Ann & Robert H. Lurie Children's Hospital of Chicago®

Ross-Konno operation in neonates and infants

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CHOP Meeting 2025 Orlando FL.
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Ann & Robert H. Lurie
Children's Hospital of Chicago®

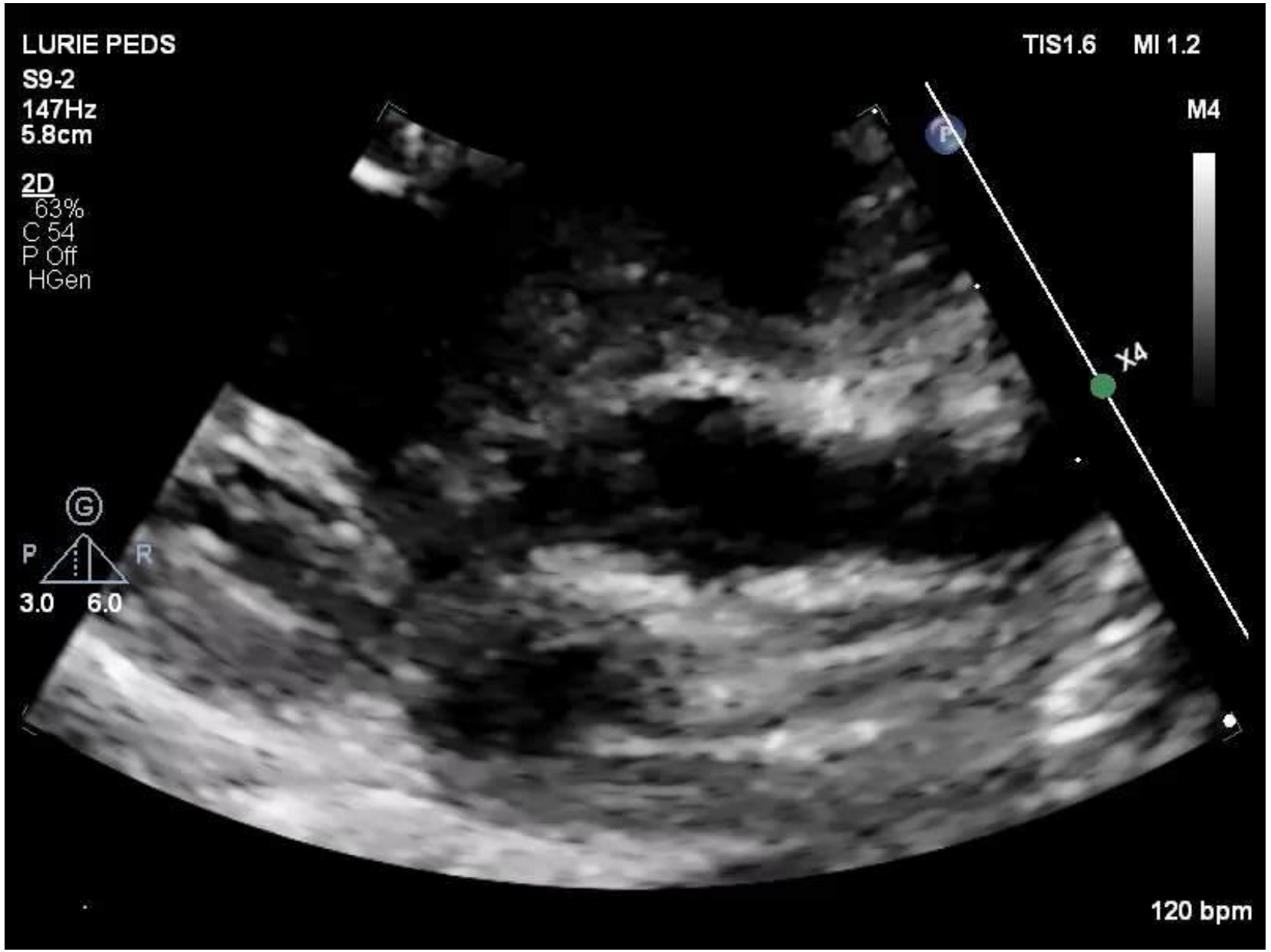
So nice to be in Orlando with you…

Who are we talking about?

- Isolated aortic valve disease
- Aortic valve disease with coarctation of the aorta / Shone associations
- Aortic interruption with VSD, narrowing of LVOT due to posterior deviation of septum

Newborn with interrupted aortic arch and posterior malalignment VSD causing left ventricular outflow tract obstruction.

Image:
Dr. Pei-Ni Jone



Newborn with interrupted aortic arch and posterior malalignment VSD causing left ventricular outflow tract obstruction.

