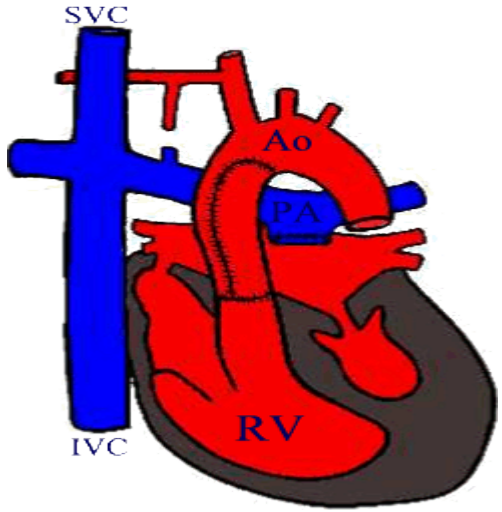
ANZ Fontan registry

Tom Gentles
Paediatric Cardiologist
Green Lane Paediatric and Congenital Cardiac Service
Starship Children's Hospital

What is a Fontan Circulation?

- Systemic venous return bypasses the heart and goes direct to the lungs
- There is no subpulmonary ventricle



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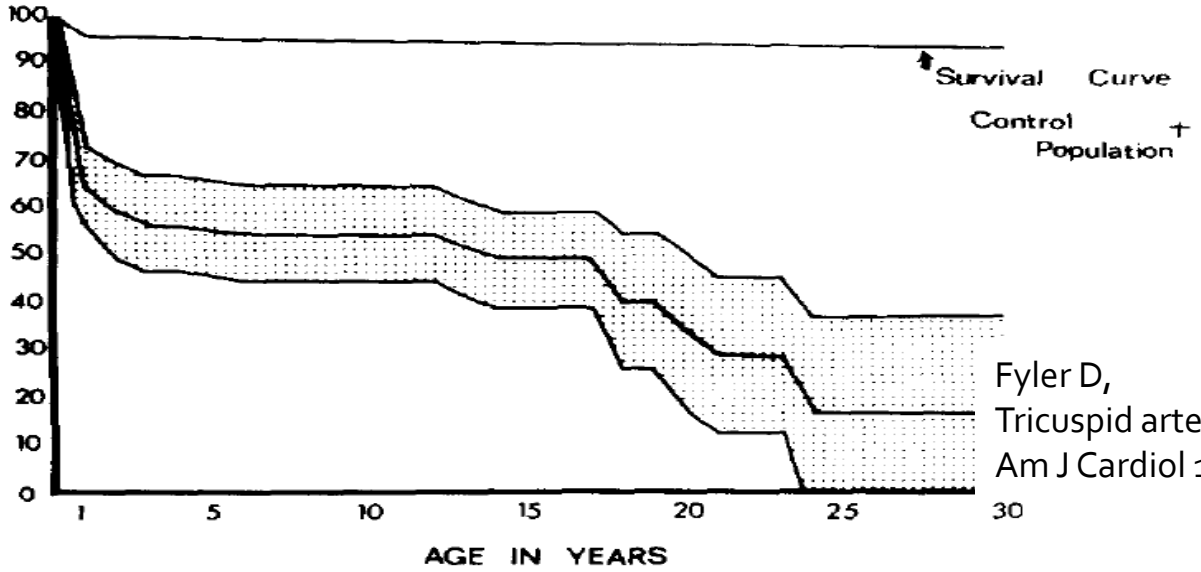
What is a Fontan Circulation?

- Response to a very poor outcome



Fyler D,
Tricuspid artesia clincal course in 101 patients
Am J Cardiol 1975

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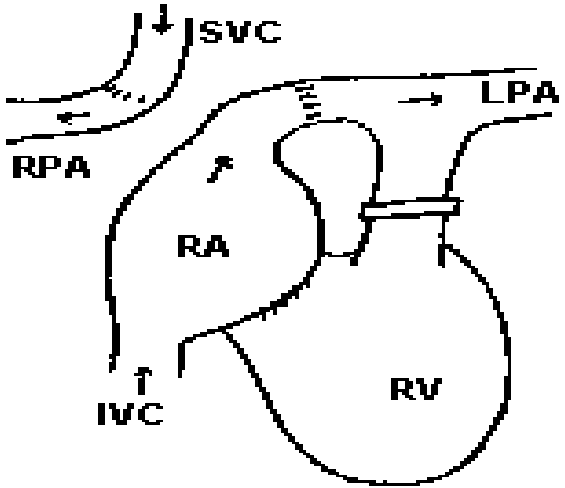
What is the Fontan procedure?

- A novel solution to a desparate situation



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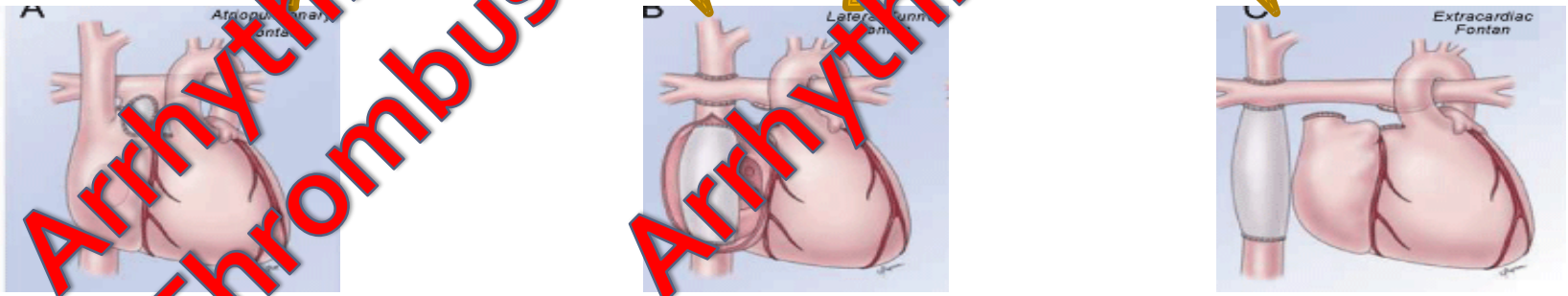
What is the Fontan procedure?

- Evolving treatment pathway
  - Technical modifications attempted to overcome adverse consequences



1970'mid/late 80's
RA-PA connection

Mid/late 80's to mid 90's
Lateral tunnel

Mid 90's to present
External conduit

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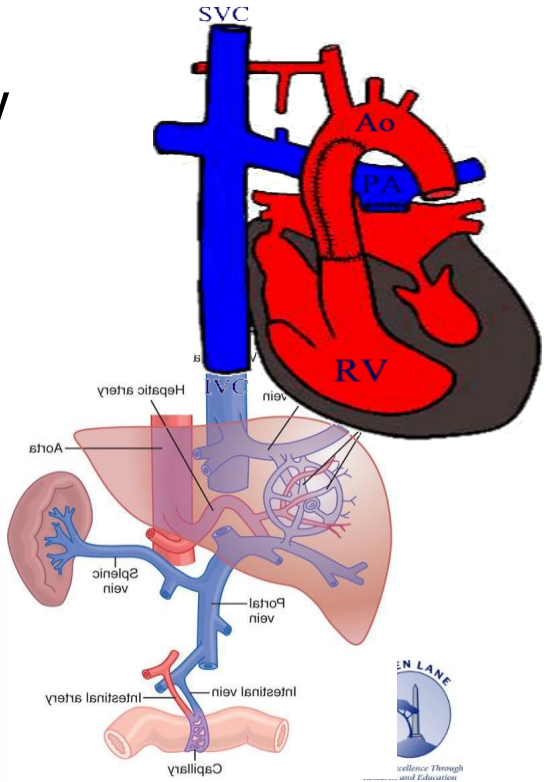
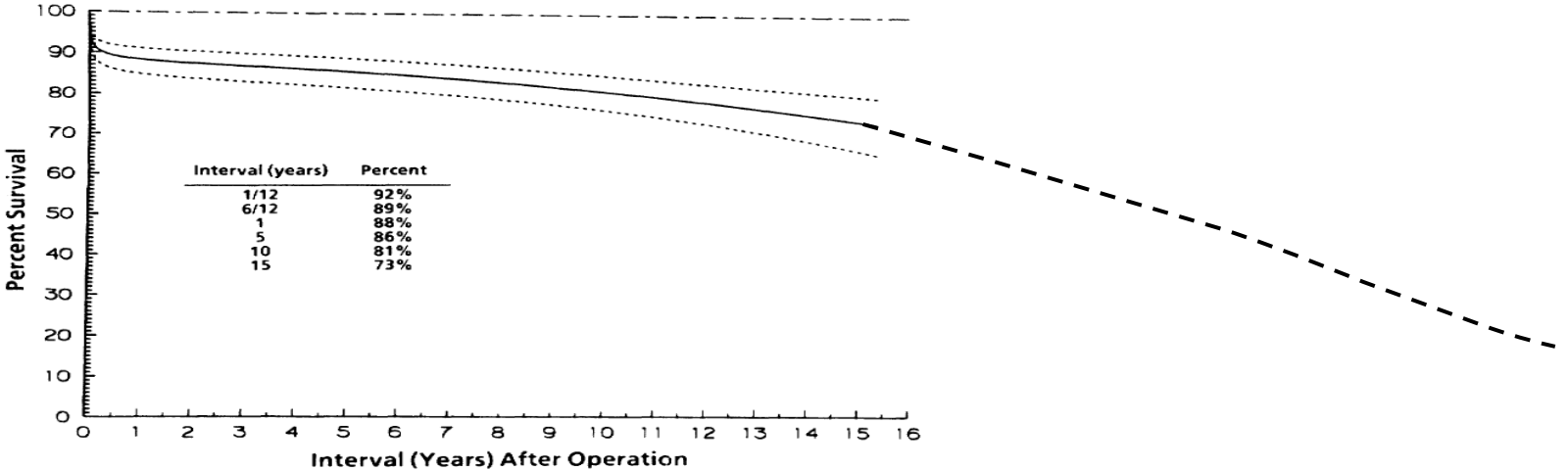
Consequences of the Fontan Circulation

