

Idiopathic VF and J-wave Syndromes

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Definition of Idiopathic Ventricular Fibrillation (IVF)

VF in patients without overt organic heart diseases

Brugada syndrome

ST/J point elevation in right precordial leads (V1 – V3)

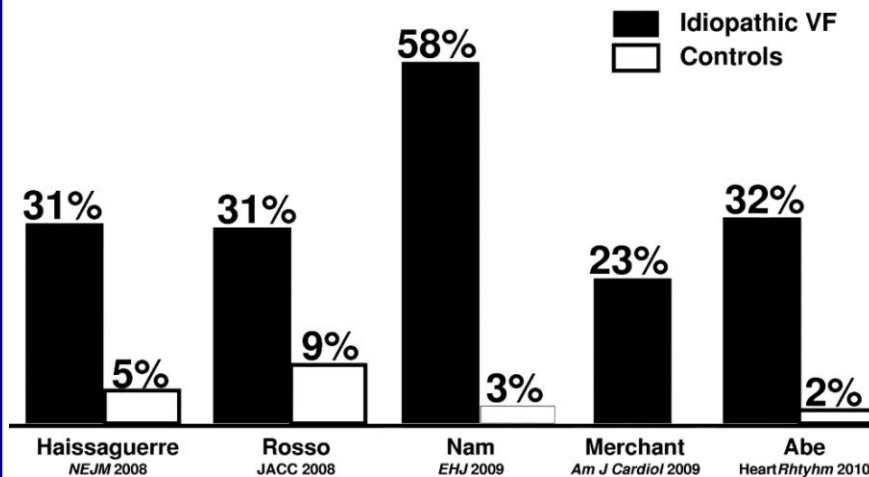
Early repolarization syndrome

J wave in infero/lateral leads (II, III, aVF, V4-V6)

IVF

no abnormalities in ECG

Prevalence of Early Repolarization in Patients with Idiopathic VF and controls.

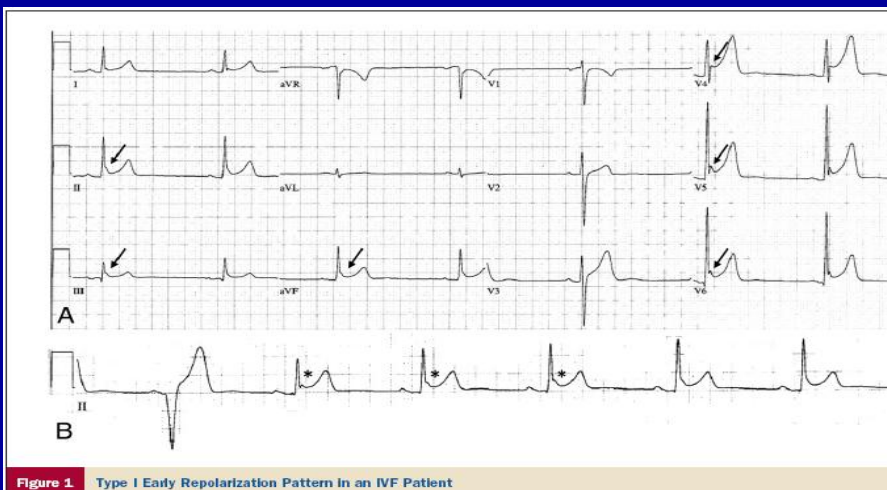


Rosso et al, Heart Rhythm 2011;8:923

Prevalence and Characteristics of ER in the CASPER study

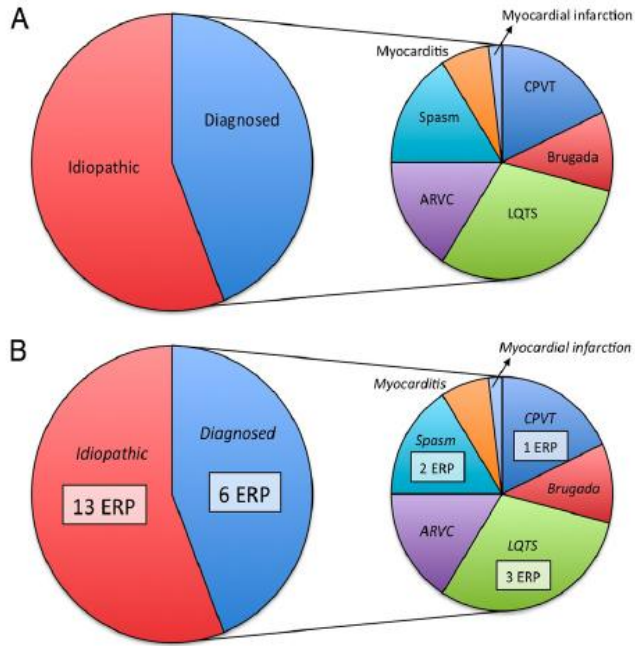
(Cardiac Arrest Survivors with Preserved Ejection Fraction Registry)

