Cardiac disease at risk in the young athlete

(Pathologies cardiaques à risque chez le jeune sportif)

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Déclaration de Relations Professionnelles

Disclosure Statement of Financial Interest

I currently have, or have had over the last two years, an affiliation or financial interests or interests of any order with a company or I receive compensation or fees or research grants with a commercial company:

<table>
<thead>
<tr>
<th>Affiliation/Financial Relationship</th>
<th>Company</th>
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<tbody>
<tr>
<td>• Grant/Research Support</td>
<td>None</td>
</tr>
<tr>
<td>• Consulting Fees/Honoraria</td>
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<tr>
<td>• Major Stock Shareholder/Equity</td>
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<td>• Other Financial Benefit</td>
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</table>

Société Française de Cardiologie
The heart of the younger than 30 years athlete: myths and realities?

A. Puerta 1984-2007

T Simpson 1937-1967

MV Foe 1975-2003

P Sauer 1977-2012

M Candau 1993-2009

F Latu 1980-2009
• **In France (2012)**

• 17.5 M
• 37% women
• 10 M competitive

A majority of young athletes
Figure 2. Distribution by age of sports-related sudden deaths (SDs) in the overall population (blue) and among young competitive athletes (red).

Marijon et al. Circulation 2011;124:672-81
2 opposite facets: The paradox of exercise

• Benefits

• Increased risk of Sudden Death?

• 1000-1500 SD /year in France

<table>
<thead>
<tr>
<th>Study population</th>
<th>Ref.</th>
<th>Study design and reporting system</th>
<th>Incidence (person-years)</th>
</tr>
</thead>
<tbody>
<tr>
<td>US Military (age 18–35)</td>
<td>Eckart et al.</td>
<td>Retrospective, mandatory</td>
<td>1:9000</td>
</tr>
<tr>
<td>Italian Athletes (age 12–35)</td>
<td>Corrado et al.</td>
<td>Prospective, mandatory</td>
<td>1:25,000</td>
</tr>
<tr>
<td>US Adolescents (age 12–19)</td>
<td>Atkins et al.</td>
<td>Prospective</td>
<td>1:27,000</td>
</tr>
<tr>
<td>US Children (age 10–14)</td>
<td>Chugh et al.</td>
<td>Prospective, EMS/Hospitals</td>
<td>1:58,000</td>
</tr>
<tr>
<td>US Athletes (age 12–35)</td>
<td>Maron et al.</td>
<td>Retrospective, public media reports</td>
<td>1:160,000</td>
</tr>
</tbody>
</table>

Maron BJ. NEJM 2000; 1409-11
SD:  - up to 80% during exercise
- Men + strenuous exercise
- survival: 15 to 30%
2 different populations

< 35 Y
0.5 to 2.5 / 100 000

> 35 Y
1 to 4 / 100 000

Ref. Population Prevalence (%)

Fuller et al. 5617 high school athletes (USA) 0.4
Corrado et al. 42,386 athletes age 12–35 (Italy) 0.2
Wilson et al. 2720 athletes and children age 10–17 (UK) 0.3
Bessem et al. 428 athletes age 12–35 (Netherlands) 0.7
Baggish et al. 510 collegiate athletes (USA) 0.6

Cardiac disease at risk in the young athlete

- **Structural cardiac disease**
  - **Inherited**
    - HCM: 10 to 40%
    - Coronary abnormalities: 10 to 30%
    - ARVC: 4 to 25%
    - Congenital heart diseases <5%
    - Valvular disease (MVP) <5%
    - Marfan <5%
  - **Acquired**
    - Active myocarditis: 10 to 30%
    - Premature CAD: 5 to 35%

- **Non structural cardiac disease**
  - Unexplained: 5 to 40%
  - Chanelopathies: QT abnormalities, catecholaminergic polymorphic VT, Brugada Sd
  - WPW
  - Early repolarisation?
Causes of sudden death in young competitive athletes

