

# 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**

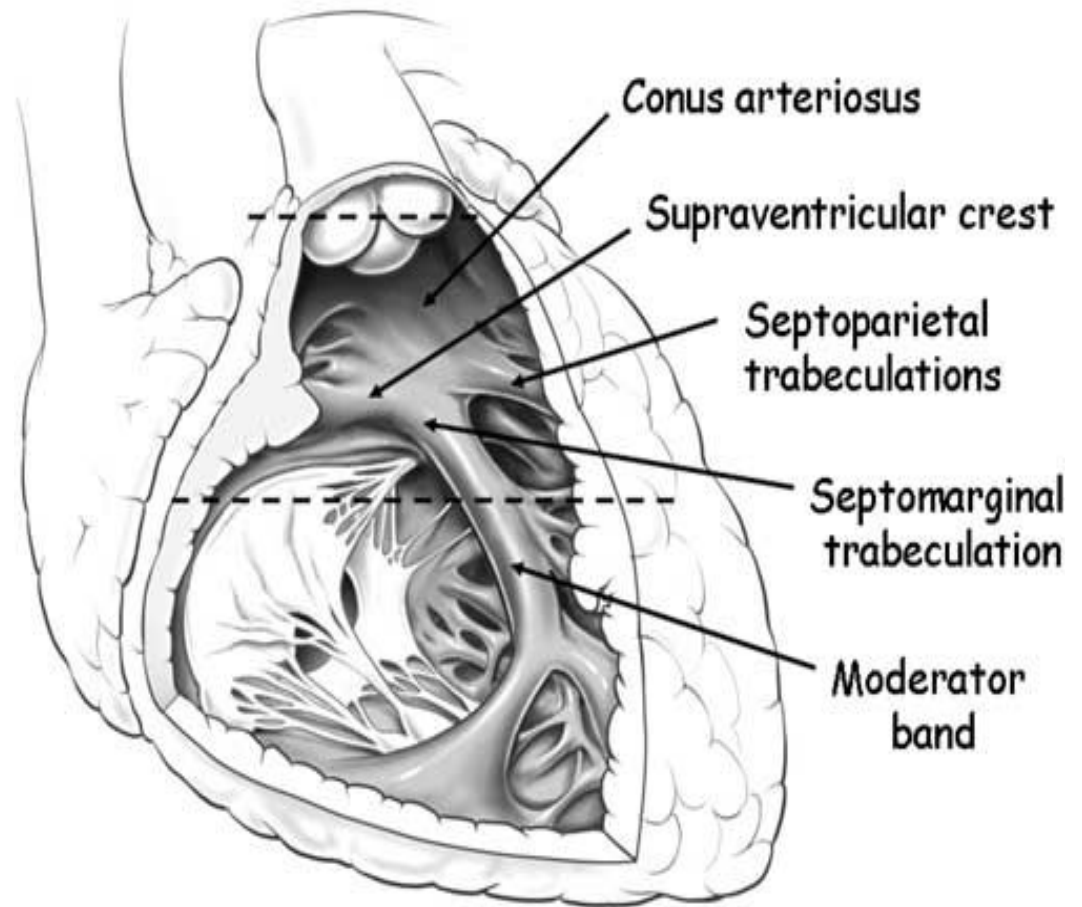
## ANATOMIC CORRELATES

**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**

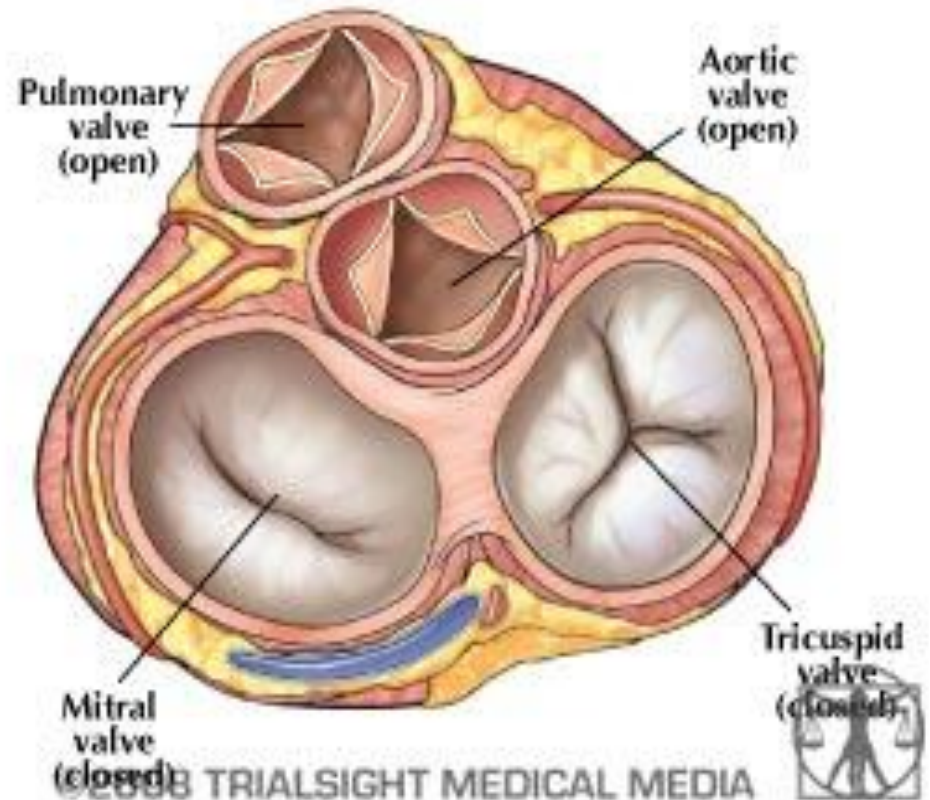


**LVOT is region of LV between anterior cusp of mitral valve and ventricular septum**

**Muscular and fibrous parts**

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

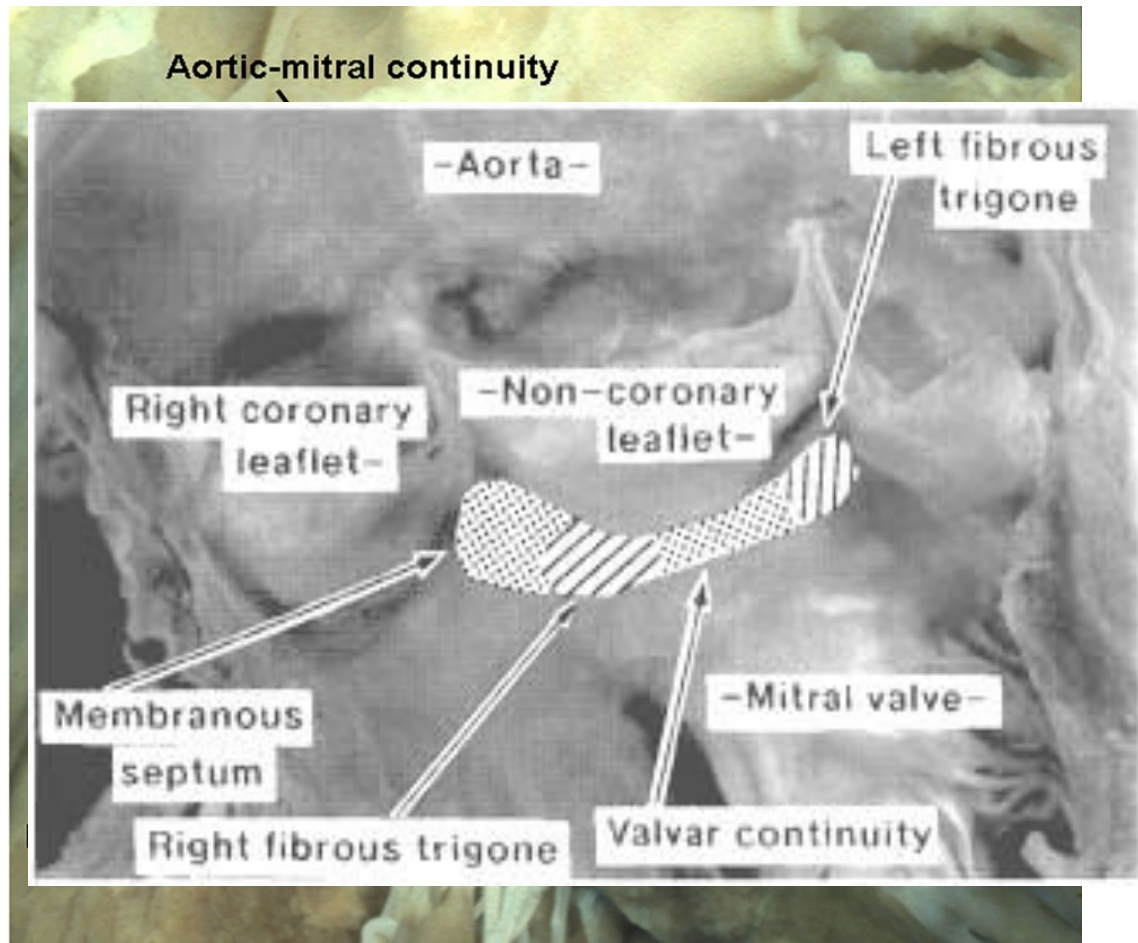
**Close proximity to AV node and His bundle -early activation in VT**





