Sudden Death - Definition

- Sudden Death
  - Typically refers to Sudden Cardiac Death
  - Two general definitions
    - Death within the first hour after onset of symptoms
    - Death within the first twenty-four hours after onset of symptoms

Learning Objectives

- Identify the common causes of sudden death in children and adolescents
- Translate knowledge about sudden death into preparticipation evaluation of athletes
- Recognize the features that characterize a valid screening program

Disclosures

- My division makes money evaluating youth for heart disease and screening youth for conditions that are associated with increased risk of sudden death.

Another disclosure

1974, Lambert and colleagues, 20 centers, retrospective review, age 1-21 years
- 254 deaths
  - 186 patients without surgery, 68 postoperative
  - 58% at rest or sleeping
  - 32% playing
  - 10% active sports
Sudden Death in Children

- Most common diagnoses
  - Aortic valve stenosis – 17%
  - Eisenmenger syndrome – 16%
  - Cyanotic congenital heart disease with low pulmonary blood flow – 14%
  - Hypertrophic cardiomyopathy – 7%

15th Bethesda Conference

- Sudden Cardiac Death-1985
  - Driscoll, Olmstead County, 515 death certificates over 33 yrs, 3-20 years old
    - 12 sudden death, 1.3/100,000 patient-years, 7 cardiac deaths
  - Garson, pediatric cardiology population, 25 yrs
    - 981 total deaths, 101 sudden death (9%)

15th Bethesda Conference

- Garson, TCH, cont
  - 101 sudden death
    - R-L shunt, postop shunt (12%)
    - Eisenmenger syndrome (11%)
    - Postop tetralogy of Fallot (11%)
    - Unoperated tetralogy of Fallot (7%)
    - Dilated cardiomyopathy (5%)
    - Postop complete AV septal defect (5%)
    - Postop Mustard (TGA) (5%)
    - Long QT syndrome (5%)
    - No one with a normal heart

Garson, TCH, 1985

- 49% NYHA class III, 23% NYHA class IV
- 87% had cardiomegaly on chest X-ray
- 89% had a recent cath: 46% had poor hemodynamics, 43% had pulmonary hypertension
- Routine ECG showed arrhythmia is 57% and correlated with poor hemodynamics
- 22% died during athletics (Mustard, PO Tet, CM, LQTS), 50% awake, 28% asleep
- Arrhythmia therapy introduced in 1978

Hypertrophic Cardiomyopathy

- National Heart and Lung Institute had an HCM (ASH) study group
  - 1976, Maron, 46 children with ASH, 35 with more than one year of follow-up
    - 11 with sudden death (4% per year)
    - Nothing found to be predictive of sudden death
- Incidence: 1:500
- Primary inheritance pattern is AD

Sudden Death in Young Athletes

- Maron, 1980, AFIP registry
- 29 competitive athletes, 13-30 yrs old with sudden death
  - 22 during or immediately after severe exertion
  - 28 had structural cardiac disease
    - 14: hypertrophic cardiomyopathy plus 5: idiopathic concentric LVH
    - 3: anomalous origin of the LCA
    - 3: atherosclerosis
    - 2: ruptured aorta
    - 1: hypoplastic coronary arteries
Transitions
- 1980s-echocardiography became standard
- 1980s-neonatal open heart surgery emerged
- 1980s-interventional catheterization emerged
- 1980s-cardiac transplantation became successful
- 1980s-electrophysiology became increasingly scientific

Nontraumatic Sports Death
- Van Camp, MSSE, 1995, registry data
- High school and college athletes
  - 160 nontraumatic deaths over a 10 year period, 137 have sufficient data for analysis
  - 100 athletes with cardiovascular conditions
  - 30 athletes with noncardiovascular conditions
  - 7 with no identifiable cause of death

Nontraumatic Sports Death
- Cardiovascular causes
  - Hypertrophic cardiomyopathy – 51
    - Probably HCM – 5
  - Coronary artery anomalies – 16 (3 had HCM)
  - Myocarditis – 7
  - Aortic stenosis – 6
  - Dilated cardiomyopathy – 5
  - Atherosclerosis-3, Aortic rupture-3, other cardiomyopathy-2, subaortic stenosis-2, coronary artery aneurysm-1, mitral valve prolapse-1, RV cardiomyopathy-1, WPW-1(also had HCM)

Nontraumatic Sports Death
- Noncardiovascular causes
  - Hyperthermia – 13
  - Sickle cell trait with rhabdomyolysis – 7 (1 also had a coronary anomaly)
  - Status asthmaticus – 4
  - Lightning – 3
  - Arnold-Chiari malformation-1, GI bleed-1, anaphylaxis-1
  - Undetermined cause - 7

Modern data on Sudden Death
- Puranik, Sydney, autopsy data, 1995-2004, ages 5-35 years
  - 427 sudden deaths, 241 cardiac deaths (56%)
    - Primary arrhythmia – 70 (29% of cardiac deaths)
    - Myocardial infarction – 59 (24.5%, 2/3 over age 30 years)
    - Myocarditis – 28 (11.6%)
    - Hypertrophic cardiomyopathy – 14 (5.8%)
    - Dilated cardiomyopathy – 13 (5.6%)
    - Aortic dissection – 12 (5.4%, 4 appeared to have Marfan syndrome)
    - Coronary anomalies – 5
    - Cardiac valve disease - 3

