Cardiac Screening for Sports Participation: What’s “Good Enough?"

Bill Drake, MD MS
Friday, April 22, 2016
Kansas AAP Meeting
Kansas City Pediatric Cardiology Associates

Disclosure

• On the Athletic Testing Solutions Medical Advisory Board. Paid to read/interpret pediatric ECGs and Echocardiograms for Athletic Testing Solutions.
• I do not intend to discuss an unapproved/investigative use of a commercial product/device in my presentation.
Objectives of the Pre-participation Sports Examination (PPE)

- **Primary**
  - Screen for conditions that may be life threatening or disabling
  - Screen for conditions that may predispose to injury or illness

- **Secondary**
  - Determine general health
  - Serve as an entry point into the health care system for adolescents
  - Provide an opportunity to initiate discussion on health related topics

Causes of sudden cardiac death in young athletes


How often do these occur in the population?

<table>
<thead>
<tr>
<th>Defect</th>
<th>Prevalence</th>
<th>Frequency</th>
</tr>
</thead>
<tbody>
<tr>
<td>Hypertrophic Cardiomyopathy</td>
<td>0.2%</td>
<td>1:500</td>
</tr>
<tr>
<td>Wolf-Parkinson-White</td>
<td>0.1-0.3%</td>
<td>1:1,000 to 1:333</td>
</tr>
<tr>
<td>Long QT Syndrome</td>
<td>0.05%</td>
<td>1:2,000</td>
</tr>
<tr>
<td>Marfan Syndrome</td>
<td>0.02-0.03%</td>
<td>1:5,000 to 1:3,000</td>
</tr>
<tr>
<td>Potential Lethal Coronary Abnl</td>
<td>0.0003%</td>
<td>1:333,333</td>
</tr>
<tr>
<td>Total Prevalence</td>
<td>0.37-0.58%</td>
<td>1:270 to 1:172</td>
</tr>
</tbody>
</table>
How can an Athlete with CV Disease be recognized?

- comprehensive evaluation by a primary care physician
- systematic screening of families with known genetic diseases
- Incidental/fortuitous finding on exam or imaging
- systematic screening of large populations
- symptoms associated or unassociated with sports

Current US Screening

- AAP recommends Personal/Family History and Physical Examination. Use the AHA 14 point evaluation.
- No ECG, per AHA/AAP consensus*
- Kansas (KSHSAA) is one of 35 states that mandate annual screening for athletes to participate in sanctioned sports.

KSHSAA requirements

- Rule 7, Sec. 1, Article 1 (for grades 7-12). No student is eligible to represent any school in interschool athletics, spirit or spirit competition groups until there is on file with the superintendent or principal, a statement certifying that the student has passed an adequate physical examination and is physically fit to participate in interschool athletics, spirit or spirit competition groups. The statement must be signed by any of the following licensed health care providers: (i) a physician; (ii) a chiropractor; (iii) a physician assistant (PA) who has been authorized to perform this examination by a Kansas licensed supervising physician; or (iv) an advance practice registered nurse (APRN) who has been authorized to perform this examination by a Kansas licensed supervising physician.

- Article 5. The Physical Examination shall not be taken earlier than May 1 preceding the school year for which it is applicable. (!)

- Article 6. No middle/junior or senior high school student is eligible to represent his/her school in an interscholastic athletic event until a Physical Examination Form is completed and filed with the school prior to the first try-out or practice in which the student participates.
AHA 14 Element Recommendations for Pre-participation Screening of Competitive Athletes


