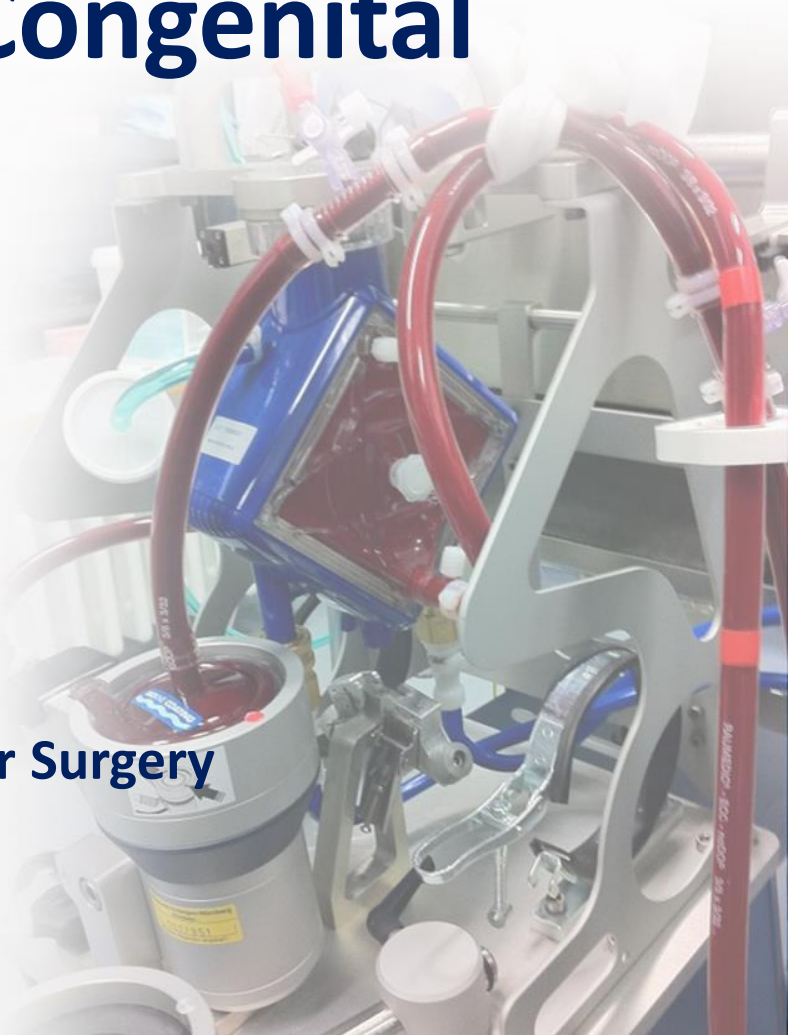
Low Cardiac Output Syndrome of Postoperative Adult Congenital Heart Disease in ICU



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COI Disclosure

Name of First Author: Hong-Gook Lim

The authors have no financial conflicts of interest to disclose concerning the presentation

Disclosure

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- **Grants/Research Support: N/A**
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- **Other: N/A**

Introduction

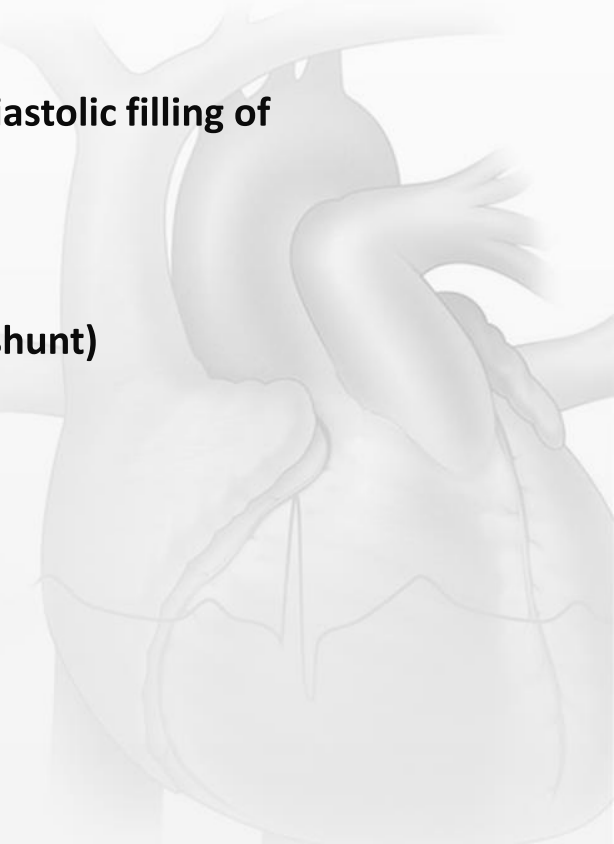
- Advances in surgical techniques and in medical Mx of children with CHD increased survival to adult, resulting in ACHD surpassing pediatric CHD.
- Many patients are not truly “repaired” but are “palliated,” so long-term effects of residual defects, deterioration of prosthetic valves or surgical conduits, or other complications are responsible for LCOS.
- Patient-specific risk factors include complex anatomy, repeated open chest surgeries and reduced baseline cardiac function.
- LCOS among ACHD grew exponentially in the last 2 decades and is among the most common reasons for morbidity and mortality in ACHD, similar to NCHD.
- ACHDs are a heterogeneous group with unique anatomy and long-term sequelae related to chronic cardiac disease.
- The mechanisms underlying LCOS in ACHD are lesion dependent and differ substantially from NCHD.

Introduction

- As a result, Tx developed to treat LCOS in NCHD have frequently failed to produce a benefit in ACHD.
- MCS in the form of VADs decrease mortality in LCOS among non-transplant candidates and appear to offer a similar benefit to transplant candidates doing poorly while awaiting transplant.
  - ACHD differed from NCHD, including younger age, greater allosensitization, more RV dysfunction, and unfavorable mediastinal anatomy.
- Although heart transplants in ACHD have increased over the past decade, these patients remain more likely to have a prolonged waiting time and to die while on the transplant waitlist.
  - ACHDR pose unique challenges due to complex anatomy, multiple previous palliative and corrective surgeries, preformed HLA antibodies, effects on other organs (kidney, liver, lungs) of long-standing cardiac dysfunction or cyanosis, pulmonary hypertension, physiology of Fontan-type circulation and malnutrition.

Classification of LCOS

- 1. Systemic morphological LV dysfunction
  - Pressure overload (sub-, supravalvular or valvular AS, CoA)
  - Volume overload (AR, VSD, PDA, or MR)
  - Myocardial injury (limited myocardial protection during bypass, ventriculotomy)
  - Altered myocardial architecture (non-compaction)
  - Altered geometry of sub-pulmonary ventricle interfering with diastolic filling of systemic ventricle (severe PR in ToF)
- 2. Sub-pulmonary morphological RV dysfunction
  - Volume overload (severe PR in ToF, ASD with large left-to-right shunt)
  - Pressure overload (severe RVOT obstruction)
- 3. Systemic morphological RV dysfunction
  - Pressure overload [corrected TGA, d-TGA after atrial switch]
  - Myocardial injury by functional ischaemia (single RCA)
  - Volume overload [corrected TGA, d-TGA after atrial switch]

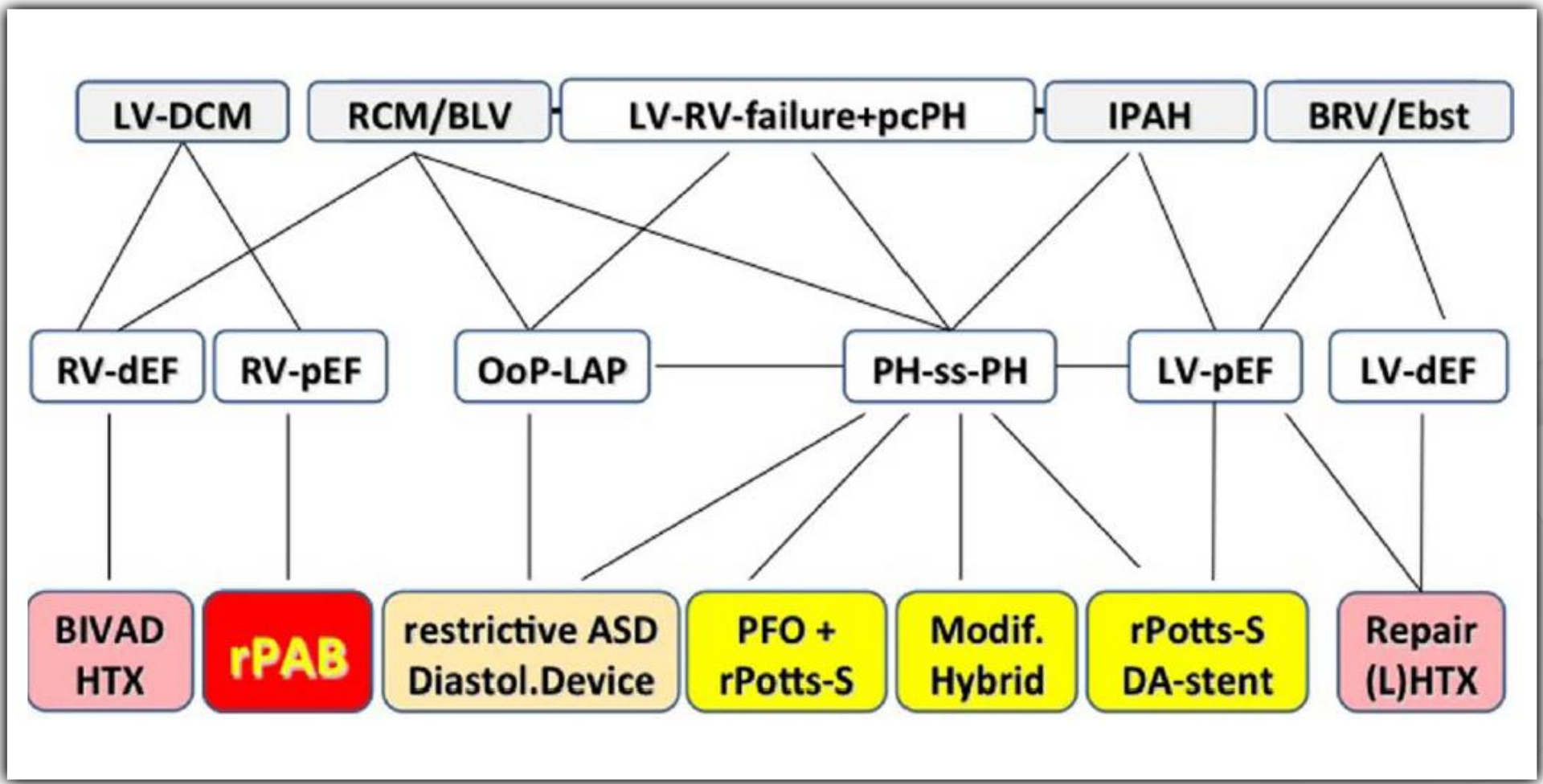


Classification of LCOS

- 4. Systemic single ventricle dysfunction
  - Volume under-load after initial volume overload (Fontan repair)
  - Myocardial injury (limited myocardial protection during bypass, ventriculotomy)
  - Myocardial architecture
- 5. Dysfunction of cyanotic systemic and/or sub-pulmonary ventricle with or without PHT
  - Myocardial injury by chronic hypoxia (VSD with PS)
  - Pressure overload (Eisenmenger syndrome)
- 6. Acquired ischaemic heart disease and ventricular dysfunction
  - Cardiovascular risk factors (HT, hyperlipidaemia, DM, smoking)
  - Congenital CA abnormalities (anomalous origin and/or course, extrinsic compression by dilated PA, kinking after re-implantation of CAs)
- 7. Systemic ventricle dysfunction due to tachyarrhythmias

Tx for LCOS by Lt-Rt interactions in ACHD (Heart 2017)

- **'End-stage' heart failure: potential surgical interventional therapies based on left-right heart interactions**



Potts Shunt (KJCTS 2015)

Korean J Thorac Cardiovasc Surg 2015;48:52-54 ☐ Case Report ☐
ISSN: 2233-601X (Print) ISSN: 2093-6516 (Online) http://dx.doi.org/10.5090/kjtcs.2015.48.1.52

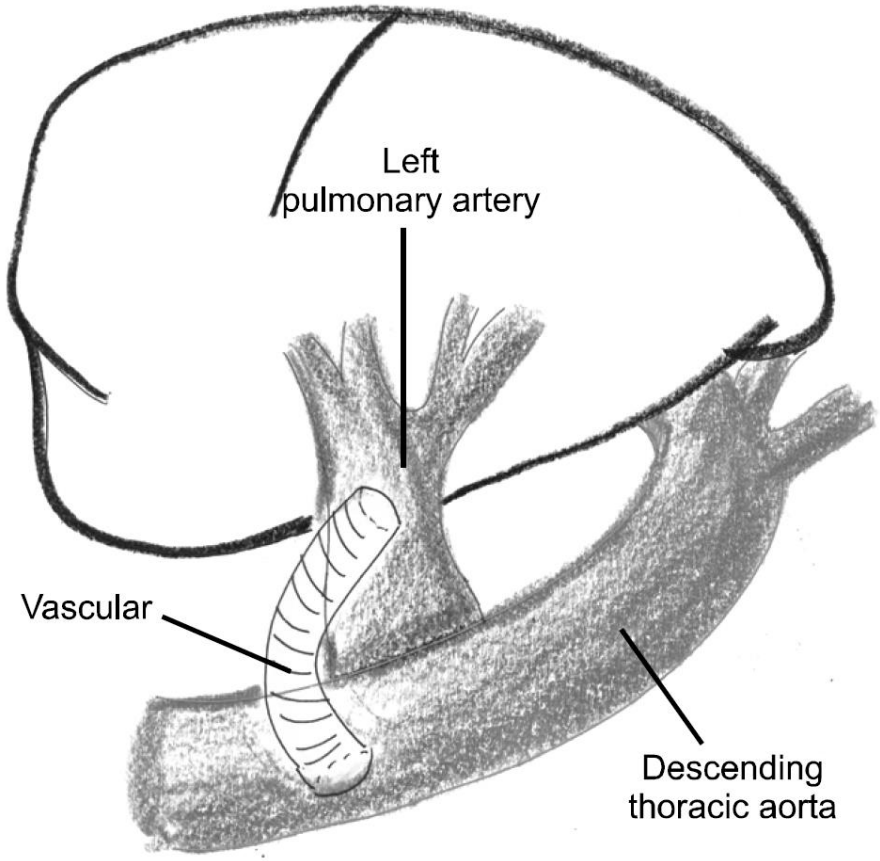
Potts Shunt in Patients with Primary Pulmonary Hypertension

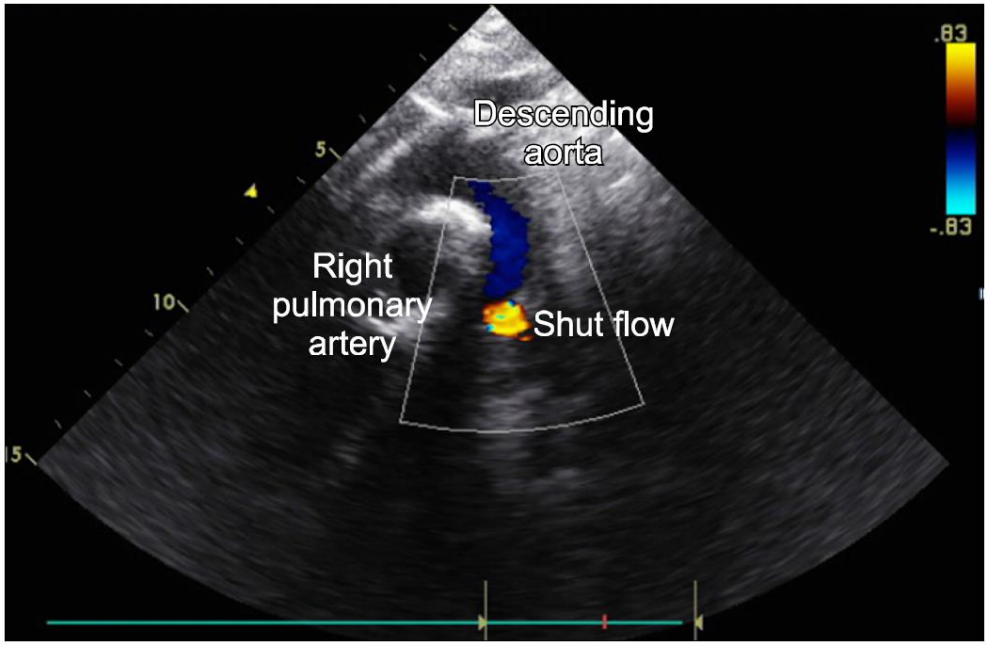
Sue Hyun Kim, M.D.[1], Woo-Sung Jang, M.D.[1], Hong-Gook Lim, M.D., Ph.D.[1], Yong-Jin Kim, M.D., Ph.D.[2]

Idiopathic pulmonary arterial hypertension eventually leads to right-sided heart failure and sudden death. Its mortality rate in children is still high, despite improvements in pharmacological therapy, and therefore novel treatments are necessary. The Potts shunt, which creates an anastomosis between the left pulmonary artery and the descending aorta, has been proposed as a theoretically promising palliative surgical technique to decompress the right ventricle. We report the case of a 12-year-old girl with suprasystemic idiopathic pulmonary hypertension and right ventricular failure who underwent a Potts shunt for palliation with good short-term results.

Key words: 1. Shunts
2. Pulmonary hypertension

Potts Shunt (KJCTS 2015)





Long-Term Result of TOF Repair (EJCTS 2010, SNUH)

ELSEVIER

European Journal of Cardio-thoracic Surgery 38 (2010) 311–317

EUROPEAN JOURNAL OF CARDIO-THORACIC SURGERY

www.elsevier.com/locate/ejcts

The long-term result of total repair for tetralogy of Fallot☆

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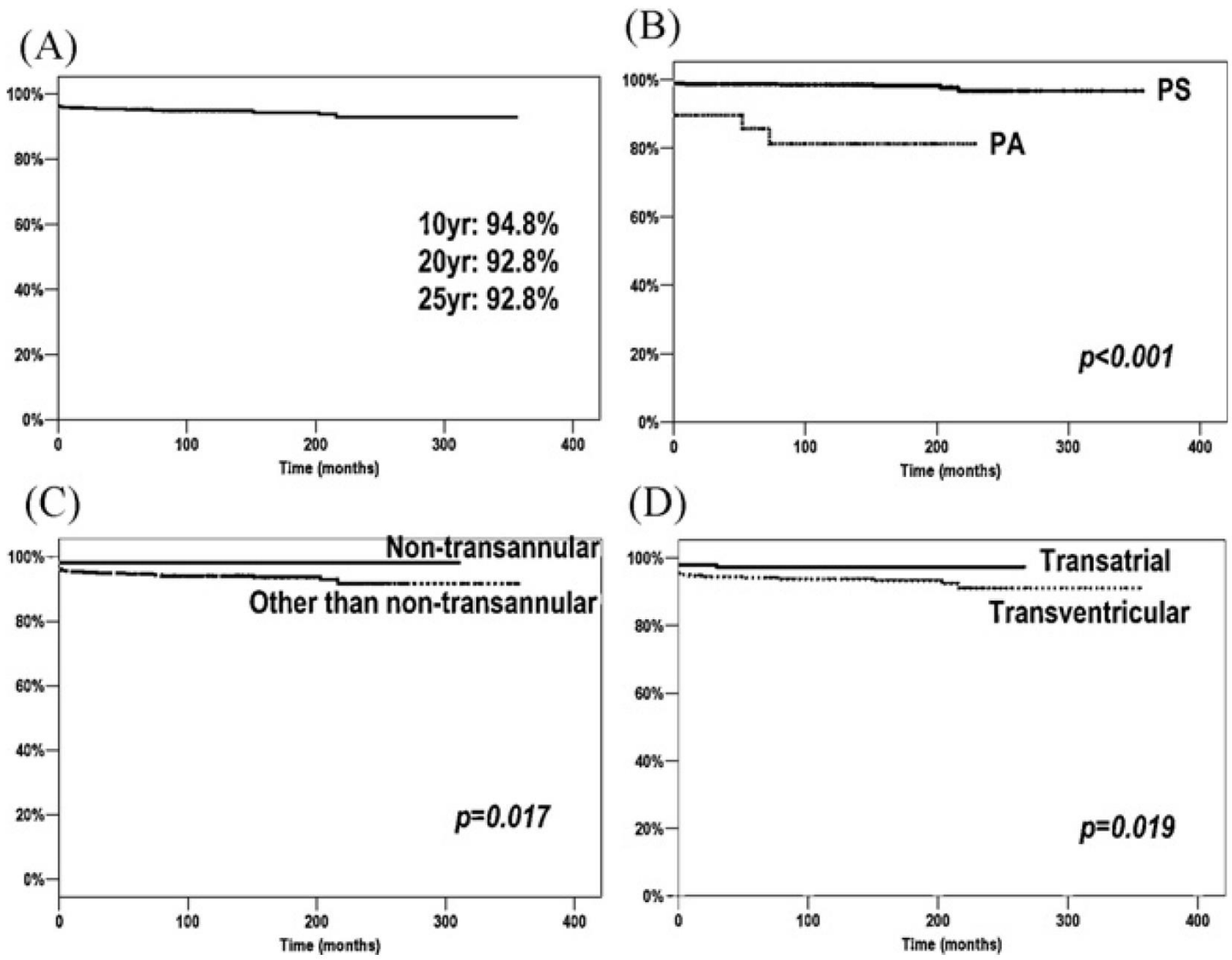
Abstract

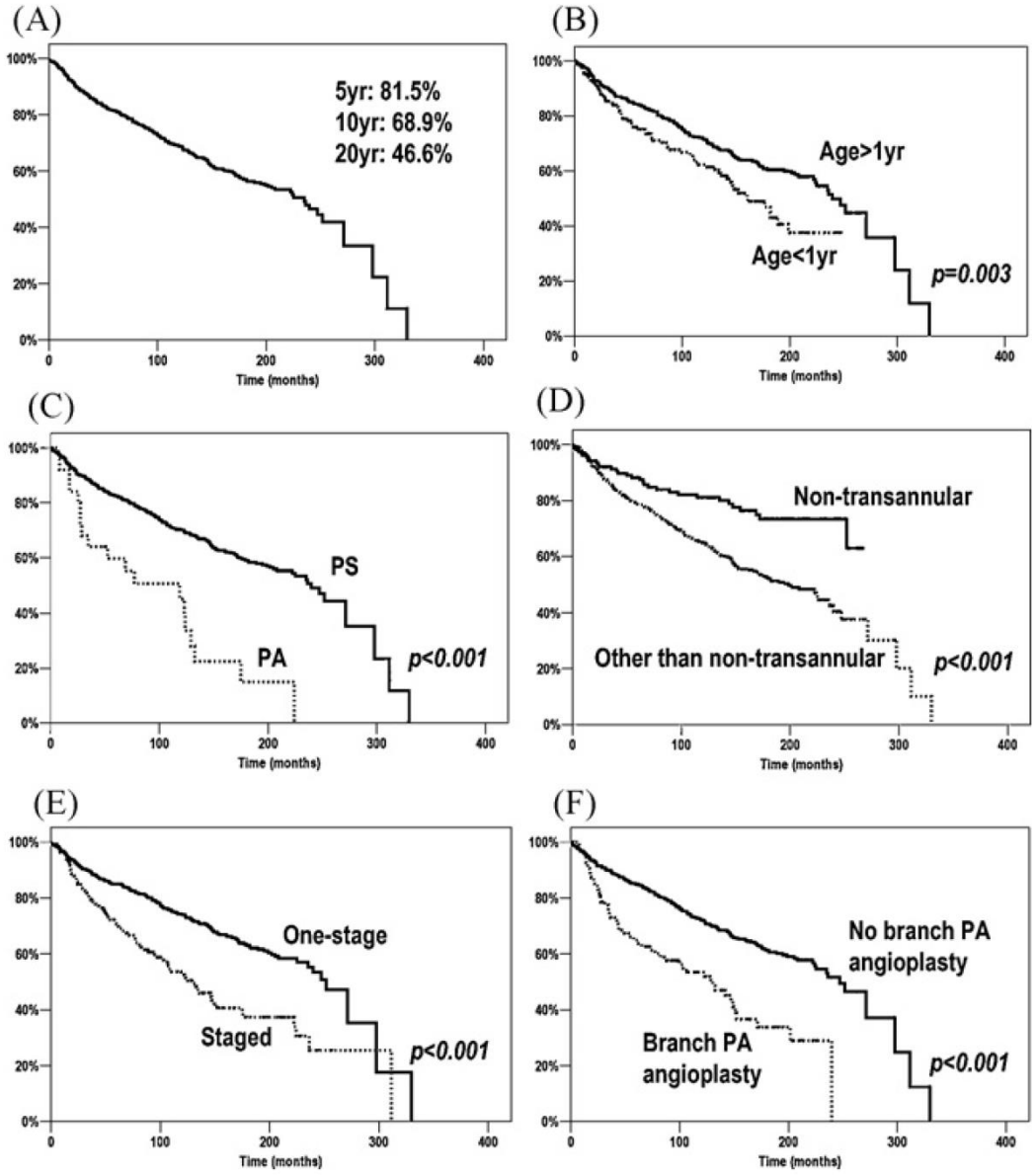
Objective: The objective of this study was to evaluate the long-term outcome of total repair for tetralogy of Fallot. **Methods:** Between April 1986 and December 2007, a total of 734 patients underwent total repair for tetralogy of Fallot. There were 444 males and 290 females. The median age and weight were 17.2 months (0.4–329.6 months) and 9.5 kg (2.6–53.5 kg). The median follow-up duration was 150.2 months (1.9–356.2 months). **Results:** There were 27 early deaths (3.7%) and 13 late deaths. A longer cardiopulmonary bypass time and the use of total circulatory arrest were risk factors for early death. The overall survival rate was 94.8%, 92.8% and 92.8% at 10, 20 and 25 years, respectively. The presence of pulmonary atresia was a risk factor for long-term survival. Re-operation or re-intervention was required in 224 patients (31.7%). The most common causes of re-operation or re-intervention were pulmonary regurgitation in 109 patients and branch pulmonary artery stenosis in 127 patients. Freedom from re-operation or re-intervention rate was 81.5%, 68.9% and 46.6% at 5, 10 and 20 years, respectively. Reconstruction of the right ventricular outflow tract with other than non-trans-annular repair and branch pulmonary arterioplasty at the time of total repair were the risk factors for late re-operation or re-intervention. The use of a monocusp patch was not associated with early mortality or re-operation. At the latest follow-up, most patients were in the New York Heart Association functional class 1 or 2. **Conclusions:** The long-term outcome of total repair for tetralogy of Fallot was satisfactory. A longer cardiopulmonary bypass time and the use of deep hypothermic circulatory arrest were associated with early mortality. The patients with pulmonary atresia have poorer late survival. Preservation of the pulmonary annulus can reduce the re-operation rate. A small pulmonary artery that requires augmentation may increase the risk of re-operation. The use of a monocusp in patients who underwent trans-annular repair has no benefit for early survival, the postoperative recovery and avoidance of re-operation. Age was not a risk factor for early mortality and re-operation.

