Rehabilitating or “Growing” the Left Ventricle: Promise and Pitfalls

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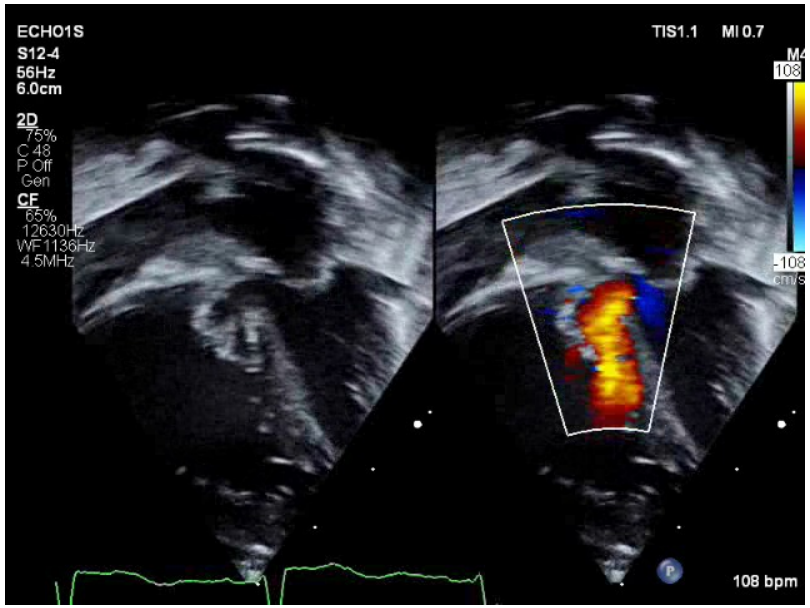
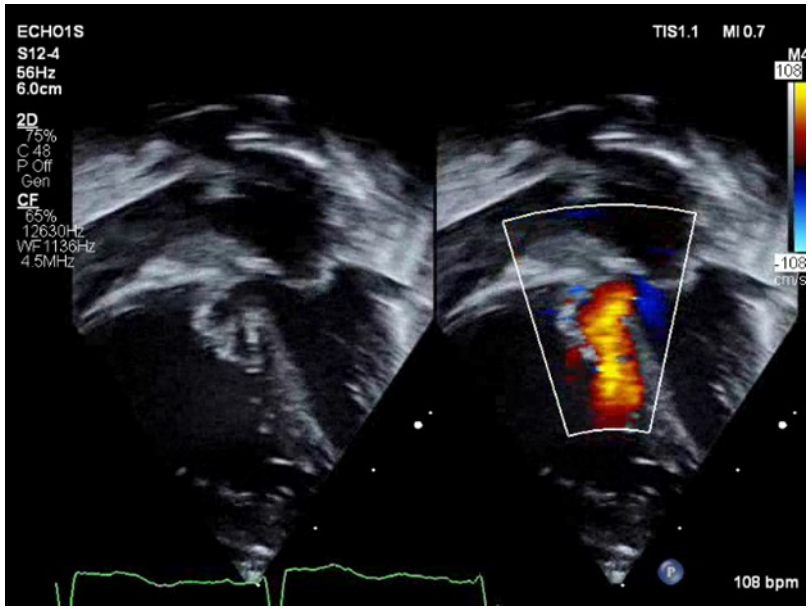
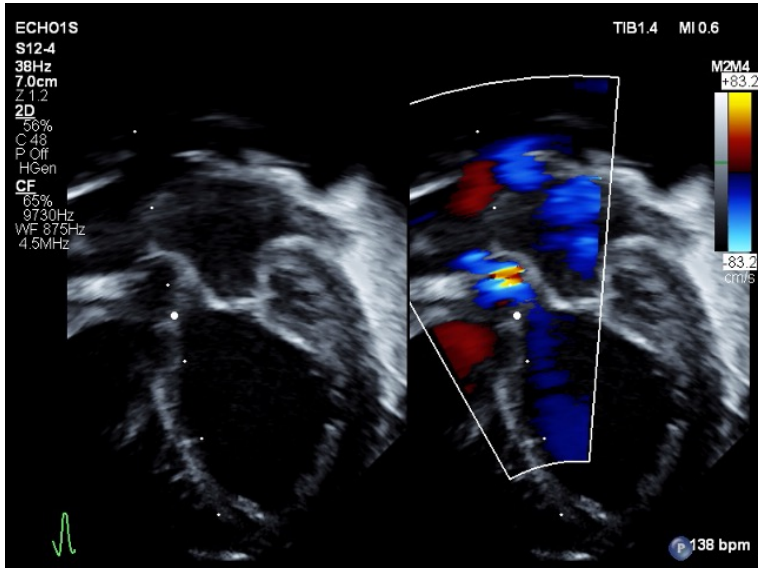
What is a Borderline LV?

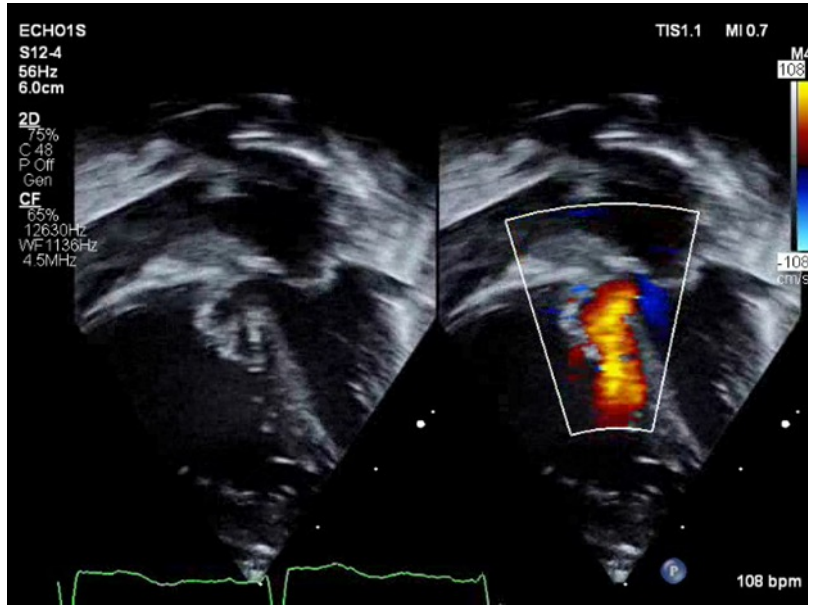


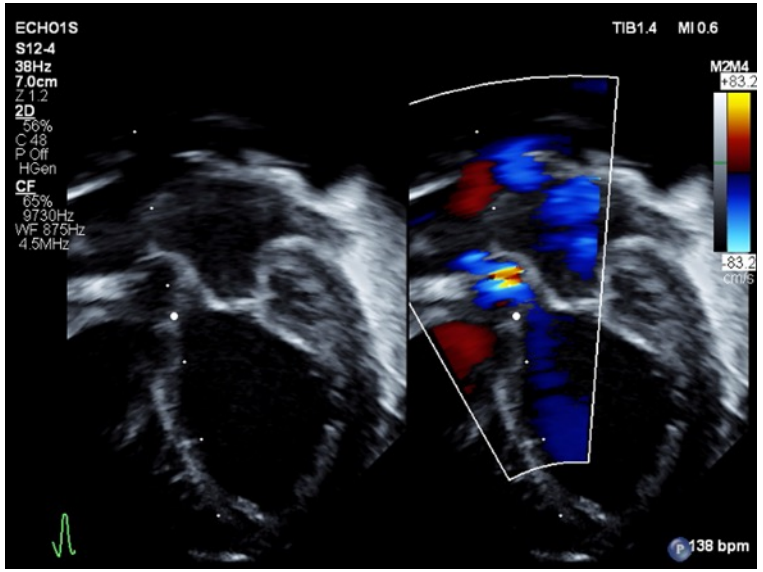
What is a Borderline LV?