- Elevated central venous pressure and low cardiac output
- Reduced exercise capacity
- Insidious progessive end-organ damage
  - Liver kidney
  - GI tract
  - Myocardium



Outcome after a "perfect" Fontan



Fontan et al, Circulation 1990

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- BUT!!
  - Perceptions are biased by patients with difficult problems
  - Most the studies are from large quaternary referral centres with a potential bias toward more complex patients

ANZ Fontan Registry

fontanregistry.com

Fontan REGISTRY

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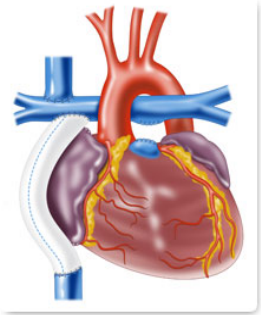

Heart disease is the leading cause of child death in Australia, accounting for one in three childhood deaths. While some patients require only minor surgical procedures, others need major operations to improve their health. Researchers are studying the outcomes of one of these operations, known as the Fontan procedure. A lot of patients born with abnormal hearts cannot have their heart repaired with two pumping chambers (ventricles) like everybody else. Most of them are born without part or all of one pumping chamber. They are said to have a single ventricle. These patients with the most complex forms of heart disease can lead an almost normal life after the Fontan operation, in which the blood to the lungs is redirected to bypass the heart.

Since its conception 40 years ago, the Fontan procedure has given a lifeline to an increasing number of children, allowing them grow and enjoy a good quality of life. However, there are still many unanswered questions about their long-term health prospects.

To find out how we can help these patients in the future, a registry has been established to collect health information on all Fontan patients living in Australia and New Zealand. It is hoped this information will help researchers and doctors determine the medical needs of patients to improve their long-term health and life expectancy.

Murdoch Childrens Research Institute
Healthier Kids. Healthier Future.

FACEBOOK

MCRI FACEBOOK

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ANZ Fontan Registry

fontanregistry.com

- How many people have a Fontan circulation in Australia and NZ?
- How are they doing?
- Are they having regular check-ups?
- Centres in Australia and NZ do things differently
  - Are any of these "variances" associated with superior outcomes?

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The ANZ Fontan Registry

- Largest population-based registry of Fontan patients
- >25 publications so far
  - Long term outcome
  - Burden of disease

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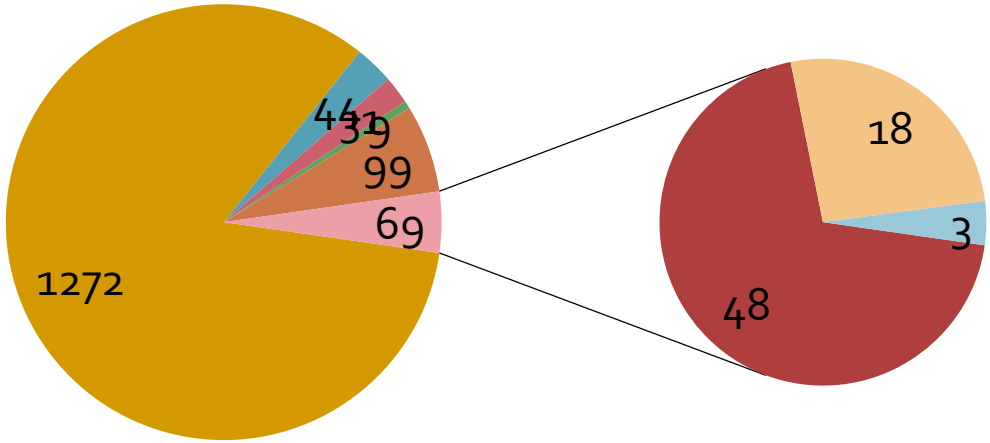
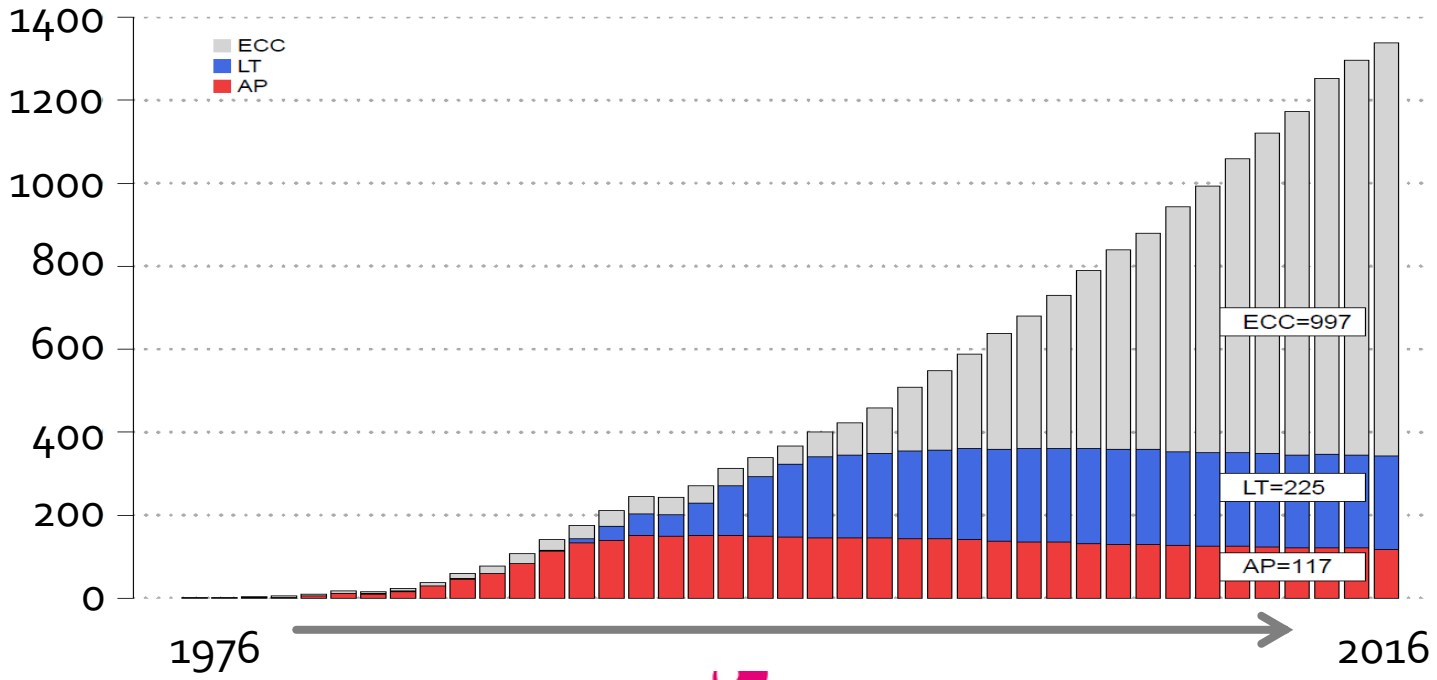
Registry Participation