Keywords: Congenital heart disease; Long-term outcome; Tetralogy of Fallot

Survival


(A)
100%
80%
60%
40%
20%
0%
10yr: 94.8%
20yr: 92.8%
25yr: 92.8%
0
100
200
300
400
Time (months)
(B)
PS
PA
p<0.001
Time (months)
(C)
Non-transannular
Other than non-transannular
p=0.017
Time (months)
(D)
Transatrial
Transventricular
p=0.019
Time (months)

Reoperation or Reintervention



PVR in PR after TOF Repair (EJCTS 2012, SNUH)

European Journal of Cardio-Thoracic Surgery 42 (2012) e1–e8
doi:10.1093/ejcts/ezs219 Advance Access publication 4 May 2012

ORIGINAL ARTICLE

Mid-term results of bioprosthetic pulmonary valve replacement in pulmonary regurgitation after tetralogy of Fallot repair

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Abstract

OBJECTIVES: Pulmonary valve replacement (PVR) is performed to reduce right ventricular (RV) volume overload, resulting in improved ventricular function and clinical status. Significant pulmonary regurgitation (PR) after tetralogy of Fallot (TOF) repair could result in RV dysfunction, exercise intolerance, arrhythmia and sudden death. The present study was conducted to investigate the mid-term clinical outcomes of PVR after TOF repair.

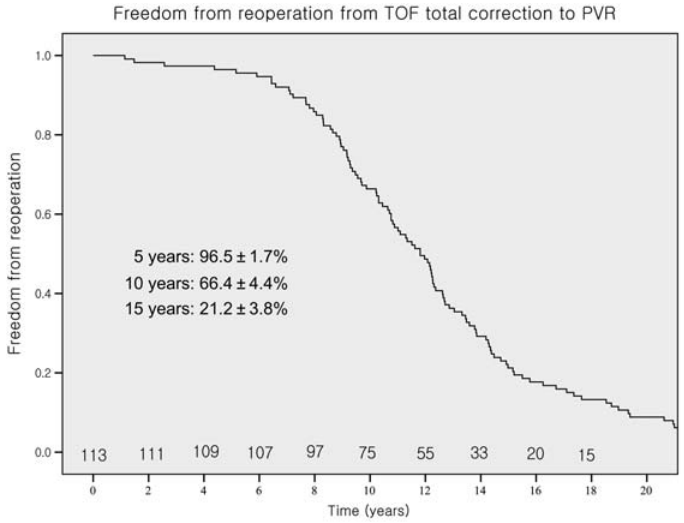
METHODS: Between 2001 and 2010, we retrospectively reviewed the outcomes of 131 (89 males and 42 females) PVRs with PR or pulmonary steno-insufficiency after TOF repair. PVR was performed at a mean age of 14.8 ± 6.7 years. The mean interval from total correction of TOF to PVR was 12.5 ± 5.2 years. Surgical indications of PVR were more than moderate PR with/without pulmonary stenosis, right ventricle dilatation, right ventricle dysfunction and reduced exercise capacity. Hancock II ($n = 58$), Carpentier–Edwards Perimount ($n = 49$) and St Jude Biocor ($n = 35$) bioprosthetic valves were used. The mean z-score at implantation was 1.1 ± 0.8. The mean valve size implanted was 25.1 ± 1.5 mm.

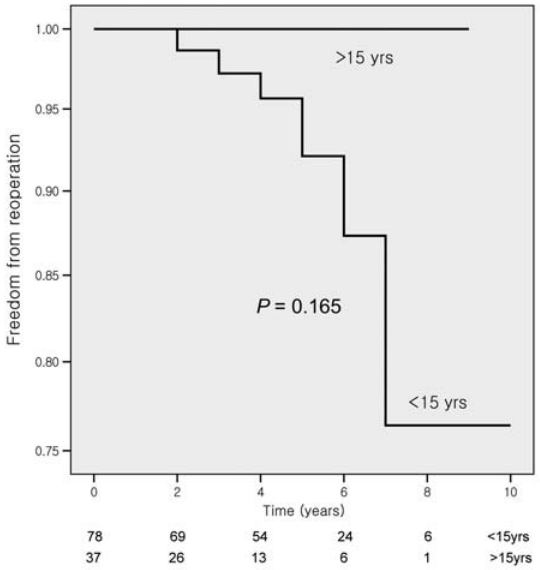
RESULTS: There was no early or late mortality in this study. RV end-diastolic and end-systolic volume indices (from 111.3 ± 34.7 to 64.6 ± 23.6, $P < 0.01$) (preoperative $n = 70$, postoperative n = 17) were markedly decreased PVR during the 13.2 ± 16.1 months follow-up period. Eleven patients (male = 10, female = 1) required a repeat PVR operation due to prosthetic valve failure. The rate of freedom from reoperation at 10 years was 66.4 ± 4.4%. Implanted valve type (Carpentier–Edwards bovine valve), young age, and large-sized valve implantation (z-score > 2.0) were risk factors for a repeat PVR in the univariate analysis. There was no risk factor in the multivariable analysis.

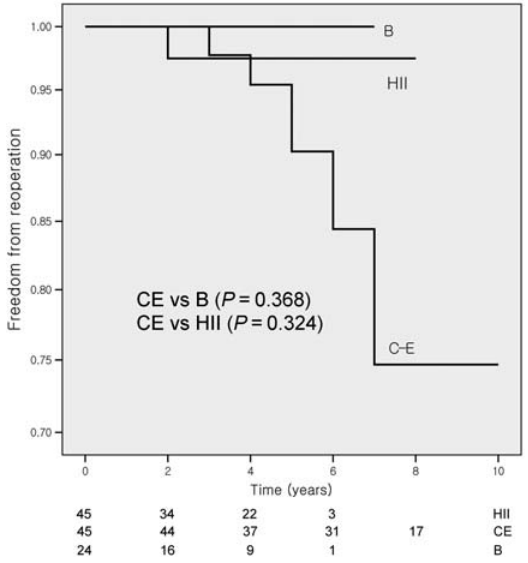
CONCLUSIONS: PVR reduced the RV volume and improved the RV function within the first postoperative year. The rate of freedom from reoperation during the 10-year follow-up period in our series was acceptable. However, a longer follow-up will be necessary to determine the long-term outcomes of bioprosthetic valves in PVR.

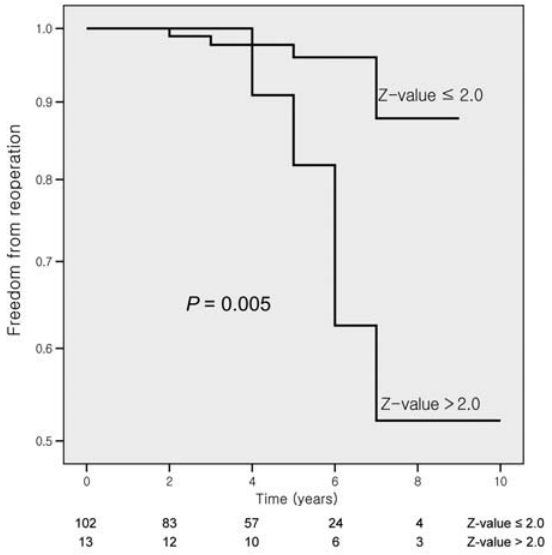
Keywords: Pulmonary valve replacement • Bioprosthetic valve • Tetralogy of Fallot

Reoperation after PVR









Transcatheter Pulmonic Valve (IJC 2014, SNUH)

International Journal of Cardiology 173 (2014) 74–79

Contents lists available at ScienceDirect

International Journal of Cardiology

journal homepage: www.elsevier.com/locate/ijcard

Novel self-expandable, stent-based transcatheter pulmonic valve: A preclinical animal study

Gi Beom Kim [a,d,1], Hong-Gook Lim [b,d,1], Yong Jin Kim [b,d,*,1], Eun Young Choi [c,d,1], Bo Sang Kwon [a,d,1], Saeromi Jeong [d,1]

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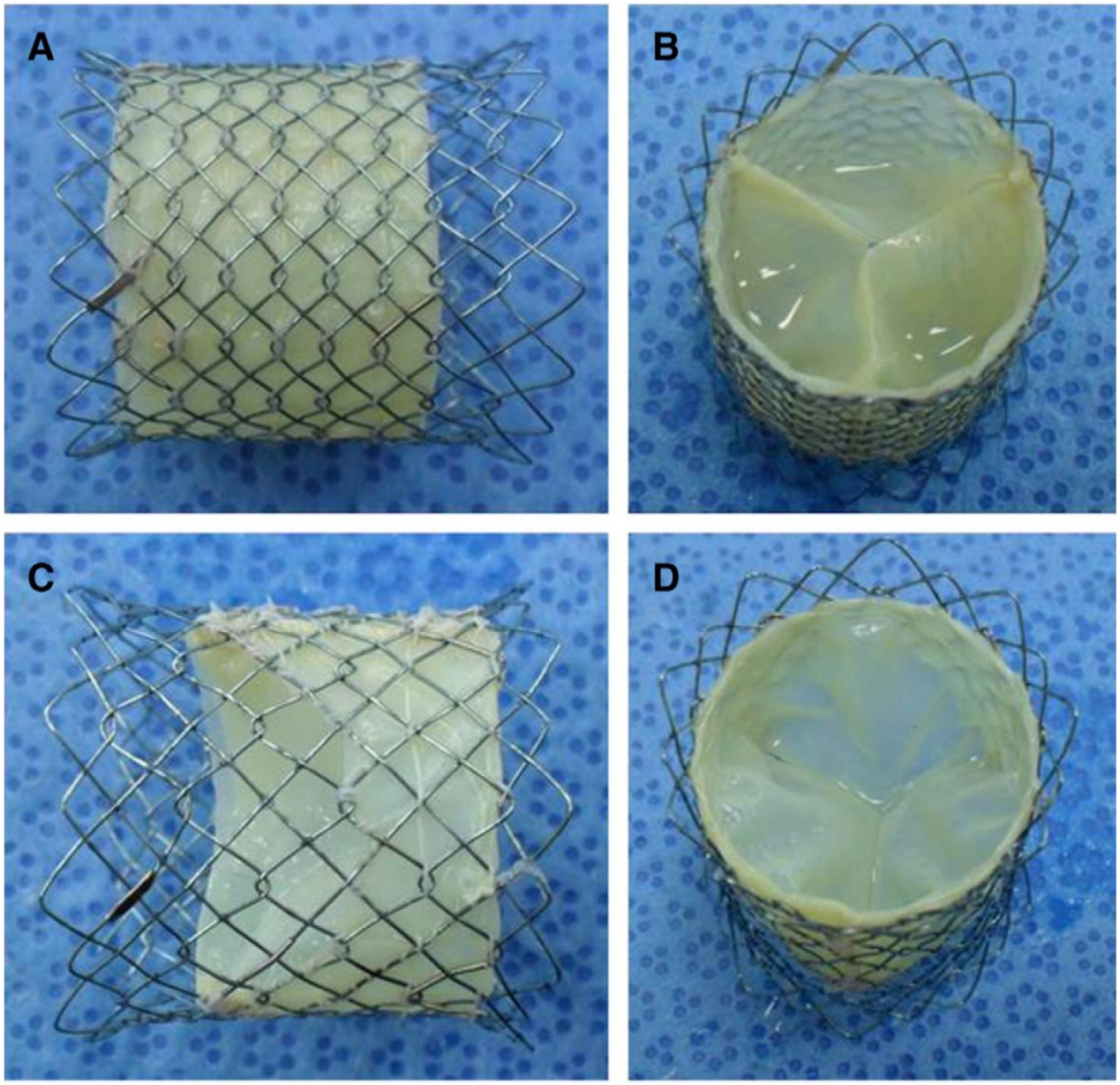
Nitinol
α-Galactosidase
Catheters

ABSTRACT

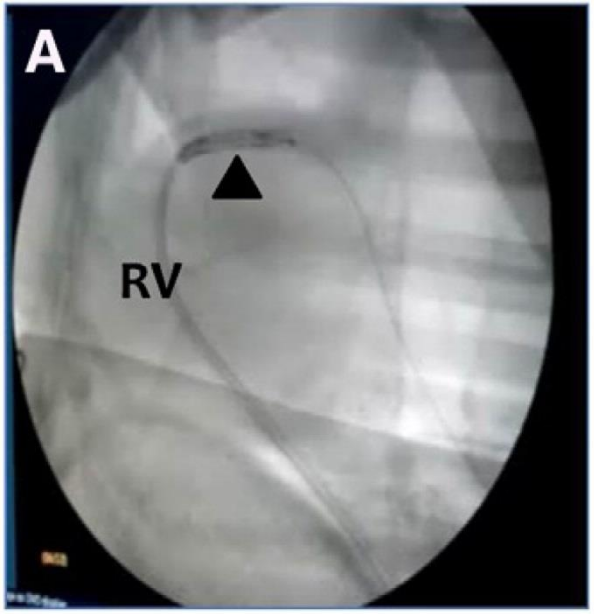
Background: Because transcatheter implantation of pulmonary valve is indicated for limited-size dysfunctional right ventricular outflow tract only as a balloon-expandable stent, we investigated the feasibility of a large-diameter self-expandable valved stent and the durability of the valve after >6 months.
Methods: We made a nitinol-wire-based, self-expandable valved stent with leaflets made from porcine pericardium. The porcine pericardium was treated with α-galactosidase, glutaraldehyde, and glycine after decellularization. After cutting the inguinal or cervical area, we implanted a valved stent in 12 sheep through the femoral or jugular vein by using an 18-Fr delivery catheter, controlling the catheter handles and hook block under fluoroscopic and echocardiographic guidance.
Results: The mean body weight of sheep was 43.9 kg. We successfully implanted valved stents (diameter: 24 mm in 7 sheep, 26 mm in 5 sheep) in good position in 8 sheep, in the main pulmonary artery (PA) in 2 sheep, and in the right ventricular outlet tract (RVOT) in 2 sheep. We sacrificed 8 sheep (6 sheep in good position, 1 sheep in the main PA, and 1 sheep in the RVOT) after >6 months. Five of the 6 sheep implanted in good position showed well-preserved valve morphology at the time of sacrifice. Histologic findings after routine sacrifice showed well-maintained collagen wave structure and no visible calcification in all explanted valve leaflets.
Conclusions: Transcatheter implantation of a nitinol-wire-based, self-expandable valved stent in the pulmonic valve was feasible, and stents implanted in good position showed well-preserved valve leaflets with functional competence in the mid-term results.

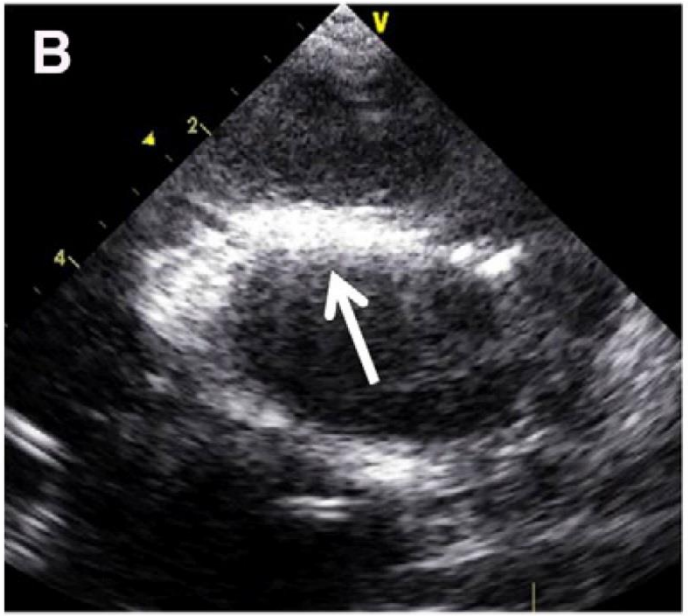
Valved Stent

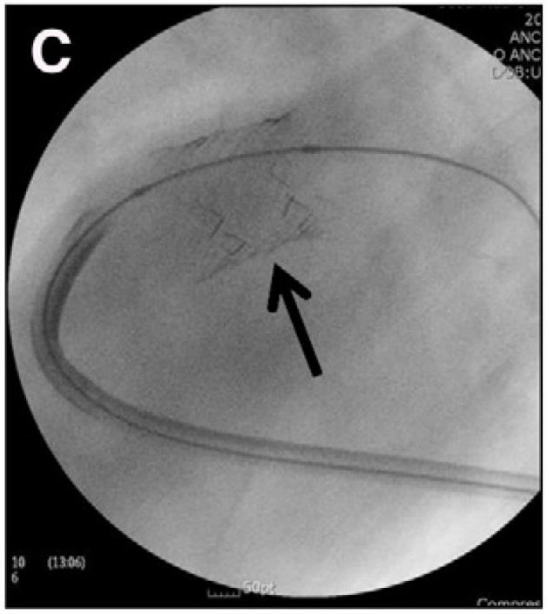


Outcome of TPV implantation

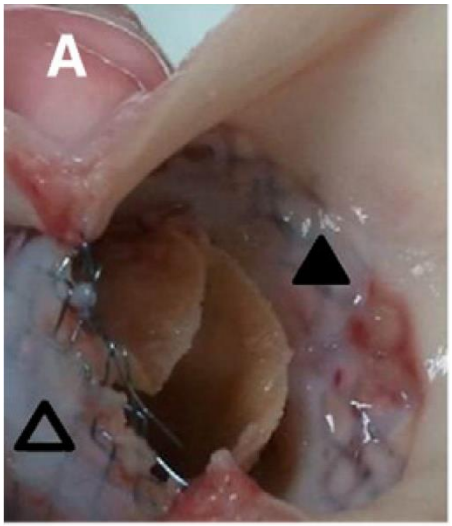
Sheep	Body weight (kg)	Valve diameter (mm)	Stent type	Hook block	Route	Valve position	F/U duration (months)	PR[a]	Peak PG[b] (RV-PA) (mm Hg)	Result	Valve morphology[c]	Calcium deposition (μg/mg)
1	34.5	24	D	−	FV	Good	6	Mild	11	Sacrificed	Tissue loss	4.4
2	45	24	D	−	FV	PA distal	6	No	3	Sacrificed	Attached to wall	4.0
3	51	26	D	−	FV	Good	2.5	No	NA	Died	Thick leaflet	6.8
4	45	26	D	−	FV	RVOT	6	No	8	Sacrificed	Attached to wall	4.4
5	47	26	D	−	FV	Good	6	No	11	Sacrificed	Good	5.38
6	41	24	D	−	FV	Good	6	Trivial	1	Sacrificed	Good	4.4
7	39	24	D	−	JV	Good	6	No	7	Sacrificed	Good	3.2
8	48	24	D	−	FV	RVOT	4	No	NA	Died	Attached to wall	3.2
9	39	24	D	−	FV	PA distal	2	No	NA	Died	Attached to wall	3.2
10	45	26	M	+	JV	Good	4	Moderate	NA	Died	Tissue loss	4.4
11	40	24	M	+	JV	Good	10	Trivial	1	Sacrificed	Good	3.92
12	53	26	M	+	JV	Good	7	No	6	Sacrificed	Good	4.07
Mean	43.9	24.8							6			

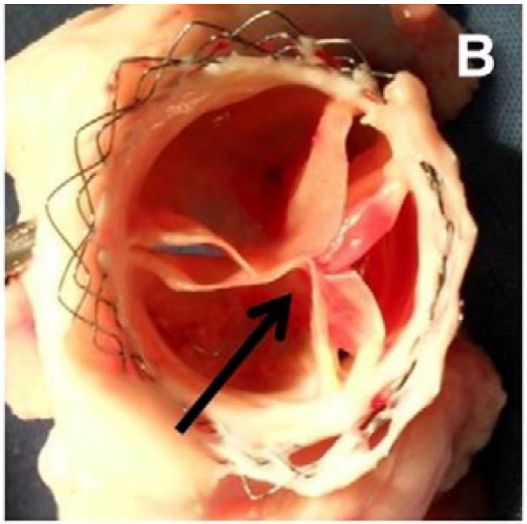


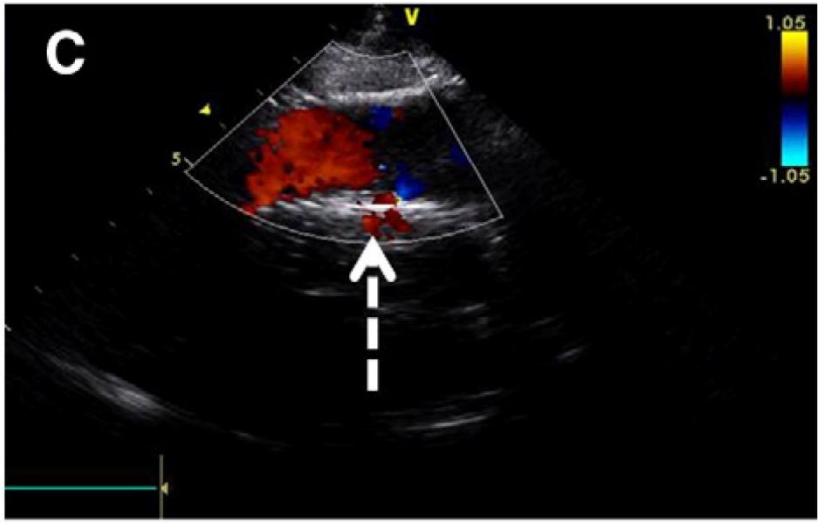


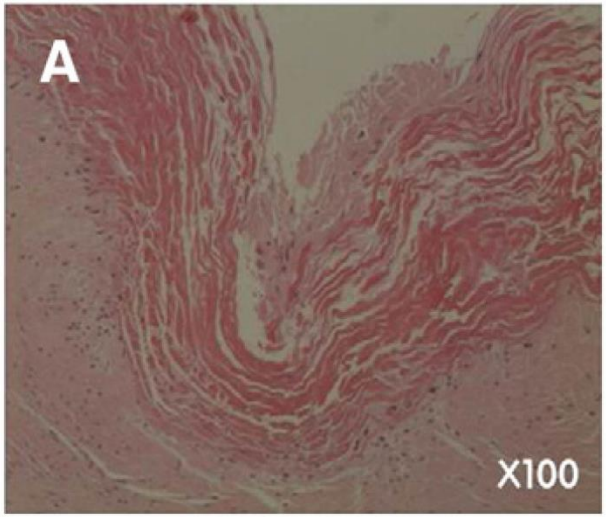


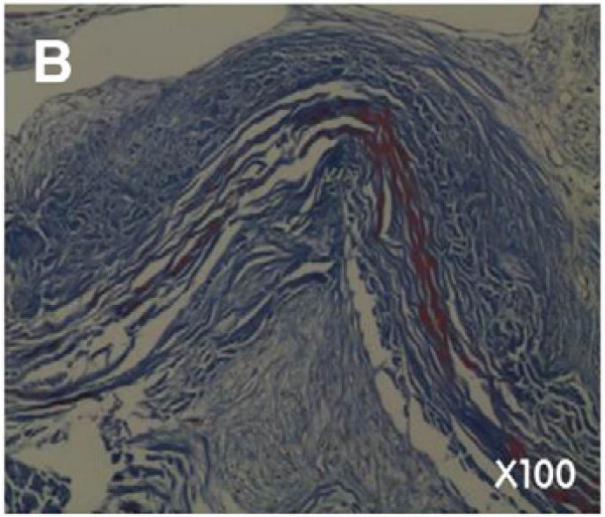
Outcome of TPV implantation

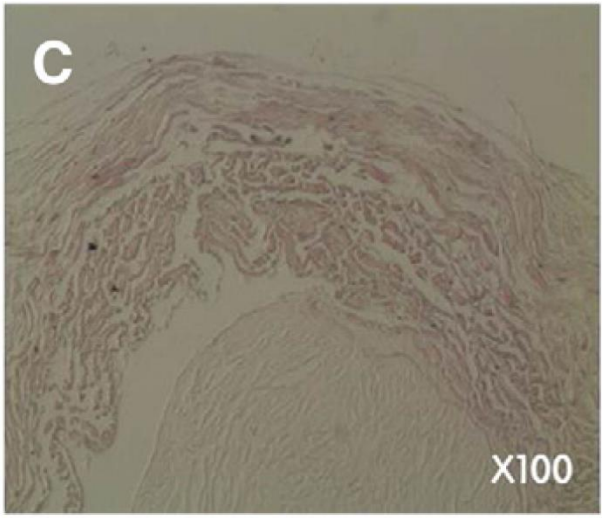












TPV in Human Experience (CCI 2017, SNUH)

Catheterization and Cardiovascular Interventions 00:00–00 (2017)

Case Report

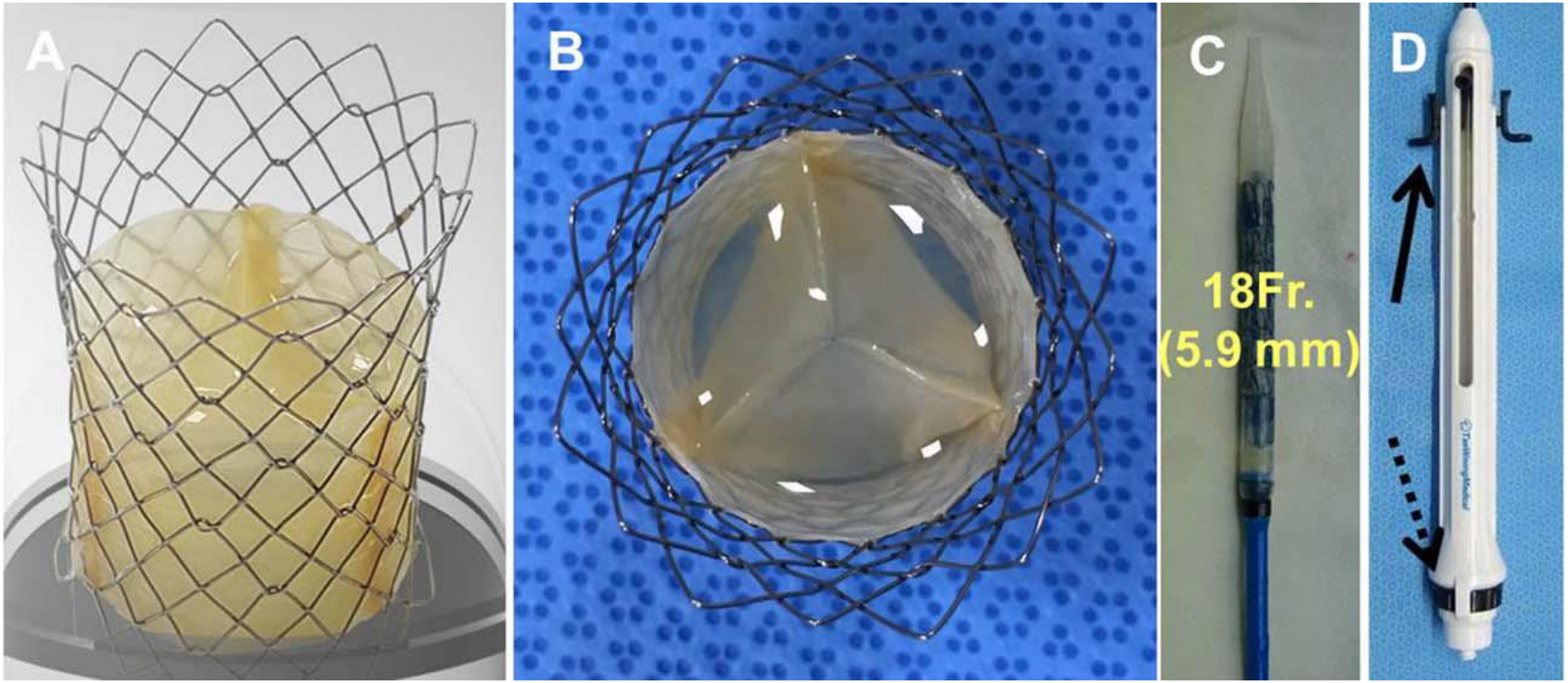
First in Human Experience of a New Self-Expandable Percutaneous Pulmonary Valve Implantation Using Knitted Nitinol-Wire and Tri-Leaflet Porcine Pericardial Valve in the Native Right Ventricular Outflow Tract