Maron et al. Circulation 2006;114:1633-44

Marijon et al. Circulation 2011;124:672-81
# SCD related to exercise

<table>
<thead>
<tr>
<th>Study</th>
<th>Location</th>
<th>Total SCD</th>
<th>Age Range</th>
<th>Causes</th>
</tr>
</thead>
<tbody>
<tr>
<td>Eckart</td>
<td>US (military)</td>
<td>108</td>
<td>18-35y</td>
<td>Unexplained (35%), coronary abn.(30%), Myocarditis (10%), HCM (6%)</td>
</tr>
<tr>
<td>Sheppard M</td>
<td>UK</td>
<td>118</td>
<td>&lt;35y</td>
<td>HCM / LVH (41%), ARVC (13%)</td>
</tr>
<tr>
<td>Tabib A</td>
<td>France</td>
<td>27</td>
<td>&lt;30y</td>
<td>HCM (29%), (ARVC 26%)</td>
</tr>
<tr>
<td>Corrado</td>
<td>Italy</td>
<td>49</td>
<td>11-35y</td>
<td>ARVC (23%) CAD 18% coronary abn (12%)</td>
</tr>
<tr>
<td>Soldberg</td>
<td>Norway</td>
<td>23</td>
<td>17-34y</td>
<td>M infarction (40%), Myocarditis (20%),</td>
</tr>
<tr>
<td>Holst</td>
<td>Denmark</td>
<td>15</td>
<td>12-35y</td>
<td>ARVC (25%), unexplained (25%), CAD (13%)</td>
</tr>
<tr>
<td>Suárez-Mier</td>
<td>Spain</td>
<td>61</td>
<td>11-65y</td>
<td>CAD (45%), unexplained (16%), ARVC (16%), HCM(10%)</td>
</tr>
<tr>
<td>Raschka C</td>
<td>Germany, Austria</td>
<td>47</td>
<td>&lt;35y</td>
<td>CAD (36%), Myocarditis (30%)</td>
</tr>
</tbody>
</table>
Ethnic differences

What about young women athletes?

- **Sex Ratio of SCD during exercise:**
  - 1/5 to 1/30!
  - Less premature CAD

- **Normal heart (arrhythmic SCD...) > 50%**
  - Afro-American > White recruits (RR 10.2).

- **Higher Survival at hospital admission (46.5 vs 30%)**

---

Eckart RE. Am J Cardiol 2006; 97:1756-8
Pre participation screening

- Physical examination
  - Cardiac murmur
  - Pulses
  - Marfan?
  - HR and Blood pressure

- ECG (French (2009) – European guidelines)
  - > 12 Y: every 3 years
  - >20 Y: every 5 years (>>>35Y)

- Echocardiography: elite athletes

Avez-vous déjà un dossier médical dans une autre structure, si oui laquelle :

Avez-vous eu connaissance dans votre famille des évènements suivants :
- Accident ou maladie cardiaque ou vasculaire survenue avant l’âge de 50 ans
  Oui Non
- Mort subite survenue avant 50 ans (y compris mort subite du nourrisson)
  Oui Non

Avez-vous déjà ressenti pendant ou après un effort les symptômes suivants :
- Malaise ou perte de connaissance
  Oui Non
- Douleur thoracique
  Oui Non
- Palpitations (cœur irrégulier)
  Oui Non
- Fatigue ou essoufflement inhabituel
  Oui Non

Avez-vous
- Une maladie cardiaque
  Oui Non
- Une maladie des vaisseaux
  Oui Non
- Été opéré du coeur ou des vaisseaux
  Oui Non
- Un souffle cardiaque ou un trouble du rythme connu
  Oui Non
- Une hypertension artérielle
  Oui Non
- Un diabète
  Oui Non
- Un cholestérol élevé
  Oui Non

Fumez-vous
- Depuis combien de temps ?

- Suivi un traitement régulier ces deux dernières années
  (médicaments, compléments alimentaires ou autre)
  Oui Non
- Eu une infection sérieuse dans le mois précédent
  Oui Non
- Avez-vous déjà eu :
  - un électrocardiogramme
    Oui Non
  - un échocardiogramme
    Oui Non
  - une épreuve d’effort maximale
    Oui Non

Carre F, Brion R, Douard H et al. SFC guidelines 2009
Pre participation screening

• ...In young athletes
  – Asymptomatic
  – Normal physical examination
  – Do not limit athletic performance
  – > 50% do not mention symptoms (intense activity – female athletes)

—>>>> What about ECG?

<table>
<thead>
<tr>
<th>Cardiac disease</th>
<th>ECG value</th>
<th>Sensitivity (%)</th>
<th>Specificity</th>
</tr>
</thead>
<tbody>
<tr>
<td>HCM</td>
<td>+++</td>
<td>73-98</td>
<td>low</td>
</tr>
<tr>
<td>ARVC</td>
<td>+++</td>
<td>25-94</td>
<td>low</td>
</tr>
<tr>
<td>Long QT</td>
<td>+++</td>
<td>83-100</td>
<td>moderate</td>
</tr>
<tr>
<td>WPW</td>
<td>+++</td>
<td>-</td>
<td>high</td>
</tr>
<tr>
<td>CMD</td>
<td>+</td>
<td>high?</td>
<td>low</td>
</tr>
<tr>
<td>Myocarditis</td>
<td>++</td>
<td>10-54</td>
<td>low</td>
</tr>
<tr>
<td>Brugada</td>
<td>++</td>
<td>&gt;20</td>
<td>high</td>
</tr>
<tr>
<td>MVP</td>
<td>+/-</td>
<td>66</td>
<td>low</td>
</tr>
<tr>
<td>CAD</td>
<td>+/-</td>
<td>low</td>
<td>low</td>
</tr>
<tr>
<td>Coronary abn.</td>
<td>-</td>
<td>low</td>
<td>low</td>
</tr>
<tr>
<td>Marfan</td>
<td>-</td>
<td>low</td>
<td>low</td>
</tr>
</tbody>
</table>