Consented
Waiver of consent
Transplanted
Taken Down
Deceased
Not Consented "Other"
Refused
Withdrawn

Total 1523 to end 2016

Increasing numbers living with a Fontan circulation



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ANZ Fontan Registry Report 2016

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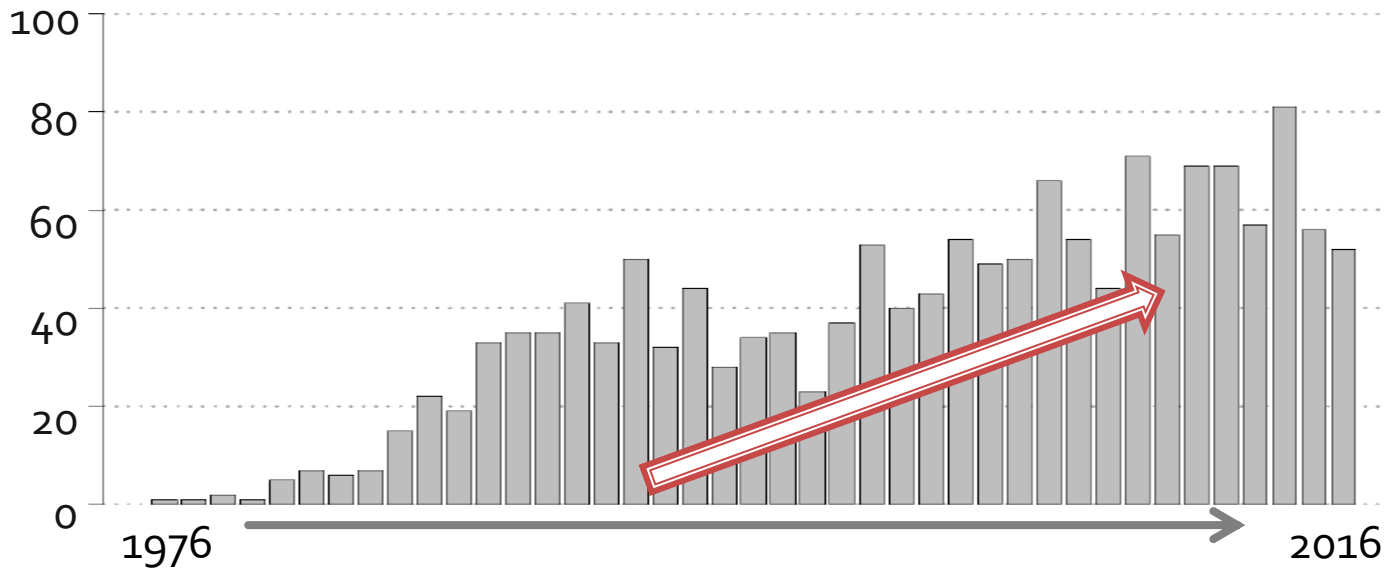
ANZ Fontan surgeries by year



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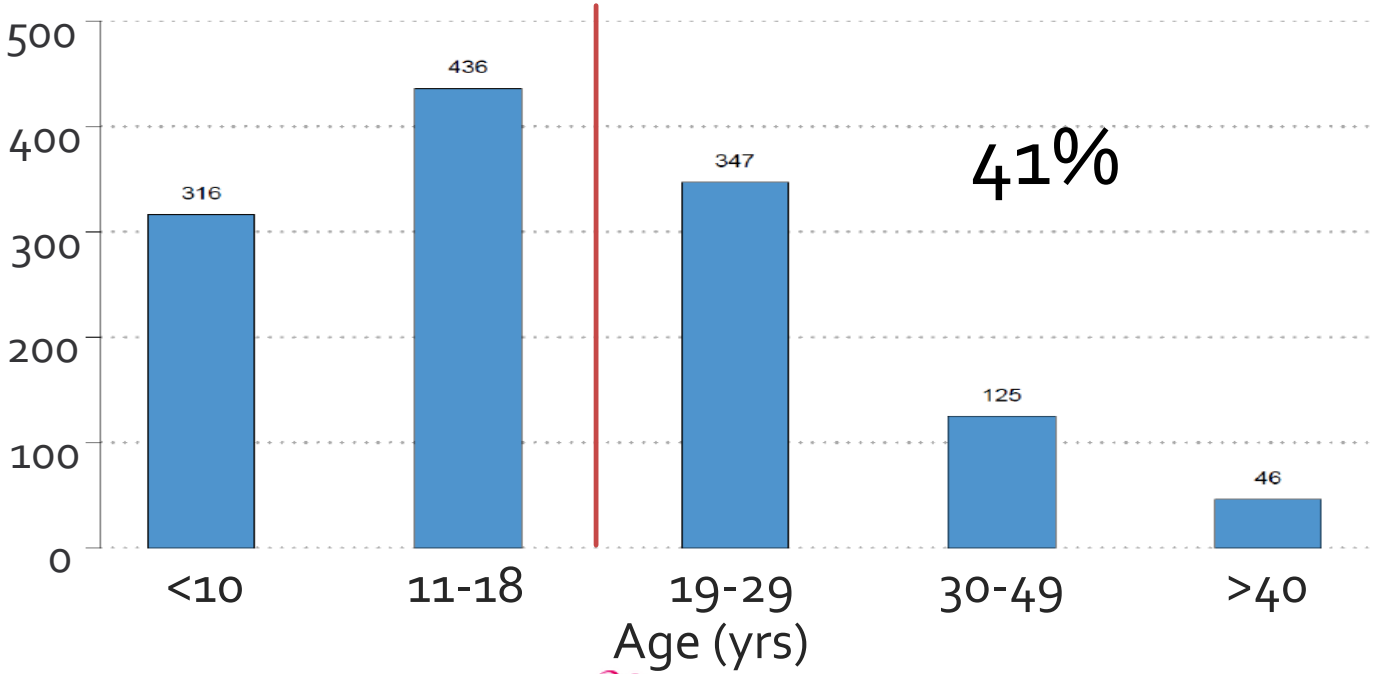
ANZ Fontan Registry: Age



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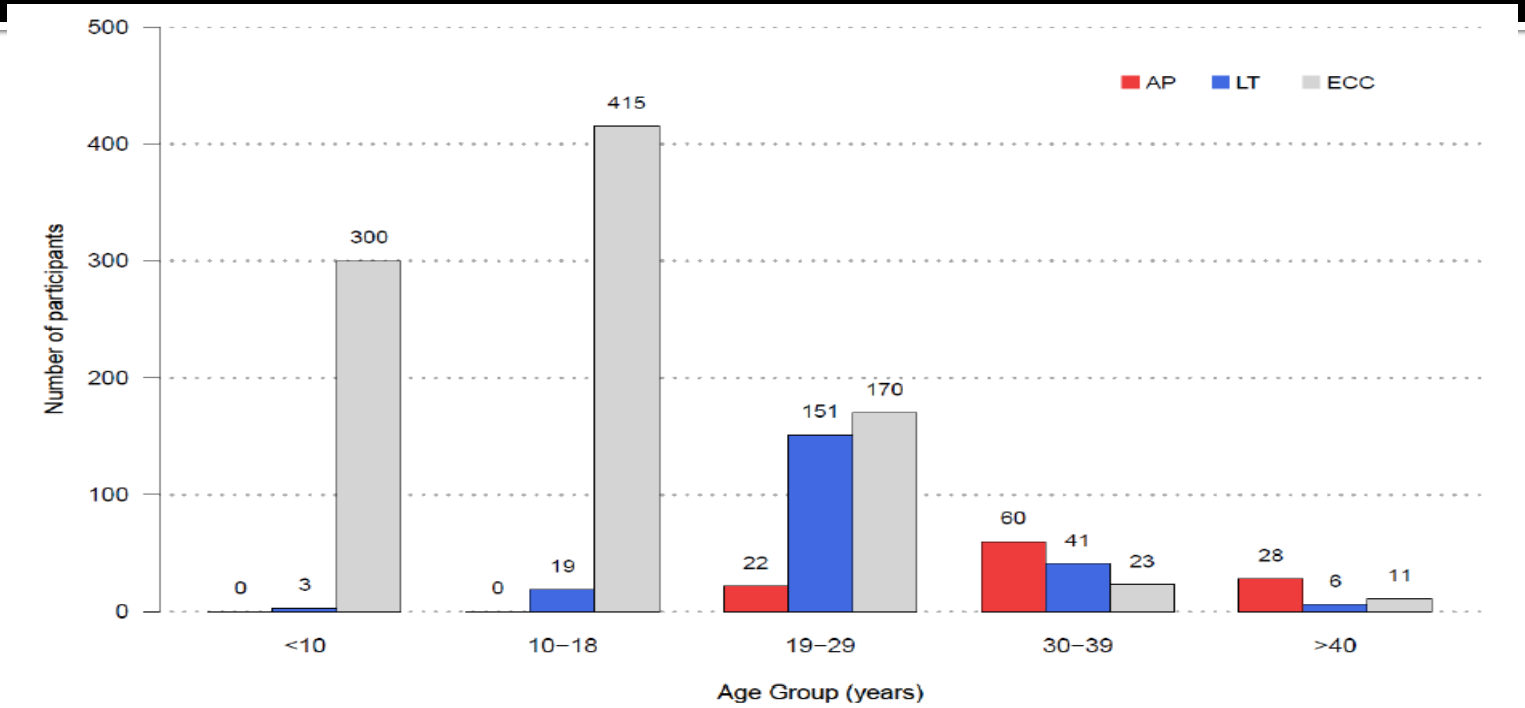