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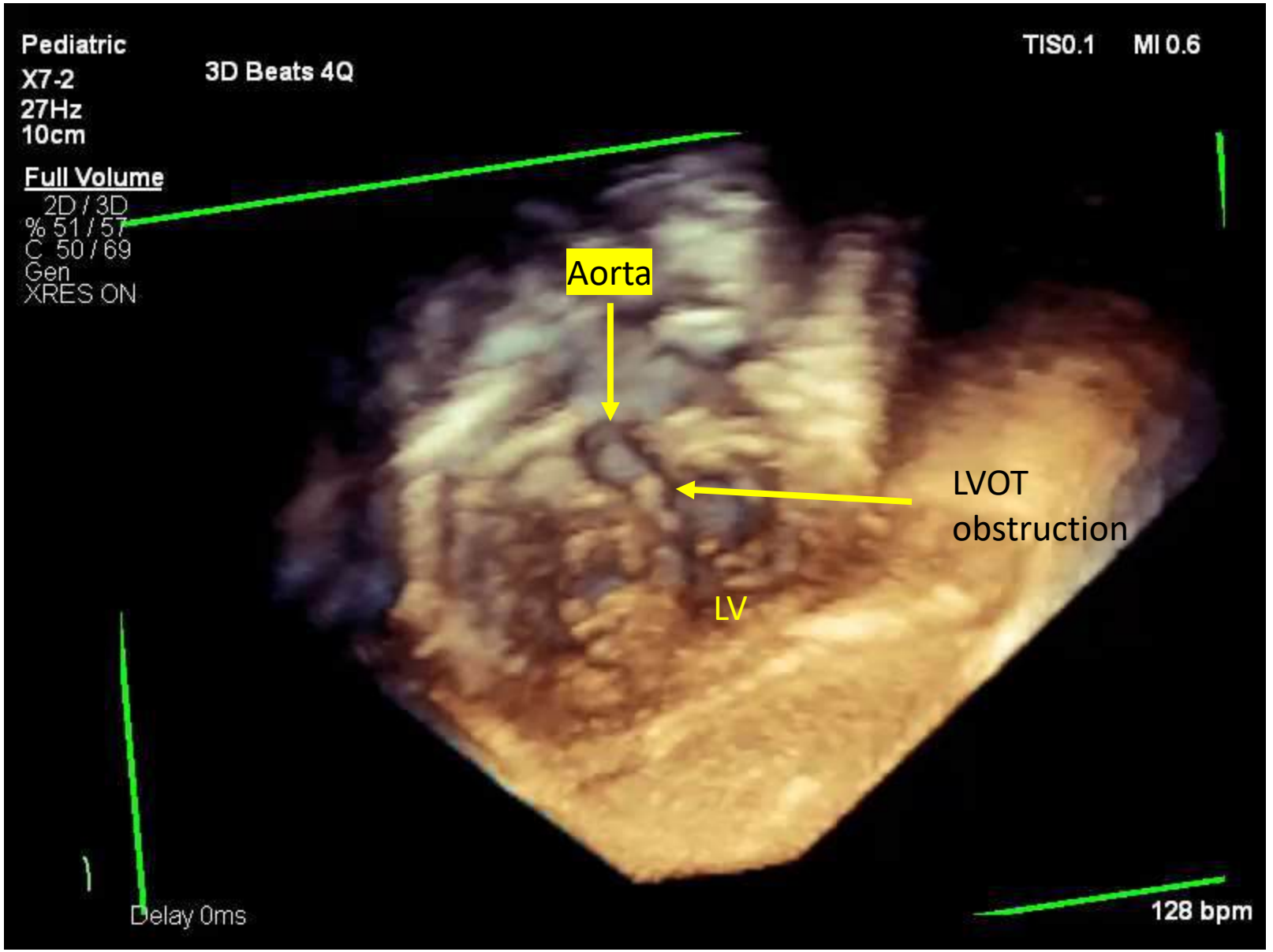
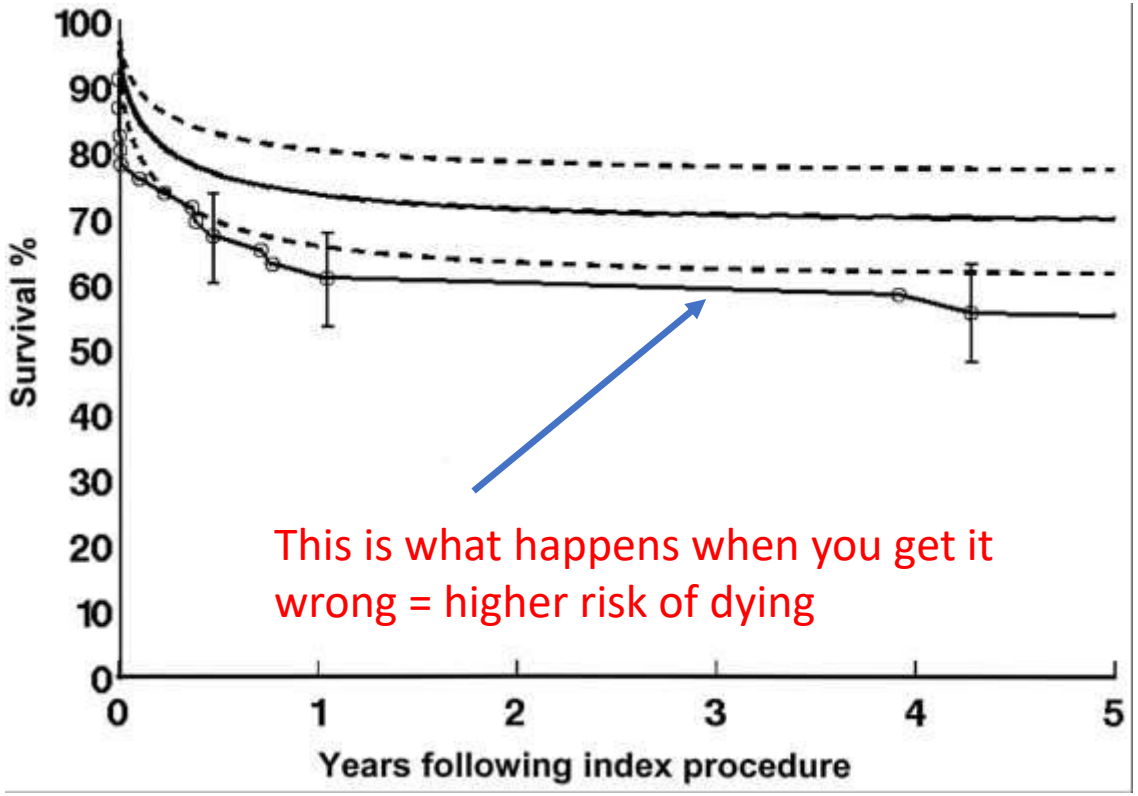


Is it big enough? Or do we have to do Single V?

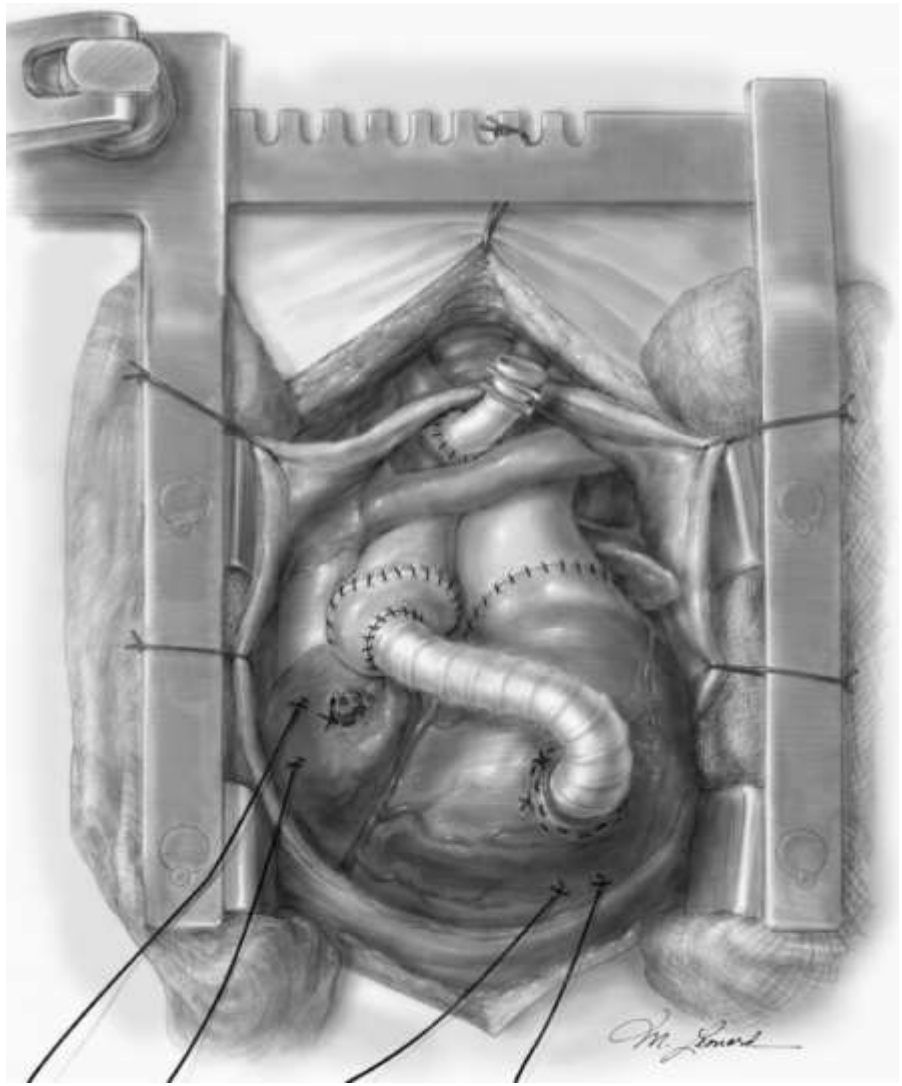
- Can the LVOTO handle the entire cardiac output?
  - Morphology of the valve
  - Presence of antegrade flow (…but usually on PGE)
  - Size of the LV
- Many algorithms
  - Single ventricle vs. biventricular circulation
  - Rhodes score, CHSS calculator etc.



