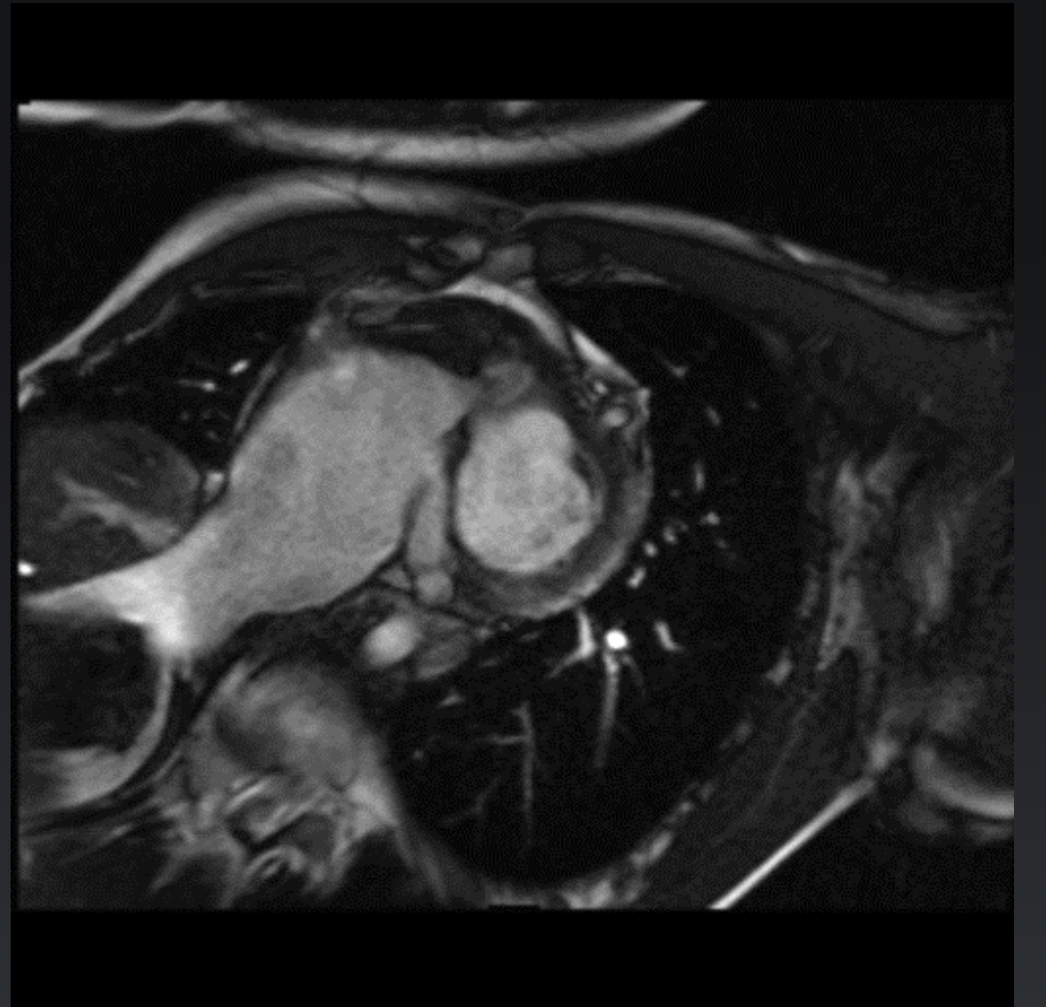
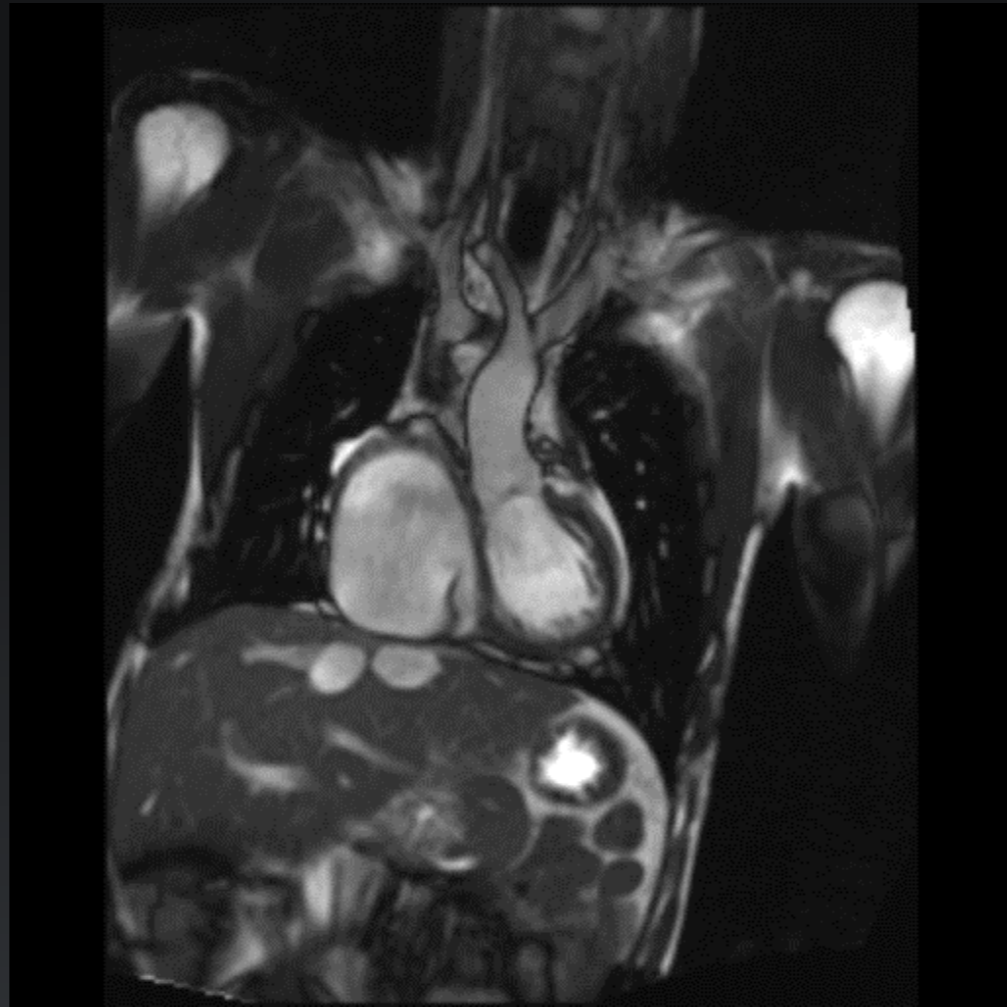
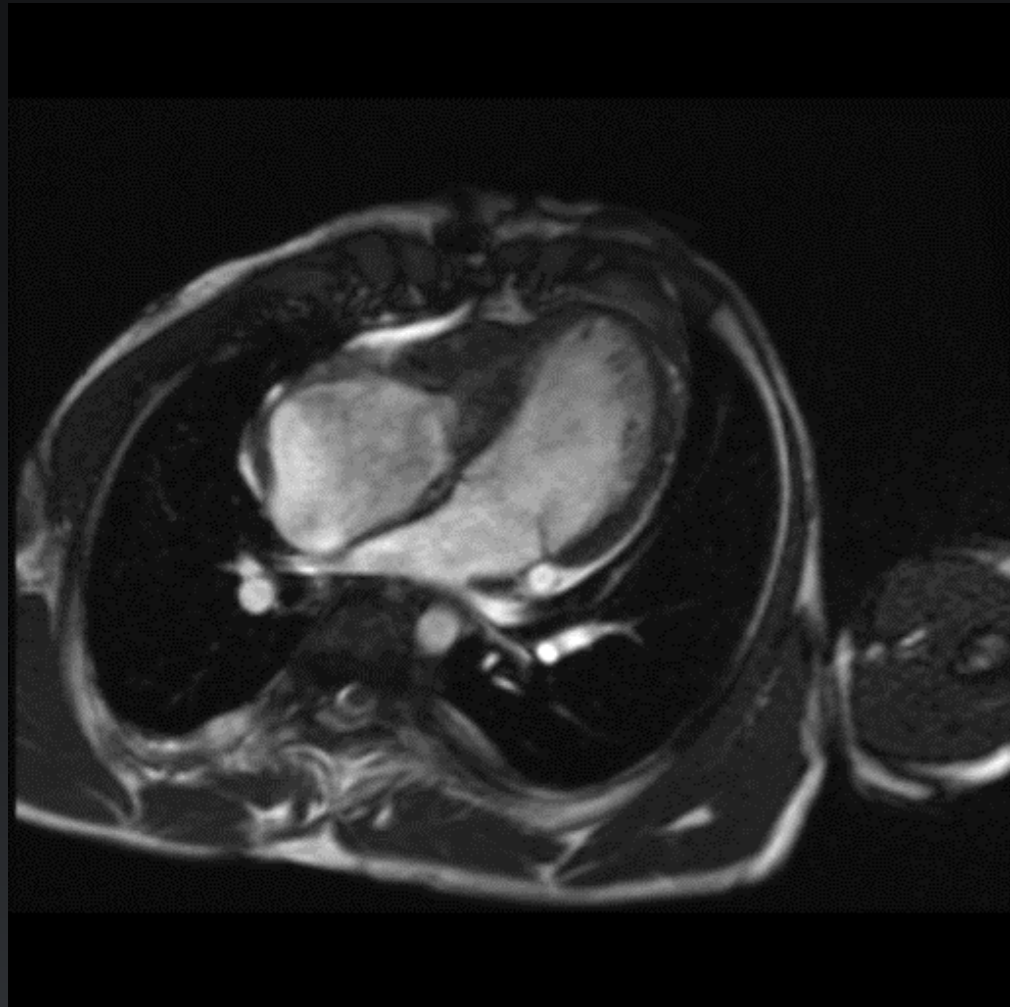