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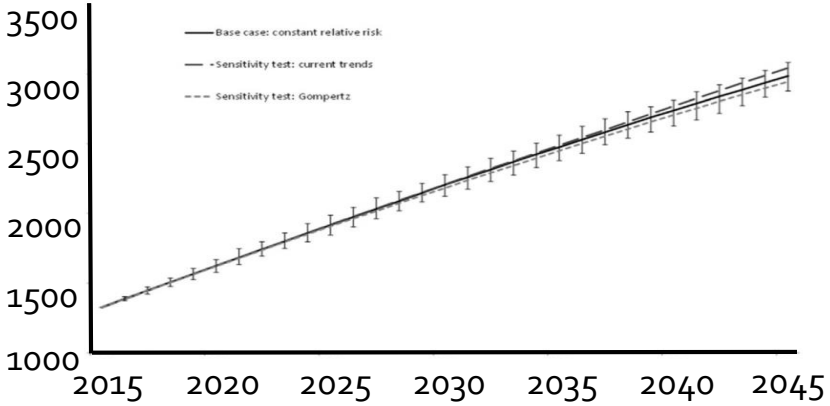
The Fontan epidemic: Population projections from the Australia and New Zealand Fontan Registry

C. Schilling et al. / International Journal of Cardiology 219 (2016) 14–19

Living population



	Survivors	Per 100,000	Average age
2014	1265	4.5	18
2025	1917	5.8	23
2045	2986	7.2	31

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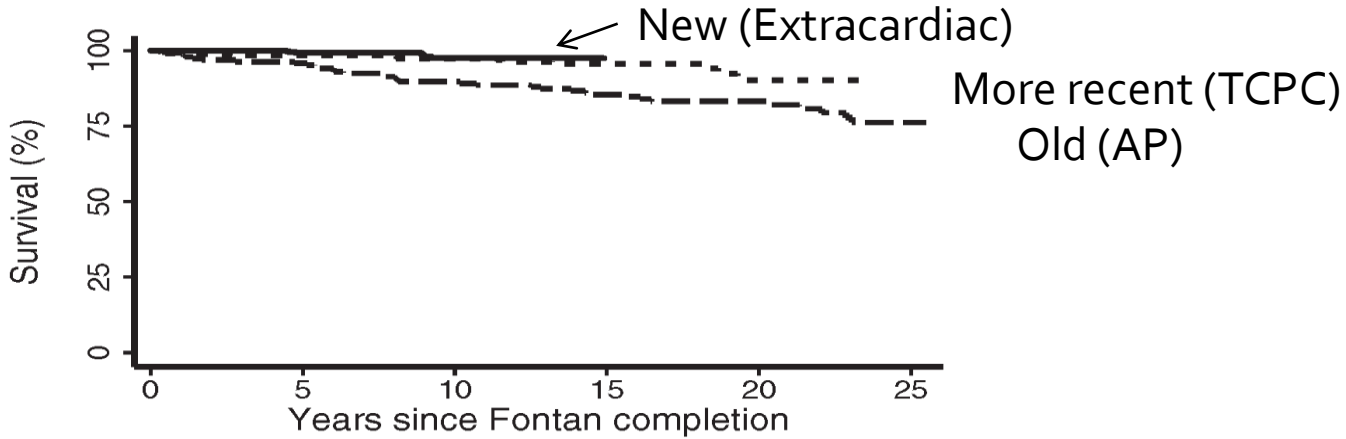
Satisfactory survival 15-20 years out

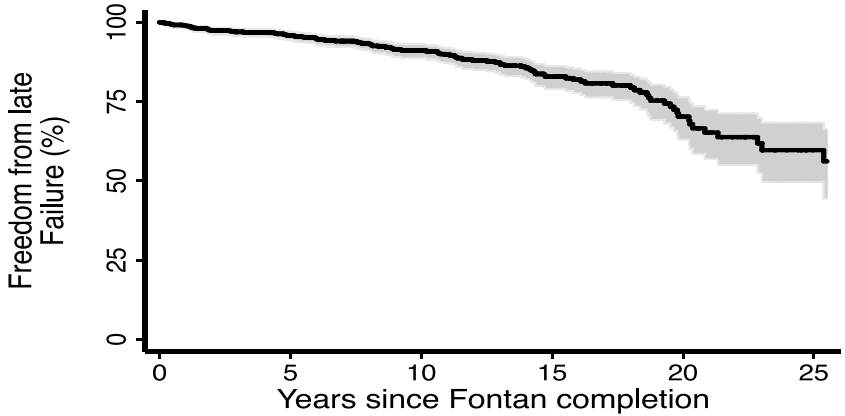
Redefining Expectations of Long-Term Survival After the Fontan Procedure

Twenty-Five Years of Follow-Up From the Entire Population of Australia and New Zealand



d'Udekem et al Circulation 2014

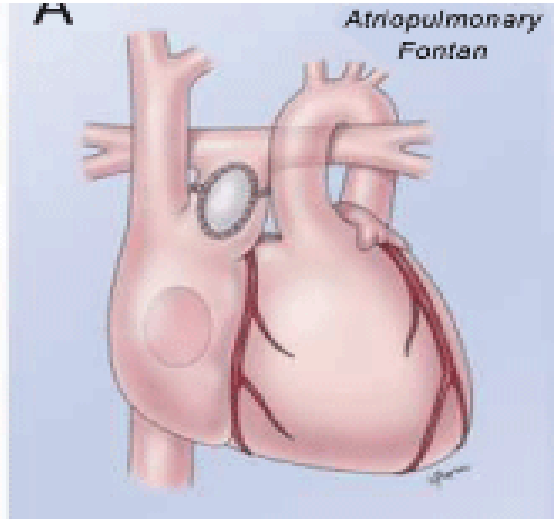
Significant on-going risk of failure

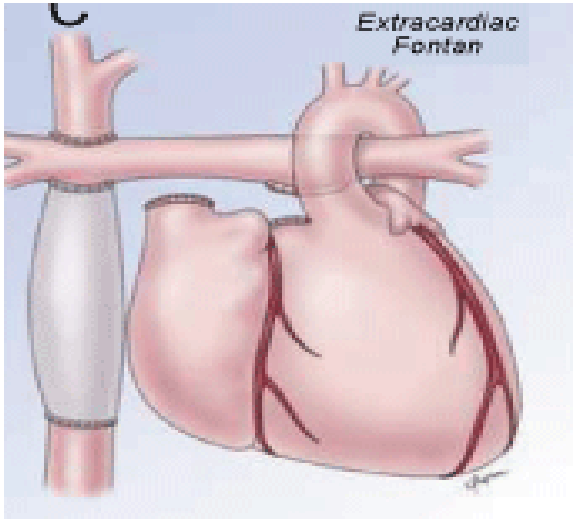


- Death/transplant
- Takedown
- Protein losing enteropathy
- Plastic bronchitis
- NYHA Class 3 and 4

Freedom from failure		95% CI
10 years	91 %	89 – 93%
15 years	83%	79 – 86%
20 years	70%	63 - 76%
25 years	56%	44 – 66%