J Am Coll Cardiol 2011;58:722-8



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Prevalence and Characteristics of ER in the CASPER Study,

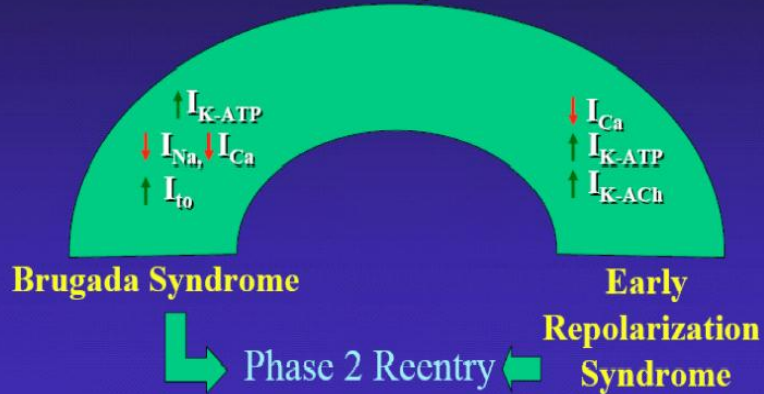
J Am Coll Cardiol, 2011;58:722-8



Distribution of Diagnoses and Early Repolarization In the Patient Cohort

Outward shift of repolarization current during early phase of the action potential

J Wave Syndromes



J Wave Syndromes, Atzelevitch and Yan, Heart Rhythm 2010

Genetic Basis of Brugada syndrome

Table 2. Genetic Basis of BS					
	Locus	Ion channel	Gene/protein	% of probands	
Causative genes					
BS1	3p21	↓ I _{Na}	<i>SCN5A, Nav1.5</i>	11–28%	
BS2	3p24	↓ I _{Na}	<i>GPD1L</i>	Rare	
BS3	12p13.3	↓ I _{Ca}	<i>CACNA1C, Cav1.2</i>	6.6%	
BS4	10p12.33	↓ I _{Ca}	<i>CACNB2b, Cavβ2b</i>	4.8%	
BS5	19q13.1	↓ I _{Na}	<i>SCN1B, Navβ1</i>	1.1%	
BS6	11q13–14	↑ I _{to}	<i>KCNE3, MIRP2</i>	Rare	
BS7	11q23.3	↓ I _{Na}	<i>SCN3B, Navβ3</i>	Rare	
BS8	12p11.23	↑ I _{K-ATP}	<i>KCNJ8, Kir6.1</i>	2%	
BS9	7q21.11	↓ I _{Ca}	<i>CACNA2D1, Cava2d</i>	1.8%	
BS10	1p13.2	↑ I _{to}	<i>KCND3, Kv4.3</i>	Rare	
BS11	17p13.1	↓ I _{Na}	<i>MOG1</i>	Rare	
BS12	12p12.1	↑ I _{K-ATP}	<i>ABCC9, SUR2A</i>	Rare	
Modulatory genes					
	15q24-q25	↓ I _r	<i>HCN4</i>		
	7q35	↑ I _{Kr}	<i>KCNH2, HERG</i>		
	Xq22.3	↑ I _{to}	<i>KCNE5 (KCNE1-like)</i>		