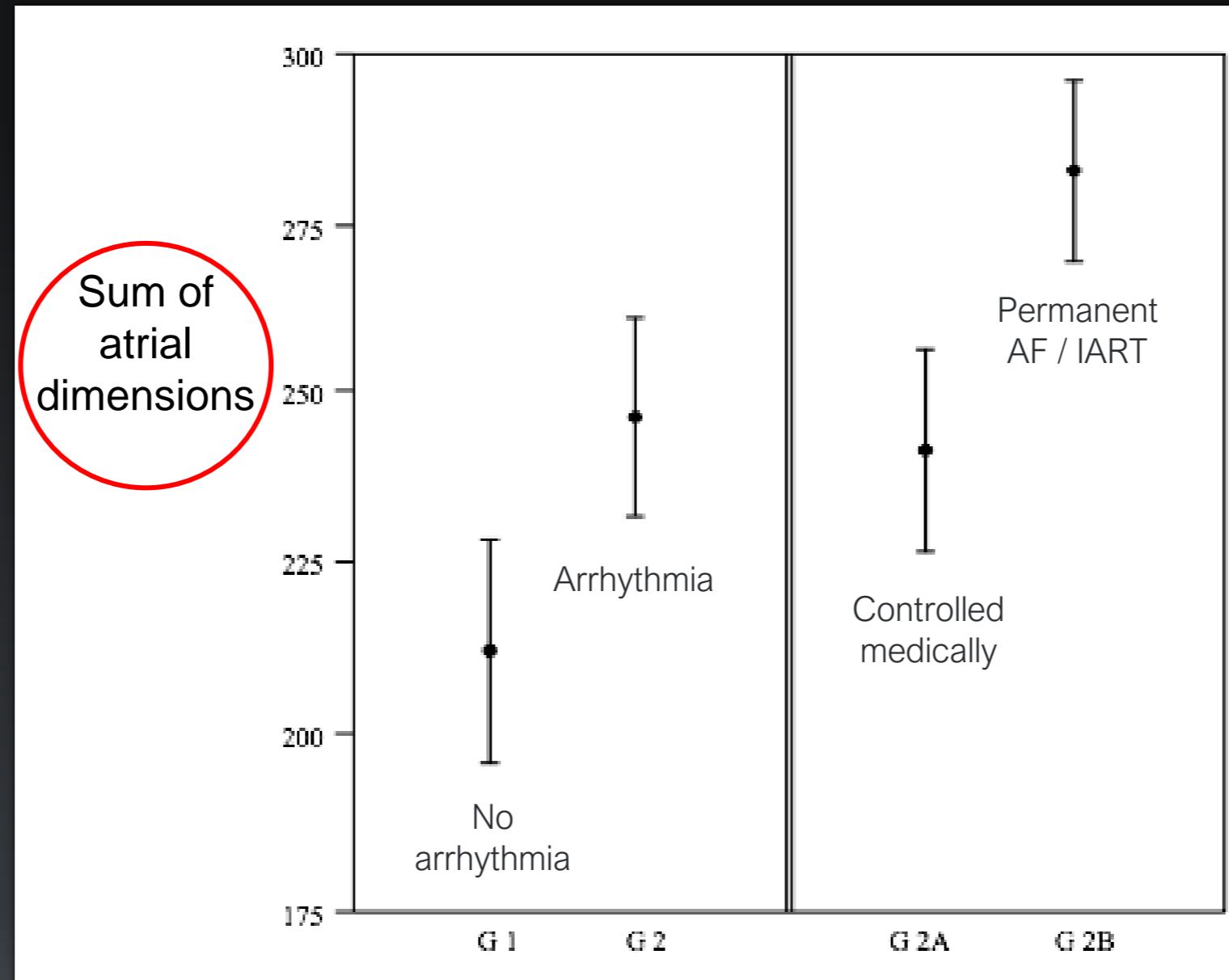
# The Role of Fontan Conversion

Tim Hornung

Green Lane Congenital Cardiac Service  
Starship & Auckland Hospital

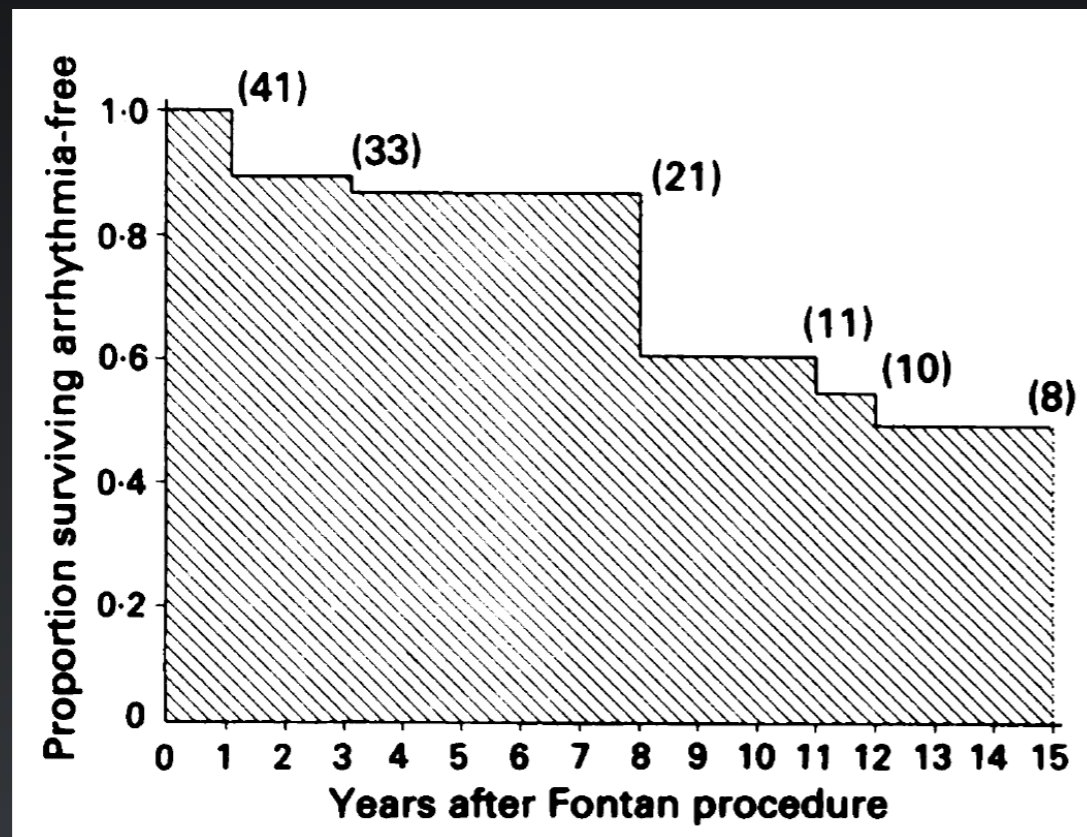


# Progressive RA Dilation → Atrial Arrhythmias

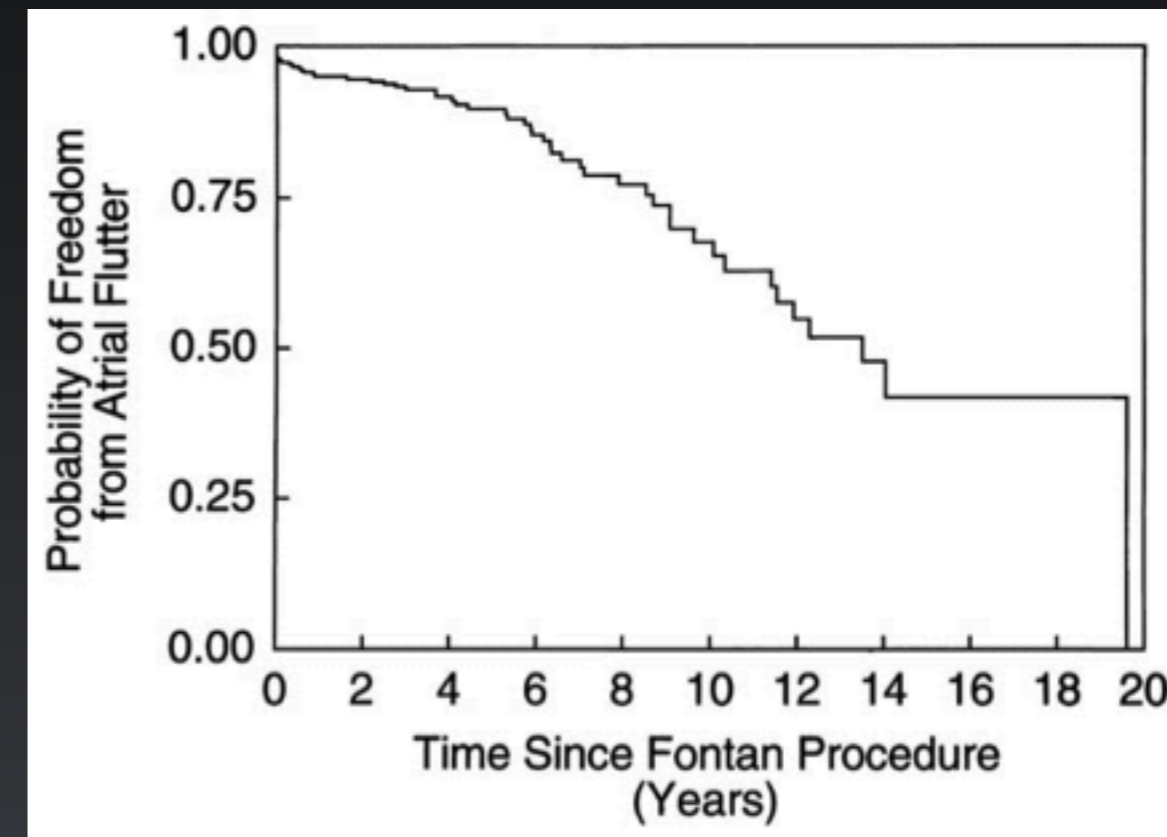


