6 Χρόνια CHALLENGE: Πού βρισκόμαστε το 2018;

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Changing demographics of CHD

- Adults with CHD
- Children with CHD (90% of live births)
- 18 y.o. (80% of children with CHD)
- Live births with CHD (0.8% of live births)

Changing mortality in ACHD

Valvular and Congenital Heart Disease

The prevalence of adult congenital heart disease, results from a systematic review and evidence based calculation

Teun van der Bom, MD, Berto J. Bouma, MD, PhD, Folkert J. Mcijboom, MD, PhD, Acilko H. Zwinderman, PhD, and Barbara J. M. Mulder, MD, PhD. Amsterdam, and Utrecht, The Netherlands
The number of adults with CHD exceeds the number of children by 2:1.
ACHD centers worldwide

Webb G et al. Int J Cardiol. 2015
Background

Up to now there are no data on the epidemiology of CHD in Greece
Purpose

- Estimation of the prevalence and long-term outcomes of congenital heart defects in Greece

- Development of an efficient organizational structure for the improvement of healthcare for patients with congenital heart disease
Methods and Materials

• Initiation of the registry in January 2012
• Informed consent
• Patients > 16 years old
• 18 expert ACHD centers
Methods and Materials

http://www.hcs.crf.gr/
Results
Πορεία της καταγραφής

N=2420
Adult congenital heart disease in Greece: Preliminary data from the CHALLENGE registry

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ABSTRACT

Background: The majority of patients with congenital heart disease (CHD), nowadays, survives into adulthood and is faced with long-term complications. We aimed to study the basic demographic and clinical characteristics of adult patients with congenital heart disease (ACHD) in Greece.

Methods: A registry named CHALLENGE (Adult Congenital Heart Disease Registry, A registry from Hellenic Cardiology Society) was initiated in January 2012. Patients with structural CHD older than 16 years old were enrolled by 16 specialized centers nationwide.

Results: Out of a population of 2115 patients with ACHD, who have been registered, (mean age 38 years (SD 16), 52% women), 47% were classified as suffering from mild, 37% from moderate and 15% from severe ACHD. Atrial septal defect (ASD) was the most prevalent diagnosis (33%). The vast majority of ACHD patients (92%) was asymptomatic or mildly symptomatic (NYHA class I/II). The most symptomatic patients were suffering from an ASD, most often the elderly or those under targeted therapy for pulmonary arterial hypertension. Elderly patients (>60 years old) accounted for 12% of the ACHD population. Half of patients had undergone at least one open-heart surgery, while 39% were under cardiac medications (15% under antiarrhythmic drugs, 16% under anticoagulants, 16% under medications for heart failure and 4% under targeted therapy for pulmonary arterial hypertension).

Conclusions: ACHD patients are an emerging patient population and national prospective registries such as CHALLENGE are of unique importance in order to identify the ongoing needs of these patients and match them with the appropriate resource allocation.

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Results until April 2017

- $N=2115$
- mean age 38 years (SD 16)
- 52% women
- 47%-> mild, 37%-> moderate, 15%-> severe ACHD
- 12% elderly (>60 years)
ACHD subtypes

CHALLENGE team. IJC 2017
Elderly = 12% of the population

More often mild CHD and underwent fewer surgeries, while almost two thirds did not undergo any surgical procedure at all.

