Prevention of Sudden Cardiac Death (SCD)

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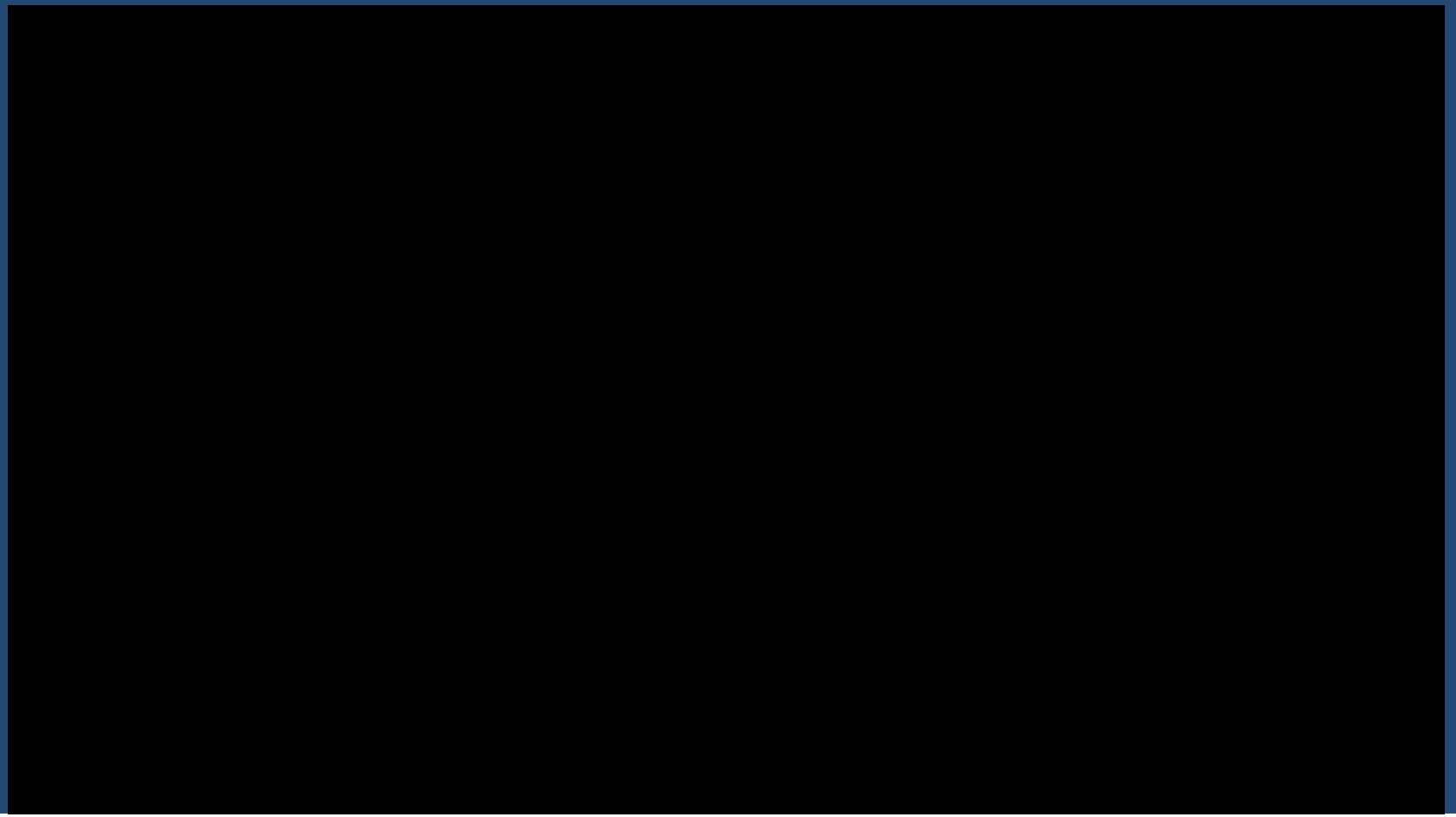
Objectives

- Be able to recognize common causes of sudden cardiac death and treatment/ prevention modalities.

Disclosures

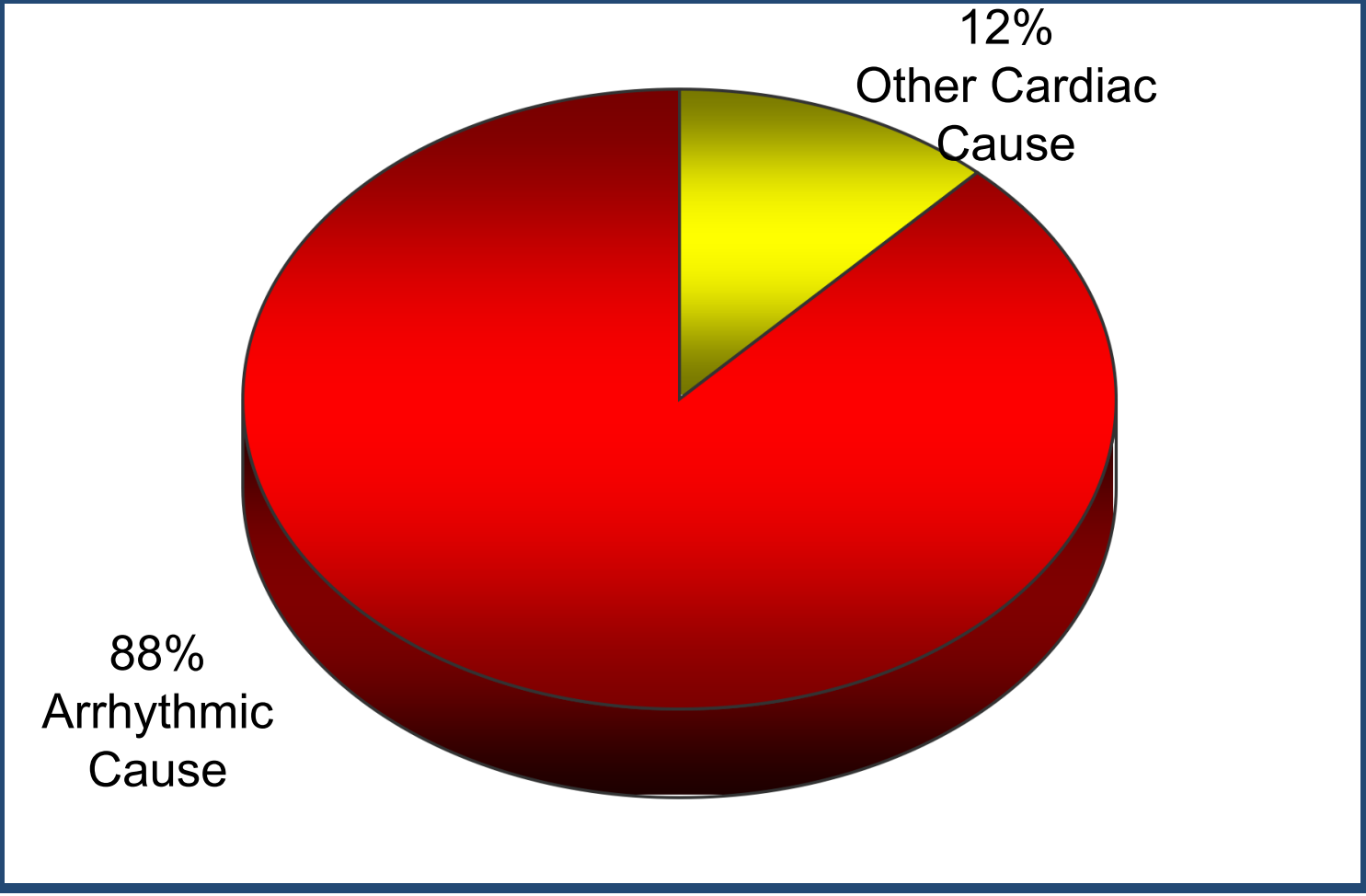
- Archer First Response, Equity, Medical Advisor
- Boston Scientific Corporation, Scientific Advisory Board, Remuneration donated to LRH Foundation.

What does SCD look like in real life?



Cause of SCD

"Sudden cardiac death: the pro-arrhythmic interaction of an acute loading with an underlying substrate."



- Luscher TF, "Sudden cardiac death: new aspects of a continuing clinical challenge," European Heart J (2017), V. 38, 2973-2976.
- Albert CM. *Circulation*. 2003;107:2096-2101.

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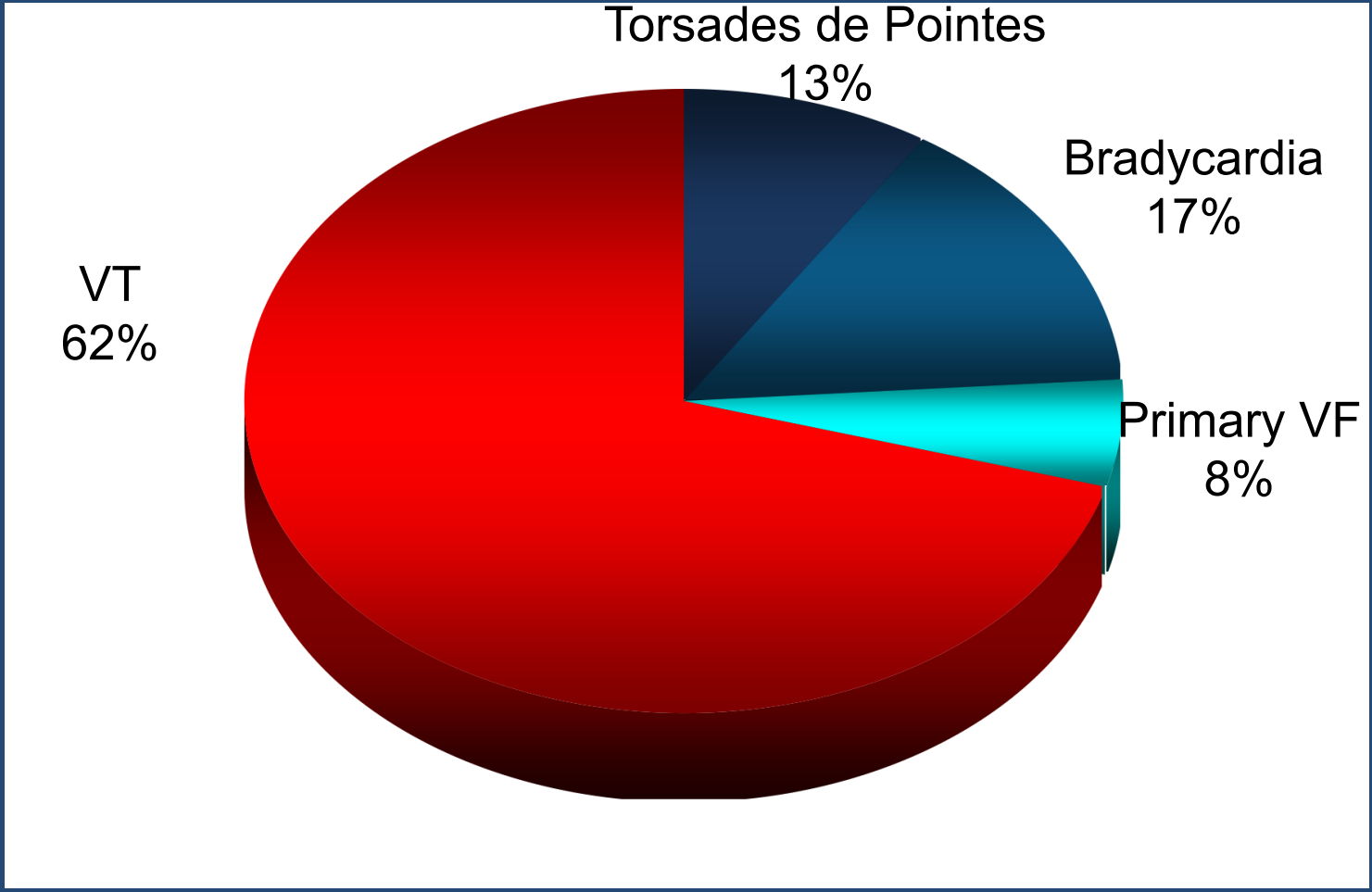
Underlying Arrhythmias of SCD



B Adapted from Bayes de Luna A. *Am Heart J.* 1989;117:151-159. ayés de Luna A. *Am Heart J.* 1989;117:151-159.

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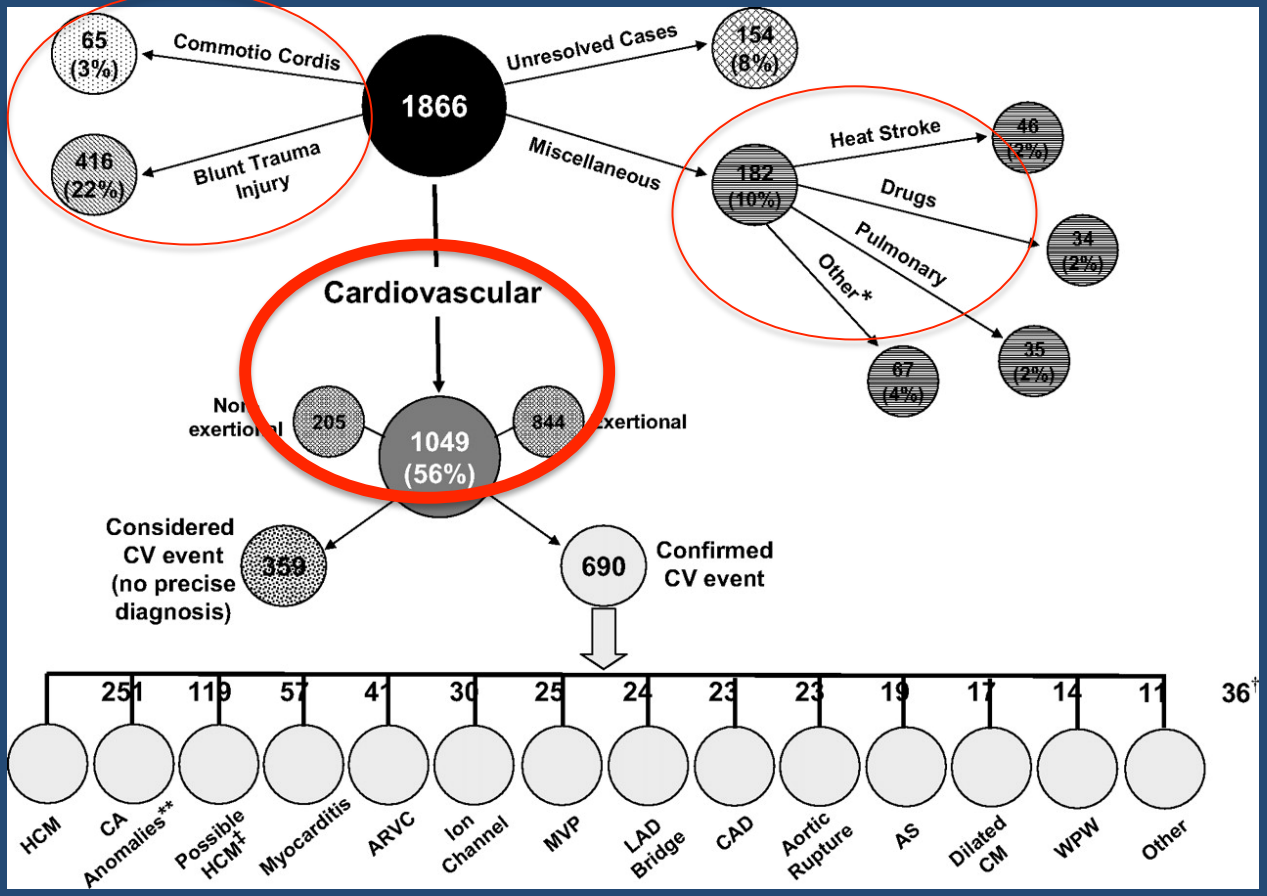
Sudden Deaths in Young Competitive Athletes



Syncope during peak exertion often indicates pathology.