Soukias N, Hornung TS, Kilner PJ, Frogoudaki A, Davlouros P, Wong T, Gatzoulis MA. Determinants of atrial arrhythmia late after atriopulmonary Fontan operation. *Hellenic J Cardiol.* 2004;45:384-90.

# Atrial Arrhythmias After Atrio-Pulmonary Fontan Operation

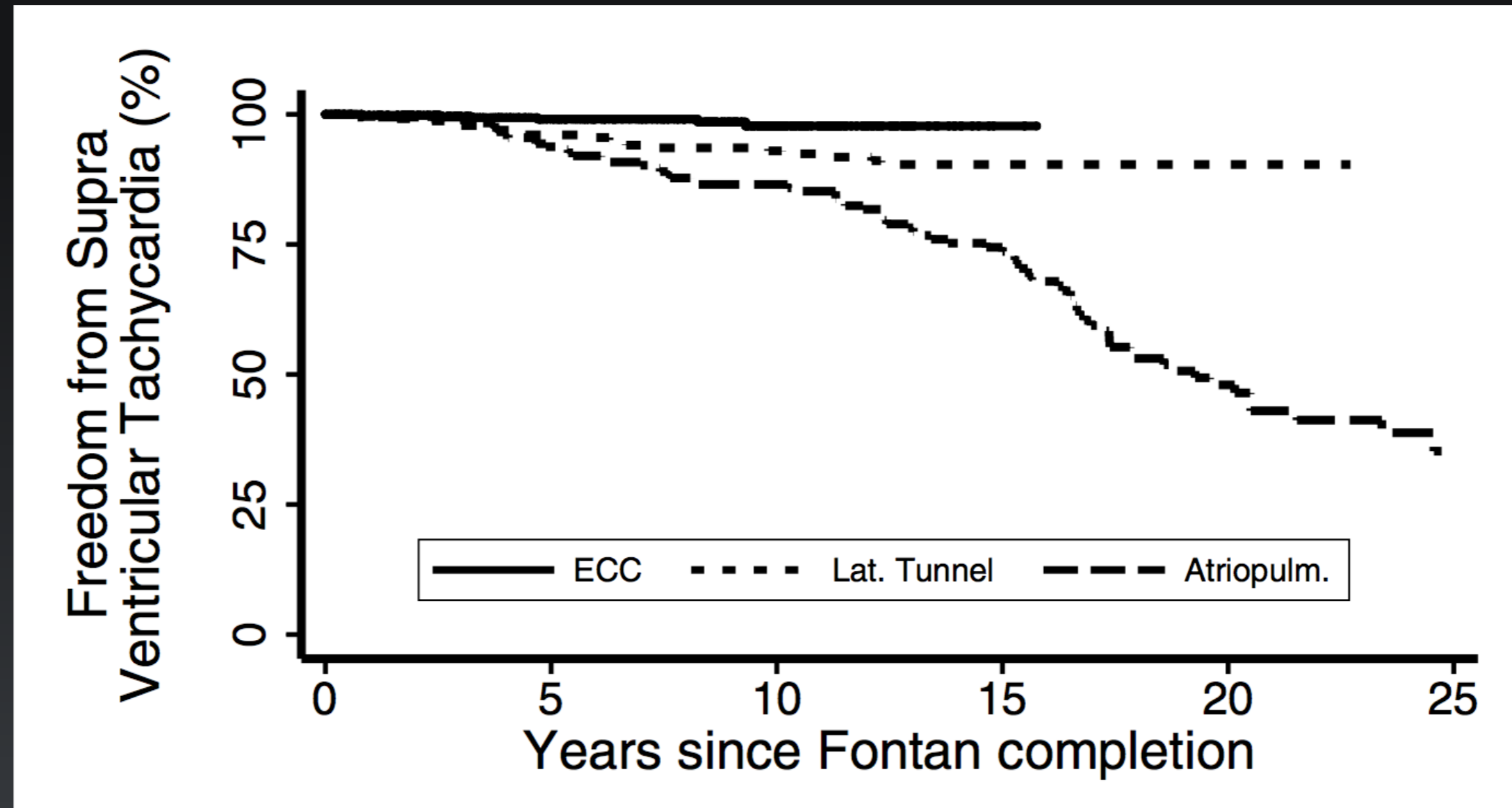


Peters, Somerville (London)  
Br Heart J 1992;68:199-204.