Gi Beom Kim,[1*] Bo Sang Kwon,[1] and Hong Gook Lim[2]

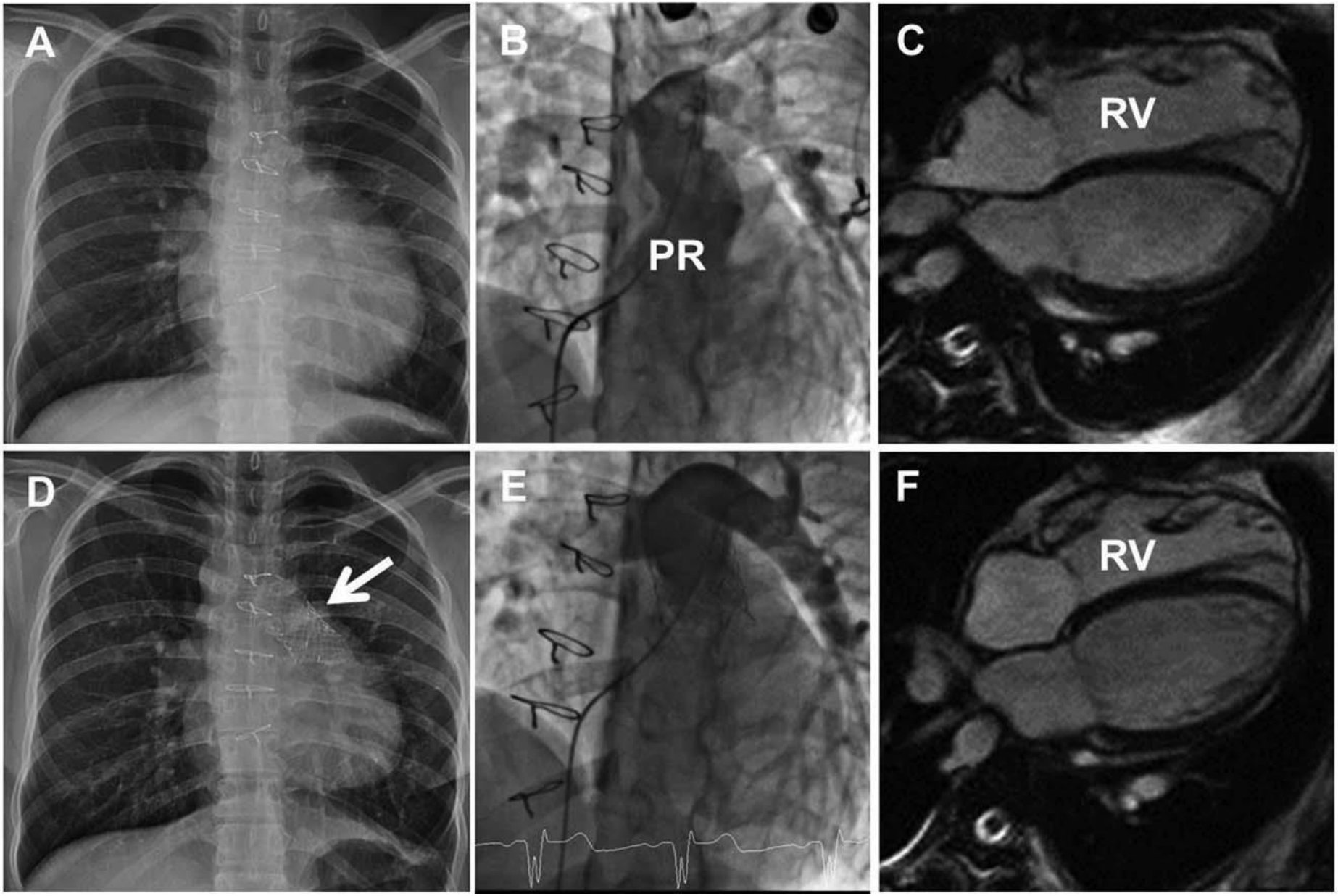
Balloon-expandable percutaneous pulmonary valve systems using the Melody and Edwards SAPIEN transcatheter heart valves have been increasingly used instead of surgically implantable pulmonary valves. However, limited patients with right ventricular outflow tract (RVOT) lesions are suitable candidates for percutaneous pulmonary valve implantation (PPVI) using these systems after surgical correction of tetralogy of Fallot. Therefore, larger self-expandable valved-stents are being developed for native RVOT lesions. We report the first-in-human case of a new self-expandable PPVI in a patient with a native RVOT lesion using a newly made knitted nitinol-wire stent mounted with a tri-leaflet porcine pericardial valve developed in South Korea.

Key words: pulmonary valve insufficiency; alpha-galactosidase; xenograft transplantation

TPV in Human Experience



TPV in Human Experience



Long-Term Results of ASO (EJCTS 2013, SNUH)

European Journal of Cardio-Thoracic Surgery 43 (2013) 325–334
doi:10.1093/ejcts/ezs264 Advance Access publication 9 May 2012

ORIGINAL ARTICLE

Long-term results of the arterial switch operation for ventriculo-arterial discordance

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Received 19 January 2012; received in revised form 28 February 2012; accepted 23 March 2012

Abstract

OBJECTIVES: The arterial switch operation (ASO) has become the standard surgical procedure for transposition of the great arteries (TGA) or variants with an excellent early outcome. However, there are concerns regarding neopulmonary stenosis, neoaortic regurgitation (neoAR) associated with neoaortic root dilatation and coronary artery disease.

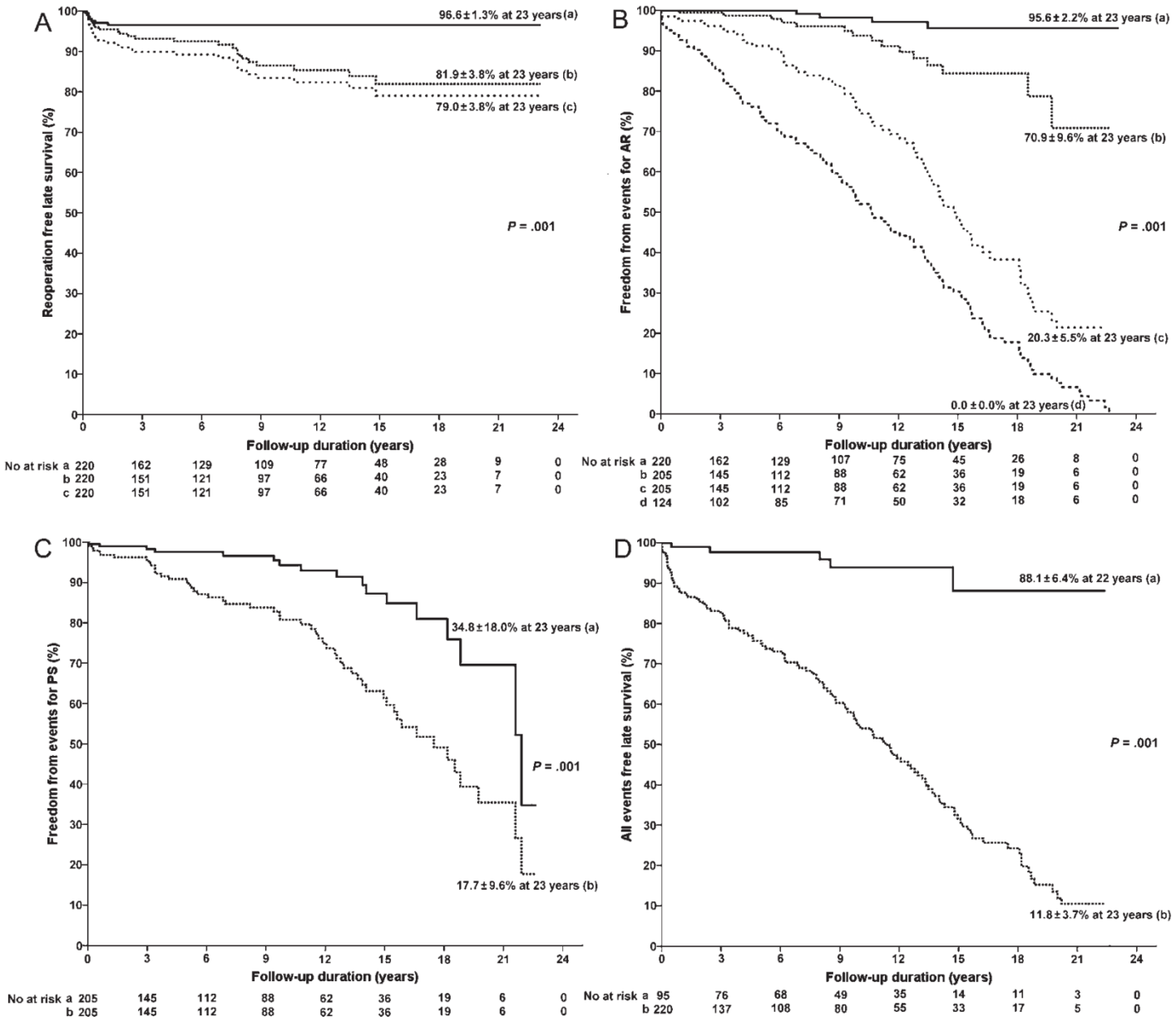
METHODS: A total of 220 early survivors of the ASO were included in this retrospective study between November 1987 and June 2011. The median age and weight at operation were 13 days (0–1768 days) and 3.52 kg (1.69–19 kg), respectively. The indications for the ASO included TGA with intact ventricular septum in 113 patients, TGA with ventricular septal defect in 90 and Taussig–Bing anomaly in 17 patients. The median follow-up period was 103.2 months (0.4–277.4 months). Statistical analyses with the Kaplan–Meier and Cox proportional hazards models were performed.

RESULTS: The actuarial late survival rate and freedom from reoperation at 23 years were 96.6 ± 1.3 and 81.9 ± 3.8%, respectively. Twenty-four (10.9%) patients underwent reoperations for right ventricular outflow tract obstruction in 10 patients, neoAR in four and coronary artery stenosis in three, etc. Freedom from neoAR of Grades IV, III and II at 23 years was 90.2 ± 6.6, 70.9 ± 9.6 and 20.3 ± 5.5%, respectively. The risk factors for neoAR were size discrepancy of the great arteries, aortic root dilatation after the ASO and follow-up duration after the ASO. NeoAR was significantly correlated with the size of aortic sinus and aortic sinotubular junction over time. Freedom from pulmonary stenosis (PS) of ≥36 and ≥20 mmHg at 23 years was 34.8 ± 18.0 and 17.7 ± 9.6%, respectively. The risk factors for PS were Taussig–Bing and arch anomalies. Coronary artery evaluation was performed in 95 (43.2%) patients with angiography, computed tomography or single-photon emission computed tomography, and five (5.3%) patients had abnormal coronary morphology or perfusion. Three patients underwent reoperation for coronary artery stenosis, and two had reversible perfusion defects in various regions, which were clinically not significant. Freedom from coronary events was 88.1 ± 6.4% at 22 years. A risk factor for coronary events was the single coronary artery.

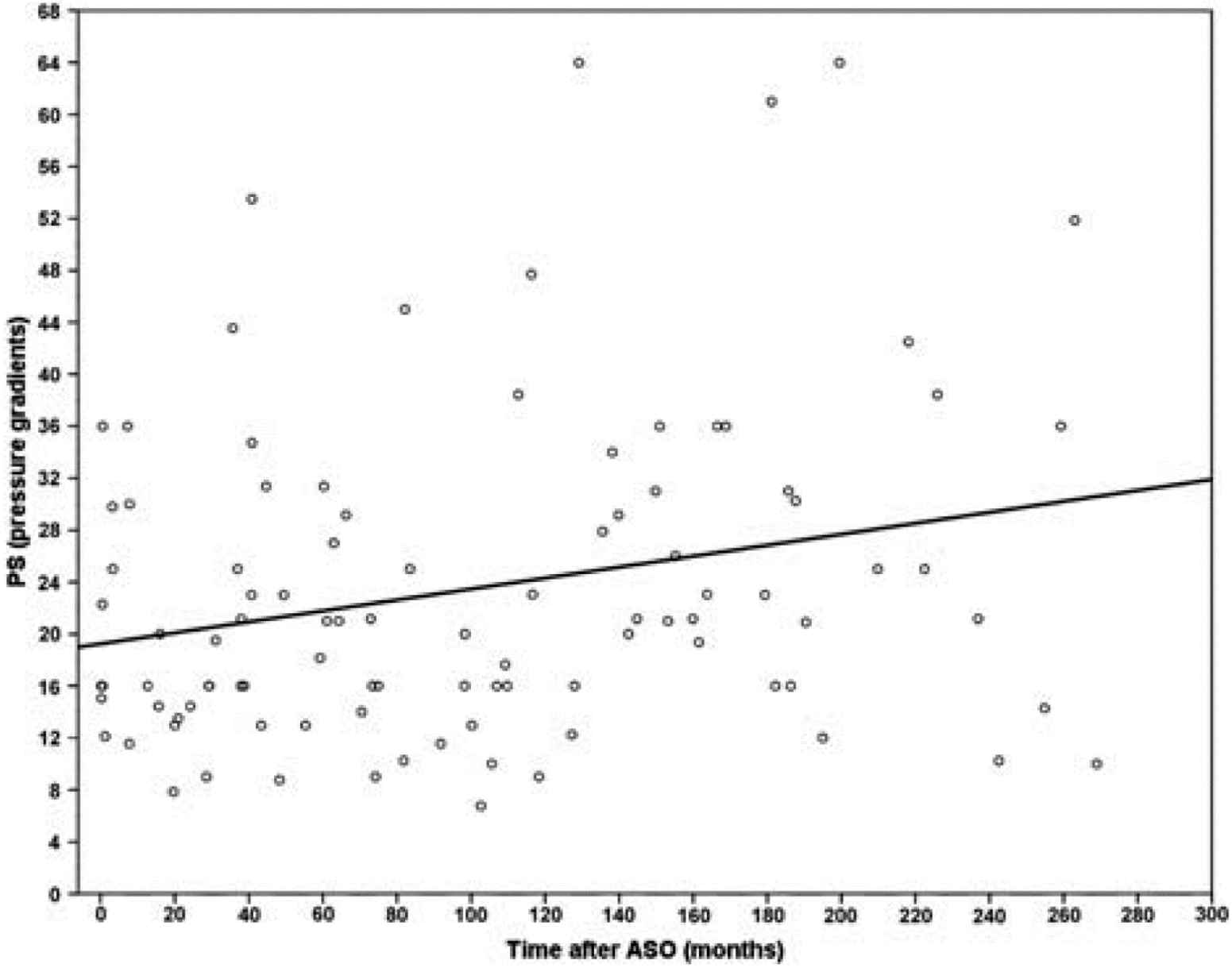
CONCLUSIONS: The survival and functional outcomes of the ASO were excellent in the long-term. Strict serial surveillance is required to evaluate the long-term functional outcome of the ASO, particularly in a high-risk anatomy.

Keywords: Arterial switch operation • Transposition of the great arteries • Taussig–Bing anomaly • Aortic regurgitation • Pulmonary stenosis

Long-Term Results of ASO (EJCTS 2013, SNUH)



Long-Term Results of ASO (EJCTS 2013, SNUH)



REV for VA discordance with VSD, PS (JTCS 2014, SNUH)

Twenty-five years' experience of modified Lecompte procedure for the anomalies of ventriculoarterial connection with ventricular septal defect and pulmonary stenosis

Hong-Gook Lim, MD, PhD, Woong-Han Kim, MD, PhD, Jeong Ryul Lee, MD, PhD, and Yong Jin Kim, MD, PhD

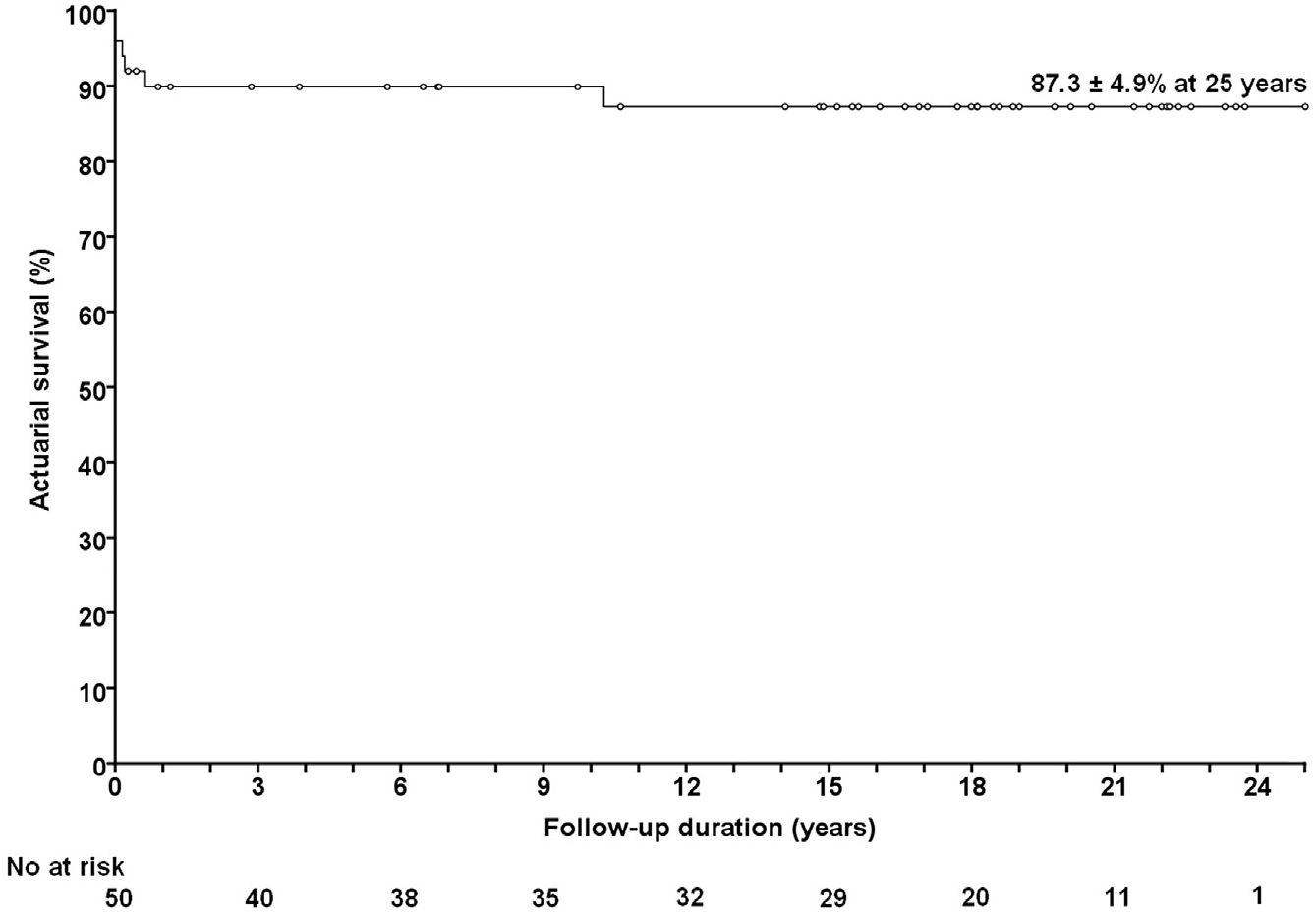
Objectives: To overcome the drawbacks of the Rastelli operation, the modified Lecompte procedure creates a connection from the left ventricle to the aorta with greater freedom from residual obstruction due to the resection of the outlet septum and avoids the implantation of an extracardiac valved conduit. We evaluated the effectiveness of this technique with analysis of our 25-year long-term results.

Methods: We reviewed the records of 50 patients who underwent the modified Lecompte procedure during the past 25 years. The median age at operation was 1.95 years (range, 0.30-12.48 years). The diagnoses involved anomalies of the ventriculoarterial connection with ventricular septal defect and pulmonary outflow tract obstruction, such as transposition of the great arteries, double outlet right ventricle, and double outlet left ventricle.

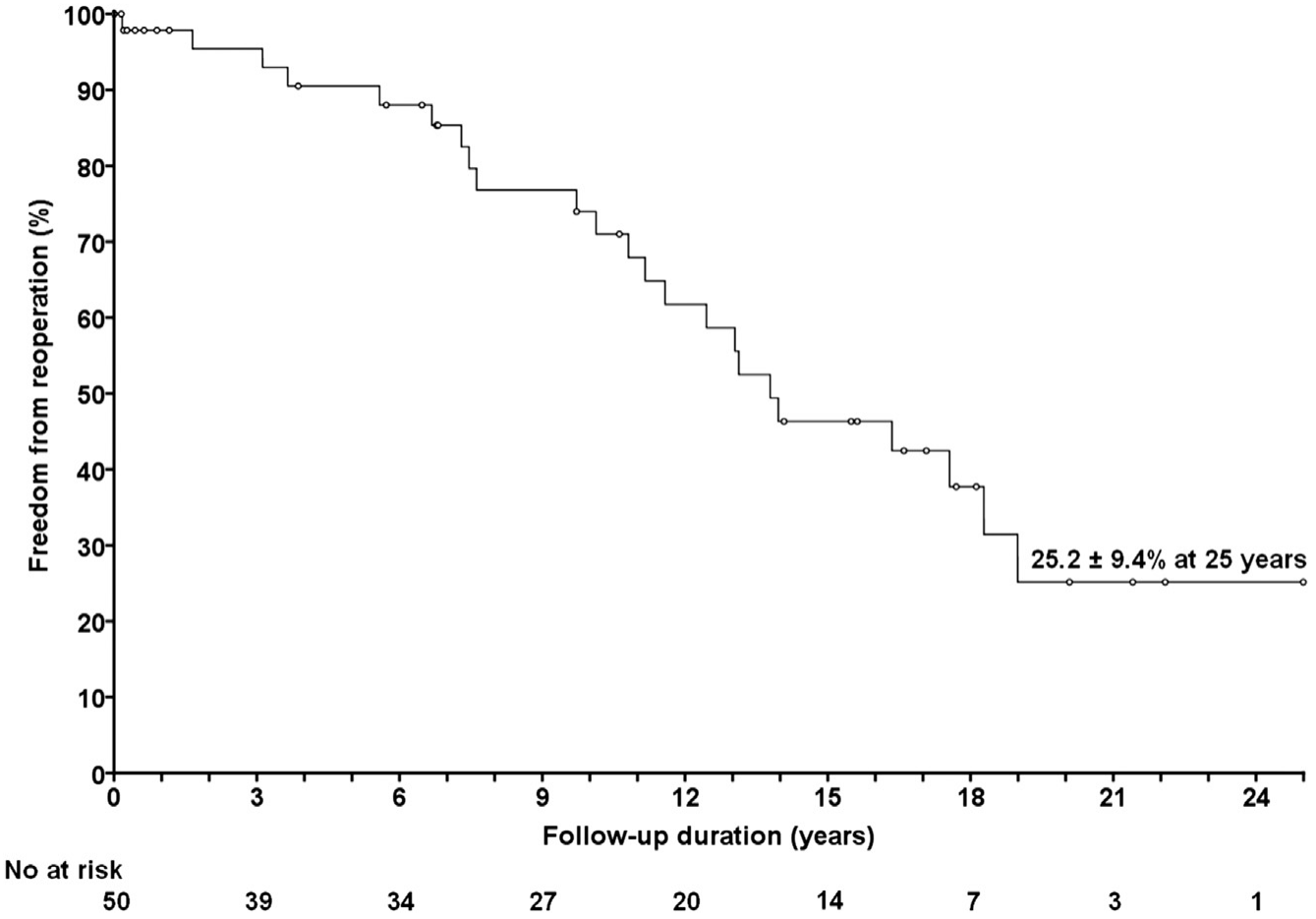
Results: There were 2 early deaths (4.0%). During a mean follow-up of 14.2 ± 7.9 years (range, 0.2-25 years), there were 4 late deaths. Actuarial survival was 87.3% ± 4.9% at 25 years. The freedom from arrhythmia and reoperation at 25 years was 87.7% ± 6.2% and 25.2% ± 9.4%, respectively. The freedom from reoperation for left ventricular outflow tract obstruction and right ventricular outflow tract obstruction at 25 years was 88.5% ± 5.4% and 49.6% ± 9.0%, respectively. At last follow-up, 43 survivors (97.7%) are in New York Heart Association class I.