Fontan Conversion





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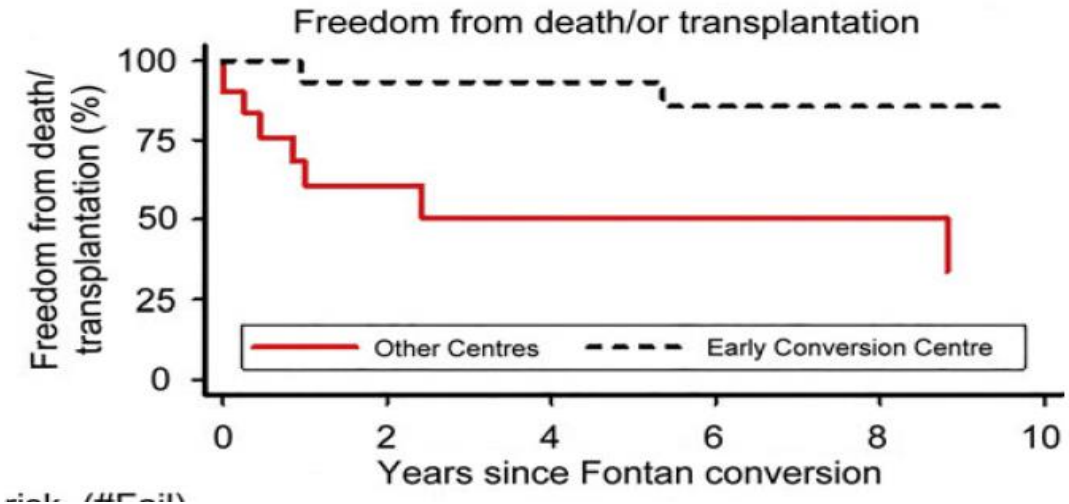
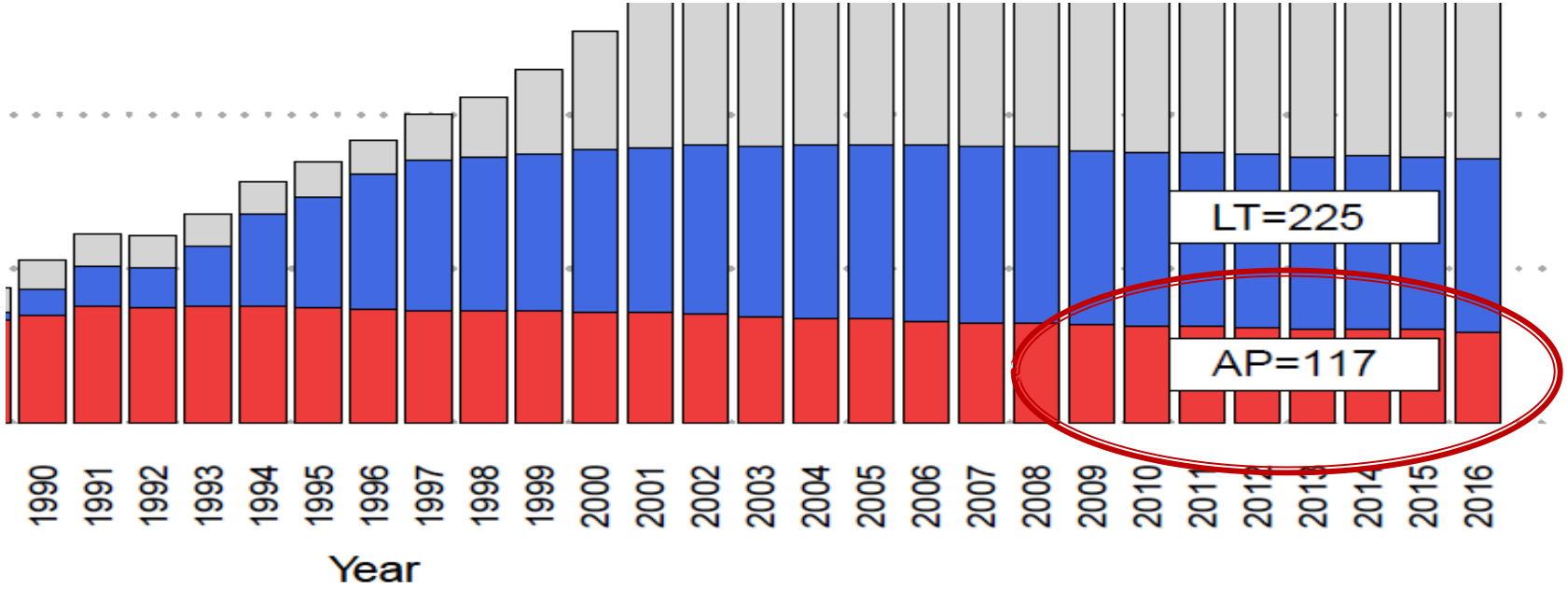
Fontan "conversion"

- Best done before there are too many complicating factors



AP Fontan survivors



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ANZ Fontan Registry Repott 2016

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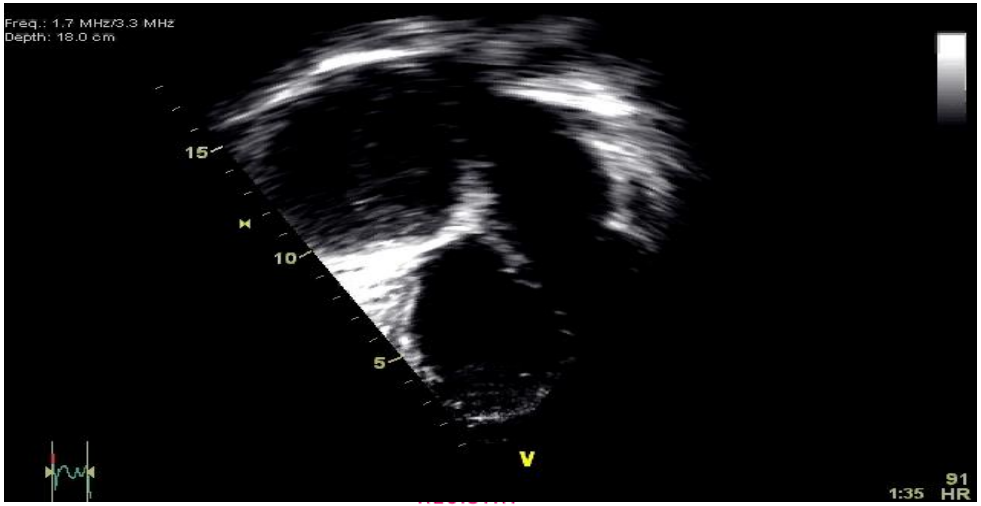
Thrombosis

- Increased risk of thrombosis
  - Venous stasis and procoagulants



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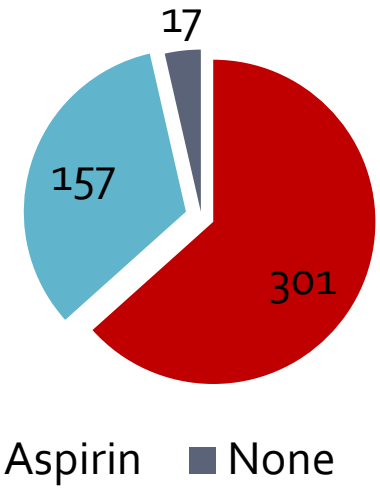
No difference between aspirin and warfarin after extracardiac Fontan in a propensity score analysis of 475 patients

Eur J Cardiothorac Surg 2016

- Institutional bias
  - Two centres predominately used warfarin.
  - Other centres used warfarin for 12 months then aspirin
- Those on warfarin tended to have less favorable anatomy