Fishberger, Walsh (Boston)  
J Th CV Surgery 1997;113:80-86.

# Australia and NZ Fontan Registry Data



D'Udekam Y et al. 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. *Circulation*. 2014;130:[suppl 1]S32-S38.

# Arrhythmia Burden:

## ANZ Fontan Registry: AP Fontan Data

- 215 hospital survivors
- 130 developed atrial arrhythmias
  - 101 atrial flutter
  - 50 atrial fibrillation
  - 3 SVT
- Freedom from arrhythmia
  - At 20 years: **45%**
  - At 28 years: **23%**
- Development of arrhythmia increased the likelihood of death or transplantation: **OR 3.1**

Poh CL et al. Three decades later: The fate of the population of patients who underwent the Atriopulmonary Fontan procedure. International Journal of Cardiology. 2017;231:99-104.



# Medical Treatment

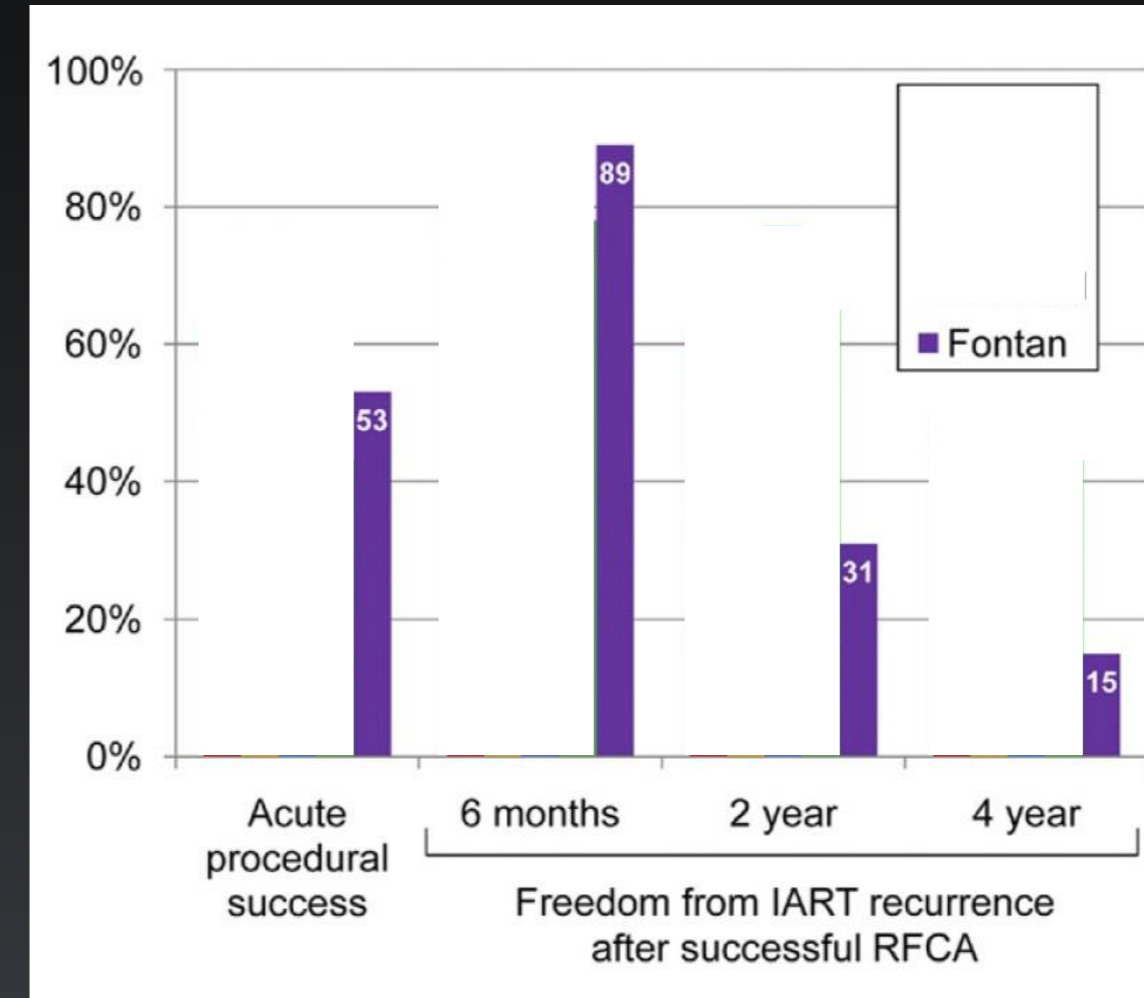
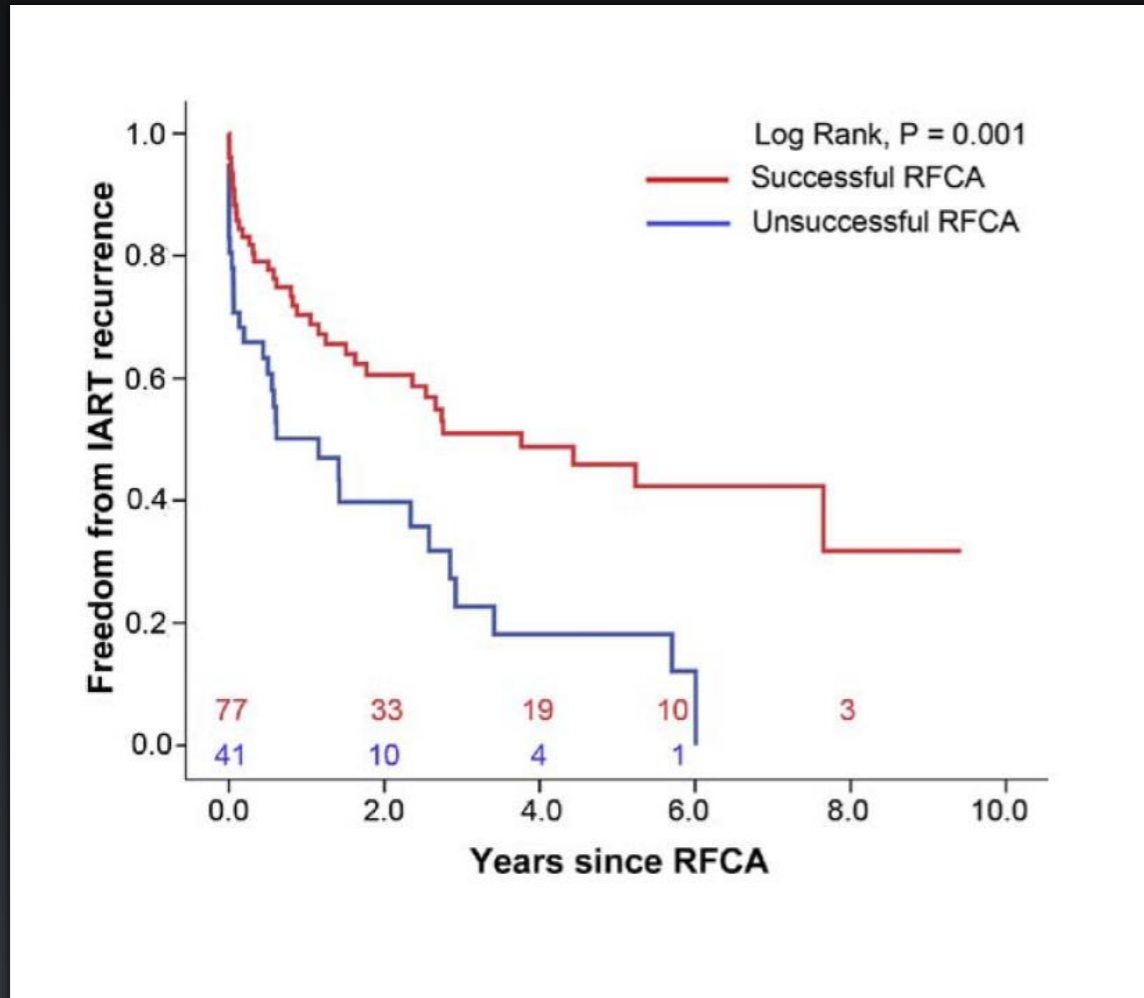
## Amiodarone-associated thyroid dysfunction: risk factors in adults with congenital heart disease.

