Burden of Adults with Congenital Heart Disease (ACHD) in the World:
A Tsunami is approaching

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In what sense can ACHD be compared with a Tsunami?

1- Fast increase in number of patients (waves)
2- Inability to cope with speed of the wave
3- ACHD patients get no timely help
   = fast increase in morbidity & mortality
4- Also for ACHD there are warning signs and systems
Definition of ACHD

Patients with Congenital Heart Disease (CHD)

12 - 14 - 18 yrs of age
Incidence of Congenital Heart Disease
- $8/1000$ live born

If left untreated
- $20$-$25\%$ die during infancy
- $60\%$ die during first years of life
- Only $15\%$ survive into adulthood
Natural & un-natural History of CHD
Atrial Septal Defect (ASD)

- Secundum ASD 70%
- Primum 15%
- Sinus Venosus 10%
- Coronary Sinus (rare)

7% (20%)
Most common CHD in Adults
Survival curve of Untreated ASD Patients vs General Population

Figure 15-9  Plot of Campbell's actuarial survival computations of the life expectancy of surgically untreated patients with atrial septal defects who reach the age of 1 year. It is based on three sets of collected data. There is some spread between the three sets of data, indicating confidence limits of modest width around the point estimates (they cannot be calculated from the data). The life expectancy of the general population 1 year of age is also from Campbell and is very close to that computed from United States life tables. The data (see "Natural History") suggest that 99.9% of patients born with atrial septal defect (ASD) reach the first year of life unless unrelated conditions cause their death.

(Data are from Campbell.29)
Natural History of ASD

25 %
dead before their 30's

75 %
dead by the age of 50 yrs

90 %
dead by the age of 60 yrs
Survival Following CHD Surgery

Daniels, CJ. Congenital Heart Disease. ACCSAP V
Coarctation of Aorta: MRI
Coarctation of Aorta: Surgery

Area of constriction to be completely removed

Post Surgery
Aortic Coarctation

Survival after surgery by age at operation

Kirklin & Barratt Boyes
Survival After Coarctation Repair

Mean Age of Death: 38 yo

% Survival

Cause of Death

<table>
<thead>
<tr>
<th>Cause of Death</th>
<th>%</th>
</tr>
</thead>
<tbody>
<tr>
<td>*CAD (mean 40 yo)</td>
<td>37</td>
</tr>
<tr>
<td>Sudden Death</td>
<td>13</td>
</tr>
<tr>
<td>CHF</td>
<td>9</td>
</tr>
<tr>
<td>CVA</td>
<td>7</td>
</tr>
<tr>
<td>Ruptured Aorta</td>
<td>7</td>
</tr>
<tr>
<td>Perioperative Death</td>
<td>7</td>
</tr>
<tr>
<td>Other</td>
<td>20</td>
</tr>
</tbody>
</table>

n = 571

Natural History

Patients with uncorrected TOF

10% reach 3\textsuperscript{rd} decade
3% reach 4\textsuperscript{th} decade
Tetralogy: Palliation

In adults

- Surgery

If anatomy does not allow complete repair

- Stent of MAPCA
- Stent of BT shunts
- BAP of RVOTO
Natural History

Survival Following Complete Repair of Tetralogy of Fallot

- Control Population:
  - 100% at 0 years
  - 96% survival at 30 years

- Sudden Death:
  - Accounted for 70%
  - Older age > 12y is a strong predictor
  - Transannular patch is associated

- n=163

- p<0.01
Only PDA Surgical ligation deserves the term “total correction“

otherwise there is no total correction or complete repair
“There is no operation for CHD that is corrective in the strictest sense, if, by corrective, we mean that all sufferers are restored to complete normality by operation.

The hard truth is ...particularly sad..for those who have undergone supposedly corrective surgery”.  

Perloff
Residua after Surgery

Residua
- Conditions that are ‘left-over,’ i.e. that are obligatory to or consequences of the pre-operative diagnosis and state.
- They are not the result of surgery

Examples
- Systemic right ventricle in congenitally corrected transposition
- Unrepaired anomalous coronary artery in tetralogy of Fallot
Sequelae after surgery

Sequelae

- Conditions that arise as a result of the operation and that are considered necessary consequences of the operation
- They are specifically ‘intrinsic’ to the operative design

Examples

- Ventricular tachyarrhythmias after ventriculotomy
- Pulmonary regurgitation after tetralogy repair
How to explain the increase in ACHD patients?

