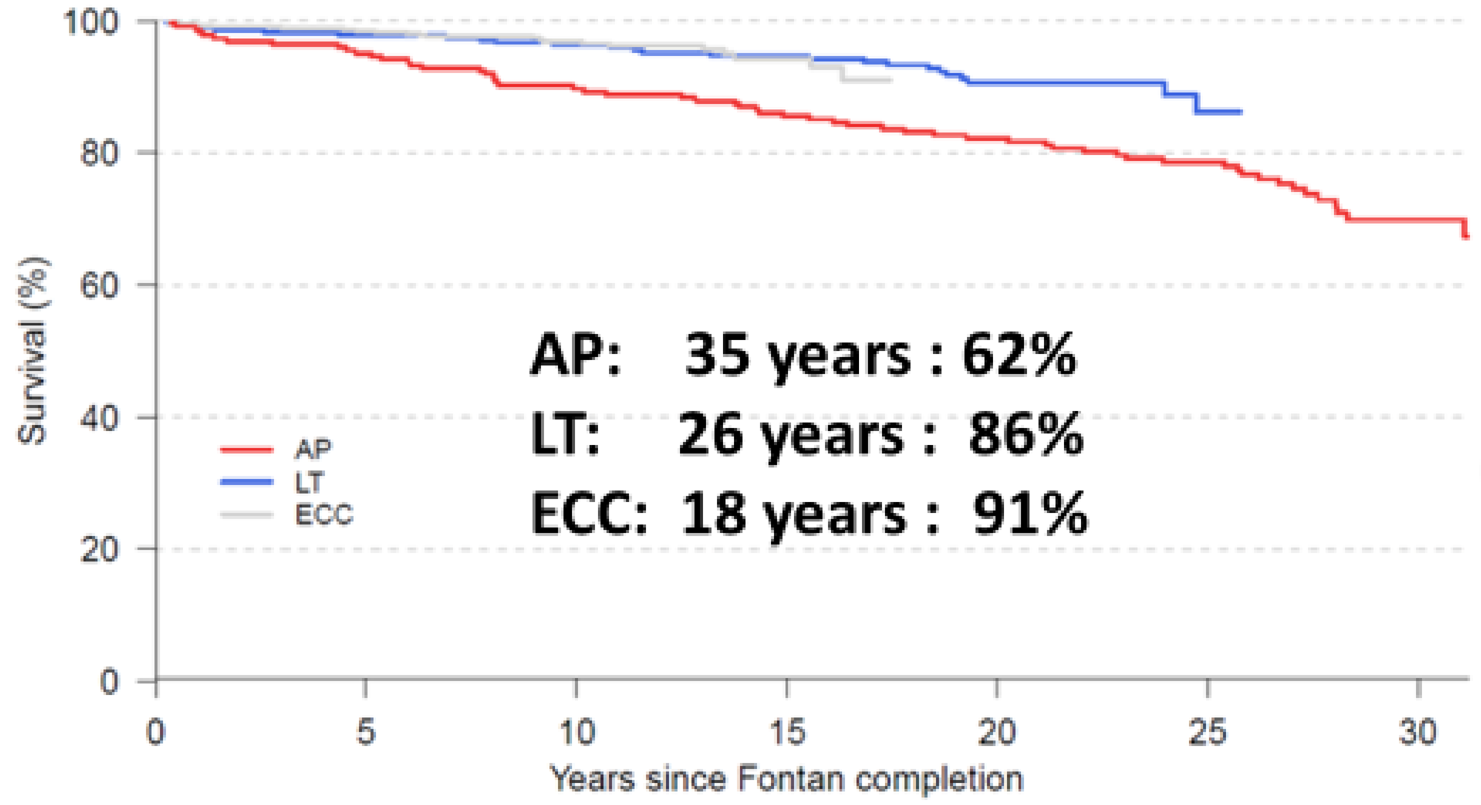
Late Reoperations in Fontan survivors

Indications and Outcomes

Yves d'Udekem, Kirsten Finucane



Number at risk

AP	225	212	198	182	166	132	39
LT	289	264	247	217	144	29	
ECC	984	596	306	96			

Figure 6 Kaplan Meier Survival Curve by type of Fontan operation

Note: AP=Atriopulmonary Connection; LT=Lateral Tunnel; ECC=Extracardiac Conduit

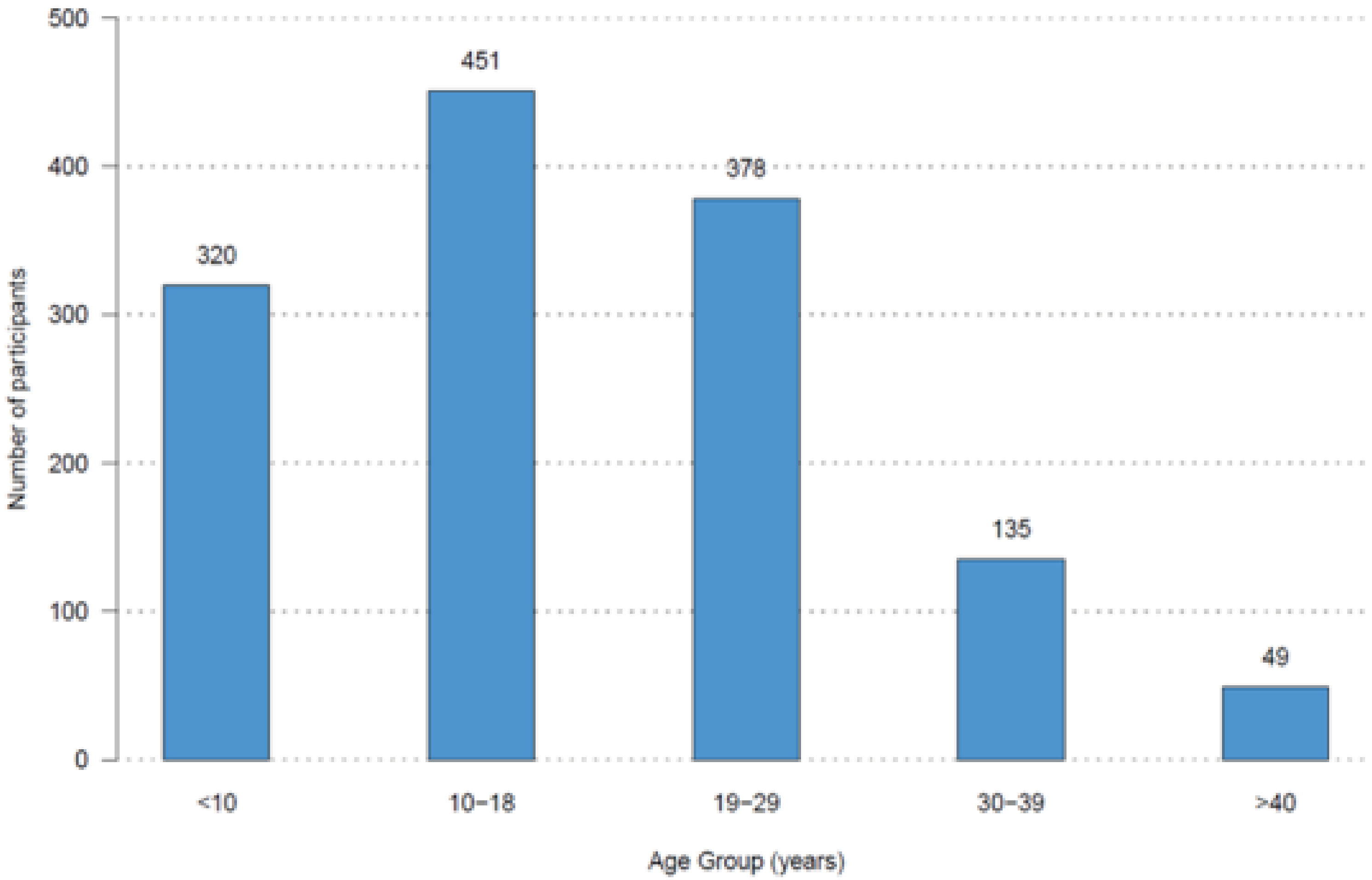


Figure 1 Current age of Fontan Registry participants (*N* = 1333)

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Longitudinal Outcomes of Patients With Single Ventricle After the Fontan Procedure

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- 546 patients → 343 patients
- 21.2 +/- 4 years of age at last follow-up
- Mean of 9.4 +/- 0.4 years following the Fontan 1 study
- Mean of 17.8 +/- 3.4 years after Fontan procedures.

Fontan REGISTRY

TABLE 1 Complications and Interventions After Fontan Procedures

	Fontan 1 (N = 546)	Fontan 2 (N = 416)	Fontan 3 (N = 373)
Time since Fontan procedure, yrs	8.7 ± 3.4	15.2 ± 3.4	17.8 ± 3.4
Cardiac surgery	23	28	32
Catheter intervention	48	57	62
Electronic device	13	13	16
Stroke	2	2	4
Seizures	3	5	7
Thrombosis	8	9	12
Arrhythmia treatment	20	28	32
Protein-losing enteropathy	4	7	8
Cirrhosis	0.4	4	8
Plastic bronchitis	0.1	0.5	1

Values are mean ± SD or %.