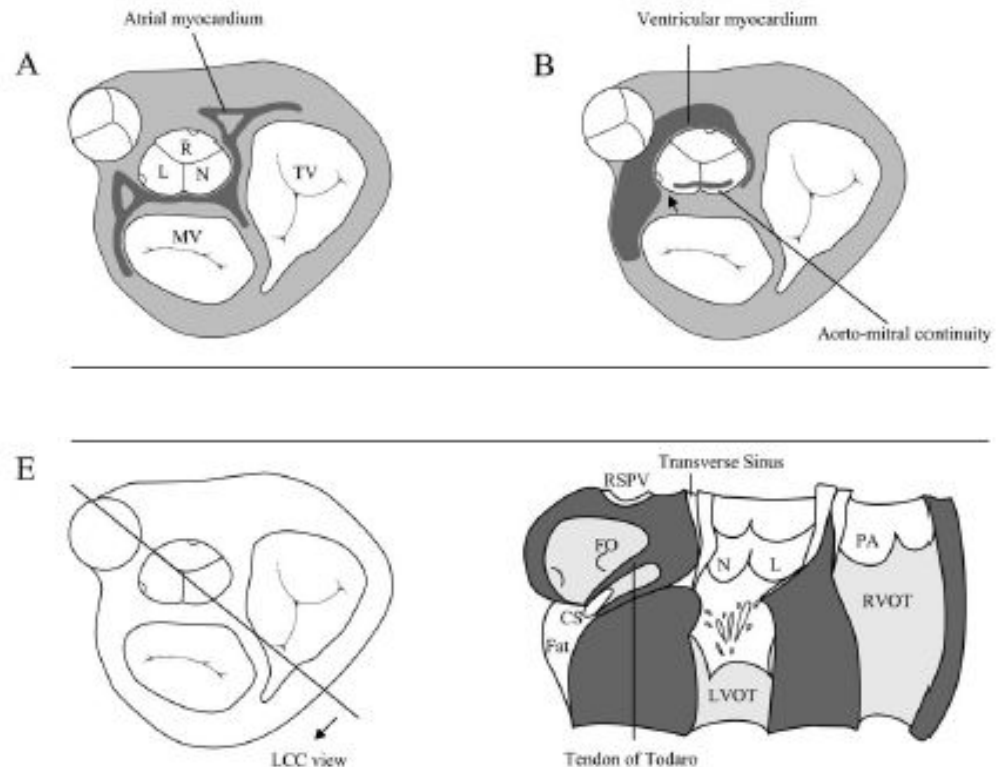
**Non-coronary cusp and posterior aspect of left coronary cusp are continuous with fibrous aortomitral continuity**

**Explain lack of VT related to the non-coronary cusp**

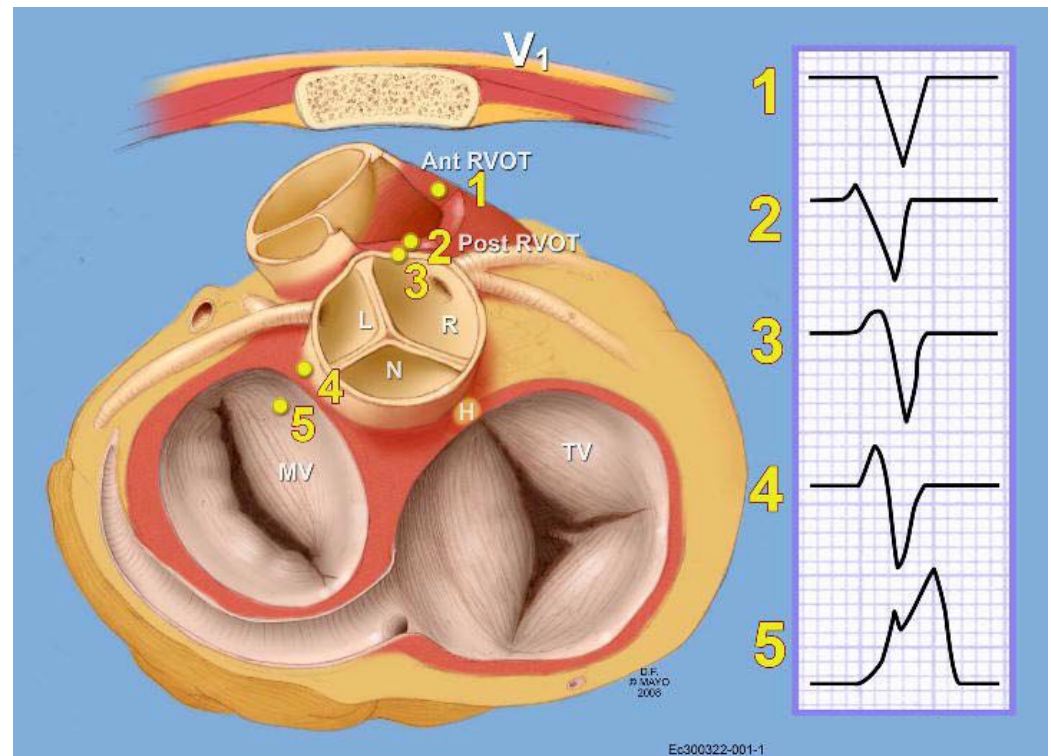


**VT from aortic sinuses of Valsalva arise from muscular extensions of the LVOT to areas above the base of the aortic valve cusps**

**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**

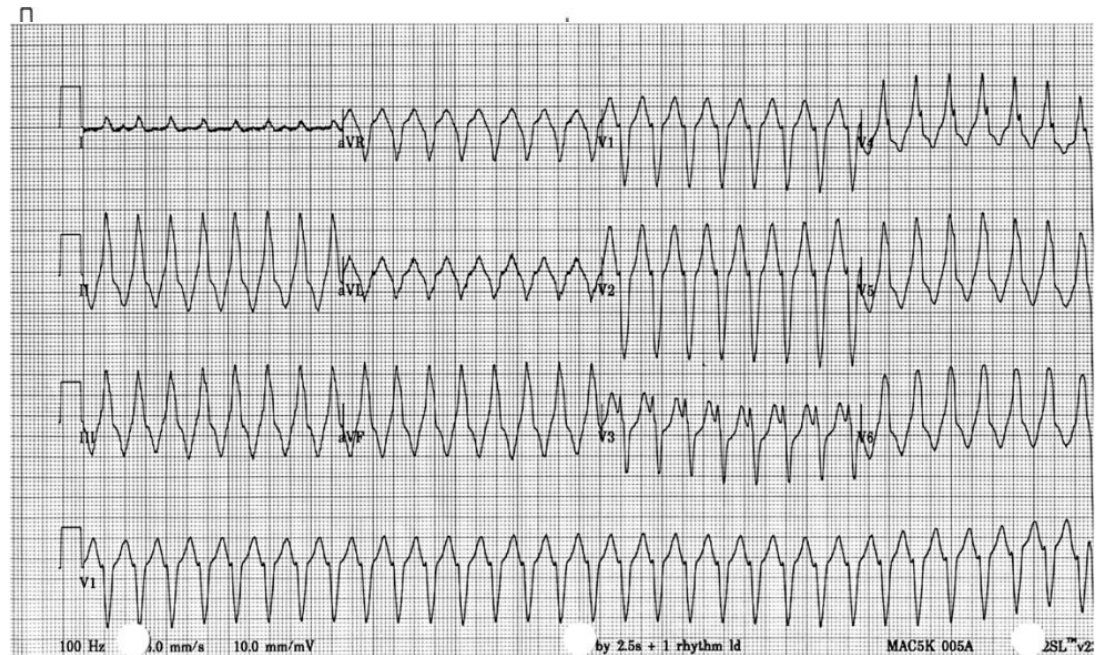


# RVOT VT

**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**





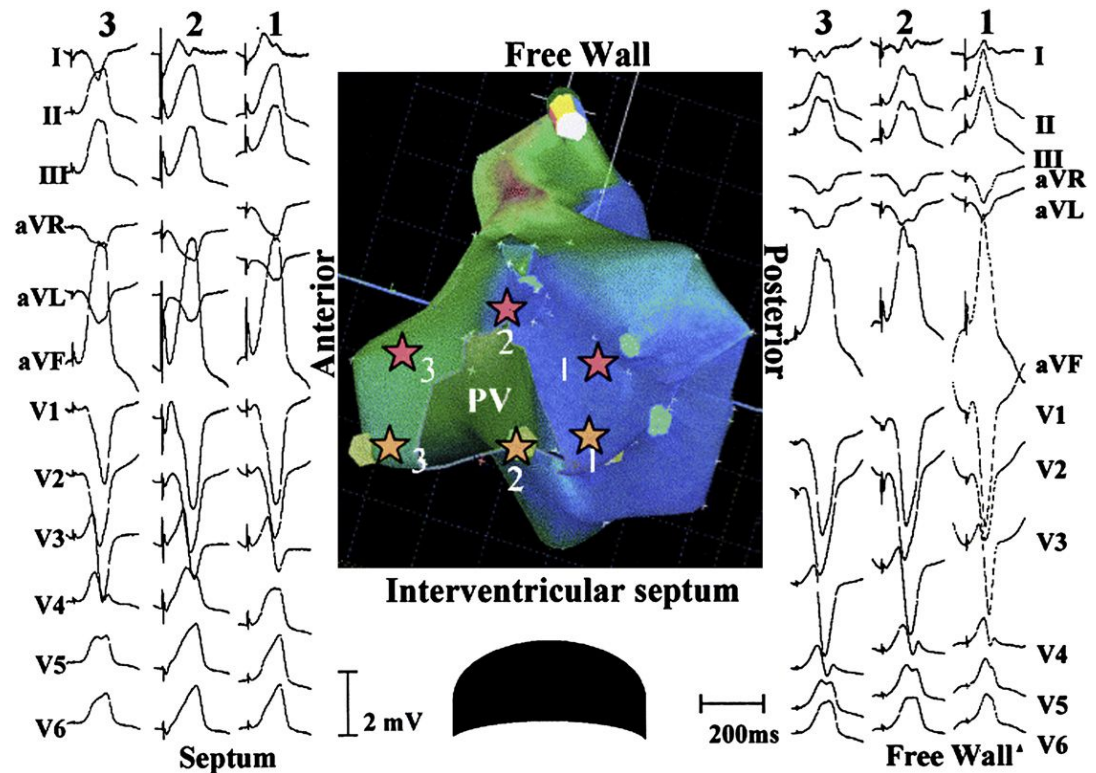


**Differentiation of septal from free wall RVOT VT**

**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)**



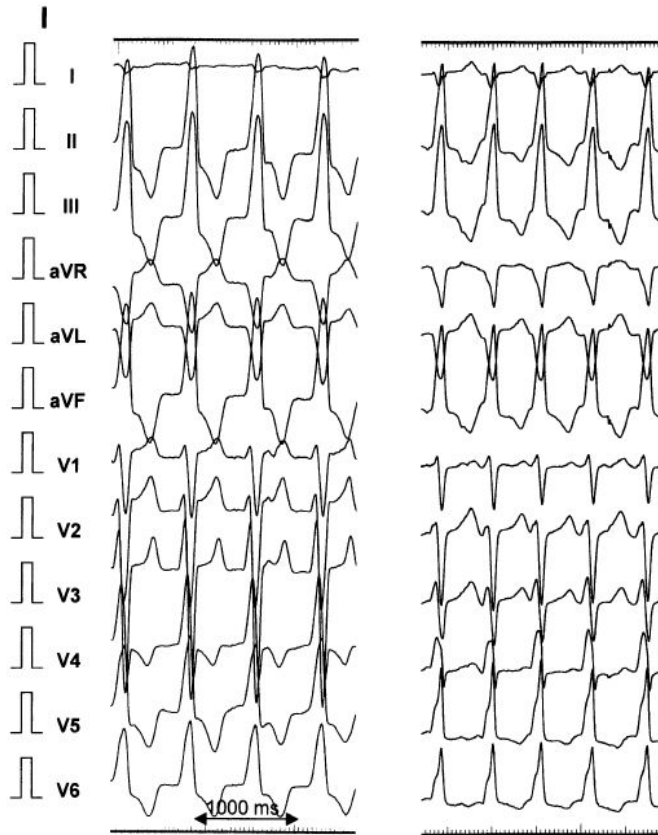
**Anterior position of free wall relative to septum -Accounts for deeper S wave in lead V2 than RVOT septum**