- 92 ACHD patients taking amiodarone for > 6 months
- Mean age 34 years
- **36% developed thyroid dysfunction**
- **Risk factors included previous Fontan surgery (OR 4.0, p=0.17)**



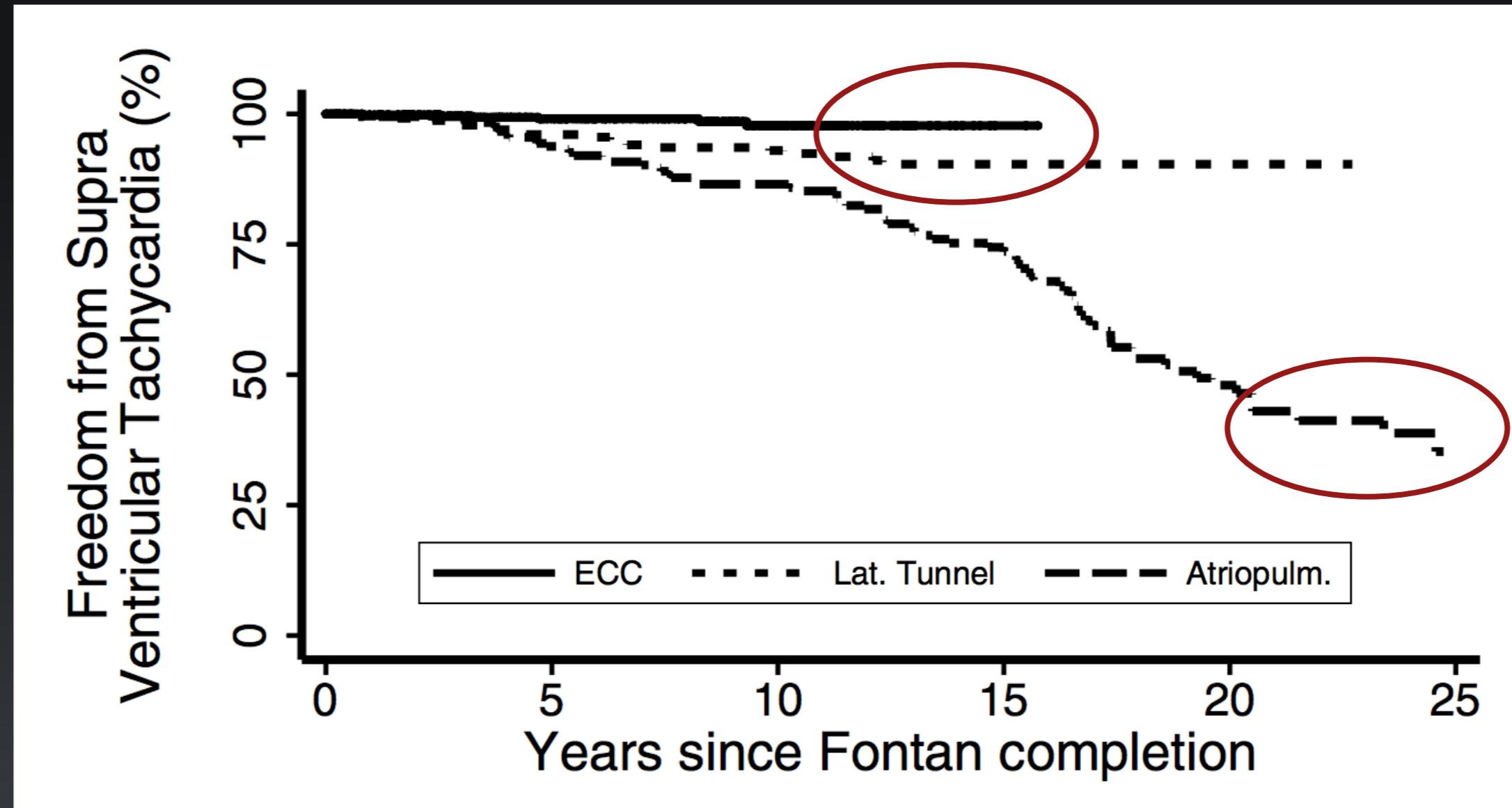
# Catheter Ablation

Outcome of Intra-Atrial Re-Entrant Tachycardia Catheter Ablation in Adults with Congenital Heart Disease. *S-C Yap et al (Toronto). JACC 2010;56:1589-96.*