Conclusions: The modified Lecompte procedure has excellent long-term results for treating anomalies of ventriculoarterial connection with ventricular septal defect and pulmonary outflow tract obstruction. Early repair is possible with low mortality and morbidity in terms of arrhythmia, reoperation for right or left ventricular outflow tract obstruction, and functional class. (J Thorac Cardiovasc Surg 2014;148:825-31)

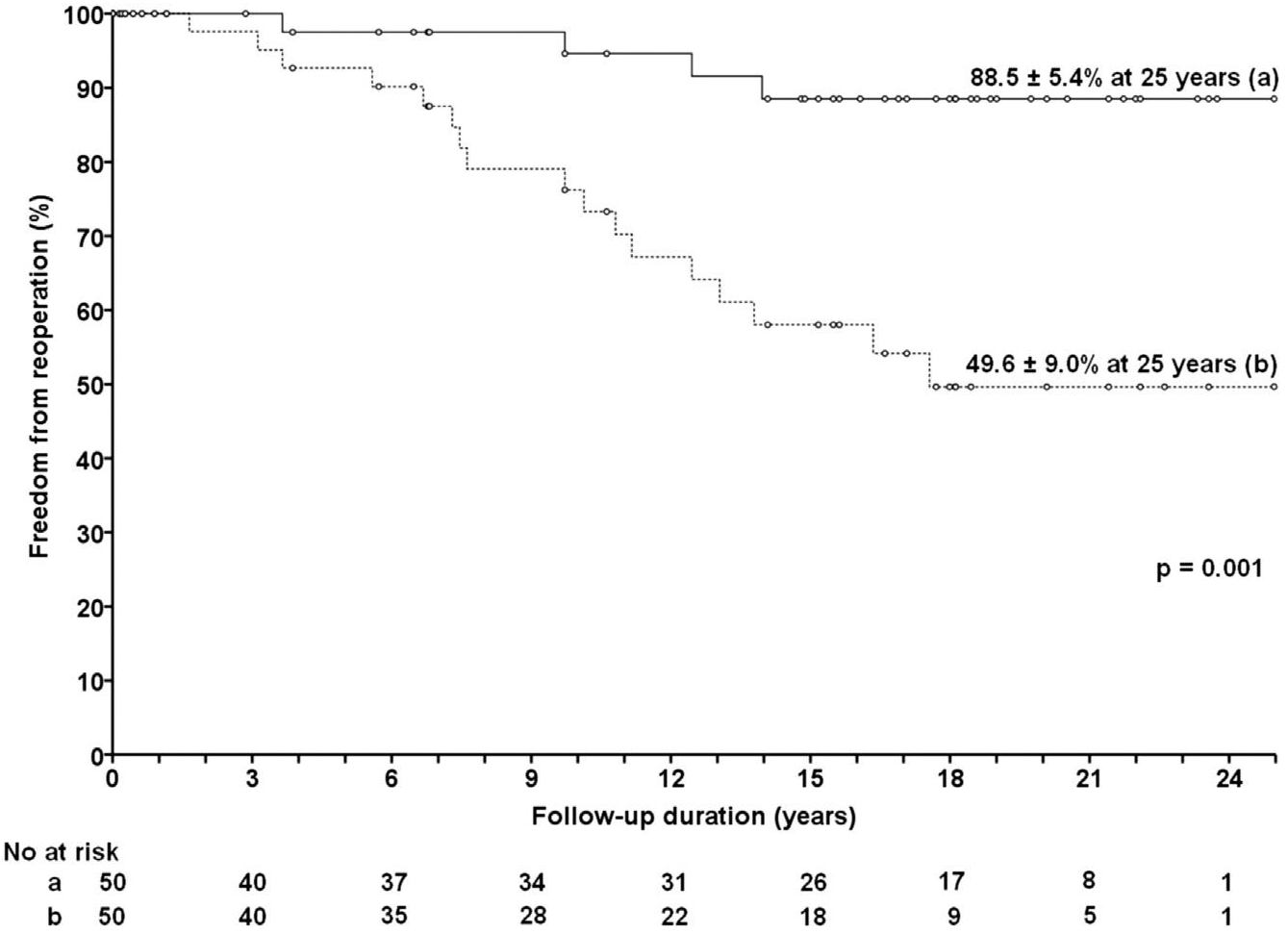
Survival


100
90
80
70
60
50
40
30
20
10
0
Actuarial survival (%)
87.3 ± 4.9% at 25 years
0
3
6
9
12
15
18
21
24
Follow-up duration (years)
No at risk
50
40
38
35
32
29
20
11
1

Reoperation



Reoperation for LVOT and RVOT obstruction



Rastelli vs REV for TGA, VSD, PS (EJCTS 2004, SNUH)

Repair of transposition of the great arteries, ventricular septal defect and left ventricular outflow tract obstruction

Jeong Ryul Lee[a,*], Hong Gook Lim[a], Yong Jin Kim[a], Joon Ryang Rho[a], Eun Jung Bae[b], Chung Il Noh[b], Yong Soo Yun[b], Curie Ahn[c]

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[b]*Department of Pediatrics, Seoul National University Children's Hospital, Seoul National University College of Medicine, Seoul National University Medical Research Center, Xenotransplantation Research Center, 28 Yongon-dong, Jongro-gu, Seoul 110-744, South Korea*
[c]*Department of Internal Medicine, Seoul National University Children's Hospital, Seoul National University College of Medicine, Seoul National University Medical Research Center, Xenotransplantation Research Center, 28 Yongon-dong, Jongro-gu, Seoul 110-744, South Korea*

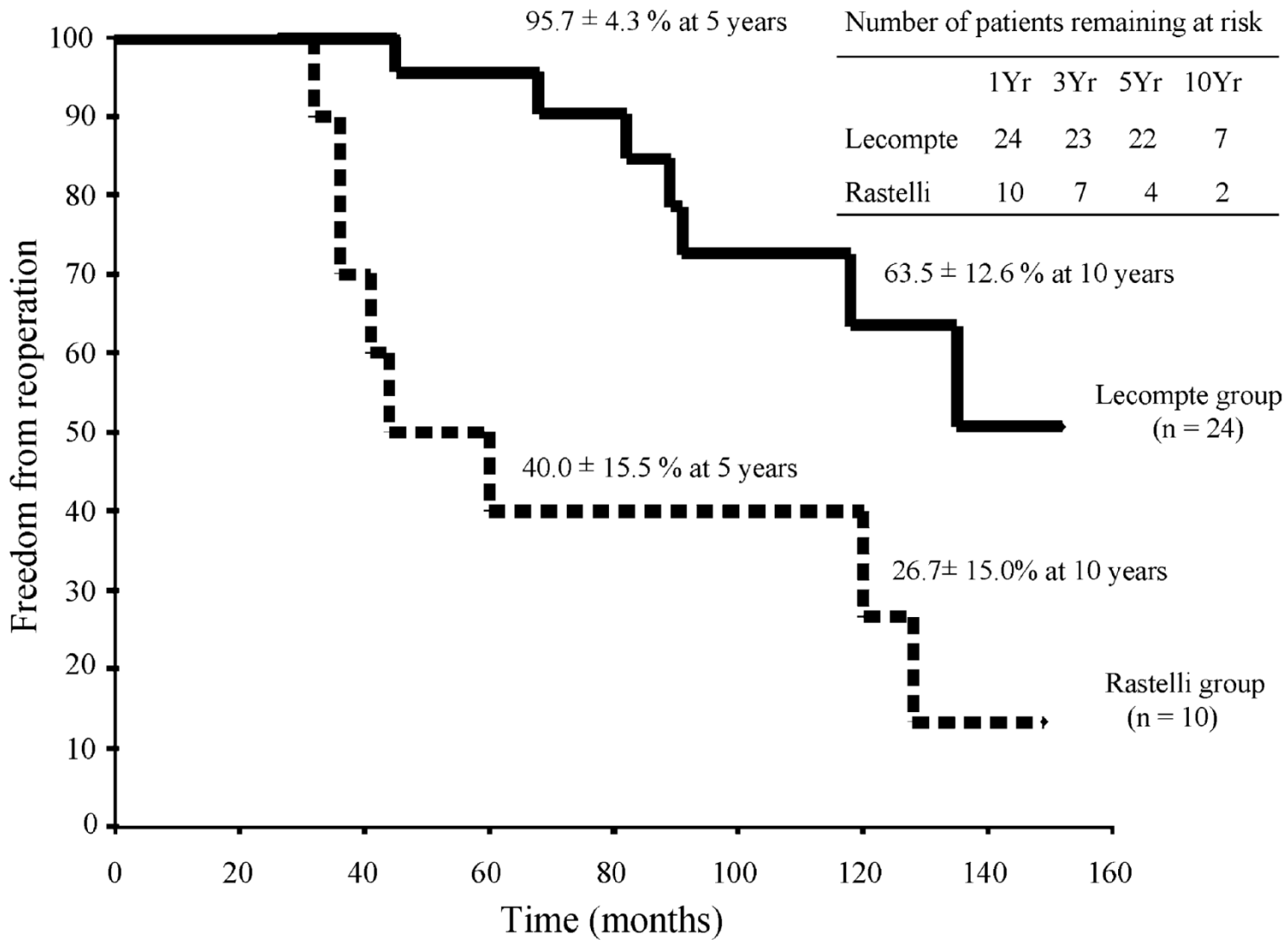
Received 17 September 2003; received in revised form 29 December 2003; accepted 5 January 2004

Abstract

Objectives: This study was undertaken to compare the outcomes of the Lecompte procedure and Rastelli repair in the transposition of the great arteries (TGA) with ventricular septal defect (VSD) and left ventricular outflow tract obstruction (LVOTO) and to determine the risk factors associated with unfavorable events. **Methods**: Over a 12-year period (April 1990–October 2002), 35 patients underwent complete repair for TGA, VSD, and LVOTO. Twenty-five patients (71%) underwent the Lecompte modification, and mean age and weight were 23.4 ± 18.2 months and 10.2 ± 3.0 kg. Ten patients (29%) underwent the Rastelli operation, and mean age and weight were 39.1 ± 36.1 months and 13.8 ± 6.8 kg. **Results**: One early death (3%) occurred after the Lecompte procedure and no late death. The mean follow-up was 5.9 ± 3.8 years. Eight patients in the Rastelli group (80%) underwent a late reoperation for obstruction of the extracardiac conduit, and in four of these patients, a reoperation for LVOTO was concomitantly required. Reoperation was also required in six patients of the Lecompte group (25%); five for right ventricular outflow tract obstruction (RVOTO) including one for LVOTO and two for VSD leakage, and one for mitral regurgitation and left pulmonary artery stenosis. The interval prior to reoperation ranged from 1.6 to 11.1 years, with a mean of 5.7 ± 3.1 years. The actuarial figures for freedom from reoperation at 5 and 10 years were 40.0 ± 15.5 and 26.7 ± 15.0% after the Rastelli operation and 95.7 ± 4.3 and 63.5 ± 12.6% after the Lecompte procedure ($P = 0.02$). Multivariate analysis by Cox regression analysis revealed that the risk factors of RVOTO were a younger age at operation, the Rastelli operation, and ductus ligation during the operation. **Conclusions**: The Lecompte procedure and Rastelli repair provide satisfactory early and late results. However, substantial late morbidity is more associated with conduit obstruction, and LVOTO in Rastelli repair rather than Lecompte procedure.

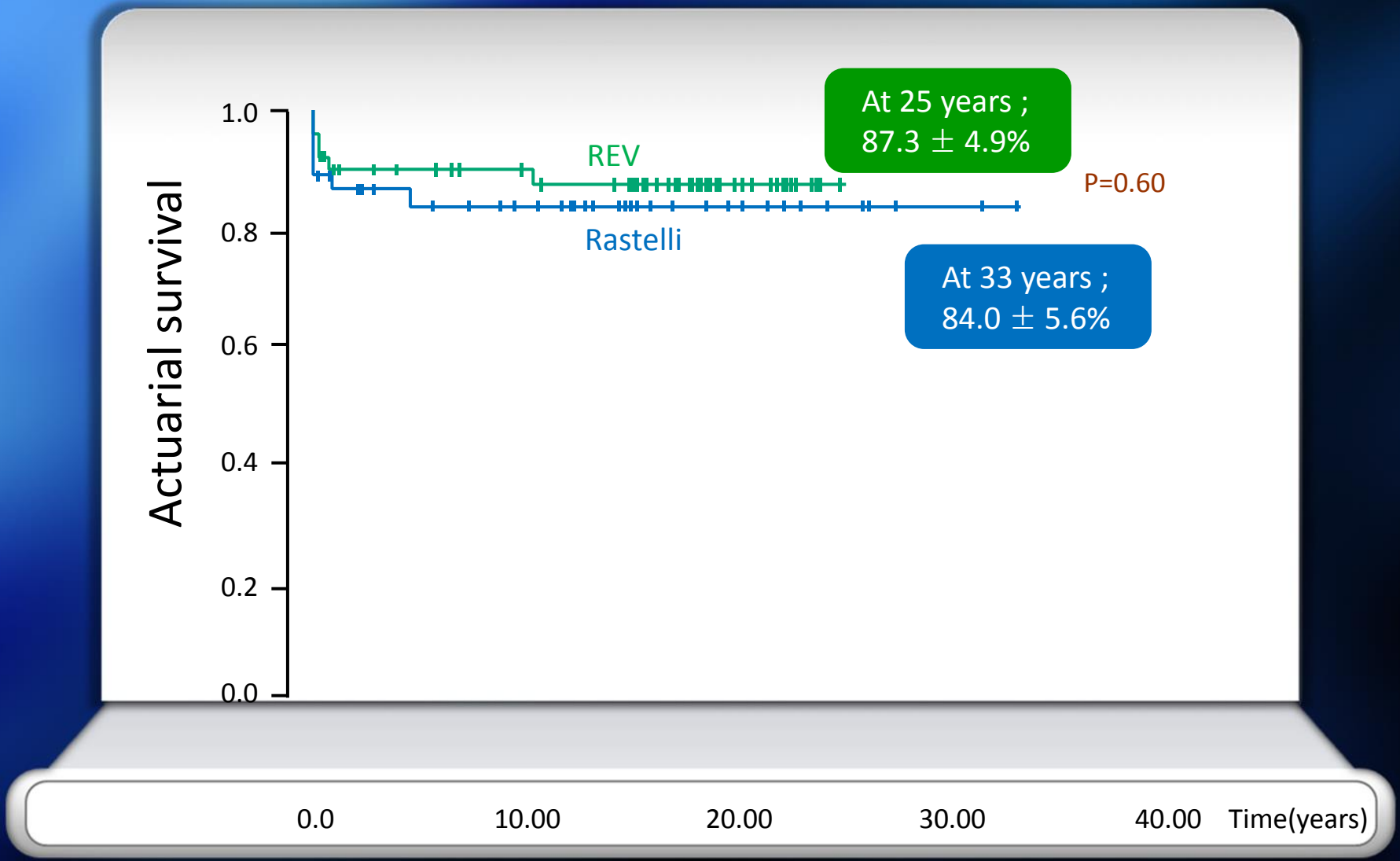
Keywords: Transposition of the great arteries; Rastelli; Lecompte

Rastelli vs REV for TGA, VSD, PS (EJCTS 2004, SNUH)



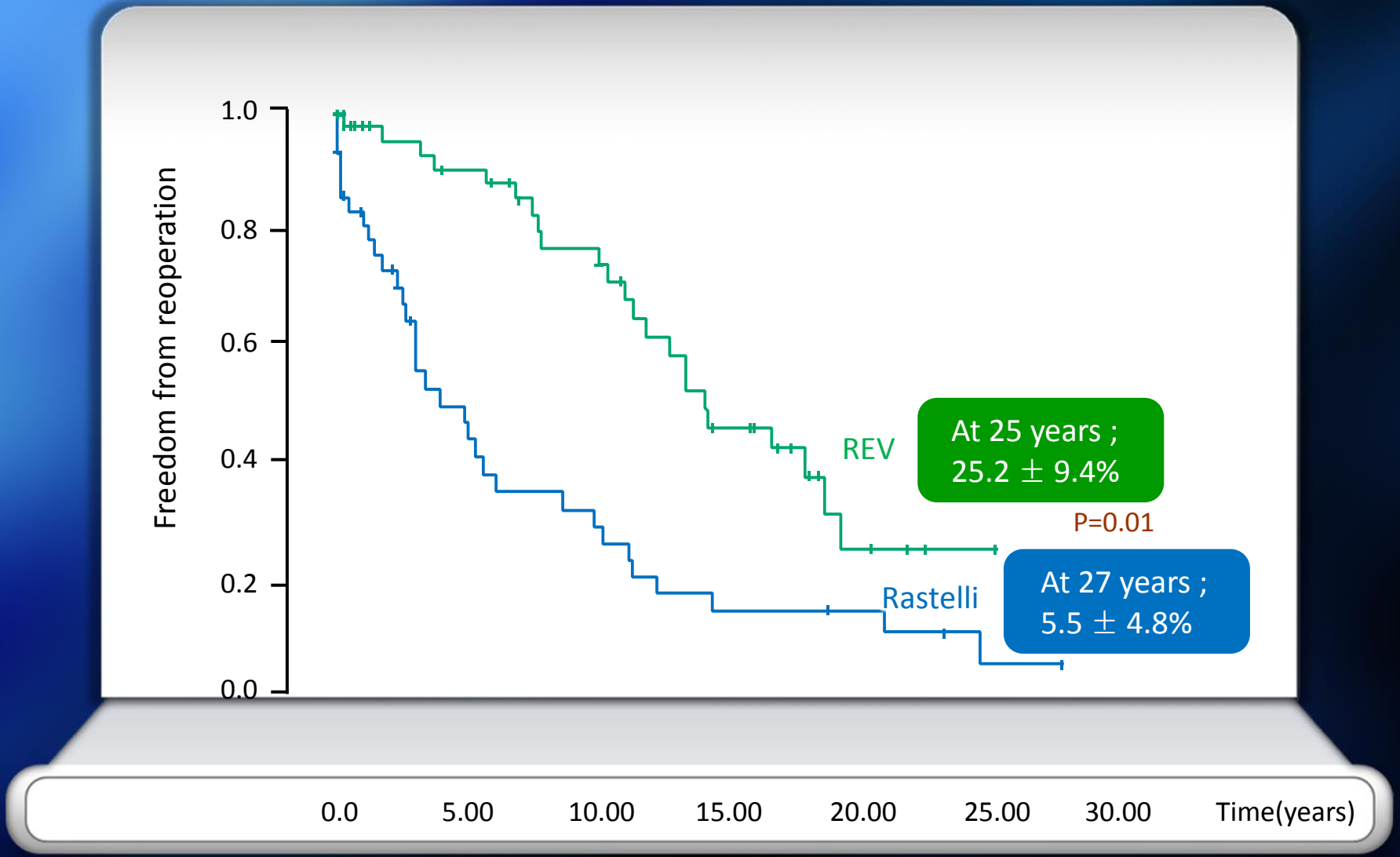
Long-Term Results of Rastelli vs REV (WSCTS 2015, SNUH)

Survival



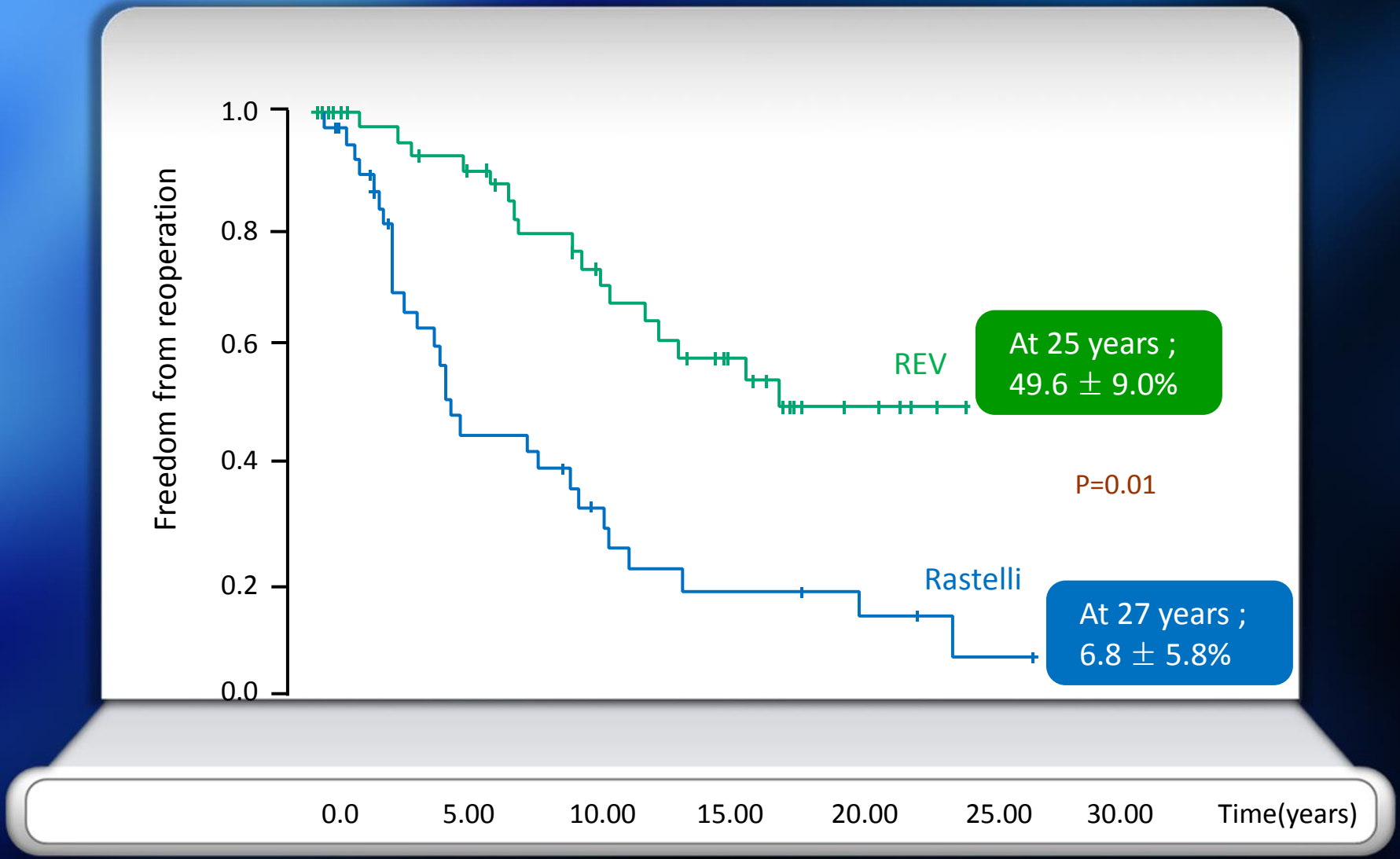
Long-Term Results of Rastelli vs REV (WSCTS 2015, SNUH)

Reoperation



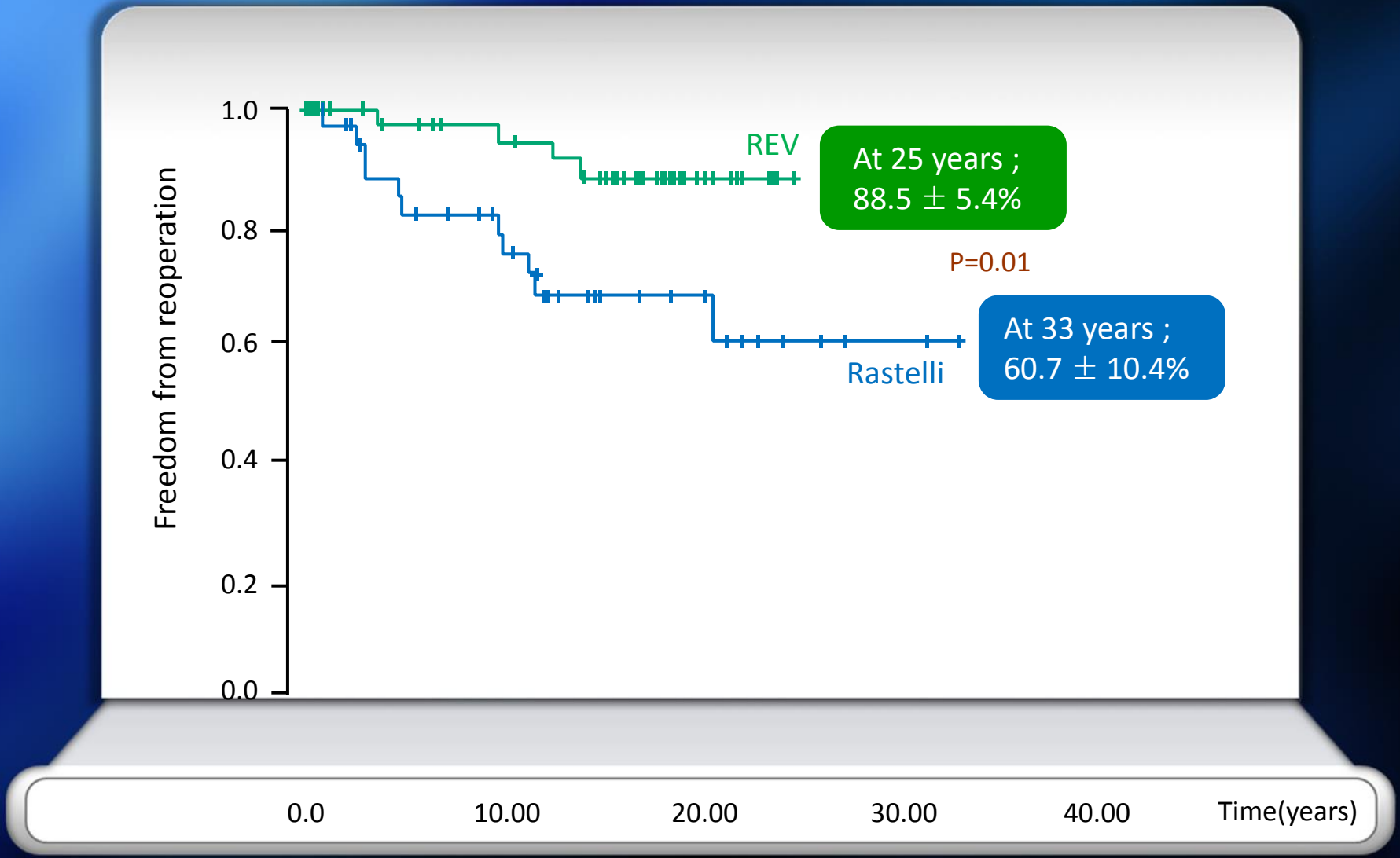
Long-Term Results of Rastelli vs REV (WSCTS 2015, SNUH)

RVOT obstruction



Long-Term Results of Rastelli vs REV (WSCTS 2015, SNUH)

LVOT obstruction



Long-Term Results for Ebstein Repair (KCJ 2016, SNUH)

Long-Term Results after Surgical Treatment of Ebstein's Anomaly: a 30-Year Experience

Min-Seok Kim, MD, Hong-Gook Lim, MD, Woong Han Kim, MD, Jeong Ryul Lee, MD, and Yong Jin Kim, MD

Department of Thoracic and Cardiovascular Surgery, Seoul National University Hospital, Seoul, Korea

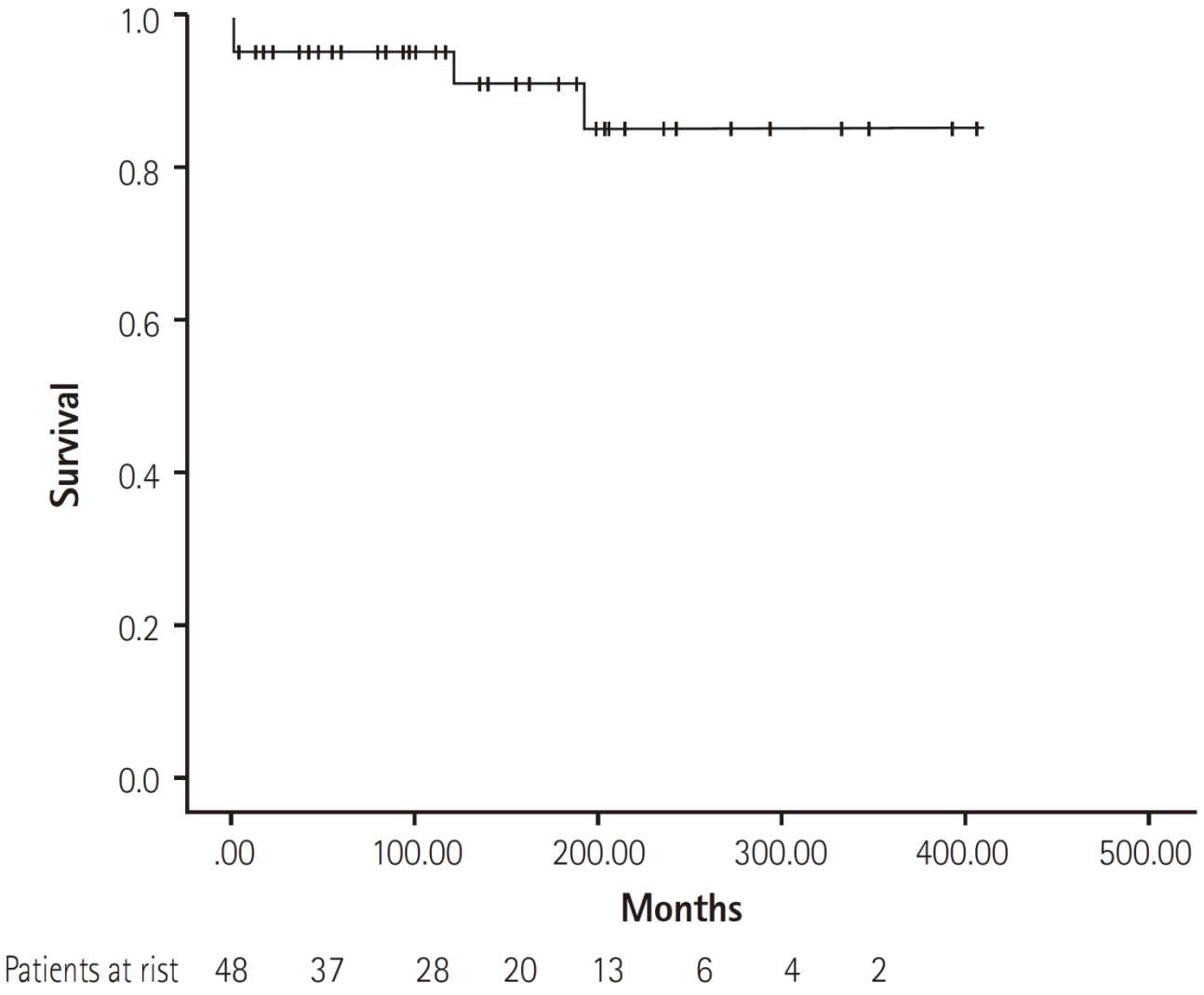
Background and Objectives: The aim of the study is to evaluate the long-term results after a surgical repair of Ebstein's anomaly.