**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

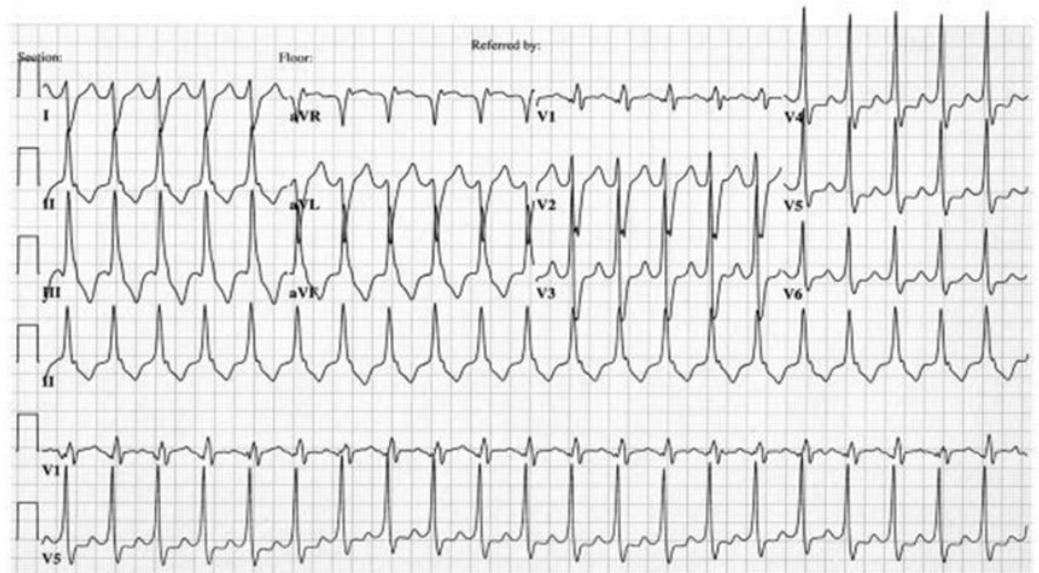
**ECG during VT shows**

**S wave in lead I**

**R-wave transition in lead V1 or 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**

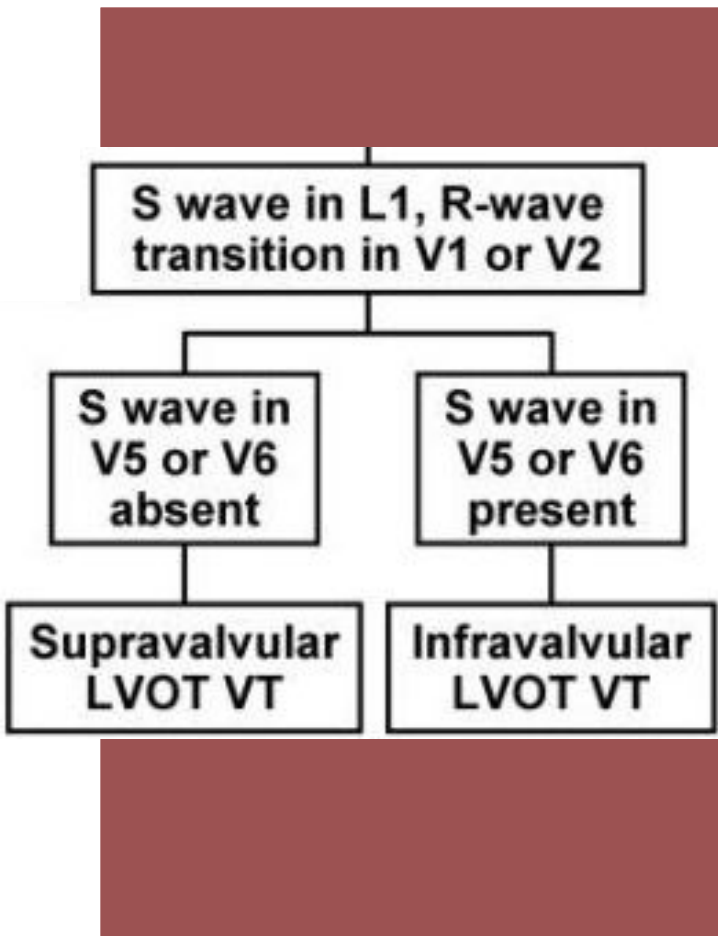
**a) Basal left interventricular or septal origin**

**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

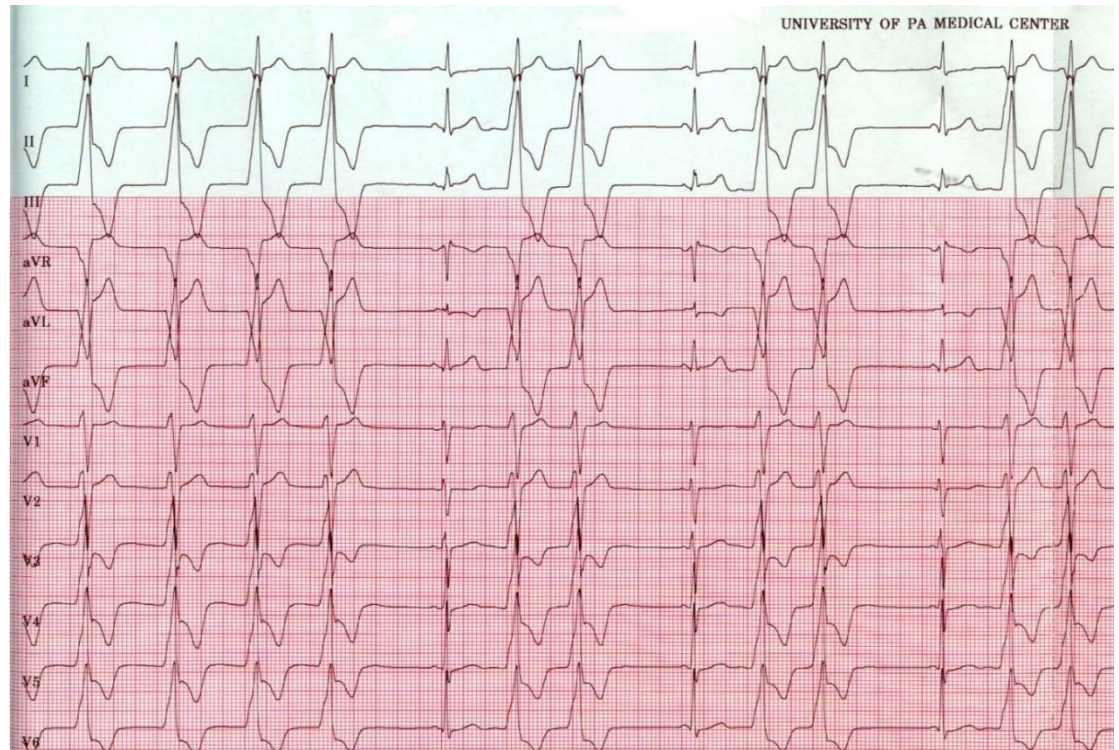
Distinction is important –RF ablation

Absence of an S wave in V5 or V6  
-supravalvular

S wave in leads V5 and V6-infravalvular

# AORTIC CUSP VT

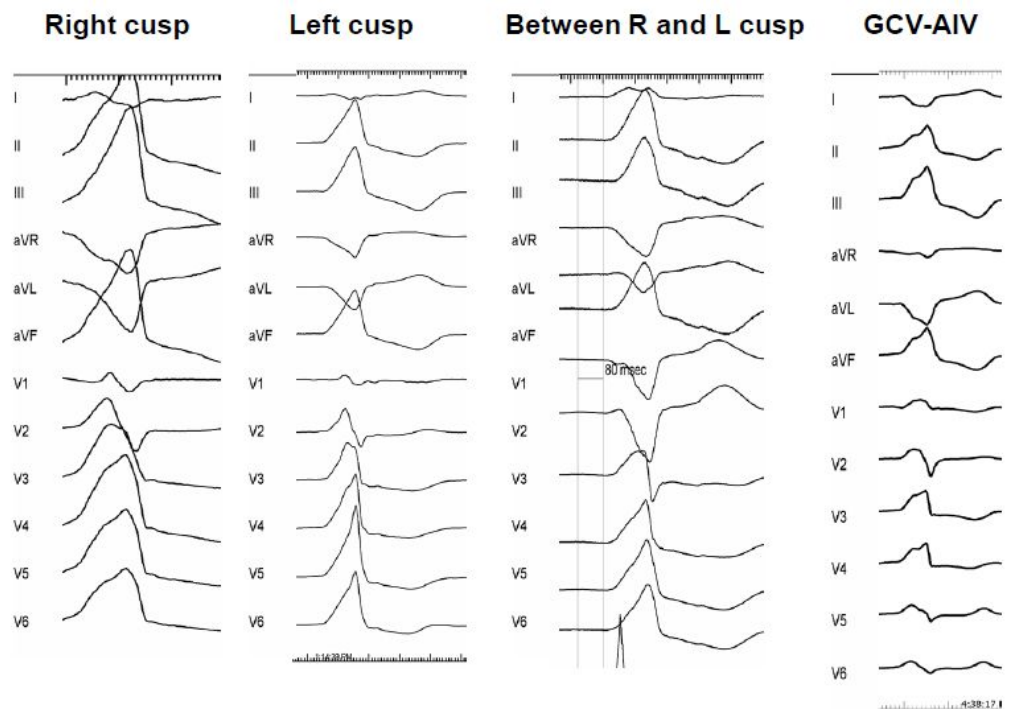
Depending on site of origin from right or left coronary cusp-LBBB or RBBB morphology