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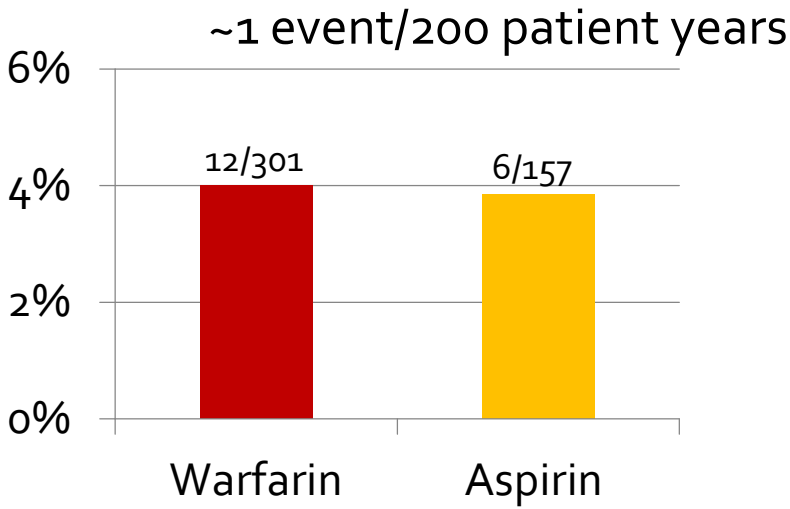
No difference between aspirin and warfarin after extracardiac Fontan in a propensity score analysis of 475 patients

Thromboembolism >1 yr after Fontan



18 thrombotic events >1 year after Fontan

- 10 conduit thrombosis (9 routine testing)
- 3 CVA strokes
- 2 TIA
- 3 Pulmonary embolism

12 serious bleeding events

- 11 warfarin (4 with INR high)

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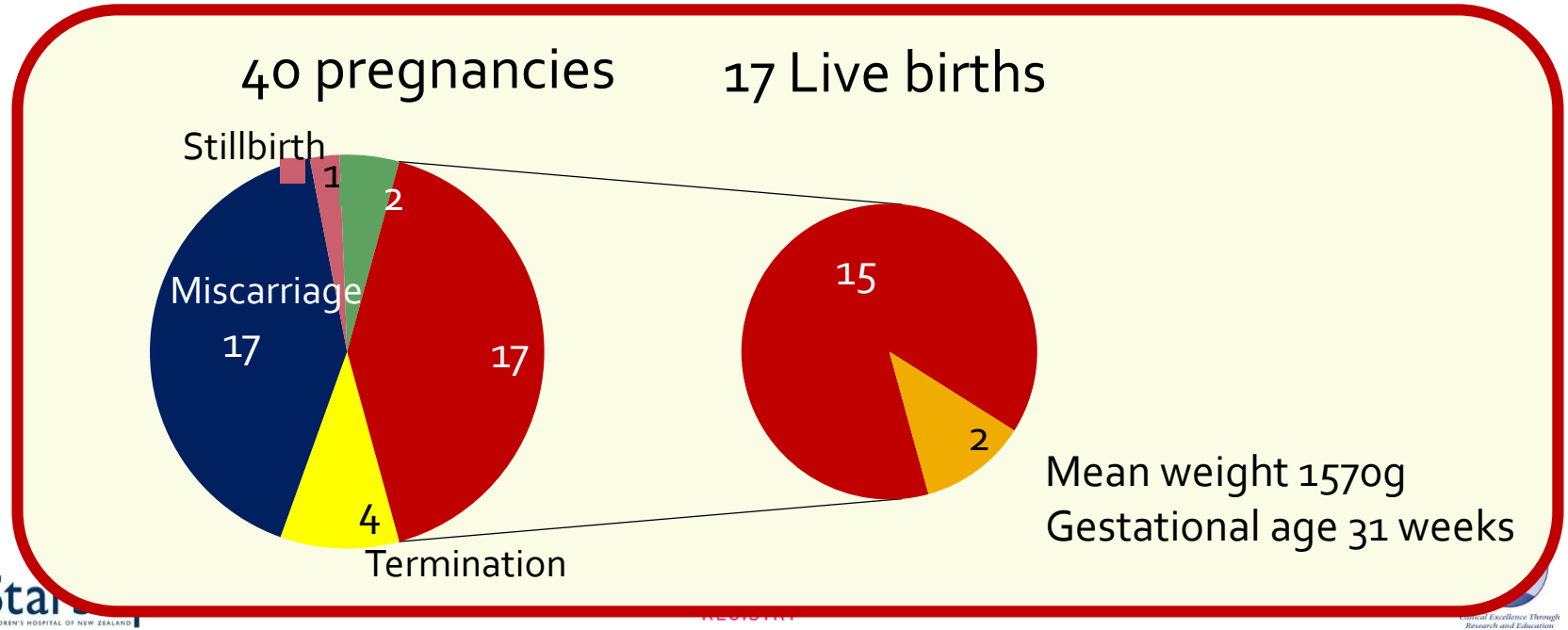
Fertility and pregnancy in the Fontan population

Int J Cardiol 2016:208:97-101

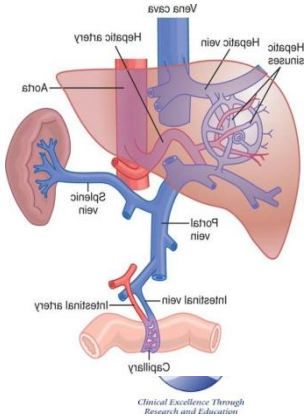
Questionnaire based study - 27 women



Liver and renal study

- Rationale
  - Liver and renal dysfunction known to occur in the Fontan circulation
  - Pattern of dysfunction, prevalence and temporal behavior poorly understood
- Cross sectional study design

Liver ultrasound and fibroscan
Serum liver function
Renal DTPA scan for GFR
Serum and urine tests of renal function



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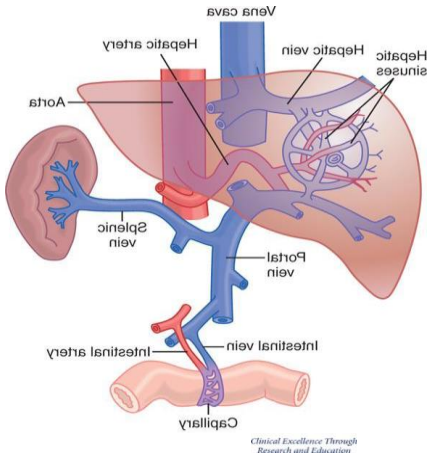
Liver and renal study

- Patient population
  - 152 patients recruited from the ANZ Fontan Registry
  - Similar characteristics vs. the remaining 977 eligible patients
  - Mean age 20 years
  - Mean time since Fontan 14 years



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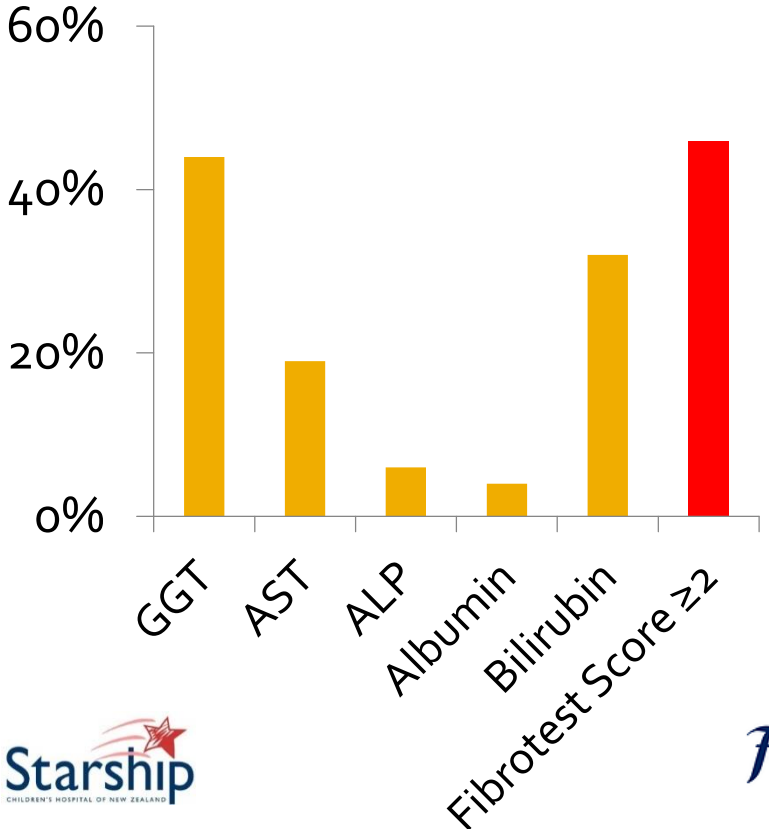
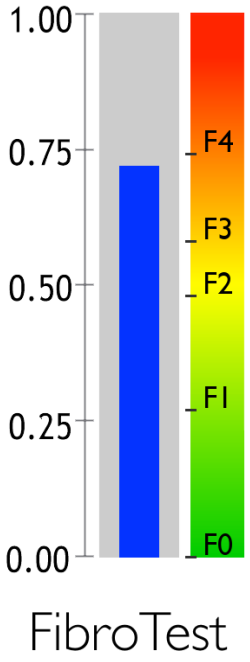
Key findings - liver





