Prevention of Sudden Cardiac Death (SCD)

Jeffrey L. Williams, MD, MS, FACC, FHRS
Co-Director, Heart Rhythm Center
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.”

Underlying Arrhythmias of SCD

- VT 62%
- Bradycardia 17%
- Primary VF 8%
- Torsades de Pointes 13%
Sudden Deaths in Young Competitive Athletes

Syncope during peak exertion often indicates pathology.

SCD in Young Athletes

- Structurally Normal (No Diagnosis)
- Hypertrophic Cardiomyopathy
- Coronary Anomaly
- Possible HCM
- Myocarditis
- ARVD
- Channelopathy
- MVP
- LAD Bridge
- Aortic Rupture
- Aortic Stenosis
- Dilated Cardiomyopathy
- WPW
- Other
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.
Hypertrophic CMP

- Deep S waves in V1, V2
- Tall R waves in V5, V6
- T wave inversion in inferior and lateral leads
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
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
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
Here’s the ECG you don’t want.

[ECG Image]

**PEDIATRIC ECG ANALYSIS**

- Normal PR interval: 124 ms
- Normal QRS duration: 78 ms
- Borderline prolonged QT: QTc = 458 msec

Technician: DAWN HASTINGS
Test ind: PROLONG QT

Referred by: BONNEY
Confirmed by: WILLIAM BONNEY M.D.
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
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

\[ QTc = \frac{QT}{\sqrt{RR}} \]
Normal versus Long QT

Corrected QT Interval (QTc, msec)

- Normal Male: 3,000
- Normal Female: 470 msec, 99th percentile in males
- Normal Female: 480 msec, 99th percentile in females

Overlap Zone
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
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
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

A. Type 1 Brugada
- Coved ST segment elevation

B. Type 2 Brugada
- Saddleback shaped

C. Type 3 Brugada
- Saddleback shaped

Taken from ecgwaves.com
Wolff-Parkinson-White

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)

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

---


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?

Total: 9 minutes
Urgency of SCD

Resuscitation Success vs. Time

Chance of success reduced 7-10% every minute

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.

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.

Maggioni AP. Circulation. 1993;87:312-322.

A

No PVBs
1-10 PVBs/h
> 10 PVBs/h

p log-rank 0.002

B

p log-rank 0.0001

Patients without LV Dysfunction (LVEF >35%)

Patients with LV Dysfunction (LVEF ≤ 35%)
EF and SCD Incidence

- 0-30%
- 31-40%
- 41-50%
- >50%

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

Average Follow-Up: 16 months

1 CIBIS-II (1999) 31%
2 MERIT-HF (1999) 54%
3 USCHFT (1996) 54%

No. Pts in Treatment Arm: N = 1327


Average Follow-Up: 12 months

Average Follow-Up: 6.5 months

N = 1990

N = 696
SCD in Heart Failure

• Despite improvements in medical therapy, symptomatic HF still confers a 20-25% risk of premature 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

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%.

- **Amiodarone vs. Placebo**: HR = 1.06, 97.5% CI = 0.86, 1.30, P-Value = 0.529
- **ICD Therapy vs. Placebo**: HR = 0.77, 97.5% CI = 0.62, 0.96, P-Value = 0.007

* Double-blind for drug therapy
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 $\leq 0.35$
  - EF $\leq 0.40$ with NSVT and EPS +
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.

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

Most Common Complications after Pacemaker Implantation
(Mean+/-Standard Deviation Reported in Prior Studies)

- Lead Dislodgement
- Pneumothorax
- Perforation
- Infection
- In-Hospital Death
- Early (<6 week) Mortality
Cost-Effectiveness

- Pacemakers for AVB or SSS over 5 years.

<table>
<thead>
<tr>
<th>Procedure</th>
<th>Cost per Life-Year Saved (U.S. $1,000)</th>
</tr>
</thead>
<tbody>
<tr>
<td>PTCA&lt;sup&gt;1&lt;/sup&gt; (Chronic CAD, severe angina, 2 VD)</td>
<td>$10.2</td>
</tr>
<tr>
<td>CABG&lt;sup&gt;1&lt;/sup&gt; (Chronic CAD, mild angina, 3 VD)</td>
<td>$18.2</td>
</tr>
<tr>
<td>Hypertension&lt;sup&gt;2&lt;/sup&gt; (mild, men, age 40)</td>
<td>$23.2</td>
</tr>
<tr>
<td>ICD&lt;sup&gt;1&lt;/sup&gt; (with EP study)</td>
<td>$25.7</td>
</tr>
<tr>
<td>Captopril&lt;sup&gt;1&lt;/sup&gt; (Post-MI, EF ≤ 40%)</td>
<td>$28.4</td>
</tr>
<tr>
<td>Cardiac Transplant&lt;sup&gt;1&lt;/sup&gt; (CHF)</td>
<td>$44.3</td>
</tr>
<tr>
<td>Peritoneal Dialysis&lt;sup&gt;2&lt;/sup&gt;</td>
<td>$57.3</td>
</tr>
</tbody>
</table>

Expensive  
Borderline Cost Effective  
Cost Effective  
Highly Cost Effective
Can we predict who will experience SCD?

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).
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

Determinants of Health

- 7% Physical Environment
- 11% Medical Care
- 22% Genetics and Biology
- 36% Individual Behavior
- 24% Social Circumstances
- 56% Genetics and Biology

This diagram is a model of all factors correlated with health outcomes for an individual.
Acknowledgements/Mentors

• The following individuals contributed to the content of this presentation:
  – Samir Saba, MD, FHRS, Chief of Cardiology, UPMC.
  – David Schwartzman, MD, FHRS, Professor of Medicine, WVU.
  – William Bonney, M.D. Children’s Hospital of Philadelphia.