KSHSAA PPE form, pp. 1, 2

Heart Health Questions

Green numbers correspond to the AHA list

- Has a doctor ever told you that you have any heart problems?________
- Do you get fatigue or become short of breath more than expected during exercise?
- Does anyone in your family have Marfan, Turner, or Noonan Syndrome, or other syndromes that may affect heart function?
- Does anyone in your family have other heart problems?
<table>
<thead>
<tr>
<th>Medical</th>
<th>Significance</th>
</tr>
</thead>
<tbody>
<tr>
<td>Appearance</td>
<td><em>Mural ecchymoses</em>, high-amplitude pulsatility, <em>paradoxic murmur</em>, <em>mitral insufficiency</em></td>
</tr>
<tr>
<td>Respiratory/Exertion</td>
<td><em>Dyspnea</em>, <em>heart rate</em></td>
</tr>
<tr>
<td>Heart</td>
<td><em>Murmurs (standing or supine, 2°/3°)</em></td>
</tr>
<tr>
<td>Palpation</td>
<td><em>Location of point of maximal impulse (PMI)</em>, <em>polypsis</em></td>
</tr>
<tr>
<td>Abdomen</td>
<td><em>Simultaneous femoral and radial pulses</em></td>
</tr>
</tbody>
</table>

**Green numbers correspond to the AHA list**

**KSHSAA PPE form, physical**

### Auscultatory Finding

- Hard, loud, basally (grade 5/6), *diastolic ejection murmur*
- Increased right upper external border
- Decreased with maneuvers that decrease venous return (i.e., squatting, or moving from squatting to standing)

### Significance

- HCM-associated LV outflow tract obstruction
- Acute valvular regurgitation
- Mitral valve regurgitation, possible dilated cardiomyopathy or HCM
- Acute valvular insufficiency; possible Marfan syndrome or bicuspid aortic valve
- Pulmonary valve insufficiency from primary pulmonary hypertension (Graham-Steel murmur)
- Physiologic (hypertensive) "flow" murmur in a well-trained athlete
AHA 14 Element Recommendations for Preparticipation Screening of Competitive Athletes

Medical history
- Personal History
  1. Headache Related to exertion
  2. Unexplained syncope or syncope
  3. Excessive and unexplained dyspnea or palpitations associated with exertion
  4. Prior hospitalization or treatment for a heart murmur
  5. Prior resection of a heart murmur
  6. Prior valvular function or replacement
  7. Prior therapy for the heart, ordered by a physician

Family history
- Premature death (sudden or unexpected, or otherwise before 50 years of age attributable to heart disease in <1 relatives)
- Diabetes mellitus or heart disease in <1 relatives <40 years of age
- Hypertrophic or dilated cardiomyopathy, HCM, or other familial diseases, Marfan syndrome, or cystic significant or tetralogy of Fallot, specific knowledge of genetic cardiac conditions in family members

Physical examination
- 11. Heart murmur
- 12. Faint pulses to exclude aortic coarctation
- 13. Physical examination of aortic aortic regurgitation
- 14. Displaced ventricular pressure (dining posterior)

KSHSAA PPE form, pp. 1, 2

Question #6
- Here it is!
Causes of sudden cardiac death in young athletes


How many times will I detect these with a thorough history and physical exam?

9% + 2% = 11%

The best History and PE alone...

- Will miss at least 2/3 of the potentially lethal abnormalities.
- ECG will help...
  - LVH on the HCM kids (60-90% have LVH)
  - Long QT Syndrome
  - WPW
  - ARVC, myocarditis

What about ECG? Echo?

- HCM
  - ECG abnormal in 60-90%
  - Echo should be abnormal in 100%
  - May have a normal PE and history
  - 25% have a family history
- WPW
  - ECG abnormal in ~100%
- Long QT Syndrome
  - ECG abnormal in 100%
- Coronary artery abnormalities
  - Resting ECG usually normal
  - Echo may be abnormal, is often at best non-diagnostic

Other causes:

- Ion channelopathies
  - 4%
- Aortic Rupture
  - 2%
- DCM
  - 2%
- AS
  - 2%
- Myocarditis
  - 6%
- ARVC
  - 4%
- MVP
  - 3%
- Tunneled LAD
  - 3%
- CAD
  - 3%
- Aortic Rupture
  - 3%
- Ion channelopathies
  - 4%
- WPW
  - 2%
- Other
  - 5%

Causes of sudden cardiac death in young athletes
Current AAP PPE Exam

- Based on History and Physical
- AAP’s stance on routine screening tests:

  **Routine Screening Tests**
  
  Routine laboratory, cardiac, and pulmonary screening tests for PPE remains controversial. However, because information from exams can be useful in making the PPE risk group decisions, no routine screening tests are required during the PPE for clearance of asymptomatic athletes.