Antzelevitch, Circulation J 2011

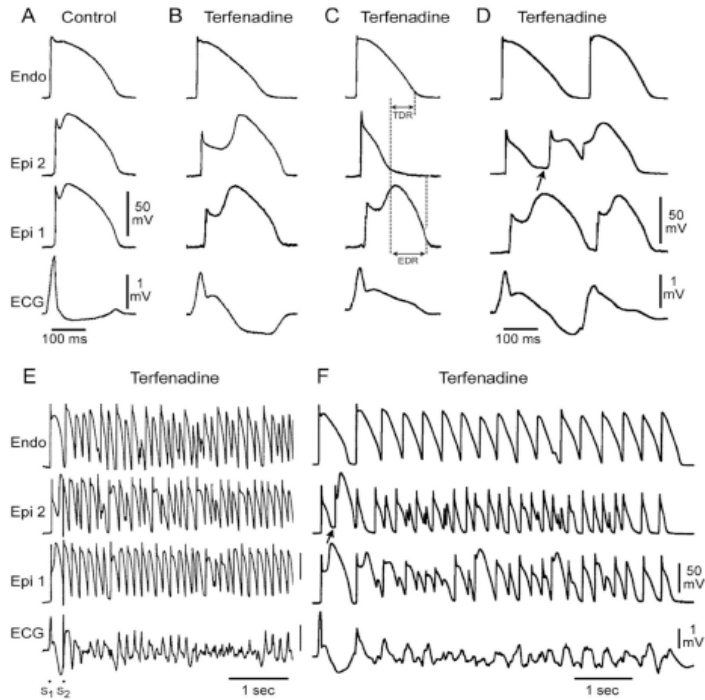
Genetic Basis of ER syndrome

Table 3. Genetic Basis of ERS				
	Locus	Ion channel	Gene/protein	% of probands
ERS1	12p11.23	I _{K-ATP}	<i>KCNJ8, Kir6.1</i>	
ERS2	12p13.3	I _{Ca}	<i>CACNA1C, Cav1.2</i>	4.1%
ERS3	10p12.33	I _{Ca}	<i>CACNB2b, Cavβ2b</i>	8.3%
ERS4	7q21.11	I _{Ca}	<i>CACNA2D1, Cava2d</i>	4.1%
ERS5	12p12.1	I _{K-ATP}	<i>ABCC9, SUR2A</i>	
ERS6	3p21	I _{Na}	<i>SCN5A, Nav1.5</i>	

Antzelevitch, Circulation J 2011

EP mechanism of Arrhythmia in both Brugada syndrome And ER syndrome

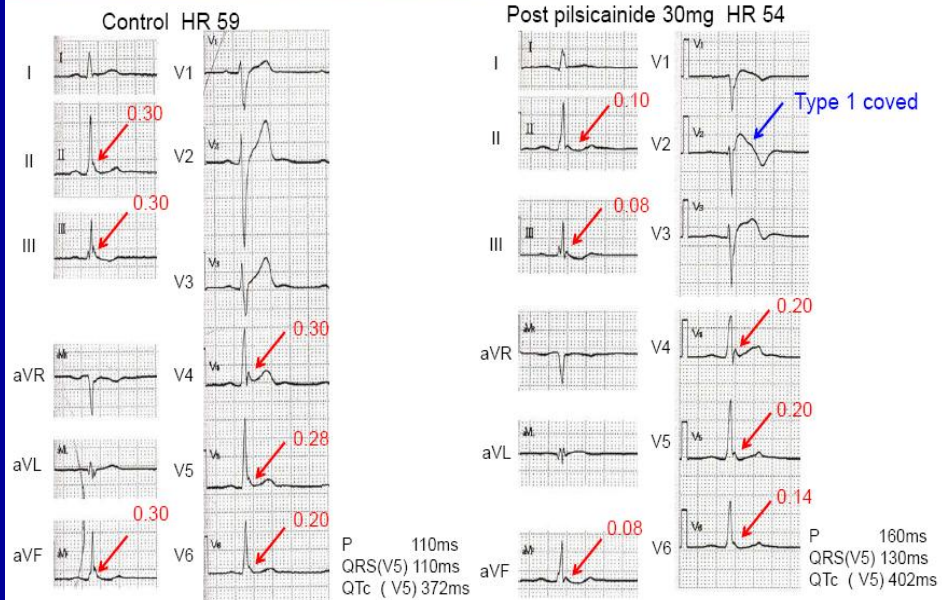
J Wave Syndromes, Atzevitch and Yan, Heart Rhythm 2010



