VENTRICULAR TACHYCARDIAS IN THE ABSENCE OF STRUCTURAL HEART DISEASE

Dr RAJESH K F

10% of patients presenting with VT have no apparent structural heart disease

VT in structurally normal hearts can be broadly considered under

- Non–life-threatening monomorphic VT
- Life-threatening polymorphic VT

NON–LIFE-THREATENING (TYPICALLY MONOMORPHIC)

Classified on basis of site of origin

Most common sites are ventricular outflow tracts and left ventricular fascicles

Outflow tract VT

Right ventricular outflow- 80%

Pulmonary artery

Left ventricular outflow-10%

- Aortic sinus of Valsalva
- Aortic cusps
- Area of aortomitral continuity
- Superior basal septum near His bundle(Peri His bundle)
- Epicardial surface of outflow tracts

Idiopathic left VT

Left posterior fascicle

Left anterior fascicle

High septal fascicle

Others

Mitral annulus

Tricuspid annulus

Papillary muscle

Perivascular epicardial

OUTFLOW TRACT VT

Idiopathic VT originate most commonly in outflow tract area

Nearly 80% of which originate from RVOT

Other outflow tract sites are rare

PHENOTYPES

Phenotypes are a continuum of the same focal cellular process

- Premature ventricular complexes (PVCs)
- Nonsustained,repetitive monomorphic VT(RMVT)
- Paroxysmal, exercise-induced sustained VT

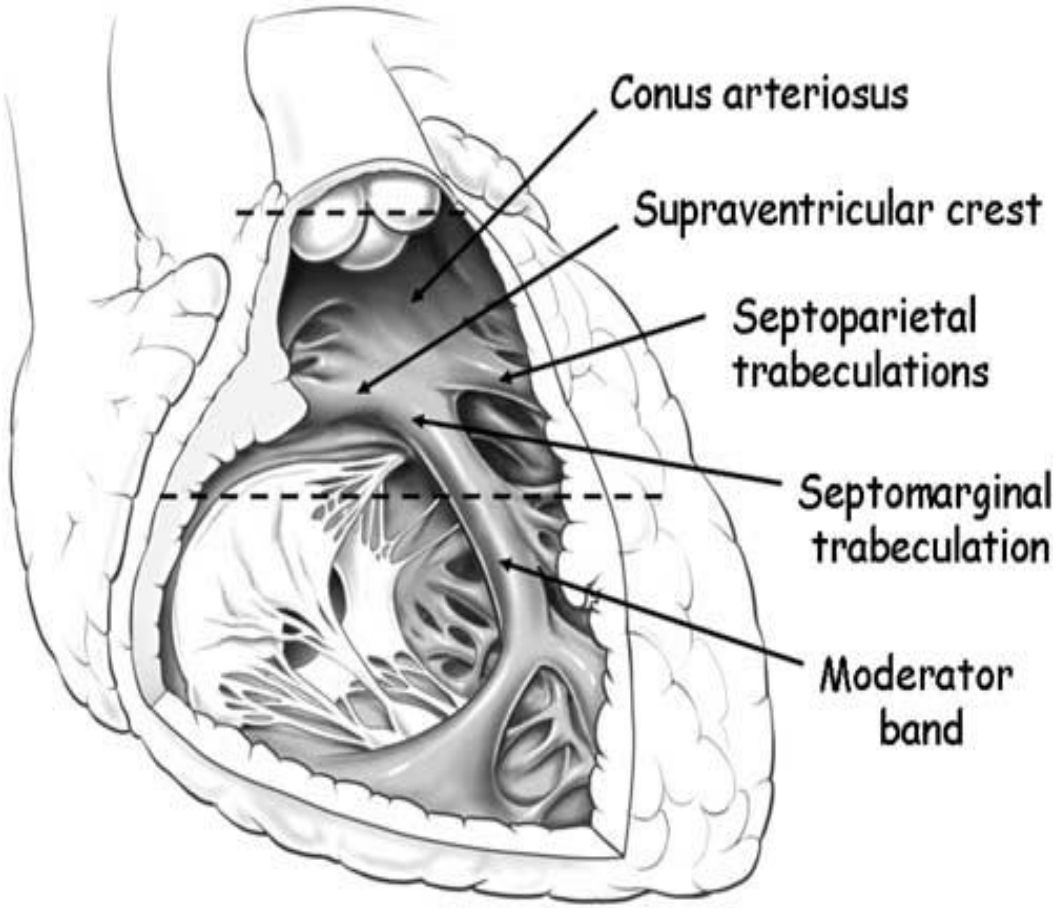
Considerable overlap observed among three phenotypes

Ablating one phenotype at a discrete site eliminates other two

Signature characteristic of sustained RVOT and LVOT is tachycardia is termination by adenosine and verapamil

ANATOMIC CORRELATES

- RVOT is bounded by pulmonary valve superiorly and superior aspect of tricuspid apparatus inferiorly
- RVOT is leftward and anterior to LVOT
- RVOT is a muscular infundibulum circumferentially
- Upper part of septal wall is the conus arteriosus, bordered below by supraventricular crest

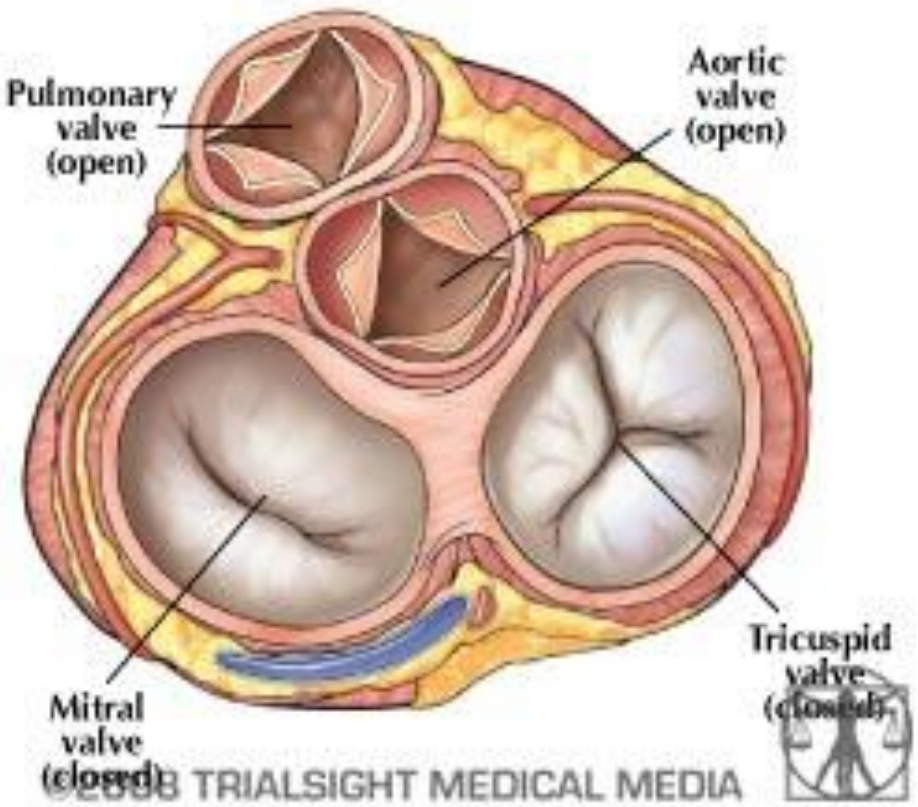


LVOT is region of LV between anterior cusp of mitral valve and ventricular septum

Muscular and fibrous parts

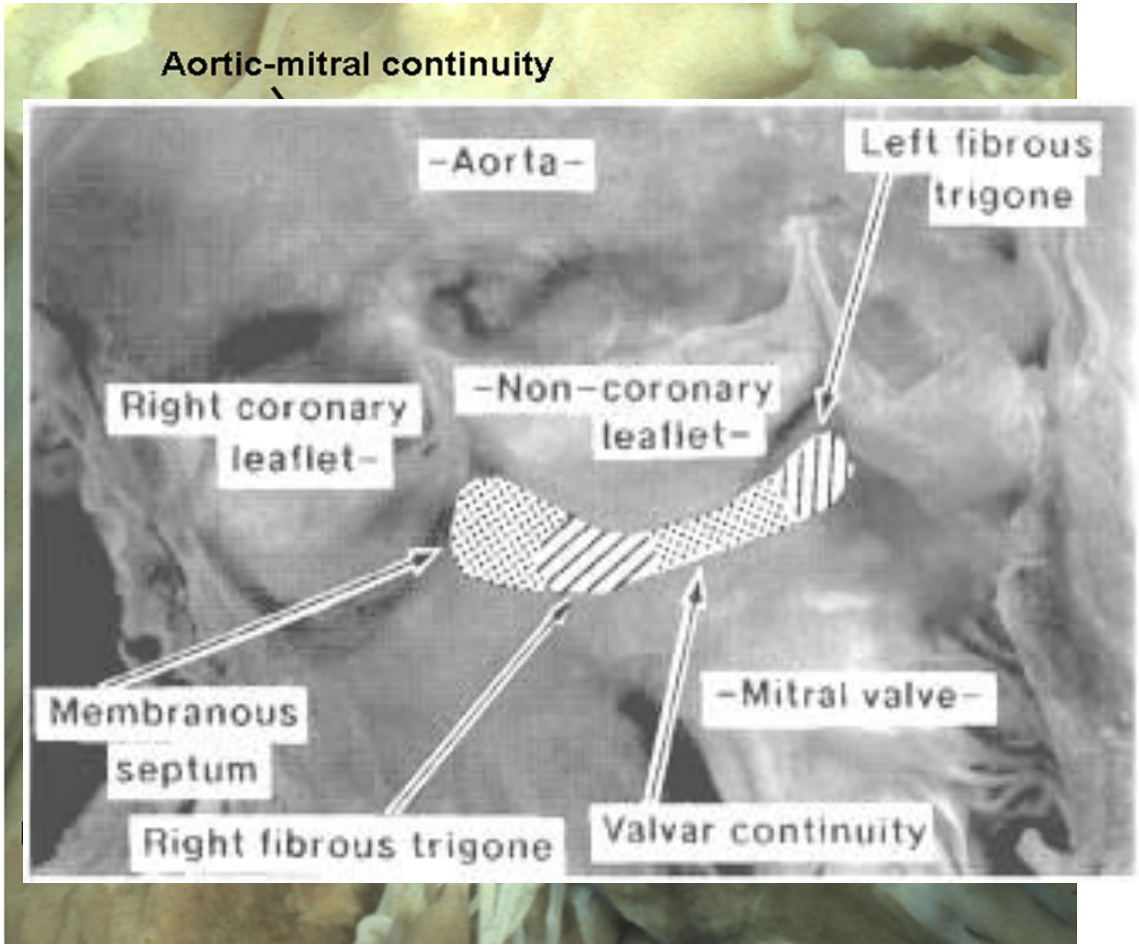
Large of part of right and some part of left aortic sinuses of Valsalva overlie muscular LVOT

Close proximity to AV node and His bundle -early activation in VT



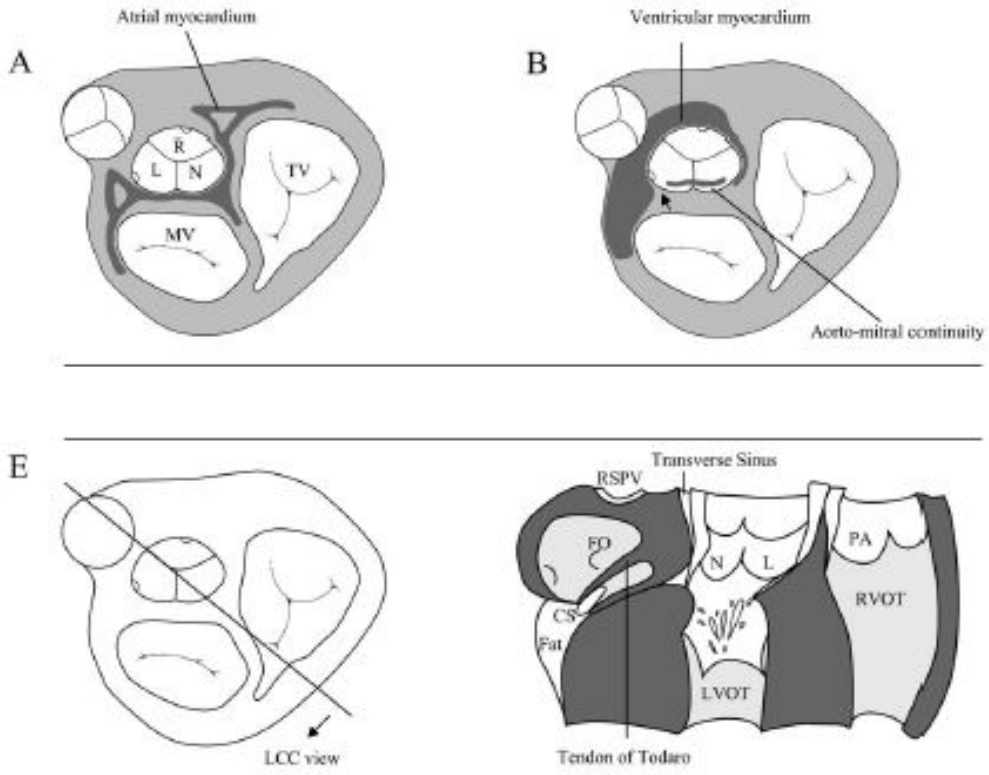
Non-coronary cusp and posterior aspect of left coronary cusp are continuous with fibrous aortomitral continuity

Explain lack of VT related to the non-coronary cusp

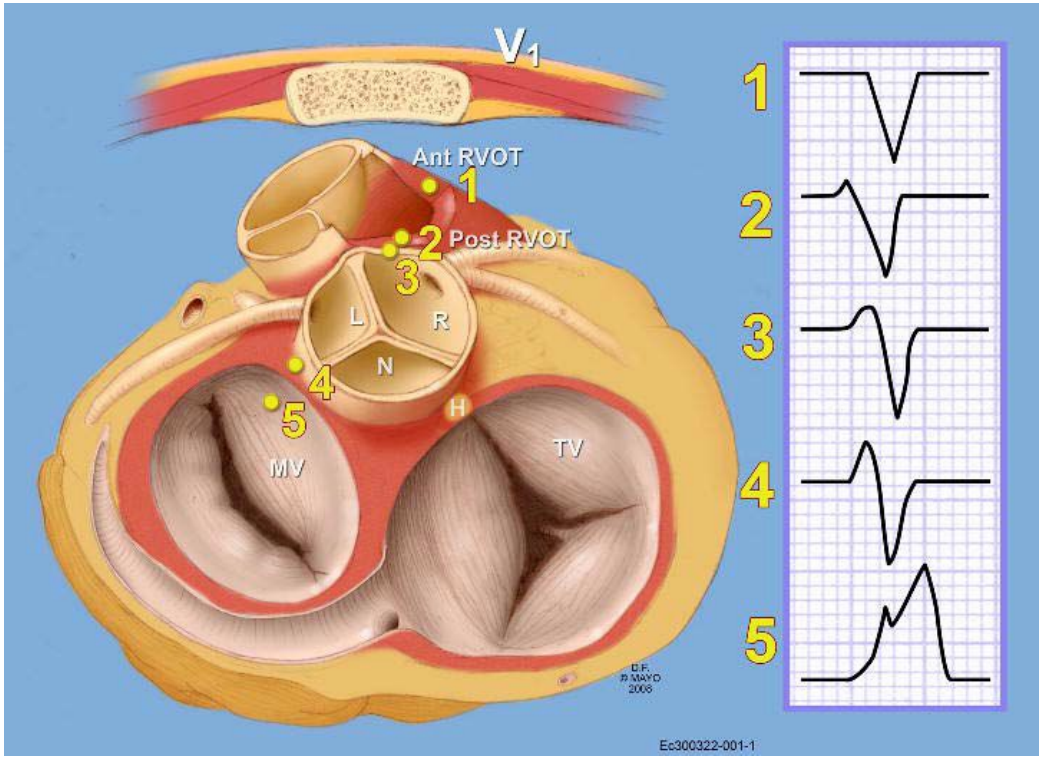


VT from aortic sinuses of Valsalva arise from muscular extensions of the LVOT to areas above the base of the aortic valve cusps

These muscle fibers often exhibit slow conduction and fractionated electrograms.