Modern data on sudden death
- Puranik, cont
  - 10.8% of sudden death occurred during exercise
  - Noncardiac deaths: epilepsy-24%, intracerebral hemorrhage-24%, asthma-16%
  - Limitations
    - People known to have disease were unlikely to have a coroner autopsy
    - More homogeneous than the US
Aortic Stenosis and Sudden Death

- Brown, Boston Children’s, 2010
  - Follow-up aortic valve stenosis after balloon aortic valvuloplasty between 1984 and 2008
    - 528 patients, 6344 patient years, median follow-up-14.6 years
    - 63 deaths, 6 sudden deaths, only 1 sudden death after age 18 months.
    - 183 exercise restricted, 220 not exercise restricted. No evidence of beneficial effect of exercise restriction. Likelihood of exercise restriction was related only to identification of the primary cardiologist

Sudden Death

- What I think I know
  - Sudden death in congenital heart disease survivors older than 1 year old has diminished markedly since the first reports-multiple factors
  - Aortic stenosis under the care of a cardiologist has a low risk of sudden death, even with exercise
  - Unrecognized hypertrophic cardiomyopathy is the most common cause of sudden death during competition
  - Anomalies of coronary origin have an uncertain risk of sudden death with exercise

Exercise

- Exercise is the application of a controlled stress on a muscle group in an effort to induce a change in structure and function
  - For most athletes, exercise includes the application of a conditioning stress on cardiac muscle

Exercise stress

- Dynamic or aerobic exercise is made up of repetitive, rhythmic muscle contractions which are performed over an extended period of time during training at submaximal levels
  - Aerobic exercise requires the delivery of large amounts of oxygen to the mitochondria to function as electron donors to produce ATP

Exercise and O2 Consumption

- Resting VO2 = 3.5 ml/kg.min
- Fit Athletes > 60 ml/kg.min
- Requirements to deliver that much O2
  - Increased cardiac capacity
  - Increased vascular conductance
  - Increased total body hemoglobin
  - Increased mitochondrial size and density
  - Increased oxygen extraction

Increased Cardiac Capacity

- Increased ventricular volume
- Increased stroke volume
- Increased LV mass
- Increased wall thickness
  - Normal wall dimensions, up to about 11 mm
  - Normal athlete wall dimensions, up to 13 or 14 mm in some
  - Normal highly elite athletes, up to 19 mm thick, half are greater than 13 mm thick
Screening Athletes for Heart Disease
- Hypertrophic Cardiomyopathy is characterized by increased LV mass
- Physiologic adaptations to high level cardiovascular fitness is characterized by increased LV mass
- Physiologic vs Pathologic
- Symptom evaluation vs Preparticipation screening

Preparticipation Screening
- ECG variations in healthy athletes
  - Sinus bradycardia
    - Sinus pauses > 2 sec in 33%
    - Wandering atrial pacemaker is common
    - Idioventricular rhythm in 20%
  - Atrioventricular conduction delay
    - 10-33% have a prolonged PR interval
    - 10% exhibit Wenckebach periodicity at rest, and 40% have periods of 2nd degree AV block on 24 hour ECG
    - “Incomplete right bundle branch block” – 14%

Preparticipation Screening
- ECG variation in healthy athletes
  - Increased P wave amplitude is common
  - Up to 80% have ECG evidence of LVH by voltage criteria
  - Vertical frontal plane QRS axis is common
  - Various ST and T wave changes are seen commonly, including T wave inversion and QT prolongation

Preparticipation Screening
- Theories behind screening
  - There are cardiovascular diseases that can be identified by screening where training and competition increase the likelihood of disease progression or sudden death
  - Disease detection will allow a decrease in the likelihood of sudden cardiac death

Sudden Death Prevention
- Screening?
  - Screening strategies are undeveloped and not evidence based
  - There is a risk of errors in screening
    - Allow athletes with disease to complete
    - Exclude athletes from sports that have no disease
  - Screening is expensive
    - Estimates range as high as $250,000 to identify one at risk youth
    - Current screening programs depend on volunteerism (loss leaders?)
Sudden Death Prevention

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- In each case, it is likely that the final common pathway is ventricular tachycardia/fibrillation

Sudden Death Prevention

- VT/VF Arrest
  - Factors associated with successful resuscitation
    - Early, effective bystander CPR
    - Early defibrillation

Unanswered Questions

- Magnitude of LVH in HCM predicts sudden death risk – Could some people with HCM compete safely?
- What are the implications of AICD therapy? Will an ICD be protective during exercise? What are the ethical questions?

Unanswered Questions

- If you revascularize an anomalous coronary artery, can you endorse competition?
- In aortic stenosis, can you stratify for safety?
- In postoperative congenital heart disease, what are the real data?