Sports Cardiology

Sports Medicine Fellows Conference

February 23rd (Tuesday), 2010

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[HUMAN PERFORMANCE LAB]
providing excellence in sport science
Why the Concern with CV Risk in Athletes?

- Deaths during sports – social impact and liability issues.
  - The Italian Experience – but reports not for all of Italy
- Recent recognition of channelopathies and diseases of the right ventricle.
- Newer medical technologies – physiological changes or pathological?
- Controversy regarding the use of the simplest technology, the ECG.
Some Facts and Questions Raised

• Young competitive athletes who die suddenly usually have had silent CV diseases, predominantly either cardiomyopathies or congenital coronary anomalies. How about channelopathies?

• Number One = CM - Hypertrophic cardiomyopathy (HCM) in most Countries with records, while Arrhythmogenic Right ventricular Dysplasia (ARVD/C) predominates in the parts of Italy where data is available.

  • About one in 500 people in the United States have HCM – but who is at risk?

  • ARVD/C – why Italy? Is it missed elsewhere? Does Exercise cause or worsen it?

• Commodus Cordis (20% deaths?) can be prevented by chest protectors but susceptibility cannot be recognized by screening (are AEDs effective?)
Screening for Sports Participation

- History of chest pain or syncope--best signs
  - Syncope during as opposed to post-exercise

- Hypertrophic Cardiomyopathy is very difficult to discern from "athlete's heart"
  - Athletic Heart Syndrome includes many abnormalities that are not dangerous
    - Gallop sounds, increased heart size/movements

- Family History - current best genetic test

Bethesda Guidelines; European Guidelines … the ECG controversy
12-Element AHA Recommendations for PPE
CV 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 50 y of age resulting from heart disease in ≥1 relative
7. Disability from heart disease in a close relative <50 y 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)§

Are the athletes being truthful?

Do they know family history?

Is auscultation a lost art?

How helpful are physical findings of Marfans?
Important Questions requiring answers prior to adding the ECG to Athletic Screening

1) what do athletes die from?
2) What is the sensitivity of the ECG (or any test) for recognizing these conditions?
AUTOPSY STUDIES OF SUDDEN DEATH IN ACTIVE YOUNG ADULTS
FOUR STUDIES REVIEWED

structural disease possibly ECG-recognized inclds HCM + ARVC/D + Myocarditis + DCM + clinically significant AS
No structural disease potentially includes channelopathies that are ECG-recognized

(EGC recognizable = 21% structural and 28% non-structural = 49%

(EGC recognizable = 41% structural and ?% non-structural = 41%

(EGC recognizable = 18% structural and 34% non-structural = 52% from mandatory autopsies

Gray Area of Overlap Between “Athlete’s Heart” and Cardiomyopathies

- **Dilated cardiomyopathy**: LV cavity: 56-70 mm
- **Frequent or complex ventricular arrhythmias**
- **Myocarditis**
- **HCM; ARVC**: Distinctly abnormal ECG
- **HCM**: LV wall thickness: 13-15 mm

**Athlete’s Heart**

**Gray-area cardiomyopathy**
Criteria to Distinguish Hypertrophic Cardiomyopathy (HCM) from Athlete's Heart

“Gray Zone” of LV Wall Thickness (13-15 mm)

HCM*

Athlete's Heart

+ Unusual Patterns of LVH†
+ LV Cavity <45 mm
− LV Cavity >55 mm
+ Marked LA Enlargement
+ Bizarre ECG Patterns
+ Abnormal LV Filling
+ Female Gender
− Thickness with Deconditioning
+ Family History of HCM
− Max. VO₂ >45 ml/kg/min. >110% predicted‡
Coronary Artery Anomalies (CAAs)

- Definition, clinical presentation, diagnostic workup (ECHO, CT angio), prognosis, and treatment.

- Ischemic mechanisms of CAAs and the incidence of these anomalies at autopsy and angiography.

- More recent studies have dealt with vexing questions related to pathophysiological mechanisms and clinical prognoses for different forms of CAAs.