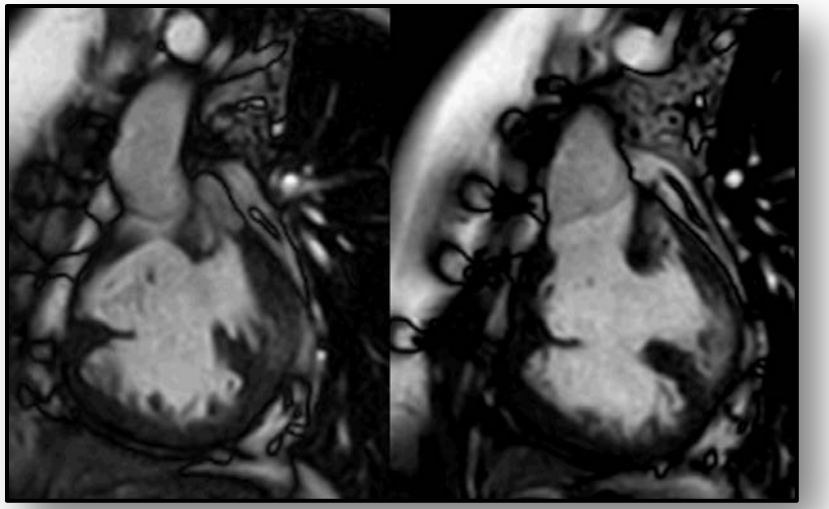




What is a Borderline LV?







What is a Borderline LV?

- HLHS (MS/AS)
- Shones

- Unbalanced AVC
- Coarctation and VSD

- Double Outlet Right Ventricle
- Complex Transposition of Great Arteries
- Upstairs Downstairs ventricles
- Double inlet Ventricle
- Straddling AV Valves

Traditional Paradigm
Borderline Left Heart

Single Ventricle Palliation

Fontan Kreutzer

Primary Biventricular Repair

Tools to assist decision ?

Biventricular Repair Approach in
Ducto-Dependent Neonates With
Hypoplastic but Morphologically Normal Left Ventricle

Alain Serraf, MD, Jean Dominique Piot, MD, Nicolas Bonnet, MD, François Lacour-Gayet, MD,
Anita Touchot, MD, Jacqueline Bruniaux, MD, Emré Belli, MD, Lorenzo Galletti, MD,
Claude Planché, MD

Biventricular Repair in Neonates With Hypoplastic Left Heart Complex

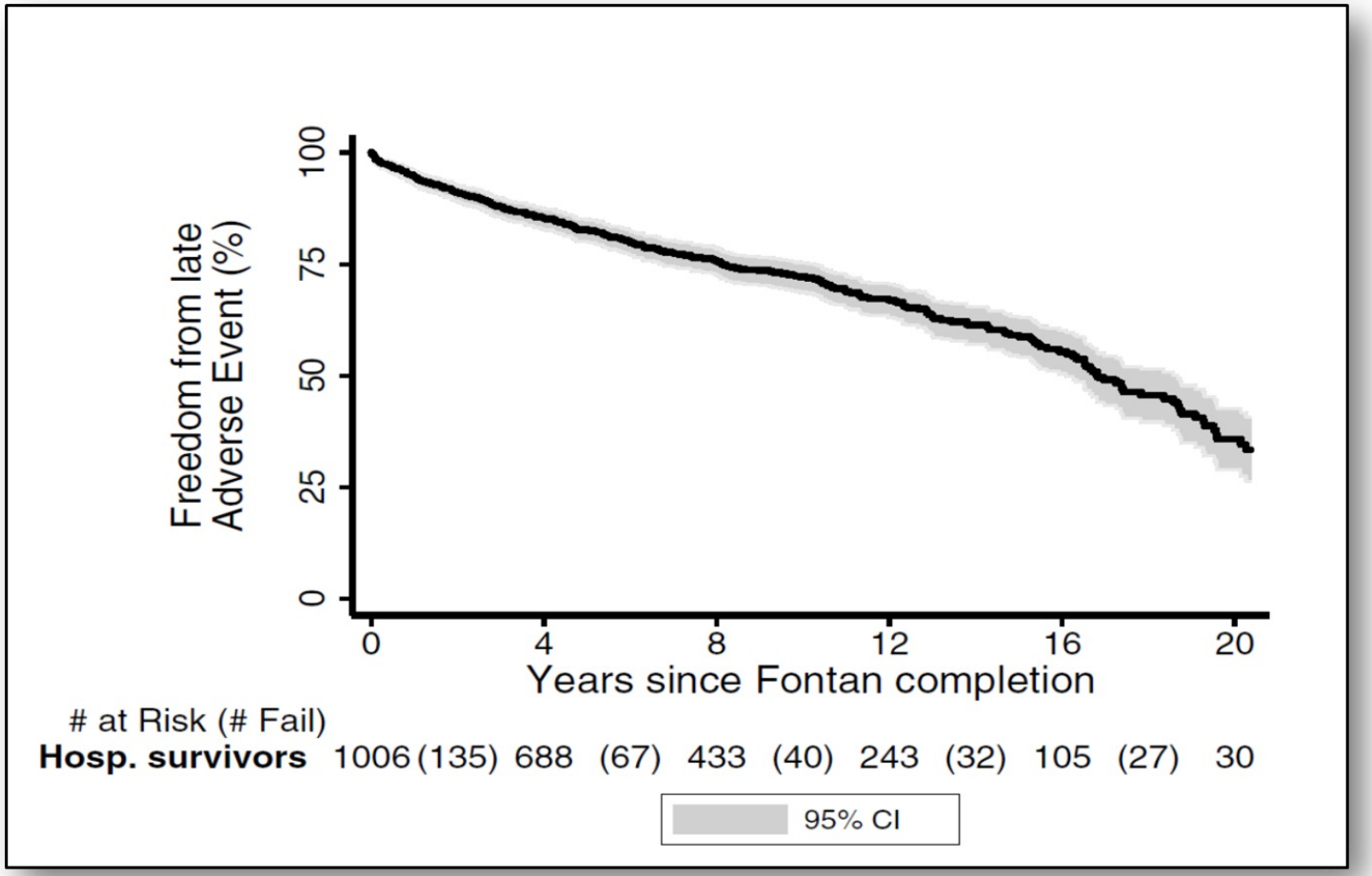
Christo I. Tchervenkov, MD, Stephen A. Tahta, MD, Luc C. Jutras, MD, and
Marie J. Béland, MD