Congenital Heart Disease

Surgical and Catheter-Based Reinterventions Are Common in Long-Term Survivors of the Fontan Operation

Tacy E. Downing, MD; Kiona Y. Allen, MD; David J. Goldberg, MD; Lindsay S. Rogers, MD; Chitra Ravishankar, MD; Jack Rychik, MD; Stephanie Fuller, MD; Lisa M. Montenegro, MD; James M. Steven, MD; Matthew J. Gillespie, MD; Jonathan J. Rome, MD; Thomas L. Spray, MD; Susan C. Nicolson, MD; J. William Gaynor, MD; Andrew C. Glatz, MD, MSCE

(*Circ Cardiovasc Interv*. 2017;10:e004924. DOI: 10.1161/CIRCINTERVENTIONS.116.004924.)

773 patients

Information available on 70% of patients

Freedom from reoperation was 69% at 15 years and 63% at 20 years

The most common operations were pacemaker placement and Fontan revision

Fontan REGISTRY

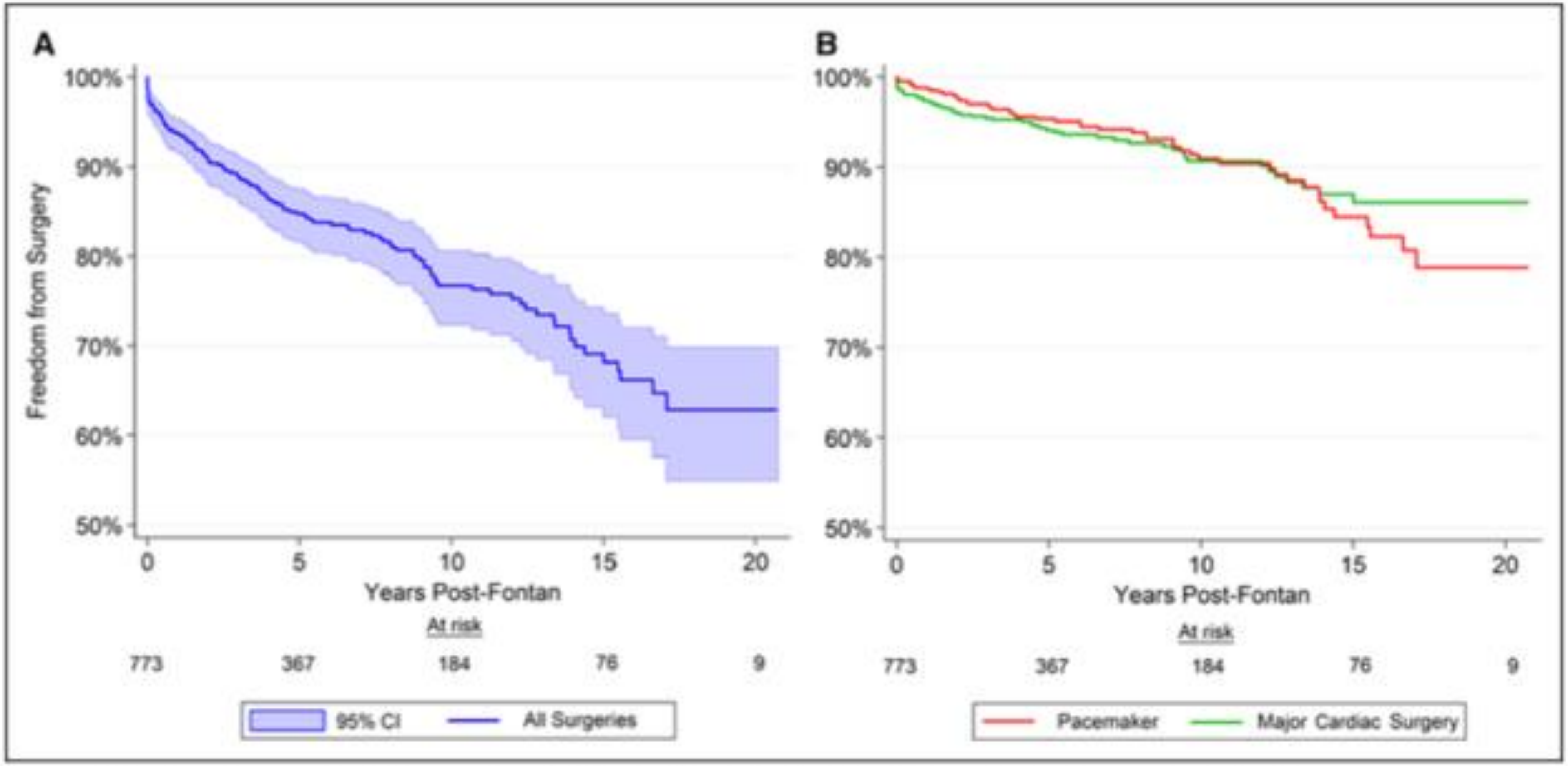


Figure 2. Freedom from surgical reintervention. **A**, Kaplan–Meier freedom from first surgical reintervention. Shaded area represents 95% confidence intervals (CIs) of the survival function. **B**, Kaplan–Meier freedom from first pacemaker and first major cardiac surgery in patients with intact Fontan. Y axis in both panels begins at 50%.

Fontan REGISTRY

The Australia and New Zealand Fontan Registry

European Journal of Cardio-Thoracic Surgery (2015) 1–6
doi:10.1093/ejcts/ezv112

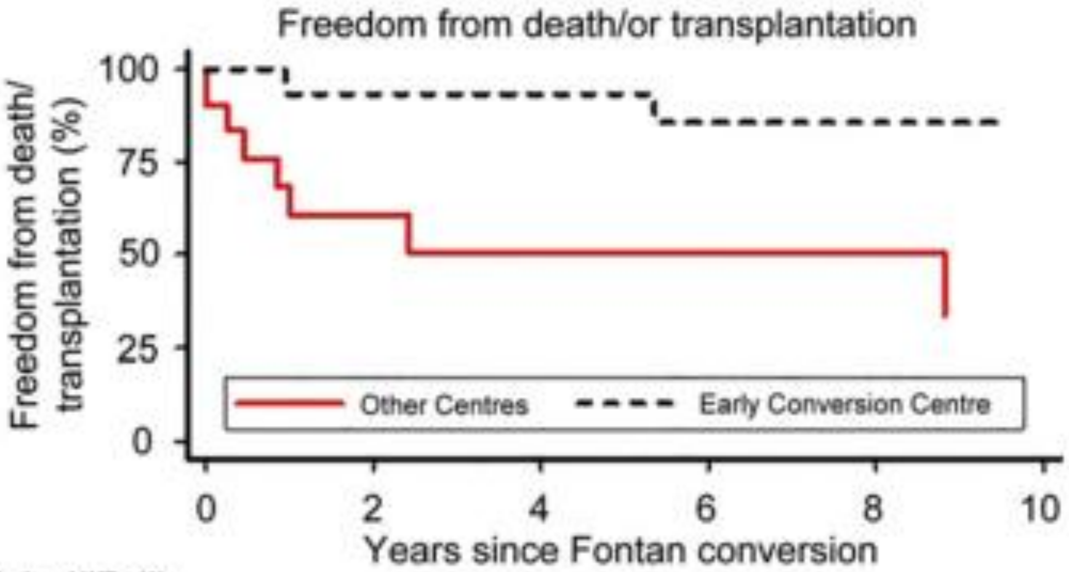
ORIGINAL ARTICLE

CONGENITAL

Cite this article as: Poh CL, Cochrane A, Galati JC, Bullock A, Celermajer DS, Gentles T *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 2015; doi:10.1093/ejcts/ezv112.

Ten-year outcomes of Fontan conversion in Australia and New Zealand demonstrate the superiority of a strategy of early conversion†

Chin Leng Poh[a,b], Andrew Cochrane[c], John C. Galati[a,d], Andrew Bullock[e], David S. Celermajer[f,g], Tom Gentles[h], Karin du Plessis[a], David S. Winlaw[i], Tim Hornung[h], Kirsten Finucane[h] and Yves d'Udekem[a,b,i,*]



# at risk (#Fail)											
Other Centres	20	(6)	6	(1)	4	(0)	4	(0)	3	(1)	1
Early Convers.	19	(1)	12	(0)	12	(1)	10	(0)	7	(0)	5

Figure 1: Kaplan–Meier survival curve of freedom of death and transplantation in the centre with a low threshold for Fontan conversion versus other centres.

