Screening Approaches for Sudden Death in Japan

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CSRC Think Tank: Prevention of SCD in the Young
February 20th, 2015
School-Based Heart Screening in Japan

- Regulated by the School Health Law
- Implementation by local authorities
- Purpose
  - Detection of heart disease
  - Precise diagnosis
  - Clarify disease severity and decide level of management
  - Appropriate follow up of children with identified disease
School-Based Heart Screening in Japan

- 1954 Heart screening in Osaka area
- 1973 School-based heart screening
  - Participation is mandatory by the School Health Law
- 1994 School-based ECG screening
  - All 1\textsuperscript{st}, 7\textsuperscript{th}, and 10\textsuperscript{th} grade children
  - Objective is to detect heart disease which may affect school life or which may cause sudden cardiac death (SCD)

- Transition of major issues
  - 195X Diagnosis and follow up of patients with rheumatic heart disease
  - 196X~197X Detection of patients with congenital heart disease (CHD) with indication for surgery
  - 198X~ Follow up of patients who have undergone cardiac surgery, Detection of arrhythmia and hypertrophic cardiomyopathy (HCM), and their control
School-based ECG screening for all 1st, 7th, and 10th graders started in 1994.
Japanese Characteristics

- **Causes of SCD in children**
  - hypertrophic cardiomyopathy (HCM), congenital heart disease (CHD), arrhythmia without structural heart disease
  - arrhythmic death under school supervision (n=58):
    - long QT synd. 13, PVC 8, Ventricular fibrillation 7, WPW synd. 7, sick sinus synd. 7, AV block 4, Ventricular tachycardia 3, idioventricular rhythm 1, AF 1, Other 8
- **Kawasaki disease**
  - high prevalence in Asia
  - increase in the number of patients (11,756 pts in 2008)
- **Brugada synd.**
  - high prevalence in Asia
  - Incidence 14.2 /100,000 • year
The First Step of the Heart Screening

• Purpose: early detection and diagnosis of heart disease

• Screening items:
  - Questionnaire of heart disease
    History of heart disease - e.g. Kawasaki disease, rheumatic fever, symptoms
  - Physical examination by school doctors
    heart murmur – e.g. Atrial septal defect (ASD)
  - ECG
    12-lead or 4-lead ECG (I, aVf, V1, V6)
    Read by pediatric cardiologists
  - (Phonocardiography )
The First Step of the Heart Screening (cont.)

- Criteria for further examination
  - Abnormal ECG findings
    - PVC, PAC, CRBBB, 1st and 2nd degree AV block, WPW synd., AV dissociation, escaped beat, severe bradycardia (HR<40bpm), bifascicular and trifascicular blocks, aberrant conduction, QS pattern, deep Q waves, ST depression, abnormal T waves
  - History of heart disease
    - e.g. Kawasaki disease, rheumatic fever, hypertension
  - Symptoms
    - e.g. arrhythmia, syncope
The Second Step of the Heart Screening

• <5% of all screened children
• Purpose: Diagnosis of heart disease
  Clarify severity of heart disease
• Screening items
  ➢ Physical examination by pediatric cardiologists
  If necessary,
  ➢ ECG
  ➢ Chest X-ray
  ➢ Echocardiography
  ➢ Exercise Stress Tests
  ➢ 24 hour Holter ECG
• Children who need further medical management or treatment are referred to hospital (the third step).
The Third Step of the Heart Screening

- **Purpose:** Diagnosis of heart disease
  Clarify severity of disease
  Control exercise tolerance and school lifestyle

- **Structured management of children with heart disease:**
  - Congenital heart disease
  - Arrhythmia without structural heart disease
  - Kawasaki disease

- **Structured management for control of exercise and school lifestyle**
  - A: Require admission to a hospital or home medical care
  - B: Exercise is not allowed
  - C: Light exercise is allowed
  - D: Moderate exercise is allowed
  - E: Heavy exercise is allowed
Outlines of the school-based screening system in Kagoshima City in 2012

