Outline

- History
- The adult population
- Do we cure CHD patients?
- Summary
1938
1st Operation in CHD

R.E. Gross
Boston, USA
1944 - Operation Aortic Coarctation

C. Crafoord
Stockholm, SE
Tetralogy of Fallot

Helen B. Taussig (1898-1986)
Baltimore, USA
1944

Helen B. Taussig (1898-1986)
Baltimore, USA

A. Blalock (1899-1964)
Baltimore, USA
Tetralogy of Fallot

Dr. A. Blalock and his team

Blalock-Taussig Shunt
1st Heart Lung Machine in 1953

- Open Heart Surgery
- Intracardiac Repair

John H. Gibbon Jr. 1903 - 1973
C.W. Lillehei, Minneapolis, USA: First Tetralogy of Fallot Repair

August 31, 1954

Cross-Circulation

Courtesy of Dr. W. Williams, Toronto
C.W. Lillehei, Minneapolis, USA: First Tetralogy of Fallot Repair

- VSD patch closure
- RVOT reconstruction
  - Transannular patch (fig.)
  - Valvotomy, infundibulectomy, RV patch enlargement
- RV-PA conduit
Åke Senning
Zürich, CH

William T. Mustard
Toronto, Ca
Åke Senning
Zürich, CH

William T. Mustard
Toronto, Ca
1975 - ARTERIAL Switch Procedure

F. Fontan
Tricuspid Atresia with ASD and VSD

Fontan Procedure

- Not suitable for biventricular repair
- Separation of the systemic and pulmonary circulation

Potts Anastomosis
1971: Fontan - Circulation
Toronto CHD History

- Hospital for Sick Children
  - Keith, Rowe, Freedom, Redington
  - Mustard, Trusler, Williams, Van Arsdell
- ACHD clinic opened 1959 at Toronto General Hospital
Survival in Tetralogy of Fallot by decade of birth at HSC, Toronto

Hickey EJ, Manlhiot C, McCrindle BW 2007
Heart and Stroke Foundation of Ontario “Functional outcomes in Tetralogy of Fallot
Survival of Complete TGA

Arterial Switch: 2000’s

Atrial Switch: 1980’s

Natural History: 1950’s

Survival (% Year)

Years

0  4  8  12  16  20

0  20  40  60  80  100

Courtesy by Dr. W. Williams
Survival Rate in CHD and Milestones in Cardiac Surgery

%  
100 80 60 40 20 0  
20 25 40 55 70 80 85  

BT-Shunt  Fallot-OP  Atrial Switch-OP  Fontan-OP  Arterial Switch-OP

Courtesy of Dr. H. Kaemmerer, Munich
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## Prevalence of Severe and Other CHD

<table>
<thead>
<tr>
<th></th>
<th>Adults Alive in 2000</th>
<th></th>
<th>Children Alive in 2000</th>
<th></th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>n (%)</td>
<td>Prevalence per 1000 Adults</td>
<td>Prevalence per 1000 Children</td>
<td></td>
</tr>
<tr>
<td>All congenital heart lesions*</td>
<td>23,563 (100)</td>
<td>4.09</td>
<td>18,979 (100)</td>
<td>11.89</td>
</tr>
<tr>
<td>Severe lesions</td>
<td></td>
<td></td>
<td></td>
<td></td>
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<tr>
<td>TOF or truncus arteriosus</td>
<td>1,001</td>
<td>0.17</td>
<td>778</td>
<td>0.49</td>
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<tr>
<td>AVCD</td>
<td>834</td>
<td>0.14</td>
<td>914</td>
<td>0.57</td>
</tr>
<tr>
<td>Transposition complex</td>
<td>235</td>
<td>0.04</td>
<td>424</td>
<td>0.27</td>
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<tr>
<td>Univentricular hearts</td>
<td>150</td>
<td>0.03</td>
<td>213</td>
<td>0.13</td>
</tr>
<tr>
<td>All severe lesions</td>
<td>2,205 (9)</td>
<td>0.38</td>
<td>2,316 (12)</td>
<td>1.45</td>
</tr>
</tbody>
</table>

Changing Age Distribution of Severe CHD 1985 - 2000

Extrapolation: Quebec Data

Prevalence of Adults with CHD
(4.09 per 1000)
~ 96,000

Annual Increase ~ 1,000 Adults per Year

Prevalence of severe CHD in Adults
(0.38 per 1000)
~ 9,000
CHD Patients in Ontario

Medium / High Risk for Complications: 50%

31,000
Children/Adolescents

38,000
Adults (>18 yrs)
Workload in Outpatient Clinic by Year

SARS closed the clinic in 2003 for several months
Age Distribution

n = 4168
Heterogenous Population

- **Diagnostic groups**
  - Shunt lesions
  - Transposition complexes, Ebstein
  - RVOTO / LVOTO, etc.
  - Pulmonary hypertension

- **Surgical procedures**
  - None
  - Palliative shunts
  - Repair
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‘The surgeon fixed my heart: 
I am cured’
Do we really correct congenital heart defects?

J. Stark, FRCS, FACS, FACC, London, England
Surgery is corrective, if……

- Ventricular function is normal
- Life expectancy is normal!
- There is no need for therapeutic measures during follow-up!
Corrective Surgery....

- Atrial septal defect
- Ventricular septal defect
- Patent ductus arteriosus

... if treated during childhood!!
Long-Term Survival after ASD Closure by Age

Emergencies

- Cardiovascular: 83%
- Infective: 6%
- Others: 11%

Kaemmerer H, Oechslin E, ... Hess J. J Thorac Cardiovasc Surg 2003; 126:1048-52
Cardiovascular Emergencies

- Arrhythmias: 53%
- Heart failure: 19%
- Cerebral ischemia: 7%
- Other cardiovascular: 6%
- Syncope: 2%
- Infections: 2%
- Others: 2%

Kaemmerer H, Oechslin E, ... Hess J. J Thorac Cardiovasc Surg 2003; 126:1048-52
Psychological Injury
Long – Term Challenges

- Arrhythmias
  - Supraventricular / ventricular

- Reoperations
  - Failure of conduits / artificial valves
  - Residual shunt / regurgitation / obstruction

- Heart failure
  - Medical therapy / heart / (lung) transplantation

- Biomedical and psychosocial concerns / barriers
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Summary

- The **demographics** of CHD population have changed:
  - Increasing number of adults with CHD
- **Distribution** of CHD:
  - Prevalence of pts with **severe CHD** is increasing more rapidly in adults than in children
  - Equalizing numbers of adults and children with severe CHD
Summary

- Mortality and morbidity are shifting away from the young and towards the adult
- **Workload** is increasing

Impact on Health Care System!
Congenital Heart Disease

- Heterogeneous population – the patient profile is changing
- Most pts are not ‘fixed’
  - Life-long risk for complications
- Expert care with knowledge and expertise in:
  - Anatomy and physiology
  - Long-term outcome
- Multi-disciplinary team approach in designated centres
Designated Centre for Pt Care

- Cardiology
- Congenital / Thoracic Surgery
- Anesthesia
- Transplantation
- Heart Failure
- Immunology
- Intensive Care
- Respirology
- Pulmonary HT
- Imaging
- Genetics
- Rheumatology
- Psychology
Patients with named conditions or operations are complex

Specialized centre
CHD is a Continuum from Fetal Life until Adulthood
ACHD TSUNAMI:
*Catch The WAVE!*

- Arrhythmias
- Heart Failure
- Pulmonary Hypertension
- Re-Intervention
- Psychosocial Issues
- Reproduction
- Premature Death