SUDDEN CARDIAC DEATH in the YOUNG

Sudden Cardiac Death in the Young

Incidence
Etiology
Identification
Intervention

Incidence Studies

- 1985 Driscoll et al (Minnesota) 1.3 cases/100,000
- 1986 Maron et al (Minnesota) most common Hypertrophic Cardiomyopathy (HCM)
- 1992 Waller et al (Indiana) 88% were cardiac in origin – retrospective screening would have detected most

Incidence Studies

- 1996 Maron et al reviewed the deaths of 158 trained athletes throughout the United States from 1985 – 1995
  - 134 were cardiac in origin
  - Median age was 17 years
  - 120 (90%) were male athletes
  - 70 (52%) were Caucasian
  - Basketball (47) and Football (45) = 69%

Incidence

- The reported incidence in children and adolescents varies from 0.8 - 6.2 cases/100,000 population depending on the study and methodology.
- The rate seems to be higher than previously thought but still relatively rare compared to the adult incidence of 1/1,000
### Incidence

- Despite the multiple studies the exact number of deaths per years is still not known. Many suffer from inherent problems.
- They often are not comprehensive.
- Since they are all retrospective, they often are not able to capture all significant data.
- Studies often include deaths that were not unexpected or cardiac in origin.

### CAUSES

#### Structural or Functional Abnormalities
- Hypertrophic Cardiomyopathy (HCM)*
- Arhythmogenic right ventricular dysplasia (ARVD)*
- Coronary artery abnormalities
- Primary pulmonary hypertension*
- Myocarditis/dilated cardiomyopathy*
- Marfan’s Syndrome*
- Aortic valve stenosis

#### Primary Electrical Abnormalities
- Long QT Syndromes*
- Brugada Syndrome*
- Wolf-Parkinson-White Syndrome (WPW)
- Primary or Idiopathic Ventricular tachycardia
- Catecholaminergic Polymorphic VT (CPVT)
- Heart Block – congenital or acquired

### CAUSES

#### Acquired
- Commotio cordis
- Drug abuse – cocaine, stimulants, inhalants, etc.
- Secondary Pulmonary Hypertension
- Atherosclerotic heart disease
CAUSES

Post-operative congenital heart defects
Tetralogy of Fallot
Transposition of the great arteries (switch / baffle)
Fontan
Hypoplastic left heart syndrome
Coarctation of the aorta
Cardiac transplantation

Hypertrophic Cardiomyopathy
HCM
Risk Factors
1. History of cardiac arrest or spontaneous VT
2. Family history of premature HCM-related death, particularly if sudden
3. Syncope—especially if exertional or recurrent
4. Multiple and repetitive or prolonged bursts of non-sustained VT on Holter
5. Hypotensive BP response to exercise
6. LV wall thickness >30 mm

ECG in HCM

Over 90% of the patients with HCM have an abnormal ECG pattern including:
- Marked increased precordial R or S voltages
- Deep Q waves
- Deeply inverted T waves
- Abnormal R wave progression in the anterior precordial leads
- Left axis deviation.
Coronary Artery Abnormalities

- LCA from the right sinus of Valsalva
- RCA from the left sinus of Valsalva
- Single coronary artery ostia
- Hypoplastic coronary artery syndrome
- William’s syndrome with coronary ostial stenosis
- Kawasaki Syndrome

Long QT Syndromes

- Romano Ward
- Jervell Lange Nielsen
- Acquired
Anderson Syndrome
Complex phenotype which includes:

a. Hypokalemic periodic paralysis
b. Dysmorphic features – syndactyly, hypertelorism, low set ears, broad forehead, micrognathia, cleft palate, clinodactyly, and scoliosis

c. QTc prolongation and ventricular arrhythmias
d. Inherited as an autosomal dominant trait

Wolff-Parkinson-White

- Pre-excitation – accessory pathway syndrome
- Second most common form of paroxysmal supraventricular tachycardia
- Prevalence is 1.5 – 3.1 / 100,000 population
- Characterized by delta waves
- May be associated with cardiomyopathies

WPW

Delta Wave

Brugada Syndrome

- Autosomal dominant familial syndrome
- Delayed right ventricular conduction
- Elevated ST in leads V1-3
- Associated with ventricular arrhythmias and
- Prevalence in 4-12% of all sudden cardiac deaths

Catecholaminergic Polymorphic Ventricular Tachycardia (CPVT)

- Characterized by salvos of bidirectional and polymorphic ventricular tachycardias
- Often in relation to adrenergic stimulation or physical exercise
- No structural myocardial disease
- Inherited autosomal dominant pattern.
Arrhythmogenic Right Ventricular Dysplasia (ARVD)

- Myocardial disease that typically affects the right ventricle with gradual replacement of myocytes with adipose and fibrous tissue.
- More common in males between 15 and 40 yrs
- Symptoms include syncope and ventricular tachycardias with LBBB pattern progressing into congestive heart failure and cardiac arrest.
- Thought to be familial

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SCD Patient Profile

- Usually a male
- High school or collegiate competitive athlete
- Multiple etiologies depending on age but usually with no known history of cardiovascular disease or symptoms

Competitive Athlete

Definition

One who participates in an organized team or individual sport systematic requiring training and regular competition against others while placing a high premium on athletic excellence and achievement.