Only 38% of elderly patients were asymptomatic.
## Results

### Table 1

<table>
<thead>
<tr>
<th>Variable</th>
<th>Overall n = 2115</th>
<th>Mild n = 1008</th>
<th>Moderate n = 790</th>
<th>Severe n = 317</th>
<th>p-value</th>
</tr>
</thead>
<tbody>
<tr>
<td>Age</td>
<td>38.0 ± 16.2</td>
<td>39.3 ± 17.3</td>
<td>37.7 ± 15.2</td>
<td>34.6 ± 13.9</td>
<td>&lt;0.001</td>
</tr>
<tr>
<td>Males</td>
<td>1008 (47.7%)</td>
<td>438 (43.5%)</td>
<td>417 (52.8%)</td>
<td>153 (48.3%)</td>
<td>&lt;0.001</td>
</tr>
<tr>
<td>NYHA I/II</td>
<td>1952 (92.3%)</td>
<td>965 (95.7%)</td>
<td>744 (94.2%)</td>
<td>243 (76.7%)</td>
<td>&lt;0.001</td>
</tr>
<tr>
<td>NrSurgeries</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>0</td>
<td>1061 (50.2%)</td>
<td>745 (73.9%)</td>
<td>232 (29.4%)</td>
<td>84 (26.5%)</td>
<td>&lt;0.001</td>
</tr>
<tr>
<td>1</td>
<td>657 (31.1%)</td>
<td>212 (21.0%)</td>
<td>344 (43.5%)</td>
<td>101 (31.9%)</td>
<td></td>
</tr>
<tr>
<td>≥2</td>
<td>397 (18.8%)</td>
<td>51 (5.1%)</td>
<td>214 (27.1%)</td>
<td>132 (41.6%)</td>
<td></td>
</tr>
<tr>
<td>Cardiac Medication Use</td>
<td>821 (38.8%)</td>
<td>311 (30.9%)</td>
<td>315 (39.9%)</td>
<td>195 (61.5%)</td>
<td>&lt;0.001</td>
</tr>
<tr>
<td>Antiarrhythmic drugs</td>
<td>314 (14.8%)</td>
<td>97 (9.6%)</td>
<td>137 (17.3%)</td>
<td>80 (25.2%)</td>
<td>&lt;0.001</td>
</tr>
<tr>
<td>Medication for HF</td>
<td>342 (16.2%)</td>
<td>109 (10.8%)</td>
<td>132 (16.7%)</td>
<td>101 (31.9%)</td>
<td>&lt;0.001</td>
</tr>
<tr>
<td>Anticoagulants</td>
<td>330 (15.6%)</td>
<td>138 (13.7%)</td>
<td>92 (11.6%)</td>
<td>100 (31.5%)</td>
<td>&lt;0.001</td>
</tr>
</tbody>
</table>

Nr: Number; NYHA: New York Heart Association, HF: Heart Failure.

- 71% WHO I/II
- NYHA III/IV mainly ASD, TOF, AV/VA abnormal connection, single ventricle
Females were older (mean age 40 years (SD 17) vs 36 years (SD 16), p<0.001), suffered more often from mild ACHD (52% vs 44%, p<0.001) and underwent fewer surgeries (no surgery was performed in 56% vs 44%, p<0.001).
Heart failure in ACHD

- AS: 14%
- VSD: 10%
- Ao Arch Abnormalities: 12%
- SV: 52%
- AVSD: 27%
- TOF: 19%
- AV/VA abnormal connections: 29%
- ASD: 12%

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Antiarrhythmic medical treatment in ACHD

- PS: 13%
- AS: 9%
- VSD: 8%
- Ao Arch Abnormalities: 8%
- SV: 29%
- AVSD: 25%
- TOF: 23%
- AV/VA abnormal connections: 27%
- ASD: 12%

CHALLENGE team. IJC 2017
WHAT'S NEXT?
Pulmonary hypertension in ACHD
Out of 2399, N=68 are in PAH treatment (≈3%)

8 excluded:
3 Fontan
5 Segmental PAH

Eisenmenger 42 pts (70%)
L-> R 12 pts (20%)
Small defects 1 pt (2%)
After repair 6 pts (10%)
Baseline Characteristics (n=60)

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<table>
<thead>
<tr>
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<th></th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Age</strong></td>
<td>46.5±14.8</td>
</tr>
<tr>
<td><strong>Males</strong></td>
<td>21 (35%)</td>
</tr>
<tr>
<td><strong>NYHA III/IV</strong></td>
<td>33 (55%)</td>
</tr>
<tr>
<td><strong>Down Syndrome</strong></td>
<td>9 (15%)</td>
</tr>
<tr>
<td><strong>PAH pharmacotherapy</strong></td>
<td></td>
</tr>
<tr>
<td>ERA</td>
<td>54 (90%)</td>
</tr>
<tr>
<td>PDE5i</td>
<td>29 (48%)</td>
</tr>
<tr>
<td>Prostanoids</td>
<td>4 (7%)</td>
</tr>
<tr>
<td><strong>Combination therapy</strong></td>
<td>26 (43%)</td>
</tr>
<tr>
<td><strong>Oxygen therapy</strong></td>
<td>13 (22%)</td>
</tr>
</tbody>
</table>
## Complications