Fontan REGISTRY

Fontan Conversion in Auckland:

- Interval since first documented arrhythmia 2.9 vs 4.0 years
- Average NYHA class 2 vs 3
- Mean no of preop antiarrhythmics 1 vs 2

ALSO

- 19 in one NZ centre vs 20 in 5 centres
- NZ has integrated paediatric/adult congenital service- CVICU, anaesthesia
- Good surveillance of adult congenital population
- 66% female in our series (vs 33% in Australian centres)

Congenital heart disease

ORIGINAL ARTICLE

Heart transplantation in Fontan patients across Australia and New Zealand

Editor's choice
Scan to access more free content

William Y Shi,[1] Matthew S Yong,[2] David C McGiffin,[2] Pankaj Jain,[3] Peter N Ruygrok,[4] Silvana F Marasco,[2] Kirsten Finucane,[4] Anne Keogh,[3] Yves d'Udekem,[1] Robert G Weintraub,[1] Igor E Konstantinov[1]

34 transplantations out of 1369 patients (2.5 %)

16 patients over 18 yrs, oldest age 51.

3 prior procedures (1-5)

11.5 yrs post Fontan (.23-33)

91% 1yr, 78% 5yr, 71% 10yr survival

To cite: Shi WY, Yong MS, McGiffin DC, *et al. Heart* 2016;**102**:1120–1126.

REINTERVENTIONS

- Out of 1524 patients (1428 pts with available data of F-up)

- 444 patients had a reintervention
  - 38 patients had **early** reinterventions (10 cath procedures)
  - 413 patients had a **late** reintervention

Reinterventions, n	Patients
1	284
2	79
3	29
4	18
5	1
6	1
12	1

Fontan REGISTRY

Catheter late reinterventions

Catheter-based reintervention, n	**214**
Fenestration closure	130
Catheter-based ablation of arrhythmia	47
Balloon dilation and stenting of PAs	27
Coiling of collaterals	29
Stenting of LAD	1
Device occlusion of left or right SVC	4
Device occlusion of ASD	2
Stenting of Fontan conduit	1
Balloon dilation of Damus-Kaye-Stansel	1

Late reoperation, n	**199**
Fontan circuit revision	16
Conversion to ECC Fontan	49
Fontan takedown	11
Transplantation	31
Fenestration creation/enlargement	8
Maze procedure/surgical cryoablation	36
AV valve repair	18
AV valve replacement	10
PA reconstruction	13
Pleurodesis	10
Diaphragm plication	3
SAS resection	15
VSD enlargement	6
Bulboventricular foramen enlargement	3
Aortic repair (root, ascending, arch)	13
Semilunar valve repair	2
Semilunar valve replacement	3
Damus-Kaye-Stansel procedure	10
Other procedures	
AV valve closure/re-closure	9
RA reduction	3
Thoracic duct ligation	2
SVC ligation	5
Conversion to lateral tunnel Fontan	1
Other	20

Fontan REGISTRY

32 pts Historical procedures

- 10 ***Damus-Kaye-Stansel*** (med 3y; 3m-9y)
  - 1 death

- 6 ***VSD enlargements*** (med 1y; 1m-29y)
  - 1 death

- 15 ***subaortic stenosis*** (med 12y; 1-26y)
  - One death

Fontan REGISTRY

Outcomes of patients born with single-ventricle physiology and aortic arch obstruction: The 26-year Melbourne experience

Melissa G. Y. Lee, BMedSc,[a] Christian P. Brizard, MD,[a] John C. Galati, PhD,[b]
Ajay J. Iyengar, MBBS, BMedSc,[a] Sandeep S. Rakhra, MBBS, BMedSc,[a]
Igor E. Konstantinov, MD, PhD,[a] Andreas Pflaumer, MD,[c] and Yves d'Udekem, MD, PhD[a]

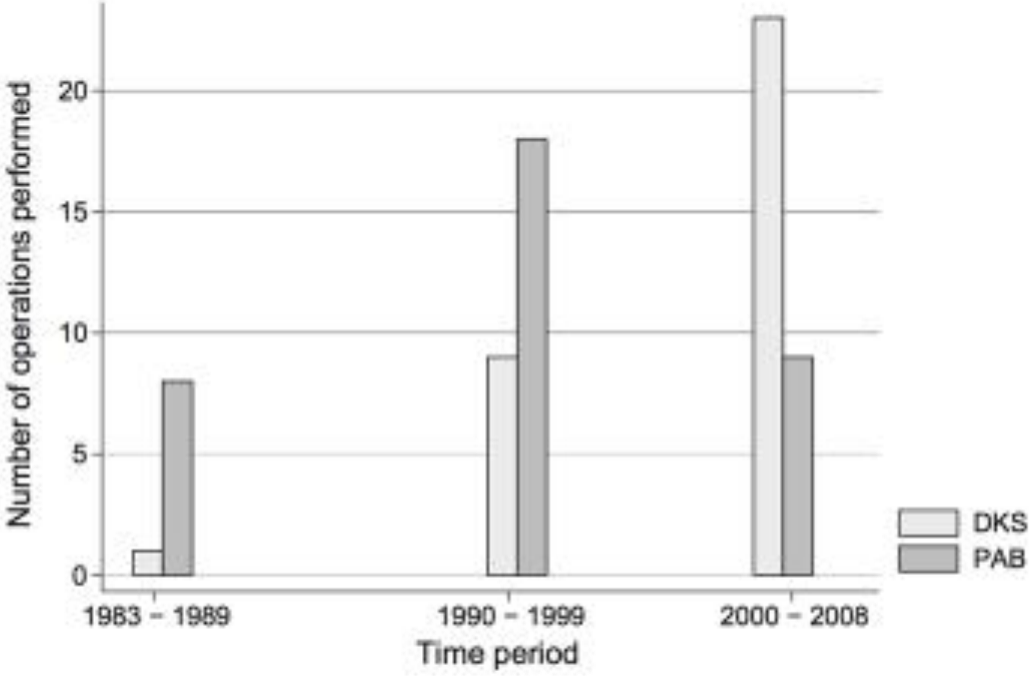


FIGURE 1. Type of initial neonatal palliation performed by era. *DKS*, Damus-Kaye-Stansel; *PAB*, pulmonary artery banding.

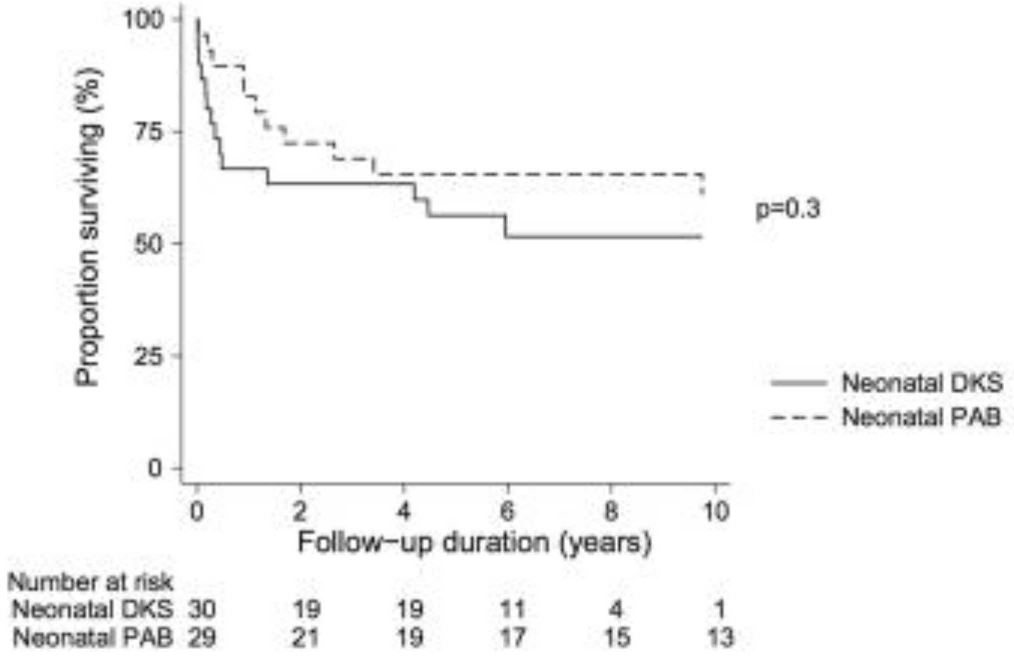


FIGURE 3. Kaplan-Meier survival curve. *DKS*, Damus-Kaye-Stansel; *PAB*, pulmonary artery banding.

Optimization of Fontan

- 16 ***Revision of Fontan circuit*** (med 3 y; 1m-16y)
  - 7 deaths (med 16 y; 5-28 y)

- 8 ***Creation/Enlargement of fenestration.***
  - 1 death early post fenestr.