- Paolo Angelini’s review (Circulation, 2007:115:1296) best review plus focus on sudden death in young athletes.
Coronary Artery Anomalies (CAAs)

- This subject is undergoing evolutionary changes related to the definition, clinical presentation, diagnostic workup, prognosis, and treatment.

- CAAs were first the subject of anatomic discussions that centered around the description and classification.
- Next, the ischemic mechanisms of CAAs and the incidence of these anomalies in the normal human population were addressed in autopsied patients and coronary angiography populations.

- More recent studies have dealt with vexing questions related to pathophysiological mechanisms and clinical prognoses for different forms of CAAs.

- Paolo Angelini’s review (Circulation, 2007:115:1296) focuses on anomalous origination of a coronary artery from the opposite sinus (ACAOS) with intussusception of the ectopic proximal vessel, which is the subgroup of CAAs that has the most potential for clinical repercussions, specifically sudden death in young athletes.
Conceptual diagram that shows most of the possible paths (1 through 5) by which the RCA, left anterior descending artery (LAD), and circumflex artery (Cx) can potentially connect with the opposite coronary cusps. Paths: 1, Retrocardiac; 2, retroaortic; 3, preaortic, or between the aorta and pulmonary artery; 4, intraseptal (supracristal); 5, prepulmonary (precardiac). AL indicates antero-left; AR, antero-right; P, posterior; M, mitral valve; and T, tricuspid valve.

## Diagnostic Criteria for ARVD/C

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| Structural or functional abnormalities | 1. Severe dilation and reduction of RVEF with mild or no LV involvement  
2. Localized RV aneurysm (akinetik or dyskinetic areas with diastolic bulging)  
3. Severe segmental dilatation of the RV |
| Tissue characterization | Infiltration of RV by fat with presence of surviving strands of cardiomyocytes |
| ECG depolarization/conduction abnormalities | 1. Localized QRS complex duration >110 msec in V₁, V₂, or V₃  
2. Epsilon wave in V₁, V₂, or V₃ |
| ECG repolarization abnormalities | Late potentials in SAECG |
| Arrhythmias | Inverted T waves in right precordial leads (V₂ to V₃ above age 12 y in absence of RBBB) |
| Family History | 1. LBBB VT (sustained or nonsustained) on ECG, Holter or ETT  
2. Frequent PVCs (>1000/24 hrs on Holter) |
| Family history of ARVD confirmed by biopsy or autopsy | 1. Family history of premature sudden death (<35 y) due to suspected ARVD  
2. Family history of clinical diagnosis based on present criteria |

The criteria state that the individual must have two major, or one major plus two minor or four minor criteria from different categories to meet the diagnosis of ARVD/C.
Newer ECG Criteria that Enhance the Value of the ECG for Screening

- **Brugada Syndrome** - recognized in 1992, ECG criteria = ST elevation in V1-2 > 2mm plus shape of ST and T wave
- **Right Ventricular Dysplasia/Cardiomyopathy (ARVD/C)** - 25 years since first described. It appears worldwide with a prevalence of about 1 in 5000 persons; ECG criteria = T wave inversion V2, slurring of S wave V1-3, epsilon waves.
- **Hypertrophic Cardiomyopathy (HC)** - 50 years since first described, it appears with a prevalence of about 1 in 500 persons; HCM is a disease of the sarcomere due to > 450 mutations in >10 genes; ECG criteria = total LVH voltage, septal Q’s, QRS duration
- **T wave Inversion (2mm) in 3 or more leads** - found in numerous risk conditions
- **Drugs/ambulatory monitoring** to bring out channelopathies
Newer ECG Criteria that Enhance the Value of the ECG for Screening

Figure 2. Example of types 1, 2 and 3 Brugada ECG patterns. Varying degrees of right bundle branch block are seen and Type 1 is characterized by coved-type ST elevations, while Types 2 and 3 depict a “saddleback” ST segment. Lead V2 is usually the most pronounced, although the patterns can extend from V1-V3.