Subjects and Methods: Forty-eight patients with Ebstein's anomaly who underwent open heart surgery between 1982 and 2013 were included. Median age at operation was 5.6 years (1 day-42.1 years). Forty-five patients (93.7%) demonstrated tricuspid valve (TV) regurgitation of less than moderate degree. When the patients were divided according to Carpentier's classification, types A, B, C, and D were 11, 21, 12, and 4 patients, respectively. Regarding the type of surgical treatment, bi-ventricular repair (n=38), one-and-a half ventricular repair (n=5), and single ventricle palliation (n=5) were performed. Of 38 patients who underwent a bi-ventricular repair, TV repairs were performed by Danielson's technique (n=20), Carpentier's technique (n=11), Cone repair (n=4), and TV annuloplasty (n=1). Two patients underwent TV replacement. Surgical treatment strategies were different according to Carpentier's types ($p<0.001$) and patient's age ($p=0.022$).

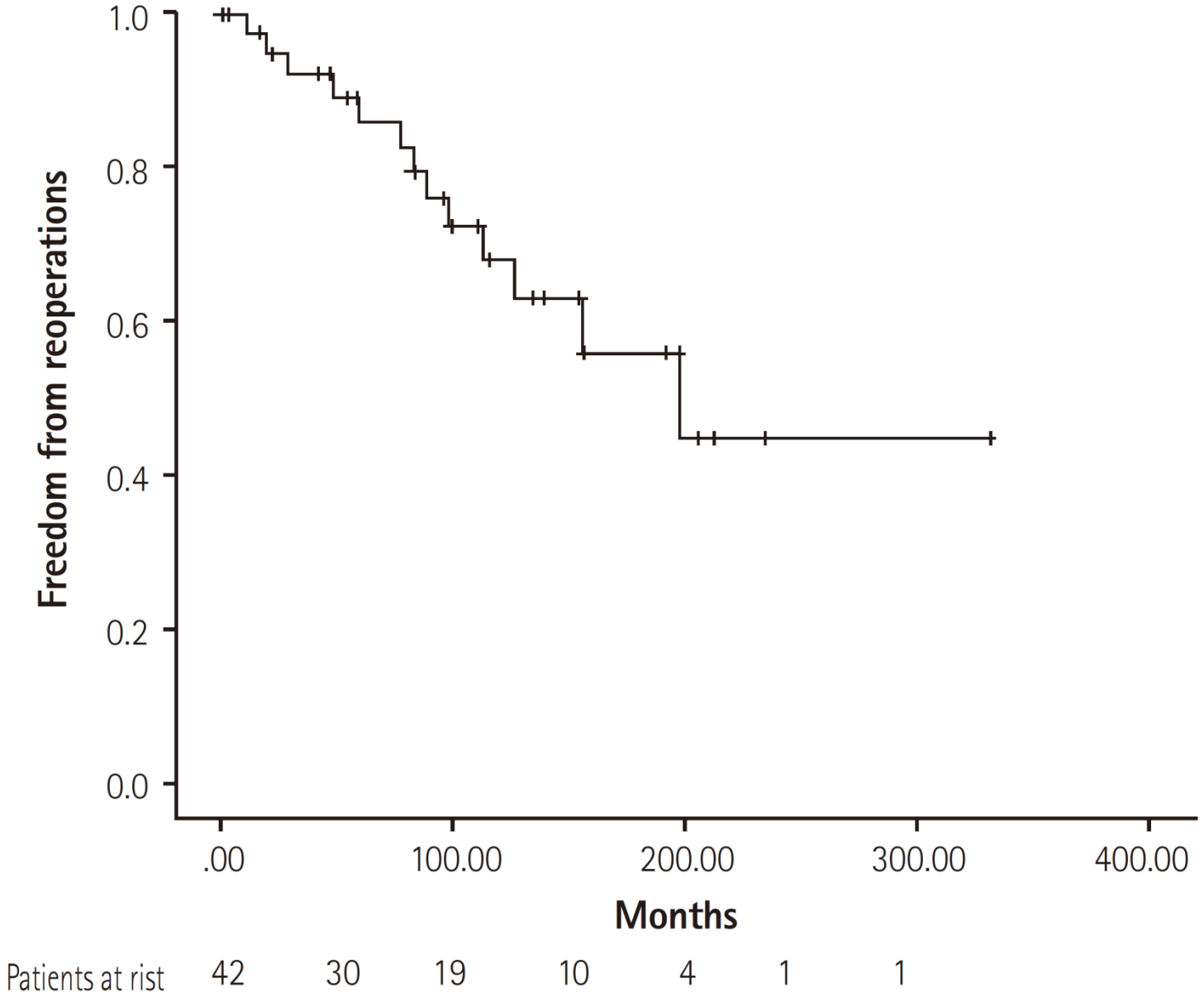
Results: There were 2 in-hospital mortalities (4.2%; 1 neonate and 1 infant) and 2 late mortalities during follow-up. Freedom from recurrent TV regurgitation rates at 5, 10, and 15 years were 88.6%, 66.3%, 52.7%, respectively. TV regurgitation recurrence did not differ according to surgical method ($p=0.800$). Survival rates at 5, 10, and 20 years were 95.8%, 95.8%, and 85.6%, respectively, and freedom from reoperation rates at 5, 10, and 15 years were 85.9%, 68.0%, and 55.8%, respectively.

Conclusion: Surgical treatment strategies were decided according to Carpentier's type and patient's age. Overall survival and freedom from reoperation rates at 10 years were 95.8% and 68.0%, respectively. Approximately 25% of patients required a second operation for TV during the follow-up. **(Korean Circ J 2016;46(5):706-713)**

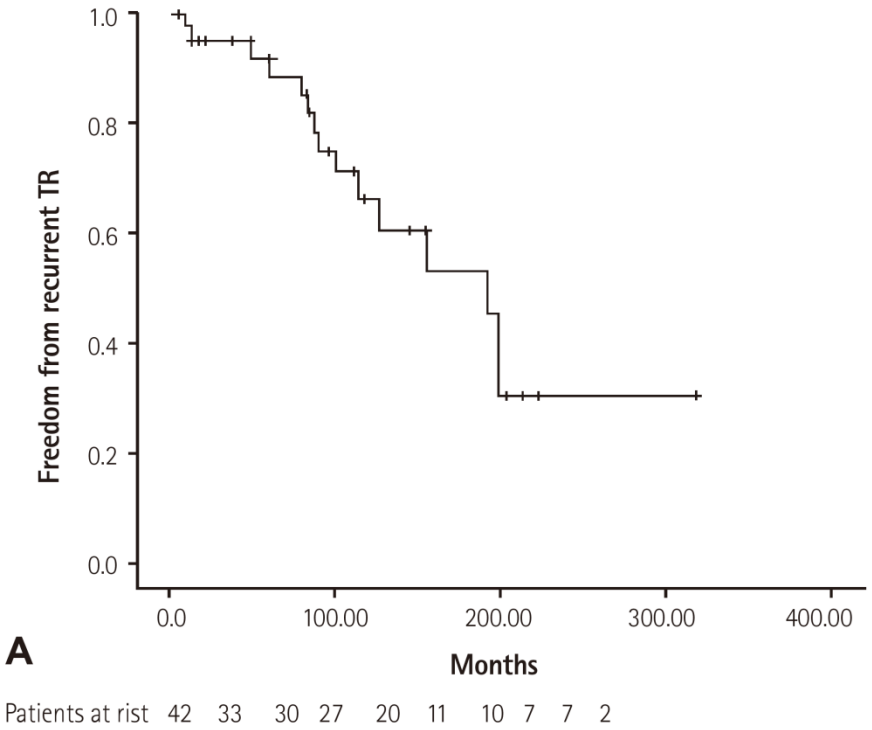
Survival


1.0
0.8
0.6
0.4
0.2
0.0
Survival
.00
100.00
200.00
300.00
400.00
500.00
Months
Patients at rist 48 37 28 20 13 6 4 2

Reoperation

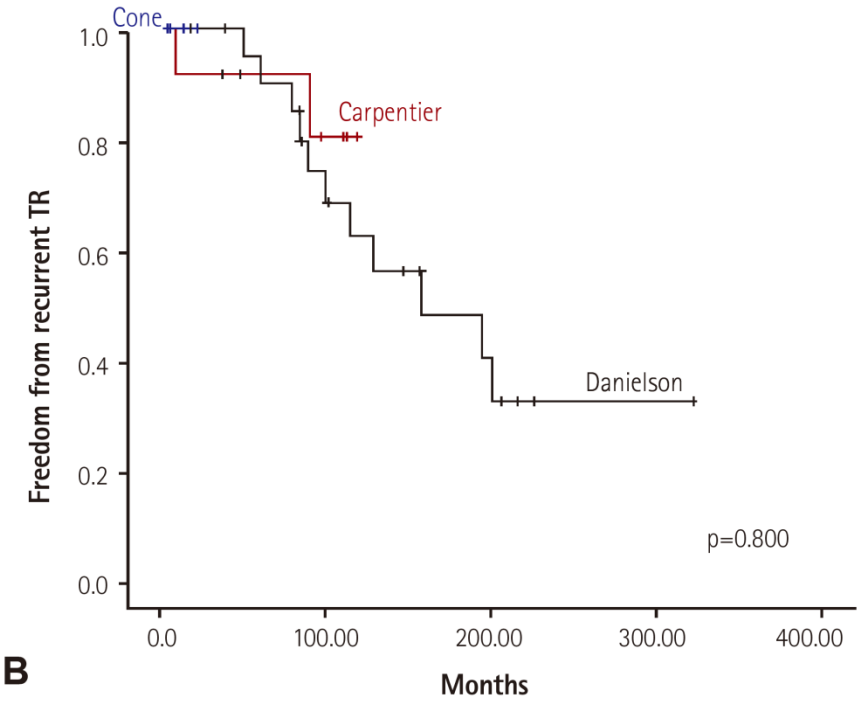


Recurrent TR



A

Patients at rist 42 33 30 27 20 11 10 7 7 2



B

TR after ACHD Surgery (JTCS 2016, Columbia)

Postoperative tricuspid regurgitation after adult congenital heart surgery is associated with adverse clinical outcomes

Matthew J. Lewis, MD, MPH,[a] Jonathan N. Ginns, MD,[a,b] Siqin Ye, MD,[c] Paul Chai, MD,[d] Jan M. Quaegebeur, MD,[d] Emile Bacha, MD,[d] and Marlon S. Rosenbaum, MD[a]

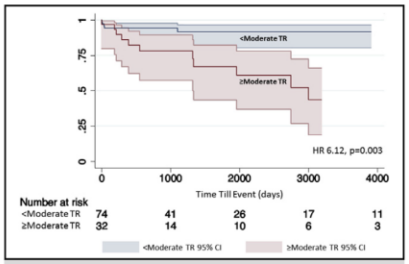
ABSTRACT

Objective: Many patients with adult congenital heart disease will require cardiac surgery during their lifetime, and some will have concomitant tricuspid regurgitation. However, the optimal management of significant tricuspid regurgitation at the time of cardiac surgery remains unclear. We assessed the determinants of adverse outcomes in patients with adult congenital heart disease and moderate or greater tricuspid regurgitation undergoing cardiac surgery for non–tricuspid regurgitation-related indications.

Methods: All adult patients with congenital heart disease and greater than moderate tricuspid regurgitation who underwent cardiac surgery for non–tricuspid regurgitation-related indications were included in a retrospective study at the Schneeweiss Adult Congenital Heart Center. Cohorts were defined by the type of tricuspid valve intervention at the time of surgery. The primary end point of interest was a composite of death, heart transplantation, and reoperation on the tricuspid valve.

Results: A total of 107 patients met inclusion criteria, and 17 patients (17%) reached the primary end point. A total of 68 patients (64%) underwent tricuspid valve repair, 8 patients (7%) underwent tricuspid valve replacement, and 31 patients (29%) did not have a tricuspid valve intervention. By multivariate analysis, moderate or greater postoperative tricuspid regurgitation was associated with a hazard ratio of 6.12 (1.84-20.3) for the primary end point ($P = .003$). In addition, failure to perform a tricuspid valve intervention at the time of surgery was associated with an odds ratio of 4.17 (1.26-14.3) for moderate or greater postoperative tricuspid regurgitation ($P = .02$).

Conclusions: Moderate or greater postoperative tricuspid regurgitation was associated with an increased risk of death, transplant, or reoperation in adult patients with congenital heart disease undergoing cardiac surgery for non–tricuspid regurgitation-related indications. Concomitant tricuspid valve intervention at the time of cardiac surgery should be considered in patients with adult congenital heart disease with moderate or greater preoperative tricuspid regurgitation. (J Thorac Cardiovasc Surg 2016;151:460-5)



Survivor function for the primary end point by degree of postoperative TR.

Central Message

TV intervention at the time of cardiac surgery should be considered in adults with CHD with moderate or greater preoperative TR.

Perspective

Approximately half of all patients with congenital disease will require repeat cardiac surgery in moderate or greater preoperative TR undergoing cardiac surgery for non–TR-related indications. TV intervention at the time of surgery may decrease the probability of death, TV reoperation, or heart transplant.

See Editorial Commentary page 466.

Outcomes of Repair for Corrected TGA (ATS 2010, Korea)

Outcomes of Biventricular Repair for Congenitally Corrected Transposition of the Great Arteries

Hong-Gook Lim, MD, PhD, Jeong Ryul Lee, MD, PhD, Yong Jin Kim, MD, PhD, Young-Hwan Park, MD, PhD, Tae-Gook Jun, MD, PhD, Woong-Han Kim, MD, PhD, Chang-Ha Lee, MD, PhD, Han Ki Park, MD, PhD, Ji-Hyuk Yang, MD, Chun-Soo Park, MD, and Jae Gun Kwak, MD

Department of Thoracic and Cardiovascular Surgery, Seoul National University College of Medicine, Seoul National University Children's Hospital, Xenotransplantation Research Center, Seoul National University Hospital, Transplantation Research Institute, Medical Research Center, Seoul National University, Department of Thoracic and Cardiovascular Surgery, Yonsei University College of Medicine, and Department of Thoracic and Cardiovascular Surgery, Samsung Medical Center, Sungkyunkwan University School of Medicine, Seoul, and Department of Thoracic and Cardiovascular Surgery, Sejong General Hospital, Sejong Heart Institute, Bucheon, Korea

Background. This study was undertaken to evaluate long-term results of biventricular repairs for congenitally corrected transposition of the great arteries, and to analyze the risk factors that affect mortality and morbidity.

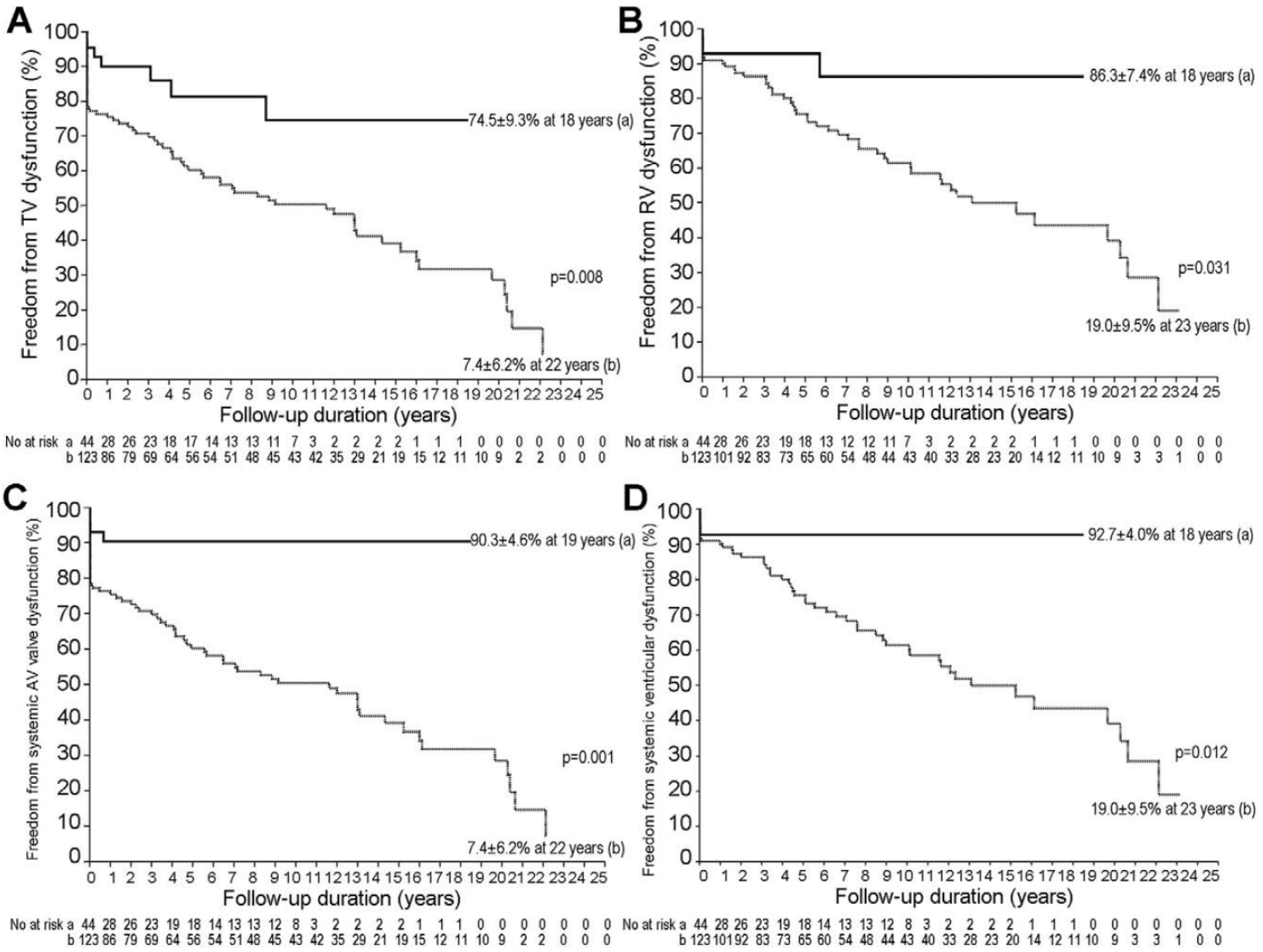
Methods. Between 1983 and 2009, 167 patients with congenitally corrected transposition of the great arteries underwent biventricular repairs. The physiologic repairs were performed in 123 patients, and anatomic repairs in 44. Average follow-up was 9.3 ± 6.6 years.

Results. Kaplan-Meier estimated survival was 83.3% ± 0.5% at 25 years in biventricular repair. In anatomic repair, left ventricular training and right ventricular dysfunction had negative impact on survival, but bidirectional cavopulmonary shunt had positive impact on survival. The reoperation-free ratio was 10.1% ± 7.8% at 22 years after physiologic repair, and 46.2% ± 12.4% at 15 years after anatomic repair ($p = 0.885$). Freedom from any arrhythmia was 49.6% ± 7.5% at 22 years after physiologic repair, and 60.8% ± 14.8% at 18 years after anatomic repair ($p = 0.458$). Freedom from systemic atrioventricular valve and ventricular dysfunction as well as tricuspid valve and right ventricular dysfunction was significantly higher in anatomic repair than in physiologic repair.

Conclusions. Long-term results of biventricular repair were satisfactory. Patients presenting with right ventricular dysfunction or need for left ventricular training represent a high-risk group of anatomic repair for which selection criteria are particularly important. Late functional outcomes of anatomic repair were excellent compared with physiologic repair. Anatomic repair is the procedure of choice for those patients if both ventricles are adequate or if surgical technique is modified with the help of additional a bidirectional cavopulmonary shunt.

(Ann Thorac Surg 2010;89:159–67)

Outcomes of Repair for Corrected TGA (ATS 2010, Korea)



TR in Corrected TGA (ATS 2015, Boston)

Tricuspid Valve Regurgitation in Congenitally Corrected Transposition of the Great Arteries and a Left Ventricle to Pulmonary Artery Conduit

Jonathan Buber, MD, Doff B. McElhinney, MD, Anne Marie Valente, MD, Audrey C. Marshall, MD, and Michael J. Landzberg, MD

Department of Cardiology, Boston Children's Hospital, Boston, Massachusetts; Department of Cardiology, Nationwide Children's Hospital, Ohio state University, Columbus, Ohio; and Lucile Packard Children's Hospital, Stanford University, Palo Alto, California

Background. The configuration of the interventricular septum can affect the function of the tricuspid valve in patients with congenitally corrected transposition of the great arteries who have a systemically functioning right ventricle. Altering septal configuration by addressing a dysfunctional conduit placed between the left ventricle (LV) and the pulmonary artery (PA) in these patients can impact septal configuration and competency of the tricuspid valve.

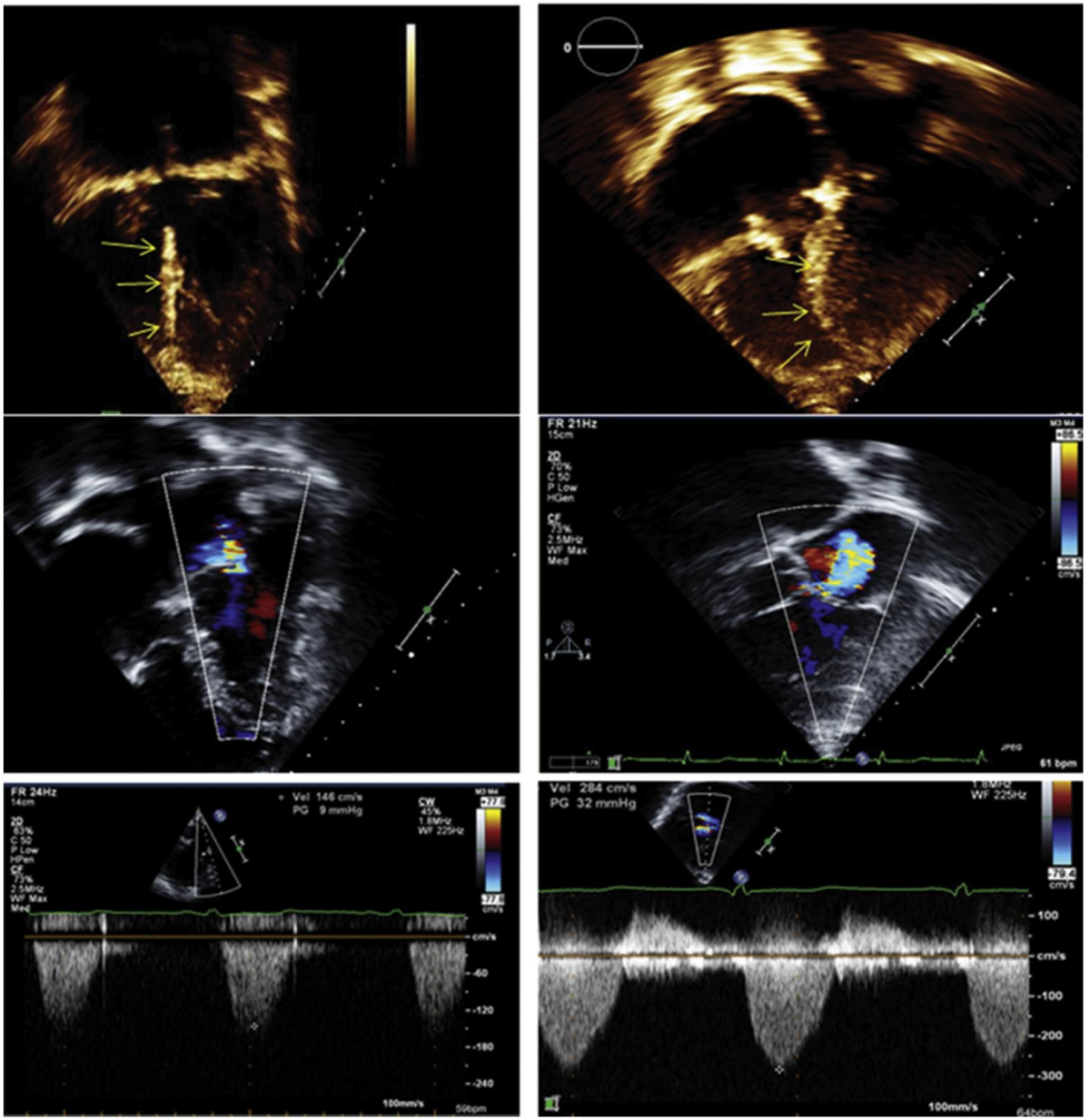
Methods. In 38 patients with an LV to PA conduit, we evaluated relationships between conduit function, RV geometry, and tricuspid valve function, and compared these variables before and after conduit intervention.

Results. Median age at conduit implant was 4.5 years (0.5 to 36) and median total follow-up was 12 years (2 to 22). Of the 38 patients, 23 (60%) underwent conduit intervention, a median of 7.5 years after implant. In 15 of these patients (65%) the degree of tricuspid regurgitation (TR) worsened, compared with only 2 patients (15%) in the non-intervention group ($p < 0.001$). Worsening TR was associated with the degree of change in RV and LV ventricular diameters, change in tricuspid annulus size and tethering distance, and the degree of septal shift, as reflected by the right ventricular sphericity index (all $p \leq 0.04$). In 8 of 15 patients with more severe TR at follow-up, there was also progressive RV dysfunction.