  This keynote brings on the difference between screening exams and case finding diagnostic tests. The value of screening tests or procedures depend on 2 variables: (1) the predictive value of the test and the test's ability to determine the likelihood of the condition being present, and (2) the ability to reduce morbidity and mortality by identifying conditions with the screening method. The screening outcome also is acceptable in terms of one's and potential side effects. The value of a case finding diagnostic test, on the other hand, is the ability to diagnose a condition for which suspicion exists and the condition is treatable. Moreover, there is often a medico-legal requirement for a diagnostic test to identify specific conditions.

  When evaluating screening tests with the above-noted criteria, analysis has not supported the use of such tests in asymptomatic, otherwise healthy young people 12 to 25 years of age for cardiovascular abnormalities in athletes (Class III; Level of Evidence C).


Mandatory ECG Screening

- Mandatory and universal mass screening with 12-lead ECGs in large general populations of young healthy people 12 to 25 years of age (including on a national basis in the United States) to identify genetic/congenital and other cardiovascular abnormalities is **not recommended** for athletes and nonathletes alike (Class III, no evidence of benefit; Level of Evidence C).

  Maron, et al., Circulation. 2014;130:1303-1334.

Role for ECG/Echo in smaller settings

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 (eg, 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 ECG/Echo 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).

  Maron, et al., Circulation. 2014;130:1303-1334.
Focused screening

- “...relatively small cohorts of young healthy people 12 to 25 years of age, not necessarily limited to athletes (eg, in high schools, colleges, universities, or local communities), provided that close physician involvement and sufficient quality control can be achieved.”
- Cost of screening borne by patient/family

In KC region, Cardiac Preparticipation Screening offered by:
- St. Luke’s Hospital
- Athletic Testing Solutions

Incremental benefit of ECG

- 510 collegiate athletes received cardiovascular screening before athletic participation.
- Each participant had routine history and examination, ECG, and TTE to detect or exclude cardiac findings.
- This performance of screening with history and examination only was compared with that of screening that integrated history, examination, and ECG.
- Hx, PE
  - Sensitivity 65% (95% CI 17-72%)
  - Specificity 94% (95% CI 92-96%)
- Hx, PE, ECG
  - Sensitivity 91% (95% CI 59-99.8%)
  - Specificity 83% (95% CI 79-86%)

Adding ECG to medical history and physical examination improved overall sensitivity of preparticipation cardiovascular screening in athletes. However, this strategy is associated with an increased rate of false-positive results when current ECG interpretation criteria are used.

Authors used transthoracic echocardiogram as the “Gold Standard.”


Incremental Benefit of ECG + Echo

- In essence, this study includes the “Gold Standard” as part of the screen.
- 964 athletes, ages 18-21 years, 52% female, 19% black.

Found 9 Athletes with significant abnormalities* having the potential to exclude from sports

<table>
<thead>
<tr>
<th>Athlete</th>
<th>History/Symptoms</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>Normal age 11 yrs</td>
</tr>
<tr>
<td>2</td>
<td>Asymptomatic</td>
</tr>
<tr>
<td>3</td>
<td>Asymptomatic</td>
</tr>
<tr>
<td>4</td>
<td>Asymptomatic</td>
</tr>
<tr>
<td>5</td>
<td>Asymptomatic</td>
</tr>
<tr>
<td>6</td>
<td>Asymptomatic</td>
</tr>
<tr>
<td>7</td>
<td>Asymptomatic</td>
</tr>
<tr>
<td>8</td>
<td>Asymptomatic</td>
</tr>
<tr>
<td>9</td>
<td>Asymptomatic</td>
</tr>
</tbody>
</table>