Maron BJ et al, "Sudden Deaths in Young Competitive Athletes
Analysis of 1866 Deaths in the United States, 1980–2006," Circulation. 2009;119:1085–1092

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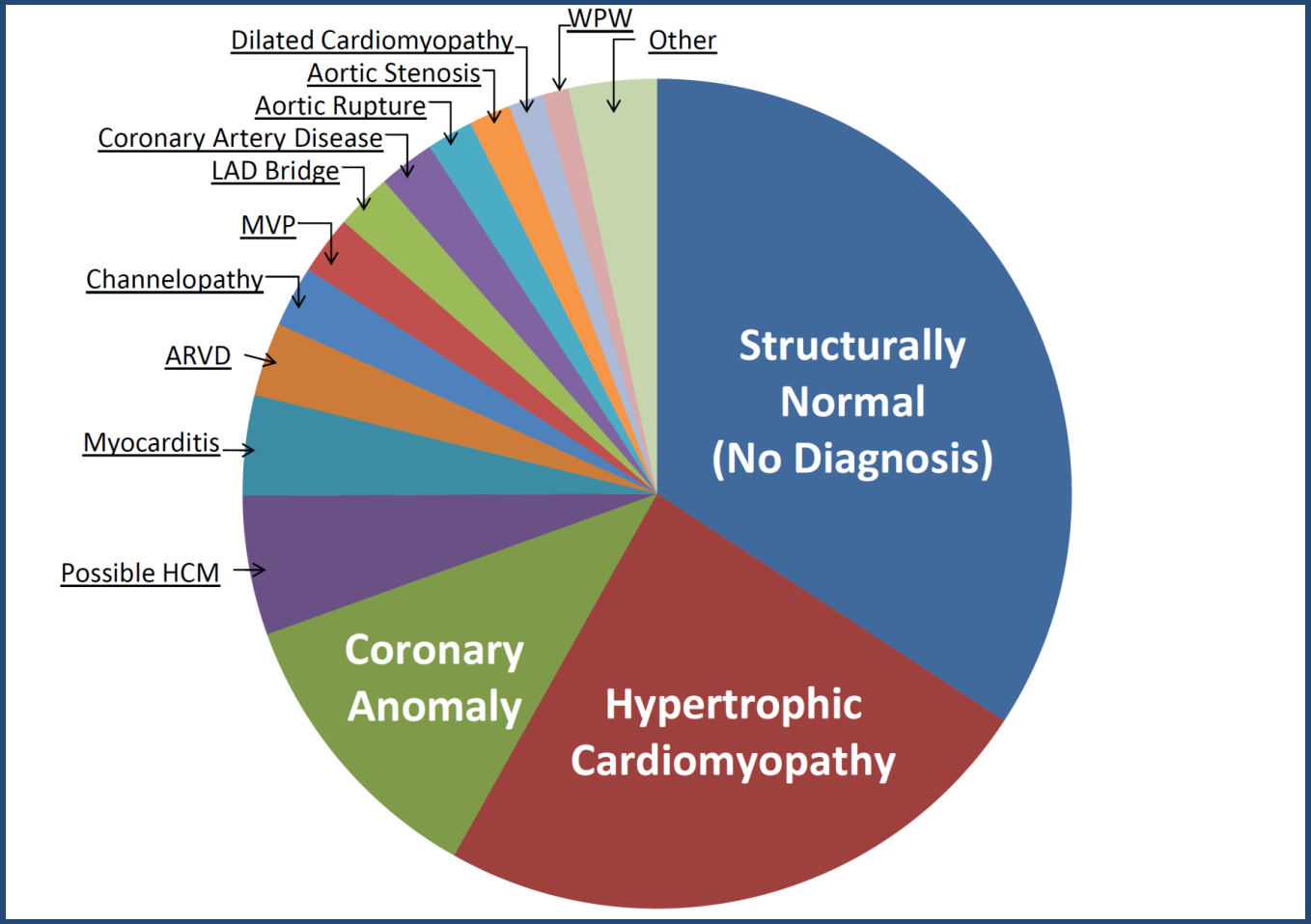
SCD in Young Athletes



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Case #1: Syncope

- 16yo hockey player come for preparticipation sports physical.
- He knocked out 2 teeth when he fainted during practice.
- You get an ECG.

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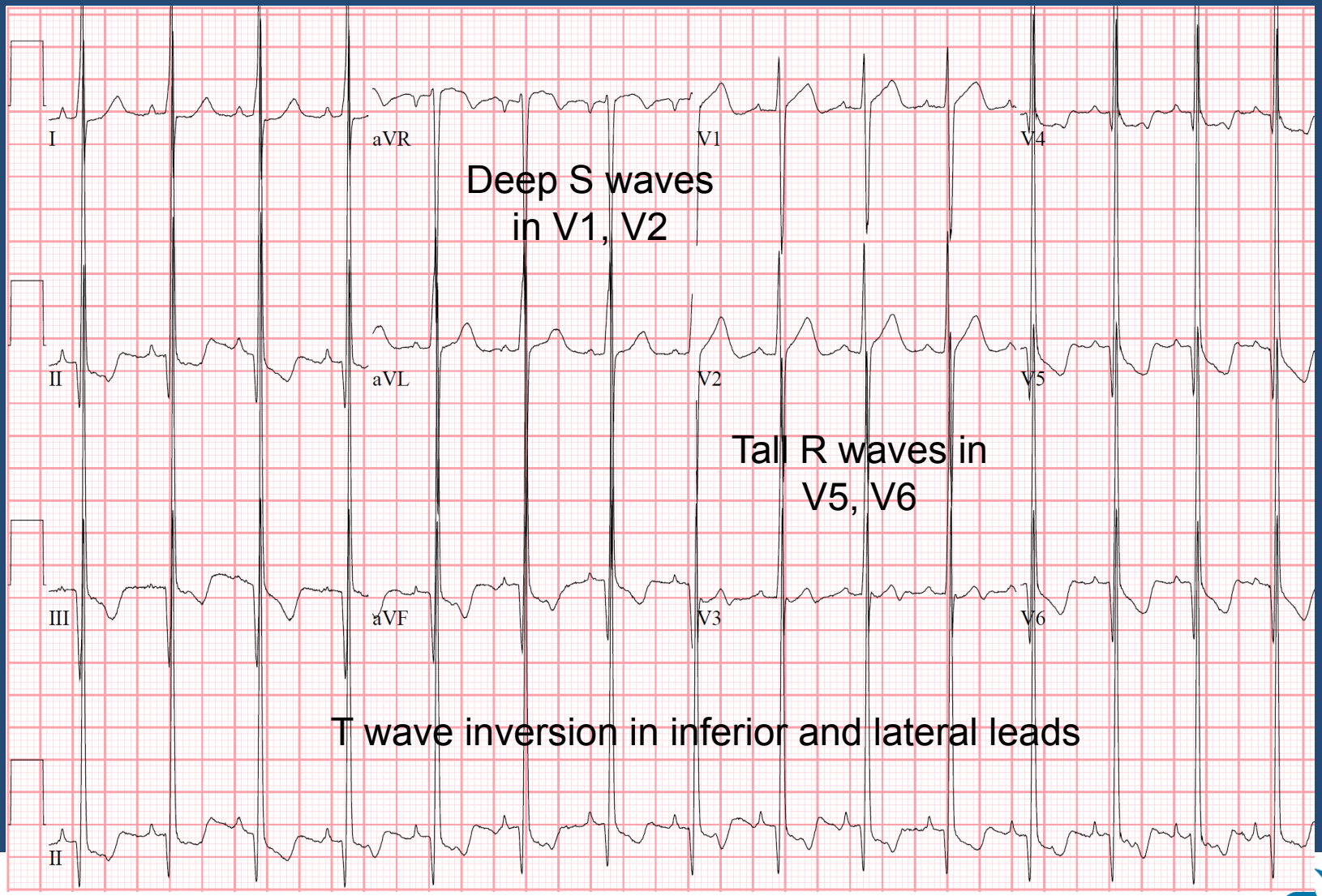
Hypertrophic CMP



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Hypertrophic Cardiomyopathy

Inheritable disorder (autosomal dominant)

- abnormal myocardial thickening
- asymmetrical enlargement of the septal surface of the left ventricular wall
- varying degrees of outflow obstruction
- risk for ventricular arrhythmias, especially with exercise
- risk for myocardial dysfunction and chronic atrial fibrillation

❖ The single most common cardiovascular cause of sudden death in the U.S.

Hypertrophic Cardiomyopathy

- Prevalence is 1:500 in the US
- Natural history: 2-8% incidence per year of sudden death in the adolescent population.
- ECG is abnormal in 75-95% of patients
- Echo is usually diagnostic
- Recommendations: exclusion from competitive sports

Hypertrophic Cardiomyopathy

Normal

HCM

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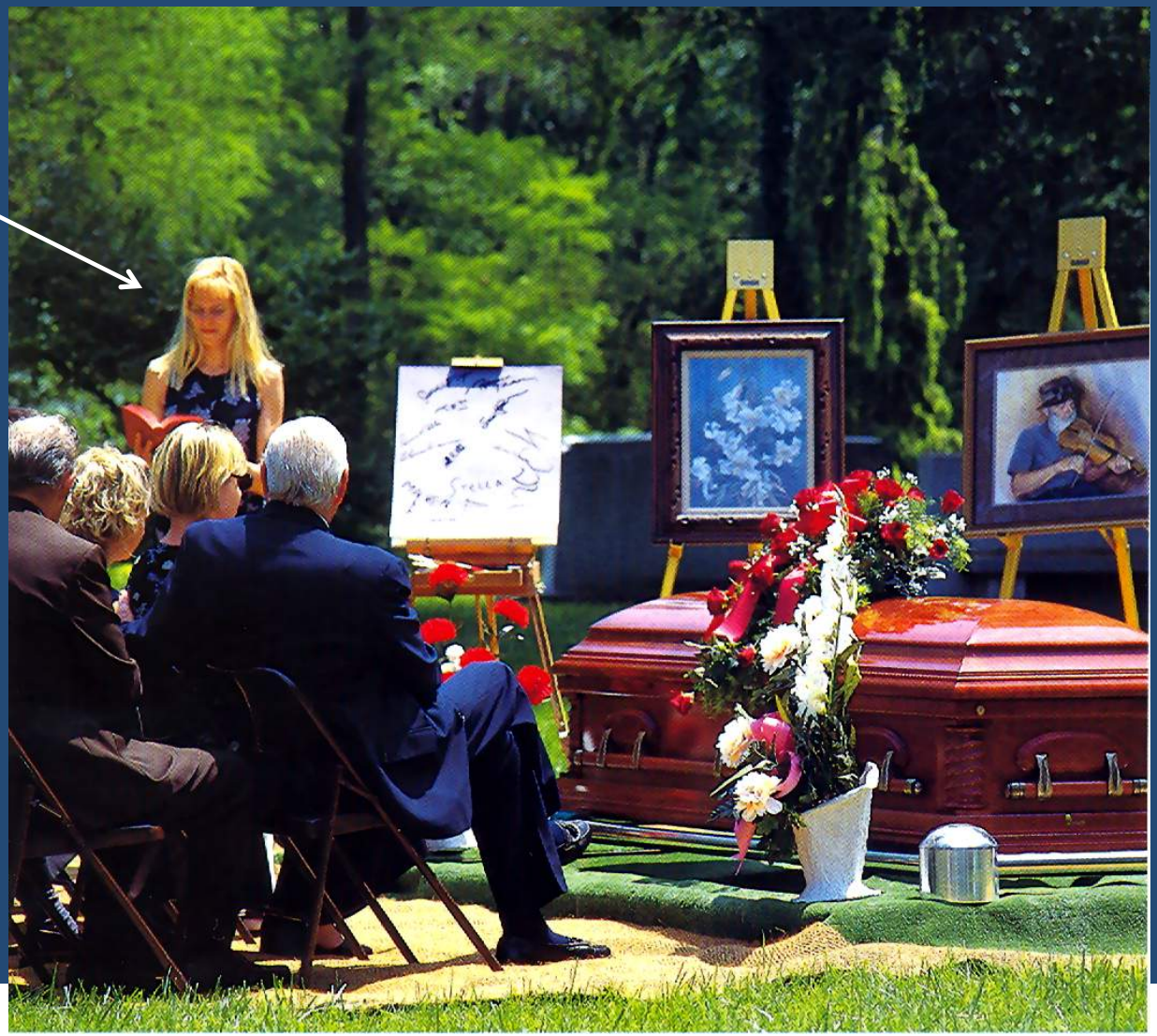
Hypertrophic Cardiomyopathy

- Symptoms: chest pain, syncope during exertion or immediately following exercise, or palpitations.
- First symptom may be sudden death.
- Physical findings:
  - heart murmur of left ventricular outflow tract obstruction or mitral regurgitation.
  - Murmur decreases from standing to squatting.
  - Then increases dramatically from squatting to standing.
  - Murmur increases with valsalva

Case #2: Brother just died.

Your patient



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Genetic Testing and SCD

About 350,000 people in the U.S. die from sudden cardiac death each year. (represented by the gray background)

About 10,000 of these are young people between the ages of 1 and 40.

Following autopsy of the 10,000 young people, **2/3** of the cases can be explained by something that shows up on autopsy.

Of those approximately 3,300 cases where the autopsy doesn't reveal anything, **25%** of the cases can be explained through genetic testing.

In the other third of the 10,000 cases, an autopsy shows the person's heart appears to be normal.

The rest of the deaths among people 1 through 40 remain unexplained.