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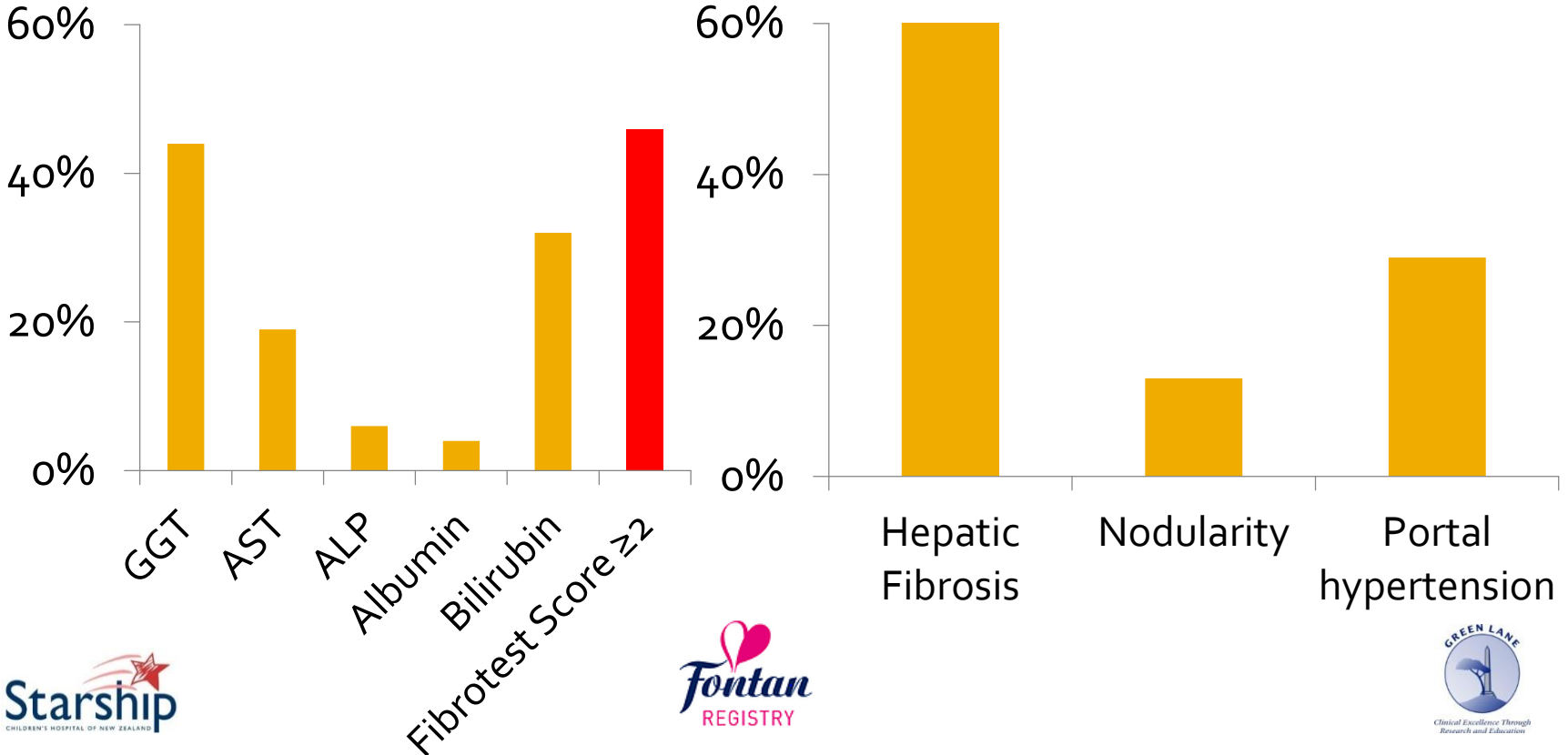
Key findings - liver



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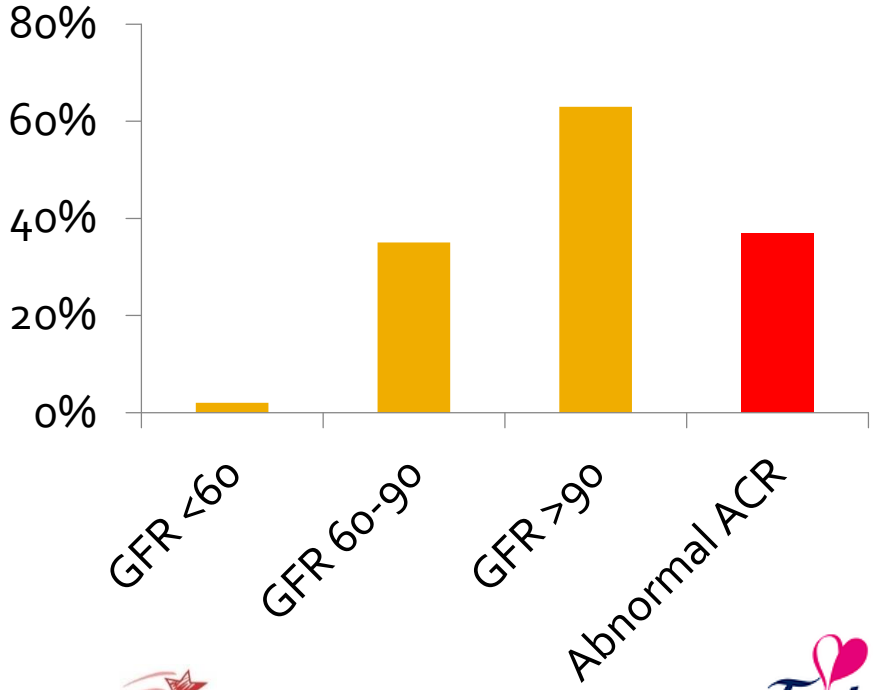
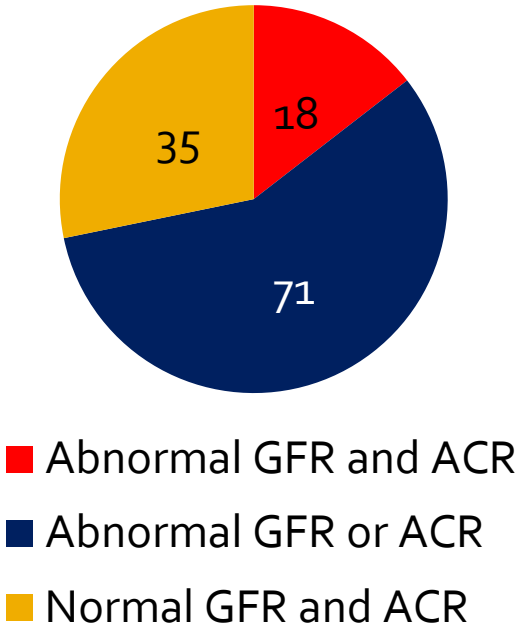
Key Findings - renal





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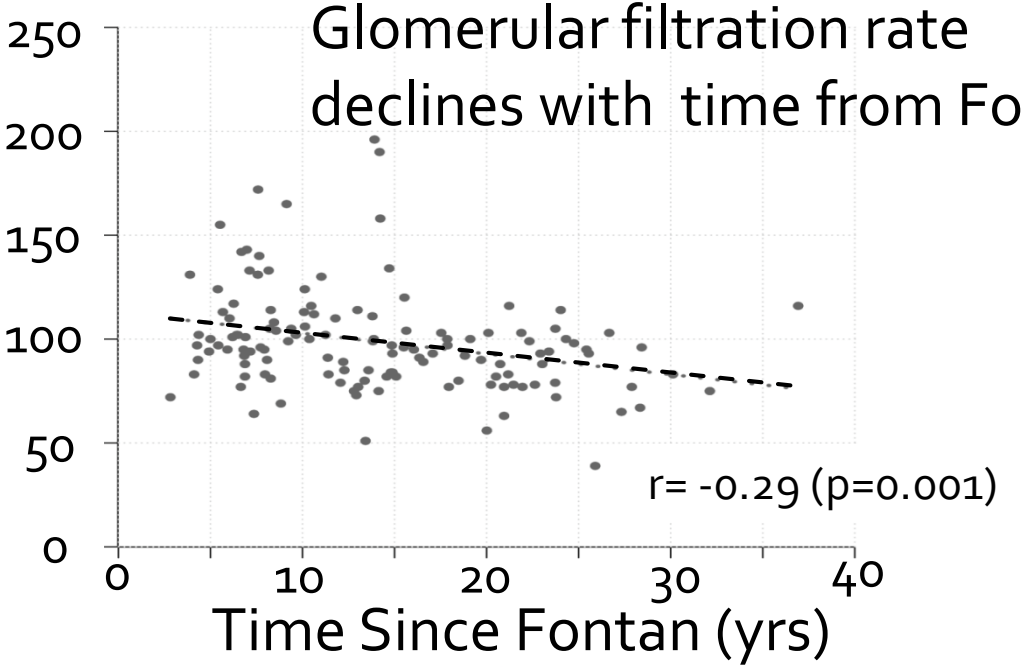
Key Findings - renal