Hickey…McCrindle, JTCVS, 2007

If the LVOT is too small 1:

- Norwood as a step to decision or to single ventricle

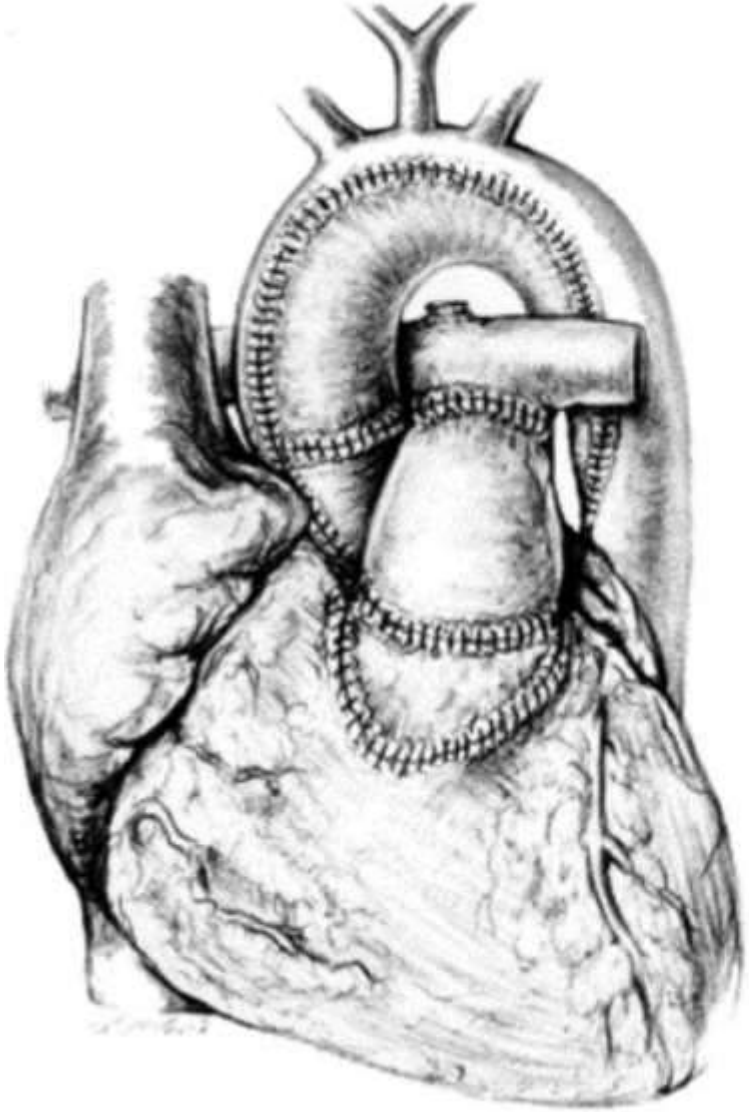


O’Donnell and Lehenbauer (Tweddell) Op Tech 2023

If the LVOT is too small and you have a big VSD 2:

- Norwood as first palliation

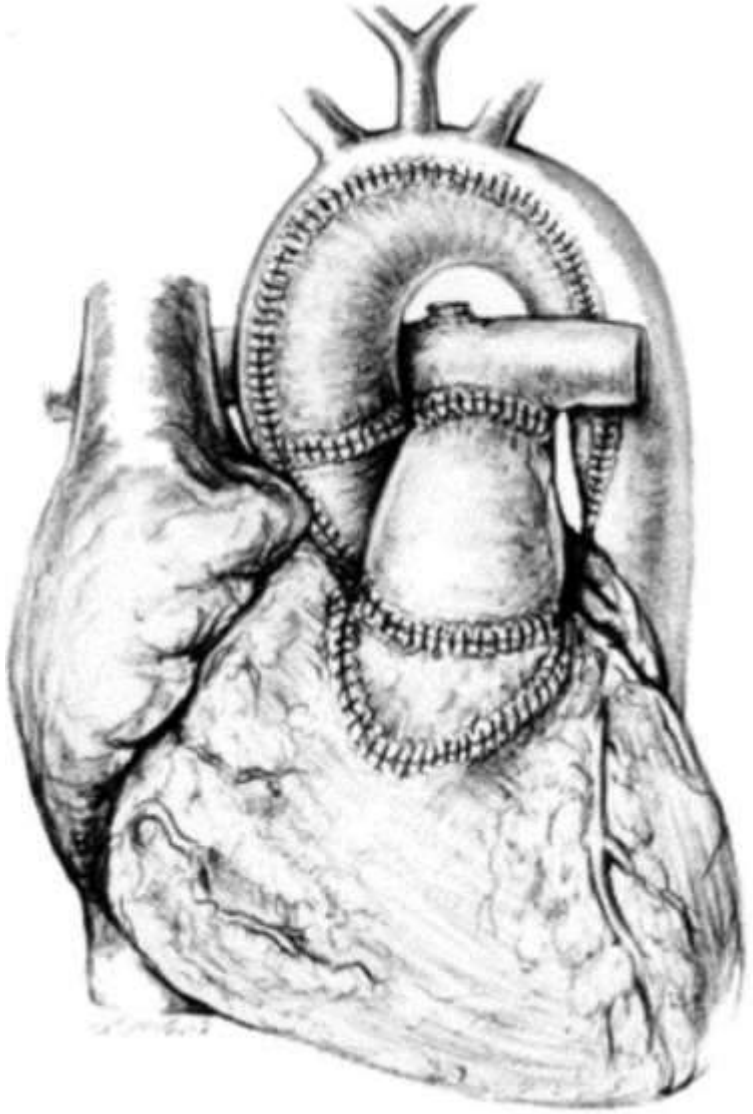
- Neonatal Yasui procedure
  (aka Norwood-Rastelli)
  - LV to PA
  - DKS
  - RV to PA connection



Nathan et al, Ann Thorac Surg 2006

If the LVOT is too small 2:

- Norwood as first palliation
- Neonatal Yasui procedure
  - LV to PA
  - DKS
  - RV to PA connection
- Yasui after initial Norwood



Nathan et al, Ann Thorac Surg 2006

Why I (now) don’t love the Yasui

Upsides – 2V

- Beautiful operation

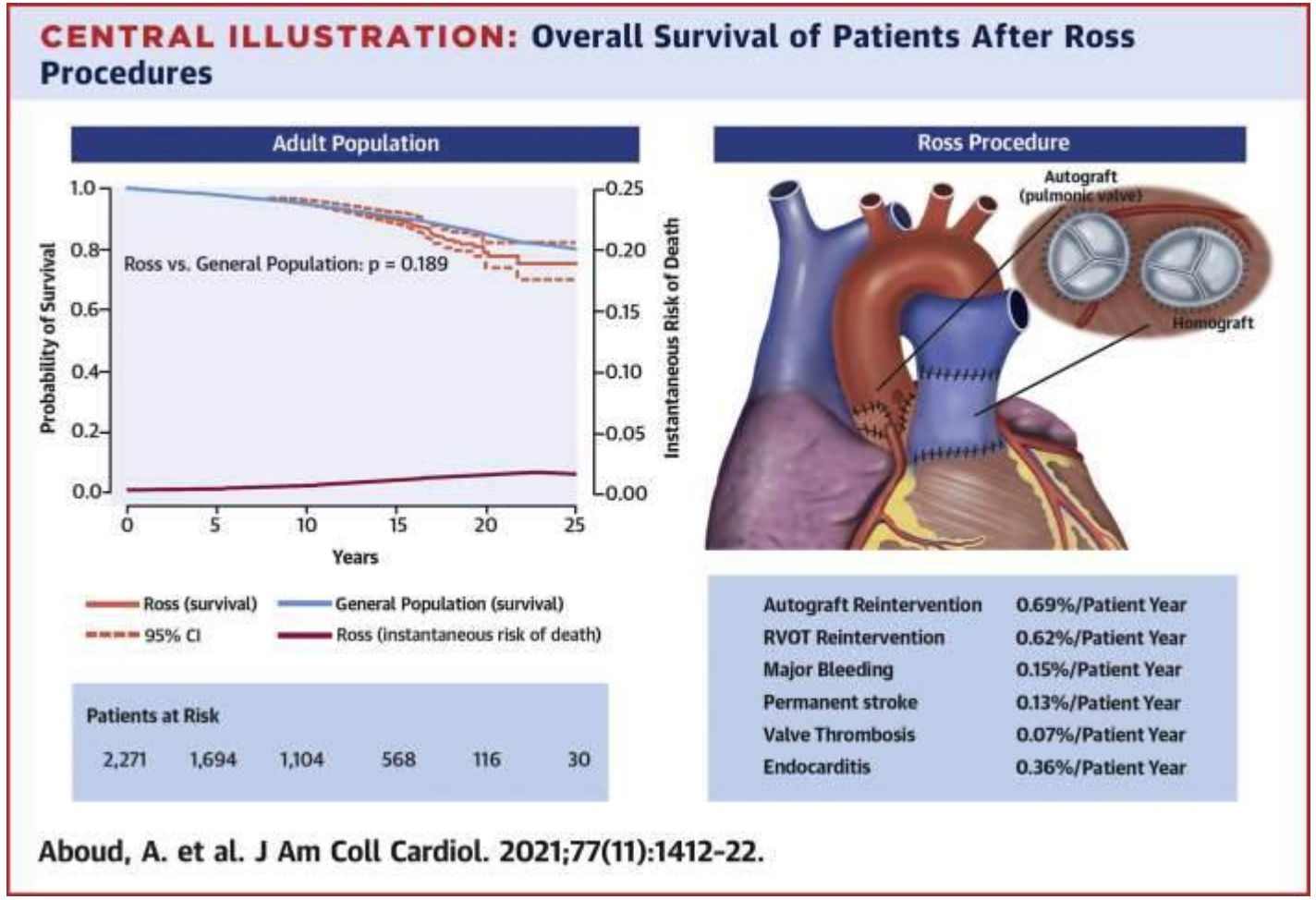
Downsides:

- big neonatal operation
- bulky Damus, impact on PAs and airways
- Indirect LV to pulmonary valve (systemic) pathway
- Indirect RV to pulmonary artery pathway for conduit
- Conduit lies beneath the sternum

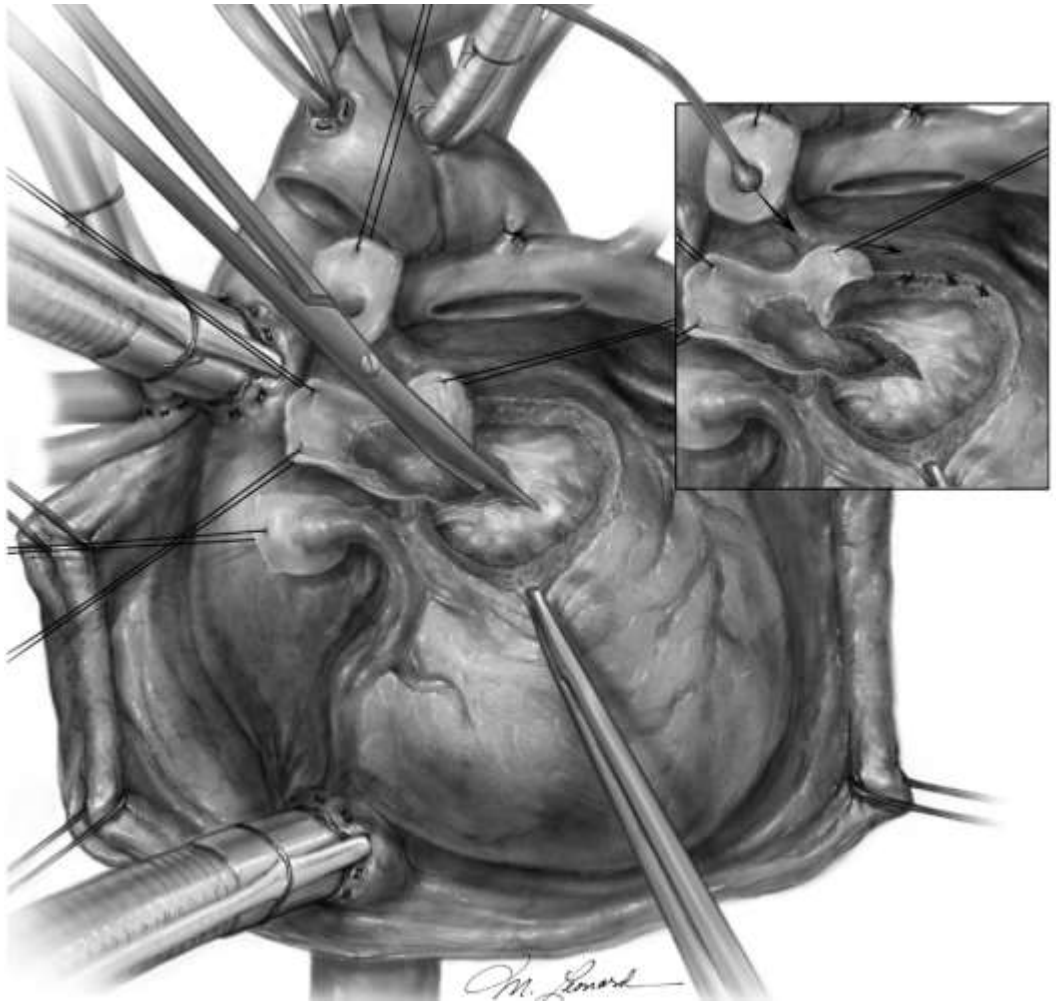
Scarlett Johansson and Adam Driver in *Marriage Story*.
Photo: Everett Collection

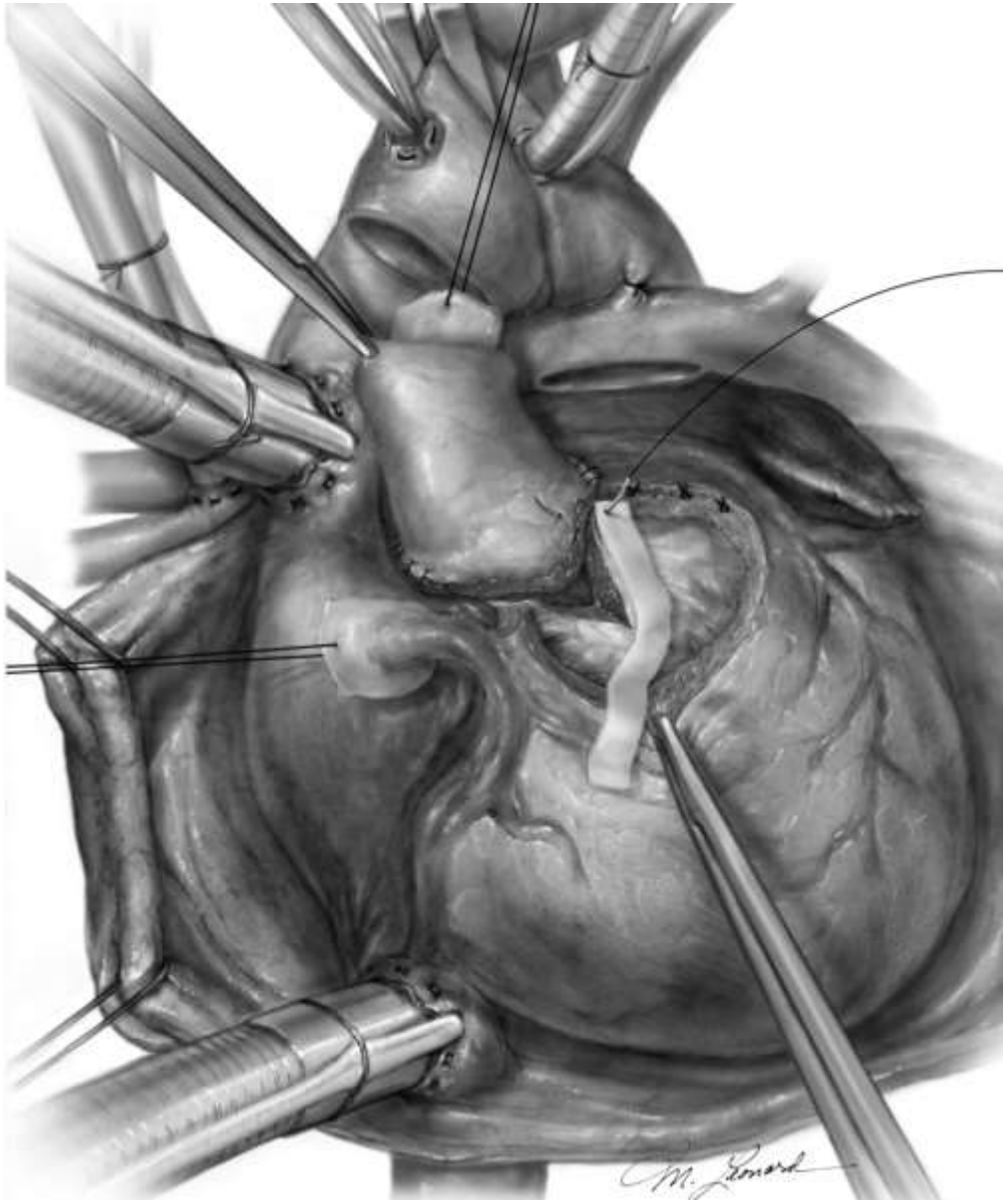
If the LVOT is too small 3: Ross Approach

- High quality aortic valve replacement
- RVOT homograft requires replacement over time
- Note survival data from the adult world!