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**

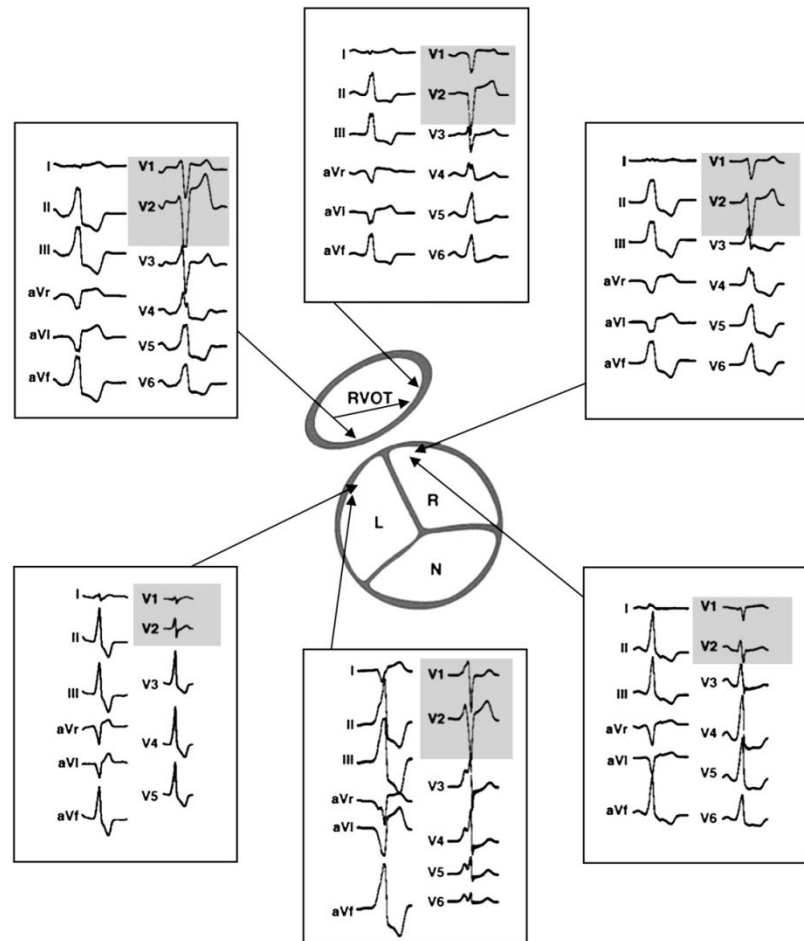


## **RVOT VT Vs aortic cusp VT**

**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**



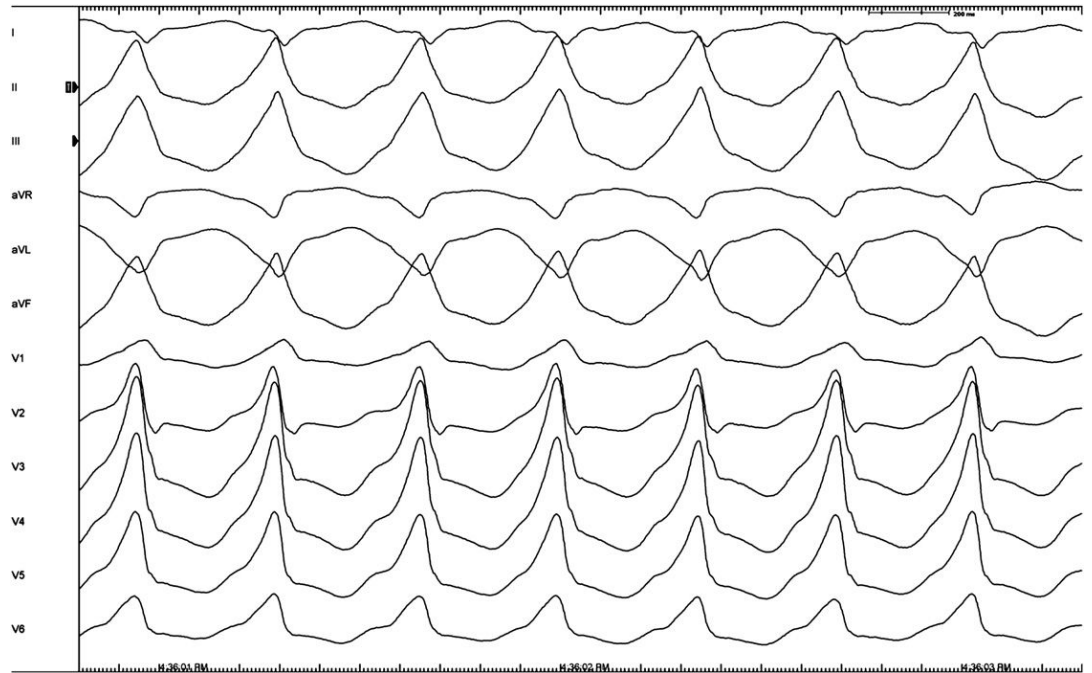


# EPICARDIAL FOCI OF VA

**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).



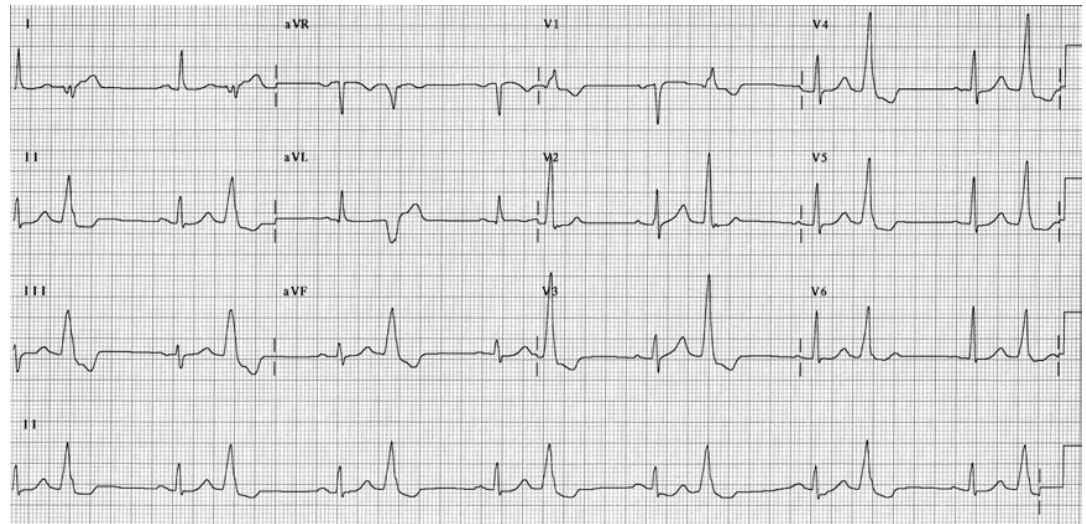
## **Psuedodelta wave**

**Interval from earliest QRS activation to earliest fast deflection in precordial leads ( $\geq 34$  ms)**

**Precordial maximum deflection index**

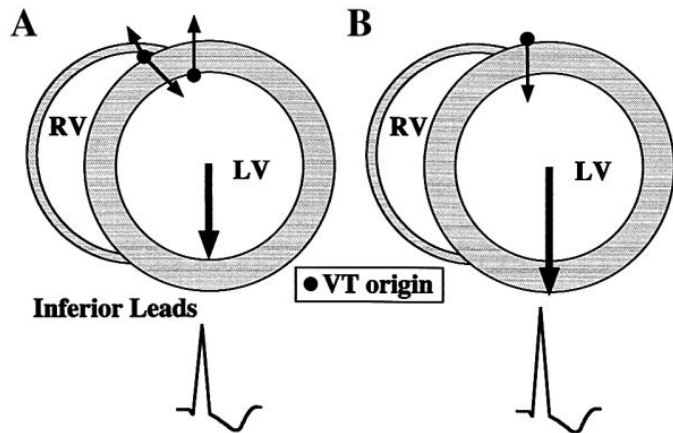
**Beginning of QRS to earliest maximal deflection in any precordial leads / QRS duration. ( $> 0.55$ )**