Base Population of Competitive Athletes

Considering the fact that there are:

- 4 million High School
- 500,000 collegiate
- 5,000 professional athletes in the United States

The major challenge is to diagnose the condition in a cost effective and accurate manner before death.

Preparticipation Screening

History and Physical Examinations

although the backbone of preparticipation screening, without non-invasive testing is not sufficient to guarantee detection of many critical cardiovascular abnormalities.

ECG in ARVD

Over 50% of the patients with ARVD have:

- Inverted T waves in V 1-3
- Increased QRS duration
- IRBBB
- Ventricular arrhythmias with left axis deviation
The inclusion of a 12 lead ECG (not to mention an ECHO) into the screening program greatly enhances the diagnostic capabilities but their inclusion are still heatedly debated due to the enormous costs and large number of abnormal patterns found in young trained athletes.

ECG changes develop in trained athletes as a consequence of sustained physical exercise. There is a misconception that these changes overlap significantly with ECG abnormalities seen in cardiovascular diseases which cause sudden cardiac death.

Elite and professional athletes have even more marked LV wall and cavity remodeling. These changes raise a diagnostic dilemma between the extreme but innocent cardiac adaption to intensive exercise training and a pathological cardiac condition.

Systematic preparticipation screening including a 12 lead ECG is a medical program established by law in Italy since 1982.

The Veneto study performed ECGs in 32,652 amateur athletes (average age 17). 28,799 (88.2%) were normal, 3,852 (11.8%) were abnormal.

The main abnormal ECG patterns noted in the Italian survey were:
- early repolarization pattern
- incomplete RBBB and prolonged PRi
These are commonly regarded as innocent ECG changes associated with the “athletic heart” and deprived of clinical significance.

Type I
- Sinus Bradycardia
- First degree AV Block
- IRBBB
- Early repolarization
- Isolated voltage criteria for LVH
ECG Changes in the Athlete

Type II
- Left atrial enlargement
- ST-segment depression
- Pathologic Q waves
- Inverted T waves in 2 or more consecutive leads
- RBBB
- LAD/RAD
- Long/short QTc
- Ventricular arrhythmias

Recommendations for Type I and II ECG Changes

In the Italian screening protocol:
- Asymptomatic athletes with Type I ECG changes and a negative family history are allowed eligibility in competitive sports without further evaluation.
- Further diagnostic work up is limited to those with Type II changes and/or positive medical history or physical examination.

American Heart Association 12 Point Recommendations

Personal History
1. Exertional chest pain or 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 before the age of 50 because of heart disease in 1 or more relatives.
7. Disability from heart disease in a close relative younger than 50
8. Specific knowledge of certain cardiac conditions in family members (i.e. Marfan etc.)

AHA 12 Point Preparticipation Screening Recommendations

Physical Examination
9. Heart murmur
10. Femoral pulses to exclude coarctation of the aorta
11. Physical stigmata of Marfan syndrome
12. Brachial artery blood pressure (sitting position) preferably in both arms.

AHA Recommendation

The AHA task force recommends that both a careful/complete personal history and physical examination be performed before participation in organized high school and collegiate sports. Screening should then be repeated every 2 years. In the intervening years an interim history should be obtained.
Caviat

Unfortunately, these guidelines should not promulgate a false sense of security because the standard history and physical examination intrinsically lack the capability to reliably identify many potentially lethal cardiovascular abnormalities.

Madison’s Foundation

- Since Madison’s death, the Foundation has funded placement of more than 300 AEDs in schools and other organizations.
- Provided training for CPR and
- Advice concerning maintenance of the AEDs
- The Foundation will inaugurate a screening program in two smaller schools this spring with plans to pursue a more vigorous program in the fall.

Sequence For Success

**Time to Defibrillation**

- AED provided by school
  - 3.6 minutes
- AED provided by EMS
  - 11.5 minutes

Larson, Ann Emerg Med, 1993

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The rest is up to EVERYONE ELSE