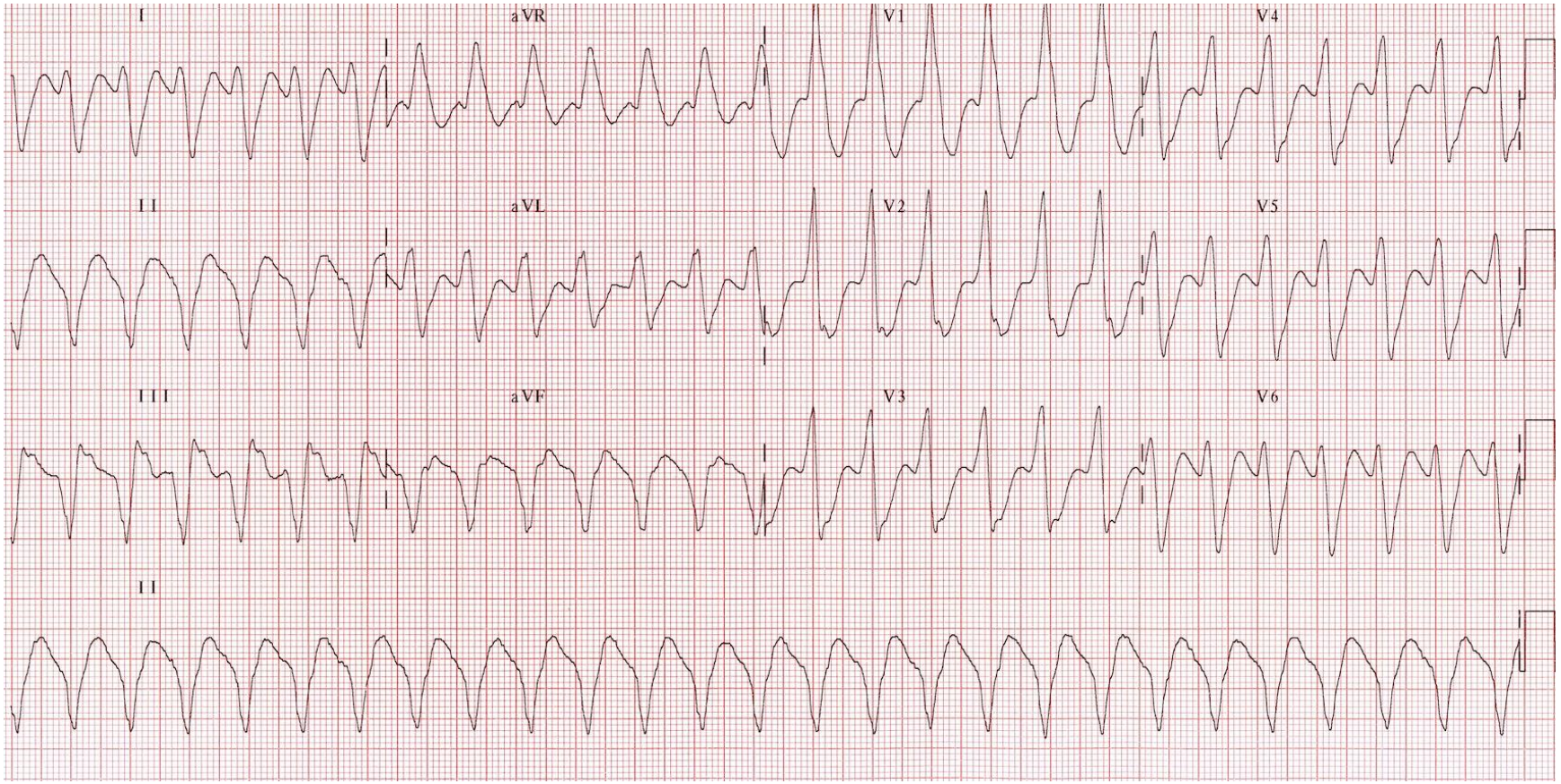
Tachyarrhythmias

- Catheter ablation is first choice treatment for long-term cure

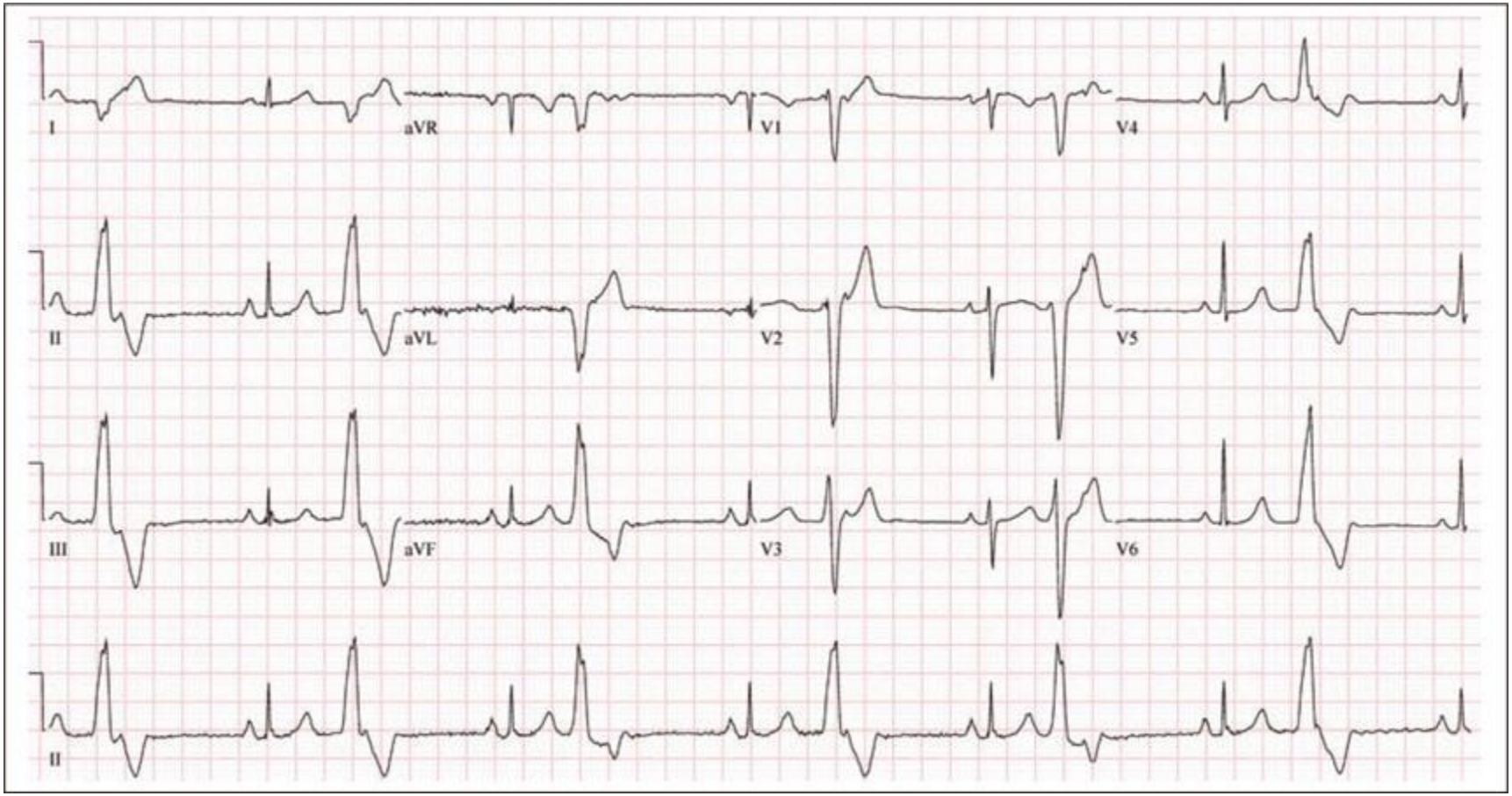




Tachyarrhythmias



Tachyarrhythmias



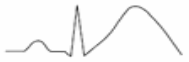
Tachyarrhythmias

- Ectopy common issue
- Not a lot to be done if patient symptomatically ok and heart structurally normal
- Can try to suppress with betablockers/verapamil
- Amenable to catheter ablation if symptomatic on meds/ventricular impairment from high ectopic burden