**(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 Avl**

- 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**

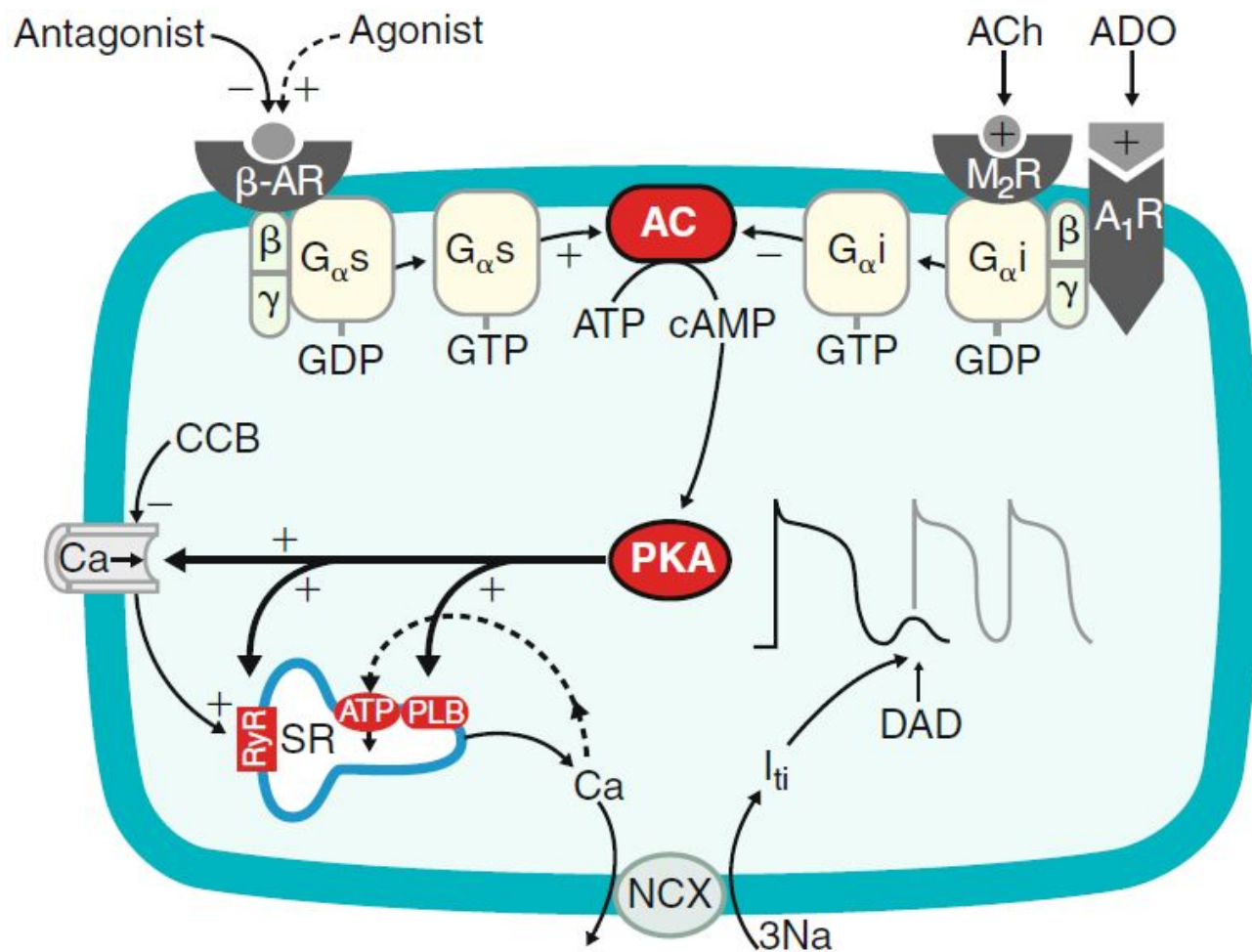
**Secondary to cyclic AMP mediated DAD**

**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**



# **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**

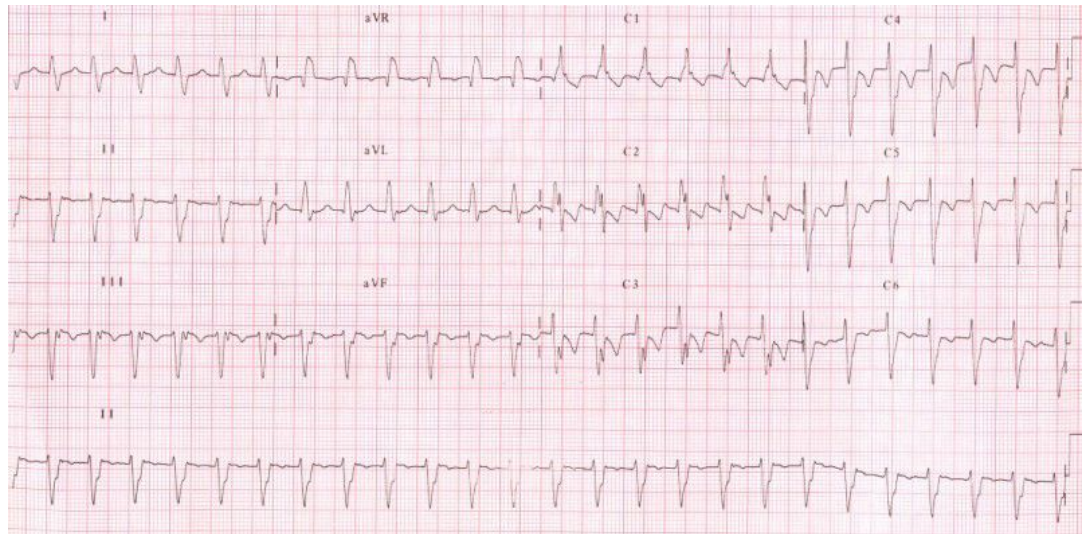
**15 to 40 years**

**More in men (60%)**

**Most occur at rest**

**Usually paroxysmal**

**Incessant forms  
leading to TCM are  
described**



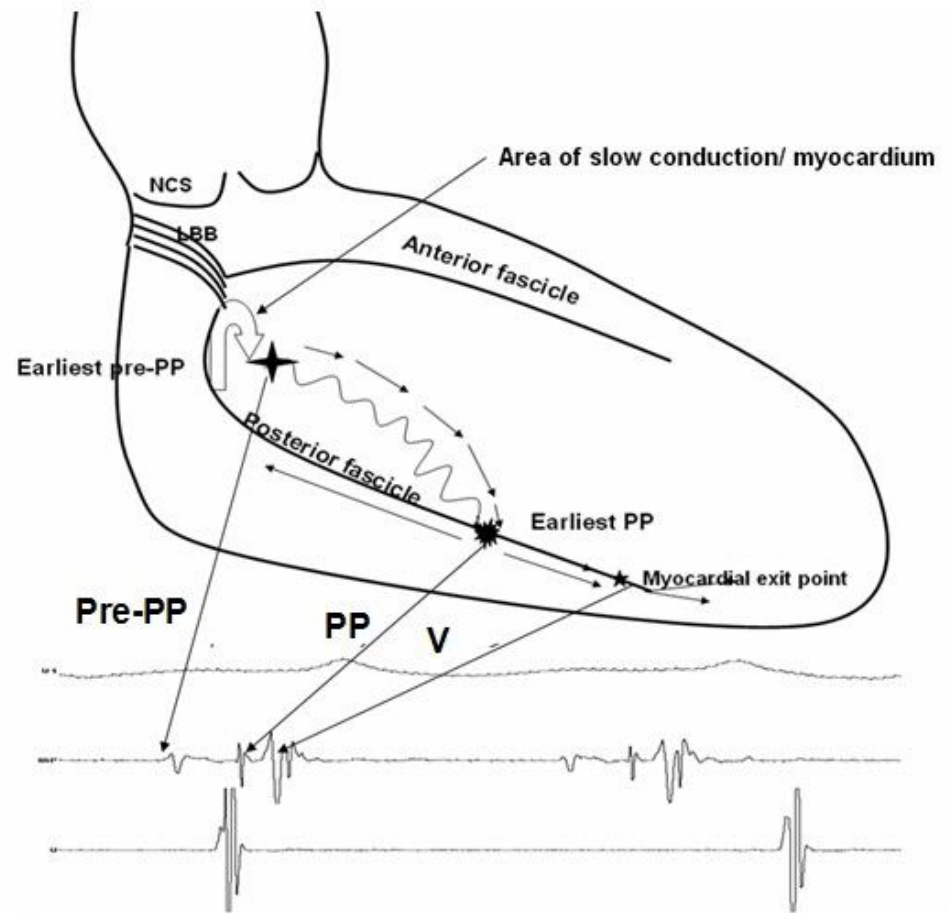
# ELECTROPHYSIOLOGIC MECHANISM

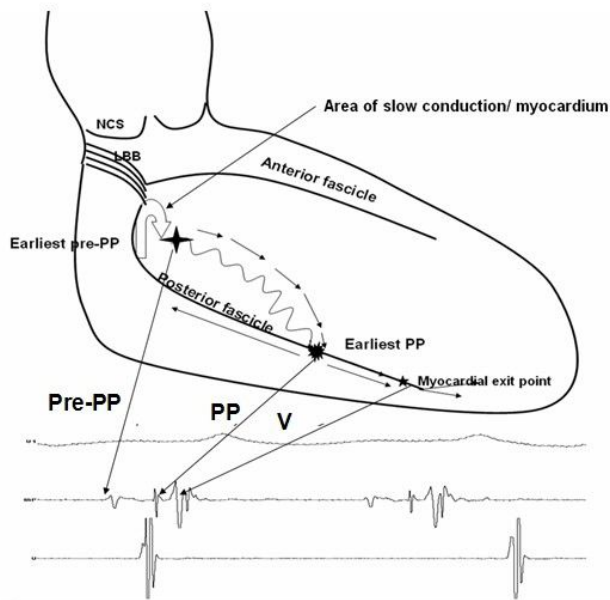
**Re-entrant mechanism**

**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**



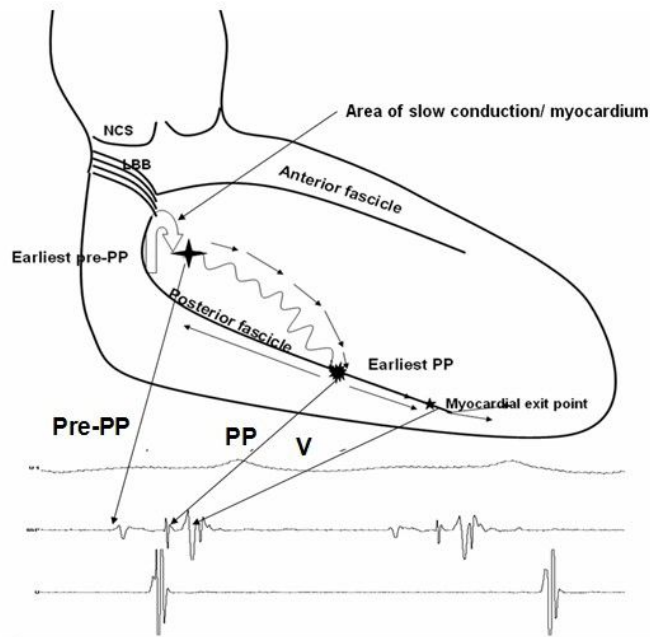


**During VT two distinct potentials can be observed before ventricular electrogram**

**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**

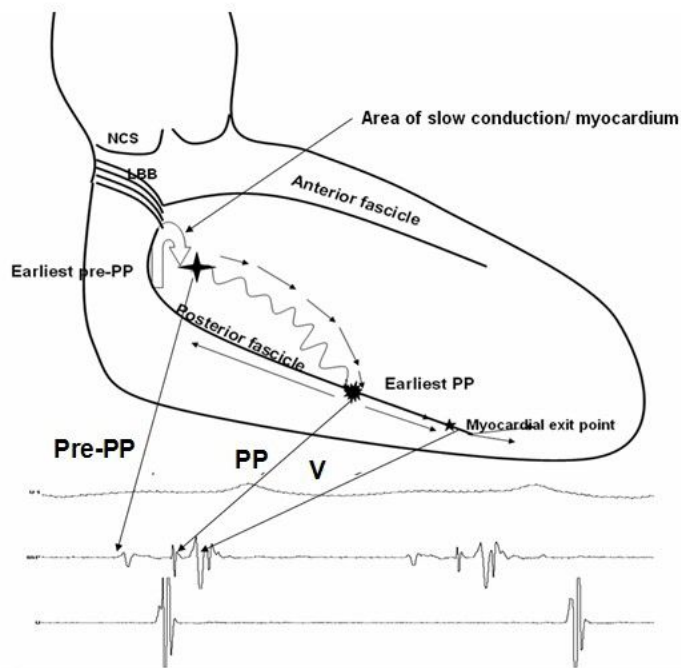
**Brief, sharp, high-frequency potential preceding onset of QRS during tachycardia**