...60% of cardiac diseases
Screening of young athletes for Cardiovascular diseases
(Center for Sports Medicine, Padua 1979-2004)

Athletes screened 42,386

Positive findings 3,914 (9%)

Heart diseases 879 (2%)

Potentially lethal heart diseases 91 (0.2%)

False positive ≈ 7%

Corrado et al JAMA 2006; 296: 1593-1601
Annual Incidence Rates of Sudden Cardiovascular Death in Screened Competitive Athletes and Unscreened Nonathletes Aged 12 to 35 Years in the Veneto Region of Italy (1979-2004)

P for trend <0.001

Corrado et al JAMA 2006;296:1593-1601
Cardiac Screening before Participation in Sports

Sanjay Sharma

Preparticipation cardiac screening should be required for all young athletes before they engage in organized competitive sports.

N.A. Mark Estes III

versial. In addition, the premise that restriction of participation in sports improves outcomes remains unproven.

gram that included ECG. The Italian, Israeli, and U.S. reports share the fundamental limitations of being retrospective, nonrandomized, observational, registry studies. Advancing potentially costly and harmful athletic screening without proven benefit would be premature.

Domenico Corrado

On the basis of the available data, the students in the vignette should undergo a preparticipation cardiovascular evaluation including ECG — the only screening strategy that has been proved to be effective.

Victoria L. Vetter

family history. However, the history and physical examination will result in fewer false positive findings than will ECG. Furthermore, a
Pre participation screening

**Cost effectiveness:**
ECG: 43000 $ / life year saved (= ICD or public access to defibrillation).

**TTE:** 200 000$

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**Maron. Circulation 2009;119:1085-92**
- 70% causes can be identified by PP screening...

**Maron. Heart Rhythm. 2013;10:374-7.**
- risk of cardiovascular SD: 1:150,000 participants/year
- 30% detectable (include ECG)...

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**Table 2. Preparticipation Athletic Screening and Athletic Restriction in Italy, the United States, and Israel**

<table>
<thead>
<tr>
<th>Country</th>
<th>Years</th>
<th>Screening</th>
<th>Initial</th>
<th>Examiners</th>
<th>Sudden Death</th>
</tr>
</thead>
<tbody>
<tr>
<td>Italy6</td>
<td>1981-2008</td>
<td>Mandatory</td>
<td>History, PE, ECG, ETT</td>
<td>Sports medicine MD</td>
<td>Decrease</td>
</tr>
<tr>
<td>United States30</td>
<td>1985-2006</td>
<td>Recommended</td>
<td>History, PE</td>
<td>MD and non-MD</td>
<td>No decrease</td>
</tr>
<tr>
<td>Israel26</td>
<td>1985 to 2009</td>
<td>Mandatory</td>
<td>History, PE, ECG, ETT</td>
<td>Certified MD</td>
<td>No decrease</td>
</tr>
</tbody>
</table>

PE indicates physical examination; ETT, exercise tolerance test.
Recommendations for competitive sports participation in athletes with cardiovascular disease

A consensus document from the Study Group of Sports Cardiology of the Working Group of Cardiac Rehabilitation and Exercise Physiology and the Working Group of Myocardial and Pericardial Diseases of the European Society of Cardiology

36th Bethesda Conference:
Eligibility Recommendations for Competitive Athletes With Cardiovascular Abnormalities

European Heart Journal (2005) 26, 1422–1445

JACC Vol. 45, No. 8, 2005
HCM
– 1/500
– Definite: Risk evaluation
  – Low risk:
    • No SD in the relatives,
    • no symptoms, mild LVH,
    • normal BP response to exercise,
    • no ventricular arrhythmia
– Uncertain/athletes’heart
HCM

Ho CY et al. NEJM 2010;363:552-63
ARVC

- 1/1000
- Sport: x 5 SD risk!
- No competitive sport
coronary arteries abnormalities

- Prevalence: 0.1 - 0.2%
- Increased risk of SD x 3 to 6 in athletes
- Premonitory Symptoms: 30% to 40%
  - Syncope
  - Chest pain
- Stress test sensitivity: 22%...

