Adult Congenital Heart Disease

They’re out there and coming to find you....

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Green Lane Paediatric and Congenital Cardiac Service
Congenital Heart Disease in New Zealand

4.5 million population

• Single Congenital Cardiac Surgical Centre
• Pioneering Congenital Cardiac Surgery from 1940s
• Approx 1:100 kids born with a congenital heart defect
• Now 60 years+ of CHD surgical treatment
Evolution of CHD surgery

- 1938  PDA ligation
- 1944-45  BT shunt, Coarctation repair
- 1954  Bypass
- 1959  Senning
- 1963  Mustard
- 1970s  Infants, Fontan
Hence

- Better diagnostic tools
- New procedures
- Better survival

Increasing Numbers!
The Adult Population with CHD

Paediatric

Adult

1980

Courtesy Tim Hornung
The Adult Population with CHD

Paediatric | Adult

2000

800,000 adults with CHD in the US,
250,000 in the UK, ? 17,500 in NZ
The Adult Population with CHD

Paediatric 2020 Adult
Congenital Heart Disease in the General Population
Changing Prevalence and Age Distribution

Ariane J. Marelli, MD; Andrew S. Mackie, MD, SM; Raluca Ionescu-Ittu, MSc;
Elham Rahme, PhD; Louise Pilote, MD, MPH, PhD

**B Severe CHD**

<table>
<thead>
<tr>
<th>Year</th>
<th>Adults</th>
<th>Children</th>
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<tbody>
<tr>
<td>1985</td>
<td>35</td>
<td>65</td>
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<tr>
<td>1990</td>
<td>36</td>
<td>64</td>
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<td>1995</td>
<td>40</td>
<td>60</td>
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<tr>
<td>2000</td>
<td>49</td>
<td>51</td>
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</table>

Number alive

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Circulation    January 16, 2007
1% birth prevalence CHD

- VSD 35%
- ASD 10%
- PDA 7%
- Pulmonary stenosis 7%
- Aortic stenosis 4%
- Coarctation 4%
- Tetralogy 4%

All cyanotic 14%
Some examples
Tetralogy of Fallot

To Body

Aorta

VSD

Left Ventricle

Right Ventricle
Tetralogy of Fallot
Tetralogy of Fallot

Late problems:

– Chronic pulmonary regurgitation
– RV dilatation and dysfunction
– Ventricular (and atrial) arrhythmias
– Impaired functional capacity
Tetralogy of Fallot
Mode of Late Death in Repaired Tetralogy

33 pts died, 4.2% mortality

- SCD: 49%
- CHF: 27%
- Re-operation: 3%
- CAD: 6%
- Non-Cardiac: 15%

None of the 16 SCD cases underwent redo surgery!
Gatzoulis - Tetralogy Multi-Centre Study

N=793 adult patients

- VT
- SD
- A Flutter
- Arrhythmia-free

% patients

RVSP (>60mmHg) (≥moderate)

TR (≥moderate)

PR (≥moderate)
Question: When to replace the pulmonary valve?

1. Symptoms
2. Important RV dilation (MRI > 150ml/m²)
3. RV dysfunction (and LV dysfunction)
3. Ventricular arrhythmias
Transcatheter pulmonary valve

Melody Transcatheter pulmonary valve (Edwards Sapien)

Short to medium term follow up
Transposition of the Great Arteries
Earlier repairs

Mustard and Senning repairs

• Patients now in mid 20s to 50

Problems

– Baffle obstruction
– Systemic Right ventricular dysfunction/failure
– Arrhythmia
– Sudden death
Issues for follow up

• Arrhythmia
  – Progressive loss of sinus rhythm
  – Bradycardia, junctional rhythm
  – Atrial arrhythmia
  – Tachy/Brady

• Baffle obstruction

• Systemic ventricular dysfunction
Arterial Switch

• Surgical ‘cure’

• Follow up
  – Narrowing of branch pulmonary arteries/main pulmonary artery
  – Can see leaks of outlet valves
  – Most do very well
  – ? Long term outcomes (eg coronary)
Coarctation of the Aorta

- Murmur heard posteriorly
- Diminished femoral pulses
- Hypertension
- Association with bicuspid aortic valve
Need careful evaluation

Bicuspid aortic valve
- Aortic stenosis
- Aortic regurgitation
- Dilation ascending aorta