Source: Mayo Clinic Windland Smith Rice Sudden Death Genomics Laboratory

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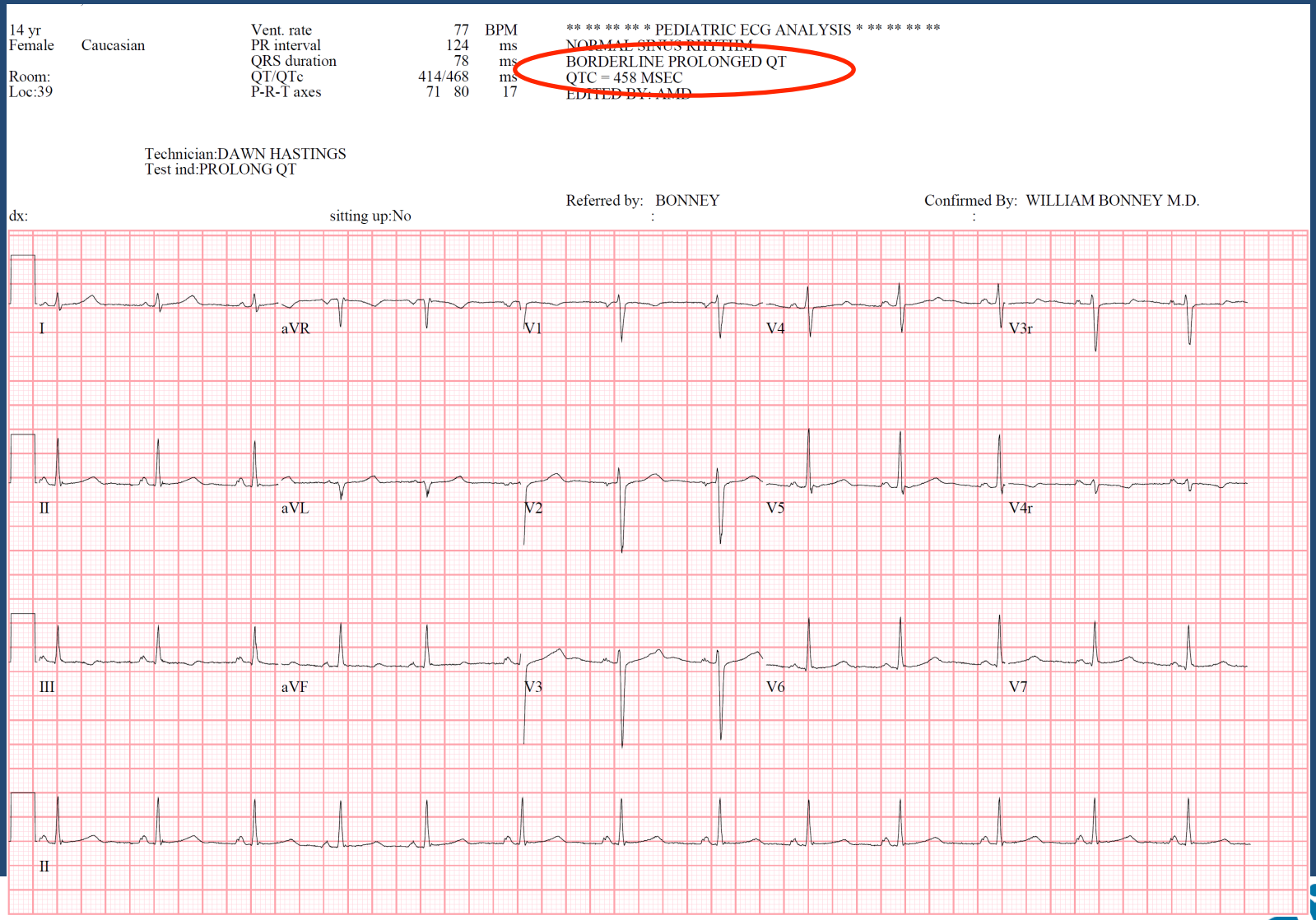
Here's the ECG you don't want.



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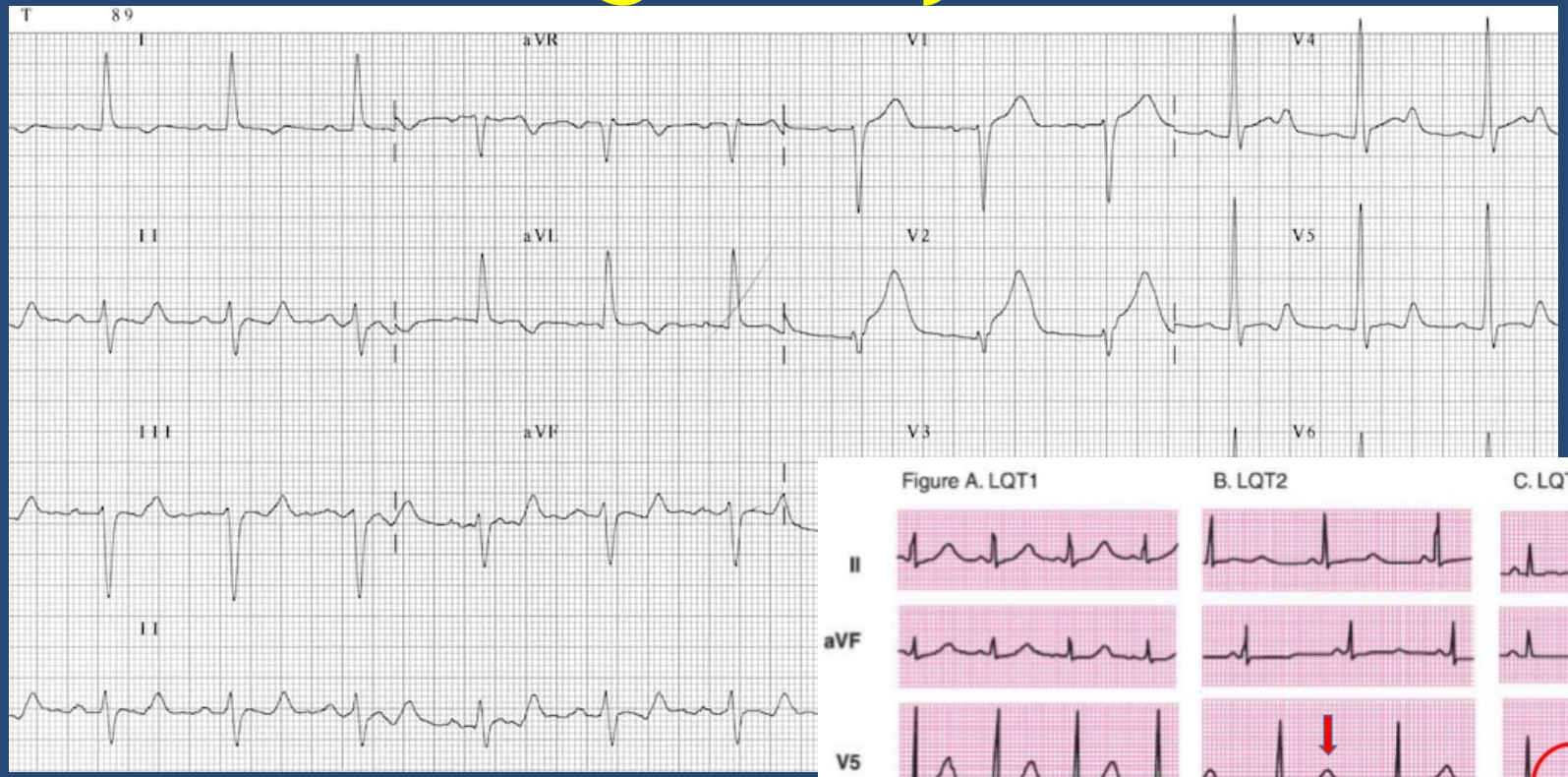
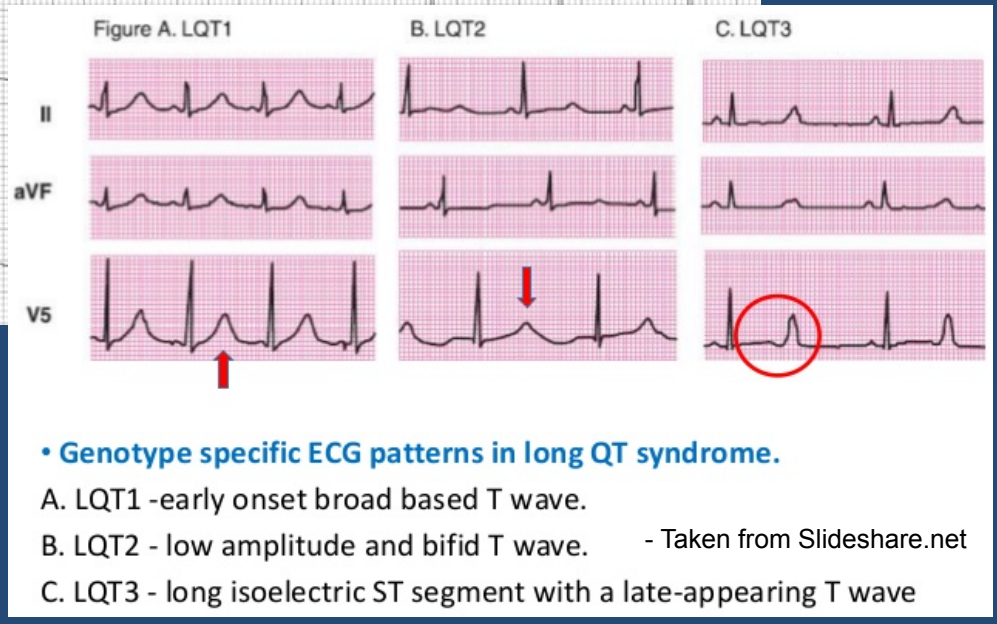
Long QT Syndromes





- **Genotype specific ECG patterns in long QT syndrome.**

A. LQT1 -early onset broad based T wave.

B. LQT2 - low amplitude and bifid T wave.

C. LQT3 - long isoelectric ST segment with a late-appearing T wave

- Taken from Slideshare.net

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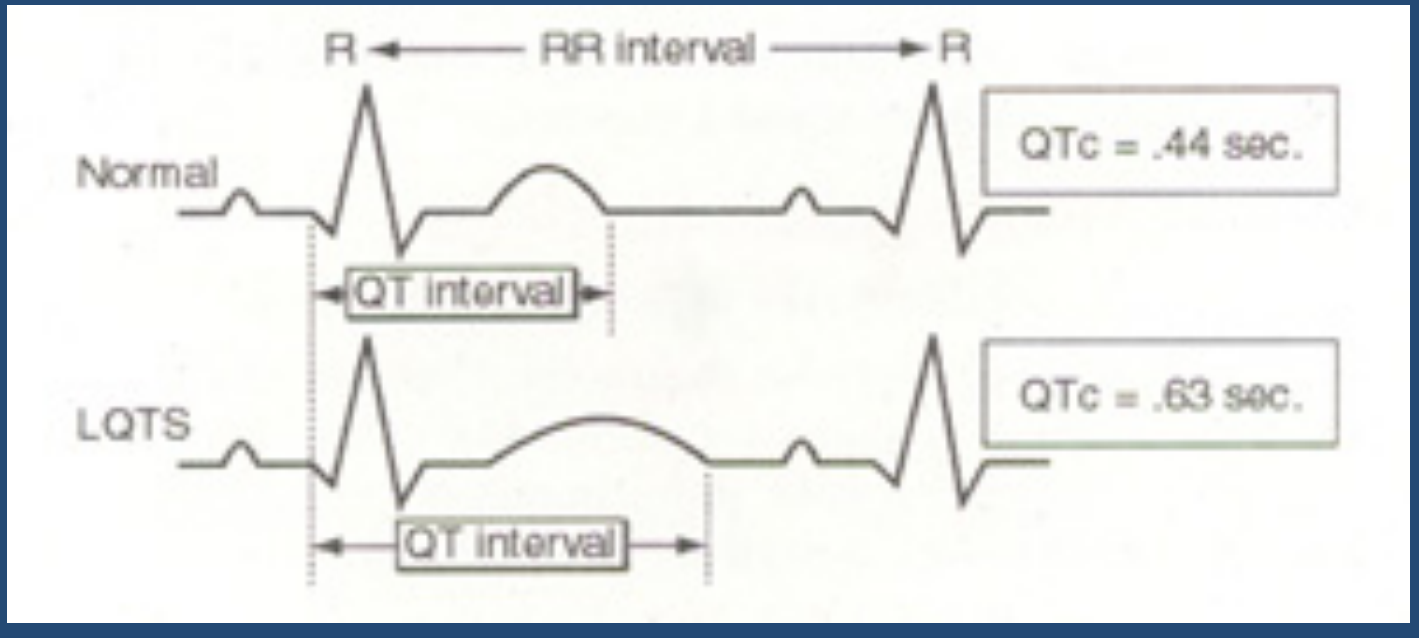
Long QT Syndrome

Inherited disorder (AD) with prolonged QTc interval.

- at least 12 different associated genotypes.
  - Sensitivity of current genotyping is ~75%
- Baseline ECG is abnormal in 90%
  - > 450 ms in males
  - > 460 ms in females
- ECG may be normal in 10 – 15% of known carriers of one of the associated gene mutations

Corrected QT Interval

$$QTc = \frac{QT}{\sqrt{RR}}$$



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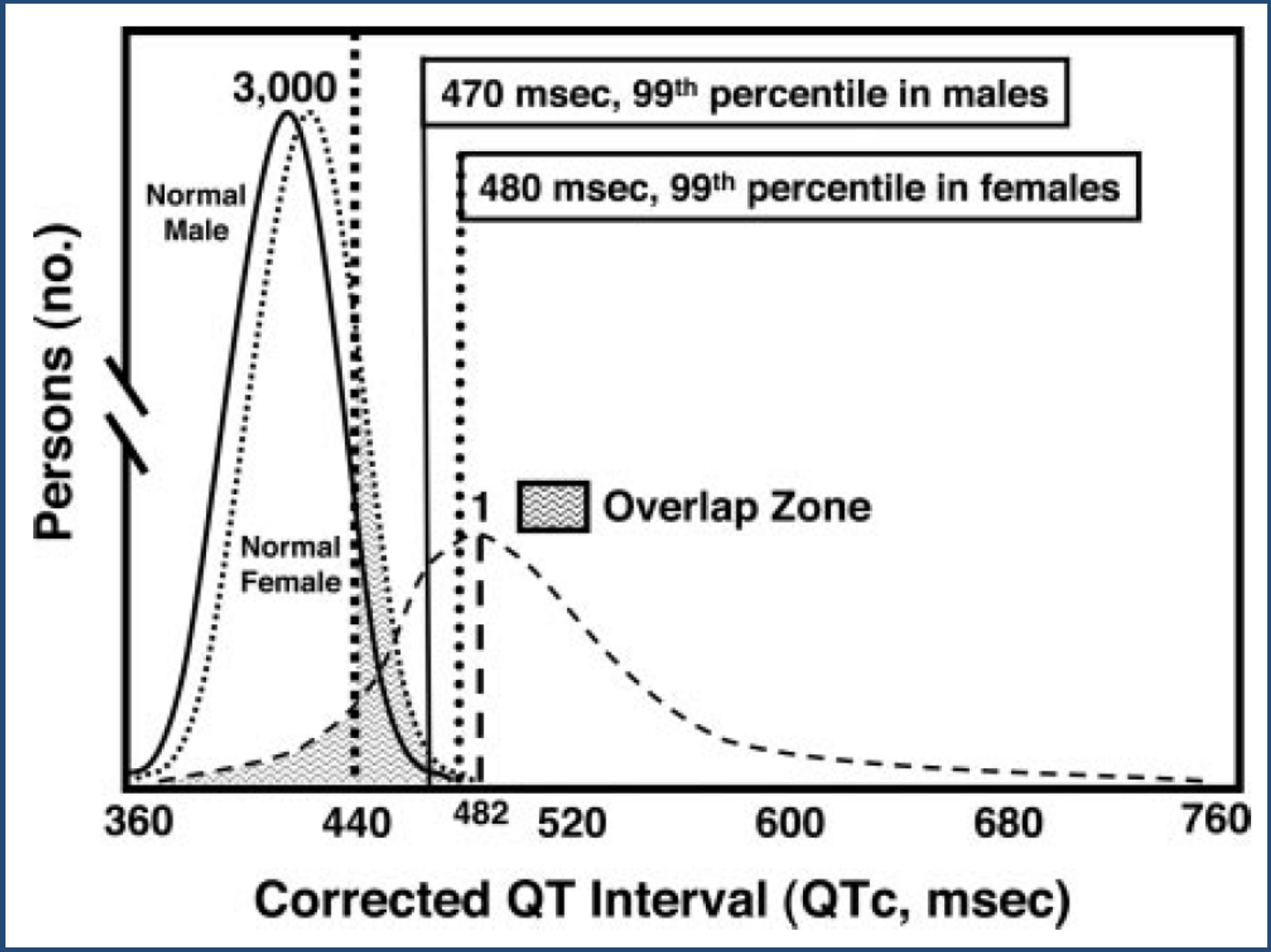
Normal versus Long QT



