Exercise Capacity in Children and Young Adults after Repair of Congenital Heart Disease

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I have no disclosure
Introduction

• CHD patients are usually followed-up using resting echocardiography/MRI, and no *functional* tests are performed.

• Cardiopulmonary exercise testing (CPET) enables evaluation of maximal cardiac capacity, providing important information on functional outcome.
Introduction
Introduction- Gas exchange
Derangements of gas exchange in disease

- Obesity
- Airflow obstruction
- Heart disease
  - Coronary
  - Myocardial
  - Valvular
  - Anemia
- Obstruction
- Restriction
- Chestwall
- Infiltrative
- Occlusion
- Hypertension
- Vasoregulatory asthenia
- Thromboemboli
- Vasculitis
- 1° Pulmonary hypertension

VO$_2$

- Oxygen uptake
  - determined by cellular demand
  - level of maximal O$_2$ transport
- Normal VO$_2$
  - Age
  - Sex
  - Body size
  - Training
  - Motivation
Cardiac Output vs. $O_2$ uptake
VO$_2$\textsubscript{max} – normalized to age and gender

- Boys 42 ml/kg/min
- Girls 38 ml/kg/min
Aerobic Capacity in Adults With Various Congenital Heart Diseases

<table>
<thead>
<tr>
<th>Age at test</th>
<th>ASD</th>
<th>CCTGA</th>
<th>Ebstein</th>
<th>Fontan</th>
<th>Mustard</th>
<th>ToF</th>
</tr>
</thead>
<tbody>
<tr>
<td>18-19 years</td>
<td>18</td>
<td>15</td>
<td>9</td>
<td>5</td>
<td>16</td>
<td>14</td>
</tr>
<tr>
<td>20-29 years</td>
<td>27</td>
<td>8</td>
<td>11</td>
<td>30</td>
<td>57</td>
<td>63</td>
</tr>
<tr>
<td>30-39 years</td>
<td>18</td>
<td>4</td>
<td>9</td>
<td>12</td>
<td>8</td>
<td>50</td>
</tr>
<tr>
<td>40-49 years</td>
<td>18</td>
<td>2</td>
<td>9</td>
<td>4</td>
<td>8</td>
<td>30</td>
</tr>
<tr>
<td>50-59 years</td>
<td>16</td>
<td>2</td>
<td>5</td>
<td>3</td>
<td>17</td>
<td>17</td>
</tr>
<tr>
<td>60-69 years</td>
<td>14</td>
<td>3</td>
<td>3</td>
<td>4</td>
<td>17</td>
<td></td>
</tr>
</tbody>
</table>

Am J Cardiol 2001;87:310–314
Distribution of peak VO2 in different diagnostic groups

Distribution of peak VO$_2$ in asymptomatic patients with ACHD (NYHA class I)

Exercise intolerance in ACHD: comparative severity, correlates, and prognostic implication

Self estimated functioning vs. VO$_2$peak

Figure 3: Correlation of objectively measured aerobic physical capacity and self-estimated physical functioning in 564 adolescents and adults with congenital heart disease ($r = 0.435$, $P = 1.72 \times 10^{-22}$); additionally depicting that many patients overestimate their physical capabilities.

European Heart Journal (2009) 30, 497–504
Self-estimated physical functioning in CHD patients
$\frac{V_E}{V_{CO2}}$ slope

- Minute ventilation / CO$_2$ production
- Index of gas exchange efficiency during exercise
  - Liters of air exhaled / 1 Liter of CO2 eliminated
- In children < 28
$\frac{V_E}{V_{CO_2}}$ in HF vs. respiratory dyspnea

Clinician’s Guide to cardiopulmonary exercise testing in adults: a scientific statement from the American Heart Association
$V_E / V_{CO_2}$ in a TOF patient before and after successful LPA balloon angioplasty

$V_E/V_{CO_2}$ in a patient with fenestrated Fontan before and after fenestration closure
$\text{VO}_2/\text{HR} = \text{Oxygen pulse (O}_2\text{P)}$

- Indirect measurement of SV (stroke volume)
- Increases throughout a ramping exercise
- Falling values during increasing workloads indicate an abnormal SV
- Should be > 80% of predicted at max exercise