**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**

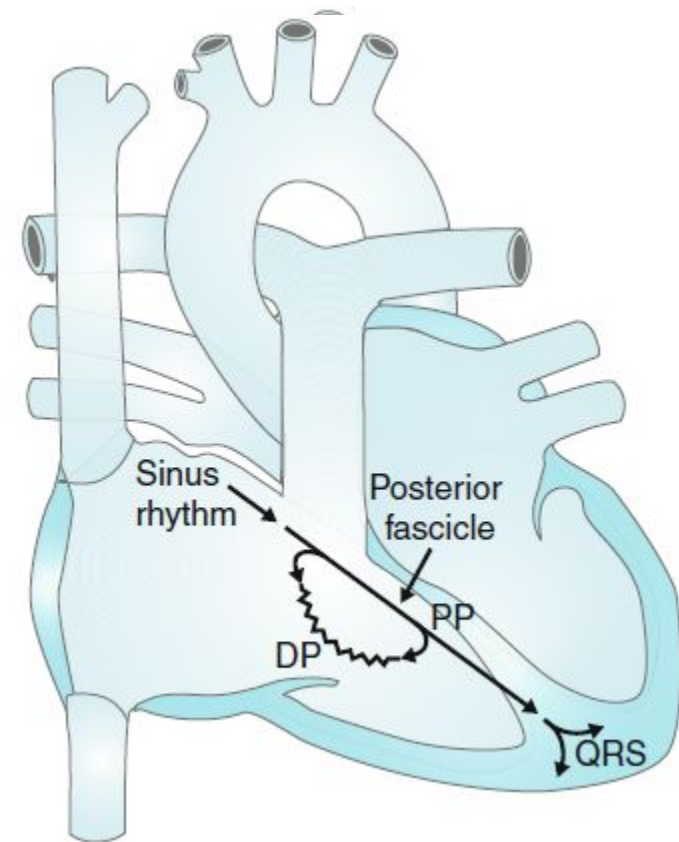
**Pre-PP is a dull, lower frequency potential preceding the PP during tachycardia**



**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**



# DD

## Supraventricular tachycardia with aberrancy

VA dissociation,

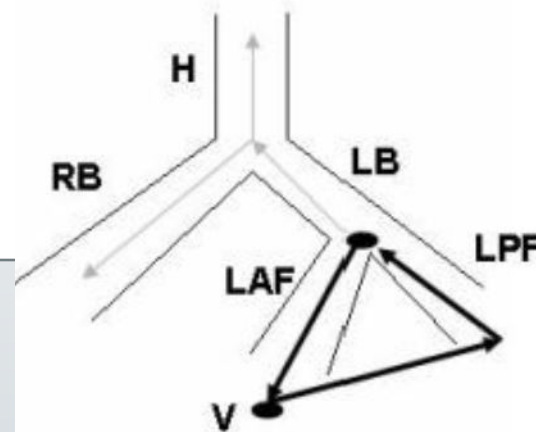
EP-Rapid atrial pacing during tachycardia can de dissociation

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

Common in patients with AWTMI 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 )

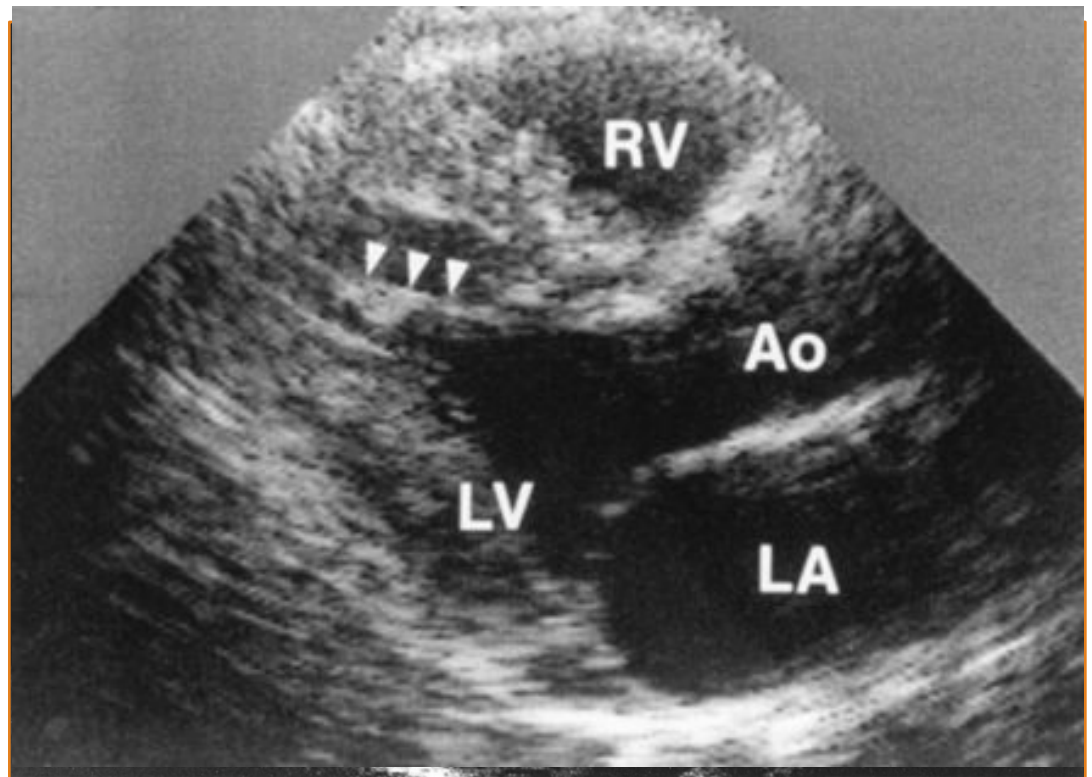


**VT originates from a false tendon extends from posteroinferior left ventricle to basal septum**

**Resection of tendon or ablation at septal insertion site eliminate tachycardia**

**Exact role tendon is unclear**

**Specificity is low**



Gallagher JJ et al. AJCardiol 1988;61(2):27A–44A

Merliss AD, Seifert MJ, Collins RF, et al 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**

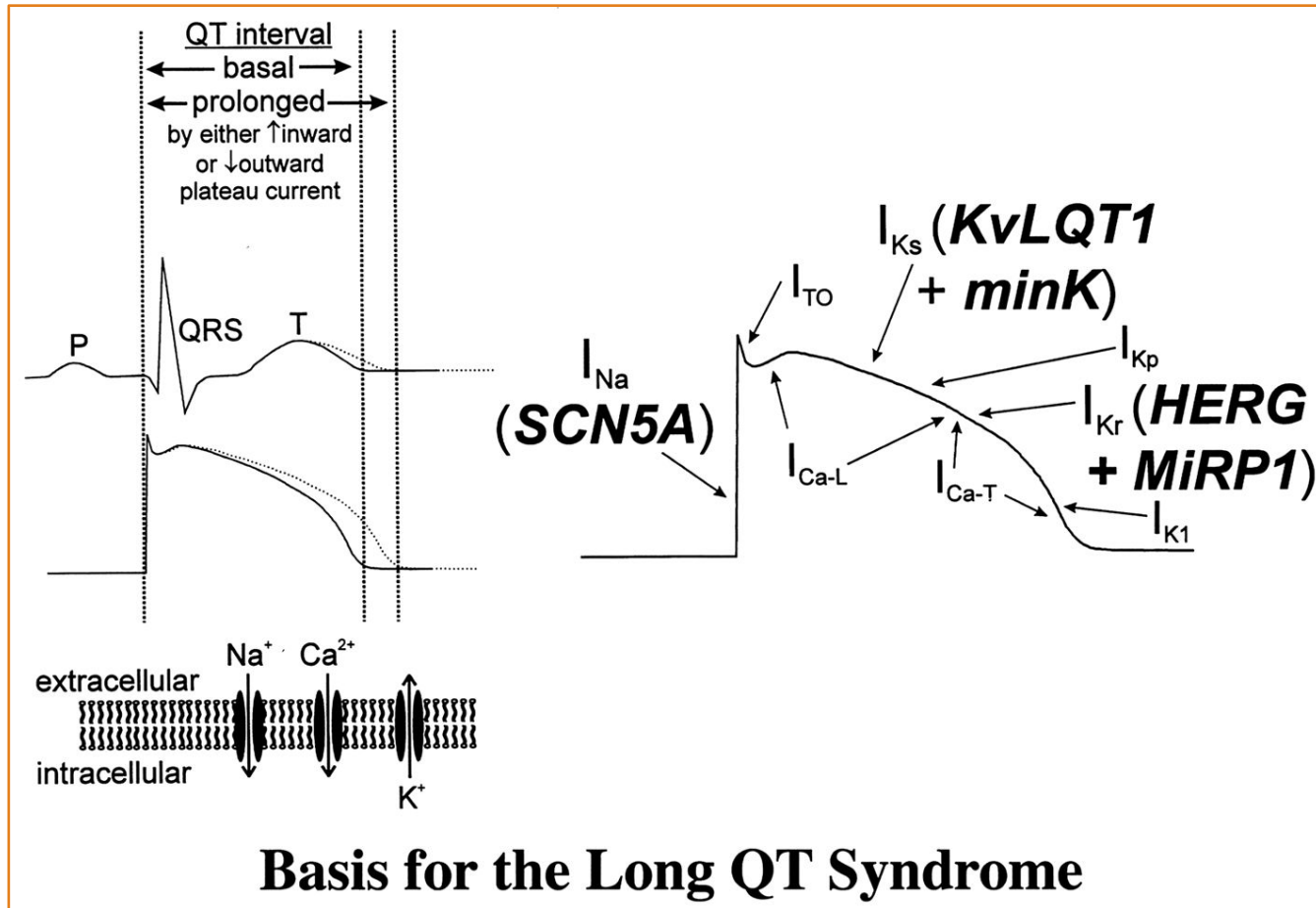
**Predisposition to EAD and torsade de pointes VT**