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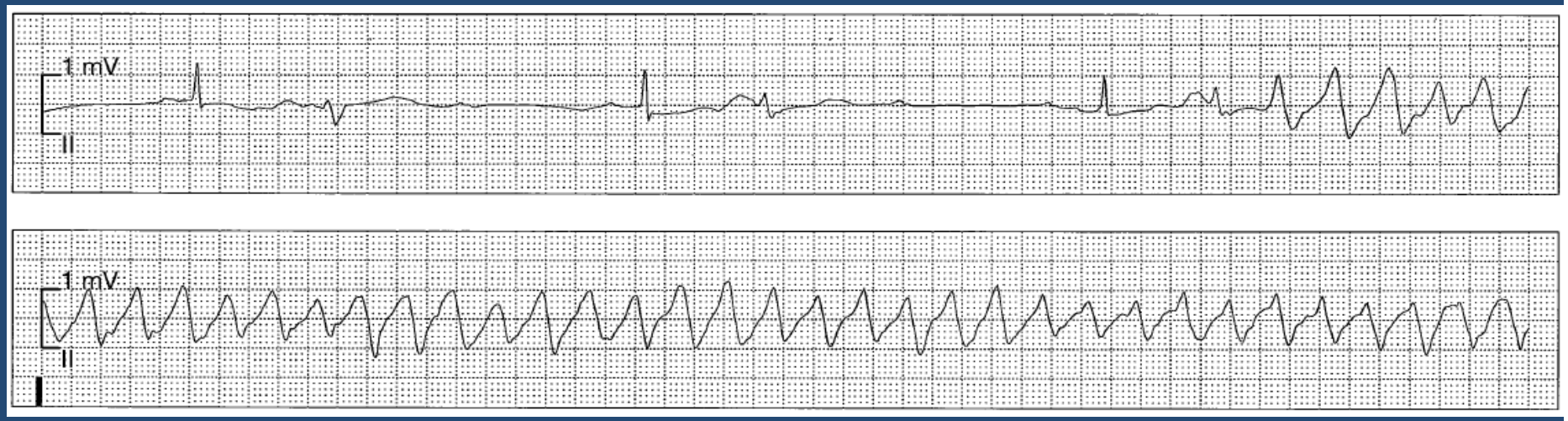
Long QT Syndrome

- Patients at risk for developing syncope and sudden death (with or without exercise)
- Risk of sudden death is 6% by 40 years of age

❖**Restriction from competitive sports.**

Torsades de Pointes



-Bradycardia

-Long QT

-Long/Short Coupling interval

-Causes immediate collapse

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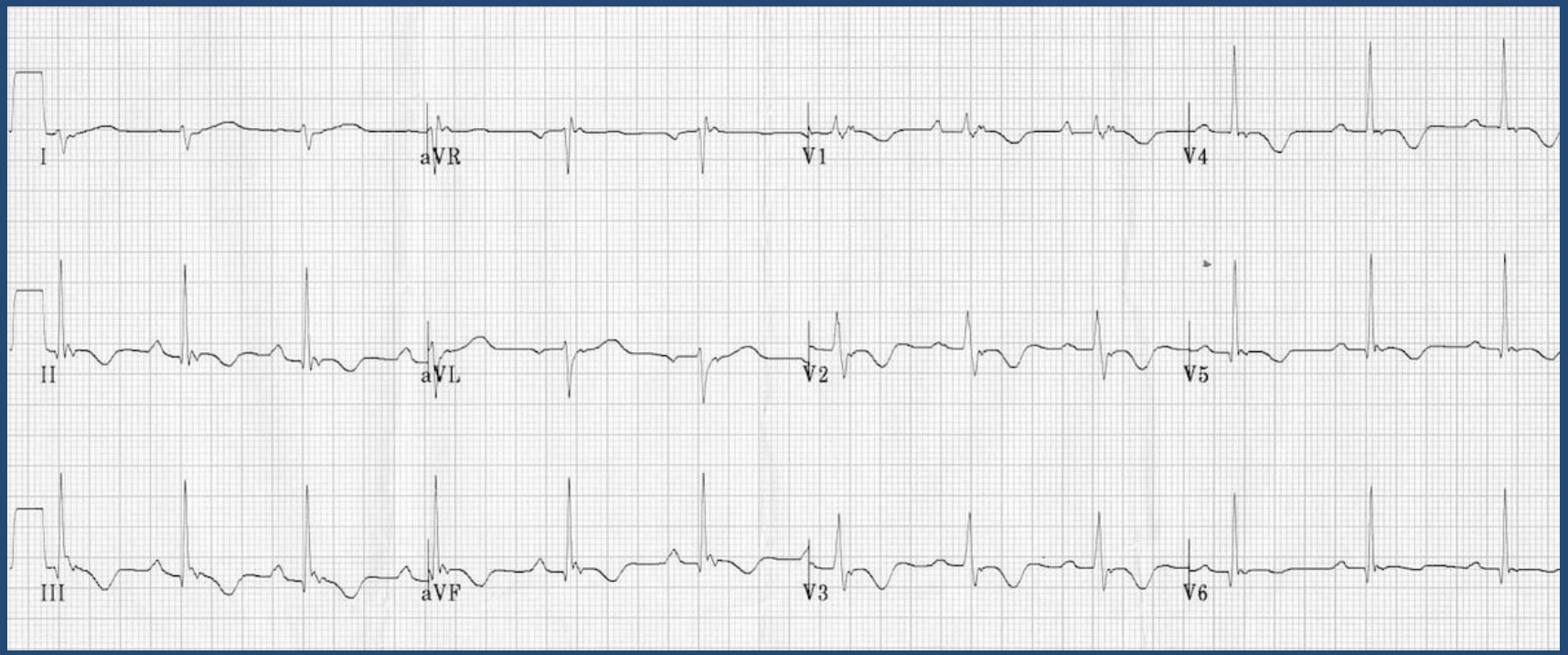
Case 3: Syncope day after running a half marathon.

- Patient reports recurrent syncope/ presyncope.
- Patient had syncope in ER and found in polymorphic VT requiring defibrillation.
- Echo with PFO but no major abnormalities. RV and LV EF normal.

? Arrhythmogenic RV Dysplasia



-Epsilon wave:
-Inverted T waves V1-V3

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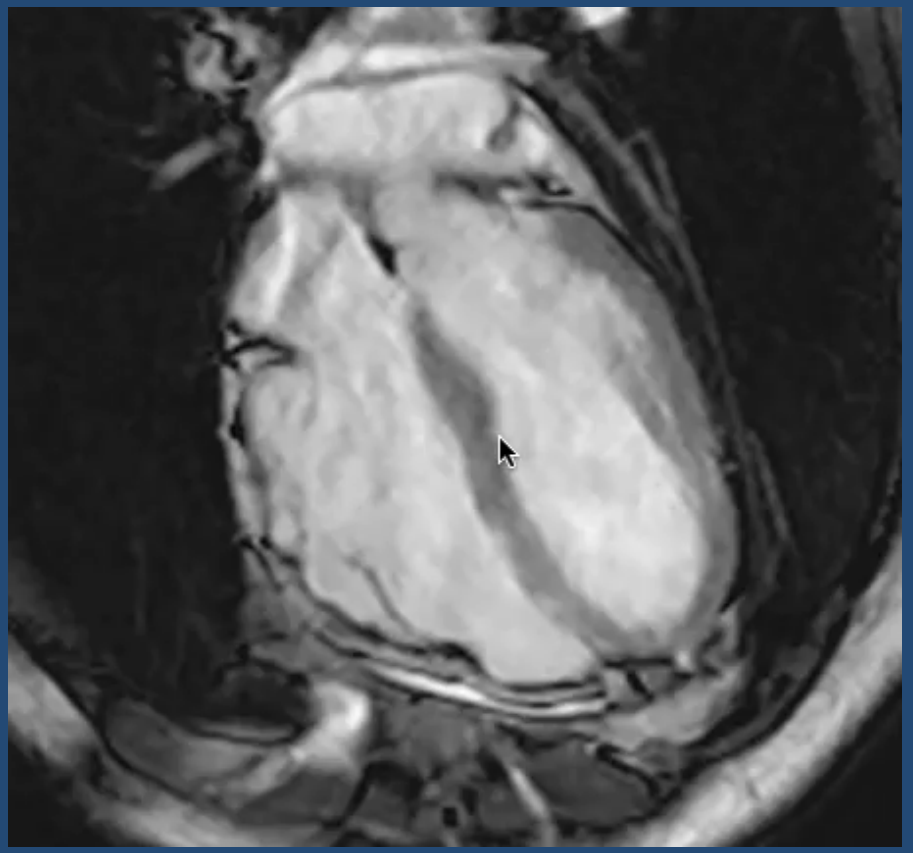
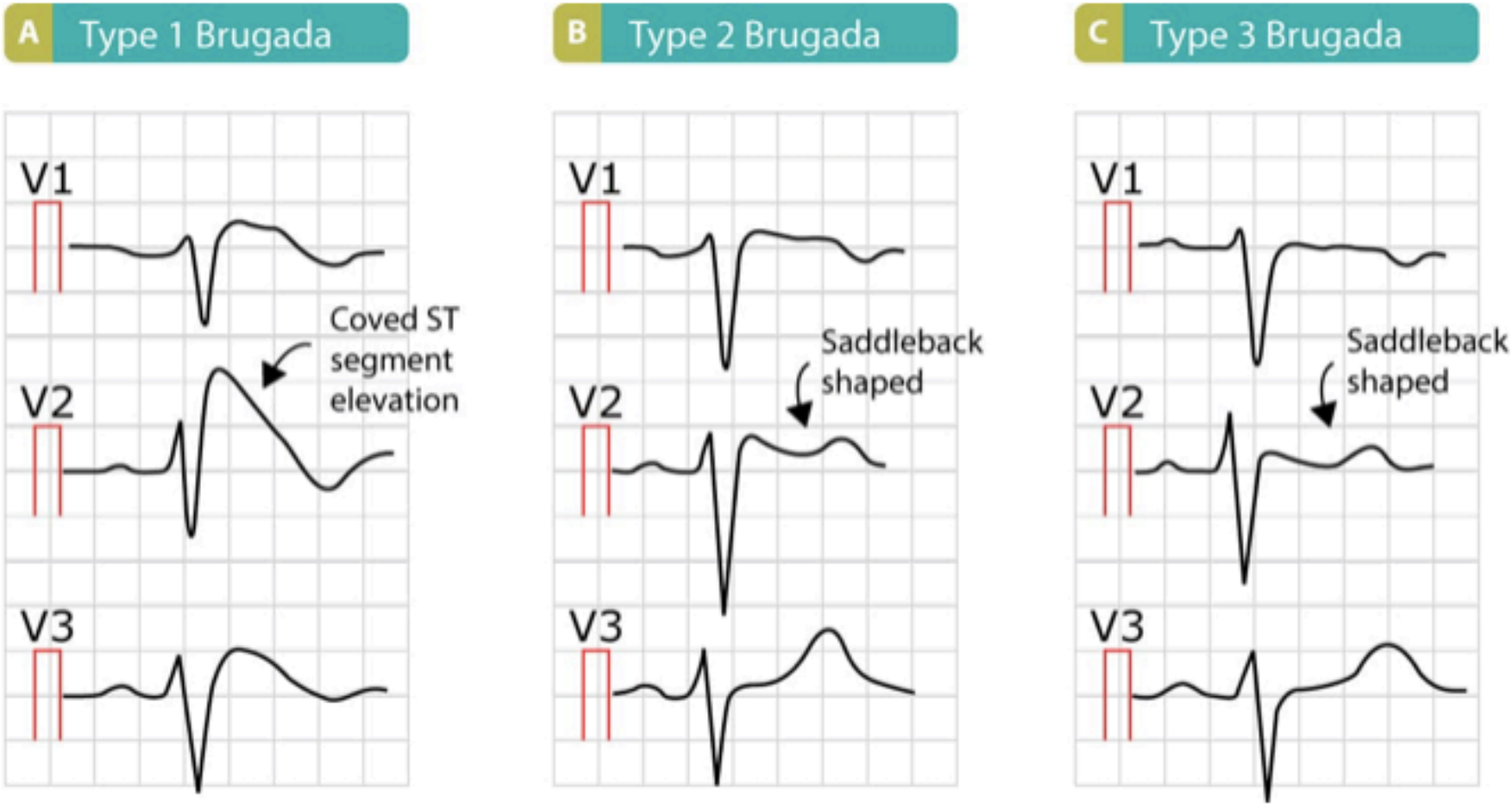
Cardiac MRI Indicates ARVD

- Cardiac MRI showed RV dysfunction in free wall near apex with RVEF<40%. RV EDV/BSA >100mL/m2. Meets major criteria for ARVD.



https://radiopaedia.org/cases/arrhythmogenic-right-ventricular-cardiomyopathy?lang=us