Localization of site of VT origin can be predicted using QRS morphology on surface ECG and anatomic relationships help to explain shared ECG patterns and subtle differences

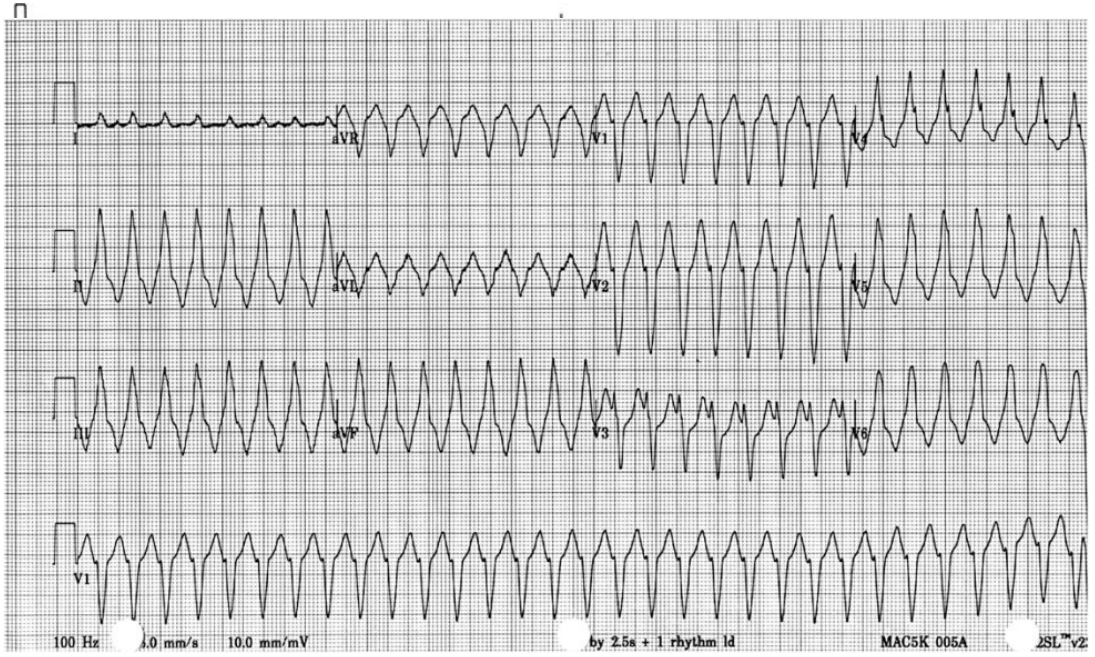


RVOT VT

LBBB and inferior axis

Right sided origin- LBBB pattern with transition from a small r-wave to a large R-wave at V3 to V4

OT site - inferior axis

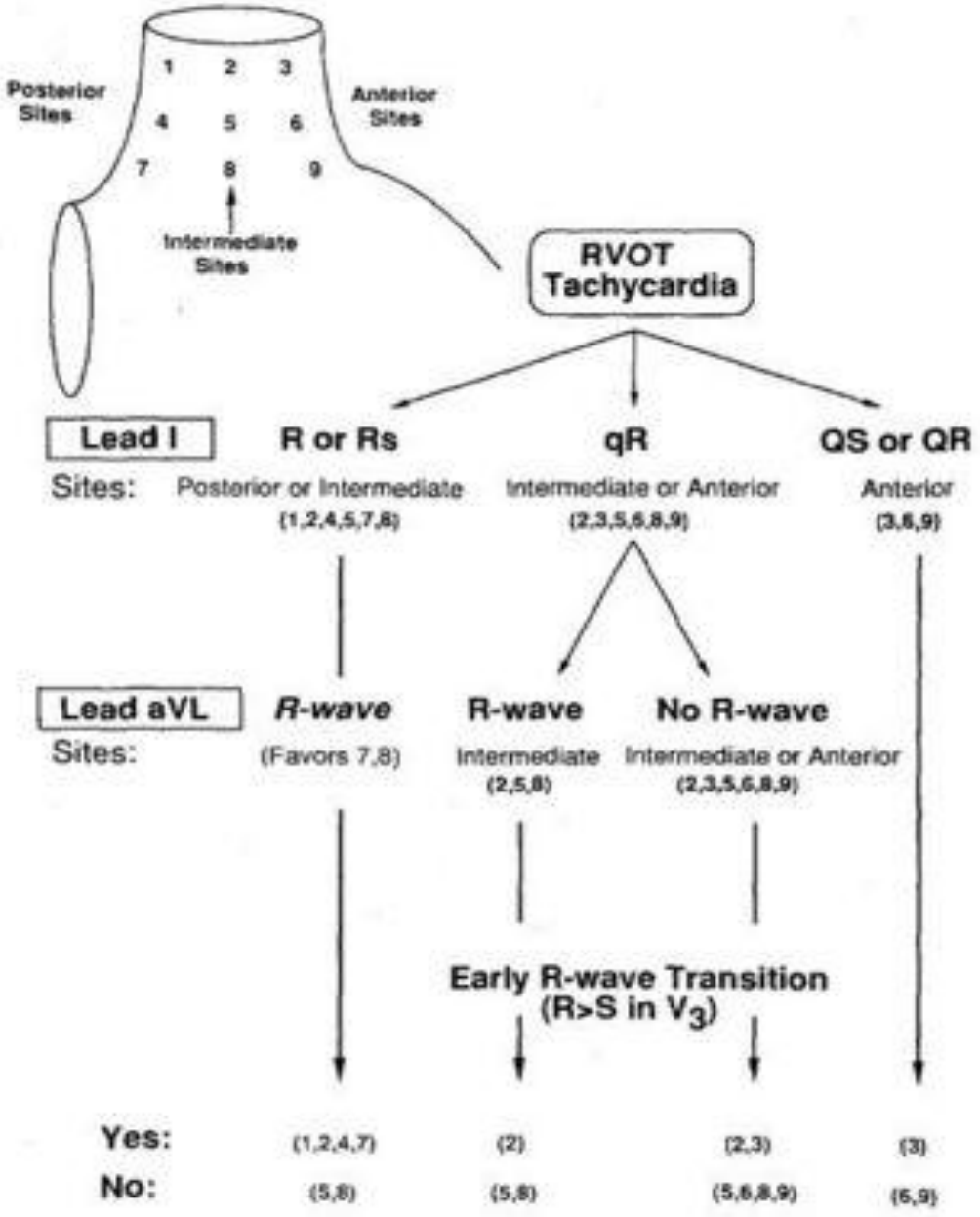


RVOT region can be divided into nine regions

Anterior sites demonstrate Q wave (Q or qR) in lead I and QS in lead aVL

Posterior sites demonstrate R wave in lead I and early precordial transition (R> S in V3)

Between anterior and posterior locations typically demonstrate a multiphasic QRS morphology in lead I



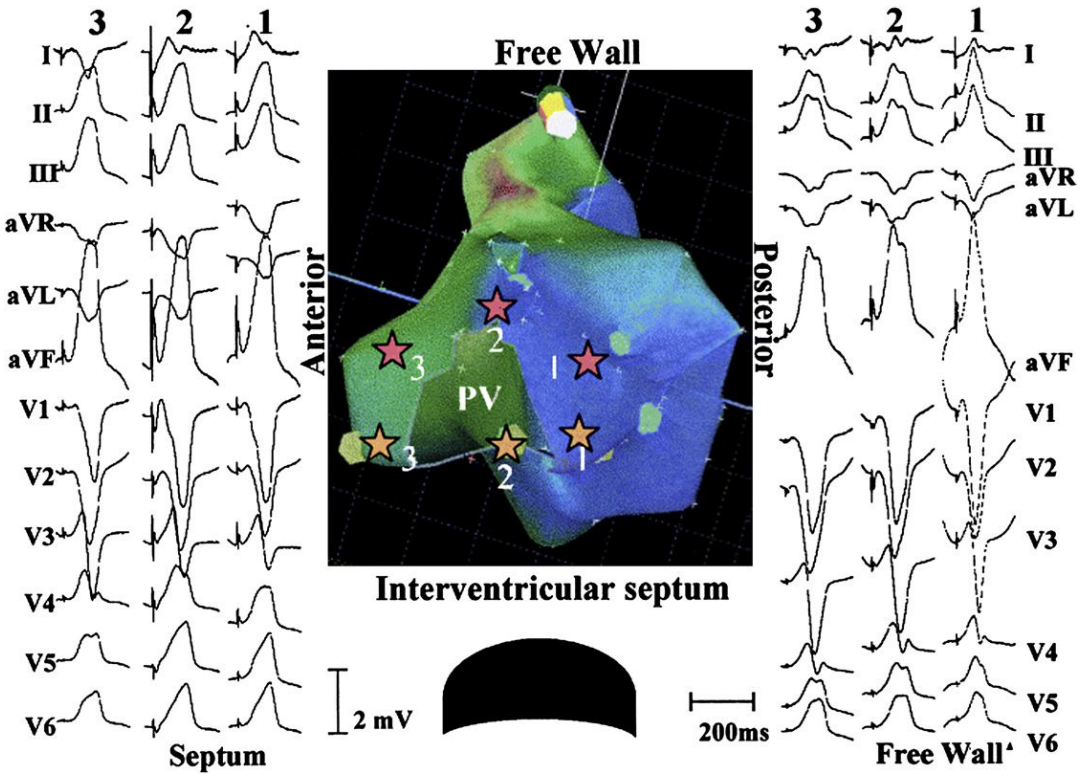
JADONATH AND COLLEAGUES

Differentiation of septal from free wall RVOT VT

RVOT VTs originating from septum - taller,narrower monophasic R waves in inferior leads

Free wall RVOT VT- typically broader QRS (>140ms) and R wave notching in inferior leads

Later transition in precordial leads (>V4)



DIXIT AND COLLEAGUES

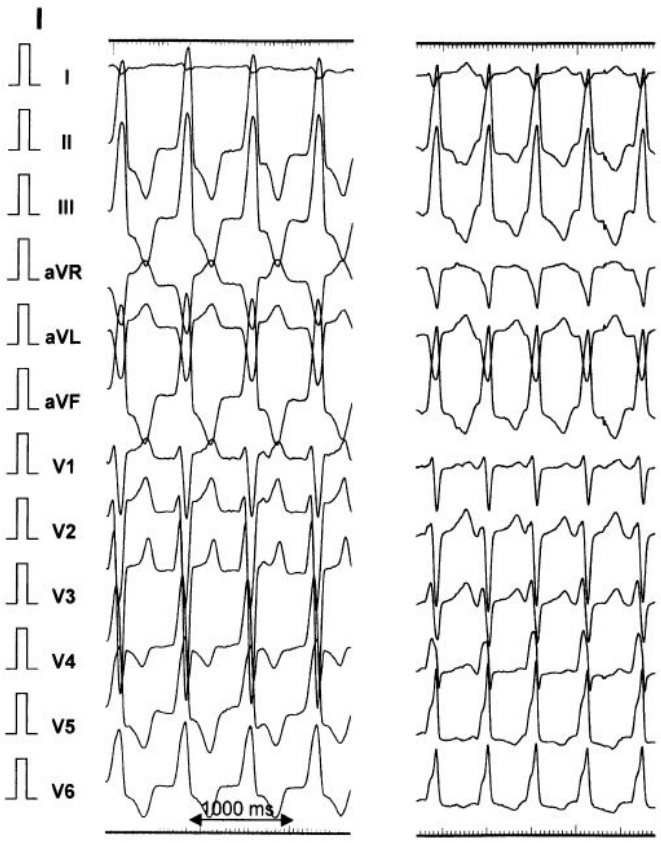
Anterior position of free wall relative to septum -Accounts for deeper S wave in lead V2 than RVOT septum

Septal site associated with a Q/q wave in lead I, whereas a free-wall site inscribes an R/r wave.