- 13 ***PA reconstruction*** (med 5y; 3m-16y)
  - 2 deaths

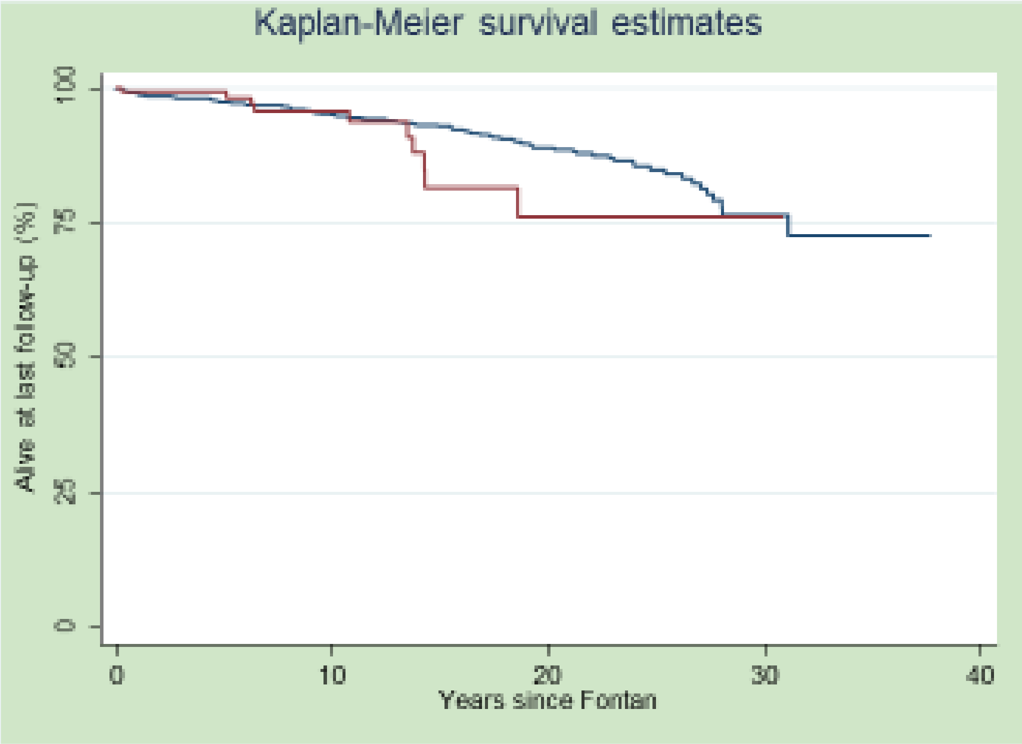
Fontan REGISTRY

AUGMENTATION OF THE PULMONARY ARTERIES IS NOT ASSOCIATED WITH WORSE LONGER-TERM OUTCOME:

A Propensity Matched Analysis From The Australia And New Zealand Fontan Registry (153 pts)

Results – Deaths (Unmatched)

- No significant difference
- 10 (6.5%) deaths in the pulmonary artery augmentation group
- 95 (7.4%) deaths in the non-augmentation group
- Hazard ratio for death 1.35, 95%CI 0.70–2.60, p = 0.366



HR 1.35, 95% CI 0.70–2.60, p=0.366
Red = pulmonary artery augmentation, Blue = non-augmentation group

29 pts Valve interventions

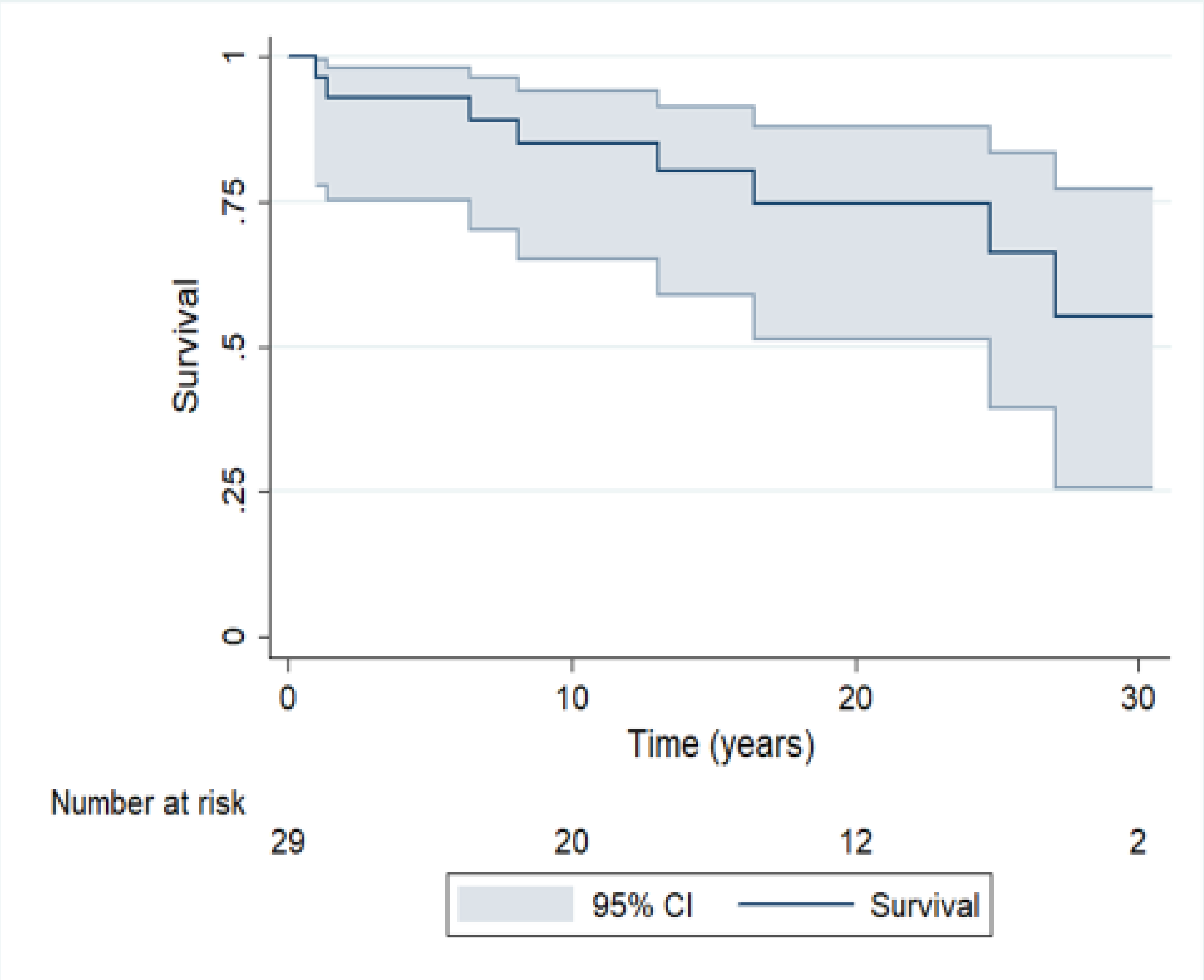
- 18 ***AV valve repairs*** (med 4y; 2m- 24y)
- 6 deaths

- 10 ***AV valve replacements*** (med 1y; 1m-29y)
- 3 deaths

- 5 ***semi-lunar valve ops*** (2 repairs/3 replacements)
- One death

Fontan REGISTRY

Survival of pts undergoing valvar reintervention



European Journal of Cardio-Thoracic Surgery 51 (2017) 1037–1043
doi:10.1093/ejcts/ezx025 Advance Access publication 23 March 2017

ORIGINAL ARTICLE

Cite this article as: King G, Gentles TL, Winlaw DS, Cordina R, Bullock A, Grigg LE *et al.* Common atrioventricular valve failure during single ventricle palliation. Eur J Cardiothorac Surg 2017;51:1037–43.

Common atrioventricular valve failure during single ventricle palliation†

Gregory King[a], Thomas L. Gentles[b], David S. Winlaw[c], Rachel Cordina[d], Andrew Bullock[e], Leeanne E. Grigg[f], Nelson Alphonso[g], Dorothy J. Radford[h], Diana Zannino[i], Edward Buratto[a,i,j] and Yves d'Udekem[a,i,j,*]