CRITICAL AORTIC STENOSIS IN THE NEONATE: A MULTI-INSTITUTIONAL STUDY OF MANAGEMENT, OUTCOMES, AND RISK FACTORS

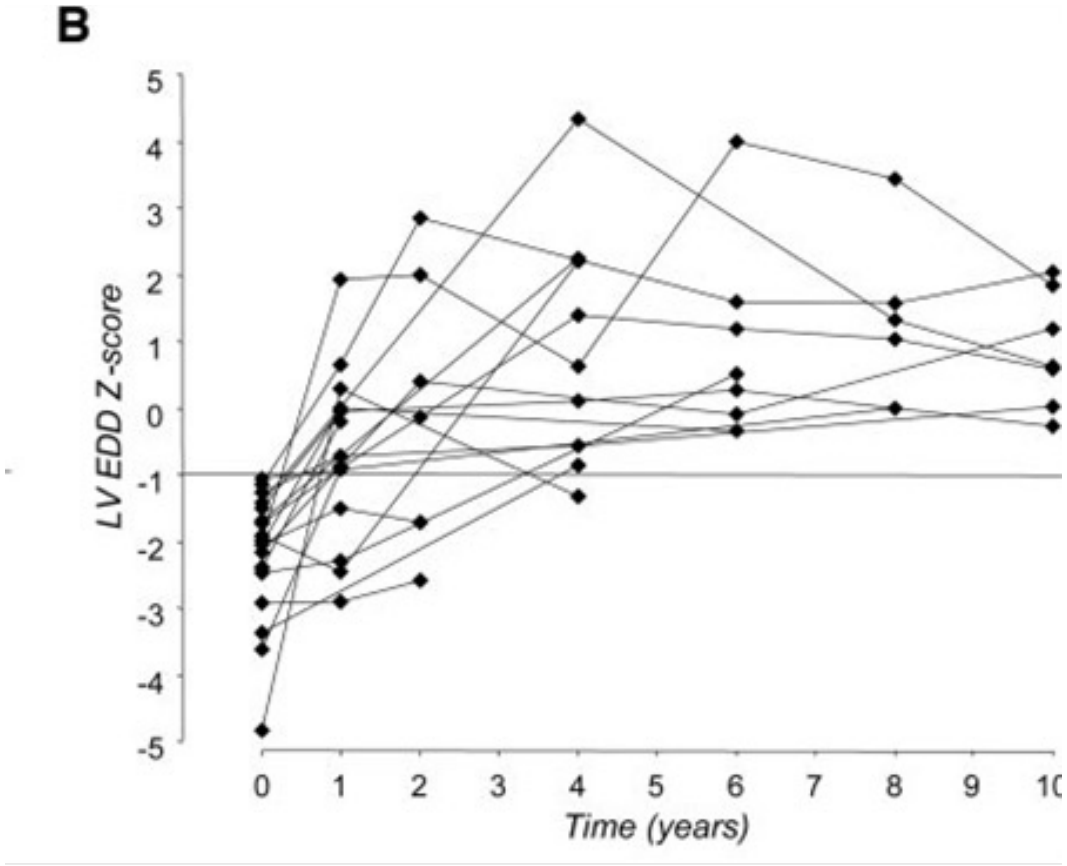
Predictors of Outcome of Biventricular Repair in Infants With Multiple Left Heart Obstructive Lesions

Marcy L. Schwartz, MD; Kimberlee Gauvreau, ScD; Tal Geva, MD

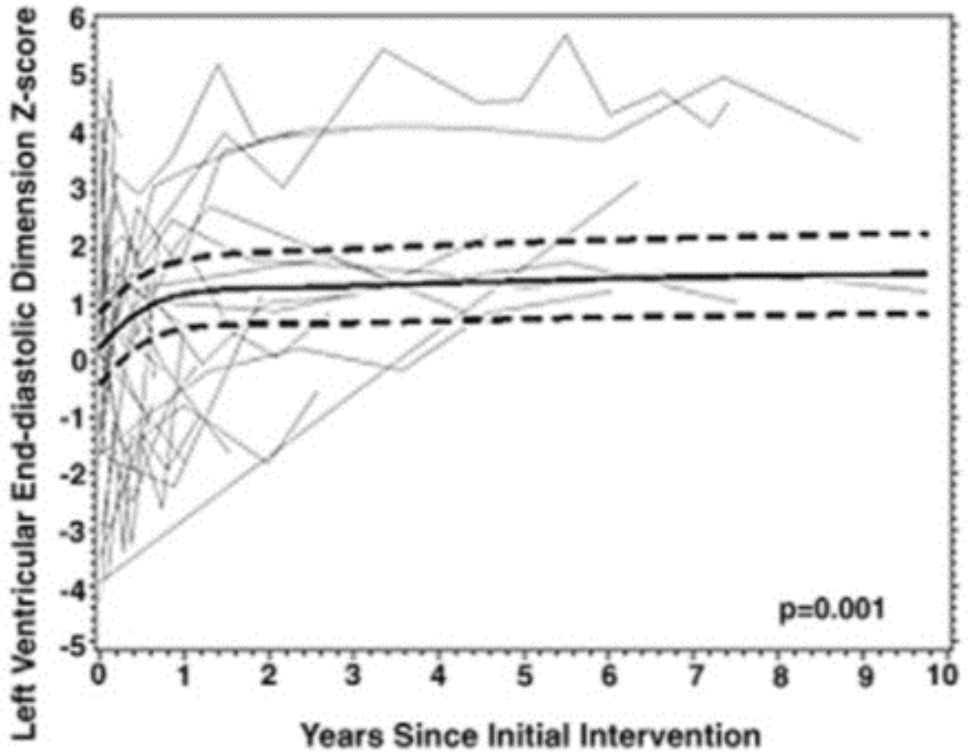
Fontan


Freedom from late Adverse Event (%)
0
25
50
75
100
0
4
8
12
16
20
Years since Fontan completion
at Risk (# Fail)
Hosp. survivors 1006 (135) 688 (67) 433 (40) 243 (32) 105 (27) 30
95% CI