ER Syndrome vs Brugada Syndrome

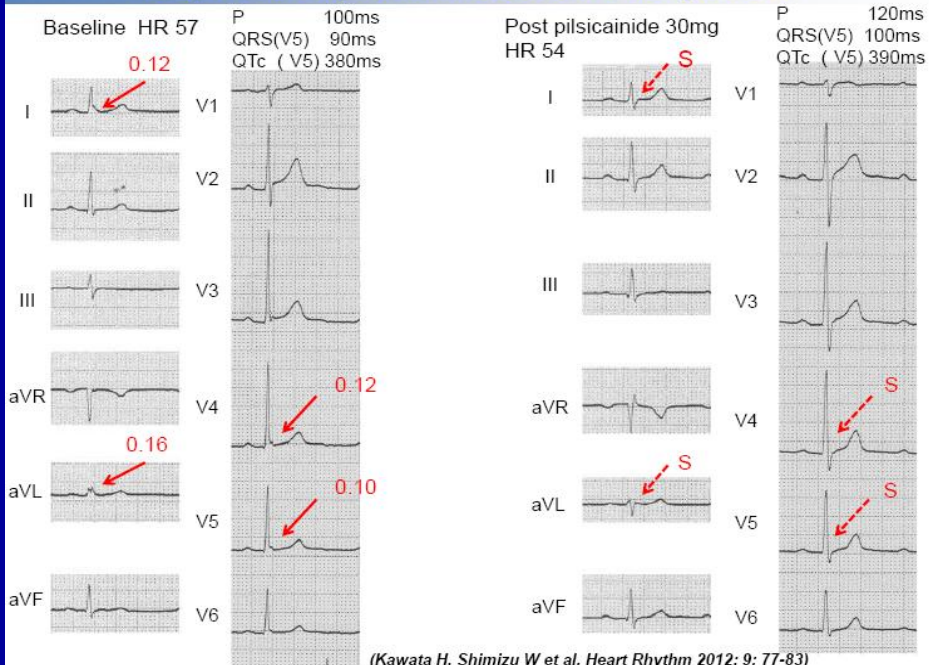
- **Similarities:**
 - Largely similar clinical, genetic, and cellular background
 - Male preponderance
 - Late adult onset
 - More VT/VF at rest/during sleep
- **Differences:**
 - Low rate of abnormal SAECG in ERS
 - Low inducibility of VF on EPS
 - Different response to Na channel blockers

Response to pilsicainide in a Brugada patient



(Kawata H, Shimizu W et al. Heart Rhythm 2012; 9: 77-83)

Response to pilsicainide in an ERS patient



(Kawata H, Shimizu W et al. Heart Rhythm 2012; 9: 77-83)

Characteristics of recurrent VF associated with ER
 Hassaguerre et al,
 J Am Coll Cardiol 2009;53:612

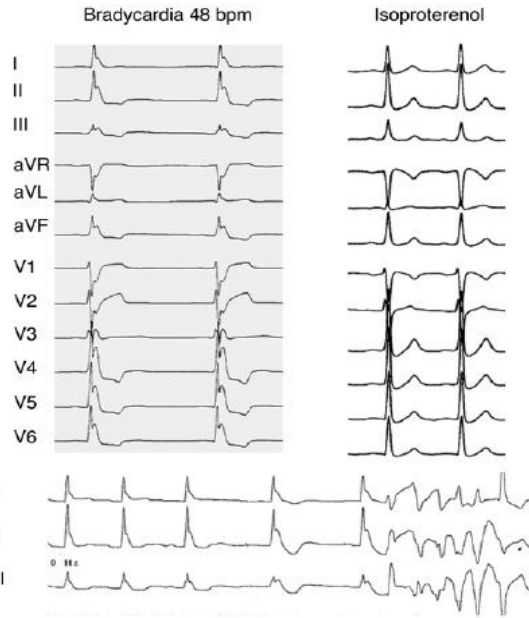
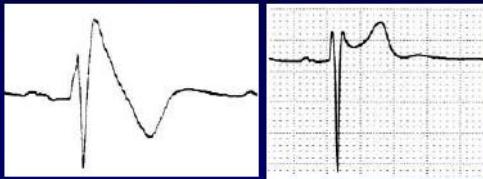