# Fontan Conversion

# Australia and NZ Fontan Registry Data

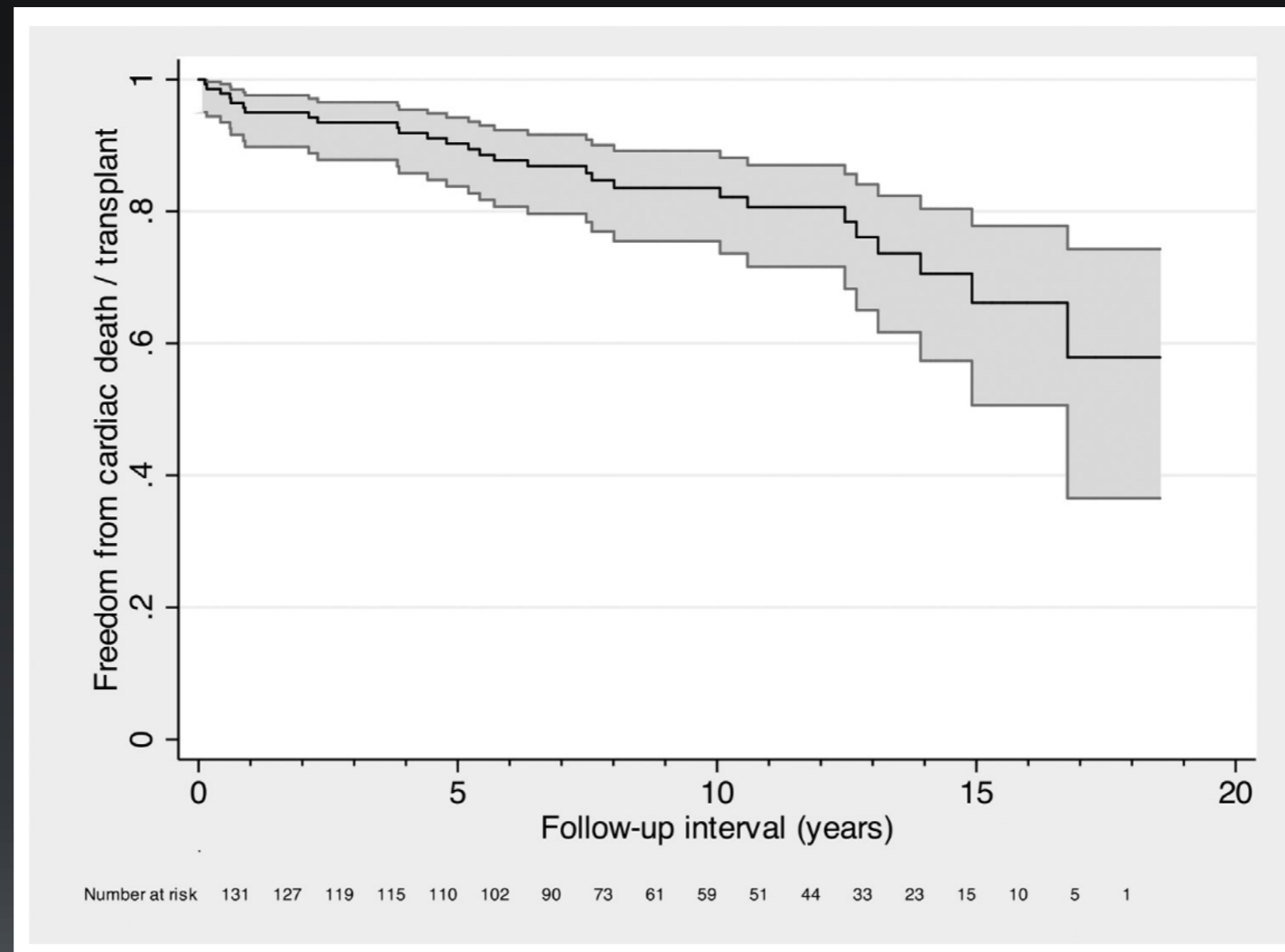


D'Udekam Y et al. Redefining Expectations of Long-Term Survival After the Fontan Procedure: Twenty-Five Years of Follow-Up From the Entire Population of Australia and New Zealand. *Circulation*. 2014;130:[suppl 1]S32-S38.

# Fontan Conversion Outcomes (Chicago)

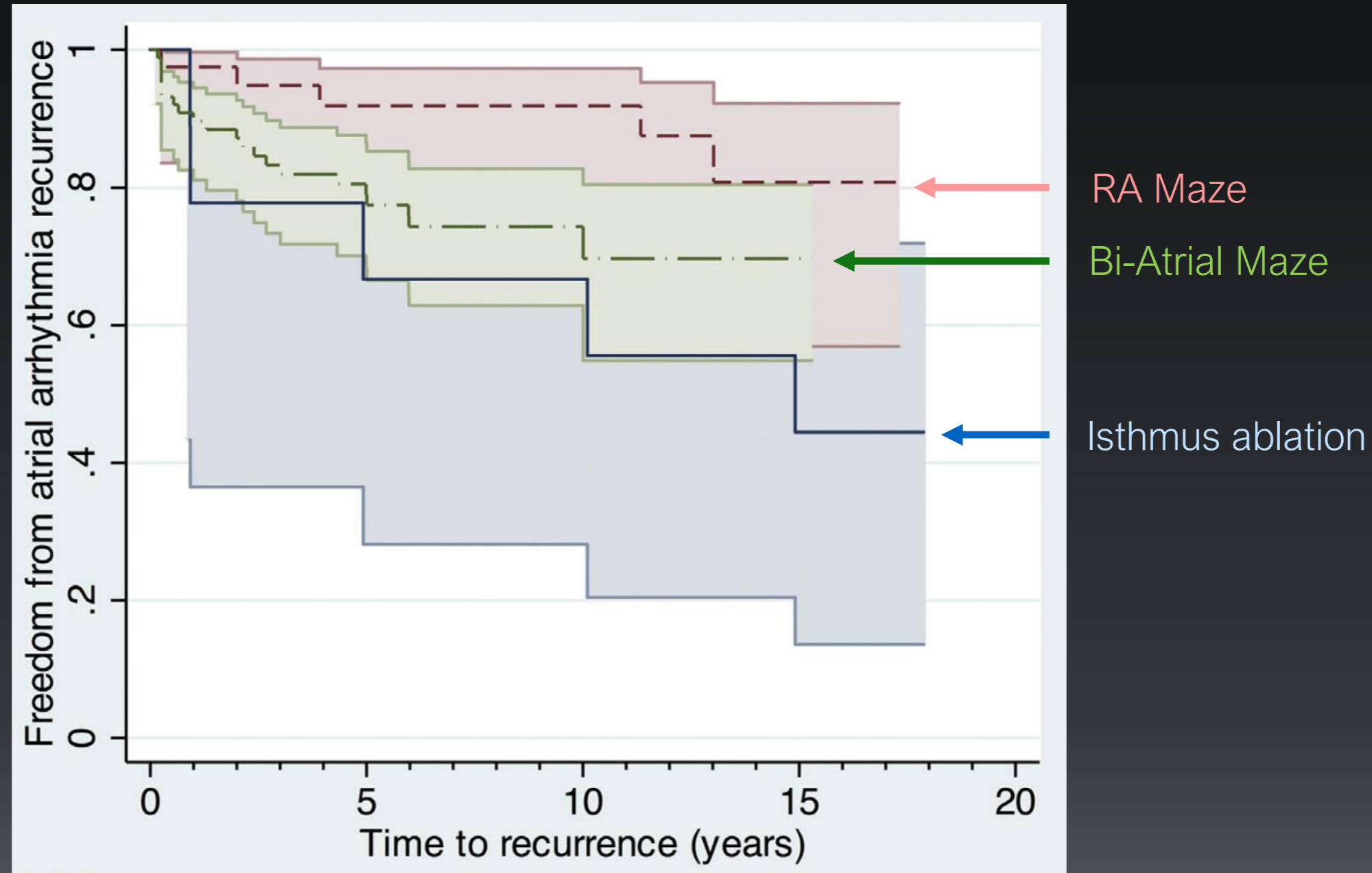
- 140 Patients
  - Median age 23 years
  - Median time since Fontan 16 years
- **Freedom from recurrent arrhythmia 77% (10 yr F/U)**
- Early death or transplant 3
- Late death or transplant 25 (non-cardiac 3)
  - **Total death or transplant 20%**

# Fontan Conversion Outcomes (Chicago)



Deal, BJ et al (Chicago). Intermediate-Term Outcome of 140 Consecutive Fontan Conversions With Arrhythmia Operations. *Ann Thorac Surg* 2016;101:717–24.

# Fontan Conversion Outcomes (Chicago)



Deal, BJ et al (Chicago). Intermediate-Term Outcome of 140 Consecutive Fontan Conversions With Arrhythmia Operations. *Ann Thorac Surg* 2016;101:717–24.