Brugada Syndrome



Taken from ecgwaves.com

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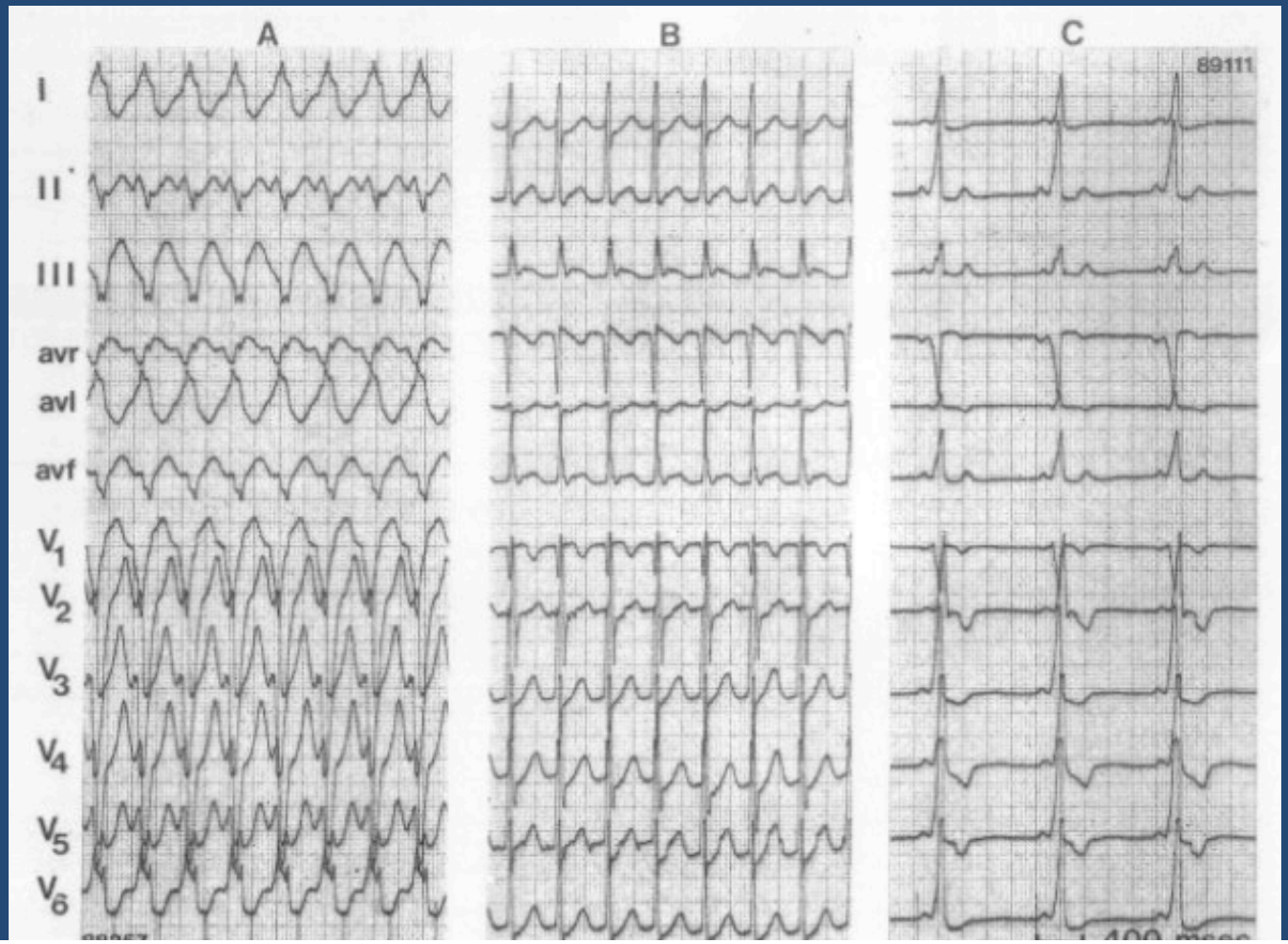
Wolff-Parkinson-White



A: Antidromic (down AP and up AVN) tachycardia, B: Orthodromic AVRT (down AV node and up L lat AP), C: SR preexcited.

AP Localization:
1. – or ± in I: Left lateral
2. QS in II: MCV left posteroseptal
3. – or ± in V1: Septal
4. Absent 1-3: R free wall (delta prior to completion of P wave)

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Wolff-Parkinson-White Syndrome

- Most common pre-excitation syndrome
- Prevalence: 1 – 3/1000
- Predisposes to re-entrant SVT and Atrial fibrillation
- SCD may occur during atrial fibrillation if rapid conduction down the accessory pathway leads to VT and VFib
- Risk stratification is done in the EP lab
- **Treatment:**
  - Beta blocker medication
  - Radio-Frequency ablation (RFA) is curative in >95% cases

Who is at risk for SCD as adult?

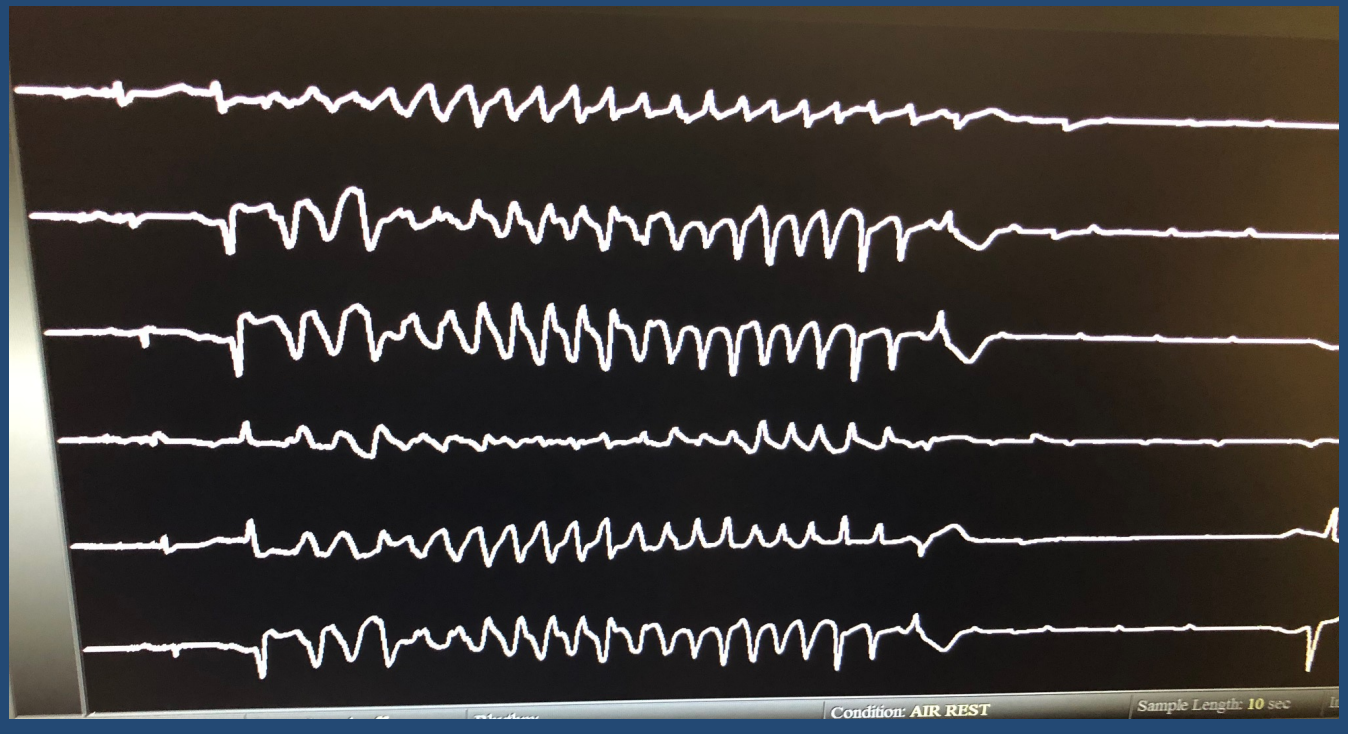
- A prior SCD
- Family history of SCD
- Congestive Heart Failure (CHF)
- Have had a Myocardial Infarction (MI)
- Ejection Fraction (EF) less than or equal to 35%

Case 4: Recurrent syncope presenting to ER. Not all heart block is created equal.

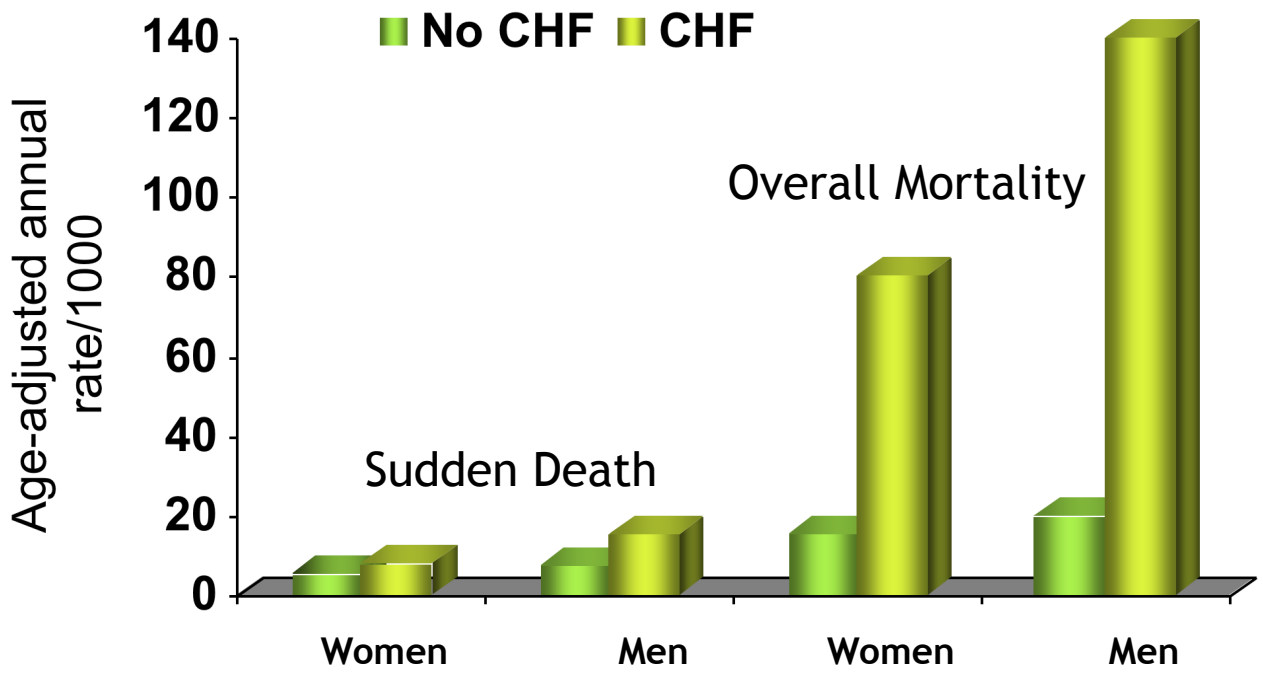
- Bradycardia-dependent VF is uncommon.
- This patient found to have complete heart block and was stable with HTN on telemetry floor. Planning on pacemaker implantation the next day.
- Called about runs of asystole on telemetry and decided to proceed with permanent device same day.

Case 4 Continued

- Recorded on EP lab monitor as patient was being prepared for pacemaker implantation.



In people diagnosed with CHF, sudden cardiac arrest occurs at 6-9 times the rate of the general population



CHF predicts increased sudden death and overall mortality.

During a 39-year follow-up of subjects in the Framingham heart Study, the presence of CHF significantly increased sudden death and overall mortality in both men and women.[2]

[1] Kannel WB, Wilson PWF, D'Agostino RB, Cobb J. Sudden coronary death in women. *Am Heart J*. 1998 Aug; 136: 205-212

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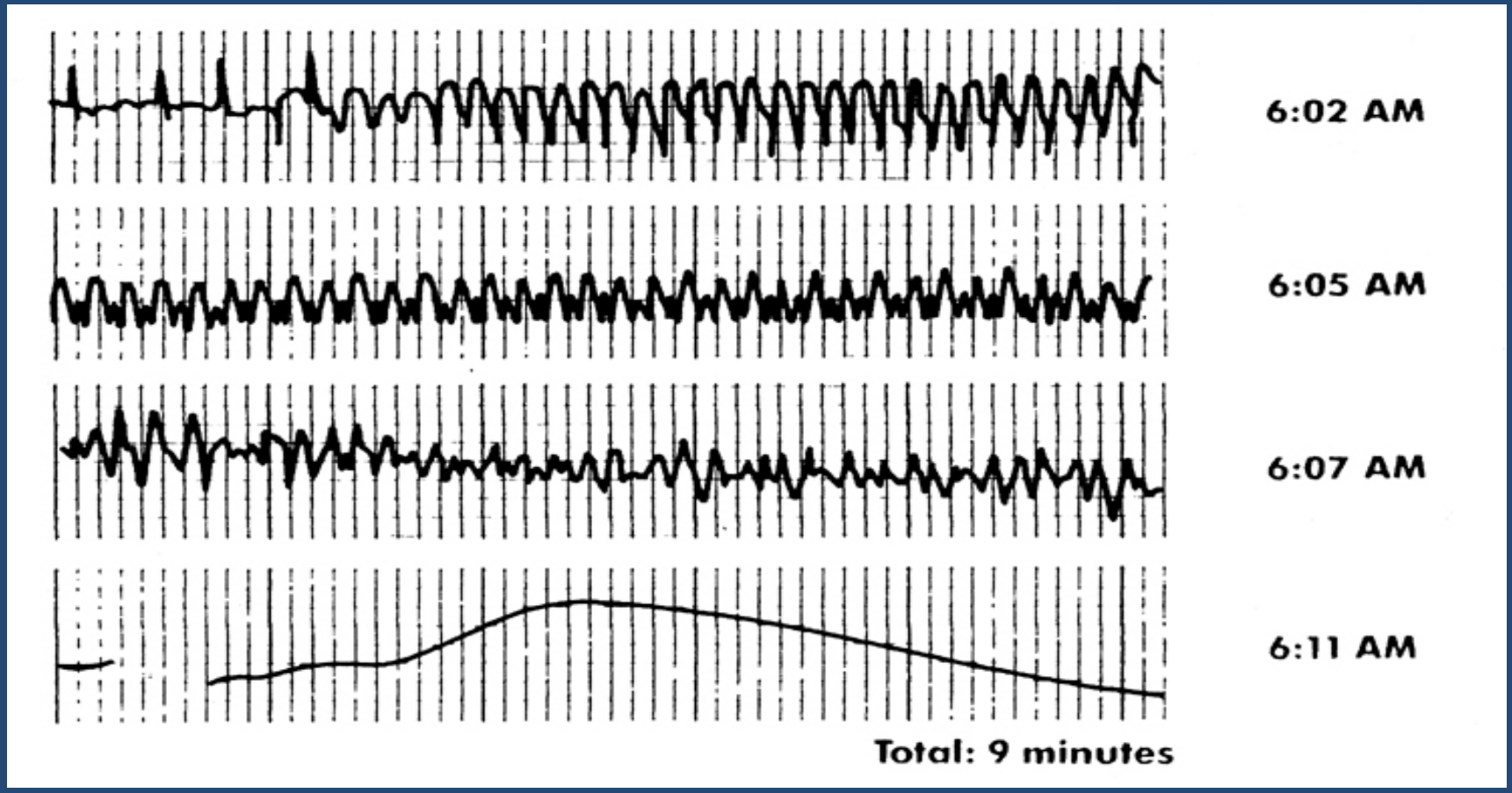
People who've had a heart attack have a sudden death rate that's 4-6 times that of the general population

- Studies show that a previous MI can be identified in as many as 75% of SCD patients
- A previous MI raises the one-year risk of SCD by 5% as a single risk factor
- The five-year risk of SCD for patients with a previous MI, non-sustained, inducible, non-suppressible VT, and a LVEF < 40% is 32%

What does SCD look like?