Ventricles can grow…

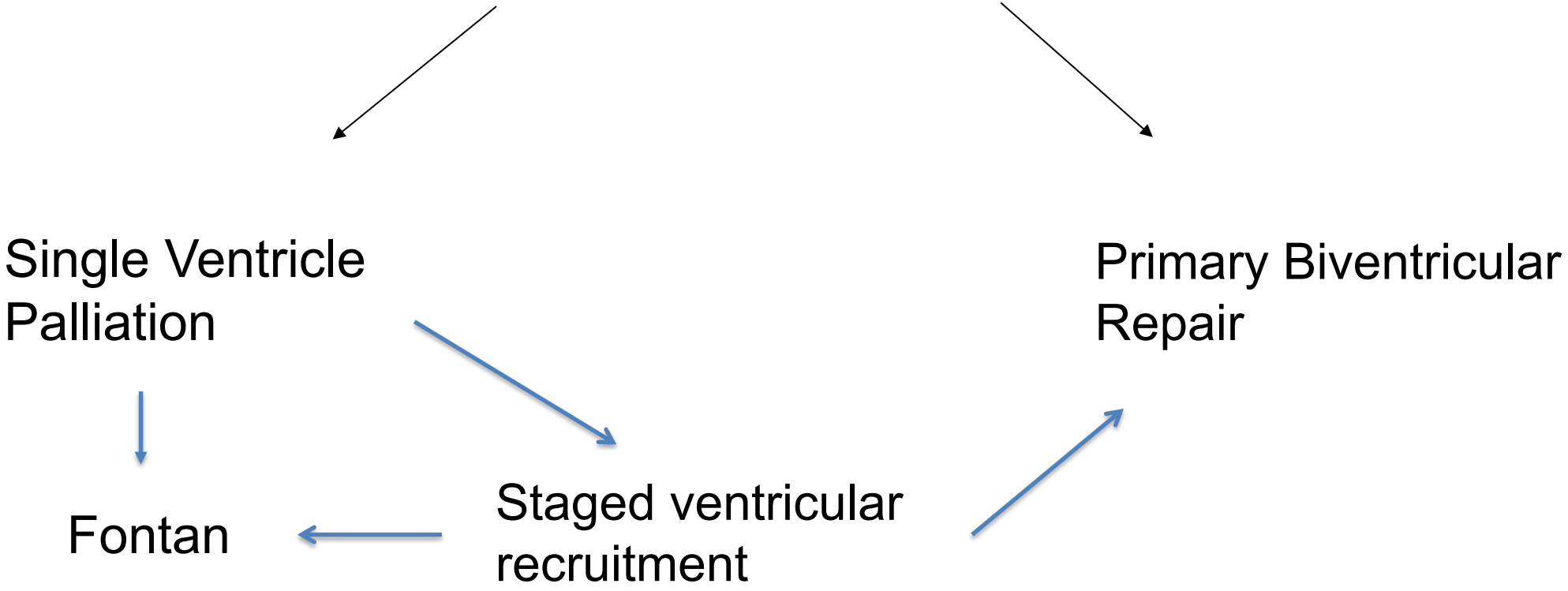


McElhenney et al. 2005 (CHB)

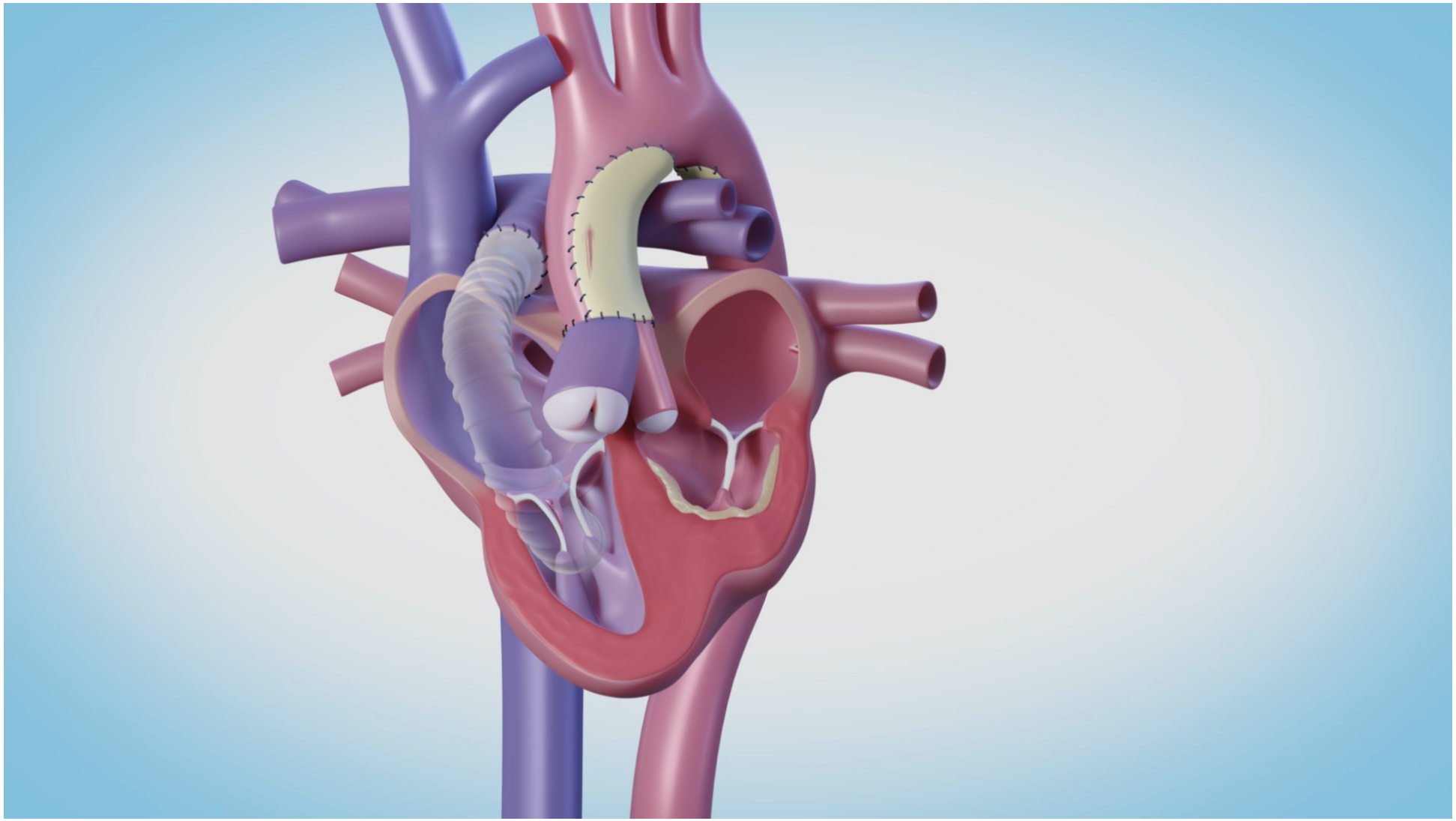


Han et al. 2007 (Toronto)