Ross-Konno in infants:





Winlaw, Greenberg, O’Donnell, OpTechs 2022

Why I like the Ross-Konno

- Utilizes the native pulmonary valve, achieves a 2V pathway
- No anticoagulation required
- Excellent LV to aortic pathway
- RVOT reconstruction is orthotopic; conduits last longer
- Can be done after a Norwood (not something to look forward to)

- Downsides:
  - Big neonatal operation
  - Mortality historically has been high (on par with a Stage I Norwood)
  - Management of associated lesions (mitral valve stenosis and regurgitation)

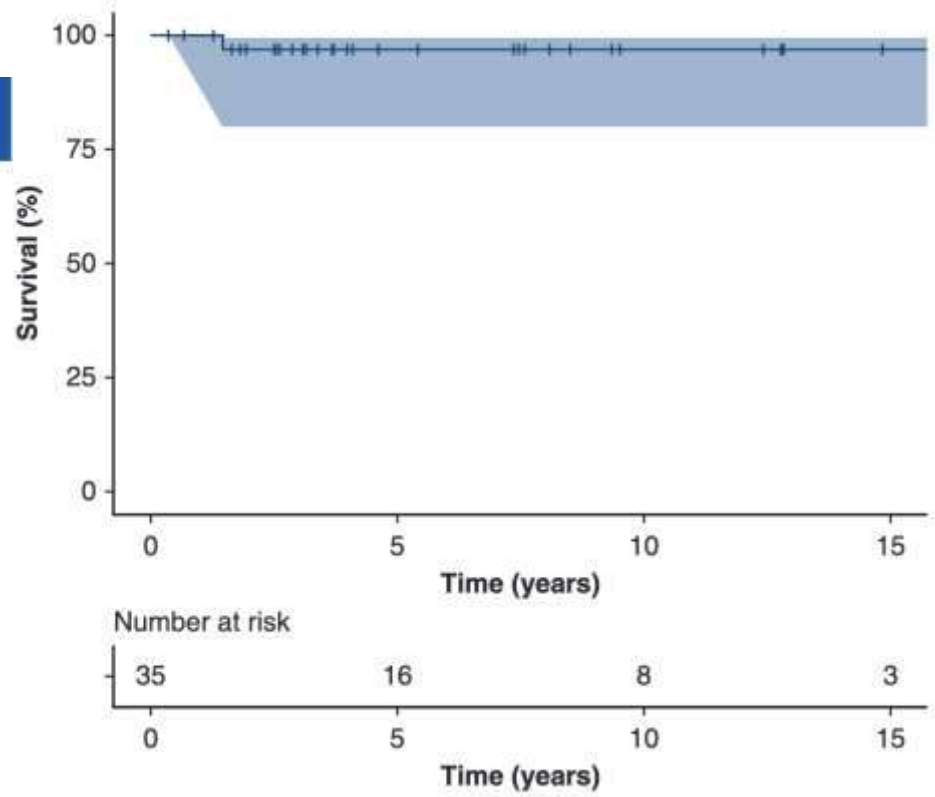
Neonatal R-K outcomes have been improving

CONGENITAL: AORTIC VALVE

The Ross/Ross-Konno procedure in infancy is a safe and durable solution for aortic stenosis

Jack C. Luxford, BA, MD,[a,b] Julian G. Ayer, MBBS, FRACP, PhD,[a,b] Kim Betts, PhD, MBiostat,[c] Gananjay G. Salve, MS, MCh,[b] Yishay Orr, MBBS, PhD, FRACS,[b] Richard B. Chard, MBBS, FRACS,[a,b] Philip Roberts, MB, ChB, FRACP,[a,b] Gary F. Sholler, MBBS, FRACP,[a,b] and David S. Winlaw, MBBS, MD, FRACS[a,b,d]

JTCVS 2022



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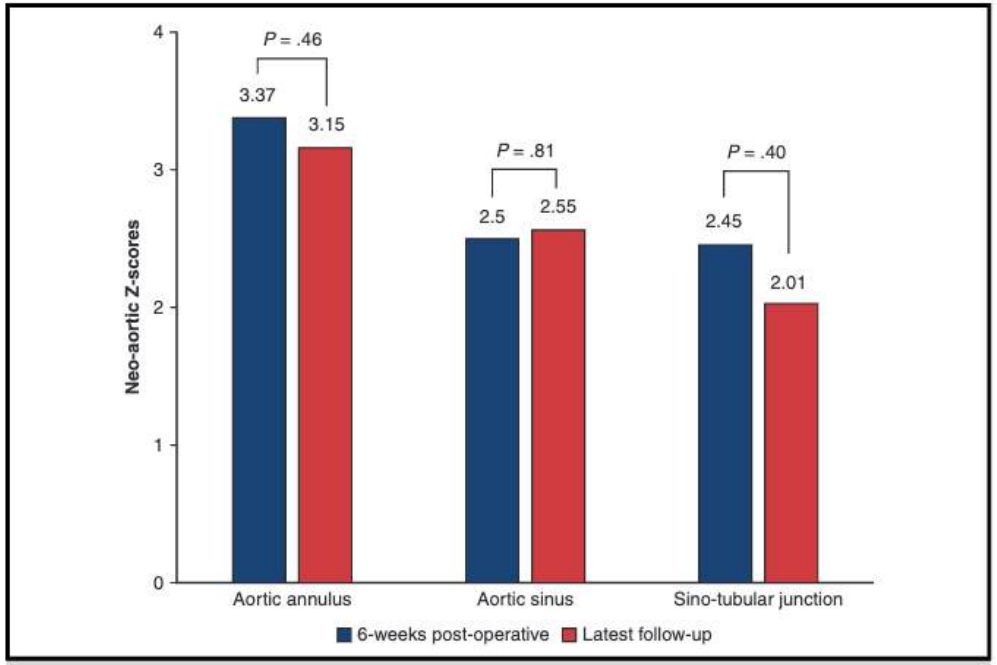


No significant change in mean Z-scores of the neo-aortic dimensions over 3 years.

What happens to the neo-aortic root?