**Cardiac Patient**

**Elite Athlete**
3.2.4 Cardiopulmonary exercise testing

Formal exercise testing has an important role in the GUCH population, in which quality of life and functional capacity are key measures of the success of intervention. Traditional exercise testing uses protocols that are largely designed for risk stratification of ischaemic heart disease and are often not appropriate in GUCH patients. CPET, including assessment of objective exercise capacity (time, maximum oxygen uptake), ventilation efficiency (VE/VCO₂ slope), chronotropic and blood pressure response, as well as exercise-induced arrhythmia, gives a broader evaluation of function and fitness, and has endpoints which correlate well with morbidity and mortality in GUCH patients. Serial exercise testing should therefore be a part of long-term follow-up protocols and interventional trials. It plays an important role in the timing of interventions and re-interventions.
Combined end point of hospitalization or death (event-free survival)

Quartile 1: peak VO₂ <15.5 ml/kg/min
- At risk: 84
- Event-free survival: 100%

Quartile 2 and 3: peak VO₂ 15.5 - 27.0 ml/kg/min
- At risk: 154
- Event-free survival: 100%

Quartile 4: peak VO₂ >27.0 ml/kg/min
- At risk: 81
- Event-free survival: 100%
Freedom From Death/Emergency Cardiac-Related Hospital Admission Stratified by Combination of VE/VCO₂ Slope and Peak VO₂

Ventilatory efficiency and aerobic capacity predict event-free survival in adults with atrial repair for TGA

Clinical Practice and Education Paper

Physical performance and physical activity in grown-up congenital heart disease
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Evaluation of children with congenital heart defects
To get an objective assessment of the functional capacity of children and adolescents with congenital heart defects, formal exercise testing should be performed with continuous measurement of gas exchange. Other methods such as history taking and questionnaires are inaccurate and not sensitive.
Guidelines for the Outpatient Management of Complex Congenital Heart Disease


1 we believe that individual assessment of the child’s cardiopulmonary function during exercise combined with the routine resting cardiovascular evaluations is essential to tailor appropriate activity level recommendations for these children and adolescents. This approach is discussed in detail in the clinical guidance sections.

Congenit Heart Dis. 2006;1:10–26
Multidisciplinary lab
Multidisciplinary team

Pediatric Cardiologist

Sport Medicine

Physiologist
Objectives

• To determine exercise capacity and cardiac function of patients with repaired CHD compared with normal controls.

• To compare measures of fitness, cardiac and pulmonary functions between CHD patients with complete or incomplete repair, as determined by resting echocardiography.
Methods

• **Design**: Retrospective analysis of prospectively-collected data

• **Population**:
  - All CHD patients <40 yrs old, with no significant additional co-morbidities,
  - After biventricular corrective interventions (surgery or catheterization),
  - CHD subgroups divided by the presence of significant anatomical residua on a resting echocardiogram
Methods

• **Controls**
• otherwise healthy children and adolescents referred to our lab for evaluation of chest pain, palpitations, arrhythmias, conduction disorders etc, and were determined to have normal cardiac function

• CPET on a cycle ergometer in our institution.
Methods- CHD subgroups

Complete repair (n=49)

- TOF – 13
- TGA (all s/p ASO)- 8
- VSD/ DCRV- 8
- COA – 4
- Ross- 3
- PS- 2
- AVC, MS, DORV, PAPVR, IHSS
Methods - CHD subgroups

Incomplete repair (n=24)

- TOF with residual PI -8
- PS/PPS (TOF, VSD+PS, DORV, Rastelli) -5
- TGA s/p Mustard -3
- LV dysfunction (TGA, TOF, ASO) - 4
- PS (PS, Ross) - 2
- AS -1
- AI -1
Methods

• Measures of cardiac function were compared between CHD (n=73) and control (n=76) groups using multiple linear regression techniques and ANCOVA, adjusting for age and sex.