GFR

ml/min/1.73m²



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Current Registry Projects

- Living with a Fontan Circulation
- Cardiovascular response to exercise
- Neurocognitive function
- Exercise training and Fontan circulation function

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Living with a Fontan circulation

- Partnership grant with NHMRC and NHF (Australia), Heart Kids Australia, ANZ hospitals, Heart Kids NZ and Starship Foundation
- Aims:
  - Design "models of care" to improve outcomes
  - Strengthen transition and avoid "lost to follow-up"
  - Measure quality of life and design family support interventions

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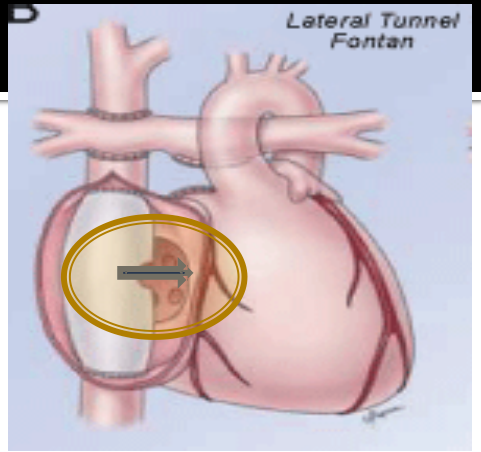
ANZ – On going research



- Heart function and brain study
  - Exercise tests, echocardiogram, MRI, psychosocial questionnaire
    - Can type of operation, fenestration and other factors predict exercise capacity and heart muscle function?
    - Does having a fenestration make any difference to exercise capacity?
    - Does having a fenestration predispose to stroke?

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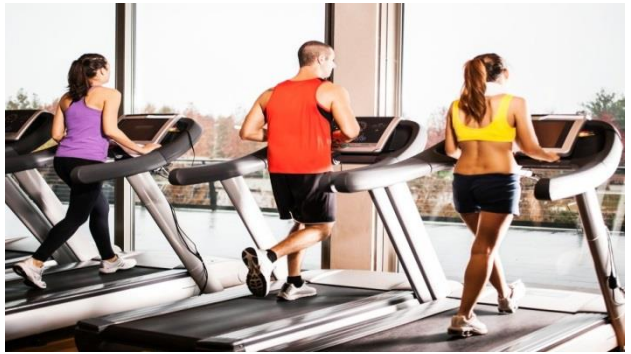
Ongoing research

- Can an exercise programme improve cardiovascular well-being?
  - Study pending funding
  - No intervention vs. training
  - Outcomes



Exercise capacity
Muscle mass
Psychological well-being

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Other questions

- ACE cessation
  - Many Fontan patients taking ACE inhibition
  - Is this type of medication useful?
  - Design phase

Engaging with the Fontan Population

Annual Fontan Education Days

- 2014 - Melbourne
- 2015 - Sydney
- 2016 - Auckland
- 2107 - Brisbane

Disseminate project results

Forum for discussion between researchers and patients

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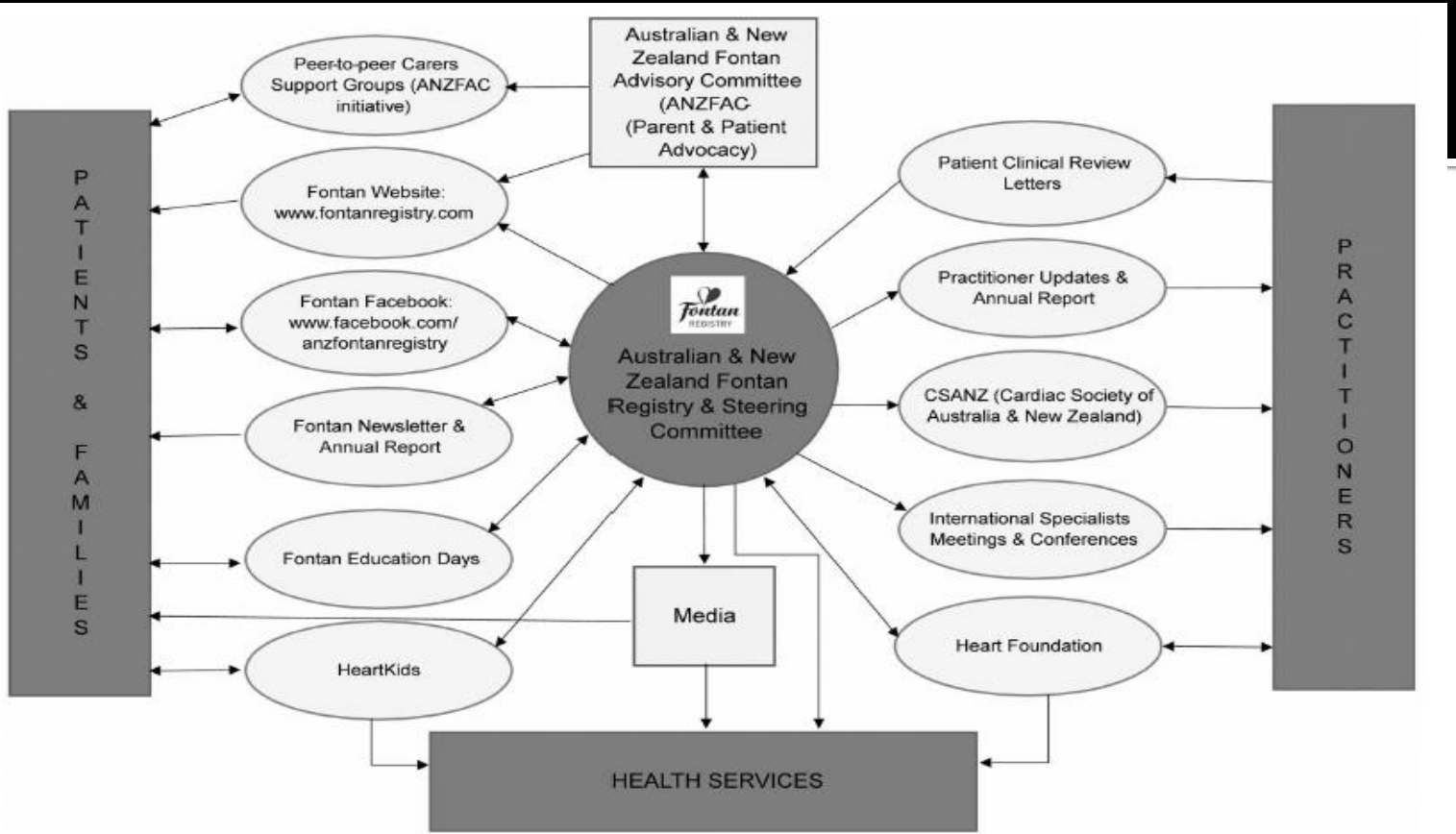

D'Udekem Y et al, Cardiol Young 2017

ANZ Fontan Registry

- Optimise long term outcome by improving
  - treatment options
  - care pathways

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Cystic fibrosis : median age of death

- 1957 : 3 years

Cystic Fibrosis Foundation investigation
31 CF centers
In the best: median age: 21 years

- 1966: 10 years
- 1972: 18 years

Standardisation of care
Centralisation of care

- 2003: 33 years
- 2004 — Identification of the best 5 centers
- 2006 — Identification of all 117 CF centers

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ANZ Fontan Registry - acknowledgements

- ANZ Fontan Registry
- HeartKids Australia and Heart Kids NZ

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