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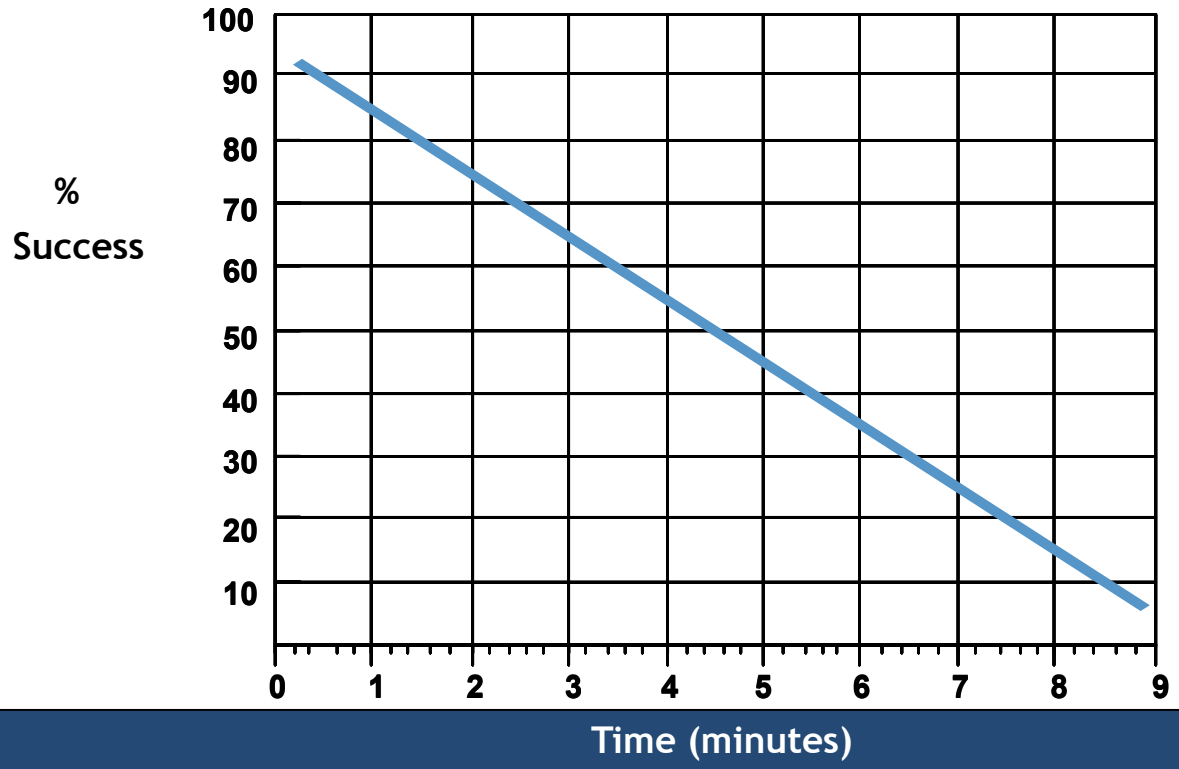
Urgency of SCD

Resuscitation Success vs. Time

Chance of success reduced 7-10% every minute



Adapted from text: Cummins RO, 1998. *Annals of Emergency Medicine.* 18:1269-1275.

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Cardiopulmonary resuscitation

- Appropriate mechanical compression during CPR essential to maintain circulation until defibrillation.
- Automated device to prevent exhaustion and inadequate CPR.
  - AutoPulse: may cause visceral damage.
  - LUCAS: no obvious visceral damage compared to manual CPR.
  - Primary outcome in 11.6% AutoPulse, 7.4% LUCAS, and 6.4% manual compressions.

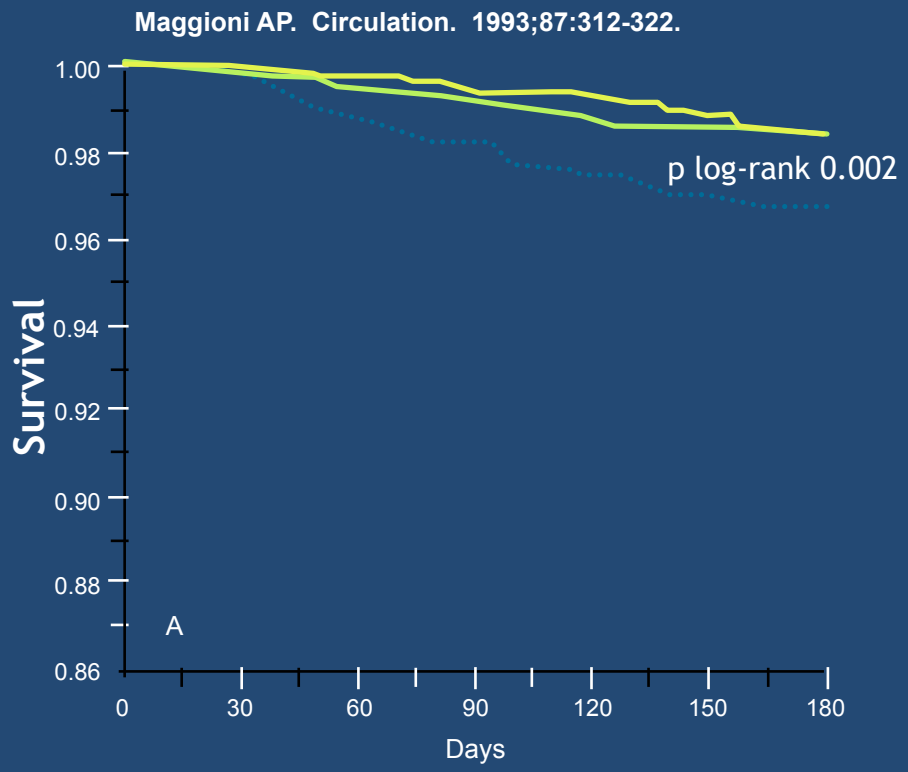
-Koster RW et al, "Safety of mechanical chest compression devices AutoPulse and LUCAS in cardiac arrest: a randomized clinical trial for noninferiority," Eur Heart J (2017), V. 38, 3006-3013.

SCD Survival = Early Defibrillation

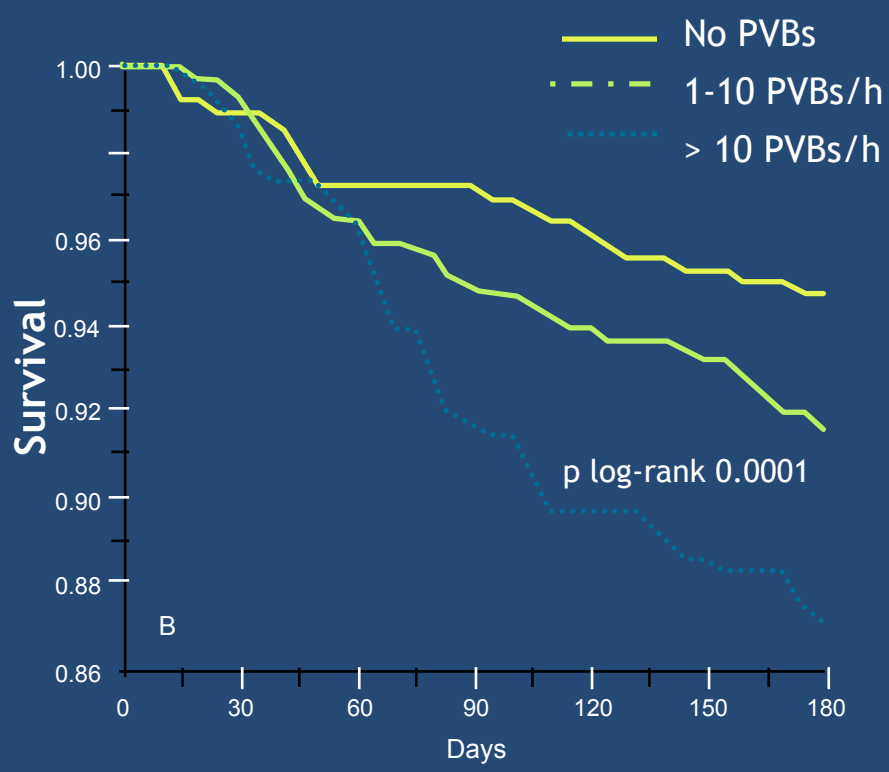
- Only effective treatment for SCD is an electrical shock delivered by:
- - Automated external defibrillator (AED) or
- - Implantable cardioverter-defibrillator (ICD)
- Time is critical – each minute of delay before defibrillation reduces survival rates by about 10%

Drone delivery of AED's.

Reduced LVEF remains the single most important risk factor for overall mortality and sudden cardiac arrest



Patients without LV Dysfunction
(LVEF >35%)



Patients with LV Dysfunction
(LVEF ≤ 35%)

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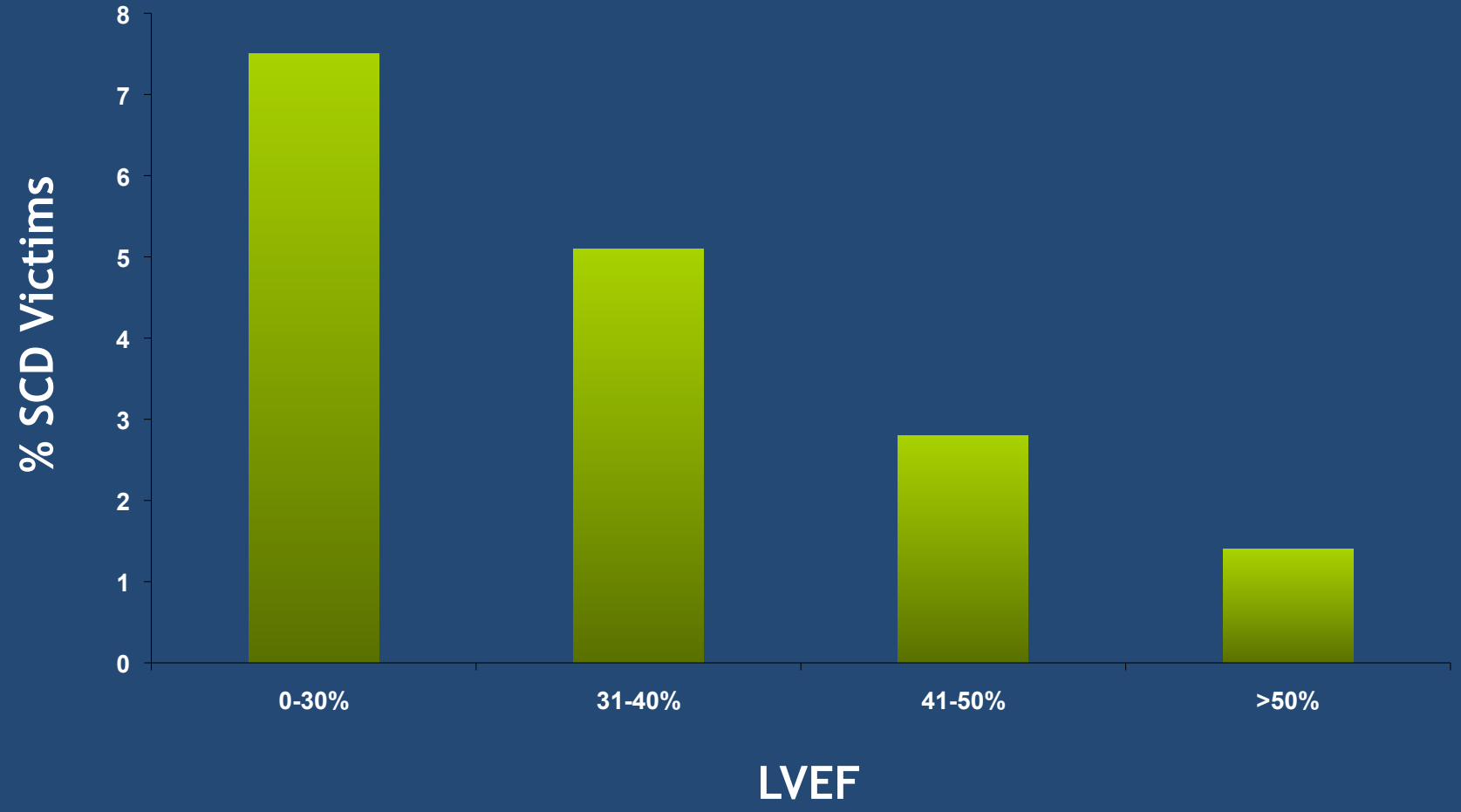
EF and SCD Incidence



Gorgels PMA. *European Heart Journal*. 2003;24:1204-1209.

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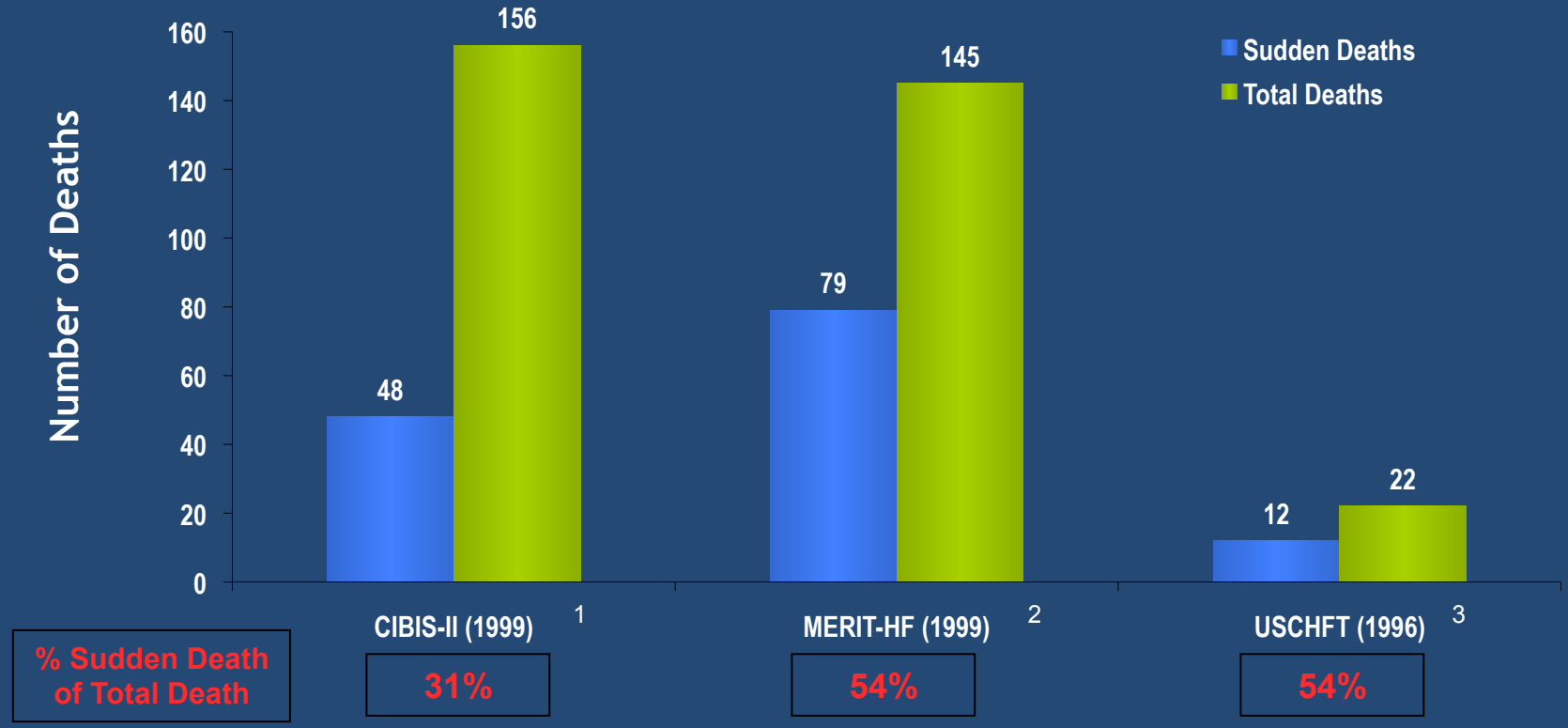
Residual Risk of SCD in Treatment Arms of CHF Beta Blocker Trials



[1] CIBIS-II Investigators. *Lancet.* 1999;353:9-13.
[2] MERIT-HF Study Group. *Lancet.* 1999;353:2001-2007.
[3] Packer M. *N Engl J Med.* 1996:334:349-355.