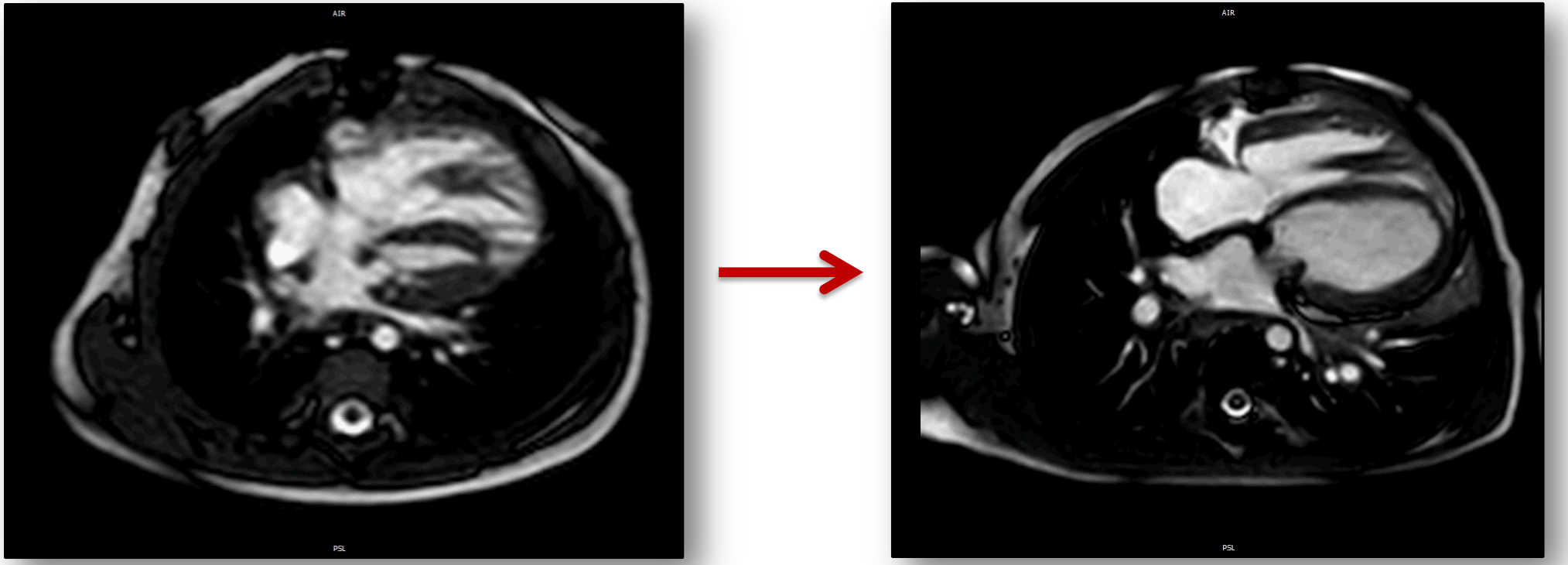
Modified Paradigm
Borderline Left Heart



HLHS variants – Staged Recruitment

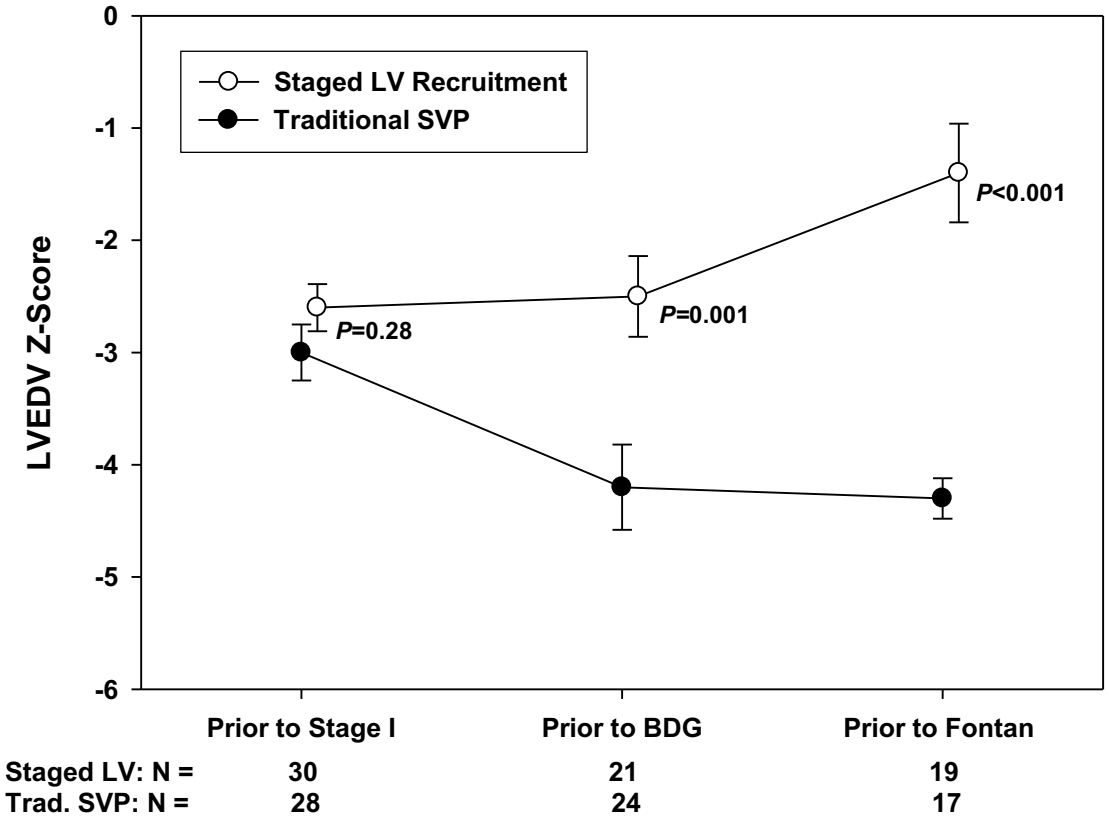


LV growth with staged recruitment



LV EDV after staged recruitment

Figure 3A



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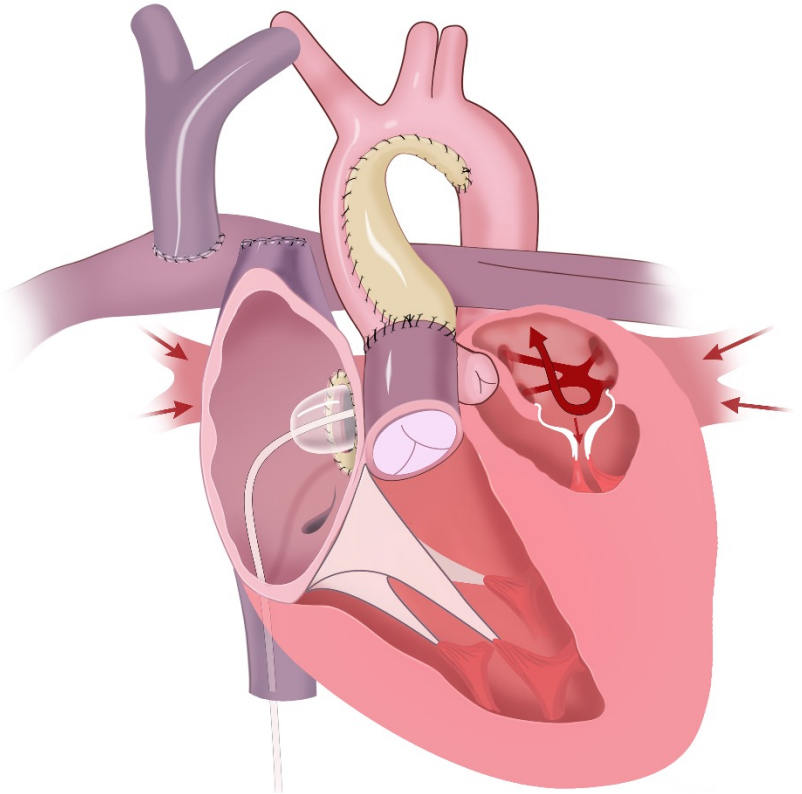
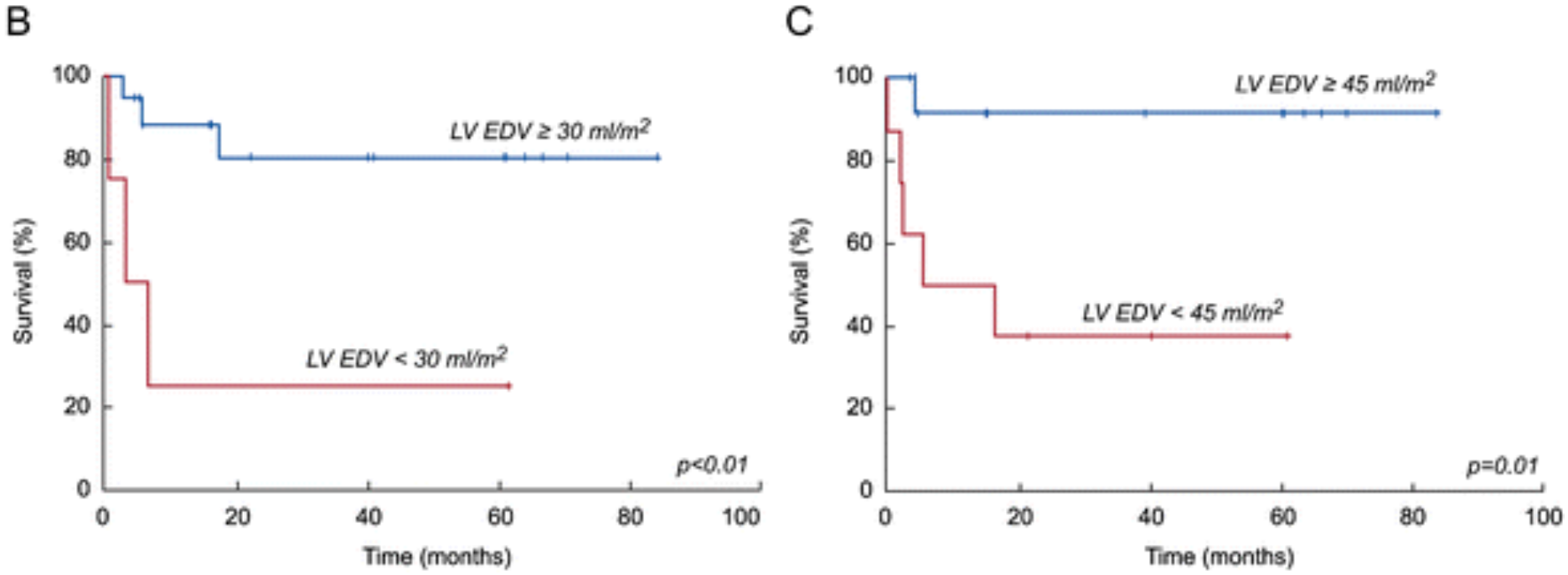
Emani et al JACC 2012