<table>
<thead>
<tr>
<th></th>
<th>Type 1</th>
<th>Type 2</th>
<th>Type 3</th>
</tr>
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<tbody>
<tr>
<td>Clinical significance</td>
<td>Diagnostic</td>
<td>Non-diagnostic</td>
<td>Non-diagnostic</td>
</tr>
<tr>
<td>J-wave amplitude</td>
<td>≥ 2 mm</td>
<td>≥ 2mm</td>
<td>≥ 2mm</td>
</tr>
<tr>
<td>T wave</td>
<td>Negative</td>
<td>Positive or biphasic</td>
<td>Positive</td>
</tr>
<tr>
<td>ST-T configuration</td>
<td>Coved-type</td>
<td>Saddleback</td>
<td>Saddleback</td>
</tr>
<tr>
<td>ST-segment</td>
<td>Gradually descending</td>
<td>Elevated ≥ 1 mm</td>
<td>Elevated &lt; 1 mm</td>
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Newer ECG Criteria that Enhance the Value of the ECG for Screening
Newer ECG Criteria that Enhance the Value of the ECG for Screening

Right Ventricular Dysplasia/Cardiomyopathy (ARVD/C)
T wave Inversion greater than 2 mm in 3 leads

Pelliccia, A, et al. Outcomes in Athletes with Marked ECG Repolarization Abnormalities. NEJM 2008:358:152-161. Positive predictive value of 36% for this ECG abnormality that occurs in 1% of athletes (immediate diagnosis in 39 and 5 in follow up [out of 129], mostly cardiomyopathies … 5 out of 90 w/o structural HD had event in FU).
T wave Inversion greater than 2 mm in 3 leads other than V1 and AVR in 33 yo 6ft 205 lb FB
EU Criteria for an Abnormal ECG

P wave
- Left atrial enlargement: negative portion of the P wave in lead V₁ ≥0.1 mV in depth and ≥0.04 s in duration
- Right atrial enlargement: peaked P wave in leads II and III or V₁ ≥0.25 mV in amplitude

QRS complex
- Frontal plane axis deviation: right ≥120° or left −30° to −90°
- Increased voltage: amplitude of R or S wave in a standard lead ≥2 mV, S wave in lead V₁ or V₂ ≥3 mV, or R wave in lead V₅ or V₆ ≥3 mV
- Abnormal Q waves ≥0.04 s in duration or ≥25% of the height of the ensuing R wave or QS pattern in ≥2 leads
- Right or left bundle-branch block with QRS duration ≥0.12 s
- R or R' wave in lead V₁ ≥0.5 mV in amplitude and R/S ratio ≥1

ST segment, T waves, and QT interval
- ST-segment depression or T-wave flattening or inversion in ≥2 leads
- Prolongation of heart rate–corrected QT interval >0.44 s in males and >0.46 s in females

Rhythm and conduction abnormalities
- Premature ventricular beats or more severe ventricular arrhythmias
- Supraventricular tachycardias, atrial flutter, or atrial fibrillation
- Short PR interval (<0.12 s) with or without delta wave
- Sinus bradycardia with resting heart rate ≤40 bpm
- First- (PR ≥0.21 s†), second-, or third-degree atrioventricular block
Title: Recommendations for Interpretation of 12-Lead Electrocardiogram in the Athlete

Perspective: The following are 10 points to remember from this position paper formulated by the European Society of Cardiology:

1. Electrocardiographic findings that are common and training-related and that do not require additional evaluation are sinus bradycardia, 1° atrioventricular block (AVB), incomplete right bundle branch block (BBB), early repolarization, and isolated voltage criteria for left ventricular hypertrophy (LVH).

2. Uncommon and training unrelated electrocardiographic findings that mandate further evaluation include T-wave inversion, ST-segment depression, pathological Q waves, atrial enlargement, a hemiblock, right ventricular hypertrophy, a BBB, or a Brugada-pattern of ST-segment elevation.