Let us go to the beginning of the problem and the “culprits”

The Cardiac Surgeons

1950 first surgical repair for CHD
Gibbon Heart-lung Machine

Lawrence H. Cohn Circulation. 2003;107:2168-2170
Adult Congenital Heart Disease

- All this improved longevity

- Population of ACHD is growing

In the last 20 years, the number of adult patients with CHD exceeded their pediatric counterpart
Adults outgrown Children with CHD

Changing Proportion of Pediatric & Adult CHD

Patients Reaching Adulthood with CHD

Adolescent CHD Patients In USA

325,000

500,000

750,000

1,000,000

1,300,000

1970

1980

1990

2000

2010

Hoffman

Fyler

Ferencz

1978

1980

1985

Kaiser

New England

Baltimore-DC

20,000 new patients/yr

5% increase/yr

Courtesy Michael Landzberg
USA
Magnitude ACHD in 320 million
2.4 million: 1 million children; 1.5 million adults

<table>
<thead>
<tr>
<th>Category and Age Group</th>
<th>CHD Severity/Race-Ethnicity</th>
<th>Estimated US Prevalence per 1000 (95% Confidence Interval), %</th>
<th>Estimated No. of Individuals (95% Confidence Interval)*</th>
</tr>
</thead>
<tbody>
<tr>
<td>CHD severity</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>All ages</td>
<td>Overall</td>
<td>7.85 (7.79–7.92)</td>
<td>2 425 000 (2 405 000–2 444 000)</td>
</tr>
<tr>
<td></td>
<td>Severe</td>
<td>0.92 (0.90–0.94)</td>
<td>283 000 (277 000–290 000)</td>
</tr>
<tr>
<td>Children</td>
<td>Overall</td>
<td>13.21 (13.03–13.39)</td>
<td>980 000 (966 000–993 000)</td>
</tr>
<tr>
<td></td>
<td>Severe</td>
<td>1.66 (1.60–1.73)</td>
<td>123 000 (119 000–128 000)</td>
</tr>
<tr>
<td>Adults</td>
<td>Overall</td>
<td>6.16 (6.10–6.22)</td>
<td>1 444 500 (1 431 000–1 459 000)</td>
</tr>
<tr>
<td></td>
<td>Severe</td>
<td>0.68 (0.66–0.70)</td>
<td>160 000 (155 000–165 000)</td>
</tr>
</tbody>
</table>

In most developing countries no clear estimates of CHD patients
Our best estimates indicate that worldwide, adults comprise 22 – 26 % of the total CHD population. Relative proportion of adults within the CHD population are 20% in Taiwan and 32 % in Thailand.
World ACHD population

Applying a prevalence range of 1.7 to 4.1 / 1,000 adults to a world population of 7.2 billion = estimated worldwide 12 to 34 million ACHD
Bibliometric approach for better estimating actual situation of ACDH worldwide
Meeting the challenge: The evolving global landscape of adult congenital heart disease

To collect information on worldwide distribution & volume of (ACHD) centers:

Centers assessed using bibliometric approach

Kempny A et al.
International journal of cardiology 168(6) · July 2013
Meeting the challenge: The evolving global landscape of adult congenital heart disease

Between 1995 and 2011 identified

94,119 ACHD publications

Kempny A et al.

International journal of cardiology 168(6) · July 2013
Meeting the challenge: The evolving global landscape of adult congenital heart disease

Results

Steady increase in number of publishing ACHD centers worldwide
Meeting the challenge: The evolving global landscape of adult congenital heart disease

Results

<table>
<thead>
<tr>
<th>Region</th>
<th>Number of ACHD centers per 10-million population</th>
</tr>
</thead>
<tbody>
<tr>
<td>Europe</td>
<td>3.6</td>
</tr>
<tr>
<td>North America</td>
<td>1.7</td>
</tr>
<tr>
<td>Oceania</td>
<td>1.5</td>
</tr>
<tr>
<td>South America</td>
<td>0.4</td>
</tr>
<tr>
<td>Asia</td>
<td>0.3</td>
</tr>
<tr>
<td>Africa</td>
<td>0.1</td>
</tr>
</tbody>
</table>

Kempny A et al.

International journal of cardiology 168(6) · July 2013
The Care of Adults with Congenital Heart Disease Across the Globe: Current Assessment and Future Perspective

Black dots identified ACHD centers.

Area of pink circle = proportion of center impact

Courtesy of Kempny A 2013

Possible explanations for differences between developed and developing countries in regard to ACHD

Apart from the obvious
1. Due to consanguinos marriage
2. Easiness of abortion
3. Yearly birth rate
Birth Rate - World

Average annual no births /year/1.000 persons

http://www.indexmundi.com/map/?v=25
Total Fertility Rate - World

Birth / woman

http://www.indexmundi.com/map/?v=31
Total Fertility Rate

Indicator for population change in country

Rate of 2 children / woman = replacement rate for population
Total Fertility Rate

Rates $<$ two children = populations decreasing in size and growing older

Rates $>$ two children = populations growing in size and median age declining
Population Pyramid in comparison USA / Saudi Arabia

2012 Population Pyramid for the United States

Data Source: U.S. Census Bureau

dshort.com
CHD burden by WHO region indexed to regional population by age

More children = < resources for CHD
What is 2020 of ACHA
Begun as initiative of ACHA in Dec 2008, the goal is to ensure that every CHD survivor be able to receive life long Adult Congenital Cardiac care.
What is the **Vision 2020**?

**Vision 2020** is: to make ACHD an established, recognized field that will deliver quality care to adults with CHD throughout the USA.
What does ACHA hope to achieve?

On turning 18, every complex CHD will have access to life-long ACHD care which is:

UTOPIA for many?

- UH Quality
- Age-appropriate
- Research-Based
- Coordinated and integrated
- Available in all regions
The data don’t predict a tsunami with absolute certainty. I still say we wait!!