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SCD in Heart Failure

- Despite improvements in medical therapy, symptomatic HF still confers a 20-25% risk of pre-mature death in the first 2.5 yrs after diagnosis.

  ≈ 50% of these premature deaths are SCD (VT/VF)

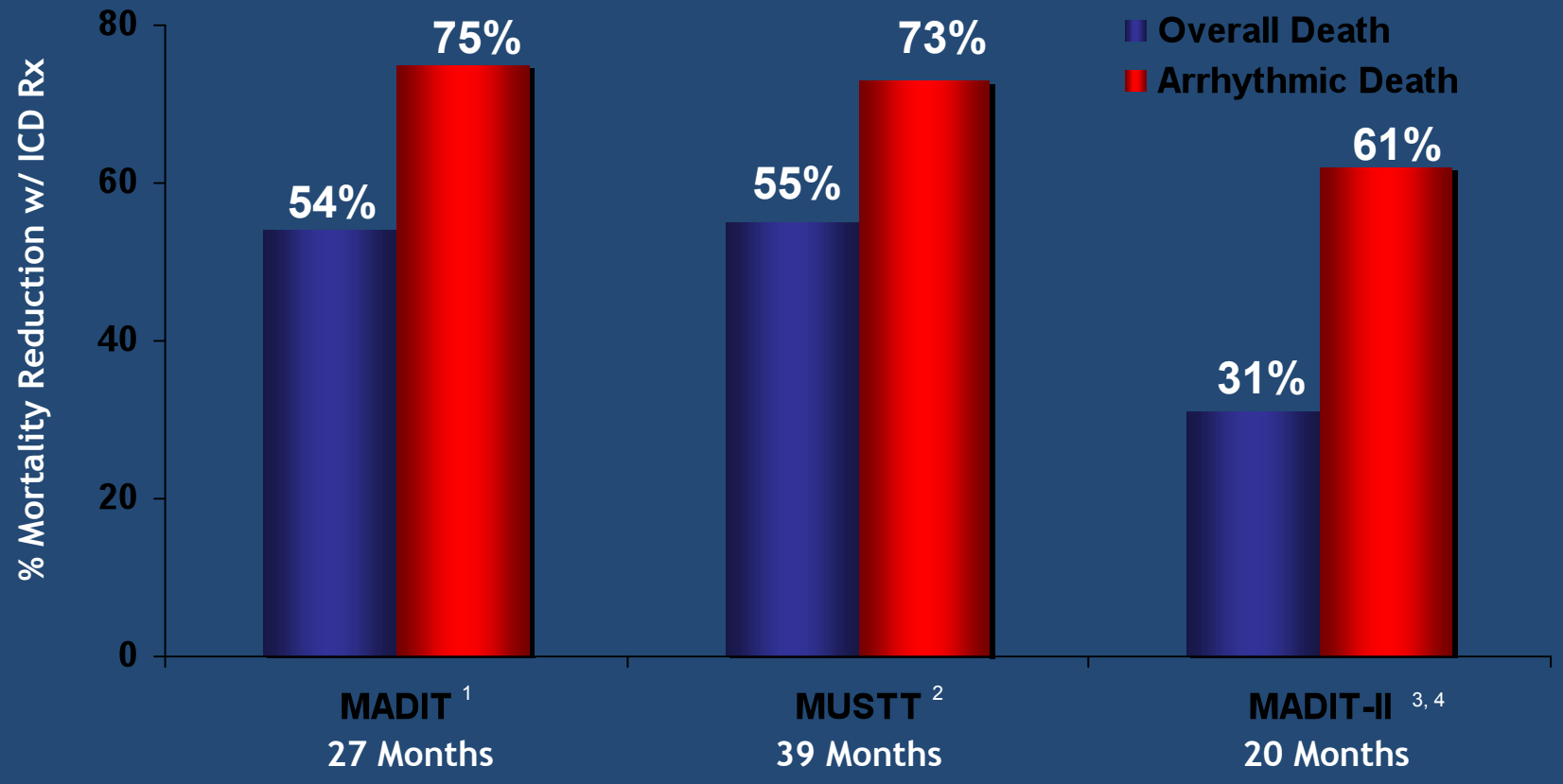
[1] Bardy G. The Sudden Cardiac Death-Heart Failure Trial (SCD-HeFT) in Woosley RL, Singh S. *Arrhythmia Treatment and Therapy.* Copyright 2000 by Marcel Dekker, Inc. , pp. 323-342,

[2] Sweeney MO. *PACE*. 2001;24:871-888.

Case 5: 65yo CAD s/p multiple MI's over the years with SOB

- Has had progressive shortness of breath despite optimal medical management.
- He has undergone coronary artery bypass surgery and a recent catheterization reveals that his native coronary arteries are still severely diseased but his bypass grafts are all still working properly.
- His EF is found to be 25% and it is recommended that he undergo a defibrillator implantation.

ICD Mortality Benefits in Post-MI Patients with LV Dysfunction



[1] Moss AJ. *N Engl J Med*. 1996;335:1933-40.
[2] Buxton AE. *N Engl J Med.* 1999;341:1882-90.
[3] Moss AF. *N Engl J Med.* 2002;346:877-83.
[4] Moss AJ. Presented before ACC 51st Annual Scientific Sessions, Late Breaking Clinical Trials, March 19, 2002.

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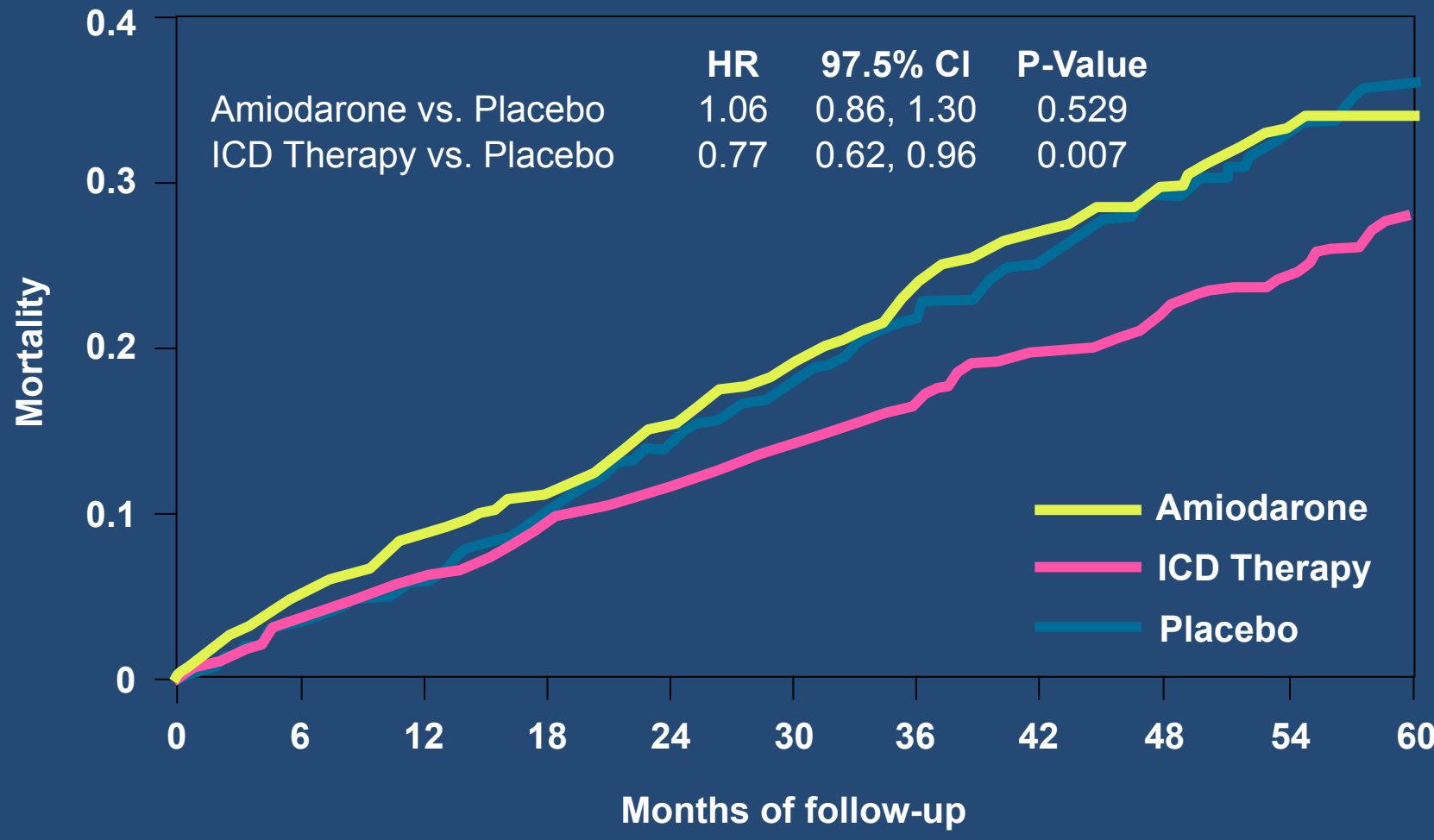
ICD effect on all-cause mortality compared to placebo* in patients with either ischemic or non-ischemic NYHA Class II and III CHF and EF < 35%.



* Double-blind for drug therapy

Bardy G. *N Engl J Med*. 2005;352:225-37.

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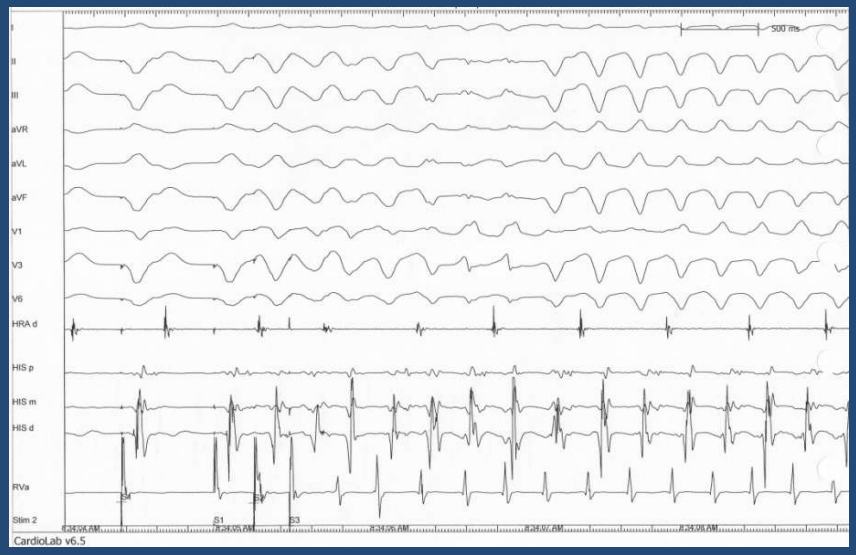
Common Indications for Implantable Cardioverter Defibrillators (ICD)

- ICD are of proven benefit in the prevention of SCD in heart failure patients with ischemic and non-ischemic cardiomyopathy.
  - EF ≤ 0.35
  - EF ≤ 0.40 with NSVT and EPS +



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ICD's are currently the most effective prevention of SCD but...

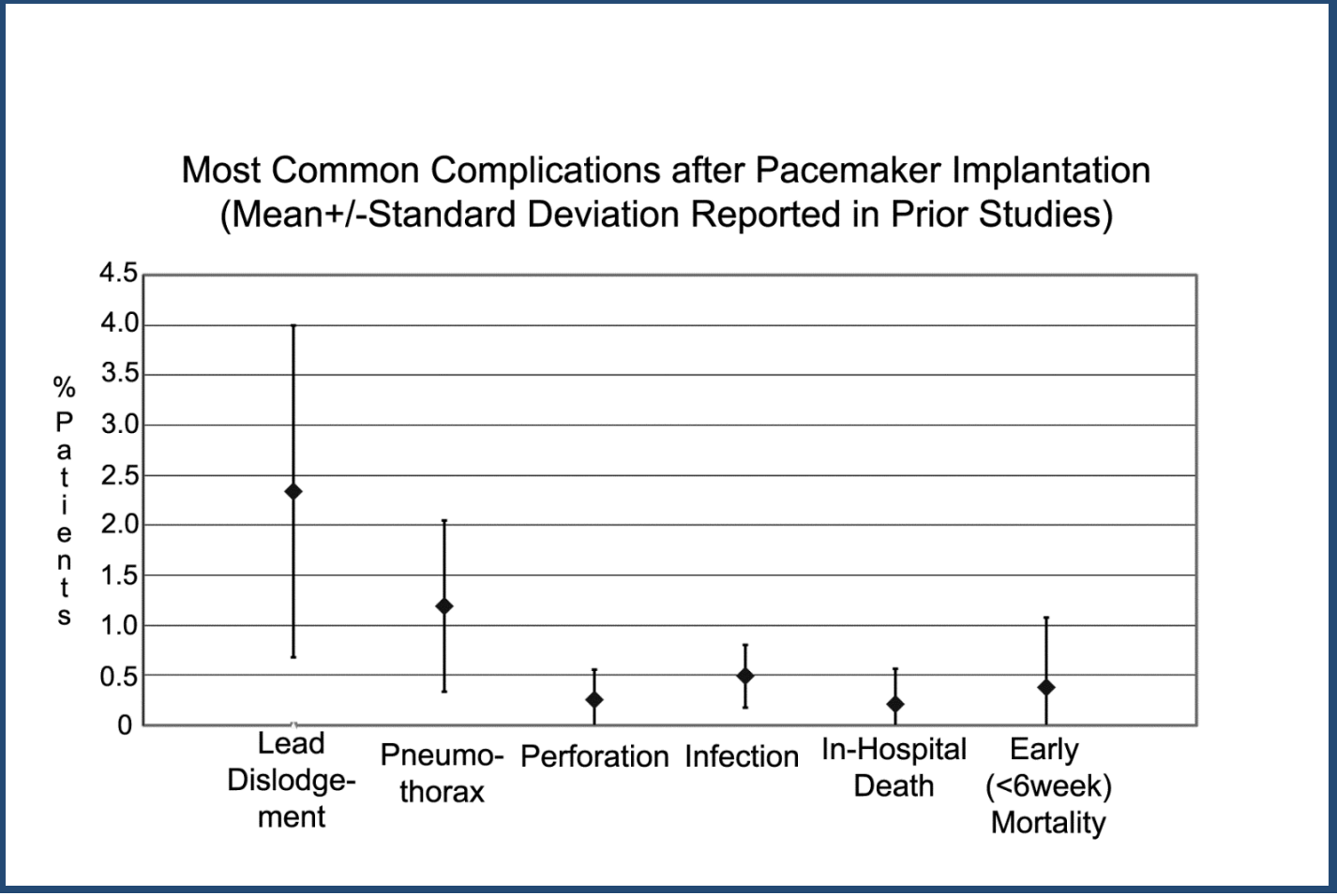
- Transvenous systems require implant in the vascular system which can cause problems with 2nd or 3rd device needed or thrombus/infection occurs
  - For extractions performed over a nearly 13-year period, the overall rate of in-hospital major complications was 10.4%, including a 4.1% rate of mortality.
  - There is debate that complication rate is not significantly lowered in high volume centers.
- Subcutaneous ICD cannot provide routine pacing functions.