Pushing the limits – LA Hypertension



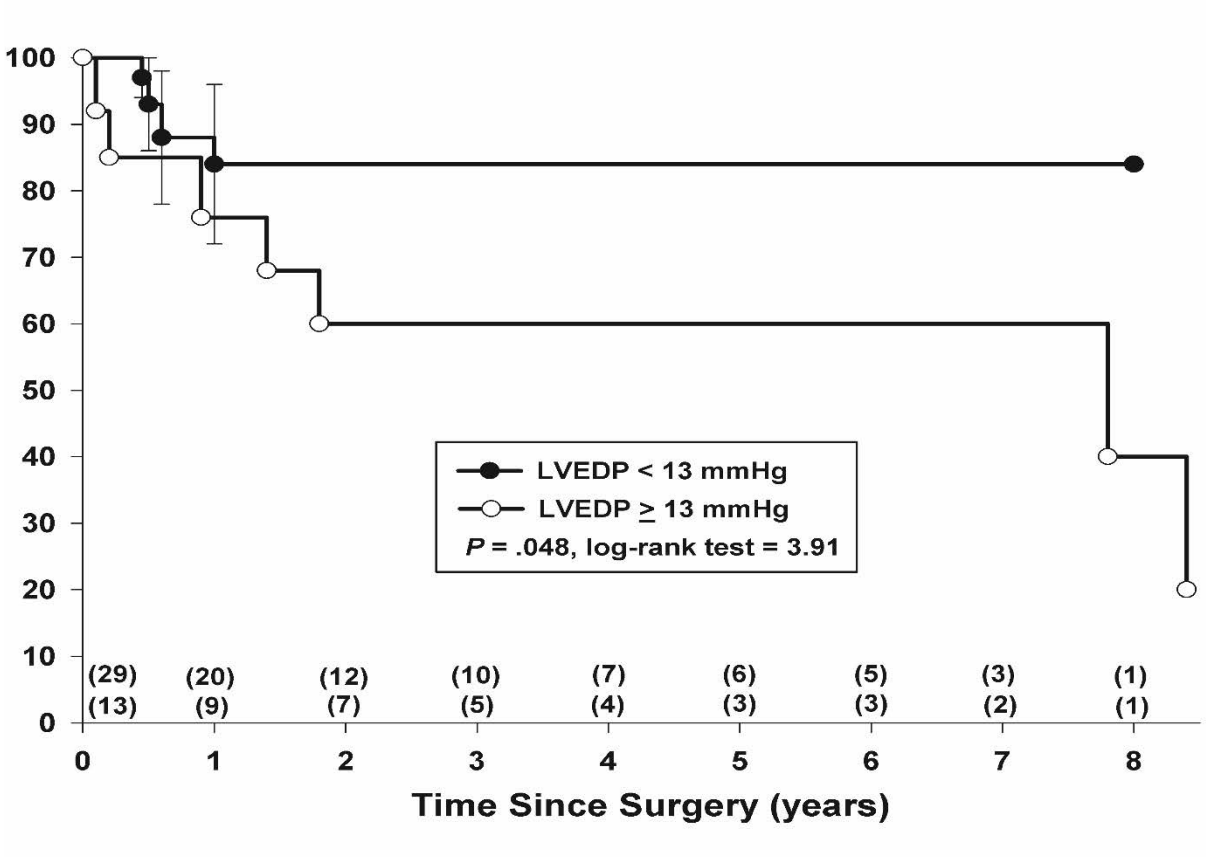
Predictor of Outcomes - LVEDV



- Banka et al JCMR 2014

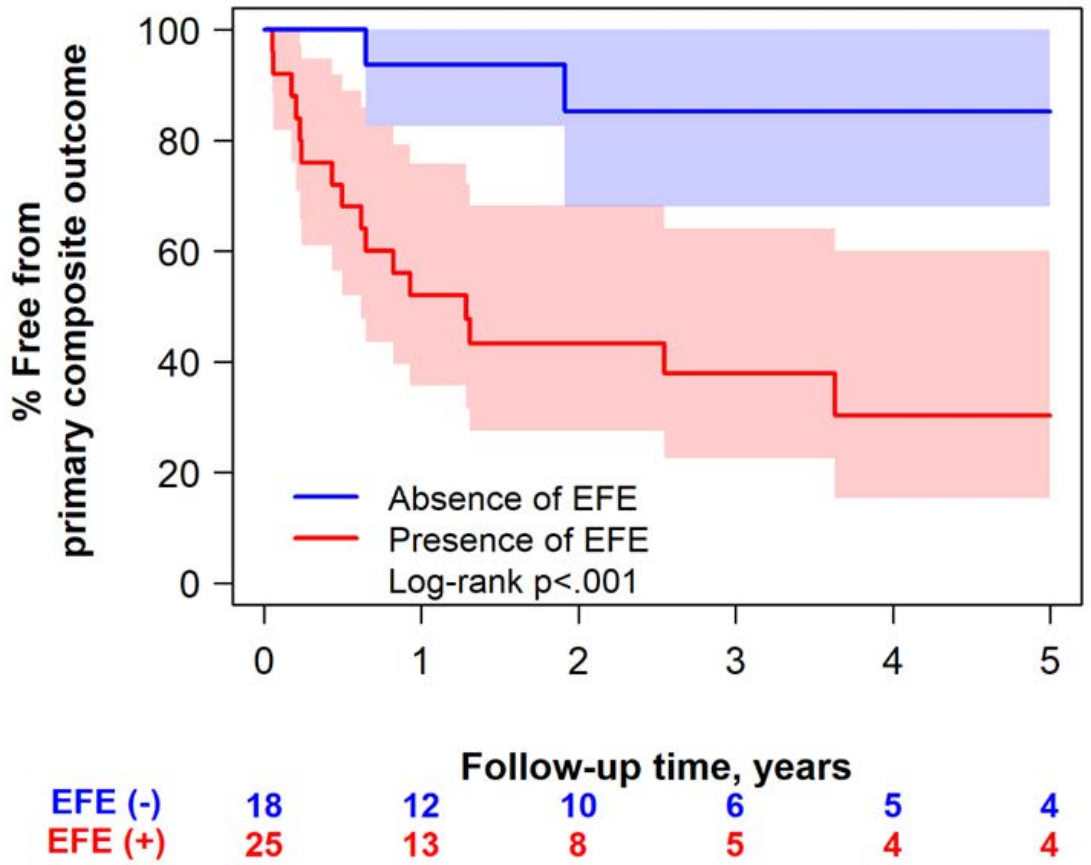
Predictor of Outcomes - LVEDP

Freedom from adverse outcome



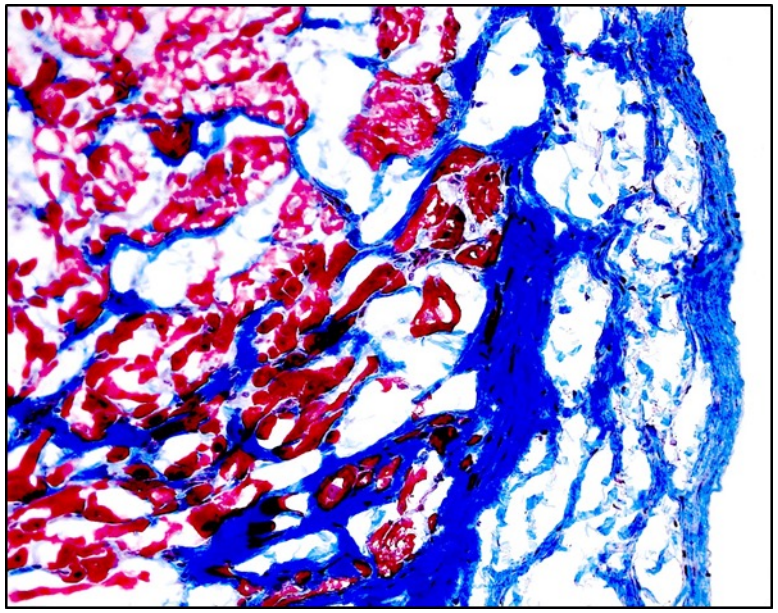
Predictor of Outcomes - EFE

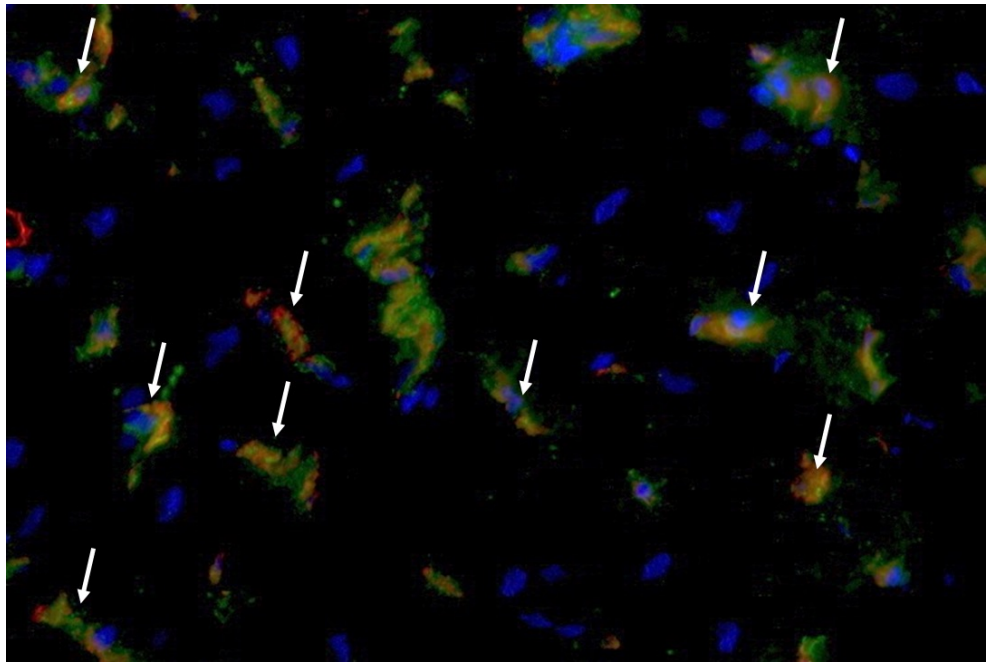
Freedom from adverse outcome