Caudal (> 2 cm from PV) Versus Cranial

VT arising >2 cm of the pulmonary valve near His bundle virtually always has a negative QRS in lead aVL

PULMONARY ARTERY VT



- Approximately 1 cm above pulmonic valve
- Associated with a precordial transition in leads V3 or V4(PA is located leftward of and anterior to RVOT)
- qR configuration in lead I
- Larger Q wave in lead aVL than in Avr
- Location superior to RVOT results in a relatively greater R wave amplitude in inferior leads
- Mapping RVOT area -low-voltage atrial or local ventricular potential of <1-mV amplitude
- RF ablation performed on PA requires more attention

DIFFERENTIAL DIAGNOSIS OF RVOT VT

Atriofascicular fibers (Mahaim fibers)

AVRT using Rt-sided accessory pathway

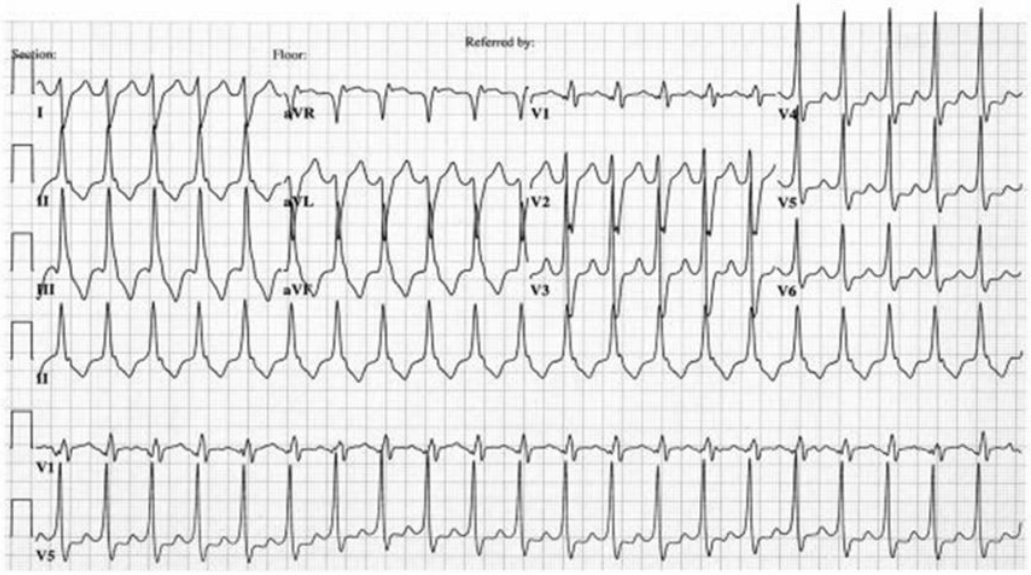
VT after repair of TOF

ARVD

LVOT VT

ECG during VT shows

- **S wave in lead I**
- **R-wave transition in lead V1or V2(Earlier precordial transition zone)**
- **More rightward axes**
- **Taller R waves in inferior leads**



S wave in LI and R-wave transition in V1 suggest LVOT VT.
R:S amplitude ratio of 30% and R:QRS duration ratio of 50%
Presence of an S wave in leads V5 and V6 suggests an infravalvular origin of the tachycardia.

Shows one of the following depending on site of origin

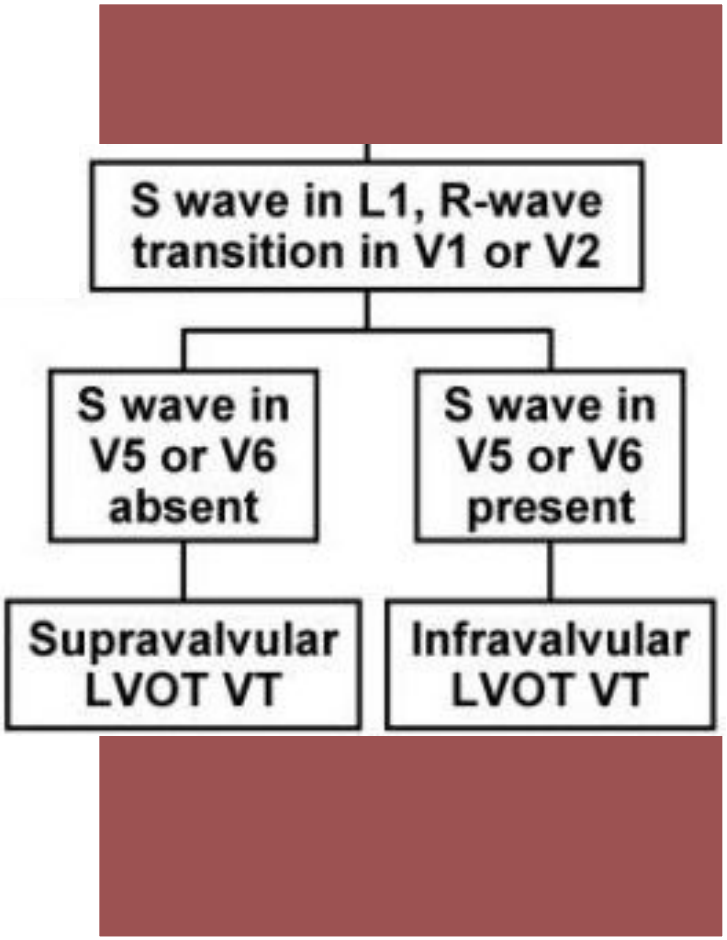
a)Basal left interventricular or septal origin

LBBB morphology with an early precordial transition in lead V2 or V3,S wave in lead V6 (due to activation of the left bundle Purkinje system) and relatively narrow QRS complex

b)VT from region of left fibrous trigone (aortomitral valve continuity)

RBBB morphology in V1 and broad monophasic R-waves across precordium

LVOT VT



May originate from supravalvular or infravalvular endocardial region of coronary cusp of aortic valve

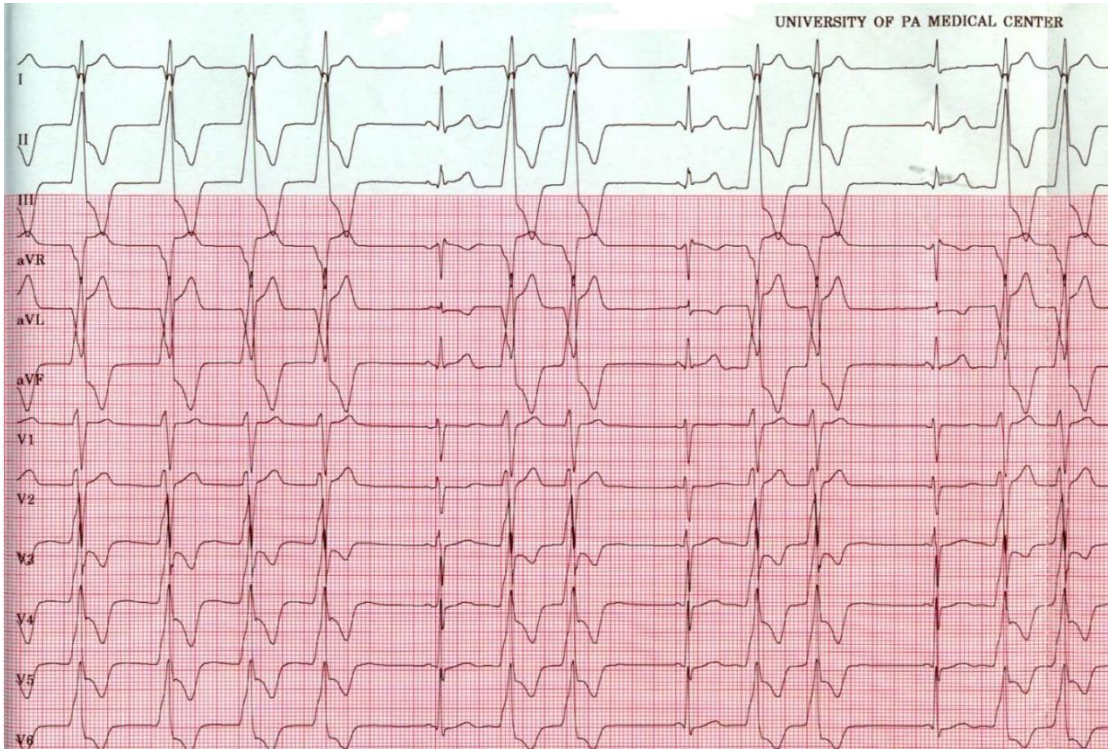
Distinction is important –RF ablation

Absence of an S wave in V5 or V6 -supravalvular

S wave in leads V5 and V6-infravalvular

AORTIC CUSP VT

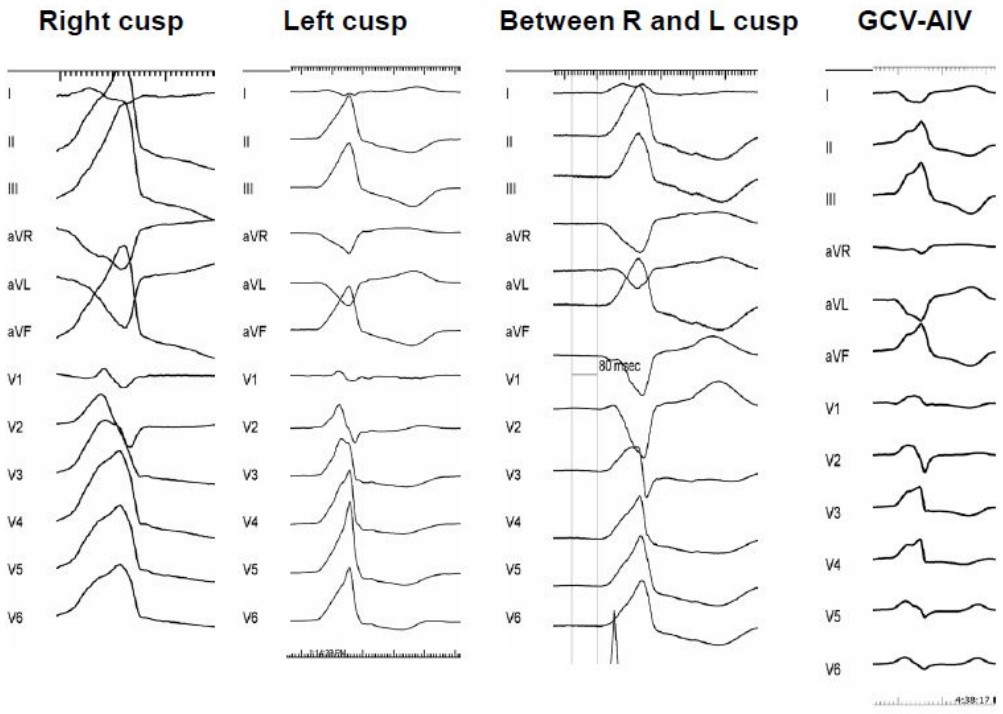
Depending on site of origin from right or left coronary cusp-LBBB or RBBB morphology



LBBB morphology with transition by V3, tall R waves in the inferior leads, and an s wave in lead I suggest the VPC from left coronary cusp.

Most VTs arise from left cusp and specifically from junction of left and right cusps

VT originating from LCC or aortomitral continuity often demonstrate terminal S wave in lead I

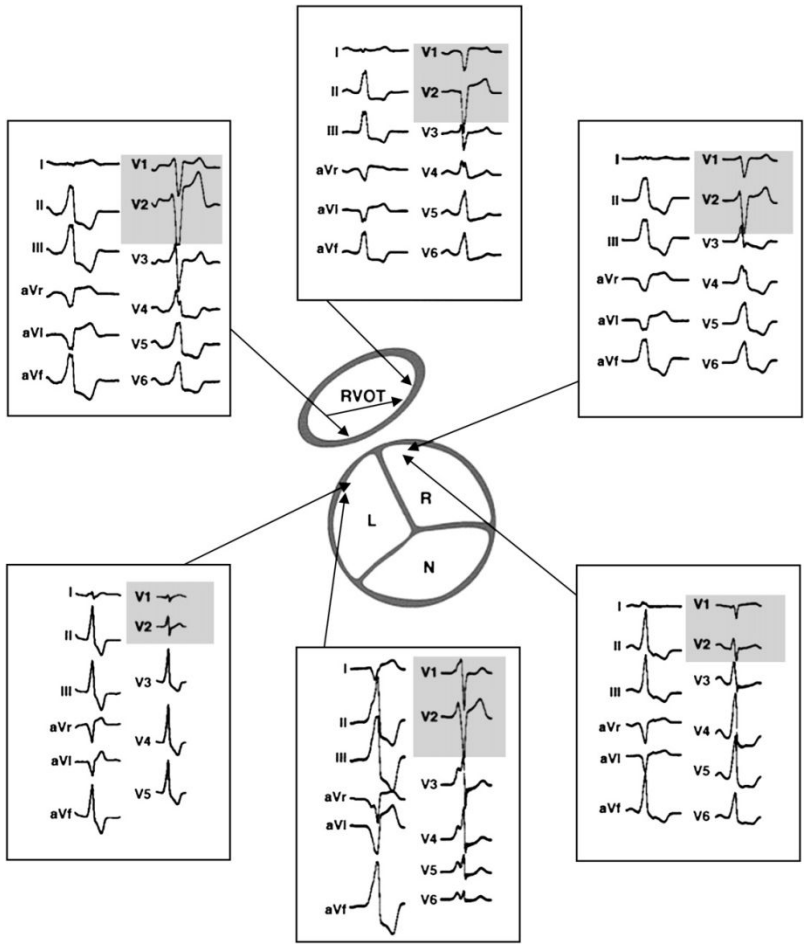


RVOT VT Vs aortic cusp VT

R wave duration and R/S wave amplitude ratio in leads V1 and V2 - Greater in tachycardias originating from cusp compared with RVOT

Precordial lead transition earlier in cusp VT occurring before lead V3

Absence of an S wave in V5 or V6 -specificity of 88% for cusp VT compared with RVOT VT



OUYANG AND COLLEAGUES

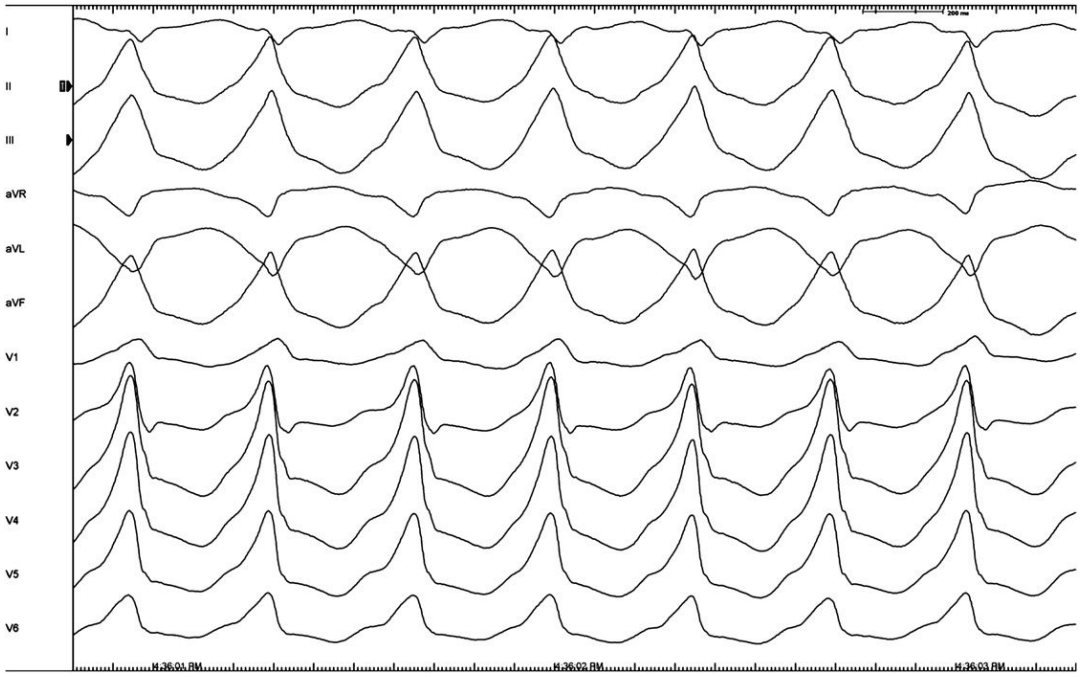
EPICARDIAL FOCI OF VA

OTVT originate from epicardial locations

9%–13% of idiopathic VT

Cluster along the course of the anterior interventricular vein and at its junction with great cardiac vein

Show catecholamine and adenosine sensitivity



Q wave in lead I and terminal S wave in V2 (Paper speed 100 mm/s).