- Hosseini SM, Rozen G, Kaadan MI, et al. Safety and in-hospital outcomes of transvenous lead extraction for cardiac implantable device related infections: analysis of 13 years of inpatient data in the United States. J Am Coll Cardiol EP. 2019;Epub ahead of print.
-Bongiorni MG et al. The European Lead Extraction ConTRolled (ELECTRa) study: a European Heart Rhythm Association (EHRA) Registry of Transvenous Lead Extraction Outcomes. Eur Heart J 2017;38:2995–3005.
-

Complications of Device Implantation

- Major complications (1-3%): death, cardiac arrest, cardiac perforation, cardiac valve injury, coronary venous dissection, hemothorax, pneumothorax, transient ischemic attack, stroke, myocardial infarction, pericardial tamponade, and arterial-venous fistula.
- Minor complications (3-7%): drug reaction, conduction block, hematoma or lead dislodgement requiring reoperation, peripheral embolus, phlebitis, peripheral nerve injury, and device-related infection.

Complications of Device Implantation



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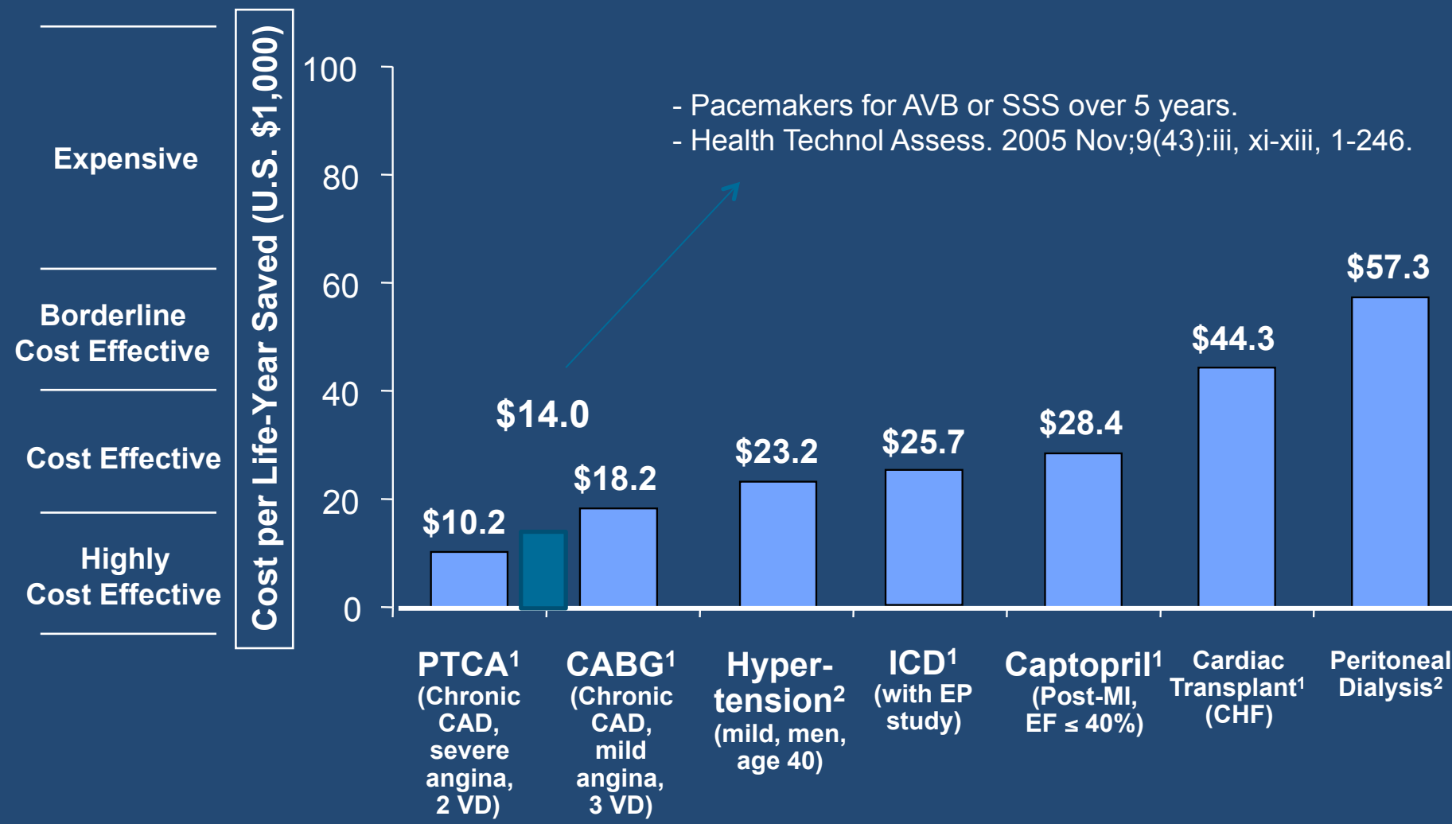
Cost-Effectiveness



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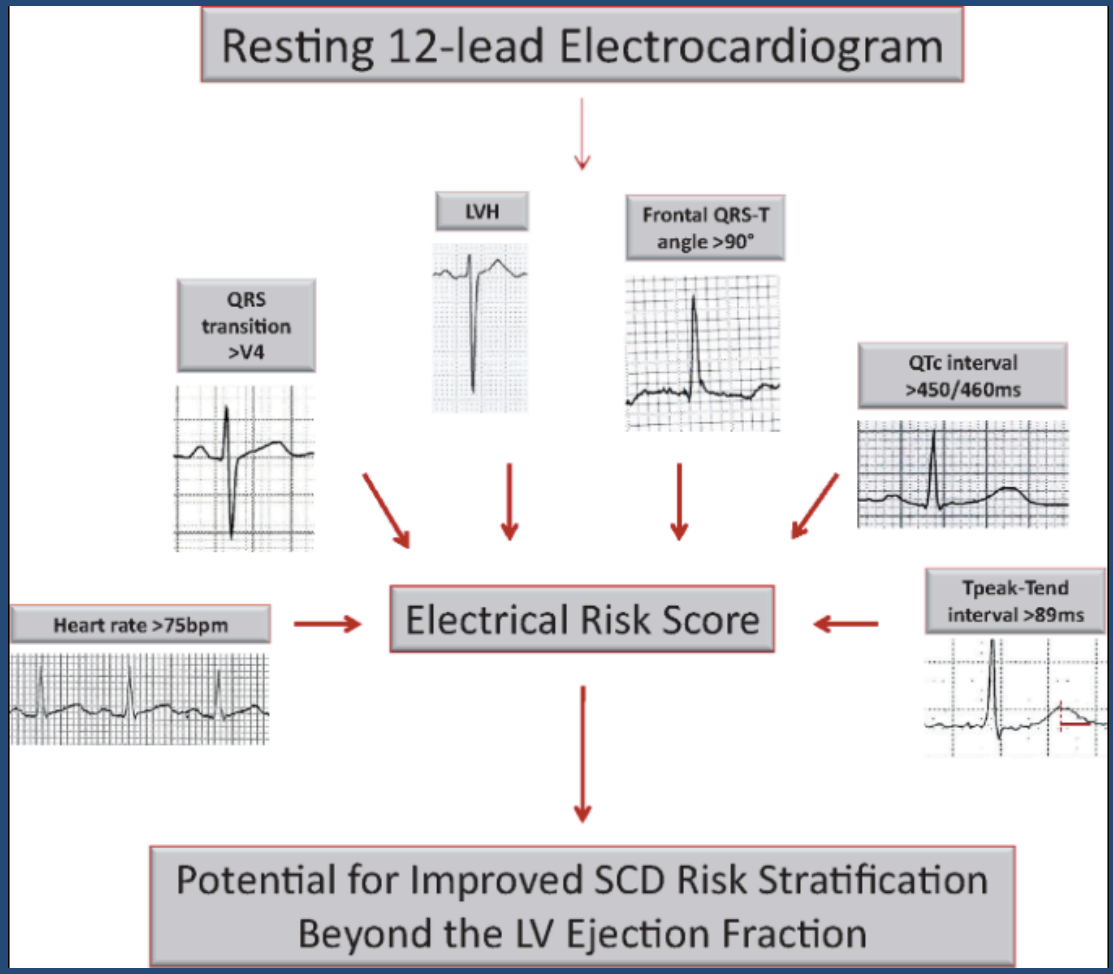
Can we predict who will experience SCD?



- Aro AL, Reinier K, Rusinaru C, Uy-Evanado A, Darouian N,, Phan D,, Mack WJ,, Jui J,, Soliman EZ,, Tereshchenko LG, Chugh SS. Electrical risk score beyond the left ventricular ejection fraction: prediction of sudden cardiac death in the Oregon Sudden Unexpected Death Study and the Atherosclerosis Risk in Communities Study. Eur Heart J 2017;38:3017–3025.

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Prevention of SCD: Young Athletes

SCIENTIFIC STATEMENT

Assessment of the 12-Lead Electrocardiogram as a Screening Test for Detection of Cardiovascular Disease in Healthy General Populations of Young People (12–25 Years of Age)

A Scientific Statement From the American Heart Association and the American College of Cardiology

Endorsed by the Pediatric and Congenital Electrophysiology Society and American College of Sports Medicine

1. It is recommended that the AHA 14-point screening guidelines (Table 1) and those of other societies, such as the Preparticipation Physical Evaluation monograph (115), be used by examiners as part of a comprehensive history-taking and physical examination to detect or raise suspicion of genetic/congenital and other cardiovascular abnormalities *(Class I; Level of Evidence C)*.
2. It is recommended that standardization of the questionnaire forms used as guides for examiners of high school and college athletes in the United States be pursued *(Class I; Level of Evidence C)*.

AHA Scientific Statement

Recommendations and Considerations Related to Preparticipation Screening for Cardiovascular Abnormalities in Competitive Athletes: 2007 Update

TABLE. The 12-Element AHA Recommendations for Preparticipation Cardiovascular Screening of Competitive Athletes

Medical history*

Personal history

1. Exertional chest pain/discomfort
2. Unexplained syncope/near-syncope†
3. Excessive exertional and unexplained dyspnea/fatigue, associated with exercise
4. Prior recognition of a heart murmur
5. Elevated systemic blood pressure

Family history

6. Premature death (sudden and unexpected, or otherwise) before age 50 years due to heart disease, in ≥1 relative
7. Disability from heart disease in a close relative <50 years of age
8. Specific knowledge of certain cardiac conditions in family members: hypertrophic or dilated cardiomyopathy, long-QT syndrome or other ion channelopathies, Marfan syndrome, or clinically important arrhythmias

Physical examination

9. Heart murmur‡
10. Femoral pulses to exclude aortic coarctation
11. Physical stigmata of Marfan syndrome
12. Brachial artery blood pressure (sitting position)§

*Parental verification is recommended for high school and middle school athletes.

†Judged not to be neurocardiogenic (vasovagal); of particular concern when related to exertion.

‡Auscultation should be performed in both supine and standing positions (or with Valsalva maneuver), specifically to identify murmurs of dynamic left ventricular outflow tract obstruction.

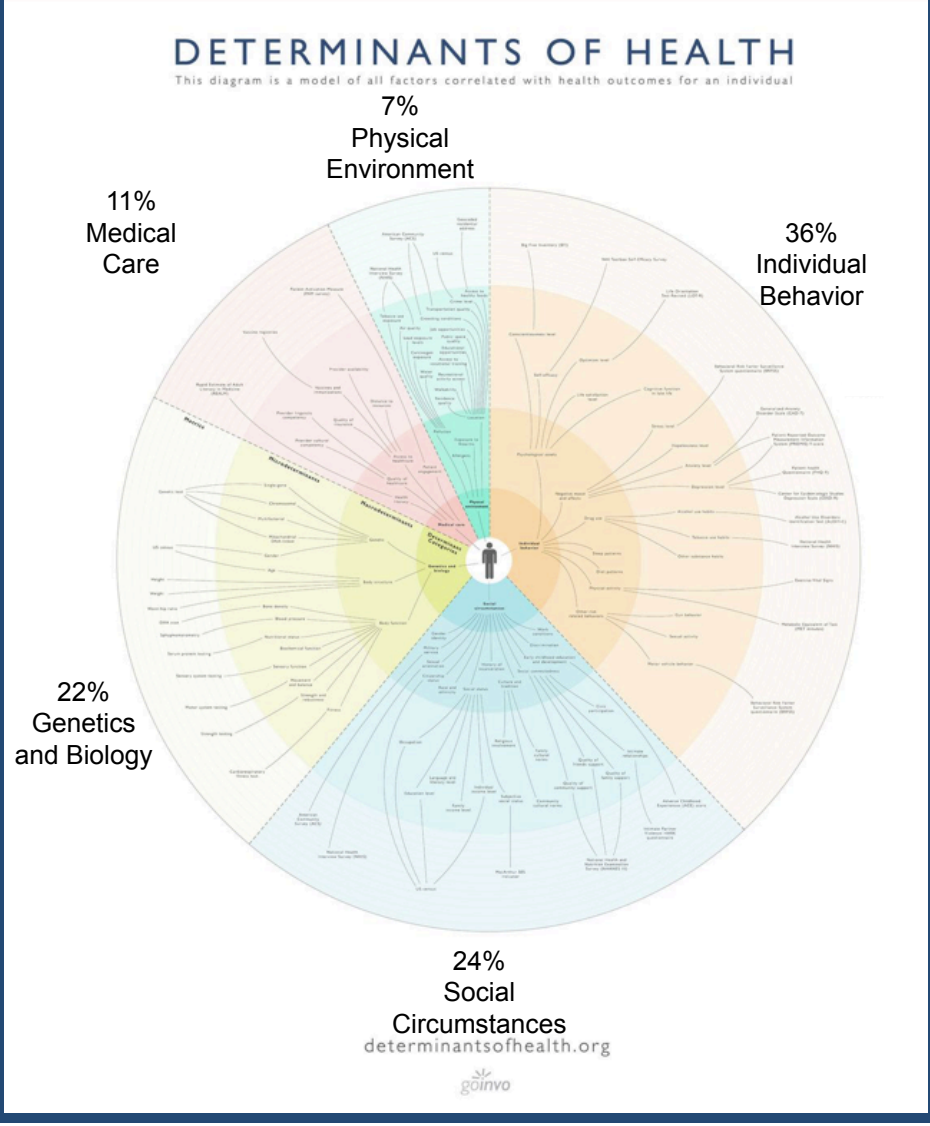
§Preferably taken in both arms.[37]

Should we get ECG's on all young athletes?

3. Screening with 12-lead ECGs (or echocardiograms) in association with comprehensive history-taking and physical examination to identify or raise suspicion of genetic/congenital and other cardiovascular abnormalities may be considered in relatively small cohorts of young healthy people 12 to 25 years of age, not necessarily limited to athletes (e.g., in high schools, colleges/universities, or local communities), provided that close physician involvement and sufficient quality control can be achieved. If undertaken, such initiatives should recognize the known and anticipated limitations of the 12-lead ECG as a population screening test, including the expected frequency of false-positive and false-negative test results, as well as the cost required to support these initiatives over time *(Class IIb; Level of Evidence C)*.

Will The Matrix be the answer to SCD?

Holy Grail: Prevention of CAD



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Acknowledgements/Mentors

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