Arch anatomy
- Arch morphology
- Arch hypoplasia
Intervention
Arch repair
Coarctation repair
Stenting

Late issues
Recoarctation
Aneurysms
Aortic dilation
Hypertension
Late follow up

Results After Repair of Coarctation of the Aorta Beyond Infancy: A 10 to 28 Year Follow-Up With Particular Reference to Late Systemic Hypertension

Twenty-four-hour ambulatory blood pressure monitoring detects a high prevalence of hypertension late after coarctation repair in patients with hypoplastic arches

Angular (Gothic) aortic arch leads to enhanced systolic wave reflection, central aortic stiffness, and increased left ventricular mass late after aortic coarctation repair: Evaluation with magnetic resonance flow mapping

The Journal of Thoracic and Cardiovascular Surgery • November 2012

The Journal of Thoracic and Cardiovascular Surgery • January 2008
Fontan circulation
Extra-Cardiac Conduit Modification
Fontan circulation

Late problems

- Ventricular dysfunction
- Arrhythmias
- Clots – stroke
- Chronic Liver congestion and cirrhosis
- Others
General points

ACHD is a **multisystem disorder**

- ? Other congenital anomalies, chromosomal
- Cyanosis, bypass, periods of low output, chest deformity, abnormal pulmonary blood supply, chronic ill health, parental and other expectations, exercise restriction, transfusion, multiple caths, cutdowns, hepatic congestion etc....

• Consider: health of kidneys, liver, brain, blood, lungs, spirits ...and the heart
Outcomes assessment

- Survival?
- Quality of life?
- Physical Functioning?
- Neuro developmental Outcome?
- Need for Reoperation?
Exercise

Fredriksen 2001 AJC

![Graph showing VO2max (ml kg^-1 min^-1) vs Age at test for different conditions.](image)

<table>
<thead>
<tr>
<th>Condition</th>
<th>18-19 years</th>
<th>20-29 years</th>
<th>30-39 years</th>
<th>40-49 years</th>
<th>50-59 years</th>
<th>60-69 years</th>
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<td>27</td>
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<td>16</td>
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<td>15</td>
<td>8</td>
<td>4</td>
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<td>Ebstein</td>
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<td>11</td>
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<td>5</td>
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<td>Fontan</td>
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<td>30</td>
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<td>Mustard</td>
<td>16</td>
<td>57</td>
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<td>ToF</td>
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<td>63</td>
<td>50</td>
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<td>Increasing Static Component</td>
<td>Increasing Dynamic Component</td>
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<tr>
<td>I. Low (≤20% MVC)</td>
<td>A. Low (≤40% Max O₂)</td>
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<tr>
<td>Archery, Auto racing†</td>
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<td>Diving†, Equestrian†</td>
<td>Table tennis, Volleyball</td>
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<td>Motorcycling†</td>
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<td>II. Moderate (20-50% MVC)</td>
<td>B. Moderate (40-70% Max O₂)</td>
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<td>Curling, Golf, Riffery</td>
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<td>Race walking, Racquetball/Squash, Running (long distance), Soccer*, Tennis</td>
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<tr>
<td>III. High (&gt;50% MVC)</td>
<td>C. High (≥70% Max O₂)</td>
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<td>Bobsledding/Luge*, Field</td>
<td>Boxing*, Canoeing/Kayaking,</td>
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<td>events (throwing), Gymnastics*†, Martial arts*, Sailing, Sport climbing, Water skiing**, Weight lifting**, Windsurfing**†</td>
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<tr>
<td>Downhill skiing†, Skateboarding†, Snowboarding†, Wrestling*</td>
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<tr>
<td>Boxing*, Canoeing/Kayaking,</td>
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<tr>
<td>Cycling†, Decathlon, Rowing, Speed-skating†, Triathlon†</td>
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* Indicates moderate intensity.
† Indicates high intensity.
Informed Encouragement to exercise and participate
Endocarditis

- Still exists!
- Focus on good dental care and dental hygiene and awareness of symptoms
- Prophylaxis in selected cases
Pregnancy

Issues
Inherited heart disease
Conception/Contraception
Physiologic challenges of pregnancy
  CV, thrombosis, arrhythmia, etc
Supervision during pregnancy
Mode and place of delivery
Care in post partum period
Longevity and Survival
Highest risk

- LV obstructive lesions
- Pulmonary hypertension
- Severe LV dysfunction
- Aortopathy/aortic dilation
- Prosthetic valves
Learning and Cognition
Neurodevelopmental outcomes