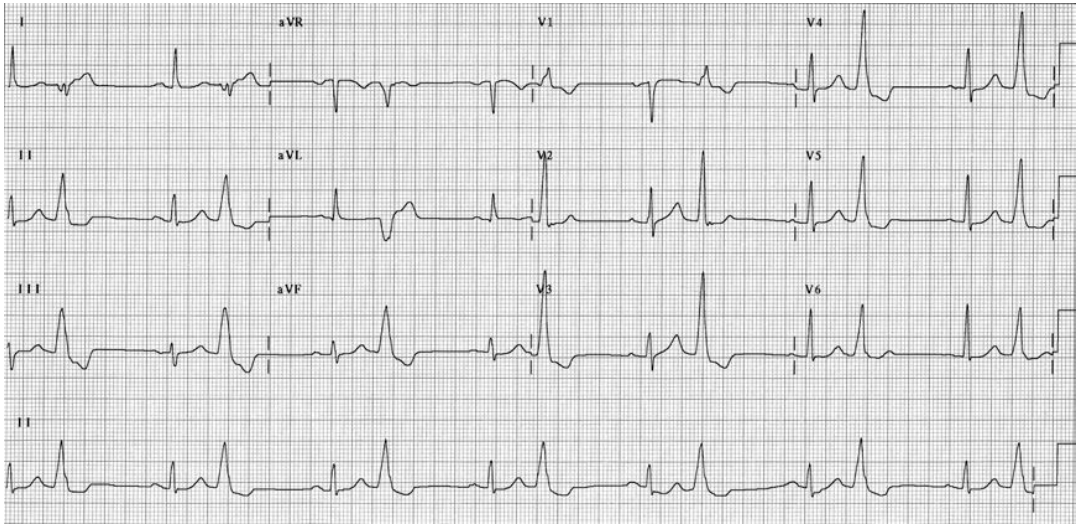
Psuedodelta wave

Interval from earliest QRS activation to earliest fast deflection in precordial leads (≥ 34 ms)

Precordial maximum deflection index

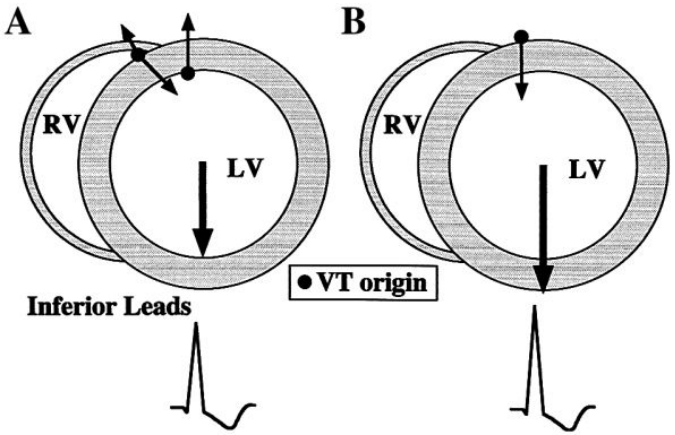
Beginning of QRS to earliest maximal deflection in any precordial leads / QRS duration. (> 0.55)

(sensitivity of 100%, specificity of 98%)



DANIELS AND COLLEAGUES

TADA AND COLLEAGUES



Epicardial compared with endocardial VT-R wave amplitude significantly greater in inferior leads

Lead I had an S wave as part of an rS or QS pattern

Q wave amplitude greater in aVL compared with aVR (ratio >1.4)

Q wave in lead I

MITRAL ANNULUS, TRICUSPID ANNULUS PAPILLARY MUSCLE PERIVASCULAR EPICARDIAL ECTOPY

MITRAL ANNULAR VT

Significant slurring of QRS complex onset resembling delta-wave

Regardless of where along circumference of mitral annulus VT originates ECG shows RBBB pattern across precordium

S wave in lead V6

More lateral site- more likely is presence of S wave in lead I and of notching in inferior leads

Posterior focus will have superior axis.

PARA-HISIAN

PVCs or VT also originate from RVOT along region of tricuspid annulus

Most common site is para-Hisian

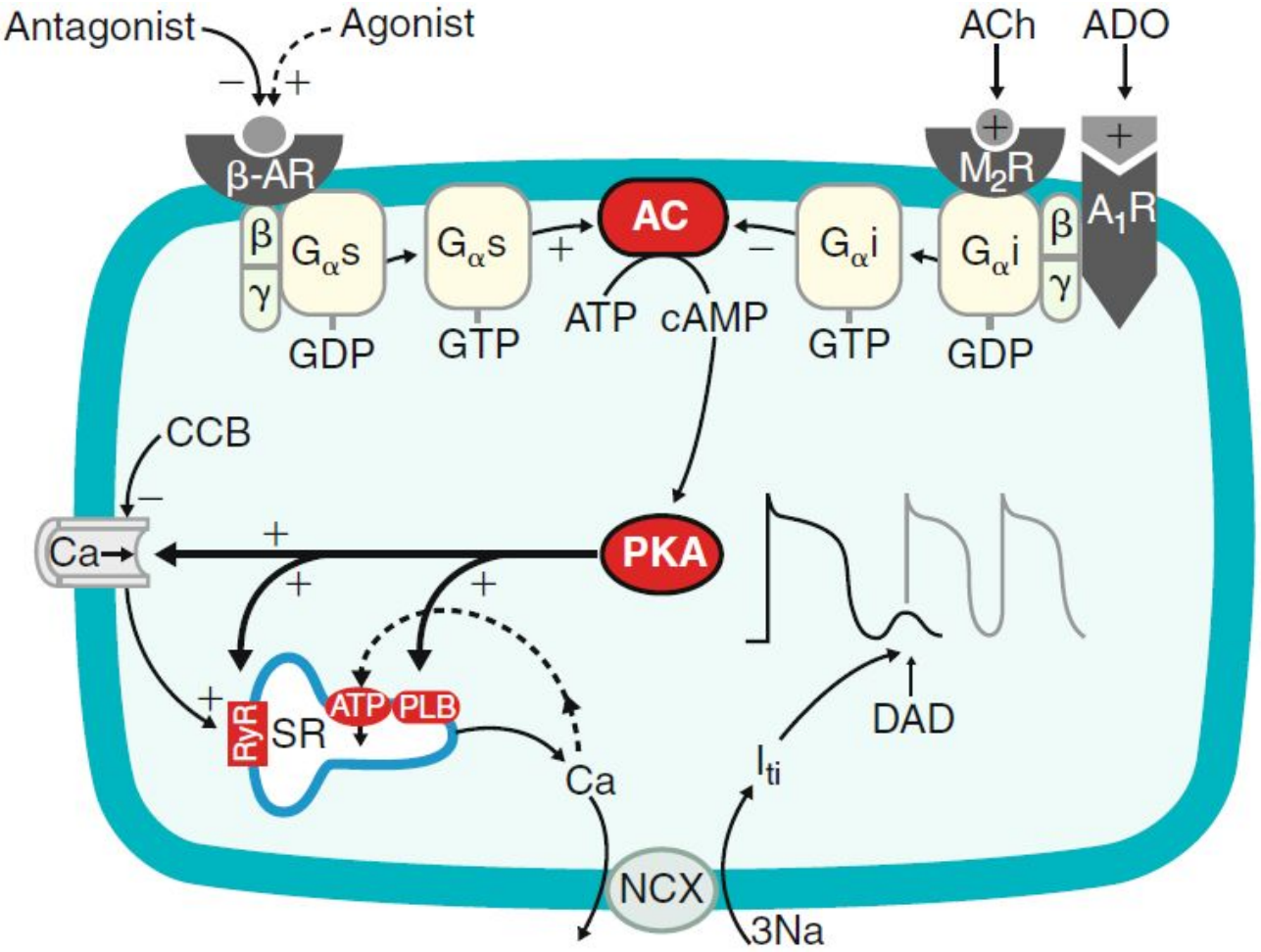
Characteristic ECG findings are

- Left bundle branch block pattern (Qs in lead V1)
- Early transition in precordial leads (V3)
- Narrower QRS complexes
- Inscription of an R wave in lead I and AvI
- Relatively small R wave in inferior leads

Sites of successful ablation record an atrial and a ventricular potential

ELECTROPHYSIOLOGIC MECHANISM

- Outflow tract VT is due to triggered activity
- Secondary to cyclic AMP mediated DAD
- Example-Exertion results in increased cyclic AMP due to beta receptor stimulation
- Release of calcium from sarcoplasmic reticulum and DAD
- Mutations in signal transduction pathways involving cAMP may be etiology for VT in some patients
- Tachycardia may terminate with Valsalva maneuvers, adenosine, BB or CCB


Antagonist
Agonist
ACh
ADO
β-AR
M2R
A1R
β
γ
Gαs
Gαs
AC
Gαi
Gαi
β
γ
GDP
GTP
ATP
cAMP
GTP
GDP
CCB
Ca
PKA
RyR
SR
ATP
PLB
Ca
Iti
DAD
NCX
3Na

CLINICAL FEATURES

- 20 and 40 years,Slight female preponderance
- May be asymptomatic but often present with palpitations, chest pain, dyspnea, presyncope and even syncope
- Occur more frequently with exertion or emotional stress
- Circadian variation- peaks at 7 AM and between 5 and 6 PM
- Women-symptoms related to changes in hormonal status
- Truly idiopathic OTVT is benign
- Small percentage of patients with very frequent VT –TCM
- Rare reports of cardiac arrest and PMVT

TREATMENT

May respond acutely to carotid sinus massage, Valsalva maneuvers or intravenous adenosine or verapamil

Long-term oral therapy with either BB or CCB

Nonresponders (33%)- class I or III antiarrhythmic agents

RFA

When medical therapy is ineffective or not tolerated

High success rate (>80%)

Ablation of epicardial or aortic sinuses of Valsalva sites is highly effective

Technically challenging and carries higher risks -proximity to coronary arteries

Tachycardia localization

12-lead ECG

Intracardiac activation

Pace mapping

BIPOLAR ACTIVATION MAPPING

OTVTs are mediated by triggered activity

Electrogram at site of origin typically precedes onset of QRS by approximately 20 msec

Exception -cusp VT, prepotentials (~50 msec) may be seen during VPCs that correspond to late potentials during sinus rhythm

PACE MAPPING

Useful because typically site of origin is focal and because underlying tissue is normal

Performed with a low output

Result in a small discrete area of depolarization

Mapping performed at site of origin of clinical arrhythmia,

ECG should mimic clinical arrhythmia perfectly (12/12, including notches)

ELECTROANATOMIC RE-CREATION OF 3D ANATOMY

Helpful for catheter mapping and localization of site of origin

Incessant VT- 3D anatomy should ideally be created during tachycardia which should be able to localize earliest site to a small region (<5 mm) with centrifugal activation

Typically pace mapping from this region should achieve a perfect match

Predictors for successful ablation

Single VT morphology

Accurate pace maps

Absence of a deltalike wave at beginning of QRS during tachycardia

Ability to use pace mapping and activation mapping

Some tachycardias arise from epicardium, necessitate ablation from great cardiac vein or epicardium itself using pericardial puncture technique

Coronary angiography is performed before ablation on epicardium or in aortic sinus

Complications during outflow tract VT ablation are rare

RBBB (1%)

Cardiac perforation

Damage to the coronary artery (LAD) - cusp region ablation

Overall recurrence rate is approximately 10%

IDIOPATHIC LEFT VT

Three varieties

left posterior fascicular VT -RBBB and LAD (90%)

left anterior fascicular VT -RBBB and RAD

high septal fascicular VT -relatively narrow QRS and normal axis

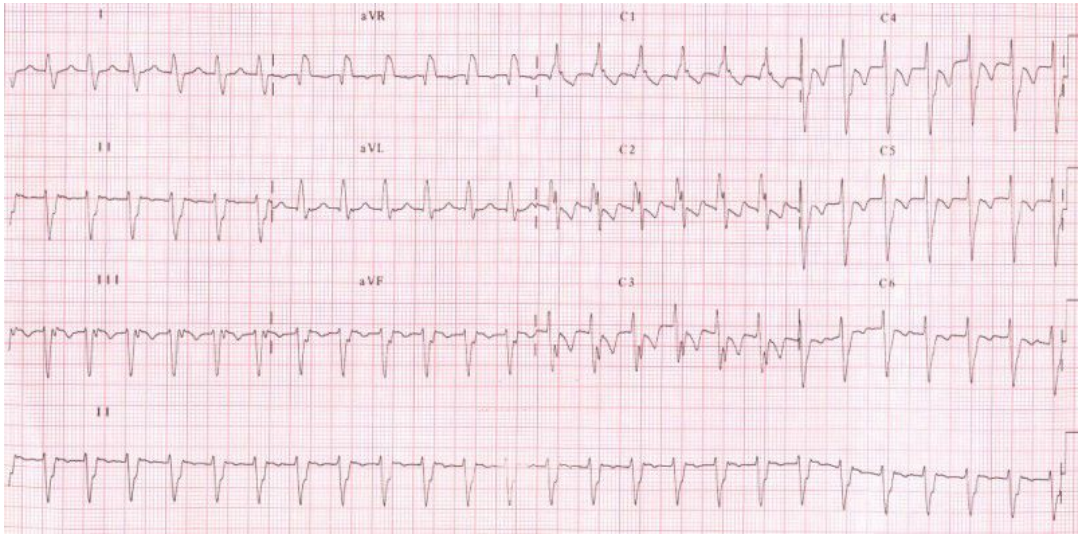
15 to 40 years

More in men (60%)