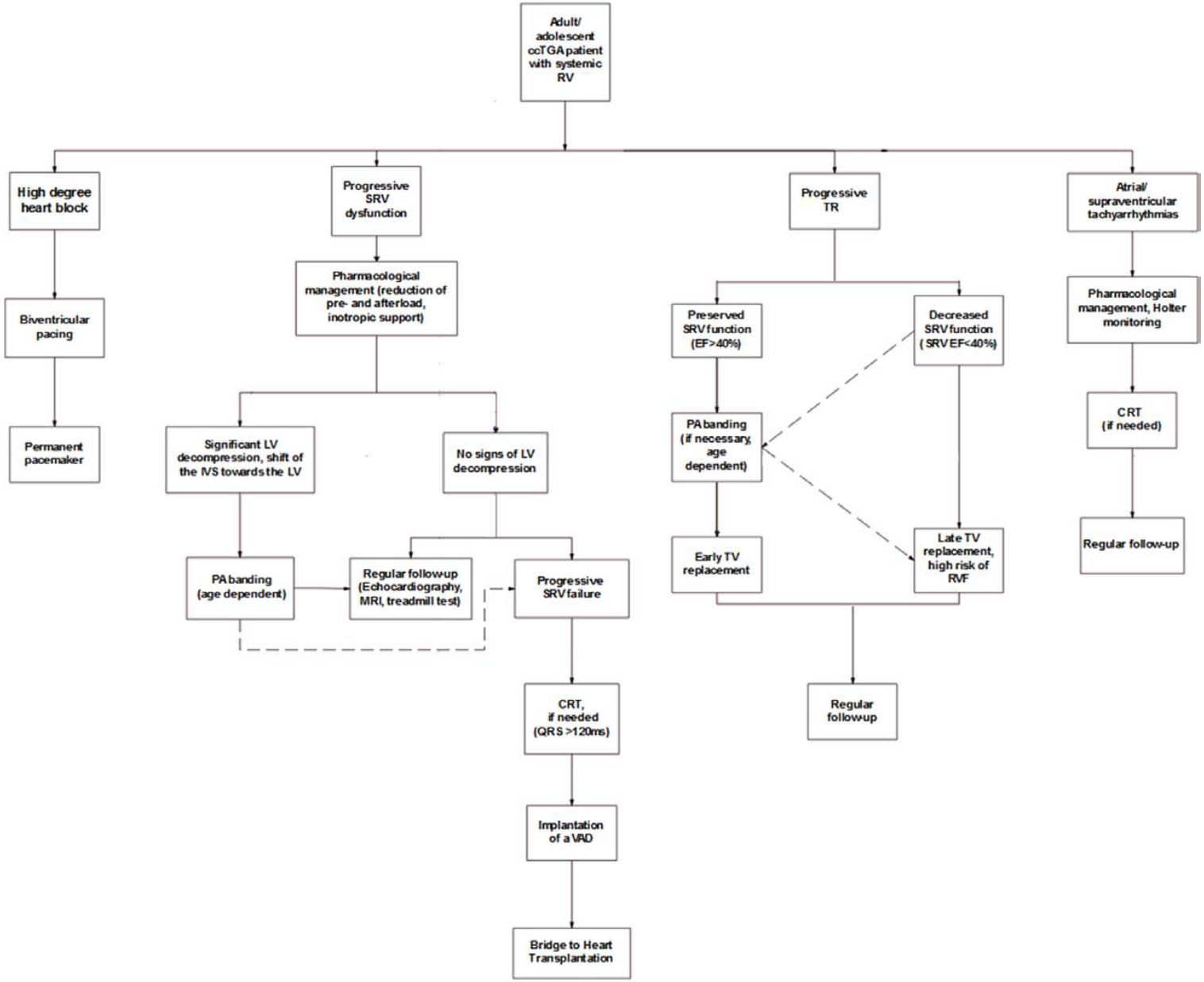
Conclusions. Intervention for LV to PA conduit dysfunction may result in worsening TR and right ventricular function, likely due in part to altered septal shift due to changes in the interventricular pressure ratio. Management of LV to PA conduit dysfunction should take these findings into account.

(Ann Thorac Surg 2015;99:1348–56)

TR in Corrected TGA (ATS 2015, Boston)



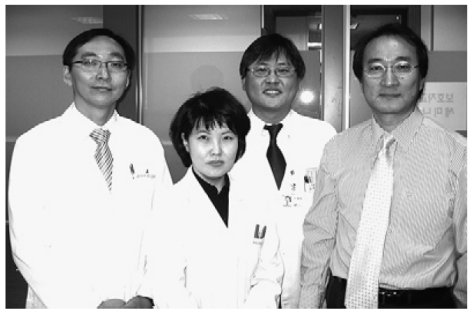
Mx of Adults with Systemic RV (Circulation 2016, Boston)



Outcome of 200 Extracardiac Fontan (JTCS 2008)

Outcome of 200 patients after an extracardiac Fontan procedure

Soo-Jin Kim, MD, PhD,[a] Woong-Han Kim, MD, PhD,[c] Hong-Gook Lim, MD,[b] and Jae-Young Lee, MD[a]



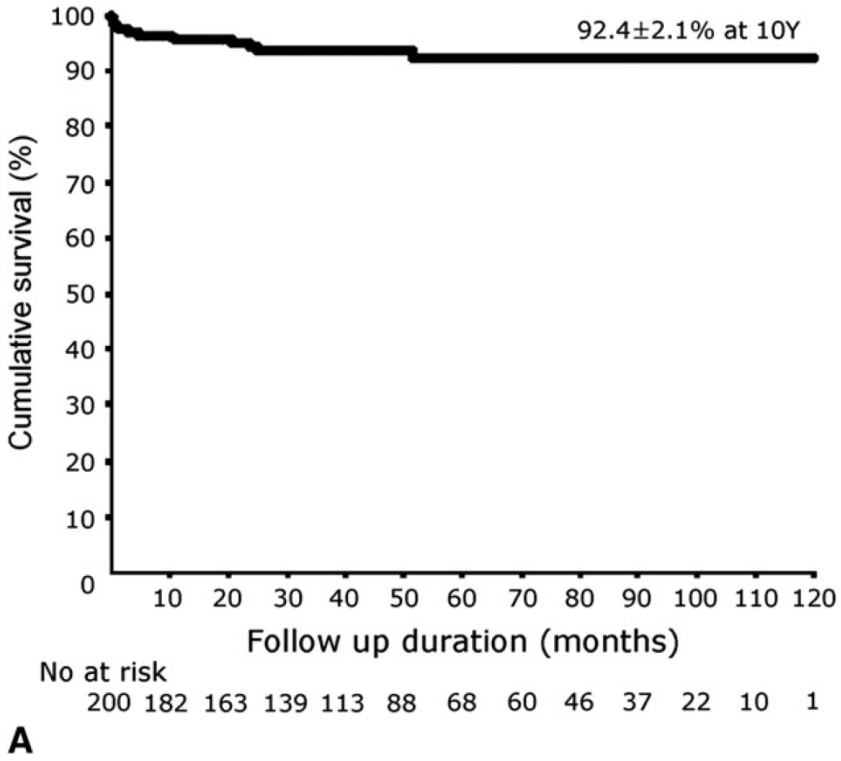
Lim, Kim SJ, Lee, Kim WH (left to right)

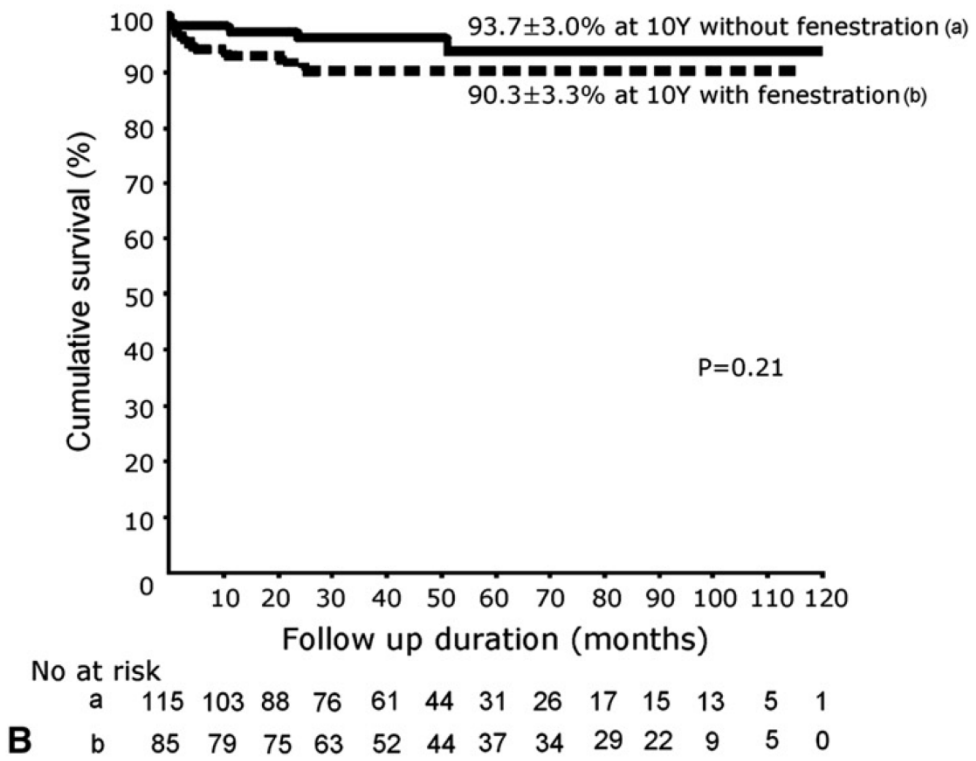
Objectives: Despite the known advantages of the extracardiac conduit Fontan procedure, the long-term outcomes related to the longevity of the conduit and anticoagulation therapy have not been determined. The purpose of this study was to evaluate the outcome of hospital survivors with an extracardiac Fontan circulation.

Methods and Results: Between 1996 and 2006, 200 patients had the extracardiac conduit Fontan operation. The median age at operation was 3.4 years. Most patients (89.5%) underwent a bidirectional cavopulmonary shunt. Fenestration was required in 85 patients. Overall, the 10-year survival was 92.4% ± 2.1%. Multivariate analysis identified severe infection during the early postoperative period and a high pulmonary arterial pressure during the preoperative period as independent risk factors for patient mortality. The Kaplan–Meier estimate for freedom from reoperation was 82.4% ± 4.1% at 10 years. Arrhythmia occurred in 32 patients after the Fontan operation; freedom from arrhythmia was 85.1% ± 4.4% at 10 years. The risk factors for arrhythmia were the heterotaxy syndrome, follow-up duration, and age at Fontan operation. Freedom from thromboembolism at 10 years was 92.9% ± 1.9%. Among all of the patients, 95.2% were classified in New York Heart Association class I.

Conclusions: The results of this study showed that during 10 years of follow-up, the overall survival and the functional status of the survivors after an extracardiac Fontan procedure were satisfactory. We might infer that fenestration provided benefit inasmuch as the high-risk Fontan patients fenestrated had similar outcomes to those not fenestrated, who were presumably low risk. The incidence of late death, reoperation, obstruction of the cavopulmonary pathway, arrhythmias, and thromboembolism was low.

Outcome of 200 Extracardiac Fontan (JTCS 2008)





AV valve regurgitation after Fontan

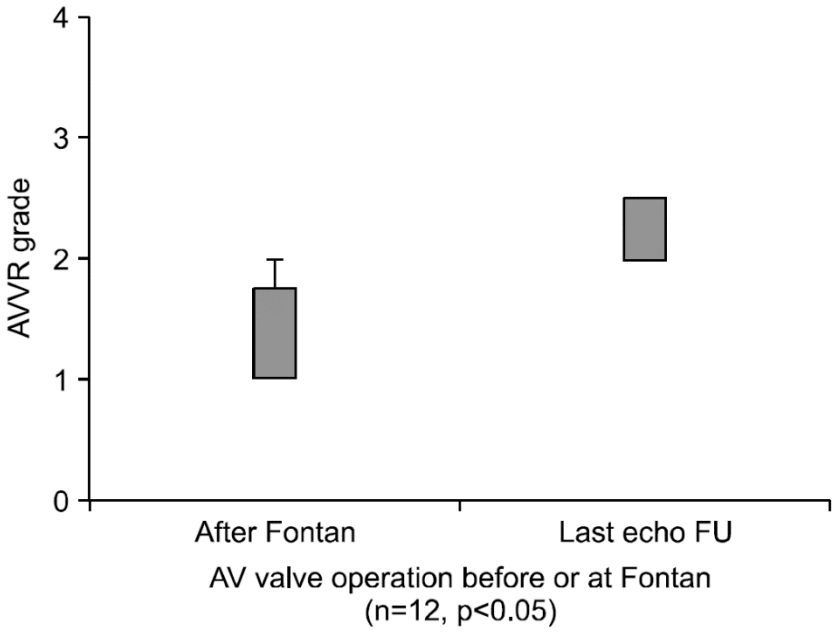
A Trend for Atrioventricular Valve Regurgitation after a Modified Fontan Operation

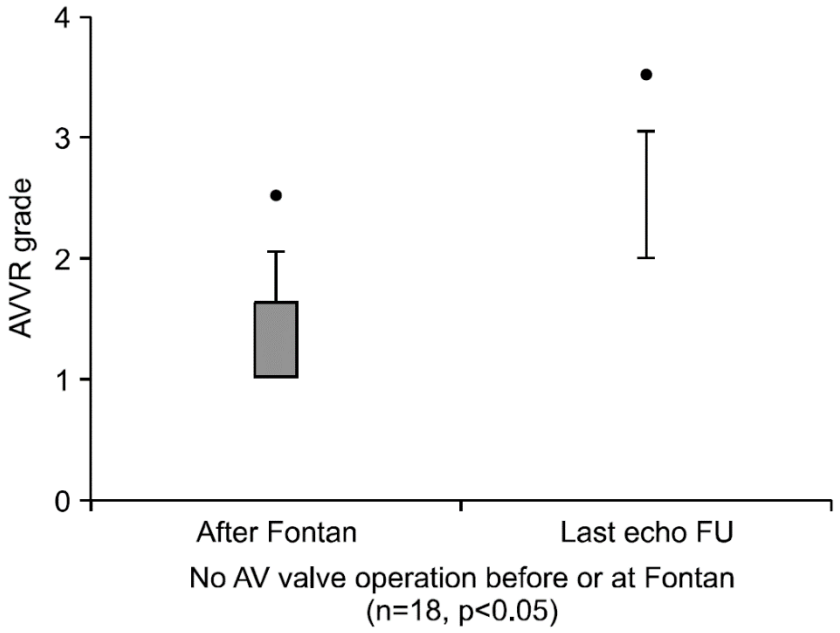
Hong-Gook Lim, M.D.*, Chang-Ha Lee, M.D.*, Hong Joo Seo, M.D.**, Woong-Han Kim, M.D.***,
Seong Wook Hwang, M.D.****, Cheul Lee, M.D.*

Background: Anatomic and functional abnormalities of the systemic atrioventricular (AV) valve are common in single ventricle pathologies and continue to be associated with poor early and late outcomes in surgically palliated single ventricle patients. We aggressively performed valvuloplasty for atrioventricular valve regurgitation (AVVR) during the course toward a Fontan operation. ***Material and Method:*** Between January 1995 and December 2004, 209 patients underwent a Fontan operation in our institution. We retrospectively evaluated the prevalence of AVVR and the influence of AV valve repair on outcome, and we analyzed the progression of AVVR after the Fontan operation for 168 patients where echocardiographic follow up results for more than 6 months after the Fontan operation were available. During the course toward a Fontan operation, 25 patients underwent 30 procedures for AVVR. These procedures were carried out during placement of a bidirectional cavopulmonary shunt (BCPS) for nine patients, between the time of placement of a BCPS and the Fontan operation for four patients, and during the Fontan operation for 17 patients. Five patients underwent procedures for AVVR twice. ***Result:*** The late mortality rate after the Fontan operation was 4.2% (n=7), with a median follow-up duration of 52 months (range, 6 ~123 months). Seven patients (4%) had unfavorable outcomes such as significant (moderate or severe) AVVR in six patients, and significant AV valve stenosis in one patient was determined at the last follow up after the Fontan operation. Among the seven patients, four patients underwent AV valve repair after the Fontan operation, and one patient underwent subsequent AV valve replacement. Progression to AVVR of equal to or greater than grade 2 was noted in 30 patients (18%) at the last follow up after the Fontan operation, including 12 patients that underwent previous AV valve procedures. Initial grading of AVVR, a previous AV valve operation, and specific AV valve morphology such as a common AV valve or mitral atresia were significant risk factors for the progression of AVVR after the Fontan operation. ***Conclusion:*** In our surgical series, a small percentage of patients showed unfavorable outcomes related to AVVR during the course toward a Fontan operation. However, a closer follow-up is required to evaluate the progression of the AVVR after a Fontan operation, especially for patients showing poor AV valve function at the first presentation and specific AV valve morphology.

(Korean J Thorac Cardiovasc Surg 2008;41:305-312)

AV valve regurgitation after Fontan





Systemic ventricular outflow tract obstruction

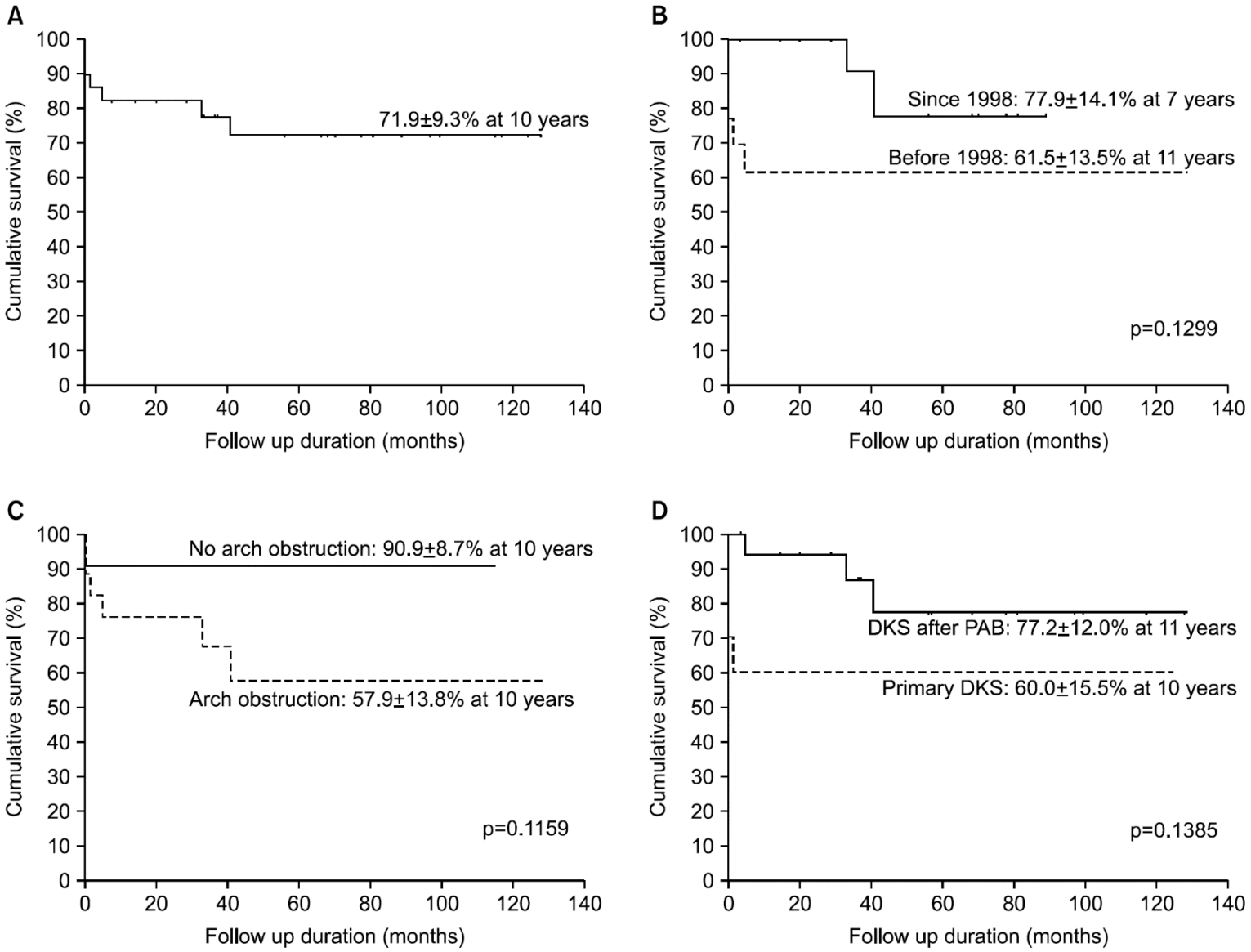
The Clinical Application and Results of Palliative Damus-Kaye-Stansel Procedure

Hong-Gook Lim, M.D.*, Chang-Ha Lee, M.D.*, Soo-Jin Kim, M.D.**, Woong-Han Kim, M.D.***,
Seong Wook Hwang, M.D.*, Cheul Lee, M.D.*, Sung-Ho Shinn, M.D.*, Kil-Soo Yie, M.D.*, Jae Woong Lee, M.D.****

Background: The Damus-Kaye-Stansel (DKS) procedure is a proximal MPA-ascending aorta anastomosis used to relieve systemic ventricular outflow tract obstructions (SVOTO) and pulmonary hypertension. The purpose of this study was to review the indications and outcomes of the DKS procedure, including the DKS pathway and semilunar valve function. ***Material and Method:*** A retrospective review of 28 patients who underwent a DKS procedure between May 1994 and April 2006 was performed. The median age at operation was 5.3 months (13 days～38.1 months) and body weight was 5.0 kg (2.9～13.5 kg). Preoperative pressure gradients were 25.3±15.7 mmHg (10～60 mmHg). Eighteen patients underwent a preliminary pulmonary artery banding as an initial palliation. Preoperative main diagnoses were double outlet right ventricle in 9 patients, double inlet left ventricle with ventriculoarterial discordance in 6, another functional univentricular heart in 5, Criss-cross heart in 4, complete atrioventricular septal defect in 3, and hypoplastic left heart variant in 1. DKS techniques included end-to-side anastomosis with patch augmentation in 14 patients, classical end-to-side anastomosis in 6, Lamberti method (double-barrel) in 3, and others in 5. The bidirectional cavopulmonary shunt and Fontan procedure were concomitantly performed in 6 and 2 patients, respectively. ***Result:*** There were 4 hospital deaths (14.3%), and 3 late deaths (12.5%) with a follow-up duration of 62.7±38.9 months (3.3～128.1 months). Kaplan-Meier estimated actuarial survival was 71.9%±9.3% at 10 years. Multivariate analysis showed right ventricle type single ventricle (hazard ratio=13.960, p=0.004) and the DKS procedure as initial operation (hazard ratio=6.767, p=0.042) as significant mortality risk factors. Four patients underwent staged biventricular repair and 13 received Fontan completion. No SVOTO was detected after the procedure by either cardiac catheterization or echocardiography except in one patient. There was no semiulnar valve regurgitation (＞Gr II) or semilunar valve-related reoperation, but one patient (3.6%) who underwent classical end-to-side anastomosis needed reoperation for pulmonary artery stenosis caused by compression of the enlarged DKS pathway. The freedom from reoperation for the DKS pathway and semilunar valve was 87.5% at 10 years after operation. ***Conclusion:*** The DKS procedure can improve the management of SVOTO, and facilitate the selected patients who are high risk for biventricular repair just after birth to undergo successful staged biventricular repair. Preliminary pulmonary artery banding is a safe and effective procedure that improves the likelihood of successful DKS by decreasing pulmonary vascular resistance. The long-term outcome of the DKS procedure for semilunar valve function, DKS pathway, and relief of SVOTO is satisfactory.

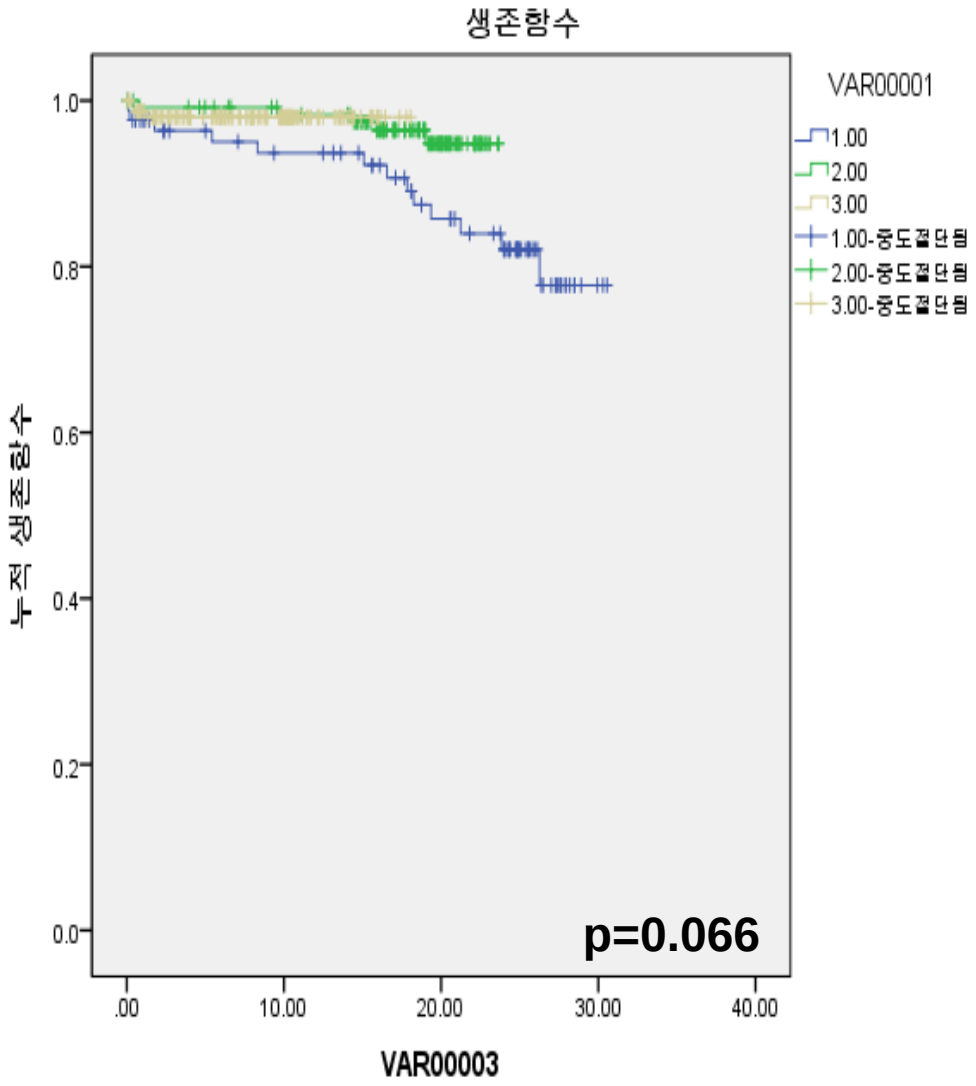
(Korean J Thorac Cardiovasc Surg 2008;41:1-11)

Systemic ventricular outflow tract obstruction

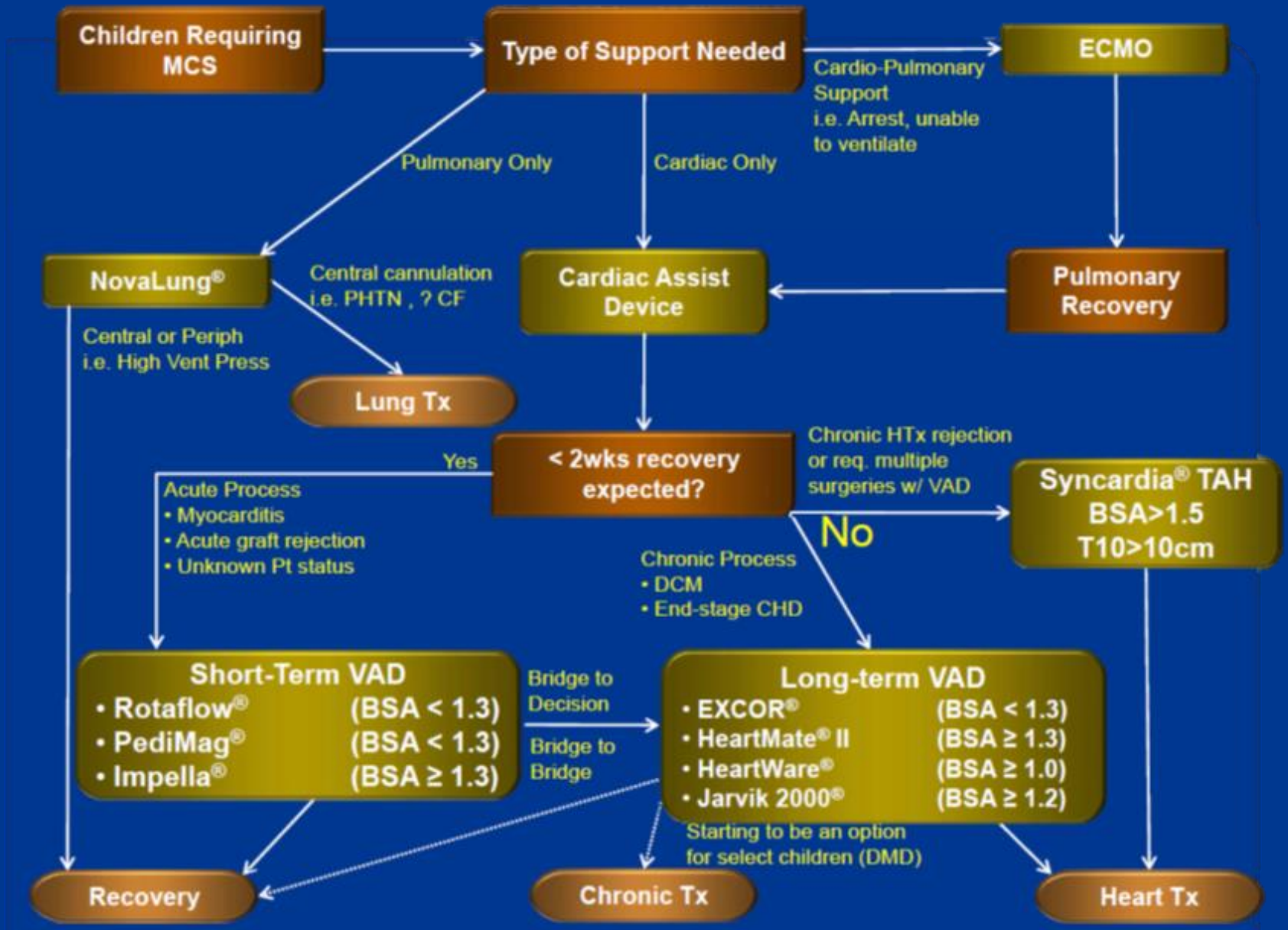


Long-Term Results of Fontan in SNUH (2015)

Overall survival rate



	5 year	10 year	15 year
AP	96.4 %	93.7 %	92.2 %
LT	99.2 %	98.3 %	97.4 %
EC	98.0 %	98.0 %	98.0 %


Children Requiring MCS
Type of Support Needed
ECMO
Cardio-Pulmonary Support
i.e. Arrest, unable to ventilate
Pulmonary Only
Cardiac Only
NovaLung®
Central cannulation
i.e. PHTN , ? CF
Cardiac Assist Device
Pulmonary Recovery
Central or Periph
i.e. High Vent Press
Lung Tx
Yes
< 2wks recovery expected?
Chronic HTx rejection or req. multiple surgeries w/ VAD
Syncardia® TAH
BSA>1.5
T10>10cm
No
Acute Process
• Myocarditis
• Acute graft rejection
• Unknown Pt status
Chronic Process
• DCM
• End-stage CHD
Short-Term VAD
• Rotaflow® (BSA < 1.3)
• PediMag® (BSA < 1.3)
• Impella® (BSA ≥ 1.3)
Bridge to Decision
Bridge to Bridge
Long-term VAD
• EXCOR® (BSA < 1.3)
• HeartMate® II (BSA ≥ 1.3)
• HeartWare® (BSA ≥ 1.0)
• Jarvik 2000® (BSA ≥ 1.2)
Starting to be an option for select children (DMD)
Recovery
Chronic Tx
Heart Tx

ECMO in ACHD (CHD 2016, Mayo)

Postcardiotomy ECMO Support after High-risk Operations in Adult Congenital Heart Disease

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ABSTRACT

Background. Cardiac operations in high-risk adult congenital heart disease (ACHD) patients may require mechanical circulatory support (MCS), such as extracorporeal membrane oxygenation (ECMO) or intraaortic balloon pump (IABP), to allow the cardiopulmonary system to recover.