# Fontan Conversion Outcomes (International)

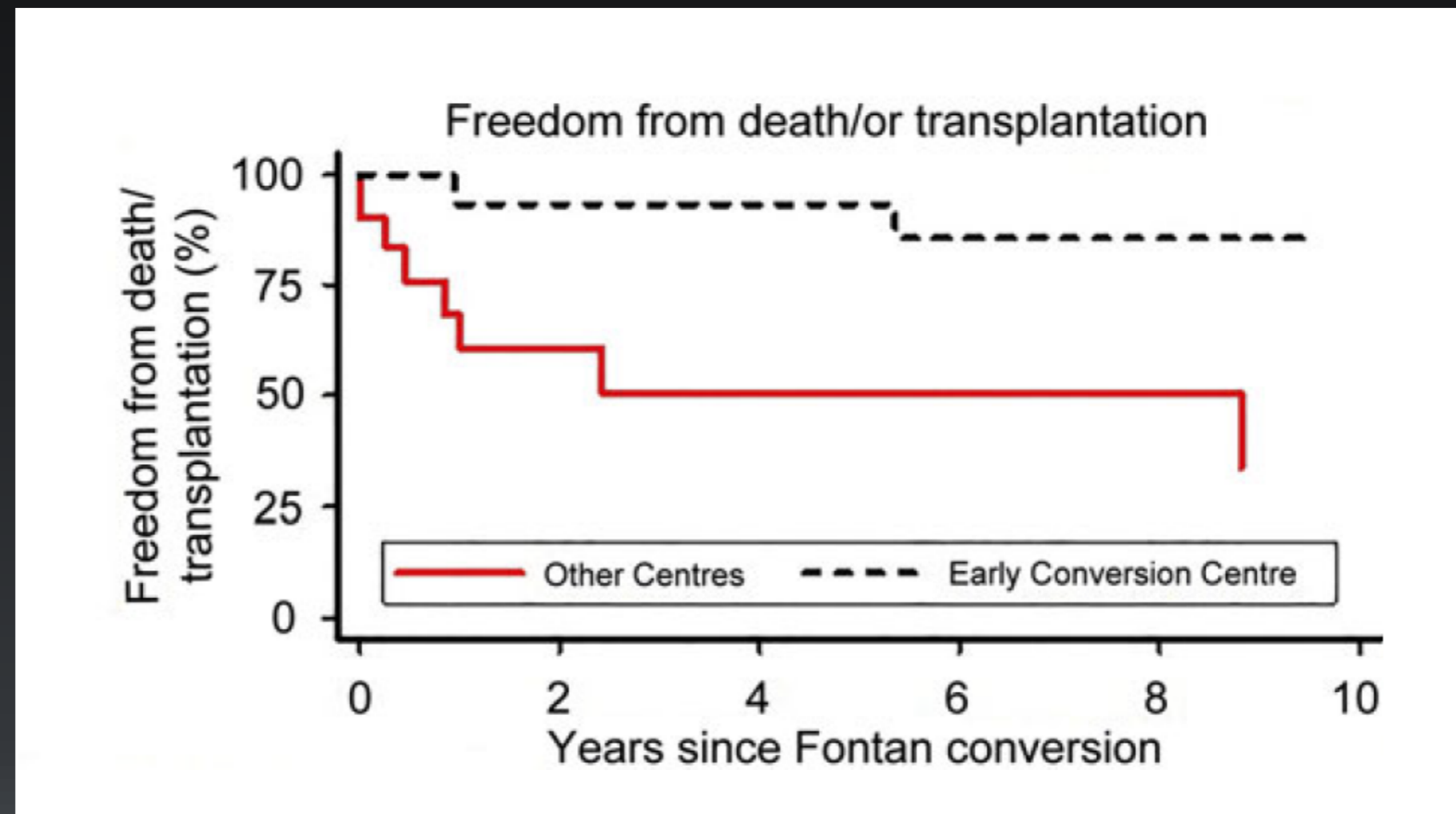
- Multi-Centre Reviews
  - Early mortality between 5 - 10%
  - Late death or transplant 10%
- Recent single centre reports (since 2010)
  - Early mortality 0 - 15%



# The Australia and NZ Experience of Fontan Conversion

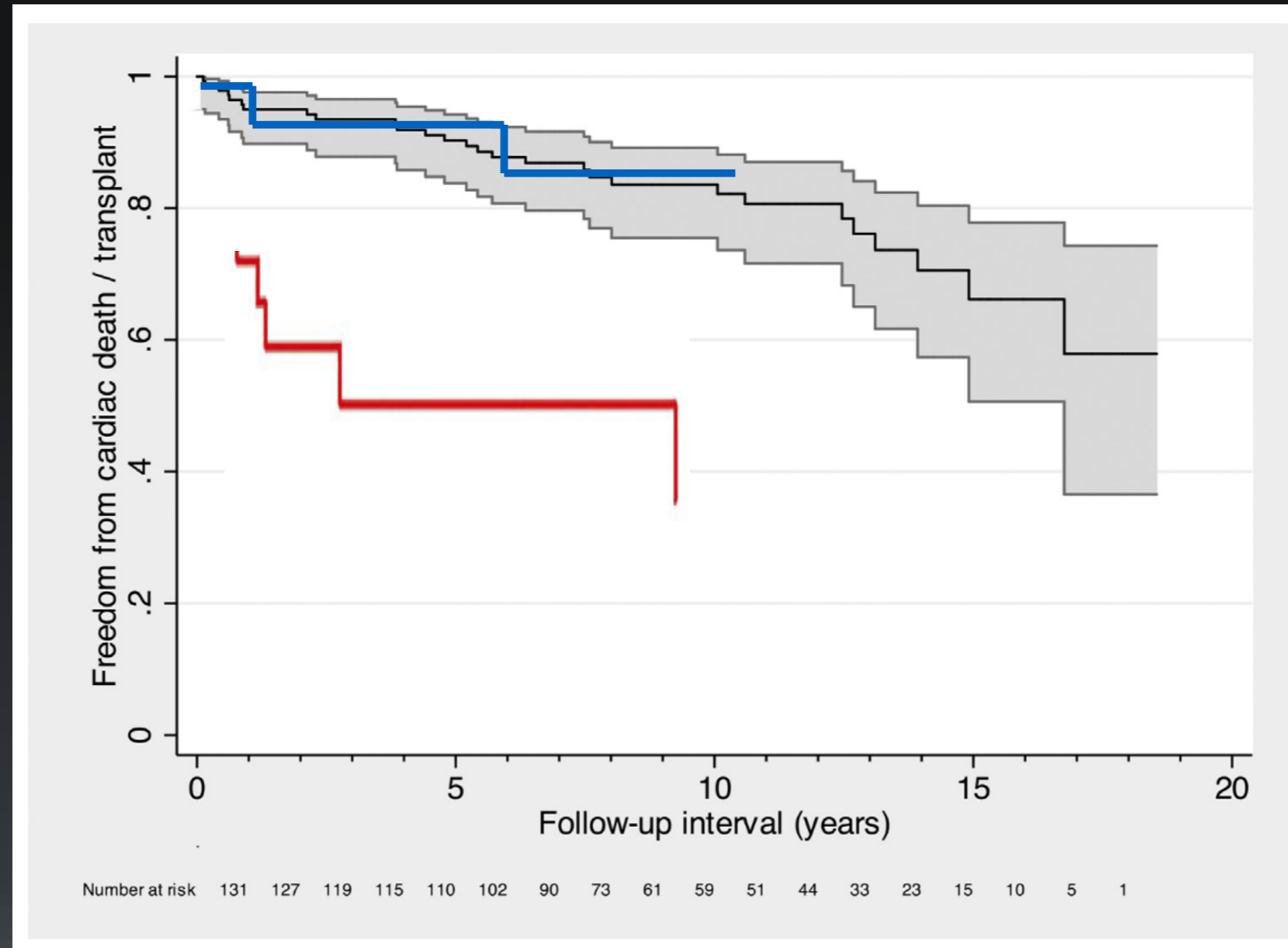
# “Early” vs “Late” Conversion Groups

## Transplant-free Survival



CL Poh et al. Ten-year outcomes of Fontan conversion in Australia and New Zealand demonstrate the superiority of a strategy of early conversion. *Eur J Cardiothorac Surg* 2016; 49; 530-535.