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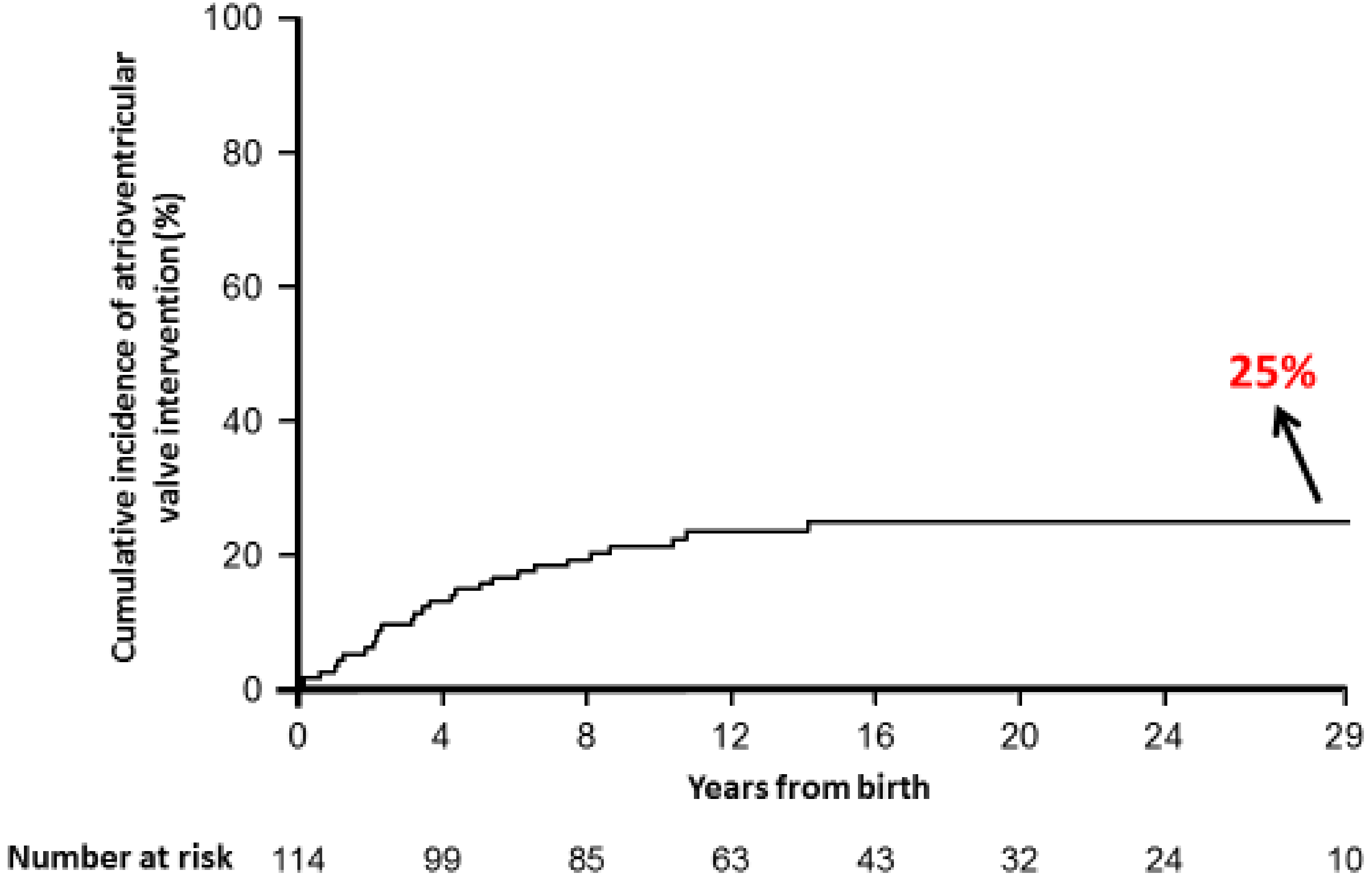
CONGENITAL