Channelopathies/Cardiomyopathies

- Long QT
- Brugada
- ARVC
- HCM
- CPVT

Channelopathies/Cardiomyopathies

Type	Current	Functional Effect	Frequency Among LQTS	ECG[12,13]	Triggers Lethal Cardiac Event[10]	Penetrance*
LQTS1	K	↓	30%-35%		Exercise (68%) Emotional Stress (14%) Sleep, Repose (9%) Others (19%)	62%
LQTS2	K	↓	25%-30%		Exercise (29%) Emotional Stress (49%) Sleep, Repose (22%)	75%
LQTS3	Na	↑	5%-10%		Exercise (4%) Emotional Stress (12%) Sleep, Repose (64%) Others (20%)	90%

Need to remember in syncope/epilepsy
Most events seem to occur after the age of 5 with LQT3 presenting latest

Table 1. Characteristics of LQTS Subtypes

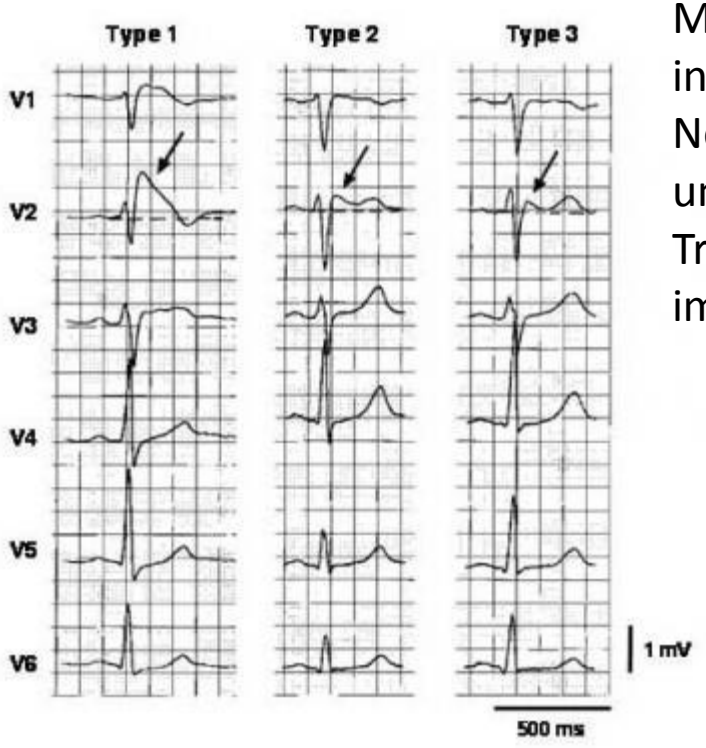
Phenotype	Frequency	Trigger(s)	Cause	Mean QTc	Treatment
LQT1	60%	Exercise (e.g., swimming, running); emotion (startle, anger, fright)	Mutation in *KCNQ1*- or *KCNE1*-defective I_{Ks} channels	490 msec	Beta-blockers
LQT2	35%	Auditory stimulation causing sudden startle (e.g., alarm clock, telephone, siren)	Mutation in *hERG*- or *KCNE2*-defective I_{Kr} channels	480 msec	Beta-blockers
LQT3	4%-5%	Sleep	Mutation in *SCN5A*- I_{Na}	510-520 msec	Beta-blockers[a]; sodium channel blockers[b]; pacemaker with defibrillator

[a] *Questionable efficacy.*
[b] *A study evaluating ranolazine for LQT3 is scheduled to be completed in September 2014.*
LQTS: long QT syndrome.
Source: Reference 9.