Figure 5 Normalization of QRST Morphology During Isoproterenol Infusion

J wave syndrome

Brugada syndrome



Coved type

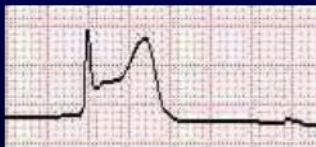
Saddle-back

Hypothermia

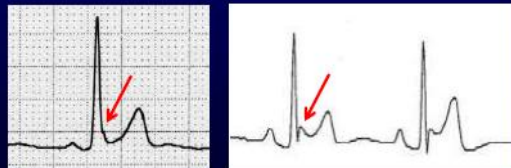


Osborn wave

Acute ischemia

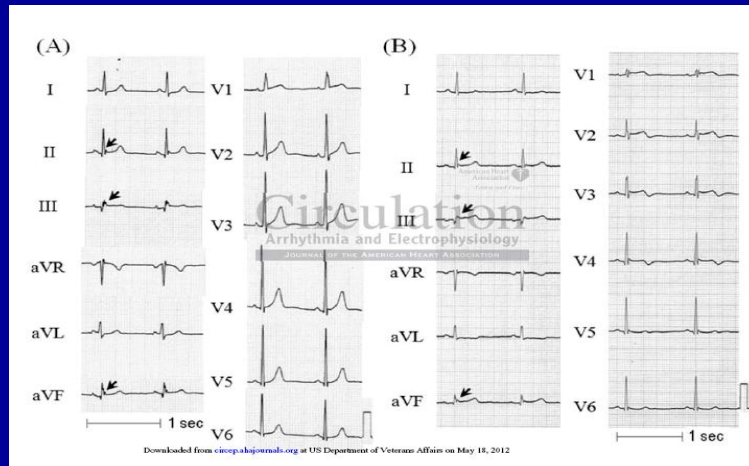


Early Repolarization Syndrome



Early Repolarization is an independent predictor of occurrence Of VF in the very early phase of acute myocardial infarction

Naruse et al, *Circulation*, April 24, 2012



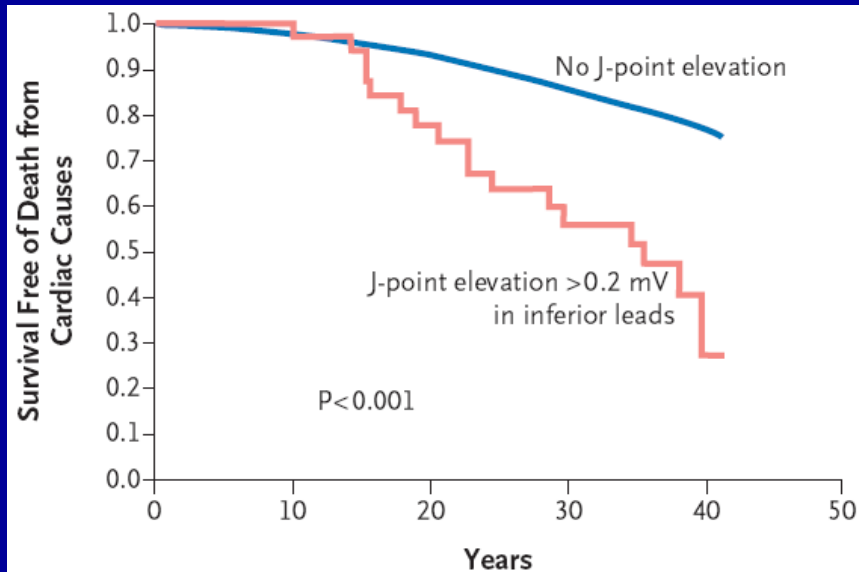
Summary: 10% of 220 consecutive patients with AMI developed VF within 48 hours. ER, time from onset to admission <180 min, and Killip class >1 were independent predictors of VF

Risk Stratification of Early Repolarization

- History of syncope/Family history of SCD
- ECG lead distribution (lateral, inferior, infero-lateral, infero-lateral+V1-3)
- Amplitude of ER
- Configuration of ST segment
- Association with short QT interval
- Dynamicity of ER amplitude/configuration

