Epidemics of adults with single ventricles: are we ready?

Yes

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Melbourne Australia
Disclosures

- Consultancy fees from MSD, Actelion
Adults with single ventricle = adults with Fontan

- Equilibrated Pts with single Ventricle will not need anything
- Failed BCPS will almost all be dead by adulthood
I had a Fontan...aarrggr

The looming plague
We are not ready

- Increasing population
- We do not have the infrastructure of care
- We do not know how to treat them
- We do not have donor hearts for transplantation
ANZ Fontan Registry

- 1,425 participant records

Number of Patients Alive with a Fontan Circulation

ECC = 851
LT = 264
AP = 157
Survival curve in 2014

Kaplan-Meier survival estimates

Number at risk

| fontan_type = AP | 208 | 197 | 183 | 173 | 166 | 135 | 34 | 4 |
| fontan_type = LT | 283 | 269 | 261 | 232 | 158 | 17  | 1  | 1 |
| fontan_type = ECC| 869 | 550 | 292 | 79  | 1   | 1   | 0  | 0 |
Projection of Fontan population in Australia and New Zealand (26 $10^6$ inhab)
We do not know how to treat them

- Medications
- Follow-up care
- Reinterventions
Let us sort out the medications !!!

$A\ 2,677,680.76\ \text{NHMRC partnership grant with National Heart Foundation, Heartkids}\
\text{MCRI leading institution and main partner funder}
No benefit of warfarin over aspirin after the extracardiac Fontan in a propensity score analysis of 478 patients.

- Ajay J Iyengar MBBS BMedSci 1,2,3, David S Winlaw MBBS MD FRACS 4,5, John C Galati BSc PhD 2,6, Gavin R Wheaton MBBS FRACP 7, Thomas L Gentles MBChB FRACP 8, Leeanne E Grigg MBBS FRACP 9, Robert N Justo MBBS FRACP 10, Dorothy J Radford MBBS MD FRACP 11, Chantal Attard BSc 12, Robert G Weintraub MBBS FRACP 2,3,13, Andrew Bullock MBBS FRACP 14, Gary S Sholler MBBS FRACP 4,5, David S Celermajer MBBS MSc PhD DSc FAHA FRACP FAA 5,15, Yves d’Udekem MD PhD FRACS 1,2,3, The Australia and New Zealand Fontan Registry
Use of ACE Inhibitors in Fontan: Rational or Irrational?

- 462/1268 (36%) of the Fontans of the Registry on ACE inhib
- Relatively strong indication in a third
  (> mild ventr. Dysfunction. > mild AV valve Regurg, semi-lunar valve Regurg, HTA)
- 27% of the treatment initiated in hospital and pursued thereafter.
Resistance training improves cardiac output, exercise capacity and tolerance to positive airway pressure in Fontan physiology

Rachael L. Cordina a,b, Shamus O'Meagher a,b, Alia Karmali a, Caroline L. Rae c,d, Carsten Liess e, Graham J. Kemp f, Raj Puranik a,b, Nalin Singh g,h, David S. Celermajer a,b,⁎
Should We Recommend Exercise after the Fontan Procedure?

Nigel Sutherland, BPhysio\textsuperscript{a}, Bryn Jones, MBBS, FRACP\textsuperscript{b,c,d}, Yves d’Udekkem, MD PhD FRACS\textsuperscript{a,b,c}\textsuperscript{a}

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\textsuperscript{b}Murdoch Childrens Research Institute, Melbourne, Vic, Australia
\textsuperscript{c}Department of Paediatrics, Faculty of Medicine, The University of Melbourne, Melbourne, Vic, Australia
\textsuperscript{d}Department of Cardiology, The Royal Children’s Hospital, Melbourne, Vic, Australia
Home and hospital-based exercise training program for Fontans
Sympathetic and vascular dysfunction in adult patients with Fontan circulation

Elisabeth Lambert a,e,*,1, Yves d’Udekel f,g,1, Michael Cheung g,h, Carolina Ika Sari a, Julia Inman a, Anna Ahimastos c, Nina Eikelis a, Atul Pathak j, Ingrid King f,g, Leanne Grigg i, Markus Schlaich b,d, Gavin Lambert a,d

Clinical microneurography

ECG

BP

MSNA

In press; Int J of Cardiol
How to treat them

• One aspirin a day and

..... 40 minutes exercise 3 times a week !!
Follow-up investigations

• Once / 2 years
  – Oxygen sat
  – ECG
  – Echo
  – Liver Function tests

• Before transition at 12-14 years
  – MRI
  – CPX testing
  – Holter
What causes failure?