Channelopathies/Cardiomyopathies

Recommendations	Class[a]	Level[b]	Ref.[c]
The following lifestyle changes are recommended in all patients with a diagnosis of LQTS: (a) Avoidance of QT-prolonging drugs (http://www.crediblemeds.org). (b) Correction of electrolyte abnormalities (hypokalaemia, hypomagnesaemia, hypocalcaemia) that may occur during diarrhoea, vomiting or metabolic conditions. (c) Avoidance of genotype-specific triggers for arrhythmias (strenuous swimming, especially in LQTS1, and exposure to loud noises in LQTS2 patients).	I	B	434
Beta-blockers are recommended in patients with a clinical diagnosis of LQTS.	I	B	435
ICD implantation with the use of beta-blockers is recommended in LQTS patients with previous cardiac arrest.	I	B	436–438
Beta-blockers should be considered in carriers of a causative LQTS mutation and normal QT interval.	IIa	B	67
ICD implantation in addition to beta-blockers should be considered in LQTS patients who experienced syncope and/or VT while receiving an adequate dose of beta-blockers.	IIa	B	439
Left cardiac sympathetic denervation should be considered in patients with symptomatic LQTS when (a) Beta-blockers are either not effective, not tolerated or contraindicated; (b) ICD therapy is contraindicated or refused; (c) Patients on beta-blockers with an ICD experience multiple shocks.	IIa	C	440
Sodium channel blockers (mexiletine, flecainide or ranolazine) may be considered as add-on therapy to shorten the QT interval in LQTS3 patients with a QTc >500 ms.	IIb	C	441–443
Implant of an ICD may be considered in addition to beta-blocker therapy in asymptomatic carriers of a pathogenic mutation in *KCNH2* or *SCN5A* when QTc is >500 ms.	IIb	C	67
Invasive EPS with PVS is not recommended for SCD risk stratification.	III	C	117