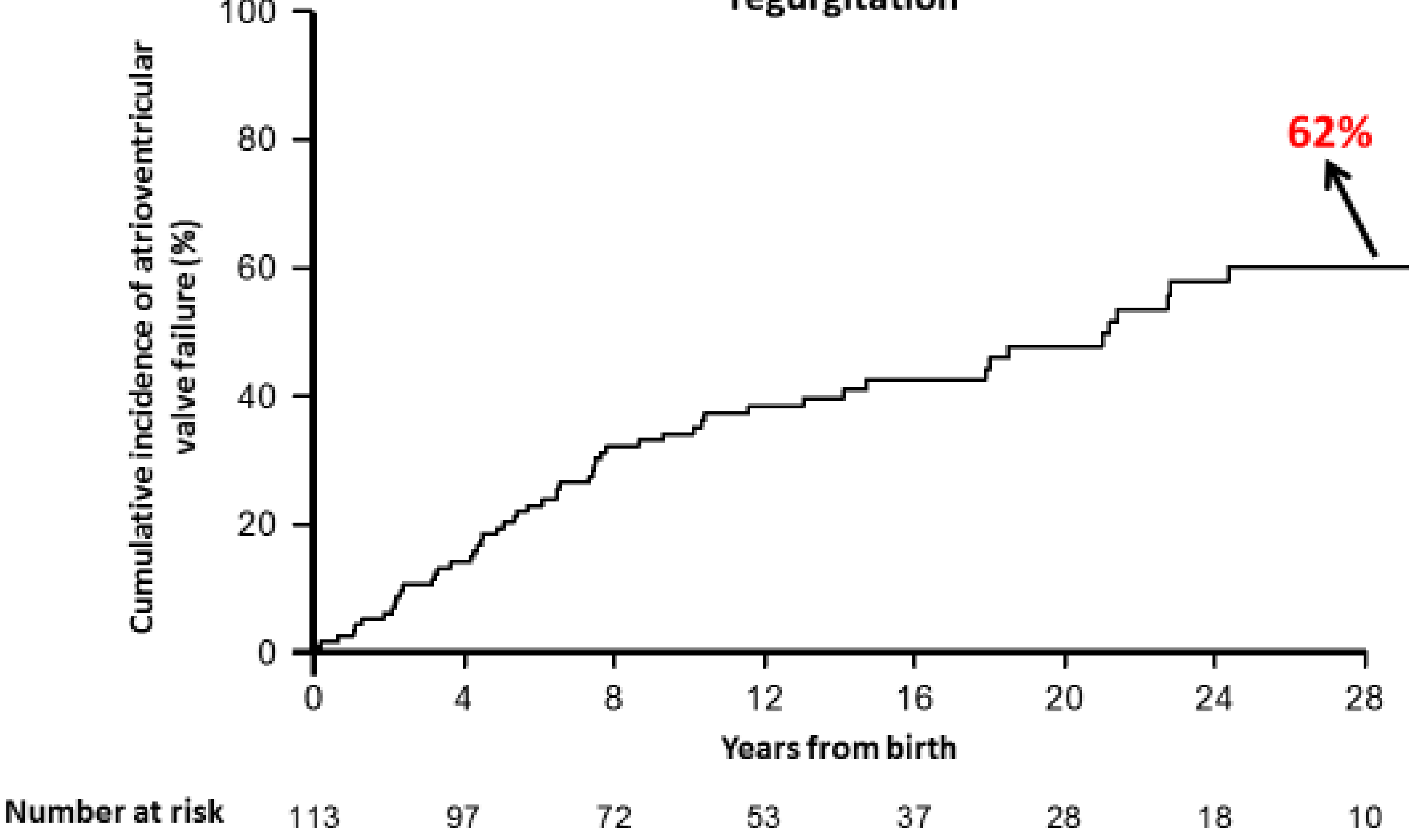
Fontan REGISTRY

Cumulative incidence of atrioventricular valve repair/replacement

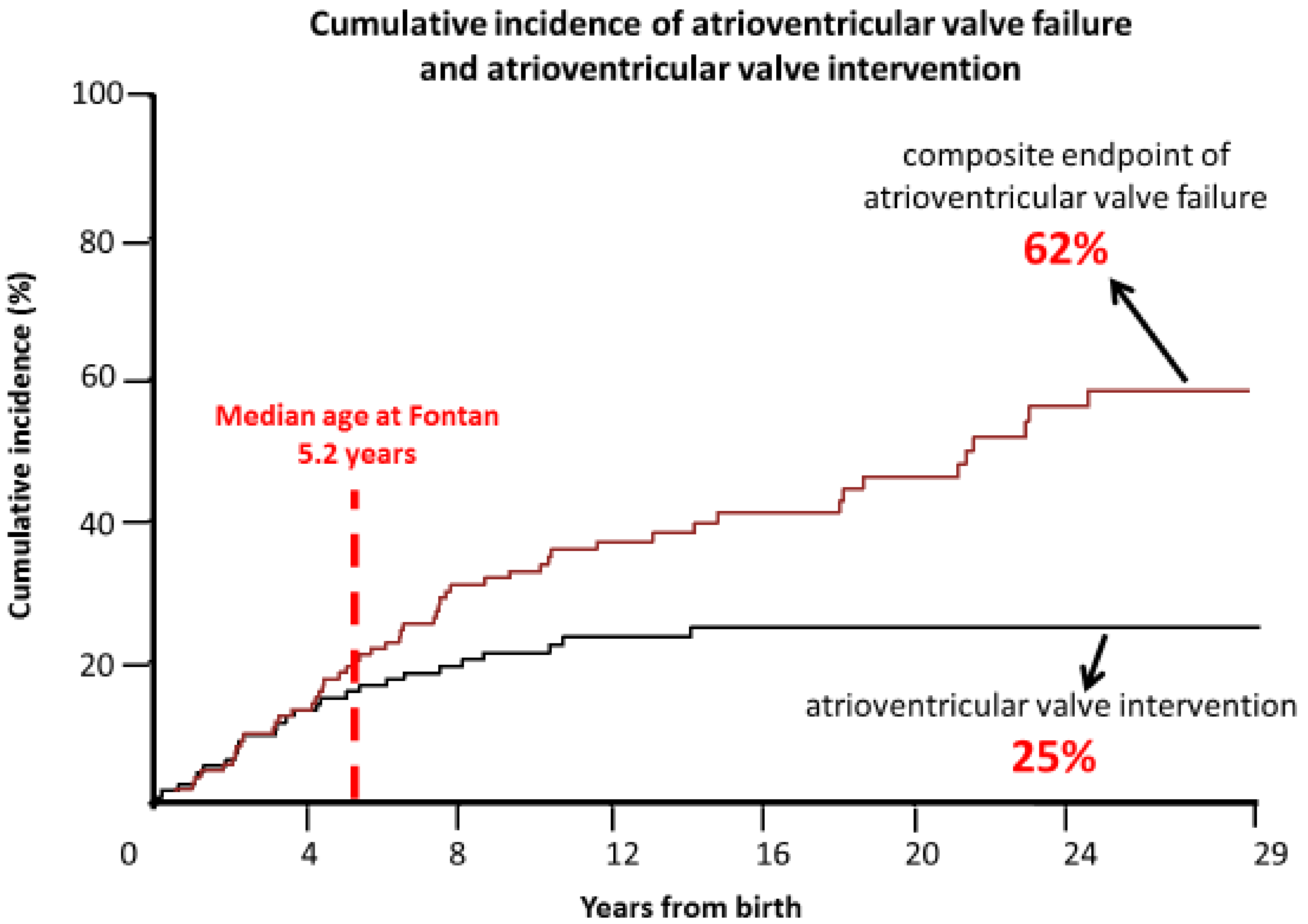


Fontan REGISTRY

Cumulative Incidence of atrioventricular valve repair/replacement or moderate or greater regurgitation



Fontan REGISTRY



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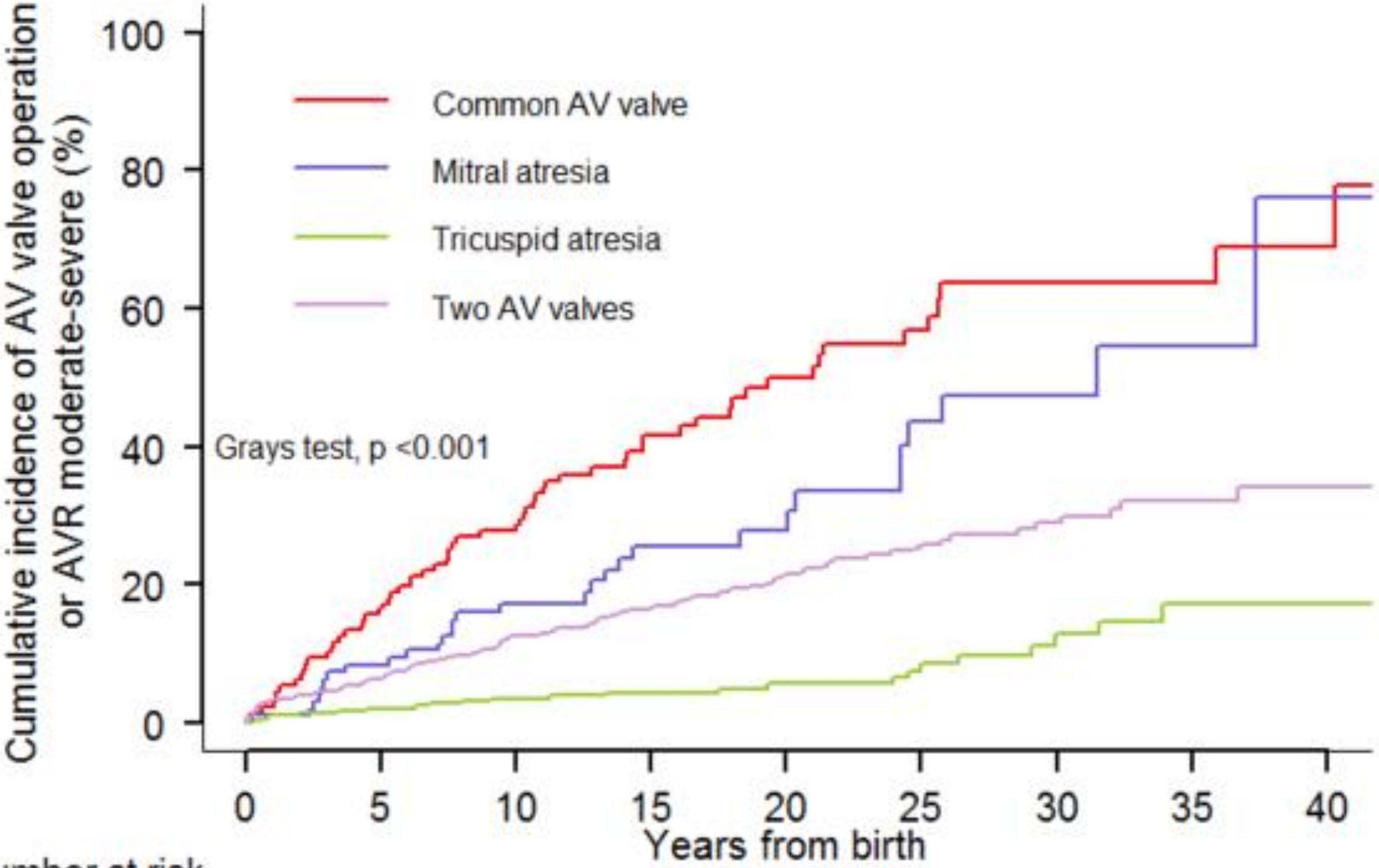


Number at risk									
Common AV valve	127	106	80	47	32	20	9	6	3
Mitral atresia	95	87	62	42	24	16	8	2	1
Tricuspid atresia	284	278	242	190	132	83	46	27	14
Two AV valves	672	627	474	322	197	116	63	31	13

Fontan REGISTRY

13 aortic interventions

- 8 aortic root/ascending aorta replacement
- 5 distal aortic arch repair
- 1 thoracic aorta repair

➢ One death

Double inlet Left Ventricle: conduction pathway

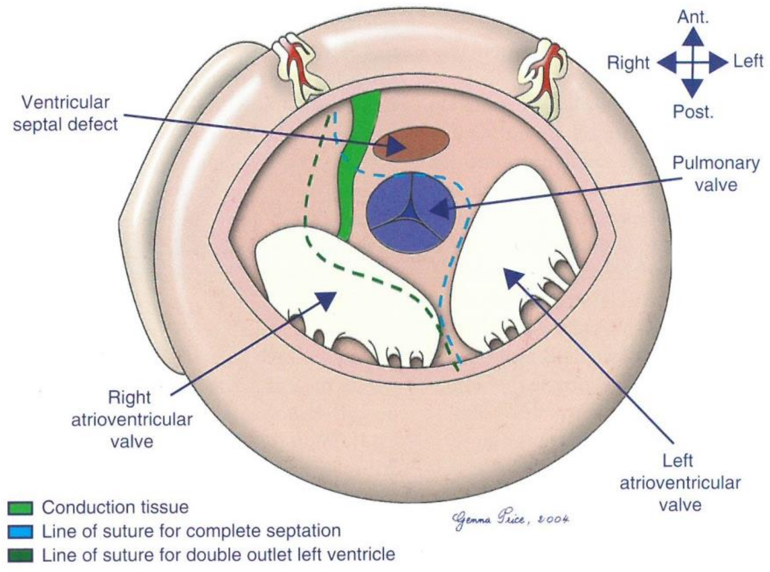


Fig. 8.14 In this cartoon, the dominant left ventricle with double inlet is seen as through a fishmouth incision in the apex of the ventricle. The conduction axis is marked in green (see also Fig. 8.4). The potential line for complete septation, placing the pulmonary trunk in communication with the systemic venous return, (blue dotted line) crosses the conduction axis. To avoid the conduction axis (green dotted line), it is necessary to deviate the site of septation so that both outflow tracts remain in communication with the pulmonary venous return through the left atrioventricular valve.