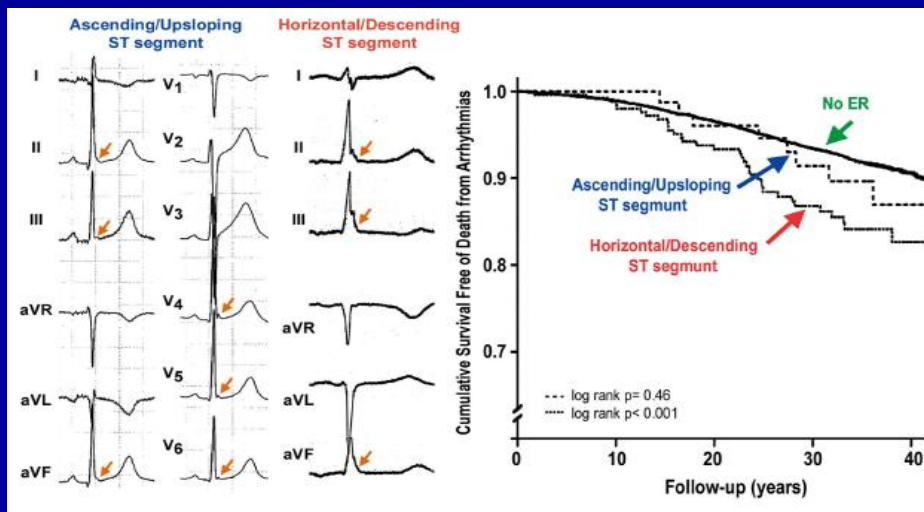
Lo **Long term outcome associated with ER on ECG**

Tikkanen et al, *N Engl J Med* 2009;361:2529



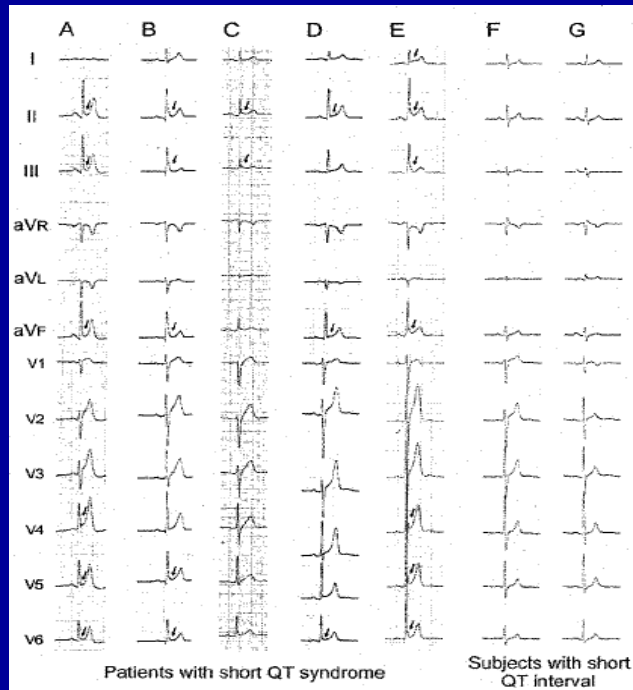
Early Repolarization: ECG phenotypes associated With favorable outcome

Tikkanen et al, *Circulation* 2011;123:2666



High prevalence of ER in short QT syndrome

Watanabe et al,
Heart Rhythm 2012



Prevalence and Characteristics of ER in the CASPER Study

J Am Coll Cardiol, 2011;58:722-8

Patient #5 (Table 2) has a family history of sudden death. He had experienced 2 syncopal episodes in conjunction with auditory stimuli, and collapsed shortly after an alarm clock sounded and was resuscitated from ventricular fibrillation. The resting QT is unremarkable, but prolonged by 60 ms with 0.10 $\mu\text{g}/\text{kg}/\text{min}$ adrenaline, and failed to shorten with exercise. The ambulance 12-lead and day 1 post-arrest ECG show J-wave elevation that is initially slurred and subsequently notched. Genetic testing for LQTS genes was negative, and there were no family members with similar findings.

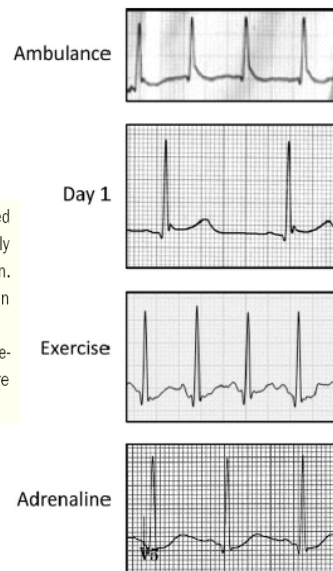


Figure 5 Lead V₆ ECG Series in a 29-Year-Old Male With Cardiac Arrest

A graphic consisting of a solid blue background on the left and a black background on the right, separated by a curved line that starts at the top left and curves towards the bottom right. The text is centered on the blue background.

THANK YOU

NABIL EL-SHERIF