Beattie et al. JTCVS in press

What is EFE?



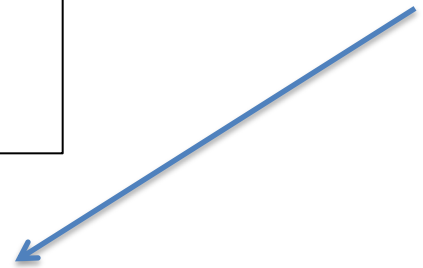


Beattie et al. JTCVS in press

Initial Single Ventricle Palliation

MRI + Cath (4-6 months)

LVEDV > 30 ml/m2
LVEDP < 13 mmHg
No EFE



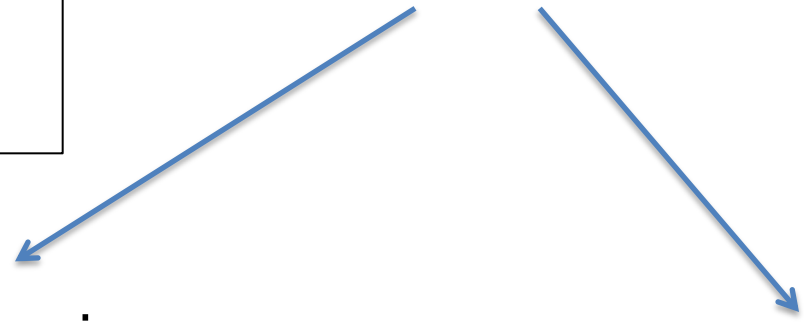
Biventricular Conversion (systemic LV)

Initial Single Ventricle Palliation

MRI + Cath (4-6 months)