<table>
<thead>
<tr>
<th>Condition</th>
<th>Count (Percentage)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Arrhythmias</td>
<td>19 (32%)</td>
</tr>
<tr>
<td>Endocarditis</td>
<td>1 (2%)</td>
</tr>
<tr>
<td>Stroke</td>
<td>3 (5%)</td>
</tr>
<tr>
<td>Pulmonary thrombosis</td>
<td>4 (7%)</td>
</tr>
<tr>
<td>HF hospitalization</td>
<td>15 (25%)</td>
</tr>
<tr>
<td>Hemoptysis</td>
<td>2 (3%)</td>
</tr>
<tr>
<td>Death</td>
<td>12 (20%)</td>
</tr>
<tr>
<td>Right heart failure</td>
<td>6 (10%)</td>
</tr>
<tr>
<td>Sepsis</td>
<td>2 (3%)</td>
</tr>
<tr>
<td>Arrhythmia</td>
<td>1 (2%)</td>
</tr>
<tr>
<td>Sudden death</td>
<td>1 (2%)</td>
</tr>
<tr>
<td>Massive pulmonary hemorrhage</td>
<td>1 (2%)</td>
</tr>
<tr>
<td>Unknown</td>
<td>1 (2%)</td>
</tr>
</tbody>
</table>
Univariable predictors for cumulative mortality risk

<table>
<thead>
<tr>
<th>Univariable predictors</th>
<th>HR</th>
<th>95% CI</th>
<th>P Value*</th>
</tr>
</thead>
<tbody>
<tr>
<td>NYHA class III/IV</td>
<td>8.88</td>
<td>1.15-68.92</td>
<td>0.04</td>
</tr>
<tr>
<td>Female gender</td>
<td>3.02</td>
<td>0.66-13.8</td>
<td>0.16</td>
</tr>
<tr>
<td>Eisenmenger</td>
<td>1.33</td>
<td>0.40-4.14</td>
<td>0.65</td>
</tr>
<tr>
<td>Age</td>
<td>1.04</td>
<td>1.00-1.08</td>
<td>0.08</td>
</tr>
<tr>
<td>Arrhythmia</td>
<td>0.75</td>
<td>0.24-2.36</td>
<td>0.62</td>
</tr>
<tr>
<td>HF hospitalization</td>
<td>7.37</td>
<td>2.2-24.66</td>
<td>0.001</td>
</tr>
<tr>
<td>Combination Therapy</td>
<td>0.76</td>
<td>0.25-2.37</td>
<td>0.64</td>
</tr>
<tr>
<td>Down Syndrome</td>
<td>0.71</td>
<td>0.15-3.28</td>
<td>0.66</td>
</tr>
</tbody>
</table>

*Based on Univariate Cox model
Future perspectives - Follow up
27600 ACHD patients wanted!!
THE CHALLENGE TEAM

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Συμβάλλοντες στην καταγραφή

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Κ. Βανταλής
Ι. Βλασερός

Νέα κέντρα
Α. Πατριανάκος
Μ. Αργυρίου

Thank you!
Back Up
Conclusions (I)

Severe ACHD patients:

• were younger
• more symptomatic
• underwent multiple surgeries
• were more frequently under medical therapy for heart failure and arrhythmias.
Conclusions (II)

The most common form of ACHD that required HF treatment was SV.

Patients with SV, AV/ VA abnormal connections, TOF and AVSD were more frequently under treatment for arrhythmias.
More to come....

• More registered patients
• More participating centers
• More investigators
• More results to publish
Conclusions (III)
Limitations

• Misclassification bias

• The entire data of the patients were not accessible

• The high proportion of expert centers might induce bias