Most of the World acts like this

already 2016

“The data don’t predict a tsunami with absolute certainty. I still say we wait!!”
Grown-up Congenital Heart Disease
Adult Congenital Heart disease

Why it remains a problem?
role of patient organisations;
a cardiologists’ view

Folkert J. Meijboom
Thoraxcentre, Rotterdam
The Netherlands
Why There is Shortage of ACHD Specialists?

- Very interesting, fascinating field
- Little evidence; a lot of research to be done
- More than enough patients
- Fast growing patient population
- Few cardiologists start their training with the idea of becoming a ACHD (GUCH) doctor
- ACHD is not a profitable subspecialty for the dept
- Often an academic career = less income for the Dr.
- No steering from Government/Health department
The Problem: Shortage of ACHD Specialists

- **Not** a problem for individual doctors
  - Work enough

- **Not** a problem for cardiology departments
  - ACHD is not a profitable subspecialty

- It is a problem for the **Dept Pediatric Cardiology**
  - Where to send their patients >18 yrs who need prolonged care

- **It is a problem for patients!**
The Problem is Bigger than just Cardiologists

- Governments?
- Health departments?
- Professional organisations of cardiologists?
- First step: awareness of this problem at those places where health care planning is done
- Who is going to take care of that?

In Poorer Countries?
The Doctor is defender of patient’s rights
Adult Cardiologist very important

Adult Cardiologist has a very important role in the management of Adults with Congenital Heart Disease because of high prevalence of ACHD and lack of enough centers dealing with these patients and because nobody else cares
What is the real problem [only?] in our area?
Experience from two failed ACHD Centers

Adult Cardiologist do not want to engage in this new specialty

They only want to close ASDs
They Need to Learn New Vocabulary

Atrial septal defect
Aortic Coarctation
Fontan, Mustard, Senning, Ross etc.
# Surgical Terminology in Adult Congenital Heart Disease

## Surgical Shunts in CHD

<table>
<thead>
<tr>
<th>Shunt</th>
<th>Procedure</th>
<th>Procedure</th>
<th>Surgery</th>
</tr>
</thead>
<tbody>
<tr>
<td>Classic Blalock-Taussig</td>
<td>Subclavian artery to pulmonary artery as end to side anastomosis.</td>
<td>Rastelli Procedure</td>
<td>Transposition with a VSD whereby the VSD is closed baffling LV to Ao and a RV to PA conduit is placed.</td>
</tr>
<tr>
<td>Modified Blalock-Taussig</td>
<td>Gore-Tex tube graft from subclavian artery to pulmonary artery.</td>
<td>Norwood Procedure</td>
<td>Hypoplastic LV syndrome. The pulmonary valve and artery are used to create neo-aorta and a BT shunt is placed to provide pulmonary blood flow.</td>
</tr>
<tr>
<td>Bidirectional Glenn</td>
<td>SVC to RPA. End to side. Second stage to fontan.</td>
<td>Mustard Procedure</td>
<td>Transposition of the great vessels. Baffle the SVC/IVC to the MV - LV -- PA and baffle the pulmonary veins to TV -- RV -- Ao.</td>
</tr>
<tr>
<td>Fontan Procedure</td>
<td>IVC to RPA to complete systemic venous circulation to PAs. Gore-Tex tube that may be in the heart (lateral tunnel) or outside the heart (extracardiac).</td>
<td>Arterial Switch</td>
<td>TGV where the aorta and MPA are switched and coronary arteries re-implanted.</td>
</tr>
<tr>
<td>Waterson</td>
<td>Ascending aorta to RPA. A punch hole between the vessels.</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Potts</td>
<td>Descending aorta to LPA. Punch hole between the vessels.</td>
<td></td>
<td></td>
</tr>
</tbody>
</table>
Examples for Problems between Pediatric Cardiologists and Adult Cardiologists
Case Presentation

21 years old lady, 117 kg, comes in a wheel chair to clinic

**History**: Primum ASD closed surgically at the age of 18 years

**Following Problems:**

Adipositas per Magna

Wheel chair bound

Atrial flutter

Systemic hypertension

Insulin dependent Diabetes mellitus

**Treating Physician**: Pediatric Cardiologist
Solutions ?
Who should we warn about the ACHD Tsunami?

All of us

Politicians, Health Care Giver
Pediatric Cardiologists
Adult Cardiologists
Cardiac Surgeon
ICUs
Causes of Tsunami?

In our case?  
Man made as we are not patient centered.
We see the Problem but
We continue doing this
Short-Term (to solve patients problems)

- Pediatric Cardiologist should treat ACHD
- Consult Adult Cardiologists for help
  (BP, Arrhythmias, Diabetes etc.)
Mid-Term Solutions

Encourage Fellows **Pediatric Cardiologist** and/or **Adult Cardiologist** to specialize in ACHD

Address issues of ACHD with **Health care giver**

**MOH / Hospital Administrators etc.**
Long-Term Solutions

Obtain better Data to understand the magnitude of the problem.

Registries are needed to at least first establish prevalence of CHD in most countries.