Most occur at rest

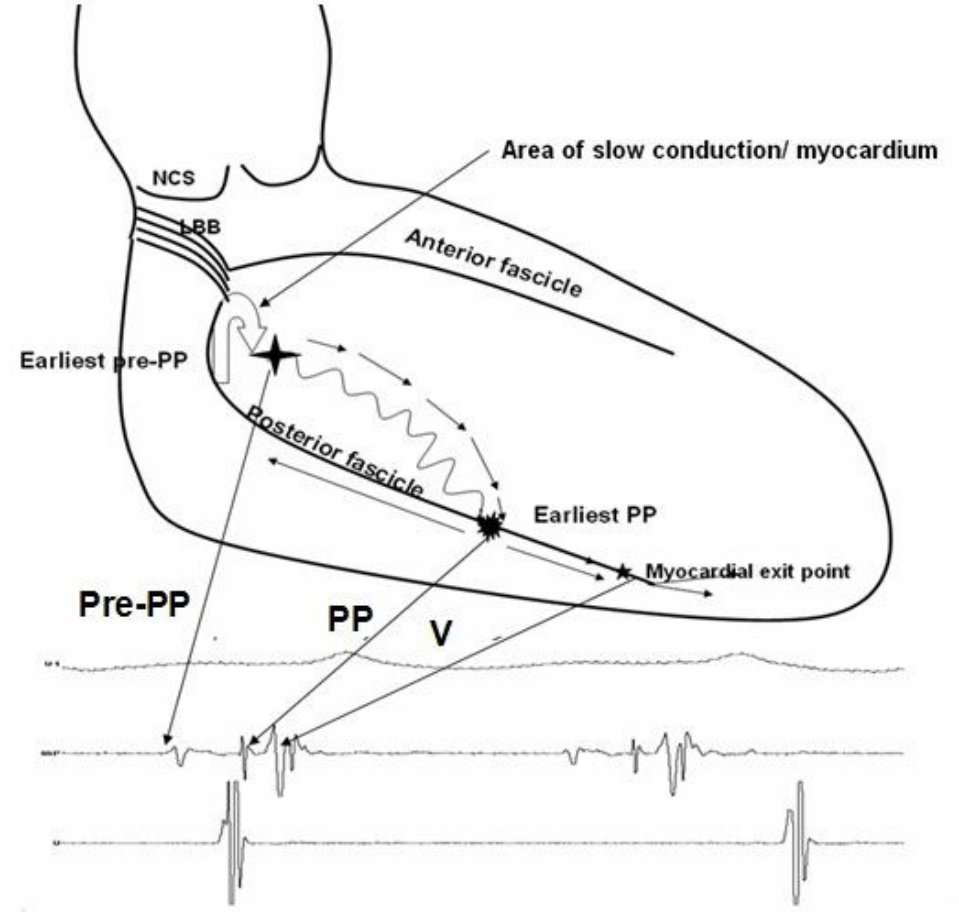
Usually paroxysmal

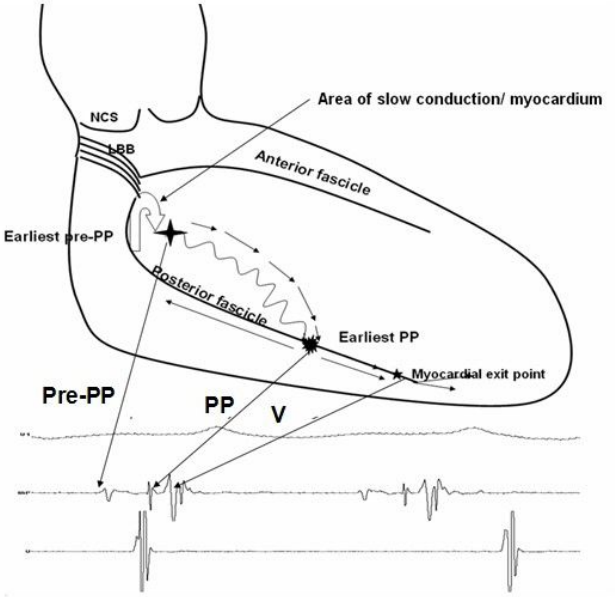
Incessant forms leading to TCM are described



ELECTROPHYSIOLOGIC MECHANISM

- Re-entrant mechanism
- Orthodromic limb -zone of slow, decremental conduction in intraventricular left septum proceeding from base to apex
- Lower turnaround point is toward the apex
- Retrograde limb is formed by Purkinje network





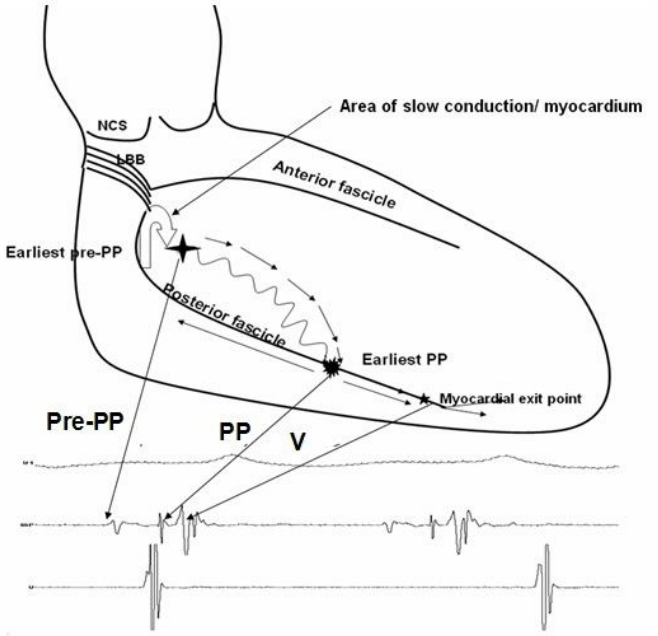
During VT two distinct potentials can be observed before ventricular electrogram

Purkinje potential (PP or P2)-activation of LPF or Purkinje fiber near LPF

Relative activation time of PP to onset of QRS complex 5-25 ms

Brief, sharp, high-frequency potential preceding onset of QRS during tachycardia

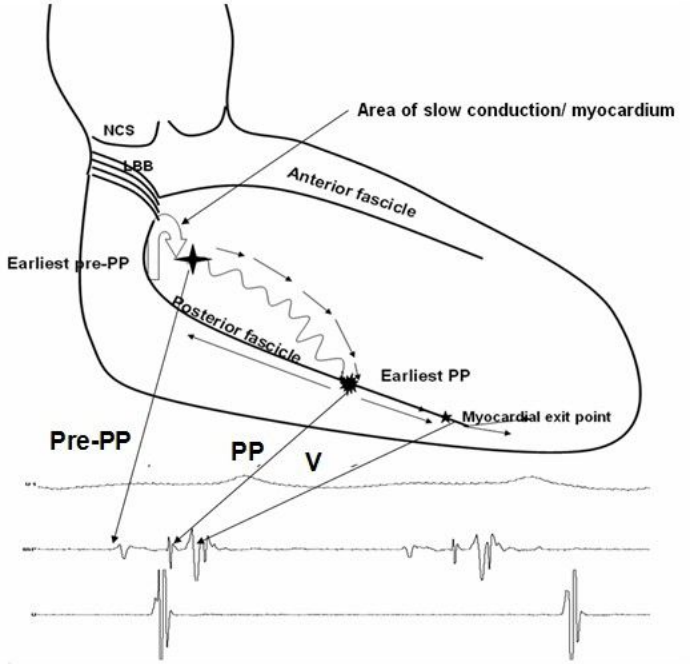
Nakagawa and colleagues



Pre Purkinje potential (Pre-PP or P1) Represents excitation at entrance to specialized zone in ventricular septum which has decremental properties and is sensitive to verapamil

Relative activation times of pre-PP to onset of QRS complex is 30-70 ms

Pre-PP is a dull, lower frequency potential preceding the PP during tachycardia

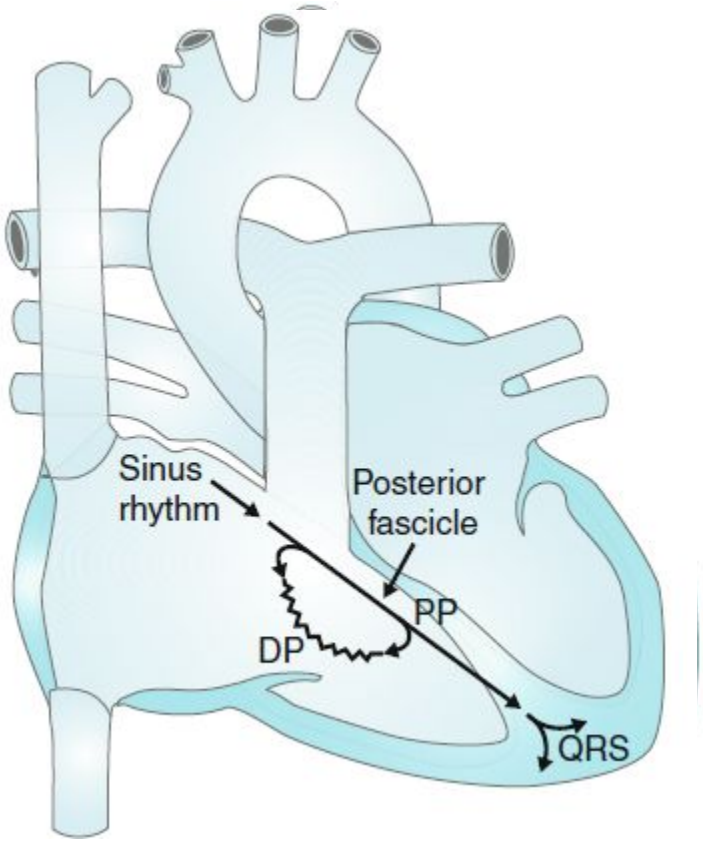


Nakagawa and colleagues

Reentrant circuit of fascicular tachycardia is completed by a zone of slow conduction between Pre PP and PP areas in basal interventricular septum

Upper turn around point of circuit

Located close to the main trunk of LBB



Area is captured antidromically during tachycardia and at higher pacing rates-pre-PP precedes PP during tachycardia.

Captured orthodromically in sinus rhythm and at relatively lower pacing rates- pre PP follows ventricular complex

Nakagawa and colleagues

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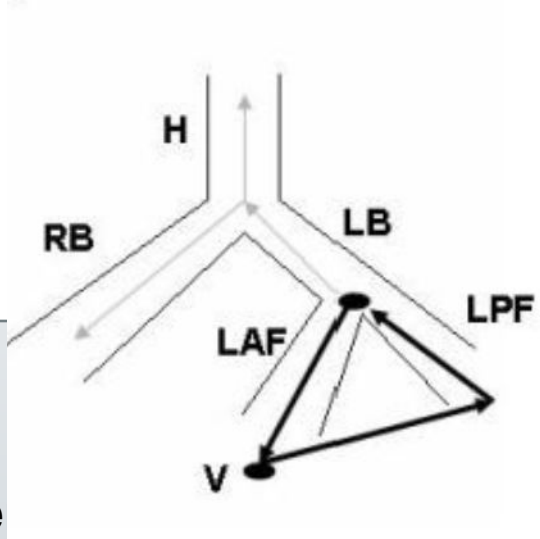


Supraventricular tachycardia with aberrancy

VA dissociation,

EP-Rapid atrial pacing during tachycardia can de dissociation

Interfascicular VT(typical RBBB morphology and left or right axis deviation)

Common in patients with AWMI and either LAHB or LPHB

EP -ventricular depolarization is preceded by His bundle depolarization in interfascicular VT which is not seen in fascicular VT

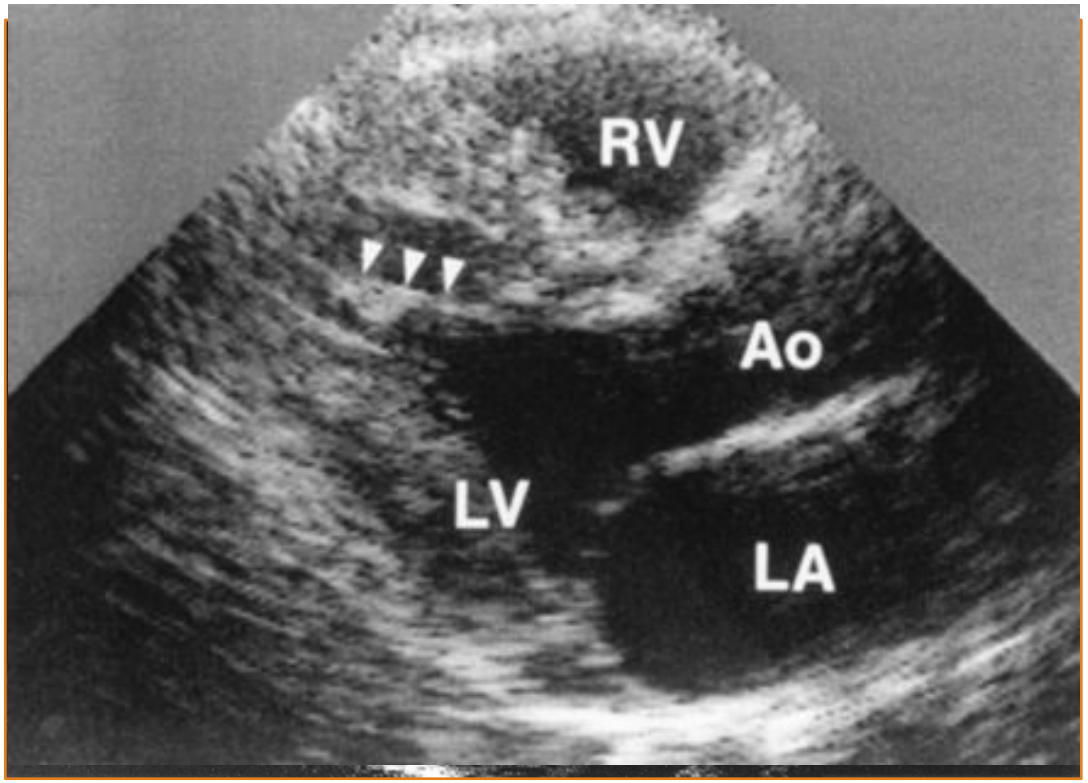
Idiopathic mitral annulus VT (RBBB morphology with right axis deviation)

VT originates from a false tendon extends from posteroinferior left ventricle to basal septum

Resection of tendon or ablation at septal insertion site eliminate tachycardia

Exact role tendon is unclear

Specificity is low



Gallagher JJet al. AJCardiol 1988;61(2):27A–44A
Merliss AD, Seifert MJ, Collins RF, etal Electrophysiol 1996;19(12 Pt 1):2144–6.
Thakur RK, Klein GJ, Sivaram CA, et al.Circulation 1996;93(3):497–501.