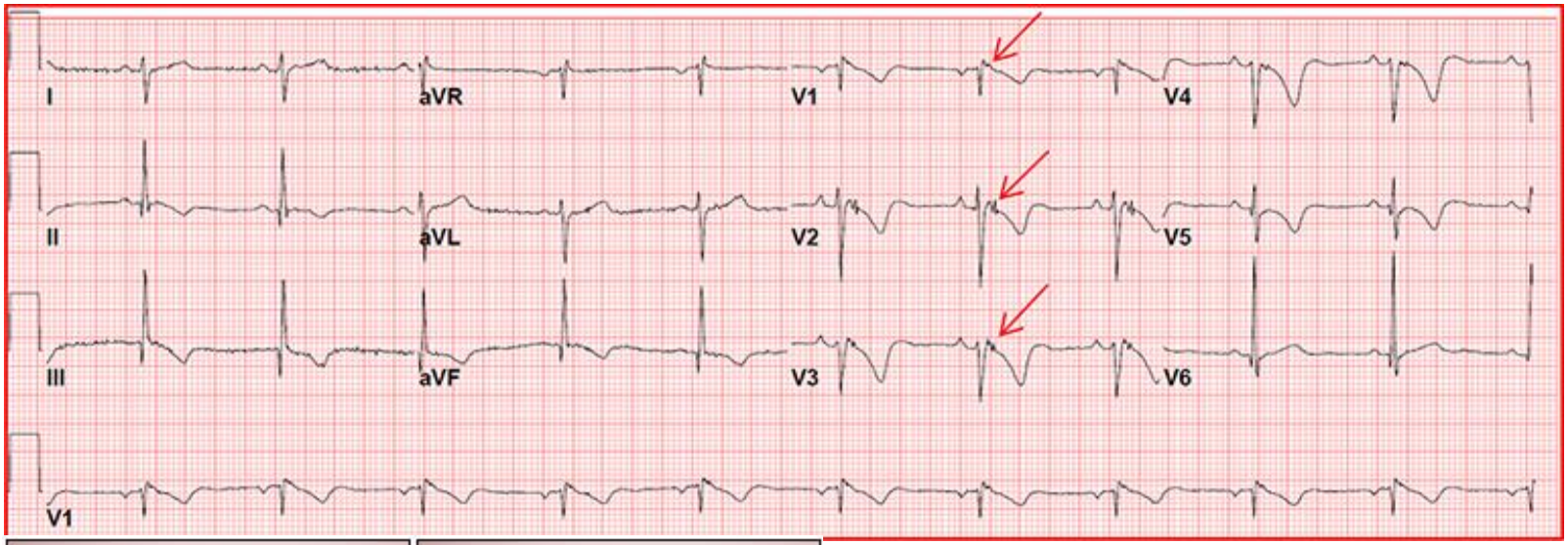
Channelopathies/Cardiomyopathies

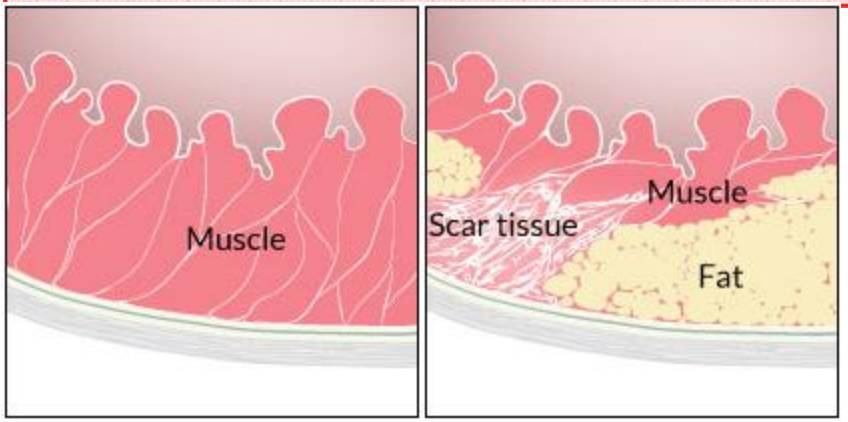


Mean age of presentation in children 8 and above
Need to consider in unexplained syncope
Treatment of fever is important

Recommendations	Class[a]	Level[b]	Ref.[c]
The following lifestyle changes are recommended in all patients with a diagnosis of Brugada syndrome: (a) Avoidance of drugs that may induce ST-segment elevation in right precordial leads (http://www.brugadadrugs.org) (b) Avoidance of excessive alcohol intake and large meals (c) Prompt treatment of any fever with antipyretic drugs.	I	C	This panel of experts
ICD implantation is recommended in patients with a diagnosis of Brugada syndrome who (a) Are survivors of an aborted cardiac arrest and/or (b) Have documented spontaneous sustained VT.	I	C	451
ICD implantation should be considered in patients with a spontaneous diagnostic type I ECG pattern and history of syncope.	IIa	C	451
Quinidine or isoproterenol should be considered in patients with Brugada syndrome to treat electrical storms.	IIa	C	453
Quinidine should be considered in patients who qualify for an ICD but present a contraindication or refuse it and in patients who require treatment for supraventricular arrhythmias.	IIa	C	454
ICD implantation may be considered in patients with a diagnosis of Brugada syndrome who develop VF during PVS with two or three extrastimuli at two sites.	IIb	C	120
Catheter ablation may be considered in patients with a history of electrical storms or repeated appropriate ICD shocks.	IIb	C	201, 455