# Survival after Fontan Conversion: Deal, Mavroudis



Deal, BJ et al (Chicago). Intermediate-Term Outcome of 140 Consecutive Fontan Conversions With Arrhythmia Operations. *Ann Thorac Surg* 2016;101:717-24.

# “Early” vs “Late” Conversion Groups

## Pre-operative Variables

	Early Conversion (n=18)	Late Conversion (n=21)
Age at conversion	25.1 yr	23.7 yr
Time since first arrhythmia	2.9 yr	4.5 yr
NYHA Pre-op	2	3
Number of anti-arrhythmias	1	2

# “Early” vs “Late” Conversion

# Case

- DORV, PS, L-TGA, mitral atresia, large VSD
- Atrio-pulmonary Fontan procedure - aged 21 years
- Atrial arrhythmias from 2008 (age 41 years)
- EPS + RFA 2011 - continued to have arrhythmias

# Case

- RA and PA pressure 11-12 mmHg
- Transpulmonary gradient 6 mmHg, PVRi 3.3 U.m<sup>2</sup>
- Cardiac index 2.1 L/min/m<sup>2</sup>
- LVEF 41%, Moderate AR
- Normal liver function tests; Normal FVC, FEV<sub>1</sub>
- **Fontan conversion aged 45 years;**
  - **Discharged day 12 post-op**
  - **Remains well 5 years later**

Lower threshold

# ~~“Early”~~ Fontan Conversion

- **Not necessarily early in terms of age**
- **Perhaps early in terms of time since Fontan**
  - One of the two patients who died was the furthest out from the Fontan operation (33 years post)
- **Early in terms of not waiting for complications**



# Risk Factors

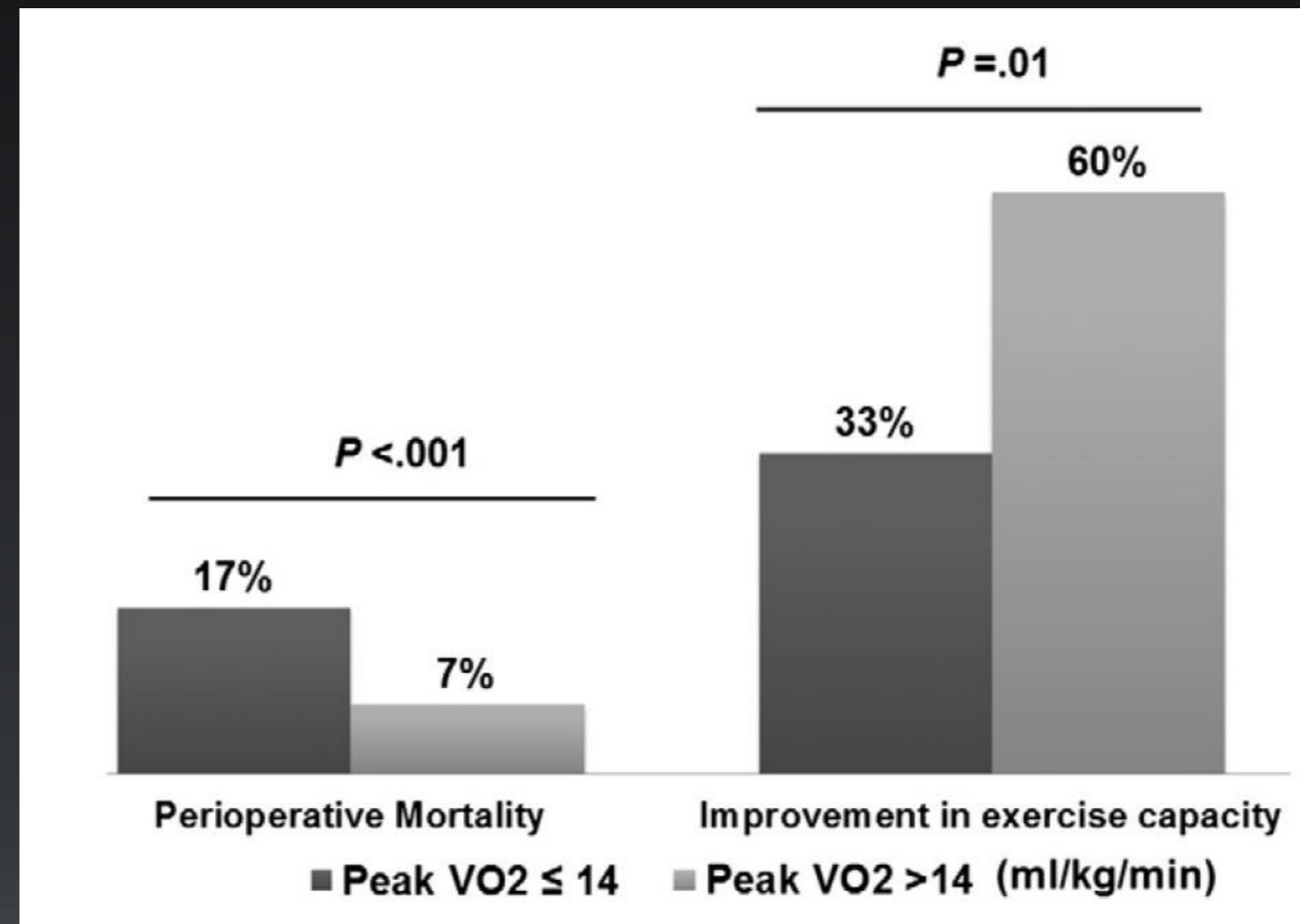
- PLE
- Severe ventricular dysfunction
- Ascites
- Cirrhosis, Liver dysfunction, HCC
- Severe AV valve regurgitation
- Non-left ventricle morphology
- Plastic bronchitis
- Renal insufficiency

# Risk Factors

*Table 3. Independent Risk Factors for Cardiac Death or Transplantation After Fontan Conversion*

Variable	Adjusted Hazard Ratio (95% CI)	<i>p</i> Value
Right or indeterminate ventricular morphology	5.71 (2.37–13.75)	<0.001
Ascites	3.69 (1.59–8.56)	0.002
Protein-losing enteropathy	4.93 (1.16–20.98)	0.03

# Peak Oxygen Uptake



Egbe AC et al (Mayo Clinic). When is the right time for Fontan conversion? The role of cardiopulmonary exercise test. International Journal of Cardiology. 2016;220:564-568.

# So when should we do Fontan Conversion?

- **Atrial arrhythmias**
  - Unless controlled easily by a single agent (?)
  - Definitely convert if amiodarone required
- **Patients requiring other surgical intervention for haemodynamic indications**
  - Valve surgery, VSD enlargement
- **Progressive symptoms / reduced  $\text{VO}_2$  etc**
  - Monitor carefully with CPET, Albumin, MRI etc

# And when should we not do Fontan Conversion?

- Factors to consider include:
  - **Protein losing enteropathy**
  - **Severe ventricular dysfunction** (unless potentially reversible)
  - Other organ system disease
    - **Cirrhosis, renal impairment, pulmonary disease**

...But remember suboptimal transplant outcomes

# Conclusions

- Atrial arrhythmias often become progressively harder to control medically or with ablation
- Fontan conversion can be performed with an acceptable mortality risk and with reasonably good medium-term outcomes
- Lower threshold for Fontan conversion is likely to reduce morbidity and mortality

# Acknowledgements

- Kathryn Rice, Chin Poh, Kirsten Finucane
- Fontan Registry: Megan Upjohn, Judith Barry
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- CAMRI: Anna Lydon and the team
- Starship / GLH Cardiologists & Cardiac Surgeons
- Starship Echo team