ECG

Baseline 12-lead ECG is normal in most patients

Exit near the area of the left posterior fascicle

- RBBB + left superior frontal plane axis
- Relatively narrow QRS duration (<140 msec)
- RS interval <80 msec

Exit near the area of the left anterior fascicle

- RBBB+ right frontal plane axis

- Long-term prognosis is very good
- Patients who have incessant tachycardia may develop tachycardia related cardiomyopathy
- Intravenous verapamil is effective in acutely terminating VT
- Mild to moderate symptoms oral –verapamil
- BB and class I and III antiarrhythmic agents useful in some
- Medical therapy is often ineffective in patients who have severe symptoms

RADIOFREQUENCY ABLATION

Associated with significant symptoms or who are intolerant or resistant to medical therapy

Strategies employed to identify the ideal site for ablation

- Pace mapping
- Endocardial activation mapping
- Identifying diastolic Purkinje potentials (MC approach)
- Identifying late diastolic potentials
- When VT is noninducible-ablation during sinus rhythm using electroanatomic mapping may be considered

LIFE-THREATENING (TYPICALLY POLYMORPHIC VT)

Rare

Generally occurs with genetic ion channel disorders

Associated with SCD

Abnormalities exist at molecular level

LIFE-THREATENING (TYPICALLY POLYMORPHIC VT)

Long QT Syndrome

Brugada Syndrome

CPVT

Short QT Syndrome

LONG QT SYNDROME

Corrected QT interval 440 ms in men and 460 ms in women with or without morphological abnormalities of the T waves

Decrease in outward potassium currents or increase in inward sodium currents

Prolongs repolarization phase of cardiac action potential

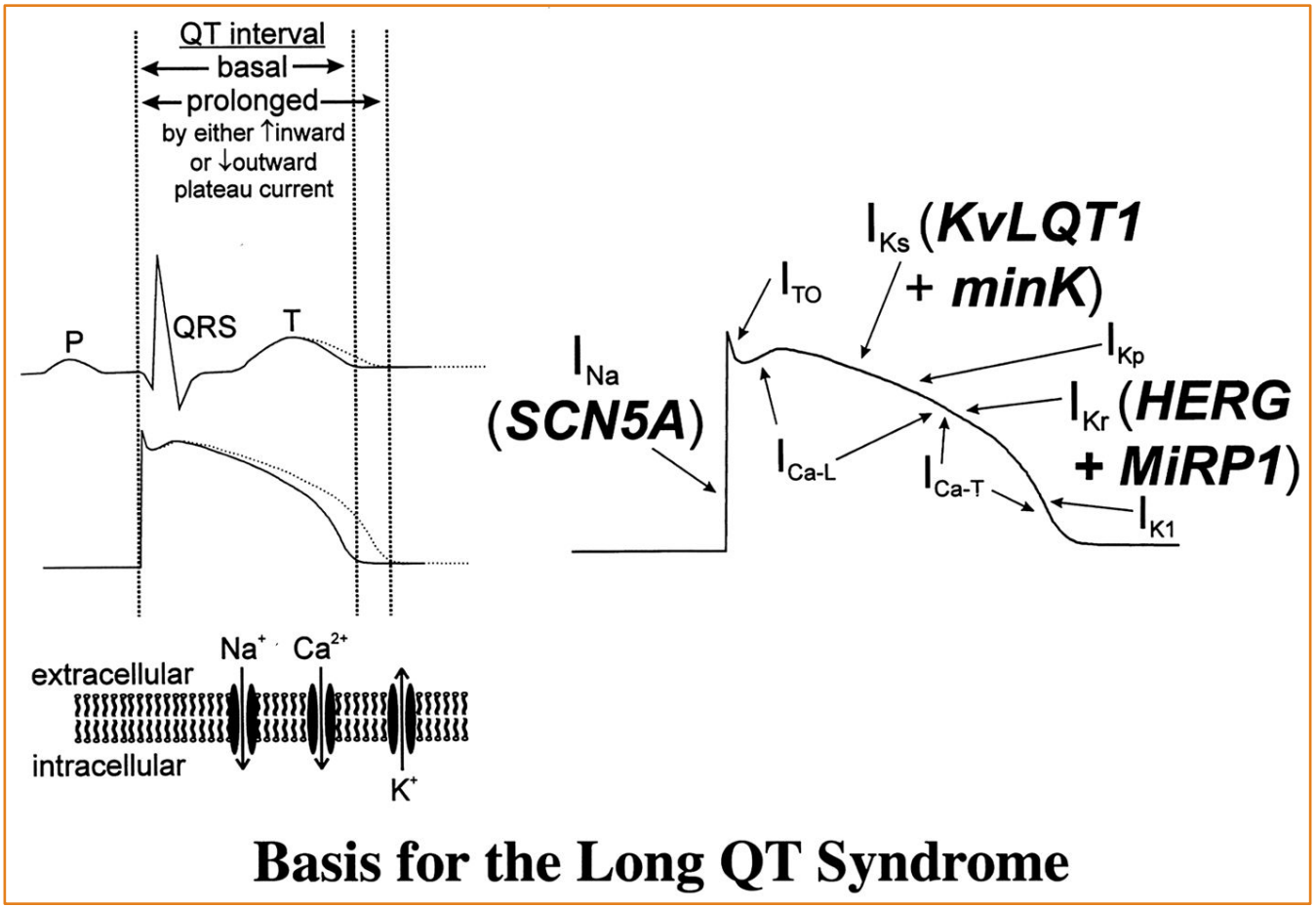
Result in prolongation of QT interval

Predisposition to EAD and torsade de pointes VT

Twelve different genes described

LQT1, LQT2, and LQT3 account for 90%

BASIS FOR THE LONG QT SYNDROME



Basis for the Long QT Syndrome

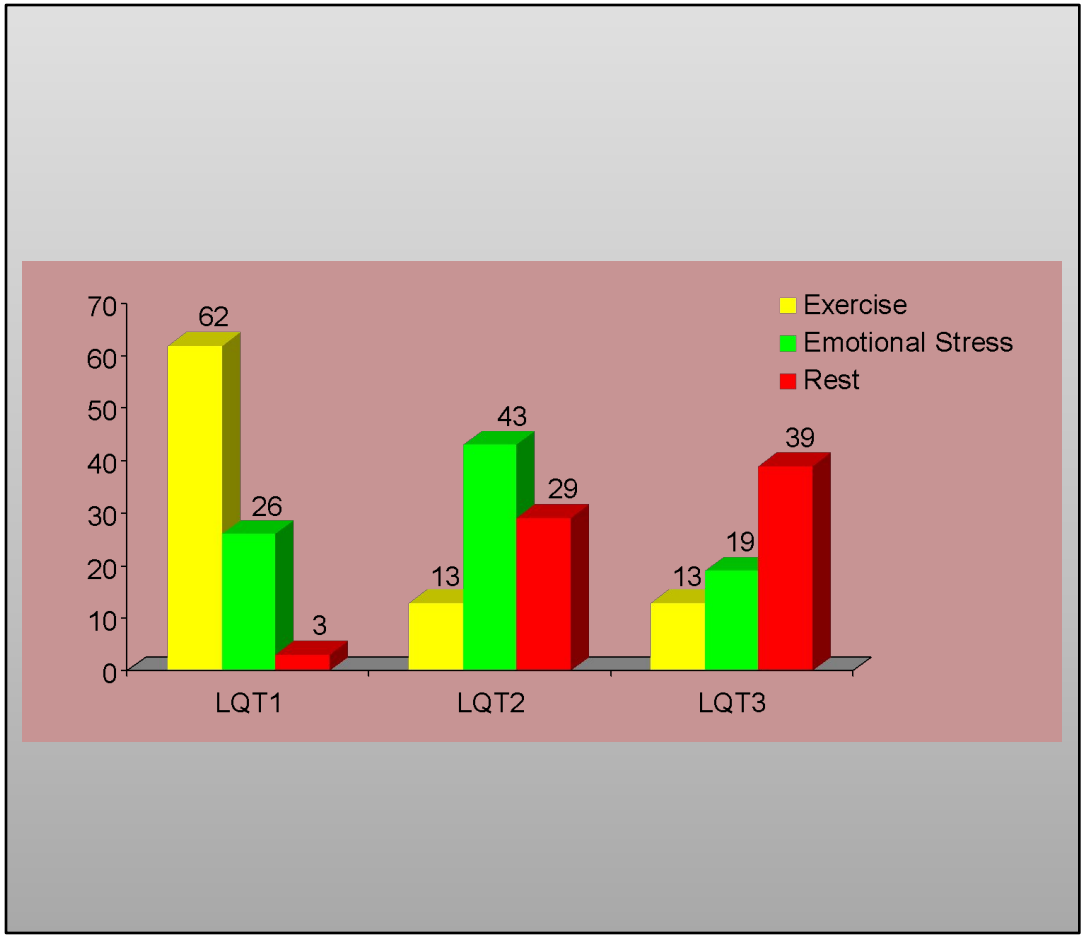
LQTS subtype	Mutated gene	Ionic current affected	Clinical frequency, %	Trigger of clinical event
LQT1†	*KVLQT1*	I_{Ks} decrease	43	Exercise
LQT2	*HERG*	I_{Kr} decrease	45	Sudden arousal
LQT3	*SCN5A*	I_{Na} increase	7	Sleep
LQT4	Ankyrin B	I_{Na}, possible late sodium current increase	Rare	
LQT5†	*KCNE1* (minK)	I_{Ks} decrease	Rare	
LQT6	*KCNE2* (MiRP1)	I_{Kr} decrease	Rare	
LQT7	*KCNJ2*	$I_{Kir2.1}$ decrease	Rare	

Approximately 25% not have identifiable gene mutations

Mean age of symptom onset is 12 years

Present with syncope, seizures, or cardiac arrest.

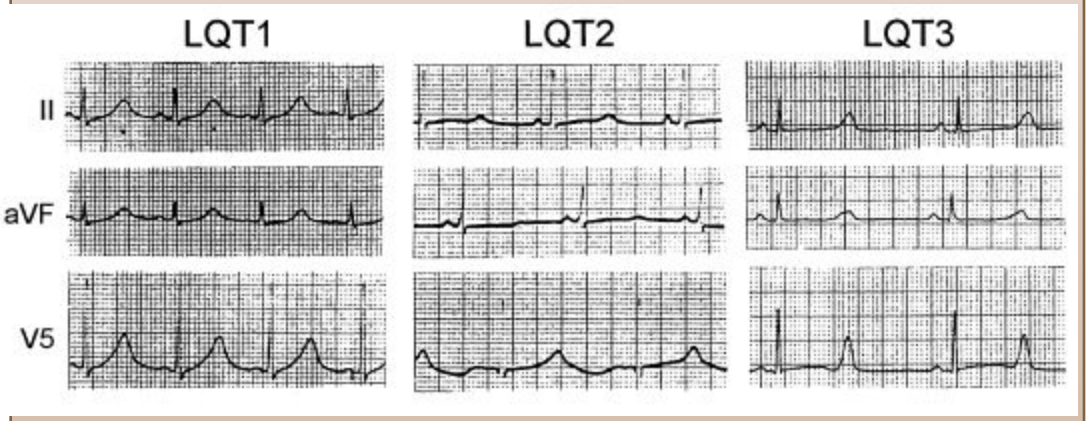
Clinical presentation and ECG repolarization (ST-T) patterns have been correlated to genotype



LQT1 -often have broad-based T waves

LQT2- T-wave is often notched in multiple leads.

LQT3- Demonstrate long ST segments



Modified Schwartz score for the diagnosis of LQTS

Variable	Points
ECG findings	
QTc ms ≥ 480	3
460–470	2
450 (in males)	1
Torsade de pointes	2
T wave alternans	1
Notched T wave in three leads	1
Low heart rate for age	0.5
Clinical history	
Syncopy	
With stress	2
Without stress	1
Congenital deafness	0.5
Family history	
Family members with definite LQTS	1
Unexplained sudden cardiac death < age 30 years among immediate family members	0.5

Score ≥3.5 : Definite LQTS
Score 2-3: Intermediate probability
Score ≤1: Low probability

Schwartz PJ. The congenital long QT syndromes from genotype to phenotype: Clinical implications. J Intern Med 2006;259(1):39–47.