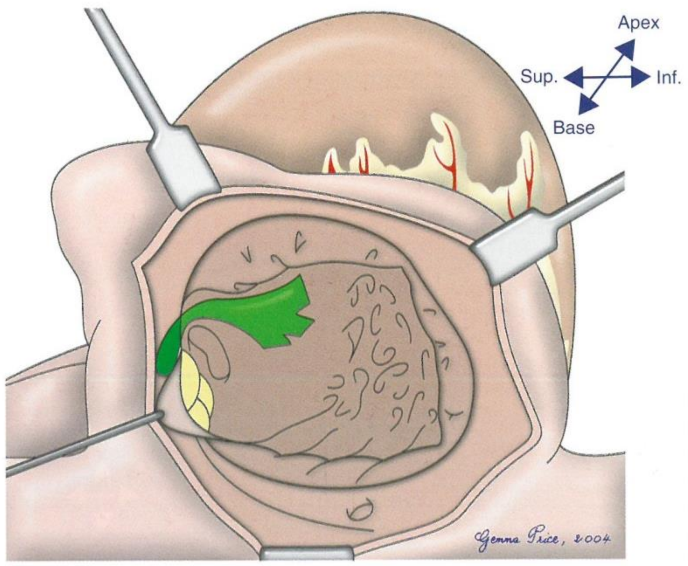
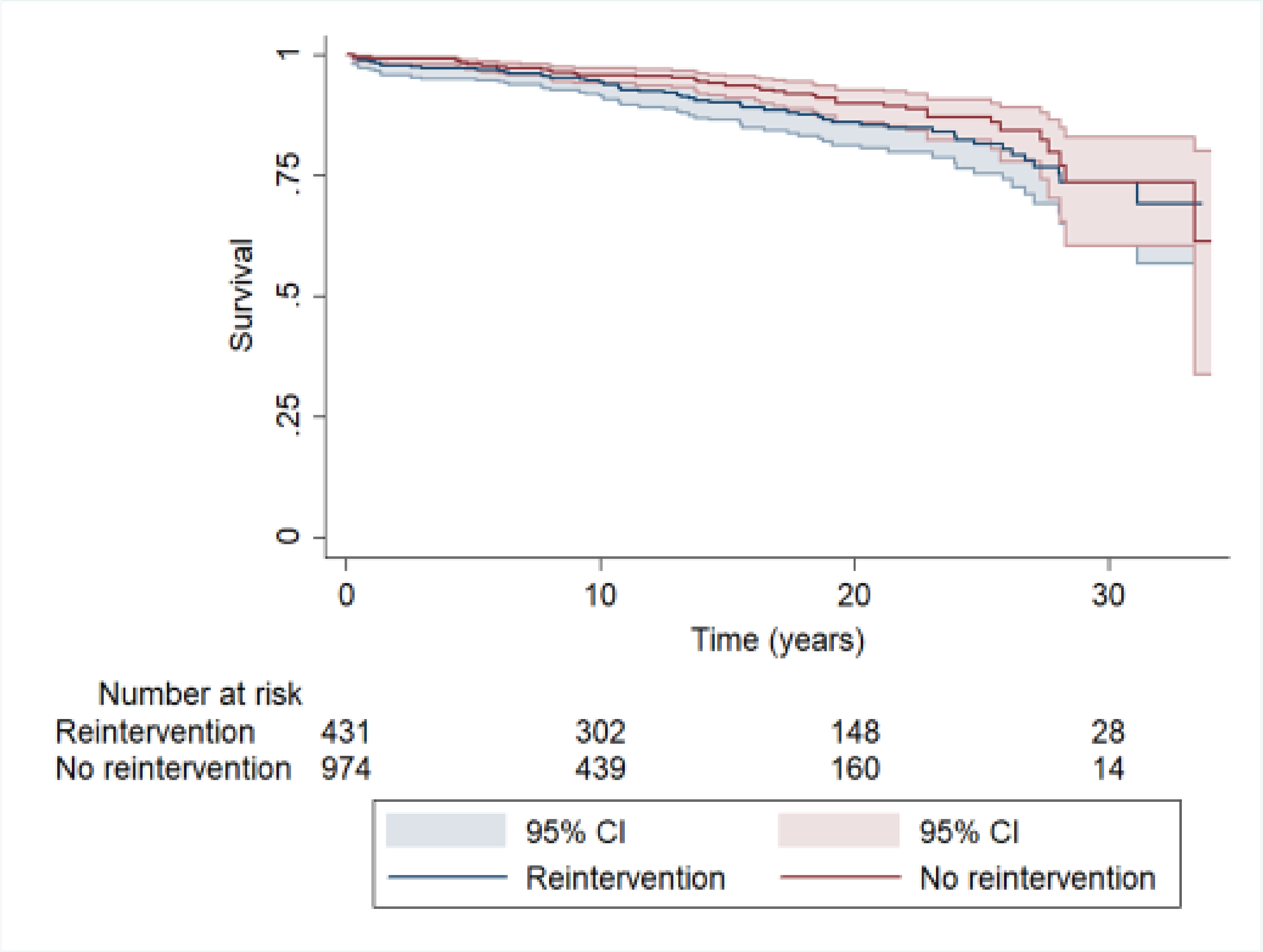


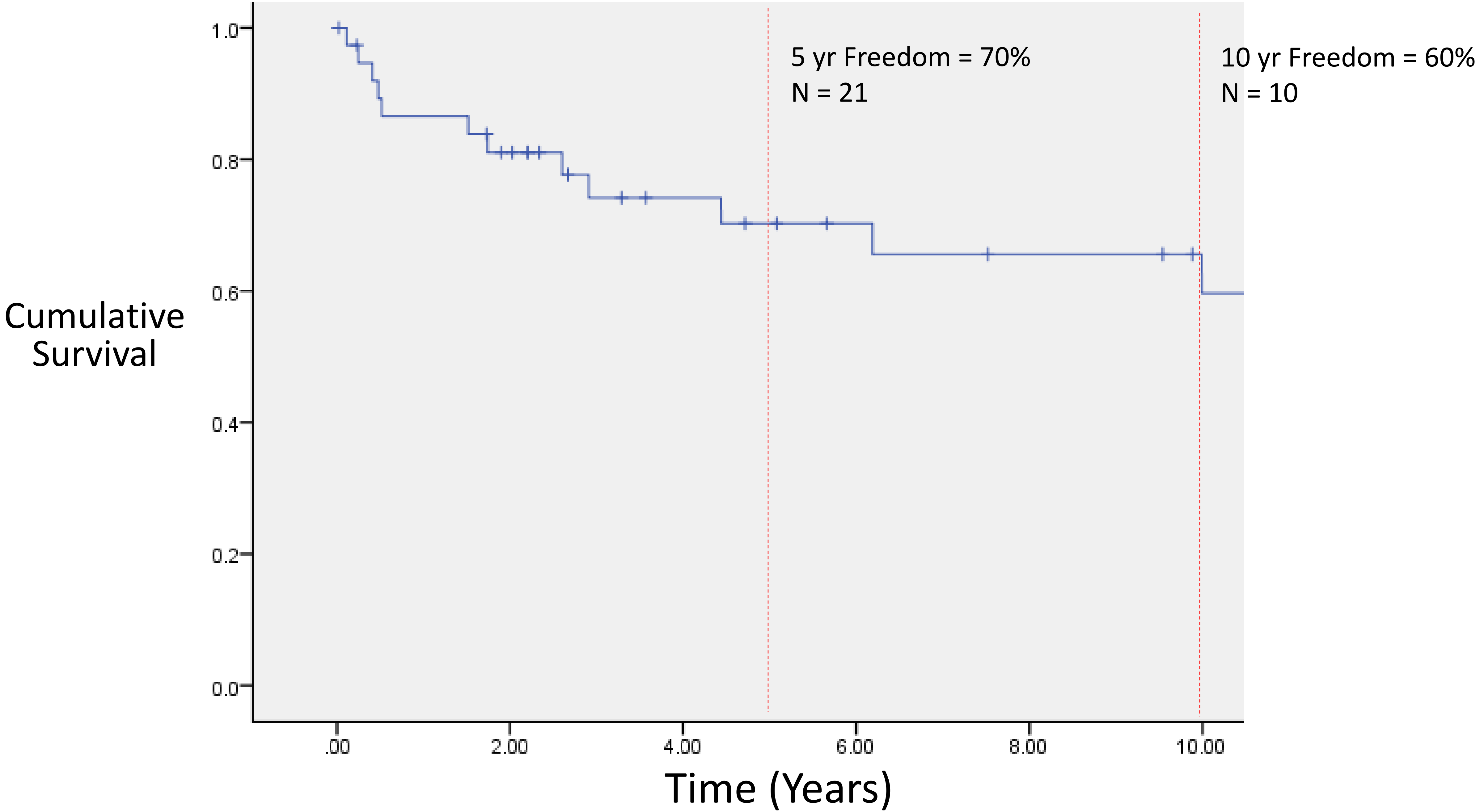
Fig. 8.15 This cartoon, again drawn in surgical orientation, shows the view of the dominant left ventricle in the setting of double inlet left ventricle with discordant ventriculo-arterial connections, and the course of the conduction axis, in green, as it would be seen by the surgeon working through the right atrioventricular valve. The leaflets of the pulmonary valve are coloured yellow.