• Similar comparisons were made between CHD patients with complete (n=49) vs. incomplete (n=24) repair

• Values also expressed as % predicted for age and sex
Methods

• CPET data analyzed for this study were:

  – Peak VO$_2$: aerobic capacity

  – Peak O$_2$ pulse: relates to stroke volume at peak exercise

  – $V_E/VCO_2$ slope: gas exchange efficiency $\approx$ cardiac function
Results- peak VO$_2$ (ml/kg/min)

Age adjusted results:

- Complete 32.7 +/-1.2  p value 0.59
- Incomplete 28.8 +/-1.9  0.03  0.05
- Control 36.4 +/-1.1
Results- peak VO₂ (ml/kg/min)

% of predicted value:

- Complete: 73.8 +/- 17.9, p value 0.96
- Incomplete: 66.3 +/- 20.1, p value 0.02
- Control: 92.9 +/- 22.0, p value 0.01
## Results- O$_2$ pulse

### Age adjusted results:

<table>
<thead>
<tr>
<th>Category</th>
<th>Mean</th>
<th>Standard Deviation</th>
<th>p value</th>
</tr>
</thead>
<tbody>
<tr>
<td>Complete</td>
<td>9.9</td>
<td>+/-0.47</td>
<td>0.059</td>
</tr>
<tr>
<td>Incomplete</td>
<td>8.0</td>
<td>+/-0.70</td>
<td>0.28</td>
</tr>
<tr>
<td>Control</td>
<td>11.0</td>
<td>+/-0.40</td>
<td>0.00</td>
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</table>
### Results - O₂ pulse

**% of predicted value:**

<table>
<thead>
<tr>
<th>Group</th>
<th>Value</th>
<th>Standard Deviation</th>
<th>p value</th>
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</thead>
<tbody>
<tr>
<td>Complete</td>
<td>90.5</td>
<td>+/- 2.9</td>
<td>0.047</td>
</tr>
<tr>
<td>Incomplete</td>
<td>78.4</td>
<td>+/- 4.3</td>
<td>0.26</td>
</tr>
<tr>
<td>Control</td>
<td>94.4</td>
<td>+/- 2.4</td>
<td>0.001</td>
</tr>
</tbody>
</table>
### Results- $V_E/VCO_2$ slope

Age adjusted results:

<table>
<thead>
<tr>
<th>Group</th>
<th>$V_E/VCO_2$</th>
<th>$\pm$</th>
<th>$p$ value</th>
</tr>
</thead>
<tbody>
<tr>
<td>Complete</td>
<td>27.7</td>
<td>+/-0.6</td>
<td>0.55</td>
</tr>
<tr>
<td>Incomplete</td>
<td>30.2</td>
<td>+/-0.9</td>
<td>0.04</td>
</tr>
<tr>
<td>Controls</td>
<td>25.8</td>
<td>+/-0.5</td>
<td>0.001</td>
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</table>
Results- Peak VO$_2$

- CHD patients had 25% lower **aerobic fitness** compared with controls:
  - Peak VO$_2$ 29±8 vs. 38±10 ml/kg/min, p=0.001

- 19% of CHD patients had normal fitness (peak VO$_2$ >85% predicted) vs.
- 62% of controls (p<0.001):

![Pie charts comparing CHD and CON groups]
Results - Peak $O_2$ pulse

- Peak $O_2$ pulse was abnormal in
  - 53% of CHD patients vs.
  - 24% of controls ($p<0.001$)
Results - VE/VCO₂ slope

- A significantly higher VE/VCO₂ slope was seen in CHD group compared to controls - 28±5 vs. 26±3 (p=0.019)

- 30% of CHD patients had an abnormal slope (>30), compared with 14% of controls (p=0.021)

- None of the measured parameters differed between CHD subgroups, except VE/VCO₂ slope:
  - An abnormal slope was found in 22% with complete repair, but 46% with incomplete repair.
Study groups vs. variables

- Cmp-Inc
- Cnt-Inc
- Cnt-Cmp

- VO2max
- VE/VCO2
- O2 pulse
Conclusions

• Patients with biventricular CHD repair have a significantly decreased exercise capacity, due to abnormal cardiac function and deconditioning

• The measured parameters were low in all CHD patients, indicating the limited ability of resting echocardiography in assessing cardiac capacity.

• Patients with incomplete repair have a significantly higher VE/VCO₂ slope
Conclusions

• Functional cardiopulmonary capacity should be determined, in order to assign our patient a safe level of activity

• Peak VO2 and VE/VCO2 are important prognostic factors
  
  – CPET should be performed routinely to help plan interventions
  
  – Patients should learn to view physical activity as an important component of their medical care
Thank you:

• Ronen Reuveny, PhD
• Omer Rosenblum, medical student
• Avshalom Koren, PhD
• Gal Dubnov-Raz, MD
QUESTIONS?