Questions to Answer

- How durable is this ?
  - Neo-aortic valve function over time
- Will the same issues we see with the Ross in adults be an issue in kids?
  - Aortic root dilation and neo-aortic valve regurgitation

Multicenter study

JTCVS 2024

CONGENITAL: AORTIC VALVE

Long-term outcomes following the Ross procedure in neonates and infants: A multi-institutional analysis

Check for updates

Jason W. Greenberg, MD, MS,[a] Madison Argo, MD, MS,[b] Awais Ashfaq, MD,[a] Jack C. Luxford, MBBS,[c] Andres A. Fuentes-Baldemar, MD,[d] Alyssa B. Kalustian, MD,[d] S. Valeria Pena-Munoz, MD,[d] David J. Barron, MD,[b] Luc L. Mertens, MD, PhD,[b] S. Adil Husain, MD,[e] Jeffrey S. Heinle, MD,[d] Lauren C. Goldie, PhD, CCRP,[d] Yishay Orr, MBBS, PhD,[c] Julian Ayer, MBBS, PhD,[c] Constantine D. Mavroudis, MD,[f] Stephanie M. Fuller, MD,[f] David L. S. Morales, MD,[a] Garick D. Hill, MD,[a] David S. Winlaw, MBBS, MD, FRACS,[a] and the Ross Collaborative*

Cincinnati Children's changing the outcome together

SickKids

the children's hospital at Westmead

Texas Children's Hospital

Intermountain Primary Children's Hospital

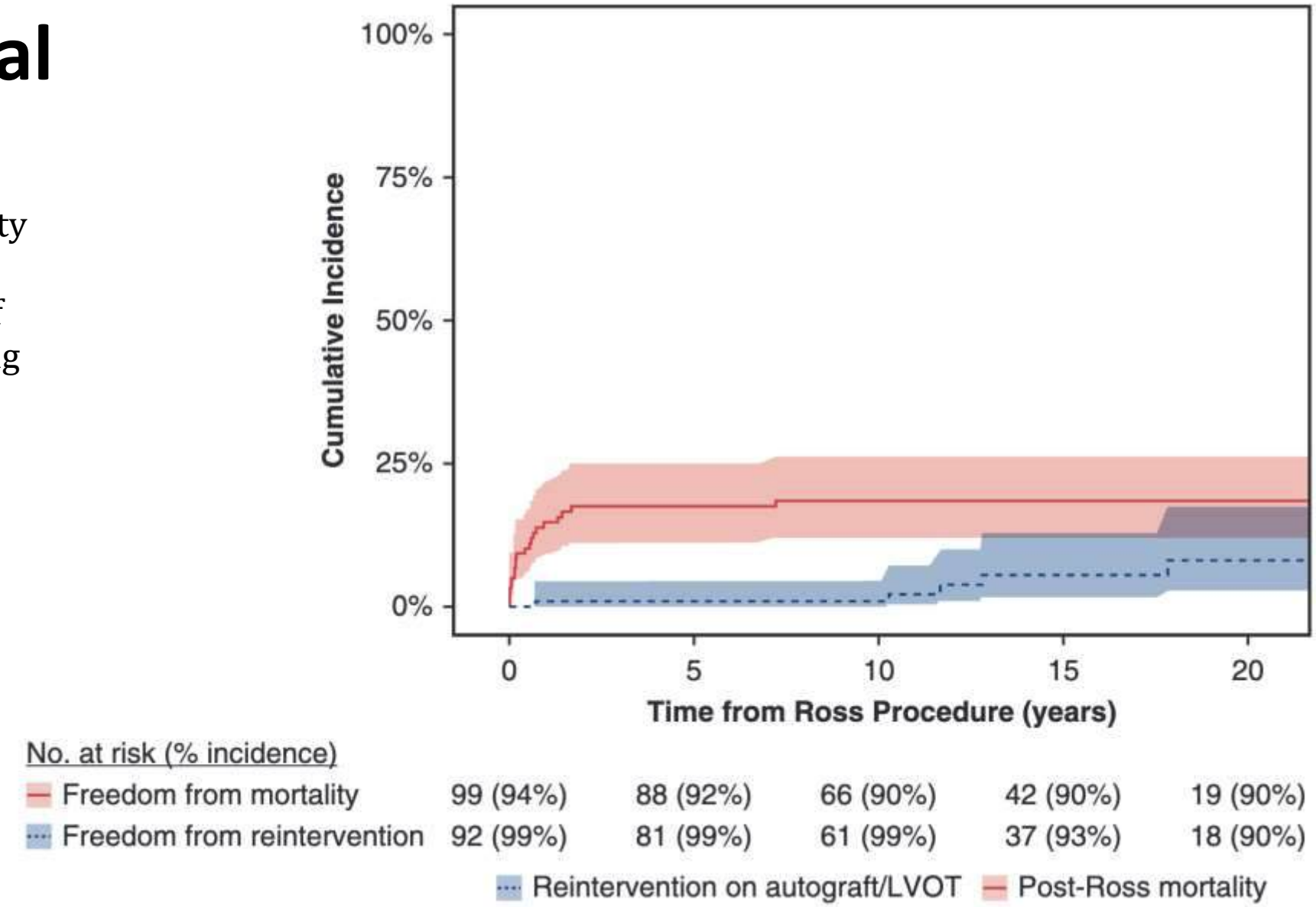
The Children's Hospital of Philadelphia

Why we constructed the study as we did

- > 5 years follow-up (1996 – 2016)
- Primary study outcomes included freedom from autograft/LVOT reintervention, autograft dilatation over time, and autograft function (for this study, defined as freedom from moderate/severe neoaortic insufficiency)
- 133 infants (n=30 neonates [23% of cohort]) were included in analysis

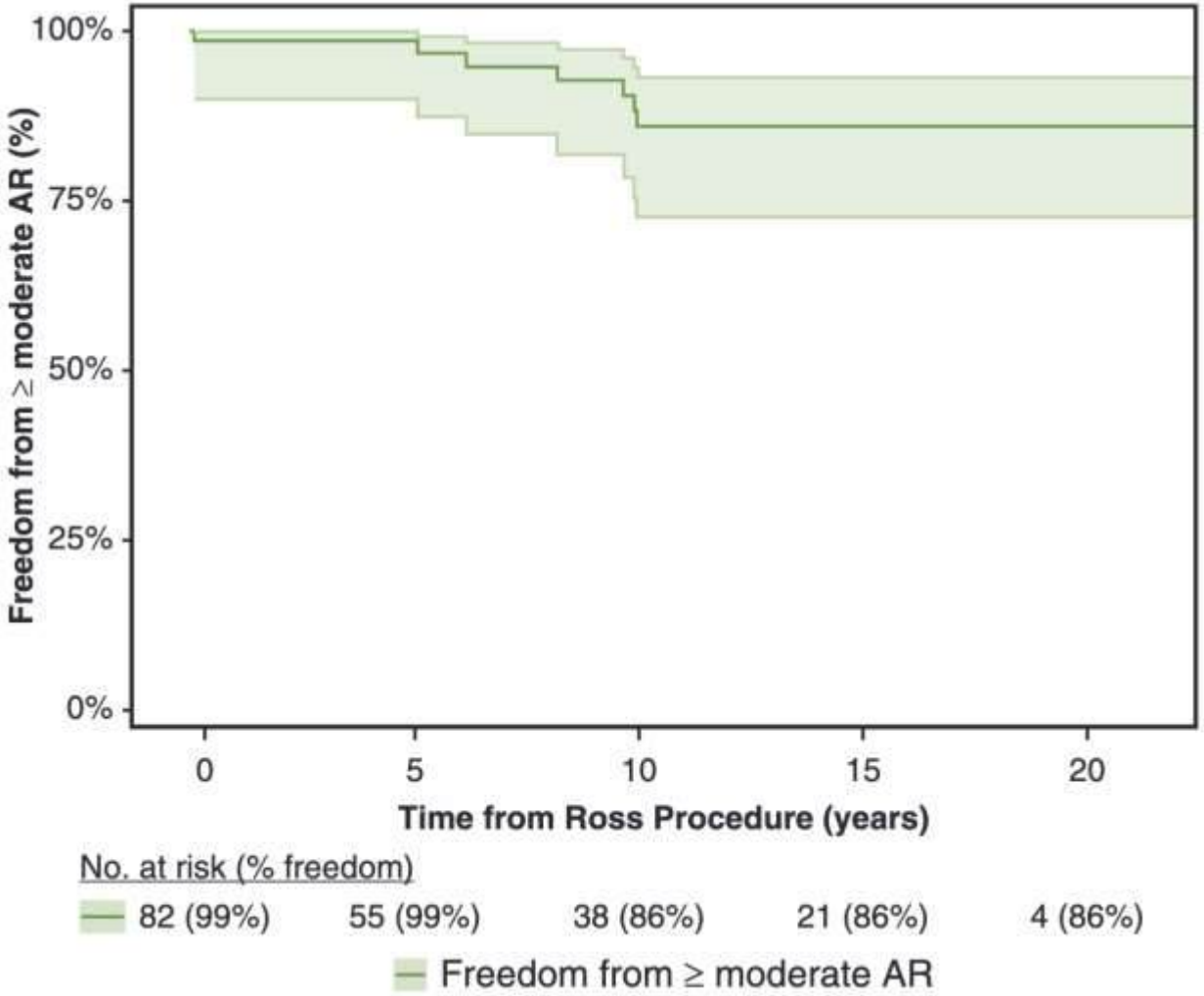
Survival

Hospital mortality occurred in 13 patients (10% of cohort), including in 4/30 (13%) neonates