3. Training-related electrocardiographic findings are more common in men than women, athletes of African descent, and high-endurance athletes such as cyclists.

4. Sinus rates <30 bpm and sinus pauses >2 seconds are common in highly trained athletes, particularly during sleep.

5. A normal chronotropic response to exertion and the absence of bradycardia-related symptoms distinguishes training-related sinus bradycardia from sinus node dysfunction.

6. 1° AVB and Mobitz I 2° AVB are common, but Mobitz II 2° AVB or 3° AVB should not be assumed to be training-related and require evaluation.

7. Early repolarization in Caucasian athletes most commonly consists of upwardly concave ST-segments and tall and peaked T waves; in black athletes, there often is convex ST-segment elevation and negative T waves, mimicking a Brugada pattern.

8. In the presence of voltage criteria for LVH, pathological hypertrophy should be suspected if there is left atrial enlargement, left-axis deviation, repolarization abnormalities, or pathological Q waves.

9. T-wave inversion ≥2 mm in ≥2 adjacent leads should prompt evaluation for structural heart disease.

10. Electrophysiological testing for risk stratification with possible catheter ablation is appropriate in athletes with ventricular pre-excitation. Fred Morady, M.D., F.A.C.C.
Key points (1):

- Exercise/Sport Related Deaths have wide social impact
- They are rare in youth but increase with age
- The public expectation of modern medicine is that they should and could be prevented
- Those that exercise have CV alterations that can be mistaken for disease
Key points (2):

- Individuals feel that they have the right to compete or to exercise while organizations/MDs have to protect themselves.
- Bayesian statistics demonstrates that rare diseases are not easily diagnosed and testing creates many false positives.
- Controversy exists as to the cost/benefit of screening.
- The recommended approach to the PPE differs between Europe and the US.
Solutions to the Athlete Screening Dilemma

1 - Standardization of the American PPE (web based and using the AHA 12-points)
2 - Optimization of the ECG criteria for further evaluation
3 - ECG screening device to allow non-medical personnel perform the first level of ECG screening (green – good, red – see your Doctor to have a 12 lead and possibly further evaluation)
The End – have a great day!!

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Historical High Points

- **1960-70's** - Early reports of unusual ECG findings in Athletes (USAFSAM, Norway, others)
- **1970** - Italian Law mandating screening of Athletes for safety ... put into action 1985 with ECG
- **1980** - 3 Univ Maryland BB deaths, NIH registry
- **1985** - Minnesota State High School League (MSHSL) mandatory insurance plan covering catastrophic injury or death to all student athletes
- **2005** – European Proposal to mandate ECG screening
- **2008** – Serial decrease in CV athletic deaths in Italy after ECG screening enacted
- **2008** – AHA 12 points for screening ... without ECG
- **2010** – IOC adds the ECG
“all citizens participating in competitive sport activities must have preventive periodical examination with the aim to evaluate them for athletic practice”*

* Italian Law # 1099-1971; inacted 1982
Medical Protection of Athletic Activity
Italian Biannual Cardiovascular “Screening” in Young Competitive Athletes

BASIC PROTOCOL:

- History
- Physical Exam
- Rest ECG (12-Leads) – mandated by whom and when?
Minnesota State High School League (MSHSL)

- This is a voluntary, nonprofit association of schools (independent of the Board of Education) that is responsible for a variety of administrative functions related to student athletes within the 440 public and private high schools of Minnesota.
- They have mandated an insurance plan covering catastrophic injury or death for all student athletes engaged in interscholastic sports programs at the varsity and junior varsity levels within the state.
- The records of this indemnity program permit an accurate assessment of the number of participants in high school sports, as well as the number of deaths during this period of time.
- The records for the 12-year period, 1985/1986 to 1996/1997 inclusive, and for grades 10 to 12 have been reported by Maron et al.
Italy – American Differences relating to Athletic Screening

Law to Screen athletes in Italy (community vs individual concerns)