**Twelve different genes described**

**LQT1, LQT2, and LQT3 account for 90%**



# BASIS FOR THE LONG QT SYNDROME



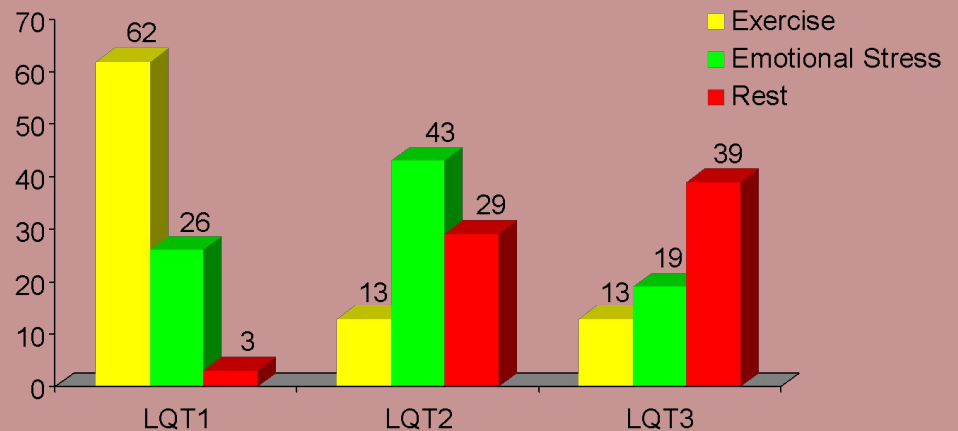
LQTS subtype	Mutated gene	Ionic current affected	Clinical frequency, %	Trigger of clinical event
LQT1 <sup>†</sup>	<i>KVLQT1</i>	$I_{Ks}$ decrease	43	Exercise
LQT2	<i>HERG</i>	$I_{Kr}$ decrease	45	Sudden arousal
LQT3	<i>SCN5A</i>	$I_{Na}$ increase	7	Sleep
LQT4	Ankyrin B	$I_{Na}$ , possible late sodium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

Approximately 25% not have identifiable gene mutations

**Mean age of symptom onset is 12 years**

**Present with syncope, seizures, or cardiac arrest.**

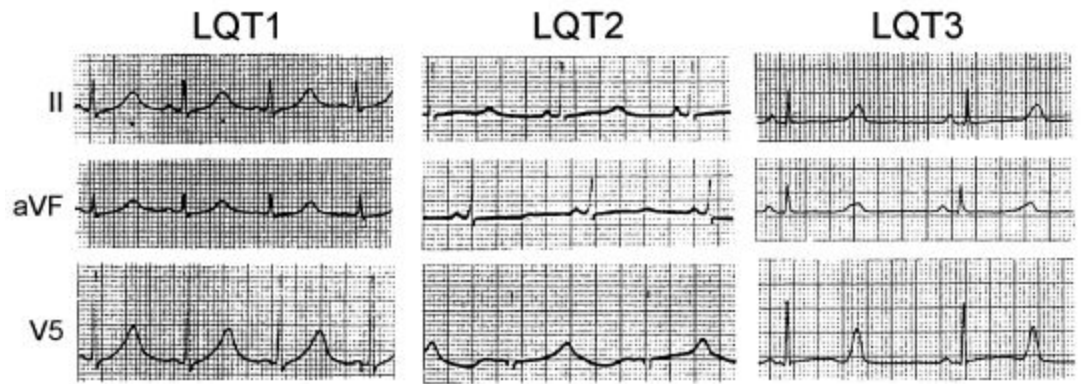
**Clinical presentation and ECG repolarization (ST-T) patterns have been correlated to genotype**



**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
<b>ECG findings</b>	
QTc ms $\geq 480$	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
<b>Clinical history</b>	
Syncopy	
With stress	2
Without stress	1
Congenital deafness	0.5
<b>Family history</b>	
Family members with definite LQTS	1
Unexplained sudden cardiac death < age 30 years among immediate family members	0.5

**Score  $\geq 3.5$  : Definite LQTS**

**Score 2-3: Intermediate probability**

**Score  $\leq 1$ : Low probability**

# 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

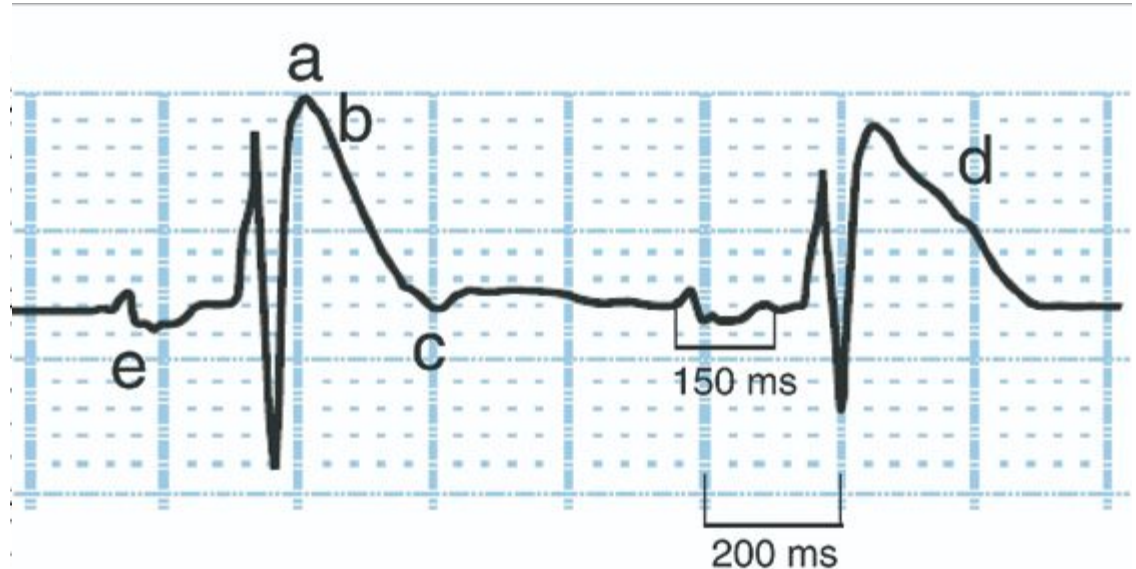
**Characterized by  
ST-segment elevation  
in V1 to V3**

**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	$\geq 2$ mm	$\geq 2$ mm	$\geq 2$ mm
T wave	negative	positive or biphasic	positive
ST-T configuration	coved type	saddleback	saddleback
ST segment (terminal portion)	gradually descending	elevated $\geq 1$ 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,  $\alpha$ -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)**