Channelopathies/Cardiomyopathies





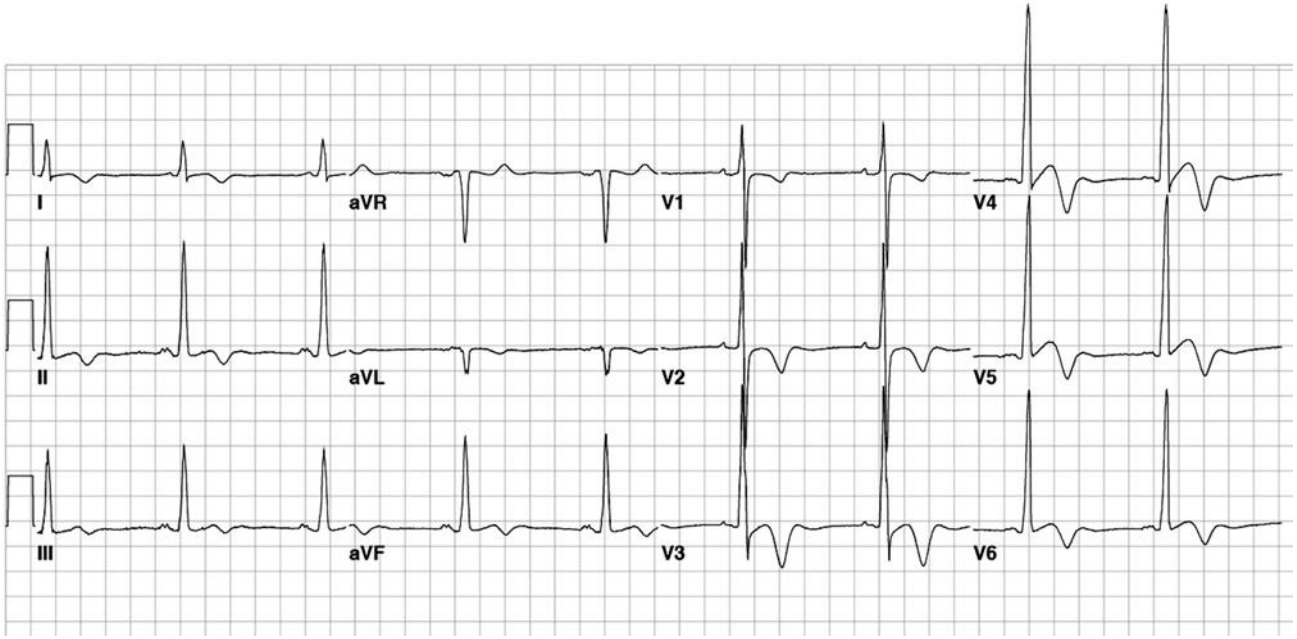
Youngest ever reported case is a 7 yr old

Normally present in teenage yrs

Channelopathies/Cardiomyopathies

Recommendations	Class[a]	Level[b]	Ref.[c]
Avoidance of competitive sports[d] is recommended in patients with ARVC.	I	C	388
Beta-blockers titrated to the maximally tolerated dose are recommended as the first-line therapy to improve symptoms in patients with frequent PVC and NSVT.	I	C	This panel of experts
ICD implantation is recommended in patients with a history of aborted SCD and haemodynamically poorly tolerated VT.	I	C	389
Amiodarone should be considered to improve symptoms in patients with frequent PVC or NSVT who are intolerant of or have contraindications to beta-blockers.	IIa	C	390, 391
Catheter ablation, performed in experienced centres, should be considered in patients with frequent symptomatic PVC or VT unresponsive to medical therapy to improve symptoms and prevent ICD shocks, respectively.	IIa	B	183, 202, 207, 392, 393
ICD implantation should be considered in ARVC patients who have haemodynamically well-tolerated sustained VT, balancing the risk of ICD therapy, including long-term complications, and the benefit for the patient.	IIa	B	387, 394, 395
ICD implantation may be considered in patients with one or more recognized risk factors for VA in adult patients with a life expectancy >1 year following detailed clinical assessment that takes into account the lifelong risk of complications and the impact of an ICD on lifestyle, socioeconomic status and psychological health.	IIb	C	This panel of experts
Invasive EPS with PVS may be considered for stratification of SCD risk.	IIb	C	113, 114