<table>
<thead>
<tr>
<th>Step</th>
<th>1&lt;sup&gt;st&lt;/sup&gt; graders</th>
<th>7&lt;sup&gt;th&lt;/sup&gt; graders</th>
<th>10&lt;sup&gt;th&lt;/sup&gt; graders</th>
<th>Special Class</th>
<th>Total</th>
<th>%</th>
</tr>
</thead>
<tbody>
<tr>
<td>1&lt;sup&gt;st&lt;/sup&gt; step</td>
<td>5,270</td>
<td>5,725</td>
<td>4,173</td>
<td>215</td>
<td>15,383</td>
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<tr>
<td>Subjects</td>
<td>5,259</td>
<td>5,689</td>
<td>4,121</td>
<td>211</td>
<td>15,280</td>
<td>99%</td>
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<tr>
<td>Participants</td>
<td>67</td>
<td>106</td>
<td>110</td>
<td>6</td>
<td>289</td>
<td></td>
</tr>
<tr>
<td>Disease +</td>
<td>20</td>
<td>26</td>
<td>36</td>
<td>1</td>
<td>83</td>
<td></td>
</tr>
<tr>
<td>2&lt;sup&gt;nd&lt;/sup&gt; step</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Subjects</td>
<td>65</td>
<td>101</td>
<td>103</td>
<td>6</td>
<td>275</td>
<td>95%</td>
</tr>
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<td>Participants</td>
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<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Disease +</td>
<td>6</td>
<td>13</td>
<td>7</td>
<td>0</td>
<td>26</td>
<td></td>
</tr>
<tr>
<td>3&lt;sup&gt;rd&lt;/sup&gt; step</td>
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<td></td>
<td></td>
</tr>
<tr>
<td>Subjects</td>
<td>8</td>
<td>14</td>
<td>8</td>
<td>0</td>
<td>30</td>
<td></td>
</tr>
<tr>
<td>Participants</td>
<td>8</td>
<td>14</td>
<td>7</td>
<td>0</td>
<td>29</td>
<td>97%</td>
</tr>
<tr>
<td>Disease +</td>
<td>6</td>
<td>13</td>
<td>7</td>
<td>0</td>
<td>26</td>
<td></td>
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## Results of the school-based screening system in Kagoshima City 2008-2012

<table>
<thead>
<tr>
<th></th>
<th>2008</th>
<th>2009</th>
<th>2010</th>
<th>2011</th>
<th>2012</th>
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</thead>
<tbody>
<tr>
<td><strong>Total subjects</strong></td>
<td>16,168</td>
<td>16,129</td>
<td>16,100</td>
<td>15,615</td>
<td>15,383</td>
</tr>
<tr>
<td><strong>2nd step</strong></td>
<td></td>
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<tr>
<td><strong>2nd step</strong></td>
<td>246 (1.52%)</td>
<td>281 (1.70%)</td>
<td>311 (1.90%)</td>
<td>338 (2.20%)</td>
<td>289 (1.88%)</td>
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<tr>
<td><strong>Abnormal</strong></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td><strong>findings in 2nd step</strong></td>
<td>83 (0.51%)</td>
<td>105 (0.65%)</td>
<td>110 (0.68%)</td>
<td>130 (0.83%)</td>
<td>109 (0.71%)</td>
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<tr>
<td><strong>CV disease+</strong></td>
<td>20</td>
<td>29</td>
<td>21</td>
<td>41</td>
<td>26</td>
</tr>
<tr>
<td><strong>Already</strong></td>
<td>316</td>
<td>250</td>
<td>309</td>
<td>263</td>
<td>305</td>
</tr>
</tbody>
</table>

* : Subjects who were diagnosed as having CV diseases by the program.
#: Subjects who had already diagnosed and followed were not included in the 2nd step.

Yoshinaga M. Pediatric Cardiology and Cardiac Surgery 2013;29:212-17
Conclusions

- Japan has established a nation-wide school-based heart screening system including ECG screening.
- Prevalence of SCD under school supervision has decreased since the school-based heart screening started.
- CHD, HCM, Kawasaki disease, WPW synd., Long QT synd., Brugada like ECG pattern has been detected.
- The School-based heart screening in Japan has been shown to be useful for identifying high-risk subjects among both athletes and non-athletes.
- The program’s cost-effectiveness is considered appropriate.
Conclusions (cont.)

• Sensitivity and specificity of the screening system needs to be improved; e.g. timing, method(-s) for screening, target population, ECG criteria

• In addition to screening, implementation of Automated External Defibrillator (AED) is necessary to prevent SCD in children.

• Creation of a nation-wide electronic ECG database is considered as a future project.

• Such ECG database might be useful for assessing cardiac safety in clinical pediatric drug development.