Physician/population ratio in Italy greater than US

Italian Sports Specialists trained re CV eval

Causes of Death (ARVD/C-Italy, HCM-US)
Roughly 10 million young competitive athletes each year in the US

Over 200 young athletes die every Year in the US (0.002% prevalence)
If a test with a 50% sensitivity/90% specificity (i.e., ECG?) used, there will be 1 million false positives requiring further testing to possibly save 100 lives (only half identified)
To consider a screening method, we need to know that the outcome can be prevented if recognized and the characteristics of the test
To test a screening method, need population randomized to screening and usual care and then assess outcomes (are lives saved?)
One Point of View regarding ECG Screening

Our Italian Colleagues deserve congratulations for presenting the Italian experience and adding to the understanding of Athletic CV screening.

They have highlighted the problems of dealing with poorly conceived legalities.

Imitating the Italian approach would divert limited health care resources from other issues with wider societal impact.
Use of the 12 lead ECG to Screen for Markers of Sudden Death in Young Athletes
Where should ECG screening be mandated?

- 26,000 High Schools
- 4,100 Colleges
- Several Hundred Professional Athletic Teams
The ECG is Mandated initially and then every 3 to 4 years as part of the PPE for all competitive athletes in the European Union but the American Guidelines are concerned with the following Obstacles
Obstacles to implementing obligatory government-sponsored national screening with ECGs:

1. The large population of athletes to screen
2. The major cost-benefit considerations
3. The recognition that it is impossible to absolutely eliminate the risks associated with competitive sports.
Gray Area of Overlap Between “Athlete’s Heart” and Cardiomyopathies

- **Dilated cardiomyopathy**
  - LV cavity: 56-70 mm

- **Frequent or complex ventricular arrhythmias**

- **Myocarditis**

- **HCM; ARVC**
  - Distinctly abnormal ECG

- **HCM**
  - LV wall thickness: 13-15 mm
Criteria to Distinguish Hypertrophic Cardiomyopathy (HCM) from Athlete's Heart

“Gray Zone” of LV Wall Thickness (13-15 mm)

- Unusual Patterns of LVH
- LV Cavity <45 mm
- LV Cavity >55 mm
- Marked LA Enlargement
- Bizarre ECG Patterns
- Abnormal LV Filling
- Female Gender
- Thickness with Deconditioning
- Family History of HCM
- Max. VO₂ >45 ml/kg/min. >110% predicted

* HCM

† LVH

‡ VO₂
LV cavity 56-70 mm: 1-8% of female athletes

LV thickness 13-15 mm: 1.7-2.5% of male athletes

Dilated Cardiomyopathy

Physiological Training Adaptation

Hypertrophic Cardiomyopathy
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Interest in deaths during sports has accelerated because of their social impact and liability issues.

The assumption that associated cardiovascular (CV) diseases should be identifiable using modern medical technologies. However, Athletic Training and Cardiac Conditions both cause changes on the ECG and imaging modalities. Controversy has arisen regarding the use of the simplest technology, the ECG.

Sudden death is rare during sport in youth: In high school, it affects roughly one in every 300,000 females and one in every 100,000 males. However, frequency of SCD increases with age particularly at “senior” status (about 40) with atherosclerosis manifests.

Sport is not a cause of this increased mortality; rather, it acts as a trigger for cardiac arrest in the presence of underlying CV diseases that predispose the young athlete to CV collapse during physical exercise.
Why the Concern with CV Risk in Athletes (2)?

- Young competitive athletes who die suddenly usually have had silent CV diseases, predominantly either cardiomyopathies or congenital coronary anomalies.

- Commodus Cordis can be prevented by chest protectors but susceptibility cannot be recognized by screening.

- In the youngest athletes, the frequency of sudden death on the athletic field tends to be dominated by hypertrophic cardiomyopathy. About one in 500 people in the United States have this condition and for some of them exercise is associated with an increased risk of life-threatening cardiac arrhythmias.

- Another common mechanism of SCD early in life is anomalous coronary arteries causing inadequate blood flow to the heart muscle during exercise.