Study Conclusions

- Structural abnormalities leading to disqualification uncommon in both studies
- Increased voltage (even >34 mm) in the absence of repolarization abnormalities are common... and are unlikely to represent structural heart disease.
- Magalinski notes: “The incremental value of echocardiography in our series was negligible in that all the players with abnormalities were identified either by history and physical examination or by abnormal ECGs. Nevertheless, it was helpful in establishing the absence of structural heart disease in the setting of ECG abnormalities.”
- KU study, 0.2% of athletes were ultimately disqualified from competition.
- These cohorts are young, college age athletes and thus may be more “self selected” to be healthy.

Is this cost effective?

- Hx, PE and ECG. A Stanford Study* showed that adding ECG to cardiovascular-focused history and physical examination would increase sensitivity but does add additional costs to testing. The addition of ECG to preparticipation screening saves 2.06 life-years per 1,000 athletes at an incremental total cost of $89 per athlete and yields a cost-effectiveness ratio of $42,900 per life year saved (95% CI, $21,200 to $71,300 per life-year saved) compared with cardiovascular-focused history and physical examination alone.
- Compared with no screening, ECG plus cardiovascular-focused history and physical examination saves 2.6 life-years per 1,000 athletes screened and costs $192 per athlete, yielding a cost-effectiveness ratio of $76,100 per life-year saved ($62,400 to $130,000).

ATS Screening – unpublished data

- Total of 3,690 screened since 2014
- 62.9% male
- Mean age 14.8 +/- 6.2 years
- N=20, 0.54% have an exclusionary abnormality
  - WPW N=8, 0.22%
  - HCM N=4, 0.11%
  - Long QT Syndrome N=3, 0.08%
  - Cardiomyopathy N=3, 0.08%
  - Marfan Syndrome N=2, 0.05%
- N=305, or 8.3% have a non-exclusionary abnormality

How often do these occur in the population?

<table>
<thead>
<tr>
<th>Defect</th>
<th>Prevalence</th>
<th>Frequency</th>
</tr>
</thead>
<tbody>
<tr>
<td>Hypertrophic Cardiomyopathy</td>
<td>0.2%</td>
<td>1:500</td>
</tr>
<tr>
<td>Wolff-Parkinson-White</td>
<td>0.1-0.3%</td>
<td>1:1,000 to 1:333</td>
</tr>
<tr>
<td>Long QT Syndrome</td>
<td>0.05%</td>
<td>1:2,000</td>
</tr>
<tr>
<td>Marfan Syndrome</td>
<td>0.02-0.03%</td>
<td>1:5,000 to 1:3,000</td>
</tr>
<tr>
<td>Potential Lethal Coronary Arbl</td>
<td>0.0003%</td>
<td>1:333,333</td>
</tr>
<tr>
<td>Total Prevalence</td>
<td>0.37-0.58%</td>
<td>1:270 to 1:172</td>
</tr>
</tbody>
</table>

Problems with adding ECG

- ECG is not a proven diagnostic tool in the setting of young, healthy populations.
- No clear outcome benefit to using ECG with respect to mortality in US. Outcome data on athlete screening and mortality have been driven primarily by only 1 database, from the Veneto region of Italy.
- These events are relatively uncommon, albeit exceedingly tragic in each case. 1:200,000 to 1:80,000 deaths per participant per year.
What’s the take away?

- AAP currently endorses History and Physical as the only mandated part of the sports pre-participation evaluation.

- Adding an ECG may not be too far off in the future. The European Society of Cardiology (ESC) endorses adding ECG in the mandated sports PPE.

- Adding ECG may be cost effective, but more studies are needed to show that it truly reduces mortality.

- Current KSHSAA forms comply with the AHA 14 point screening recommendations.

What’s the take away?

- More in-depth screening with ECG +/- echo of smaller, more focused cohorts is not inappropriate.

- Families’ concern about their childrens’ cardiac status with sports participation has driven increased availability of third party cardiac sports screening.