Survival comparison: Reintervention vs no Reintervention



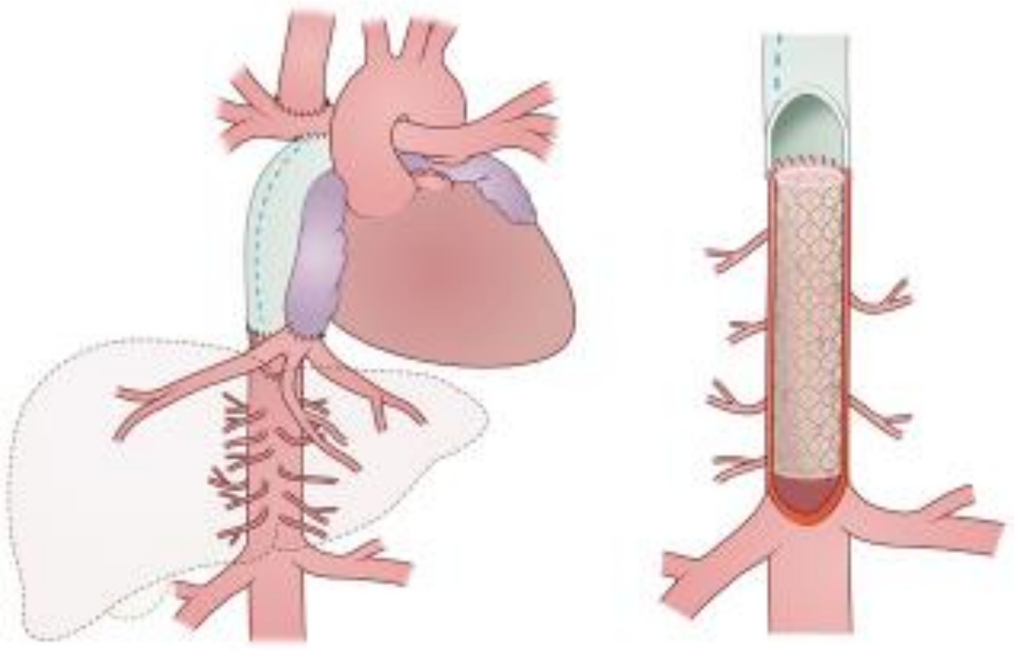
Fontan REGISTRY

Freedom from failure after developing PLE/PB (n=44)



Hepatic Vein Diversion

- Hepatic Veins are connected straight to RA
- Patch repair or stent to close entry of hepatic veins.
- 1 case in this series, 2 done in total at our centre
- Drastic resolution in symptoms (NYHA >III → NYHA II or I)
- Saturations at 75%



Courtesy C. Brizard, Department of Cardiothoracic Surgery, Royal Children's Hospital Melbourne.

Decompression of Thoracic Duct: New Approach for the Treatment of Failing Fontan

Viktor Hraška, MD

German Pediatric Heart Center, Sankt Augustin, Germany

Ann Thorac Surg 2013;96:709–11

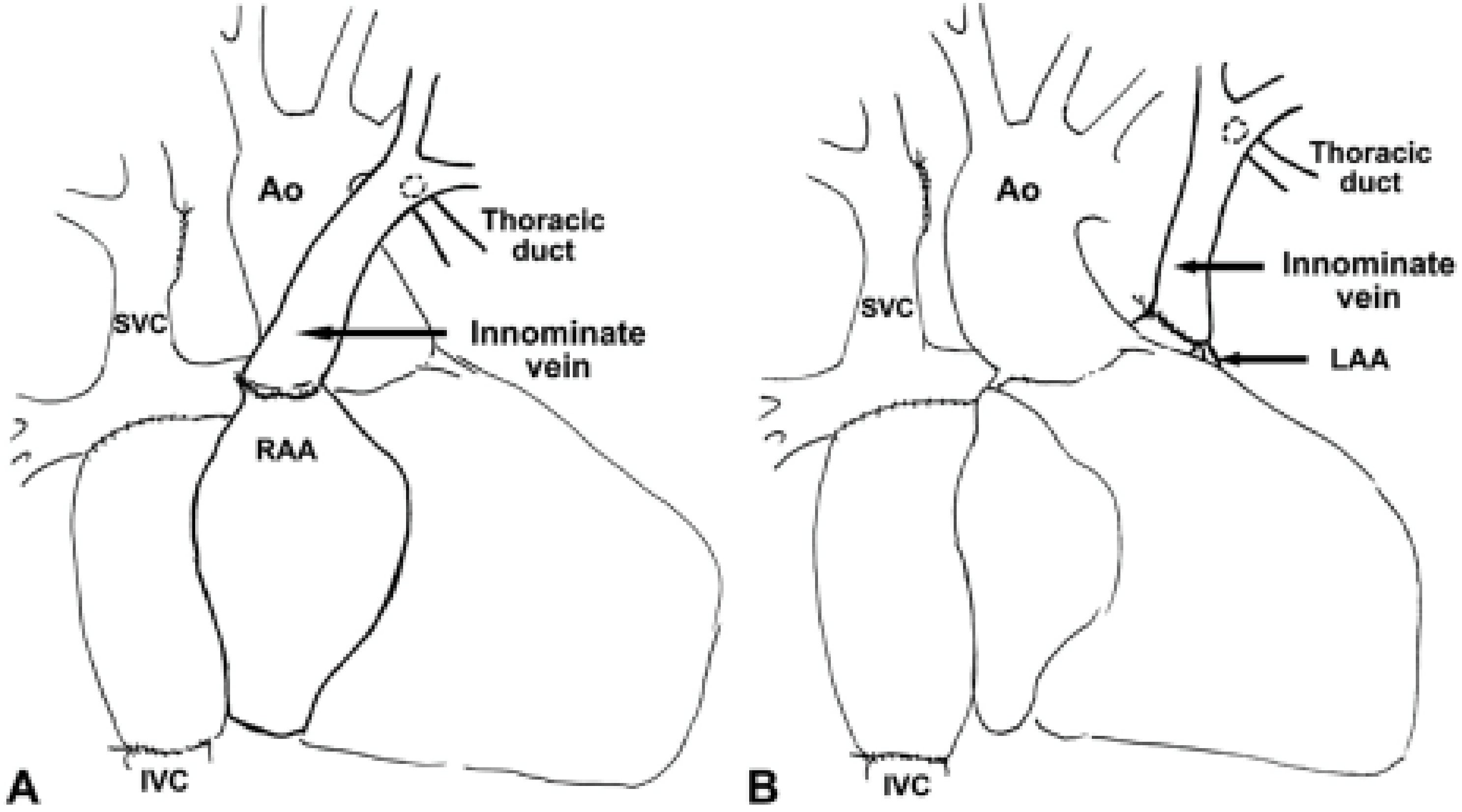


Fig 1. Innominate vein is directly anastomosed with (A) right or (B) left atrial appendage. (Ao = aorta; IVC = inferior vena cava; LAA = left atrial appendage; RAA = right atrial appendage; SVC = superior vena cava.)

Lymphopenia

MR lymphangiogram

Evaluation of the lymphatic circulation

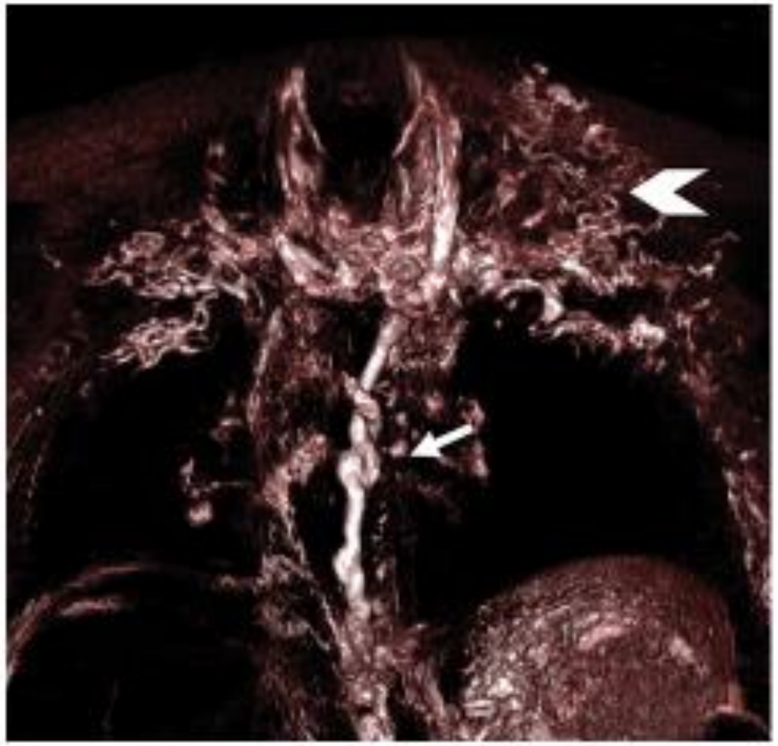


FIGURE 2 Pulmonary Lymphatic Perfusion Syndrome

Y. Dori

J Am Coll Cardiol 2017;69:2929–37

J Am Coll Cardiol 2017;69:2410–22

Preparation for Fontan Reoperation:

- Study the original morphology: position of AV bundle, valve leaflets
- Read the old operation notes
- Check for Aquired vascular lesions, eg venous occlusions, false aneurysms, diaphragm eventration, patch calcification
- Check for occult liver or renal dysfunction- high flow perfusion
- Strategise , keep crossclamp time to a minimum
- Consider arrhythmia surgery for every case but preserve node function
- Consider pacing leads with/without unit insertion for every case
- Optimise pacing to promote ventricular synchrony


MOSSIMO
Originals
NO SENSE

Fontan
REGISTRY

“Are we reoperating our patients with a Fontan circulation a lot?”

“Are we reoperating our patients with a Fontan circulation ….

……. enough ???”

Fontan REGISTRY