Methods. We reviewed records for all ACHD patients who required MCS following cardiotomy at our institution from 1/2001 to 12/2013.

Results. During the study period, 2264 (mean age 39.1 years, females ~54.1%) operations were performed in ACHD patients of whom 24 (1.1%) required postoperative MCS (14 males; median age 41 years, range 22–75). Preoperatively the 24 patients had a mean systemic ventricular ejection fraction of 47% (range 10–66%); 72% of these patients were in NYHA class III/IV heart failure. The common underlying diagnoses included pulmonary atresia with intact ventricular septum (20%), tetralogy of Fallot (16%), Ebstein anomaly (12%), cc-TGA (12%), septal defects (12%), and others (28%). Operations performed were valvular operations with/without maze (58.2%), Fontan conversion (21%), coronary bypass grafting with valvular operations (12.5%), and heart transplant (8.3%). Indications for MCS were left-sided (systemic) heart failure (32%), right-sided (subpulmonary) heart failure (24%), biventricular heart failure (36%), persistent arrhythmia (4%), and hypoxemia (4%). Forty-two percent were placed on ECMO only; in the second group, IABP was attempted and subsequently followed by ECMO initiation. The mean duration of MCS was 8.4 days (range 0.8–35.4). Common morbidities included coagulopathy (60%), renal failure (56%), and arrhythmia (48%). Overall, 46% of patients survived to hospital discharge. Deaths were due to either multi organ failure or the underlying cardiac disease; sepsis was the primary cause of death in one patient. Median follow-up for survivors was 41 months (maximum 106 months). NYHA functional class was I/II in all 8 late survivors.

Conclusions. Following complex operations in high-risk ACHD patients, MCS may be required. Despite significant morbidity, nearly half of patients survive to hospital discharge.

Outcomes for MCS in ACHD (JHLT 2017)

ELSEVIER

The Journal of Heart and Lung Transplantation
http://www.jhltonline.org

ORIGINAL CLINICAL SCIENCE

Outcomes following implantation of mechanical circulatory support in adults with congenital heart disease: An analysis of the Interagency Registry for Mechanically Assisted Circulatory Support (INTERMACS)

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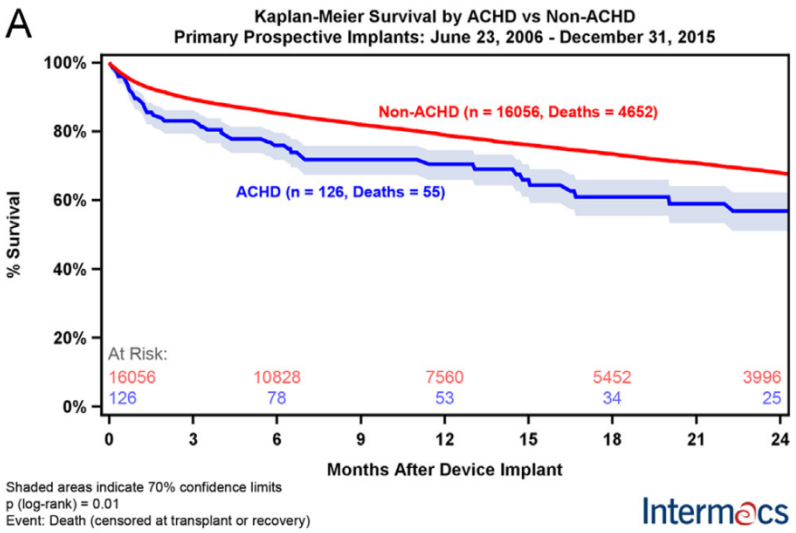
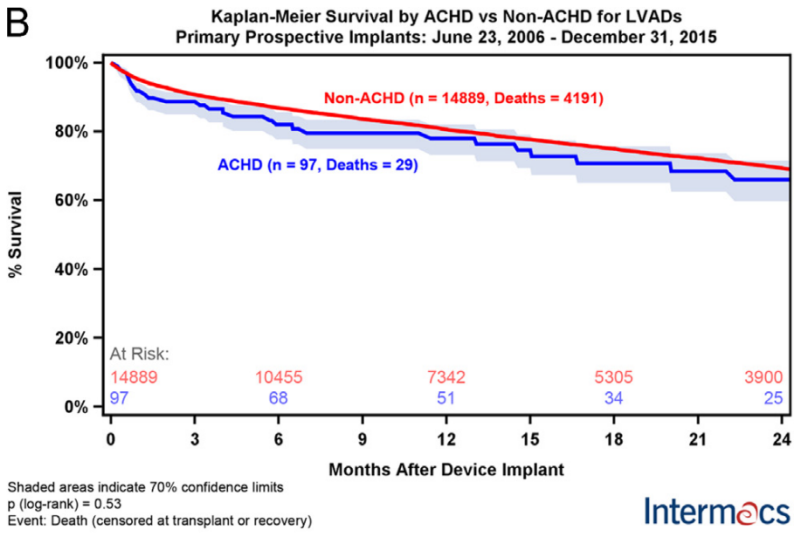
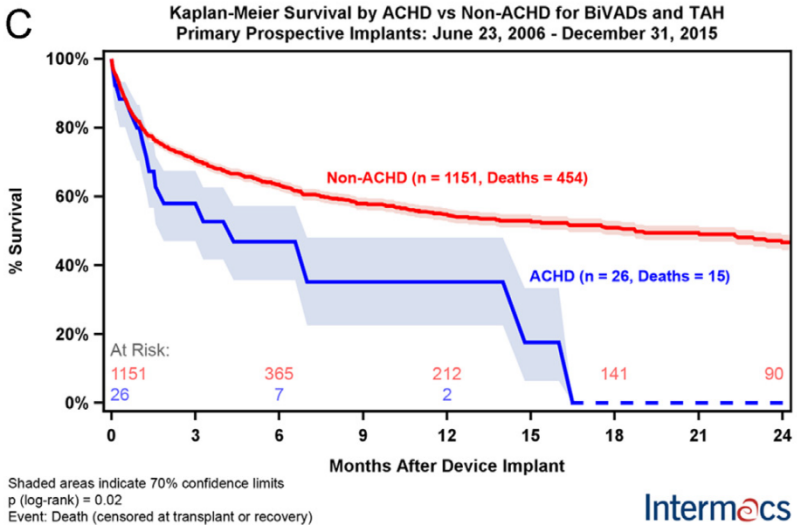
From the [a]Department of Cardiology, Boston Children's Hospital, Boston, Massachusetts; [b]Department of Cardiology, Baylor University Medical Center, Dallas, Texas; [c]Department of Cardiothoracic Surgery, Washington University School of Medicine, St. Louis, Missouri; [d]Department of Anesthesiology, Randall Children's Hospital, Portland, Oregon; [e]Knight Cardiovascular Institute, Oregon Health and Science University, Portland, Oregon; [f]Department of Cardiovascular Surgery, Mayo Clinic School of Medicine, Rochester, Minnesota; [g]Department of Cardiothoracic Surgery, University of Alabama at Birmingham, Birmingham, Alabama; and the [h]Department of Epidemiology, School of Public Health, University of Alabama at Birmingham, Birmingham, Alabama.

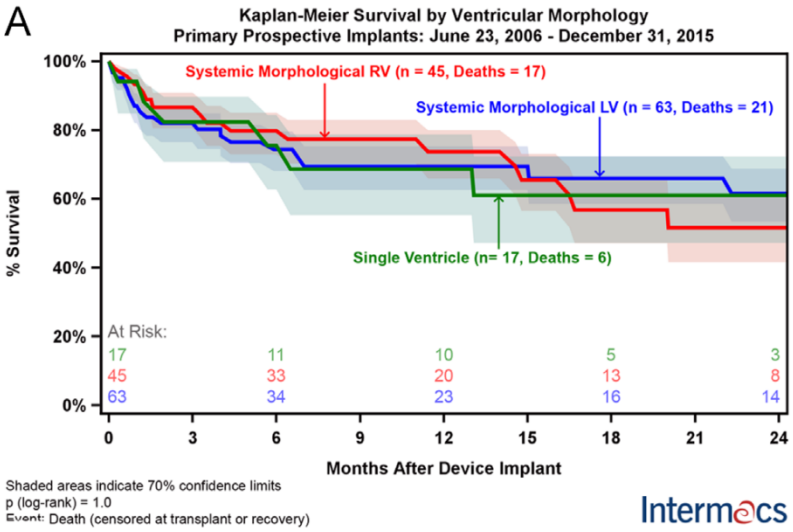
ACHD vs non-ACHD for Type of MCS (JHLT 2017)

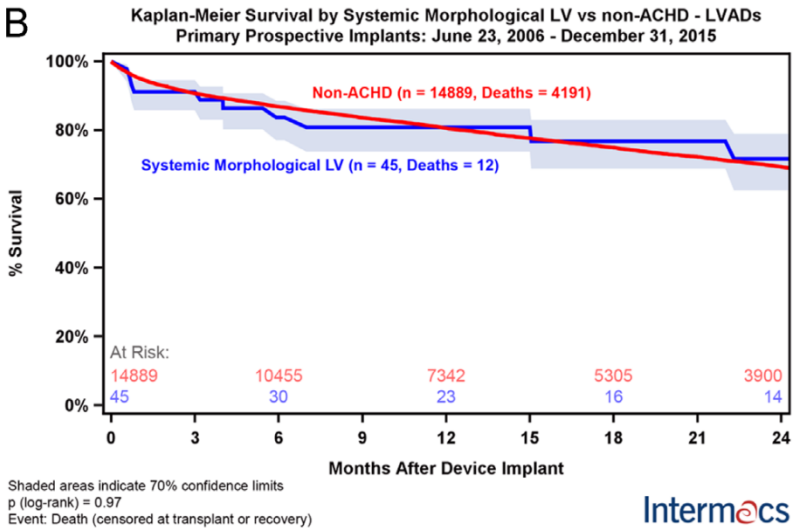


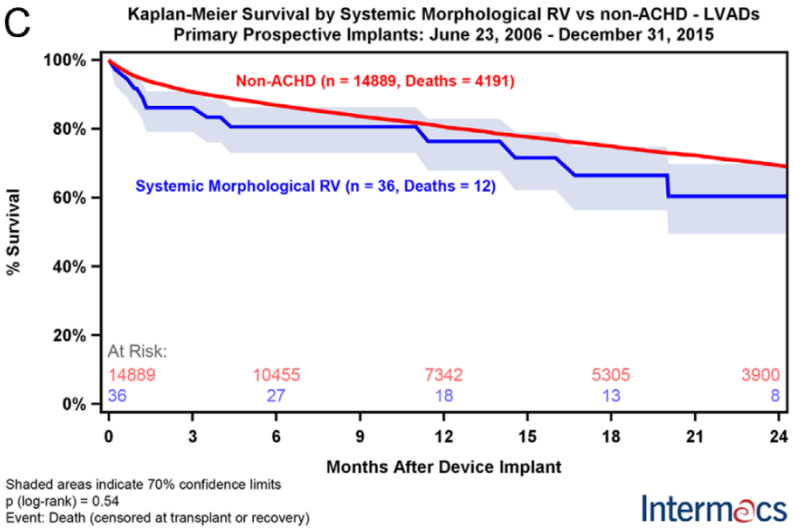


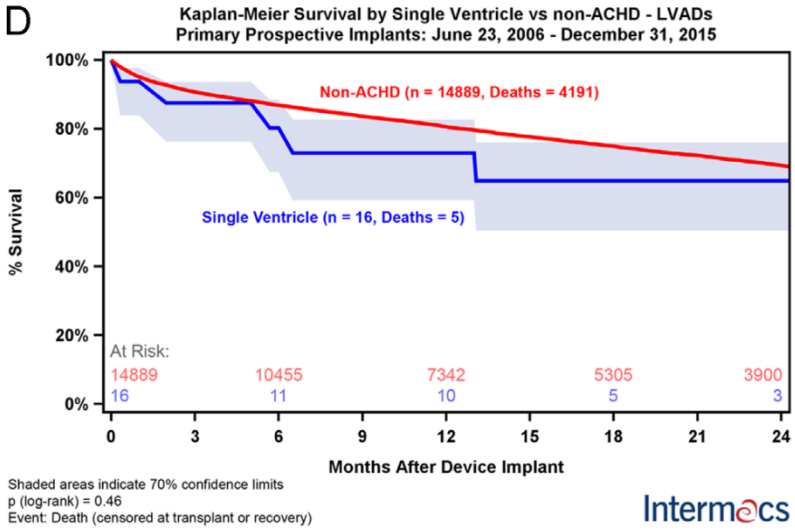


MCS in ACHD for Ventricular Morphology (JHLT 2017)

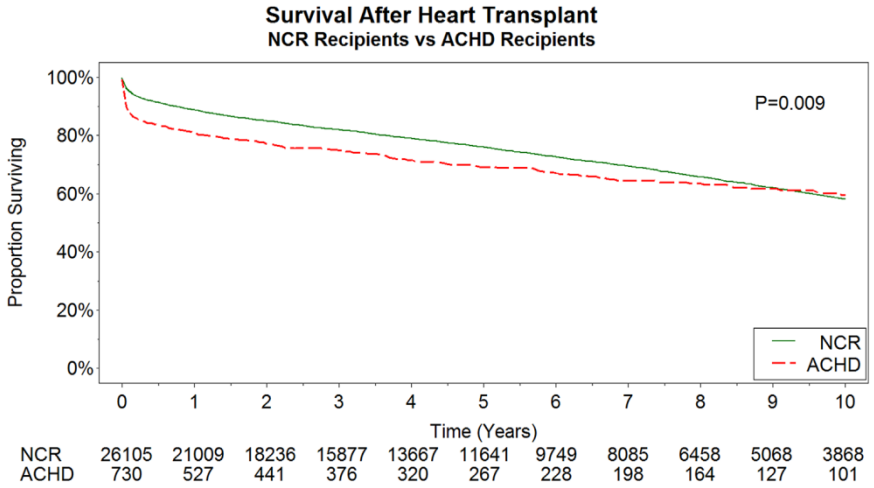


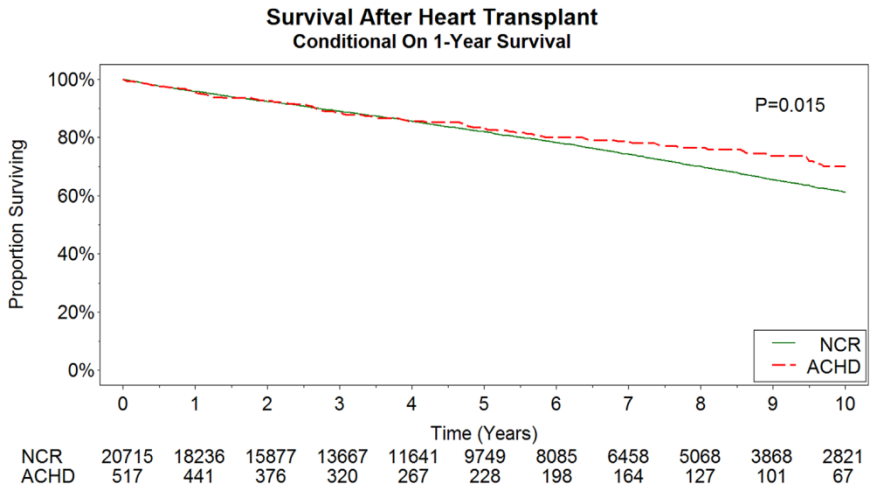


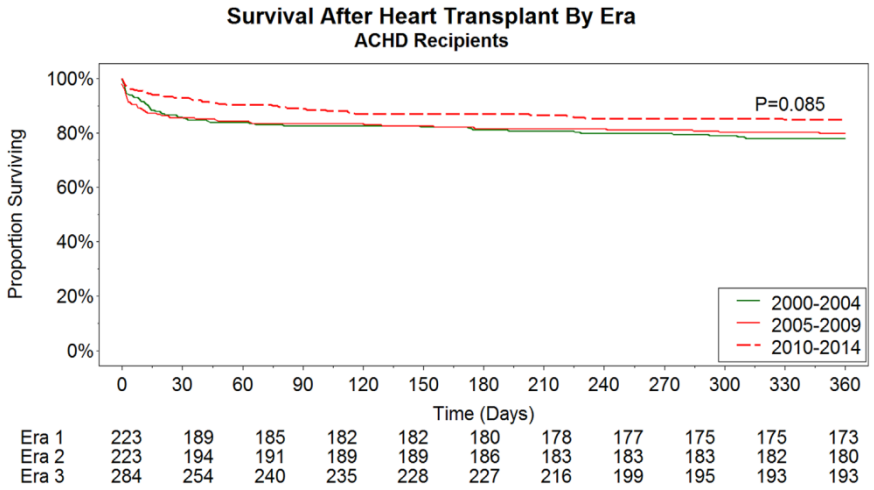


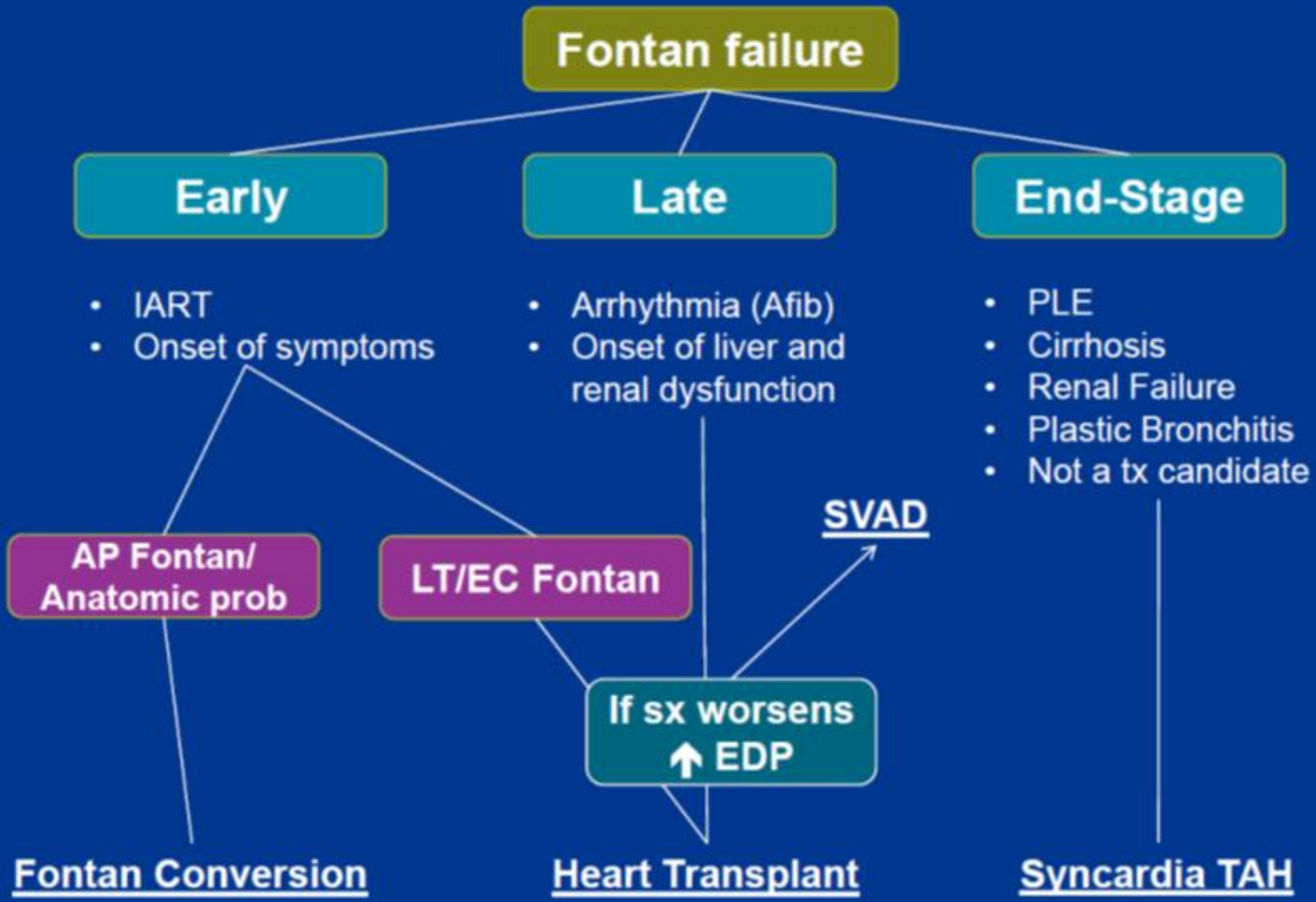


Heart Transplantation in ACHD (JCS 2016)








Fontan failure
Early
Late
End-Stage
• IART
• Onset of symptoms
• Arrhythmia (Afib)
• Onset of liver and renal dysfunction
• PLE
• Cirrhosis
• Renal Failure
• Plastic Bronchitis
• Not a tx candidate
SVAD
AP Fontan/ Anatomic prob
LT/EC Fontan
If sx worsens ↑ EDP
Fontan Conversion
Heart Transplant
Syncardia TAH

Fontan Conversion (EJCTS 2005, SNUH)

ELSEVIER

European Journal of Cardio-thoracic Surgery 27 (2005) 250–257

EUROPEAN JOURNAL OF CARDIO-THORACIC SURGERY

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Fontan conversion with arrhythmia surgery☆

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Abstract

Objective: Hemodynamic abnormalities and refractory atrial arrhythmias in patients late after the Fontan operation result in significant morbidity and mortality. We reviewed our experience with Fontan conversion and concomitant arrythmia surgery. **Methods**: Between January 1996 and February 2004, 16 patients underwent Fontan conversion and arrhythmia surgery. Mean age at the initial Fontan operation was 5.1±3.5 (range: 2–15) years and mean age at Fontan conversion was 17.0±5.8 (range: 6–30). The initial Fontan operations were atriopulmonary connections in 14 patients, extracardiac lateral tunnel in 1, and intracardiac lateral tunnel in 1. The types of arrhythmia included atrial flutter in 10 patients and atrial fibrillation in 3. Fontan conversion operation was performed with intracardiac lateral tunnel in 5 patients and extracardiac conduit in 11. Arrhythmia surgery included isthmus cryoablation in 10 patients and right-sided maze in 3. **Results**: There has been no mortality. At Fontan conversion operation, 7 patients required permanent pacemaker. All patients have improved to New York Heart Association class I or II. With a mean follow-up of 26.9±30.6 (range:1–87) months, 16 patients had sinus rhythm, 2 patients had transient atrial flutter which was well controlled, and 2 patients required permanent pacemaker during follow-up. **Conclusions**: Fontan conversion with concomitant arrhythmia surgery and permanent pacemaker placement is safe, improves New York Heart Association functional class, and has a low incidence of recurrent arrhythmias. In most patients, concomitant permanent pacemakers are needed.

Keywords: Fontan conversion; Arrhythmia surgery

Fontan Conversion (ATS 2016, Chicago)

Intermediate-Term Outcome of 140 Consecutive Fontan Conversions With Arrhythmia Operations

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Divisions of Cardiology and Cardiovascular-Thoracic Surgery, Departments of Pediatrics and Surgery, Ann & Robert H. Lurie Children's Hospital of Chicago, Northwestern University Feinberg School of Medicine, Chicago, Illinois; and Congenital Heart Institute, Florida Hospital for Children, Orlando, Florida

Background. Atrial arrhythmias and progressive circulatory failure frequently develop in patients with a Fontan circulation. Improvement of flow dynamics and revision of the arrhythmia substrate may improve outcomes in selected patients. We sought to determine intermediate-term outcomes after Fontan conversion with arrhythmia operations and identify characteristics associated with decreased transplant-free survival.