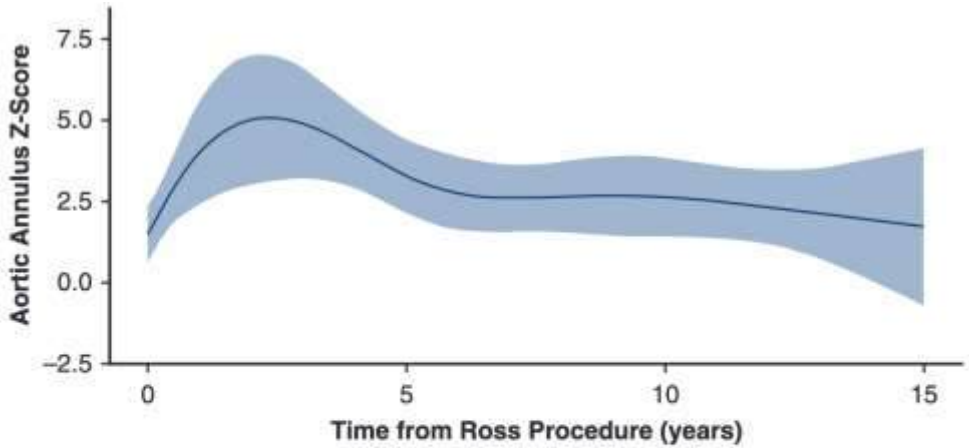
Autograft (neo-aortic valve) function

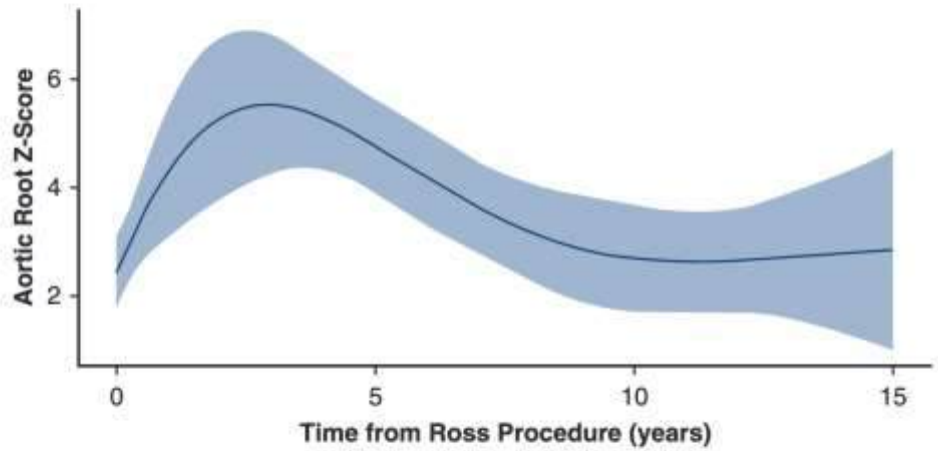
Autograft or LVOT reintervention was required in just five patients (4% [5/120] of hospital survivors) at a median 10.3 (2.4-15.3) years after Ross

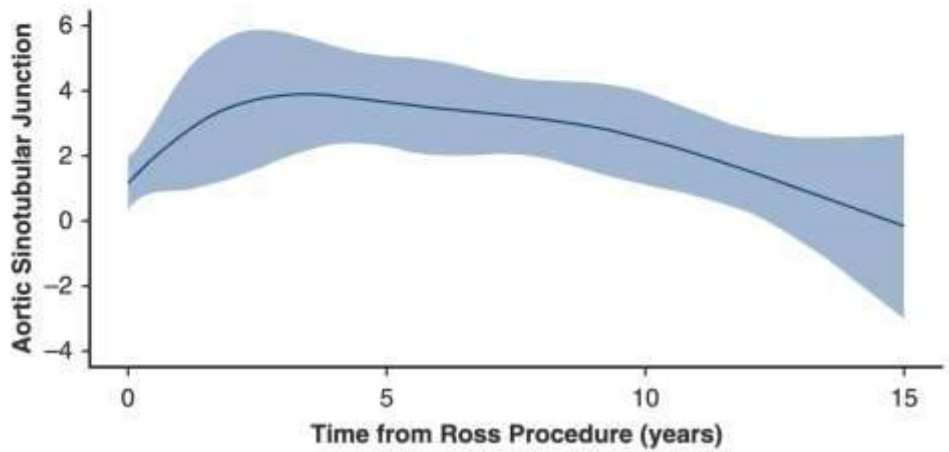


Autograft size over time

Babies grow into their new aortic root, unlike adults where the root keeps growing…







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Take homes:

- The Ross-Konno is a good option in centers with high neonatal volumes.
- Re-thinking our approach to IAA VSD with LVOTO
- There is a window of opportunity for adaptation of the autograft to the systemic circulation.
- Issues specific to the Ross remain a lifelong concern.

Supplementary – baseline characteristics

Variable	Cohort (n=133)
Male sex	90 (68%)
White (non-Hispanic) race/ethnicity*	66 (78%)
Genetic syndrome	12 (9%)
Non-cardiac comorbidity	28 (21%)
Primary cardiac diagnosis	
Shone complex	19 (14%)
Isolated AS	19 (14%)
AS + other	95 (71%)
Arch obstruction (IAA, coarctation)	55 (58%)[a]
LV hypoplasia	9 (9%)[a]
Mitral disease	31 (33%)[a]
VSD	40 (42%)[a]
Prior arch surgery	53 (72%)[b]
Prior balloon angioplasty – yes	85 (64%)
Number of balloon angioplasties	1 (1-2)
Moderate/severe AR following balloon	32 (38%)

Table 1. Baseline characteristics of neonates and infants undergoing the Ross. Values expressed as median (IQR) or n (%) as appropriate.

Abbreviations: AR, aortic regurgitation; IAA, interrupted aortic arch; LV, left ventricle.

[a]Denominator includes only patients with "AS + other" diagnosis, excluding Shone complex (listed separately above).

[b]Denominator includes only patients with arch obstruction, including Shone complex (n=74).

*Data not reported in all patients.

Supplementary – operative details

Variable	Cohort (n=133)
Age at Ross (days)	96 (36-186)
Neonatal Ross	30 (23%)
Weight at Ross (kg)	4.4 (3.6-6.5)
Inotrope requirement at Ross	30 (24%)
Ventilator requirement at Ross	31 (25%)
ECMO requirement at Ross	2 (2%)
CPB time (min)	181 (111-270)
Aortic cross-clamp time (min)	127 (73-182)
Concomitant procedures	
Konno incision	111 (83%)
Arch surgery	25 (34%)[a]
Mitral surgery	26 (52%)[b]
EFE resection	22 (65%)[c]
VSD closure	18 (35%)[d]
Intraoperative ECMO requirement	7 (5%)

Table 2. Operative details of Ross procedure. Values expressed as median (IQR) or n (%) as appropriate.

Abbreviations: CPB, cardiopulmonary bypass; EFE, endocardial fibroelastosis; ECMO, extracorporeal membrane oxygenation; VSD, ventricular septal defect.

[a]Denominator includes only patients with arch obstruction, including Shone complex (total n=74).