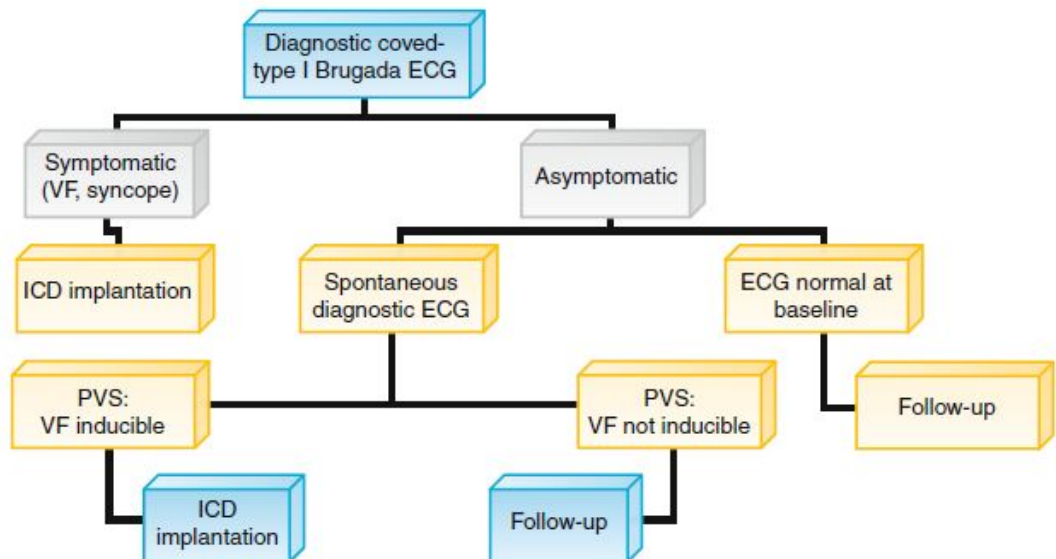
**cilostazol**

# 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**

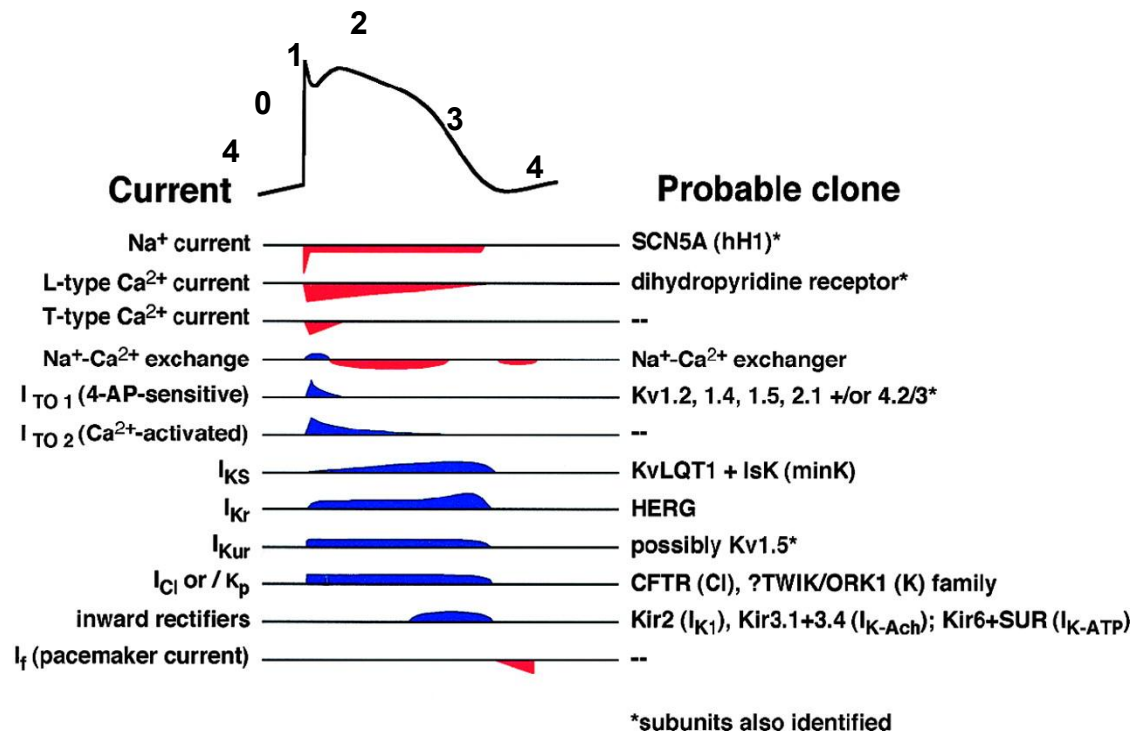
# SCN5A GENE

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

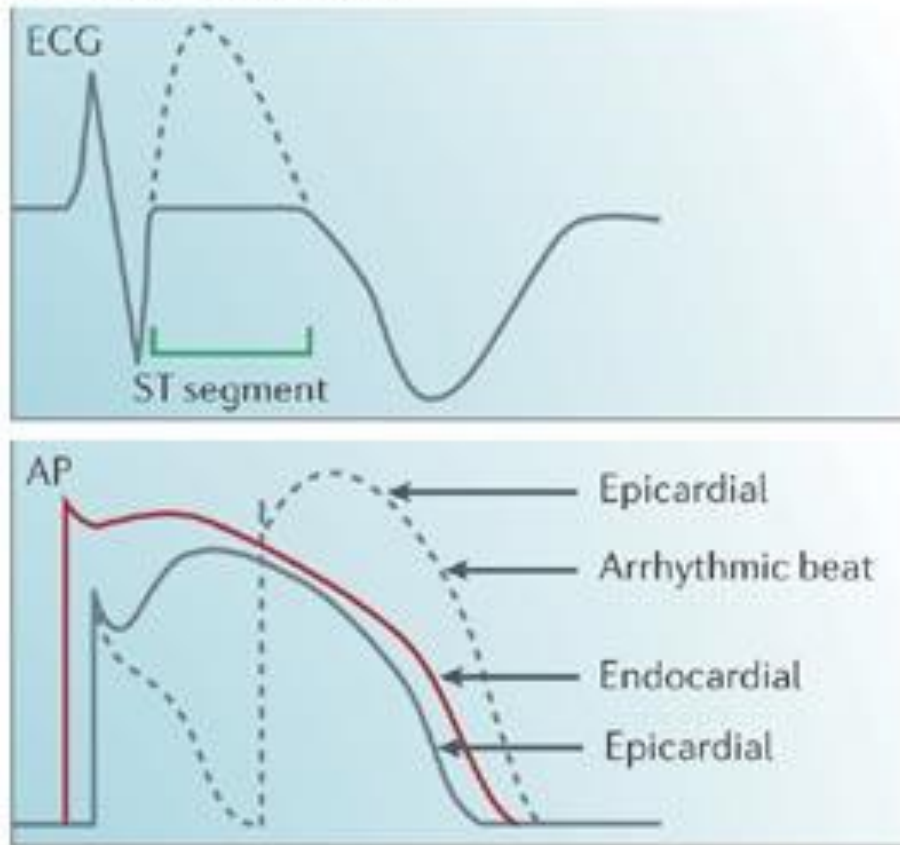
In Brugada, it opens poorly in RV epicardial cells

Autosomal dominant inheritance

20-30% of cases have abnormal SCN5A gene



## b Brugada syndrome



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Nature Reviews | Drug Discovery

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

REENTRY, 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.

# 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 sequesters 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 <sub>Kr</sub>	<i>KCNH2, HERG</i>	Shift of voltage dependence of inactivation of I <sub>Kr</sub> by +90 mV out of the range of the action potential	Gain of function of I <sub>Kr</sub>
SQT2	AD	11p15	I <sub>Ks</sub>	<i>KCNQ1, KvLQT1</i>	Shift of voltage dependence of activation of I <sub>Ks</sub> by −20 mV and acceleration of activation kinetics	Gain of function of I <sub>Ks</sub>
SQT3	AD	17q23.1–24.2	I <sub>K1</sub>	<i>KCNJ2, Kir2.1</i>	Increase in outward I <sub>K1</sub> at potentials between −75 mV and −45 mV	Gain of function of I <sub>K1</sub>
SQT4	AD	12p13.3	I <sub>Ca</sub>	<i>CACNA1C, Ca<sub>v</sub>1.2</i>	Decrease in amplitude of the inward calcium current	Loss of function of I <sub>Ca</sub>
SQT5		10p12.33	I <sub>Ca</sub>	<i>CACNB2b, Ca<sub>v</sub>β<sub>2b</sub></i>		

**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 IPV**

**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

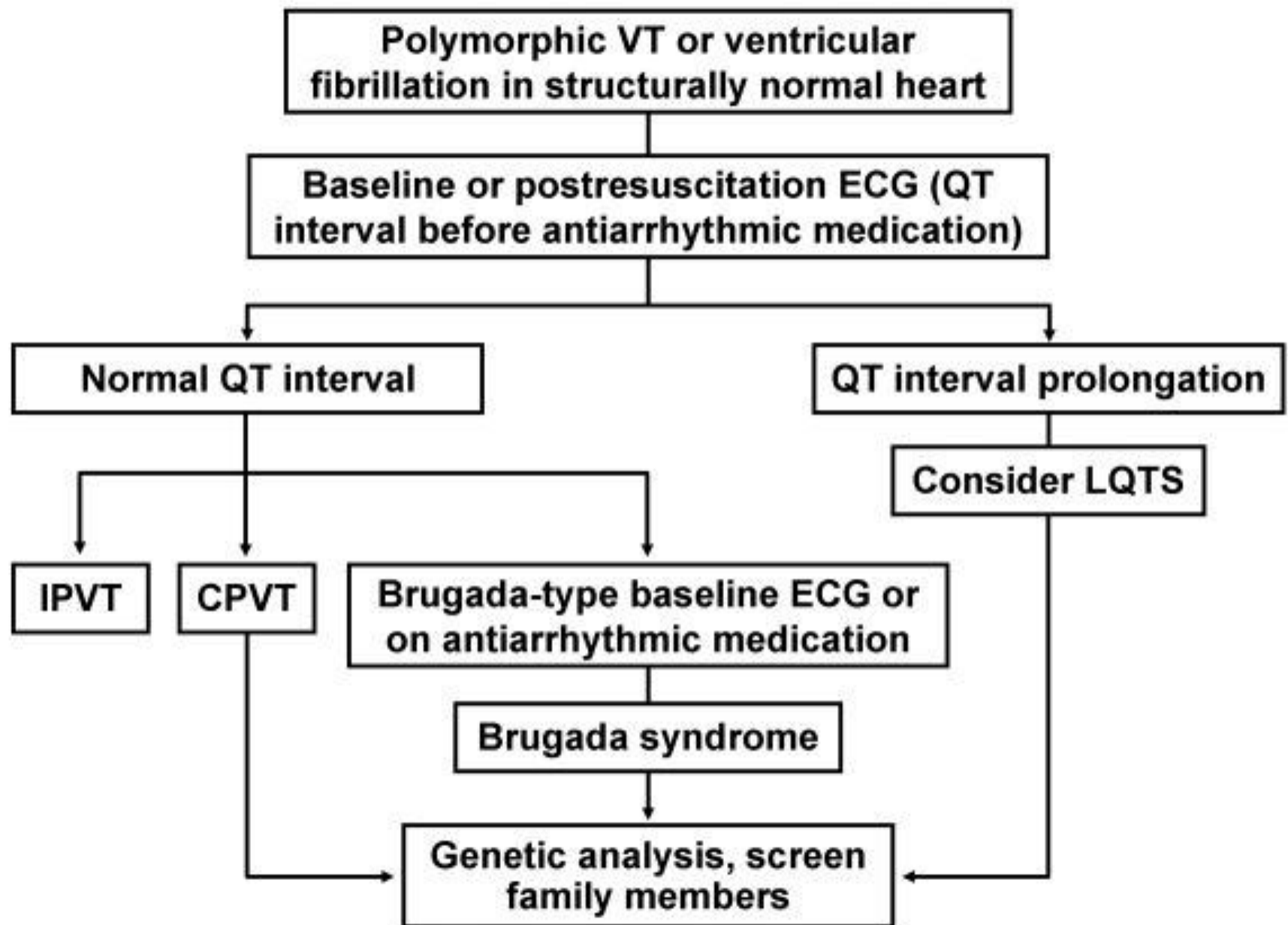
Supravalvular  
LVOT VT

Infravalvular  
LVOT VT

ILVT

None of the above morphology, sensitive to  $\beta$ -blocker

Idiopathic propranolol-sensitive (automatic) monomorphic VT (IPVT)





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THANK U