Methods. The first 140 Fontan conversions with arrhythmia operations at a single institution were analyzed for predictors of cardiac death or transplant and incidence of arrhythmia recurrence.

Results. The median age at the Fontan conversion operation was 23.2 years (range, 2.6 to 47.3 years). Preoperative arrhythmias were present in 136 patients: right atrial tachycardia in 48 patients, left atrial tachycardia in 21, and atrial fibrillation in 67. Freedom from cardiac death or transplant was 90% at 5 years, 84% at 10 years, and 66% at 15 years. The median age at the last follow-up among survivors was 32 years (range, 15 to 61 years). By multivariable analysis, risk factors for cardiac death or heart transplantation were a right or indeterminate ventricular morphology, cardiopulmonary bypass time exceeding 240 minutes, ascites, protein-losing enteropathy, or a biatrial arrhythmia operation at the time of conversion. Freedom from recurrence of atrial tachycardia was 77% at 10 years. Among 67 patients with atrial fibrillation undergoing biatrial arrhythmia operations, none had recurrent atrial fibrillation.

Conclusions. Freedom from cardiac death or transplant for patients undergoing Fontan conversion with an arrhythmia operation is 84% at 10 years. The effects of atrial arrhythmia operations are durable in most patients. These outcomes may serve as useful benchmarks for alternative management strategies.

(Ann Thorac Surg 2016;101:717–24)

Approach to Device Decision in the UVH Based on Patient Weight and Mechanism of Failure

	Systolic Dysfunction	Diastolic Dysfunction
	Elevated EDP, low CO	**Elevated EDP, normal CO**
< 15 kg	Para-corporeal pulsatile VAD (systemic ventricle-aortic cannulation)	Extra-corporeal continues flow VAD Para-corporeal pulsatile VAD (atrio-aortic cannulation)
15-50 kg	Long-term intra-corporeal centrifugal VAD	Long-term intra-corporeal centrifugal VAD
> 50 kg	Long-term intra-corporeal axial flow VAD	Long-term intra-corporeal axial flow VAD TAH

Approach to Device Decision in the UVH Based on Patient Weight and Mechanism of Failure

	Increased PVR/Fontan Failure	Mixed Type
	Elevated CVP, hepatic congestion	**Elevated CVP + EDP, pulmonary congestion, low CO**
< 15 kg	Glenn ± extra-corporeal continuous-flow VAD transitioned to a Para-corporeal pulsatile VAD	Glenn ± extra-corporeal continuous-flow VAD transitioned to a Para-corporeal pulsatile VAD
15-50 kg	Glenn ± long-term intra-corporeal centrifugal VAD	Glenn ± long-term intra-corporeal centrifugal VAD
> 50 kg	TAH	TAH
	Bi-ventricular para-corporeal VAD	Bi-ventricular para-corporeal VAD

Heart transplantation in Fontan (ATS 2013, Chicago)

Heart Transplantation for the Failing Fontan

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Divisions of Cardiovascular Thoracic Surgery, Cardiology, and Anesthesiology, Ann & Robert H. Lurie Children's Hospital of Chicago, and Departments of Surgery and Pediatrics, Northwestern University Feinberg School of Medicine, Chicago, Illinois

Background. Patients with failing Fontan circulation are at high risk for complications after heart transplantation (HTx) because of multiple prior operations, elevated panel reactive antibody, hepatic dysfunction, coagulopathy, protein-losing enteropathy (PLE), and poor nutrition. The purpose of this review was to evaluate the outcome of HTx for these patients, including those who are status post-Fontan conversion.

Methods. Of 206 heart transplants at Ann & Robert H. Lurie Children's Hospital of Chicago from 1990 to 2012, 22 patients had a failing Fontan. Median age at HTx was 12.2 years, median interval from initial Fontan to HTx was 7.1 years. Potential preoperative risk factors included PLE (n = 15), mechanical ventilation (n = 8), prior Fontan conversion (n = 7), renal failure (n = 3), and plastic bronchitis (n = 2) Median number of prior operations was 3. Donor branch pulmonary arteries were used in 17 patients.

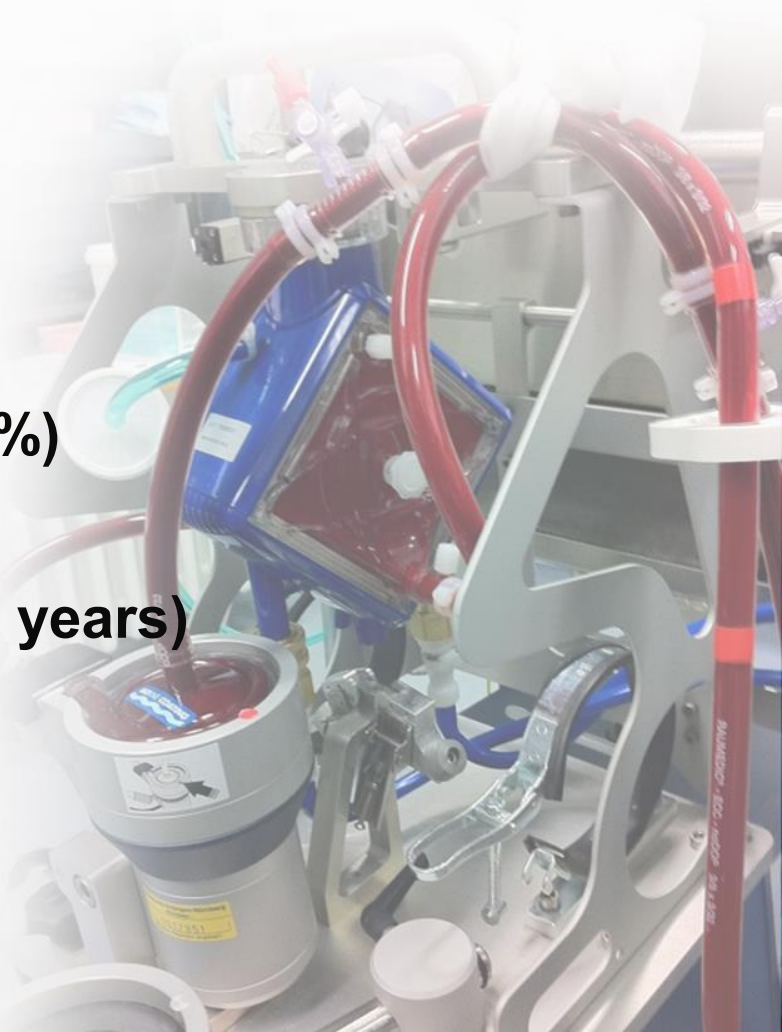
Results. There were 5 early deaths (23%), due to graft failure (1), pulmonary hypertension (1), and infection (3). There were 3 late deaths (13%) at 1, 5, and 8 years. Two of 3 patients with preoperative renal failure died. Survivors who had preoperative PLE (n = 11) and preoperative plastic bronchitis (n = 2) experienced complete resolution of these pathological conditions after heart transplantation. Median length of stay was 30 days. Five of 7 Fontan conversion patients survived, and 6 of 8 preoperative ventilator-dependent patients survived. One-, 5-, and 10-year survival was 77%, 66%, and 45%, respectively.

Conclusions. The operative mortality of HTx for patients with a failing Fontan is high. Using the donor branch pulmonary arteries greatly facilitated the transplant. Because infection caused the majority of early deaths, lower intensity initial immunosuppression may be warranted. Transplantation was successful in treating PLE in all survivors. Prior Fontan conversion was not a risk factor. Preoperative mechanical ventilation was not a risk factor. Preoperative renal failure may be a relative contraindication. Earlier referral of failing Fontan patients may improve results.

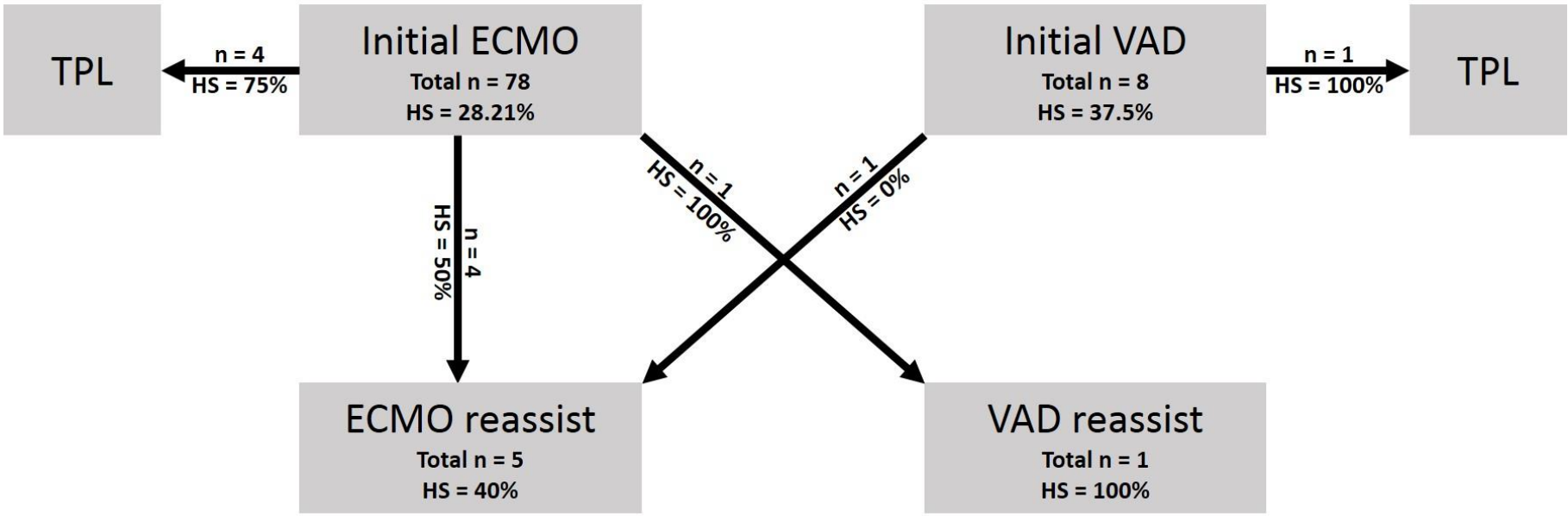
(Ann Thorac Surg 2013;96:1413–9)

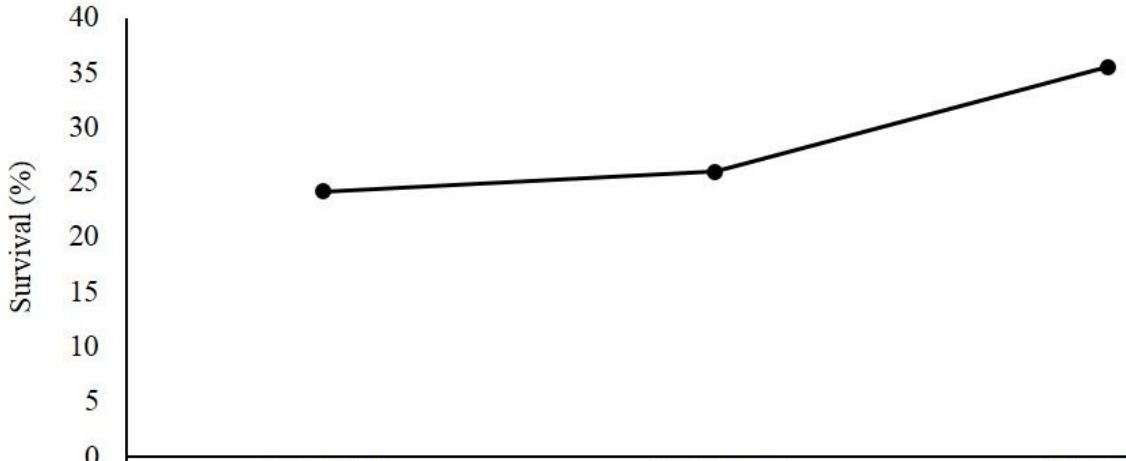
MCS Experience in SNUCH (KCJ 2017 in press)

- Duration 1999.- 2014.
- No 86
  - VA ECMO 77
  - VAD 7
  - ECMO to VAD 1
  - VAD to ECMO 1
- Sex (M:F) 51(59.3%) : 35(40.7%)
- Age 5.82 ± 8.55 years
  (range: 1 day - 41.6 years)
- CHD:ACHD 77 : 9 (> 18 years)



Flowchart of Mode Selection and Era Effect

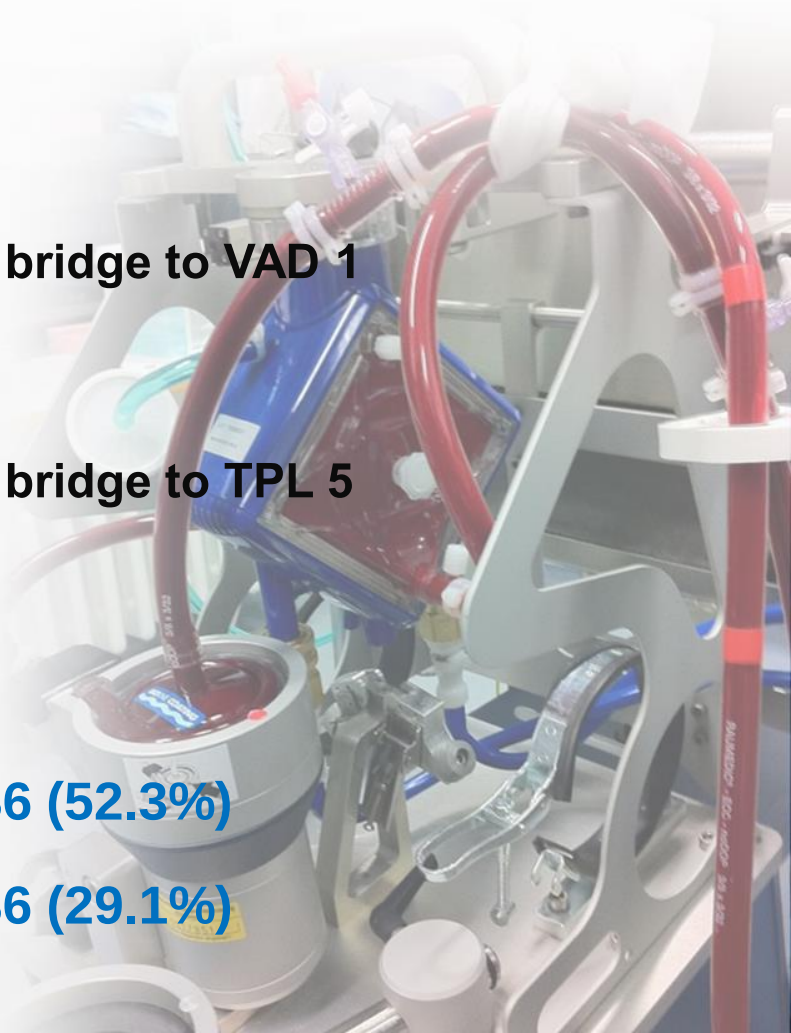




	1999-2005	2006-2010	2011-2014
Survival rate	24.14	25.93	35.48
Total number	29	27	31

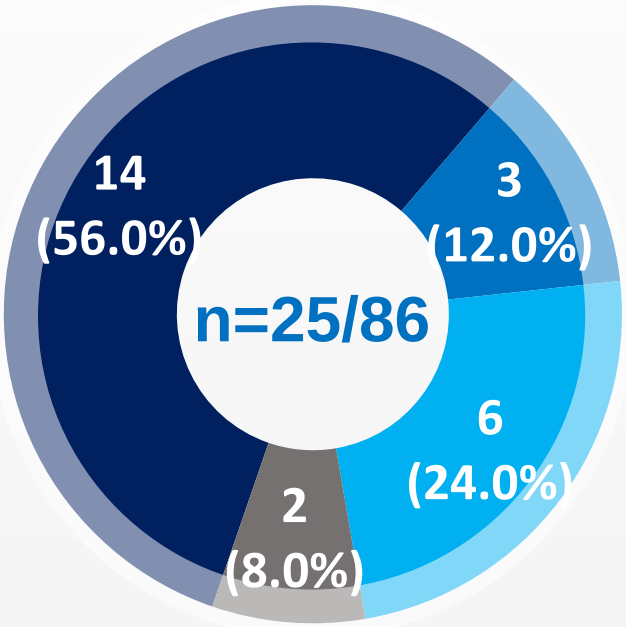
According to Indication

- **Post-cardiotomy** — **60**
  - Weaning — 32 (53.3%)
  - Survival to discharge — 14 (23.3%)
- **Acute myocarditis** — **5**
  - Weaning — 4 (80.0%)
  - Survival to discharge — 3 (60.0%) - bridge to VAD 1
- **DCMP** — **13**
  - Weaning — 6 (46.2%)
  - Survival to discharge — 6 (46.2%) - bridge to TPL 5
- **As rescue during CPR** — **8**
  - Weaning — 3 (37.5%)
  - Survival to discharge — 2 (25.0%)
- **Weaning** — **45/86 (52.3%)**
- **Survival to discharge** — **25/86 (29.1%)**

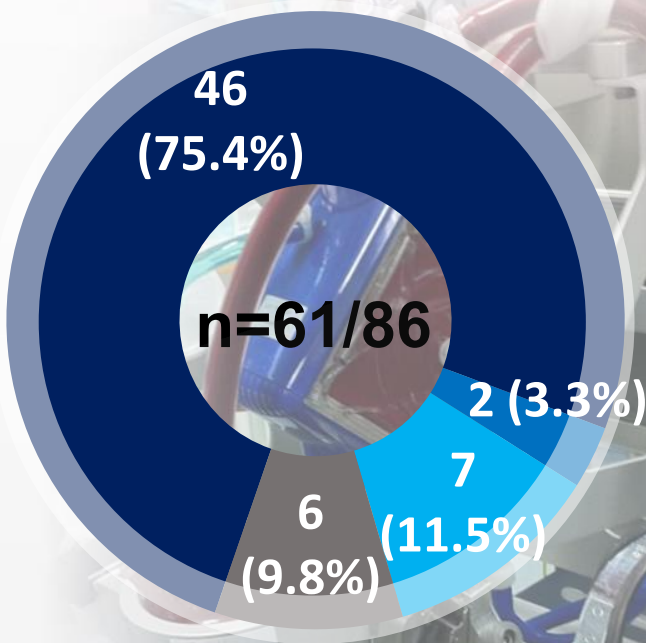


Survivors vs Nonsurvivors

Survivors
Age (years) 6.32± 8.08



Nonsurvivors
Age (years) 5.62±8.86



Postcardiotomy Myocarditis DCMP Rescue

According to Age

Trend

Neonate	3 / 24 (12.5%)
1 M ~ 1 Y	8 / 21 (38.1%)
1 Y ~ 10 Y	9 / 22 (40.9%)
10 Y ~ 18 Y	3 / 10 (30.0%)
> 18 Y	2 / 9 (22.2%)

p=0.19

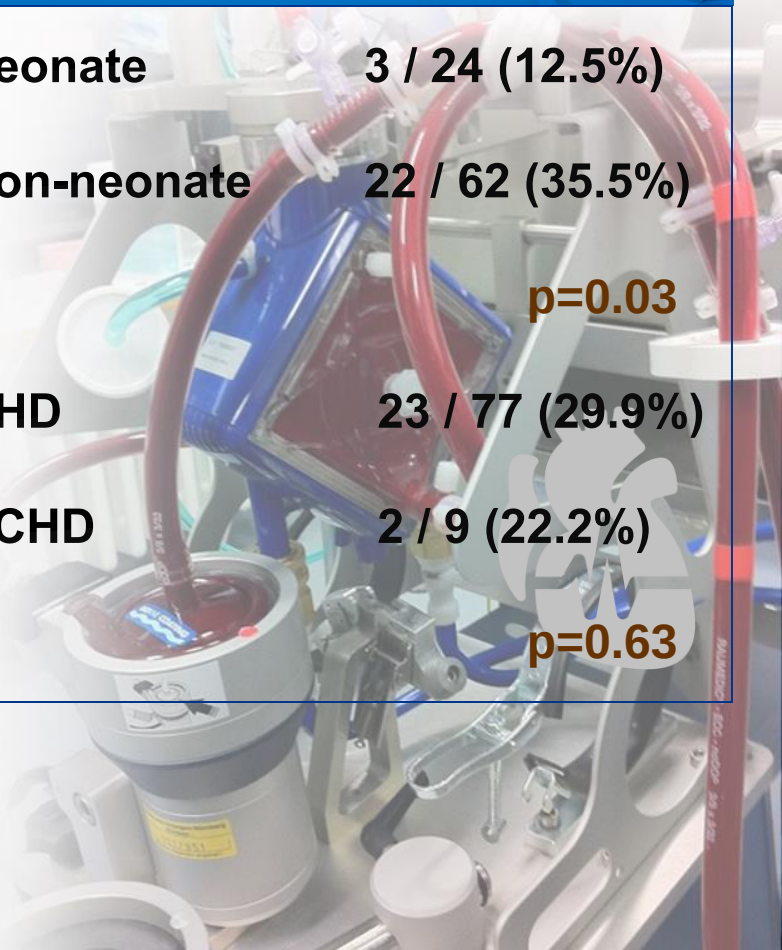
Comparison

Neonate	3 / 24 (12.5%)
Non-neonate	22 / 62 (35.5%)

p=0.03

CHD	23 / 77 (29.9%)
ACHD	2 / 9 (22.2%)

p=0.63



Risk Factors for MCS

Ventricle type

- Single ventricle — 1 / 14 (7.1%)
- Biventricle — 24 / 72 (33.3%)

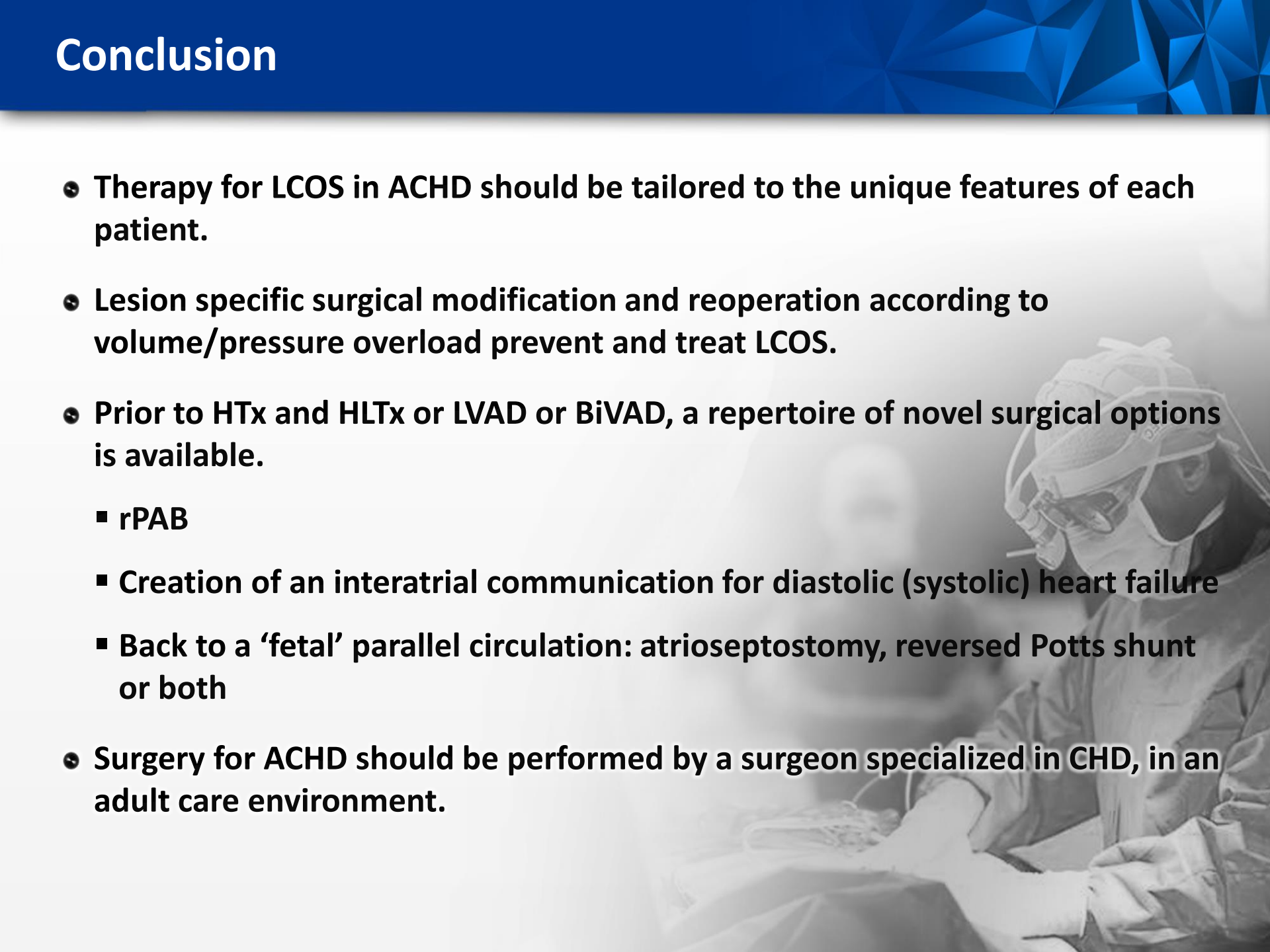
p<0.05

- **Multivariable analysis of mortality risk factors**

Variables	OR (95% CI)	p-value
PaO_2 < 100 mmHg before assist	4.314 (1.082-17.201)	0.04
Support timing	0.549 (0.317-0.951)	0.03
Neurologic complication	0.044 (0.003-0.720)	0.03
Hemorrhage	0.114 (0.016-0.812)	0.03
Peak serum bilirubin during assist	0.758 (0.603-0.953)	0.02
Peak serum AST during assist	0.999 (0.998-1.000)	0.03

Conclusion

- Therapy for LCOS in ACHD should be tailored to the unique features of each patient.
- Lesion specific surgical modification and reoperation according to volume/pressure overload prevent and treat LCOS.
- Prior to HTx and HLTx or LVAD or BiVAD, a repertoire of novel surgical options is available.
  - rPAB
  - Creation of an interatrial communication for diastolic (systolic) heart failure
  - Back to a ‘fetal’ parallel circulation: atrioseptostomy, reversed Potts shunt or both
- Surgery for ACHD should be performed by a surgeon specialized in CHD, in an adult care environment.



Conclusion

- Short-term MCS with ECMO is a reasonable approach of support to improve survival to discharge when other therapeutic options such as VAD and heart transplant are not immediately feasible or practical.
  - While survival to discharge for adults with cardiac causes from the ELSO registry is reported to be 40%, nearly half of patients survive to discharge following ECMO after high-risk cardiac operations in ACHD.
- Broader use of LVADs in ACHD may result in benefits similar to NCHD.
- ACHD undergoing BiVAD/TAH support have worse survival compared with NCHD due to worse INTER-MACS profile and greater comorbid burden.
- We should refine indications, time of listing and pre-, per-and postoperative care, to reduce perioperative mortality, which remains ‘‘the Achilles heel’’ of heart TPL in ACHD. Further studies are needed to investigate impact of immunosuppressive regimens, underlying congenital abnormalities, and status of hepatic function, particularly in failing Fontan.

감사합니다.
Thank you!