[b]Denominator includes only patients with mitral valve abnormalities, including Shone complex (total n=50).

[c]Denominator includes only patients with EFE (n=34).

[d]Denominator includes only patients with VSD (n=51).

Supplementary – hospital survivors vs. non-survivors

Variable	Hospital Survival (n=120)	Hospital Mortality (n=13)	p-value
Baseline characteristics			
Male sex	84 (71%)	6 (46%)	0.113
White (non-Hispanic) race/ethnicity*	57 (75%)	9 (100%)	0.198
Genetic syndrome	11 (9%)	1 (8%)	0.999
Extra-cardiac comorbidity	25 (21%)	3 (23%)	0.999
Primary cardiac diagnosis			
Shone complex	15 (12%)	4 (31%)	0.076
Isolated AS	21 (18%)	0 (0%)	
AS + other	84 (70%)	9 (69%)	
Arch obstruction (IAA, coarctation)	49 (57%)[a]	6 (67%)[a]	0.729
LV hypoplasia	7 (8%)[a]	2 (22%)[a]	0.209
Mitral disease	28 (33%)[a]	3 (33%)[a]	0.999
VSD	37 (44%)[a]	3 (33%)[a]	0.727
Prior arch surgery	46 (72%)[b]	7 (70%)[b]	0.999
Prior balloon angioplasty – yes	75 (63%)	10 (77%)	0.378
Number of balloon angioplasties	1 (1-2)	1 (1-1)	0.653
Moderate/severe AR following balloon	29 (39%)	3 (30%)	0.736
Operative details			
Age at Ross (days)	104 (37-181)	73 (20-231)	0.399
Neonatal Ross	26 (22%)	4 (31%)	0.489
Weight at Ross (kg)	4.5 (3.5-6.5)	4.0 (3.6-6.1)	0.261
Inotrope requirement at Ross	27 (24%)	3 (23%)	0.999
Ventilator requirement at Ross	26 (23%)	5 (38%)	0.306
ECMO requirement at Ross	1 (1%)	1 (8%)	0.191
CPB time (min)	176 (108-261)	267 (195-288)	0.070
Aortic XC time (min)	123 (73-178)	169 (110-199)	0.227
Concomitant procedures			
Konno incision	98 (82%)	13 (100%)	0.125
Arch surgery	21 (33%)[b]	4 (40%)[b]	0.727
Mitral surgery	20 (57%)[c]	6 (86%)[c]	0.222
EFE resection	21 (66%)[d]	1 (50%)[d]	0.999
VSD closure	15 (33%)[e]	3 (60%)[e]	0.331
Intraoperative ECMO requirement	6 (5%)	1 (8%)	0.531
Postoperative course–index hospitalization			
Ventilator days	4 (1-10)	11 (4-37)	0.061
Reintubation	13 (12%)	5 (38%)	**0.022**
ICU length of stay (days)	10 (5-22)	15 (5-46)	0.743
Hospital length of stay (days)	18 (9-34)	15 (6-54)	0.978
Cardiac reintervention[f]	16 (13%)	8 (62%)	**<0.001**

Table 3. Characteristics and outcomes of hospital survivors versus non-survivors following Ross.

Supplementary – neonatal subgroup

Variable	Neonatal Subgroup (n=30)
Baseline characteristics	
Male sex	16 (53%)
White (non-Hispanic) race/ethnicity*	18 (75%)
Genetic syndrome	2 (7%)
Non-cardiac comorbidity	7 (23%)
Primary cardiac diagnosis	
Shone complex	2 (7%)
Isolated AS	2 (7%)
AS + other	26 (87%)
Arch obstruction (IAA, coarctation)	15 (58%)[a]
LV hypoplasia	3 (12%)[a]
Mitral disease	9 (32%)[a]
VSD	11 (42%)[a]
Prior arch surgery	2 (12%)[b]
Prior balloon angioplasty – yes	16 (53%)
Number of balloon angioplasties	1 (1-1)
Moderate/severe AI following balloon	6 (38%)
Operative details	
Age at Ross (days)	9 (6-13)
Weight at Ross (kg)	3.3 (3.0-3.6)
Inotrope requirement at Ross	12 (44%)
Ventilator requirement at Ross	12 (44%)
ECMO requirement at Ross	1 (3%)
CPB time (min)	139 (108-241)
Aortic XC time (min)	103 (73-142)
Concomitant procedures	
Konno incision	29 (97%)
Arch surgery	13 (81%)[b]
Mitral surgery	7 (64%)[c]
EFE resection	6 (67%)[d]
VSD closure	10 (91%)[e]
Intraoperative ECMO requirement	5 (17%)
Postoperative course – index hospitalization	
Ventilator days	6 (2-15)
Reintubation	3 (11%)
ICU length of stay (days)	21 (14-35)
Hospital length of stay (days)	24 (17-49)
Cardiac reintervention[f]	8 (27%)
Hospital mortality	4 (13%)

Table S1. Baseline characteristics, operative details, and outcomes of the neonatal Ross sub-group.

Supplementary – autograft reinterventions

Variable	Reintervention on Autograft/LVOT		p-value
	No (n=108)	Yes (n=5)	
Baseline characteristics			
Male sex	79 (73%)	2 (40%)	0.137
White (non-Hispanic) race/ethnicity*	53 (77%)	2 (67%)	0.560
Genetic syndrome	10 (9%)	0 (0%)	0.999
Extra-cardiac comorbidity	21 (19%)	2 (40%)	0.268
Primary cardiac diagnosis			
Shone complex	12 (11%)	0 (0%)	0.764
Isolated AS	18 (17%)	0 (0%)	
AS + other	78 (72%)	5 (100%)	
Arch obstruction (IAA, coarctation)	49 (56%)[a]	4 (80%)[a]	0.391
LV hypoplasia	8 (9%)[a]	1 (20%)[a]	0.410
Mitral disease	29 (33%)[a]	1 (20%)[a]	0.999
VSD	36 (41%)[a]	3 (60%)[a]	0.647
Prior arch surgery	37 (69%)[b]	4 (100%)[b]	0.310
Prior balloon angioplasty – yes	68 (64%)	1 (20%)	0.071
Number of balloon angioplasties	1 (1-2)	1 (1-1)	0.538
Moderate/severe AR following balloon	27 (40%)	0 (0%)	0.999
Operative details			
Age at Ross (days)	104 (36-187)	142 (124-178)	0.166
Neonatal Ross	24 (22%)	1 (20%)	0.999
Weight at Ross (kg)	4.4 (3.4-6.5)	5.4 (4.8-6.0)	0.159
Inotrope requirement at Ross	22 (22%)	1 (20%)	0.999
Ventilator requirement at Ross	22 (22%)	0 (0%)	0.577
ECMO requirement at Ross	1 (1%)	0 (0%)	0.999
CPB time (min)	170 (109-260)	97 (96-185)	0.649
Aortic XC time (min)	114 (73-178)	74 (64-103)	0.172
Concomitant procedures			
Konno incision	87 (81%)	5 (100%)	0.582
Arch surgery	20 (37%)[b]	0 (0%)[b]	0.288
Mitral surgery	18 (55%)	1 (100%)[c]	0.999
EFE resection	21 (68%)	n/a*	n/a*
VSD closure	15 (38%)	0 (0%)[c]	0.541
Intraoperative ECMO requirement	6 (6%)	0 (0%)	0.999
Postoperative course – index hospitalization			
Ventilator days	4 (1-10)	1 (1-4)	0.266
Reintubation	13 (13%)	0 (0)	0.999
ICU length of stay (days)	11 (5-22)	4 (3-19)	0.320
Hospital length of stay (days)	18 (8-35)	9 (9-14)	0.213
Cardiac reintervention[f]	16 (15%)	1 (20%)	0.565

Table S2. Characteristics and outcomes of patients with and without autograft/LVOT reintervention requirements following Ross. Denominator includes hospital survivors only. Values expressed as median (IQR) or n (%) as appropriate.