MANAGEMENT

- Avoid trigger events and medications prolong QT interval
- Risk stratification -degree of QT prolongation, genotype and sex
- Corrected QT interval >500 ms -high risk
- LQT1 and LQT2 -higher risk of events than LQT3
- Risk of events-higher during adulthood in females and during adolescence in males
- Number of mutations increase the risk
- Once a clinical event occurs (syncope or survival after sudden cardiac death), recurrence is frequent

MEDICATIONS PROLONG QT INTERVAL

Antiarrhythmic: procainamide, quinidine, amiodarone, sotalol

Antihistamine: astemizole, terfenadine

Antimicrobial/antifungal: trimethoprim sulfa, erythromycin, ketoconazole

Psychotropics: haloperidol, risperidone, thioridazine, tricyclics

Other: epinephrine, diuretics, cisapride, bepridil, ketanserin

MANAGEMENT

BB -patients with syncope and asymptomatic patients with significant QT prolongation

Role of BB in asymptomatic with normal or mildly prolonged QT -uncertain

BB are highly effective in LQT1, less effective in others

Role of BBs in LQT3 is not established

Preferable is non selective BB

MANAGEMENT

ICD are indicated for secondary prevention of cardiac arrest and for patients with recurrent syncope despite BB therapy

Less defined therapies

Gene specific therapy -mexiletine , flecainide or ranolazine (LQT3)

PPI for bradycardia-dependent torsade depointes

Surgical left cardiac sympathetic denervation for recurrent arrhythmias resistant to BB therapy

Catheter ablation of triggering PVCs-abolish recurrent VT/VF

BRUGADA SYNDROME

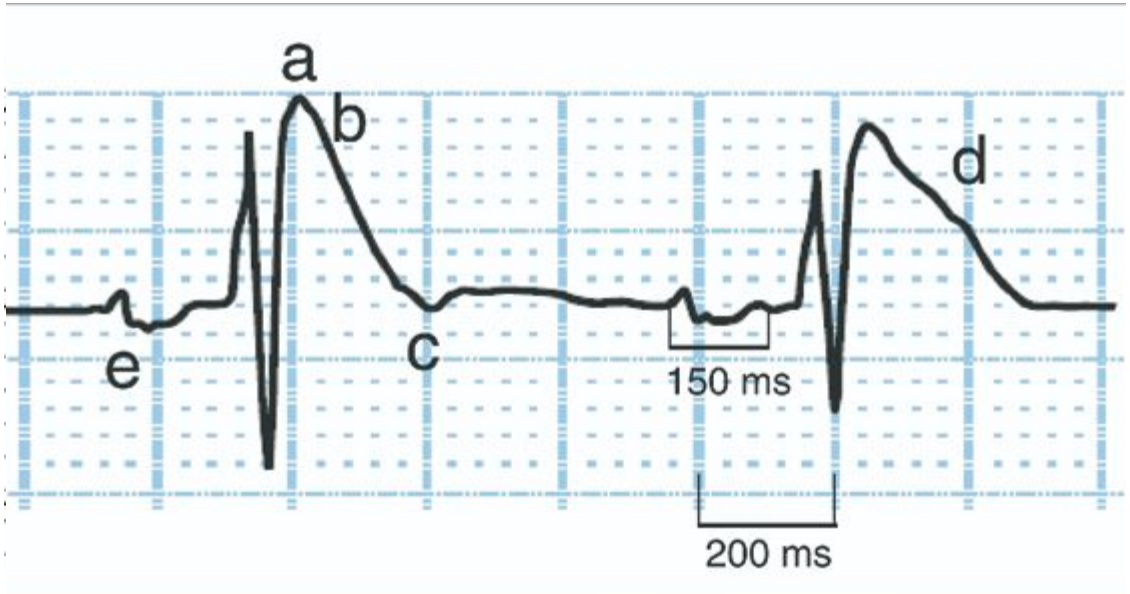
Characterized by ST-segment elevation in V1 to V3

Inverted T wave

2 mm in 2 of these 3 leads are diagnostic

Complete or incomplete RBBB pattern

Abnormally prolonged and biphasic P



ST-SEGMENT ABNORMALITIES IN LEADS V1 TO V3

	Type 1	Type 2	Type 3
J wave amplitude	≥2 mm	≥2 mm	≥2 mm
T wave	negative	positive or biphasic	positive
ST-T configuration	coved type	saddleback	saddleback
ST segment (terminal portion)	gradually descending	elevated ≥1 mm	elevated <1 mm

1 mm=0.1 mV. The terminal portion of the ST segment refers to the latter half of the ST segment.

Typical ECG pattern can be transient and may only be detected during long-term ECG monitoring

Methods to document type-1 ECG

Move V1 lead from fourth intercostal space to second

Take an ECG after a large meal -positive in approximately 50% of patients

Provoked by sodium-channel blocking agents-

ajmaline, flecainide or procainamide

CLINICAL PRESENTATION

- 0.12% to 0.14% in general population
- Syncope or cardiac arrest
- Predominantly in men in third and fourth decade
- SCD in young men,typically occurs at night
- Prone to atrial fibrillation and sinus node dysfunction
- Precipitated by a febrile state, vagotonic agents, a-adrenergic agonists,BBs, TCAs, hypokalaemia,alcohol and cocaine toxicity

Risk of SCD with Brugada syndrome is substantial

Risk of recurrent events during 4 years of follow-up

62% for those with cardiac arrest

19% for those with syncope.

Asymptomatic group -8% event rate during 2 years

Brugada P, Brugada R, Brugada J. Sudden death in patients and relatives with the syndrome of right bundle branch block, ST segment elevation in the precordial leads V(1) to V(3) and sudden death. Eur Heart J 2000;21:321-6.

TREATMENT

Drugs inhibit Ito (such as quinidine) and increase calcium current (such as isoproterenol) are effective

Lowdose quinidine may be used to treat frequent VAs in patients who already have an ICD

Quinidine and isoproterenol may be useful in VT storms

Catheter ablation of triggering PVCs and ablation of RVOT epicardial musculature successful in abolishing recurrent VT/VF in a small number of patients

Dimethyl lithospermate B (Danshen's extract)

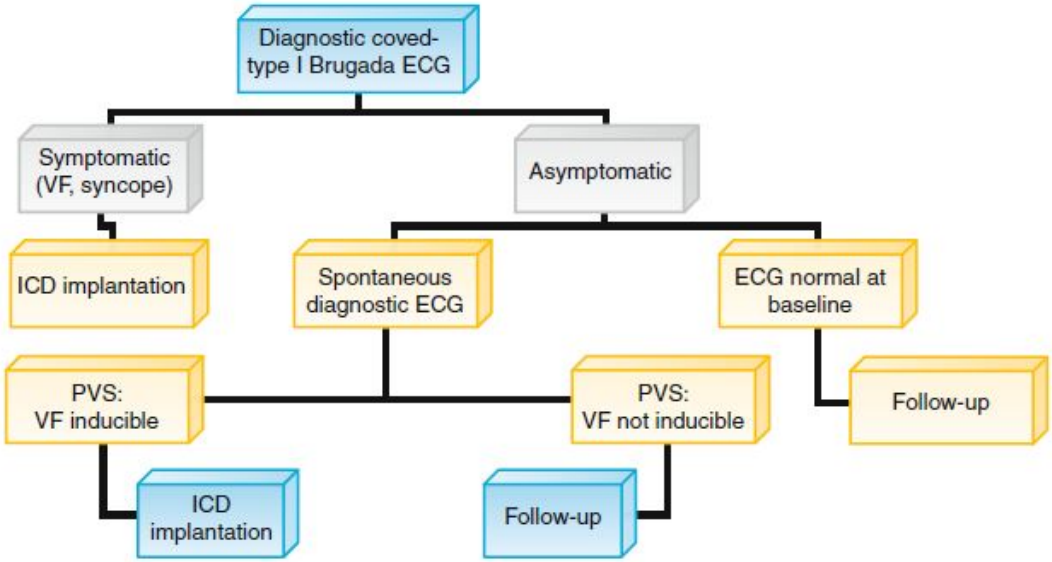
cilostazol

ICD

ICD are effective in preventing SCD

Indicated for cardiac arrest survivors

Patients with spontaneous ECG pattern and syncope are at high risk - ICD insertion is generally recommended for primary prophylaxis



Different genes involved

SCN5A gene mutations (BrS1) - loss of function of cardiac sodium channel (NaV 1.5) account for majority

BrS1 and LQT3 share SCN5A mutations

Mutation of the ankyrin-binding motif of Nav1.5

Mutation of glycerol-3-phosphate dehydrogenase 1-like (GPD1-L) gene

Mutations cardiac L-type calcium channel genes

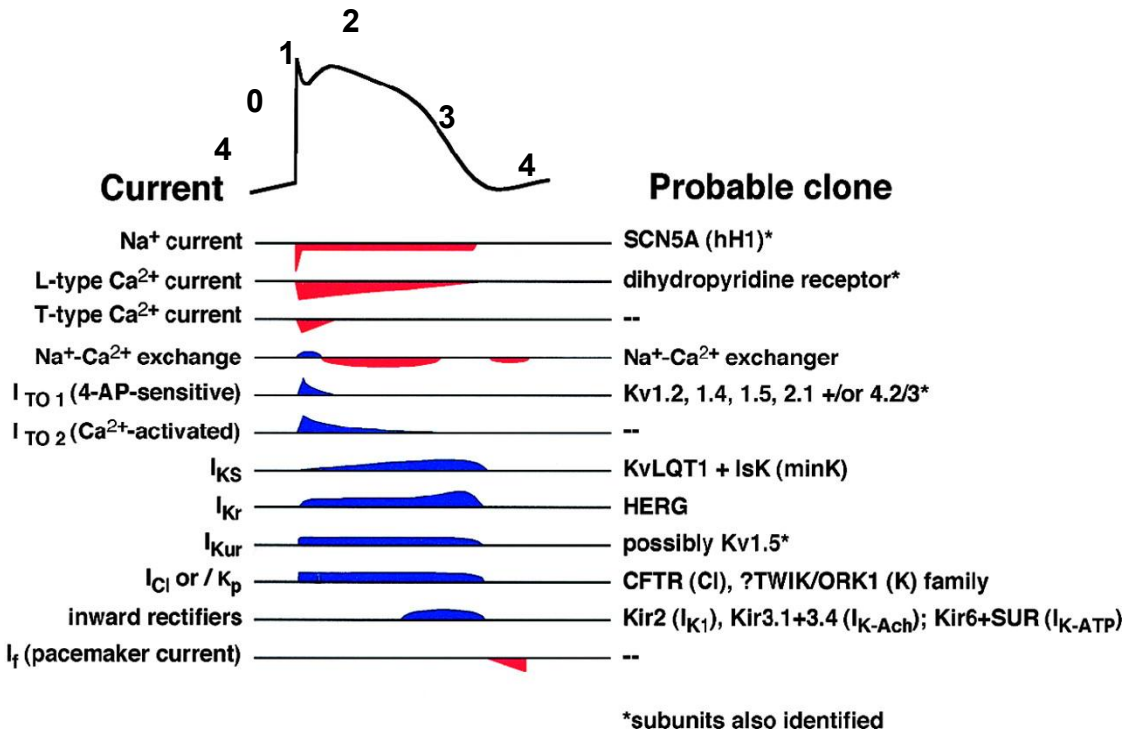
SCN5A GENE

Codes for cardiac sodium channel that opens during phase 2 of the action potential

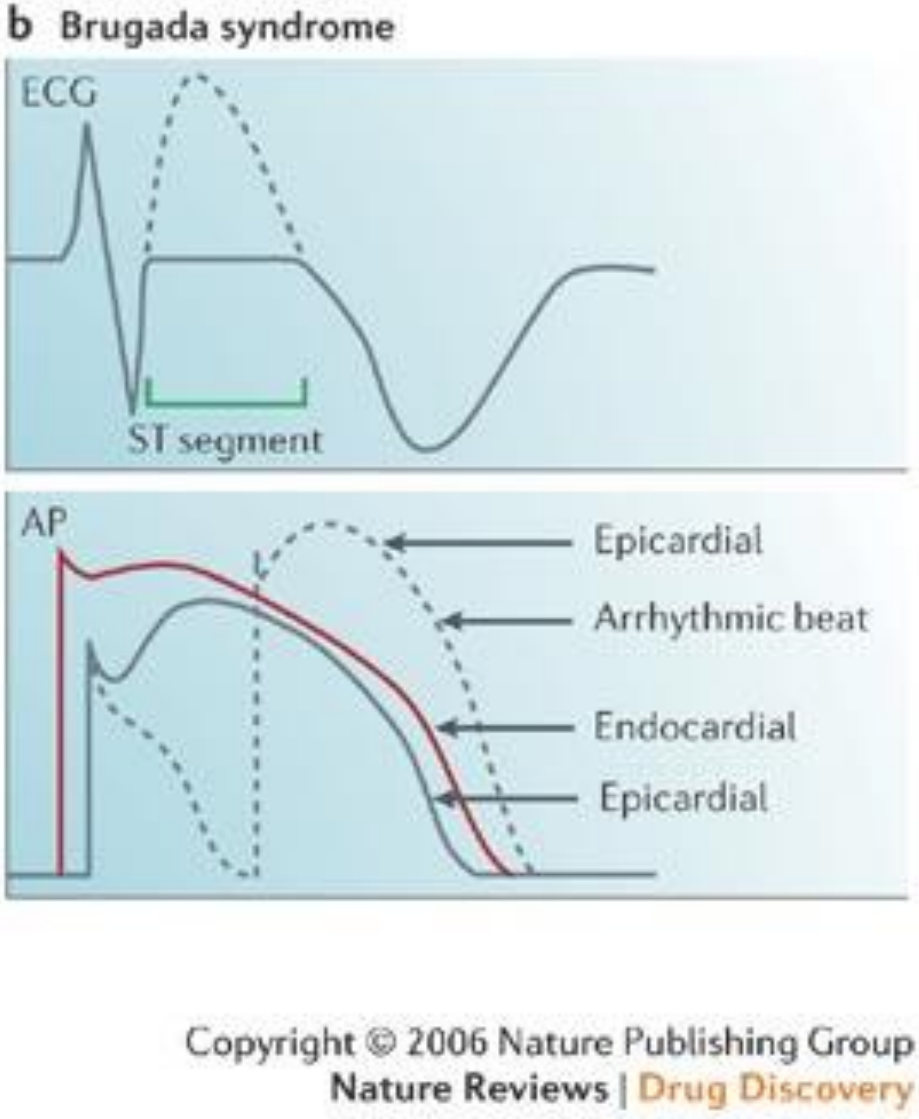
In Brugada, it opens poorly in RV epicardial cells

Autosomal dominant inheritance

20-30% of cases have abnormal SCN5A gene



Priori, S. G. et al. Circulation 1999;99:674-681



Defective sodium channels: shorter AP (phase 0), deeper notch (phase I), and shorter phase 2.

