The modified natural history of congenital heart disease

Matthias Greutmann, MD
Adult Congenital Heart Disease Program
University Hospital Zurich, Switzerland
matthias.greutmann@usz.ch
Are we ready for the future?

Matthias Greutmann, MD
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Objectives

- Natural history of complex CHD
- Effect of surgical modification
- Current situation
- Outlook and needs for the future
Survival

Chance of survival to adulthood with complex CHD

Modified from Dr. H. Kaemmerer, Munich
Norwood

Gross
Crafoord
Taussig
Blalock
Lillehei

Senning
Mustard
Fontan
Jatene
Norwood
Transposition of the great arteries
Natural history of TGA

Survival (%) vs. Age (years)

- PFO only
- ASD
- VSD + PS

Liebman, Circ 1969
Improved Survival

Modified from Dr. H. Kaemmerer, Munich
Number of patients with CHD

Number of patients


Marelli, Circ 2007
Shift to Adults: complex CHD

Number of patients

- children
- adults


Marelli, Circ 2007
Prevalence

- All congenital heart defects: 4.1 / 1000 adults
- Complex defects: 0.38 / 1000 adults

Marelli, Circ 2007
Number of adults with CHD

Switzerland 7.8 Mio
32’000
3000

Europe 730 Mio
3’000’000
280000
Mortality Shift

Age at death (years)

1987-88

2004-05

Proportion of all deaths

CHD deaths

General population

Khairy, JACC 2010
Mortality Shift

1987-88 vs 2004-05

Age at death (years)

Proportion of all deaths

CHD deaths

General population

Khairy, JACC 2010
Who are these patients?
Case 1: Female, 23 years old

Left atrial isomerism

- single right ventricle, common atrium, severe pulmonary stenosis
- Palliated with left modified BT-shunt
Palliated cyanotic CHD

![Diagram showing a single atrium and RV with hypoplastic LV.](image)

- Single atrium
- RV
- Hypoplastic LV

![Diagram of the heart showing the pulmonary artery and aortic branch.](image)
Case 2: Male, 45 years old

Tricuspid atresia
- Classic Glenn shunt
- Fontan operation
- Now: ‘Failing Fontan’
Fontan procedure

- single LV
- LA
- RA
- tricuspid atresia
- ‘giant’
Case 3: male, 23 year old

Transposition of the great arteries

Atrial switch

PV-baffle
end-stage heart failure
What is the outcome of adults with complex CHD?
506 deaths among 12,499 patients

- 32% SCD
- 34% Cardiac - not sudden
- 13% Perioperative
- 18% Noncardiac
- 3% Unknown

AHA abstract, 2010
Deaths per decade

- 1980-89: 68 deaths
- 1990-99: 187 deaths
- 2000-09: 230 deaths
Deaths vs. total patients

Year | Total patients | Deaths
---|----------------|------
1980-89 | 100 | 40
1990-99 | 200 | 300
2000-09 | 300 | 700
Disease Complexity

<table>
<thead>
<tr>
<th>Year</th>
<th>Simple</th>
<th>Moderate</th>
<th>Severe</th>
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<td>50</td>
<td>150</td>
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<tr>
<td>1990-99</td>
<td>100</td>
<td>100</td>
<td>50</td>
</tr>
<tr>
<td>2000-09</td>
<td>200</td>
<td>100</td>
<td>50</td>
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</table>
Mode of death

<table>
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<th>Simple</th>
<th>Moderate</th>
<th>Severe</th>
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<tbody>
<tr>
<td>Non-Cardiac</td>
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<tr>
<td>Cardiac</td>
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</tbody>
</table>

$p < 0.001$
Changes in age of death

- 1980-89: $p = 0.7$
- 1990-99: $p = 0.01$
- 2000-09: $p = 0.002$

Age of death

- Simple
- Moderate
- Severe
Most common entities

- Fontan palliation (9%)
- Tetralogy of Fallot (11%)
- Subaortic right ventricles (13%)
- Cyanotic defects / Eisenmenger (29%)

13% of all patients but 62% of all deaths
Active & alive Patients

Tetralogy of Fallot

Number of pts


TGA
Fontan
Cyanotics
Conclusions I

- There is an emerging population of young adult survivors with complex CHD
- These patients are at high risk of premature death at young age
Outlook and needs

- Transplantation
- Communication
Is transplantation the solution?
Number of adults with CHD

Switzerland 7.8 Mio
32’000
3000

Europe 730 Mio
3’000’000
2800000
506 deaths among 12,499 patients

- SCD: 34%
- Cardiac - not sudden: 18%
- Perioperative: 13%
- Noncardiac: 3%
- Unknown: 32%

AHA abstract, 2010
No. of Transplantations

Year of Transplant

- Listings
- Transplants

Davies, Circ 2011
Reasons not to transplant CHD
Outcome after Transplantation

- Non-CHD
- CHD
- Fontan

Davies, Circ 2011
Lamour, JACC 2009
Communication

- Do we communicate enough?
What do patients think

Self-estimated life expectancy

- Students
- CHD – mod.
- CHD - complex

Reid, JACC 2006
Communication

- 200 Adults with CHD
  - 35 ± 15 years, 80% mod. or great disease complexity

- 48 Health care provider

Tobler et al, IJC 2010
Have you ever discussed EOL planning with a member of your medical team?

- Yes: 99%
- No: 1%
Patients

Do you think a member of your medical team should bring up EOL discussions?

- No: 23%
- Yes: 77%
How often do you typically discuss EOL issues with outpatients?

- Simple: 13%
- Moderate: 49%
- Great: 89%

*p < 0.001 compared to patient group*
Patients

Do you think a member of your medical team should bring up EOL discussions?

- Simple: 77%
- Moderate: 79%
- Great: 77%
When do you think is the **best time** to bring up EOL discussions with patients?

- **Patients**
  - Before getting sick, while healthy
  - First life-threatening illness or complication
  - First admitted with life-threatening complication or dying

- **Provider**
  - Before getting sick, while healthy
  - First admitted with life-threatening complication or dying

*p < 0.001*
Are we ready for the future?

- Novel therapeutic approaches
- Better prognostication
- Better communication
Thank you!

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