Channelopathies/Cardiomyopathies

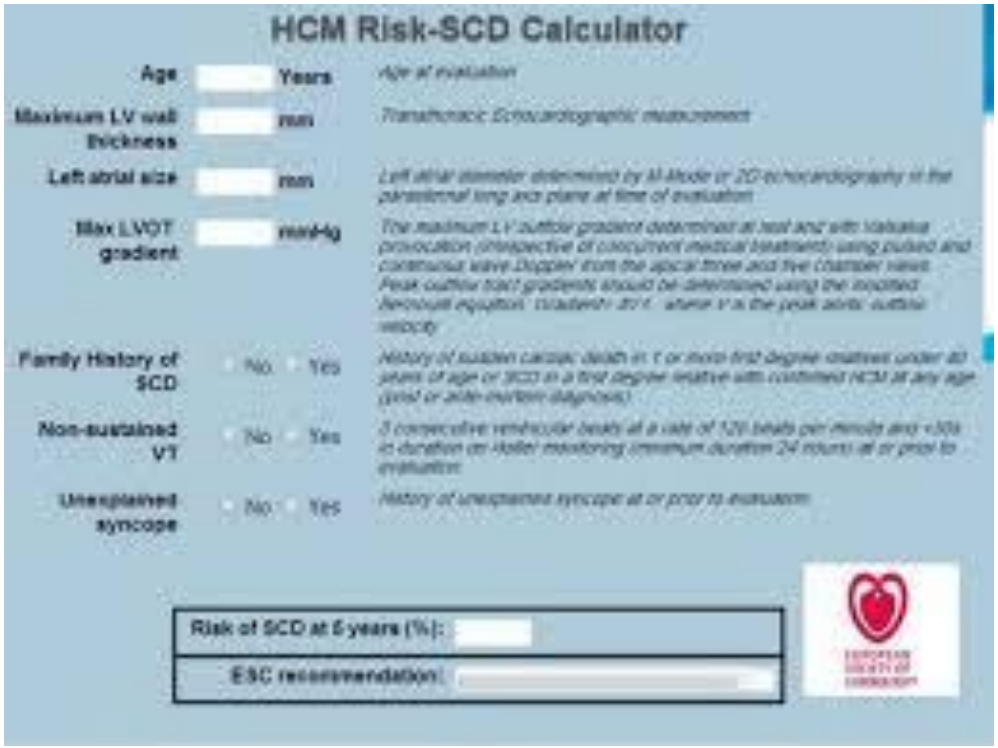


HCM more common in teenagers/young adults

Most common genes are myosin genes

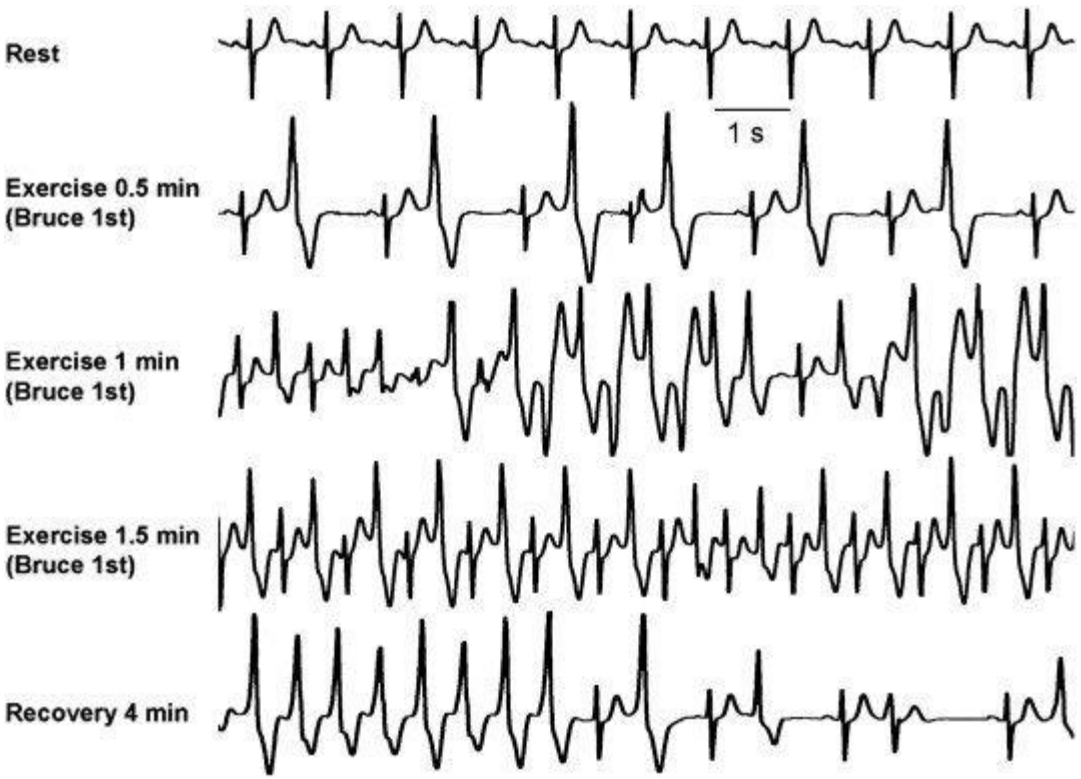
Screening does not normally begin till after 12yrs unless worried