Increasing interest 'Hearts and Minds'

• Improvement in bypass techniques
• Additional contributors
  – Genetic / Congenital Brain abnormalities
  – Hypoperfusion and acidosis
  – Prematurity
  – Environmental
  – Anticoagulation strategies – embolic events
  – Additional periprocedural insults – low cardiac output
MRI pre and post bypass
Mahle et al, Philadelphia, Circulation 2002

Full term neonates with CHD
• Sept 1 2000- Jan 31 2001
• Excl genetic abnormality, birth asphyxia, preop cardiac arrest, post op mechanical support, reop requiring bypass
• Enrolled 24/36 eligible
• Serial MRI preop, early post, late post
Neurodevelopmental assessment of school aged and adolescent children with hypoplastic left heart

Mahle et al, 2000 Pediatrics

• QoL, school performance, incidence of medical complications
• 115/138 eligible children mean age 9yrs
• Parent/Guardian report health as ‘good’ 34%, ‘excellent’ 45%, 84% school performance average or above average
• 1/3 receiving some form of special education
• Standardised testing in 28, median full scale IQ 86 (50-116), IQ < 70 in 18%
Psychological

- Anxiety and Depression – increased frequency in ACHD population
- Multiple additional life challenges
  - Family and parenting style
  - Illness in childhood
  - Physical and Intellectual challenges
  - Body image
  - Uncertainties re future health and survival
Quality of Life
Adult CHD survivors
Kamphuis et al, Leiden  Heart 2002

78 pts with previously operated complex congenital heart disease (now aged 18-32)
• 91% participation
• 5% with learning disabilities ineligible
• Specifically developed questionnaire
• Generic tool (SF-36)
• Physical indexes
Adult CHD Kamphuis et al

Descriptors

– 46% had a job for at least 12h/wk
– 15 receiving full/partial disability
– 11 still at school
– 43% lived with a partner, 40% with parents

• Health related quality of life significantly lower than general population
  – Gross motor functioning
  – Vitality

• Objectively measured medical variables correlated only weakly to QoL
QoL adults CHD
Lane et al, Birmingham, Heart 2002

Sent SF-36 to 471 ACHD pts

• Completed by 58.5%, median age 31, range 16-85
• Pts deemed ‘cured’ had lower QoL than general population
• Inoperable pts lower than all others
• Cyanotic pts worse than acyanotic
Maintaining follow up
Pattern of care

Paediatric → Adult

Transition

Loss to follow up
Moving house, Moving to train or study, New job, New relationship, missed appt -> d/c

RePresent
Injury, Illness, Pregnancy, New partner
Maintaining follow up

Need to capture the interest of the patients and maintain contact
Promote understanding and appropriate self management
Develop expectations of the ACHD service and the Health service
Provider awareness

“Coach on the sideline”
More on loss to follow

- Transition process
- Understanding need for follow up
- Denial
- Hospital systems
- Socioeconomic
- Provider education and awareness
Major challenges

- Geography (and impact on funding)
- Prioritising ACHD
- Loss to follow up
- Cognitive and Social Impacts of ACHD
Paediatric Cardiology

Approx 110 Visiting Clinics per year
**ACHD**

Approx 18 Visiting Clinics per Year

Some areas with coordinated local ACHD clinic eg Tauranga Graeme Porter, John Lainchbury Chch, Andrew Aitken Wellington, TV Liew Hamilton

Transition Clinic Christchurch
Auckland ACHD team

Cardiac Nursing Staff
Ward 31
Ward 42
CVICU
PICU

High risk obstetric team
Obstetricians/Physicians

Surgeons: John Artrip,
Kirsten Finucane

Ivor Gerber       Tim Hornung
Boris Lowe       Clare O'Donnell
Annette Rief NP   Registrar

EP/Pacing
Jon Skinner
Fiona Riddell

Cardiac Radiology
Chris Occleshaw

Clinical Psychologist
Gudrun Court

Cath Lab

Family Planning
Helen Roberts

Congenital echo service

Cardiac Intensivists
Cardiac Anesthetists
Theatre staff

John Beca
Mark Edwards
David Buckley

Starship
Children's Hospital of New Zealand
Guidelines and resources

So...

Increasing population of adolescents and adults
Increasing expectations for children and families
Need for ongoing follow up
May have complex health needs
Thank you