Cardiac valve disease

- **Mild**
  - All sports

- **Moderate to severe (+LV dysfunction)**
  - No competitive sports

- **Mitral valve prolapse**
  - Syncope, history of SD, arrhythmias

- **Prosthetic valve**:
  - I,II - A, B / risk of bodily collision/ VKA

Myocarditis

- **Active...**
  - no competitive sport

- **Within 6 months:**
  - Symptoms
  - LV function
  - Arrhythmias

- **Follow up**
  - 6 months

- **Prevention**
  - 58% P Activity while febrile...

References:
Marfan

- 1/5000

- Aortic rupture/dissection

- Despite normal (near normal) aortic root dimension

- Full phenotype/
- Genotype + /
- Family history:
  >>> No competitive sport

- Follow up+++

Athlete’s heart?

- Structural, functional, electrical remodeling.
- Physiological changes make a good athlete great!
- Proarrhythmic effect of endurance exercise training?

Baggish AL. *Circulation* 2011;123:2723-35
La Gerche. *Eur Heart J* 2012; 33:998-1006
Exercise induced ARVC: Myth or reality?

- 6 (27%) endurance athletes with Ventricular Arrhythmia: ARVC criteria
- Lower (12%) than expected desmosomal gene mutation prevalence in endurance athletes with complex ventricular arrhythmias of right ventricular origin.

Conclusion

• careful screening (ECG) can detect a majority of diseases

• Unexpected events: coronary abnormalities...

• Public access to defibrillation

At risk situations...
ECG Screening

<table>
<thead>
<tr>
<th>Normal athlete’s heart</th>
<th>Abnormal finding</th>
</tr>
</thead>
<tbody>
<tr>
<td>Sinus bradycardia</td>
<td>T wave inversion</td>
</tr>
<tr>
<td>First degree AV block</td>
<td>ST segment depression</td>
</tr>
<tr>
<td>Incomplete RBBB</td>
<td>Pathological Q waves</td>
</tr>
<tr>
<td>Early repolarisation</td>
<td>Left atrial enlargement</td>
</tr>
<tr>
<td>Isolated QRS voltage criteria for LVH</td>
<td>Left axis deviation/left anterior fascicular block</td>
</tr>
<tr>
<td></td>
<td>Right axis deviation/left posterior fascicular block</td>
</tr>
<tr>
<td></td>
<td>Right ventricular hypertrophy</td>
</tr>
<tr>
<td></td>
<td>Ventricular pre-excitation</td>
</tr>
<tr>
<td></td>
<td>Complete LBBB or RBBB</td>
</tr>
<tr>
<td></td>
<td>Long or short QT interval</td>
</tr>
<tr>
<td></td>
<td>Brugada-like early repolarisation</td>
</tr>
</tbody>
</table>

...60% of cardiac diseases
Exercise induced cardiomyopathy?

10 to 50% of healthy marathon runners... > 50Y.


What about young women athletes?

- **Sex Ratio of SCD during exercise:**
  - 1/5 to 1/30!
  - Less premature CAD

- **Normal heart (arrhythmic SCD...)** > 50%
  - Afro-American > White recruits (RR 10.2).
  - Impact of exercise in regular and elite athletes (MRI):
    - LV EDV > 105 ml/m², RV EDV > 110 ml/m².
    - RV EDD> 50 mm, LV EDD > 60 mm in 16% (short-axis)

- **Higher Survival at hospital admission** (46.5 vs 30%)

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Eckart RE. Am J Cardiol 2006; 97:1756-8
Prakken NH. Eur J Cardiovasc Prev Rehabil. 2010
Arrhythmias

• **Supraventricular**
  – Symptomatic AF, Flutter, AVNRT…. Ablation
  – Permanent AF (absence of heart disease, WPW) Individual basis

• **WPW**
  – Ablation (AF, symptoms…)
  – Not ablated: all sports except those with increased risk if syncope occurs.
Arrhythmias

- **Ventricular**
  - PVB, NSVT... Underlying cardiomyopathy?
- **Long QT, Brugada**: no competitive sport
- **Early repolarization**: 
  - so frequent in athletes (40%?)
- **Pacemaker**: IA IB except bodily collision
- **ICD**: IA IB (+6 months after implantation).

Safety of Sports for Athletes With Implantable Cardioverter-Defibrillators
Results of a Prospective, Multinational Registry

77 individuals/315: 121 Shocks
48 (13%): appropriate
40 (11%): inappropriate
Congenital heart diseases

- Small or repaired septal defect
- Operated PDA
- Repaired coarctation
- Treated pulmonary stenosis
- Mild moderate aortic stenosis

- Eisenmenger Sd
- Secondary Pulmonary Hypertension
- Univentricular hearts
- Congenitally, or Mustard, Senning, Rastelli Corrected TGA

Guidelines cover < 1% CHD population

...Leisure time activities

Budts et al. Eur Heart J 2013; 34:3669-74