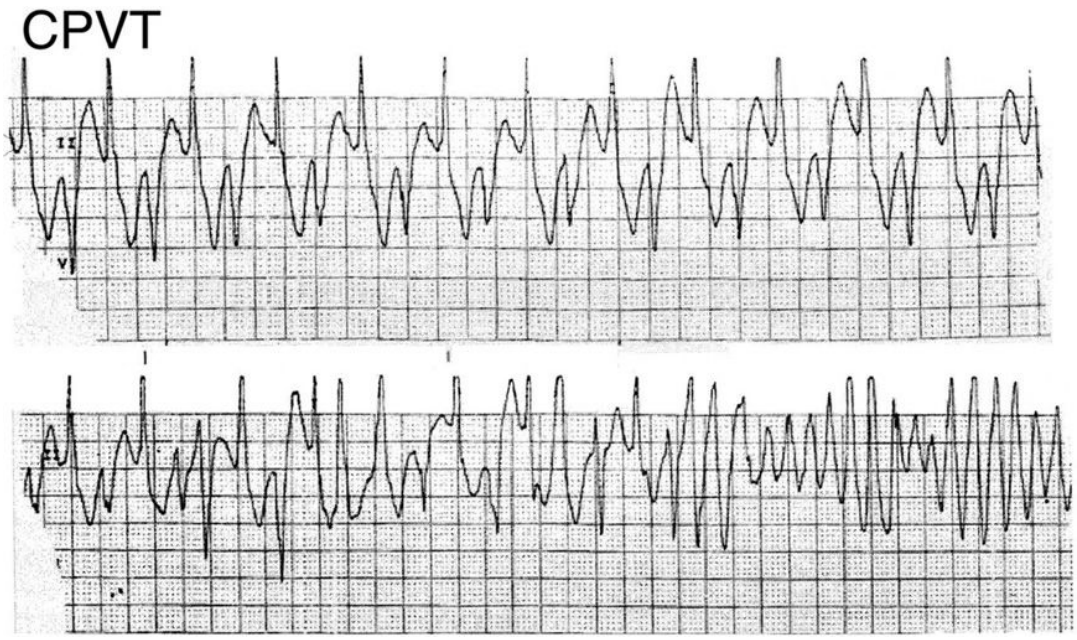
Creates juxtaposition of depolarized and repolarized cells, setting up possibility of PHASE 2 RENTRY, closely grouped PVCs, and VT or V Fib.

On EKG, ST segment not at baseline because no longer have uniform depolarization of the entire ventricle.

Nattel and Carlsson *Nature Reviews Drug Discovery* **5**, 1034–1049 (December 2006) | doi:10.1038/nrd2112

CATECHOLAMINERGIC PMVT

- Disorder of myocardial calcium homeostasis
- Clinically manifested as exertional syncope and SCD due to exercise induced VT
- Often polymorphic or bidirectional



Autosomal dominant (50%)-mutation of cardiac ryanodine receptor (RyR2 gene)

Autosomal recessive (3% to 5%)-mutations of calsequestrin 2 gene (CASQ2)

Ryanodine receptor spans membrane of sarcoplasmic reticulum

Releases calcium triggered by calcium entry into cell through L-type calcium channels

Calsequestrin-protein sequestrates calcium ions within sarcoplasmic reticulum

RyR2 and CASQ2 mutations cause intracellular calcium overload and DAD -basis of arrhythmogenesis

Resting ECG is unremarkable, prominent U waves may be seen

Typical VT patterns are reproducible with exercise or catecholamine infusion

VAs typically appear during sinus tachycardia rates of 120 beats/min to 130 beats/min, with progressive frequency of PVCs followed by bursts of polymorphic or bidirectional VT

Mean age for presentation with syncope is 4 years

EP study is not helpful in risk stratification

Medical management-BB

46% may have recurrent events while receiving therapy

CCB -limited effectiveness

Flecainide (blocks RyR2 receptor) also used

ICD

Cardiac arrest

Life-threatening VA despite maximal medical therapy

Initial ICD shock with its accompanying pain and anxiety may trigger further VAs

Surgical left cardiac sympathetic denervation -resistant cases

SHORT QT SYNDROME

- Rare disorder
- Characterized by short QT intervals of 300 to 320 ms
- Shortening or absence of the ST segment, with T wave initiating immediately from S wave
- Diagnostic criteria involving corrected QT interval, clinical history, and genotyping
- Syndrome is associated with SCD and atrial fibrillation
- Patients may present early in childhood

Subtype	Inheritance	Locus	Ion Channel	Gene	Electrophysiologic Characteristics of Mutant Current/Channel	Net Effect of Mutation
SQT1	AD	7q35	I_{Kr}	*KCNH2, HERG*	Shift of voltage dependence of inactivation of I_{Kr} by +90 mV out of the range of the action potential	Gain of function of I_{Kr}
SQT2	AD	11p15	I_{Ks}	*KCNQ1, KvLQT1*	Shift of voltage dependence of activation of I_{Ks} by −20 mV and acceleration of activation kinetics	Gain of function of I_{Ks}
SQT3	AD	17q23.1–24.2	I_{K1}	*KCNJ2, Kir2.1*	Increase in outward I_{K1} at potentials between −75 mV and −45 mV	Gain of function of I_{K1}
SQT4	AD	12p13.3	I_{Ca}	*CACNA1C, $Ca_v1.2$*	Decrease in amplitude of the inward calcium current	Loss of function of I_{Ca}
SQT5		10p12.33	I_{Ca}	*CACNB2b, $Ca_v\beta_{2b}$*		

ICD implantation for secondary and primary prevention

Preliminary observations suggest quinidine might be useful

IDIOPATHIC PROPRANOLOL-SENSITIVE VT (IPVT)

Usually occurs by fifth decade of life

Can arise from LV or RV

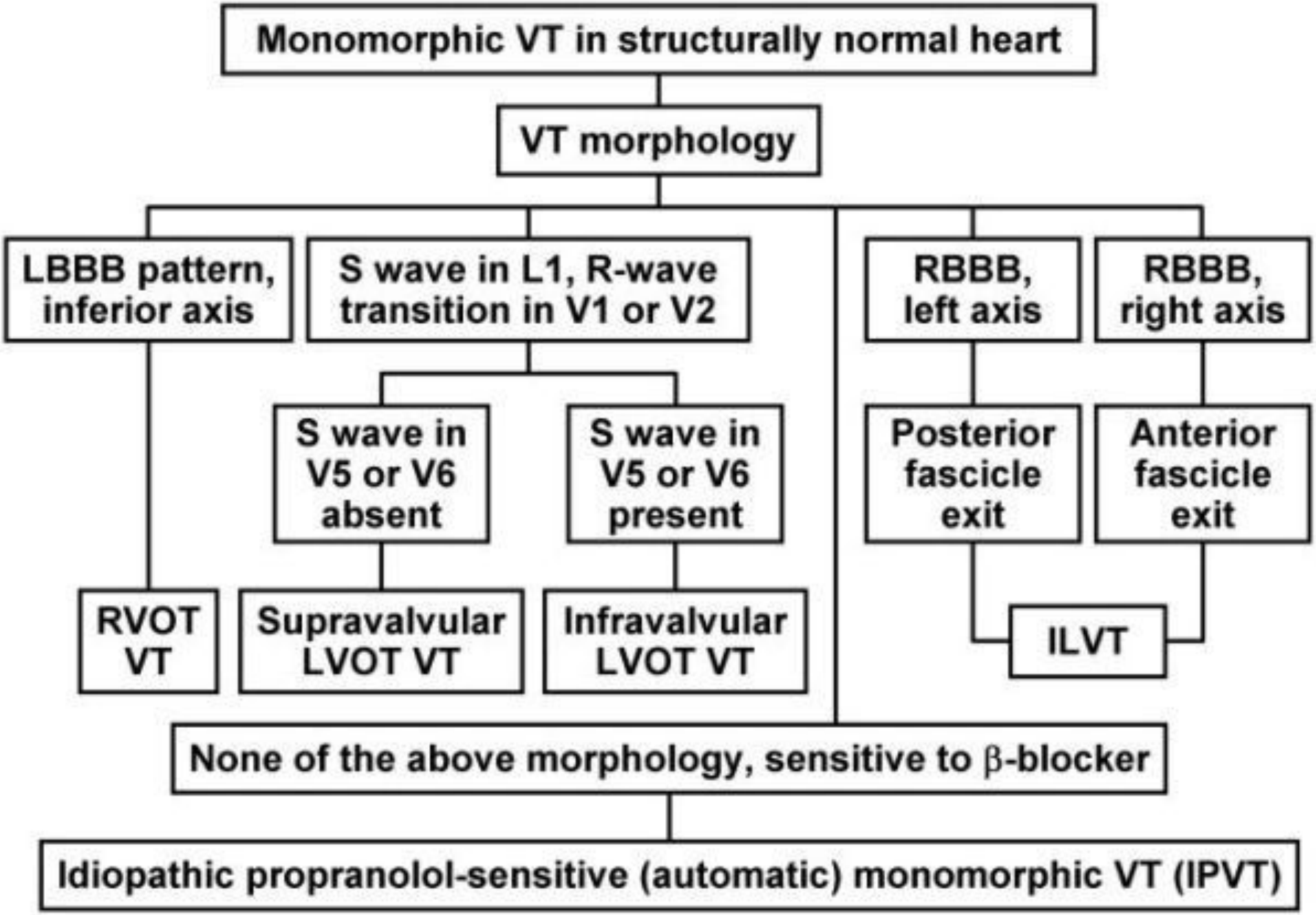
Morphology may be monomorphic or polymorphic

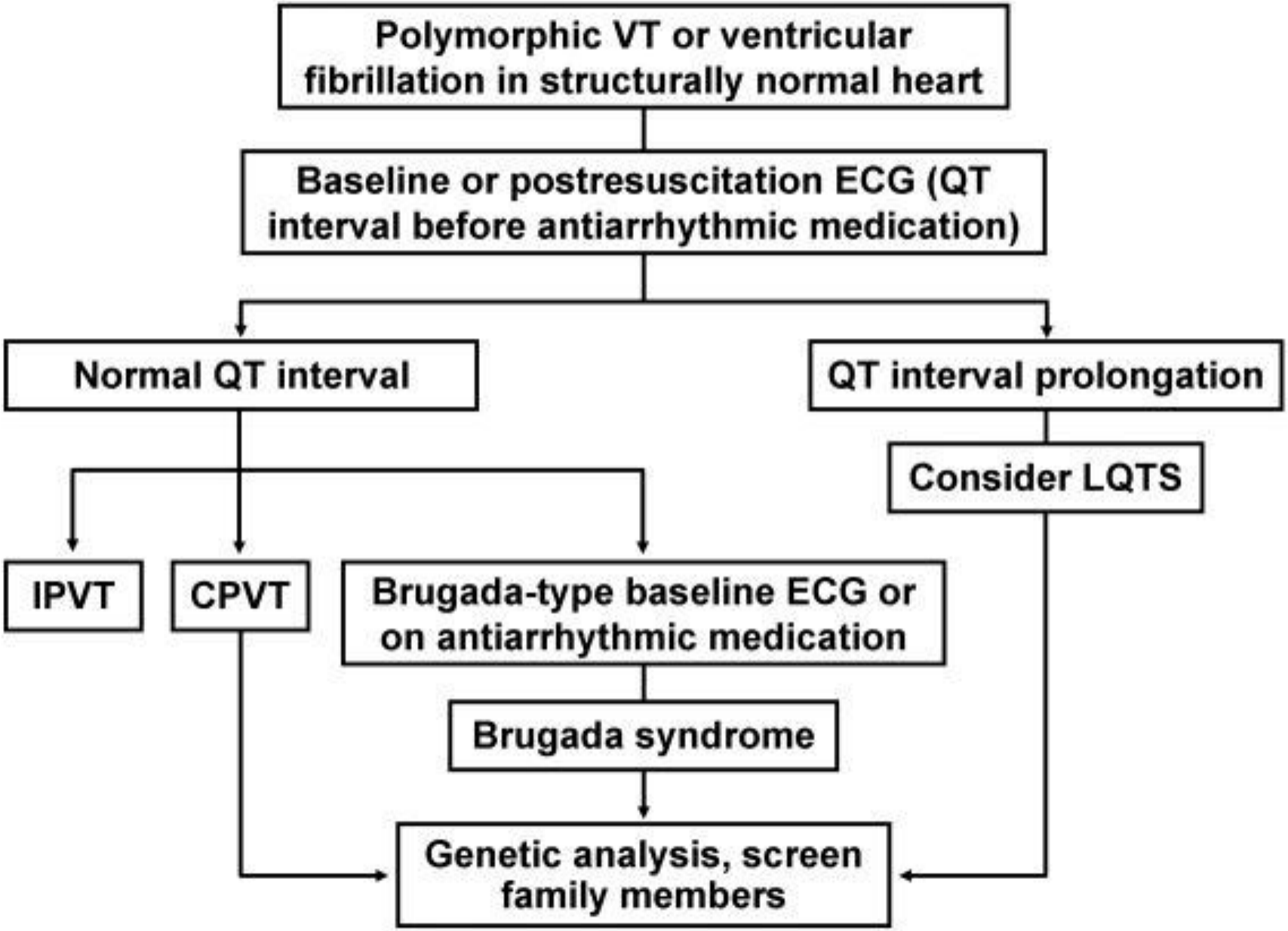
Not inducible with programmed stimulation

Isoproterenol infusion usually induces

TREATMENT OF IPVT

- BBs effective in acute situations
- Insufficient information available regarding long-term management
- Survivors of sudden cardiac death may receive ICD


Monomorphic VT in structurally normal heart
VT morphology
LBBB pattern, inferior axis
S wave in L1, R-wave transition in V1 or V2
RBBB, left axis
RBBB, right axis
S wave in V5 or V6 absent
S wave in V5 or V6 present
Posterior fascicle exit
Anterior fascicle exit
RVOT VT
Supravalvular LVOT VT
Infravalvular LVOT VT
ILVT
None of the above morphology, sensitive to β-blocker
Idiopathic propranolol-sensitive (automatic) monomorphic VT (IPVT)


Polymorphic VT or ventricular fibrillation in structurally normal heart
Baseline or postresuscitation ECG (QT interval before antiarrhythmic medication)
Normal QT interval
QT interval prolongation
Consider LQTS
IPVT
CPVT
Brugada-type baseline ECG or on antiarrhythmic medication
Brugada syndrome
Genetic analysis, screen family members

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Vol. 59, No. 20, 2012 ISSN 0735-1097 JACC


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