Channelopathies/Cardiomyopathies



Recommendations	Class[a]	Level[b]	Ref.[c]
Avoidance of competitive sports[d] is recommended in patients with HCM.	I	C	366
ICD implantation is recommended in patients who have survived a cardiac arrest due to VT or VF or who have spontaneous sustained VT causing syncope or haemodynamic compromise and a life expectancy >1 year.	I	B	116, 367–372
Risk stratification with the HCM Risk-SCD calculator is recommended to estimate the risk of sudden death at 5 years in patients ≥16 years of age without a history of resuscitated VT or VF or spontaneous sustained VT causing syncope or haemodynamic compromise.	I	B	116, 365
It is recommended that the 5-year risk of SCD is assessed at first evaluation and at 1- to 2-year intervals, or when there is a change in clinical status.	I	B	116, 365
ICD implantation should be considered in patients with an estimated 5-year risk of sudden death ≥6% and a life expectancy >1 year following detailed clinical assessment that takes into account the lifelong risk of complications and the impact of an ICD on lifestyle, socioeconomic status and psychological health.	IIa	B	116, 368
ICD implantation may be considered in individual patients with an estimated 5-year risk of SCD of ≥4 to <6% and a life expectancy >1 year following detailed clinical assessment that takes into account the lifelong risk of complications and the impact of an ICD on lifestyle, socioeconomic status and psychological health.	IIb	B	116, 365, 368
ICD implantation may be considered in individual patients with an estimated 5-year risk of SCD <4% when they have clinical features that are of proven prognostic importance and when an assessment of the lifelong risk of complications and the impact of an ICD on lifestyle, socioeconomic status and psychological health suggests a net benefit from ICD therapy.	IIb	B	116, 365, 368
Invasive EPS with PVS is not recommended for stratification of SCD risk.	III	C	116

Channelopathies/Cardiomyopathies



Recommendations	Class[a]	Level[b]	Ref.[c]
The following lifestyle changes are recommended in all patients with a diagnosis of CPVT: avoidance of competitive sports, strenuous exercise and stressful environments.	I	C	This panel of experts
Beta-blockers are recommended in all patients with a clinical diagnosis of CPVT, based on the presence of documented spontaneous or stress-induced VAs.	I	C	458, 460
ICD implantation in addition to beta-blockers with or without flecainide is recommended in patients with a diagnosis of CPVT who experience cardiac arrest, recurrent syncope or polymorphic/bidirectional VT despite optimal therapy.	I	C	458, 461
Therapy with beta-blockers should be considered for genetically positive family members, even after a negative exercise test.	IIa	C	461, 462
Flecainide should be considered in addition to beta-blockers in patients with a diagnosis of CPVT who experience recurrent syncope or polymorphic/bidirectional VT while on beta-blockers, when there are risks/ contraindications for an ICD or an ICD is not available or rejected by the patient.	IIa	C	463
Flecainide should be considered in addition to beta-blockers in patients with a diagnosis of CPVT and carriers of an ICD to reduce appropriate ICD shocks.	IIa	C	463
Left cardiac sympathetic denervation may be considered in patients with a diagnosis of CPVT who experience recurrent syncope or polymorphic/ bidirectional VT/several appropriate ICD shocks while on beta-blockers or beta-blockers plus flecainide and in patients who are intolerant or have contraindication to beta-blockers.	IIb	C	464, 465
Invasive EPS with PVS is not recommended for stratification of SCD risk.	III	C	14