• ? Progressive raise in the pulmonary vascular resistances
• ? Progressive increase in diastolic dysfunction
• Progressive Atrial dilatation

• …Or did we mess it up?
Causes and mechanisms of death

• Lateral tunnel and extra-cardiac Fontan
  – Atrio-pulmonary connection
  – Av valve regurgitation
  – Small LPA / One lung Fontan
  – Pacemaker !!
  – Residual arch obstruction
  – RV dependent coronary circulation

• Atrio-pulmonary connection ….
Determinants of Atrial Arrhythmia Late After the Atriopulmonary Fontan Operation

Nikos Soukias¹, Tim S. Hornung¹, Philip J. Kilner², Alexandra Frogoudaki¹, Periklis Davlouros¹,², Tom Wong¹, Michael A. Gatzoulis¹

¹Adult Congenital Heart Program and ²Cardiovascular Magnetic Resonance Unit, Royal Brompton Hospital and National Heart & Lung Institute, Imperial College London, UK.
Yield of investigations

- ECG
- Echo
- Liver function tests
- Holter

⇒ Hopeless
Fontan smartphone App

- Personal history
  - Morphology
  - Operations
  - Latest investigations results
- Agenda
  - Appointments
  - Medications/prescriptions
- Useful links/Social media
- Bilateral connection to the Registry
My way for follow-up

• Ongoing monitoring of Sat and Rhythm on Fontan app
• 5 yearly MRI / CPx study

• Closer monitoring of
  – AV valve regurgitation
  – Decreased Ventricular Function

• Stroke/TE events will go to the Emergency department like everybody else
Fontan conversion
Long-term outcomes: 29 Conversions

Freedom from death and/or transplantation (%)

- Early Conversion Centre: 86%
- Other Centres: 51%

p=0.007

# at Risk (# Fail)

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Heart transplantation: a solution?
ANZ Fontan transplantation: 34 pts

- **Fontan**: 91±9.6% at 1 year, 78±16% at 5 years, 71±20% at 10 years
- **Other univentricular**: 85±13% at 1 year, 70±18% at 5 years, 59±20% at 10 years
- **Biventricular**: 90±7.6% at 1 year, 88±8.4% at 5 years, 78±12% at 10 years

Survival rates are shown with statistical significance for different time points.
NUMBER OF ALL TRANSPLANT TYPES BY YEAR 1984 - 2013

HEARTS  HEART-LUNG  SINGLE LUNG  B-L LUNG

ANZCOR 2013
Comparison of risk factors and outcomes for pediatric patients listed for heart transplantation after bidirectional Glenn and after Fontan: An analysis from the Pediatric Heart Transplant Study

- 36 US centers
- 1993 – 2008
- 269 Fontans listed
Starship Children’s Hospital, Greenlane Cardiothoracic Centre Auckland, NZ
Figure 2: Cumulative proportion of heart transplantation in 2 catchment areas for 1369 patients with Fontan circulation in Australia and New Zealand.

199x112mm (300 x 300 DPI)
Follow-up care in ANZ

- Half of the patients are followed by cardiologists who are seeing less than 10 patients with a Fontan circulation.
The ultimate solution: VAD??
“... heart transplantation (HTx) is still regarded as the gold standard in the surgical treatment of terminal heart failure. Whether this is still justified in the year 2012 in Europe, and especially in Germany, needs to be seriously questioned.”

- 22 % mortality at 1 year for heart transplantation due to more liberal use of donor
- 20 -30 % mortality on the waiting list.
HVAD® System Demonstrates Long-Term Survival

HeartWare is first to present multicenter long-term data (>2 years) in a peer-reviewed forum.

Kaplan Meier Continued Survival in Patients Supported on HVAD >2 years

- Survival at: 3 years = 89%
- 4 years = 77%

The HVAD System successfully supported 74 of 382 patients in the BTT & CAP study for more than two years.

- Patients on support for >2 years demonstrate excellent quality of life outcomes and stable adverse event profiles.
- The world’s longest-supported HVAD patient has been on support for over 7 years and counting.

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1. Aaronson K et al. Patients awaiting Heart Transplantation on HeartWare Ventricular Assist Device Support for Greater than Two Years. AHA Poster, 2014.
3. Data on file with manufacturer, HeartWare, Inc, Framingham, MA.
When should we intervene?
Let us get ready:
Fontan Think Tank Bordeaux 2016

Brainstorming on 4 themes

• End-organ damages
• Risk stratification for death
• Prevention of failure
• Interventions for failure
We are not ready

- Increasing population
- We do not have the infrastructure of care
- We do not know how to treat them
- We do not have donor hearts for transplantation
The solution

• Let us keep doing what we do today
The solution

• Let us keep doing what we do today

...... Nothing

• Centralize care for failing Fontans
• And find an artificial heart

• We are ready