LVEDV > 30 ml/m2
LVEDP < 13 mmHg
No EFE

LVEDV < 30 ml/m2
LVEDP > 13 mmHg
EFE



Biventricular Conversion (systemic LV)

Staged LV recruitment

- EFE resection
- MV / AoV repair
- ASD restriction
- Glenn + shunt

Initial Single Ventricle Palliation

MRI + Cath

LVEDV > 30 ml/m2
LVEDP < 13 mmHg
No EFE

LVEDV < 30 ml/m2
LVEDP > 13 mmHg
EFE

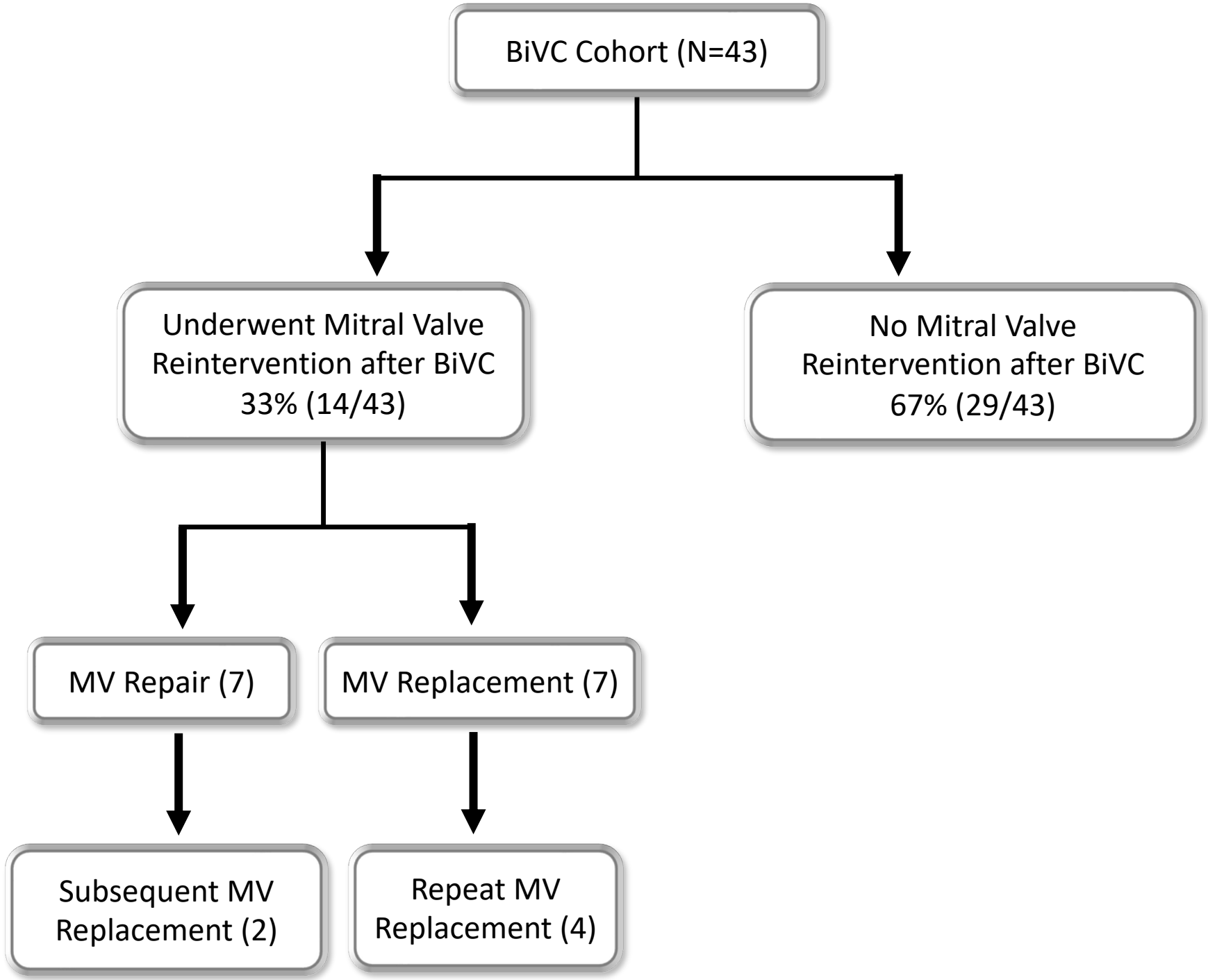
Biventricular Conversion (systemic LV)

Staged LV recruitment

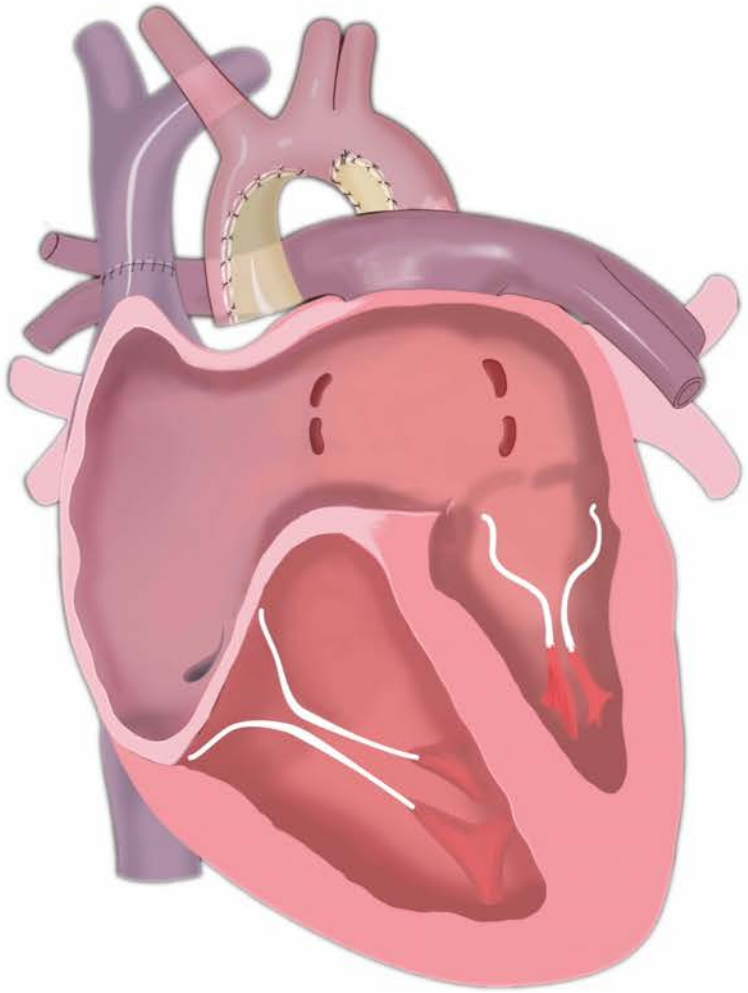
- EFE resection
- MV / AoV repair
- ASD restriction
- Glenn + shunt

Fontan Kreutzer

Repeat MRI + Cath in 1 year


BiVC Cohort (N=43)
Underwent Mitral Valve Reintervention after BiVC 33% (14/43)
No Mitral Valve Reintervention after BiVC 67% (29/43)
MV Repair (7)
MV Replacement (7)
Subsequent MV Replacement (2)
Repeat MV Replacement (4)

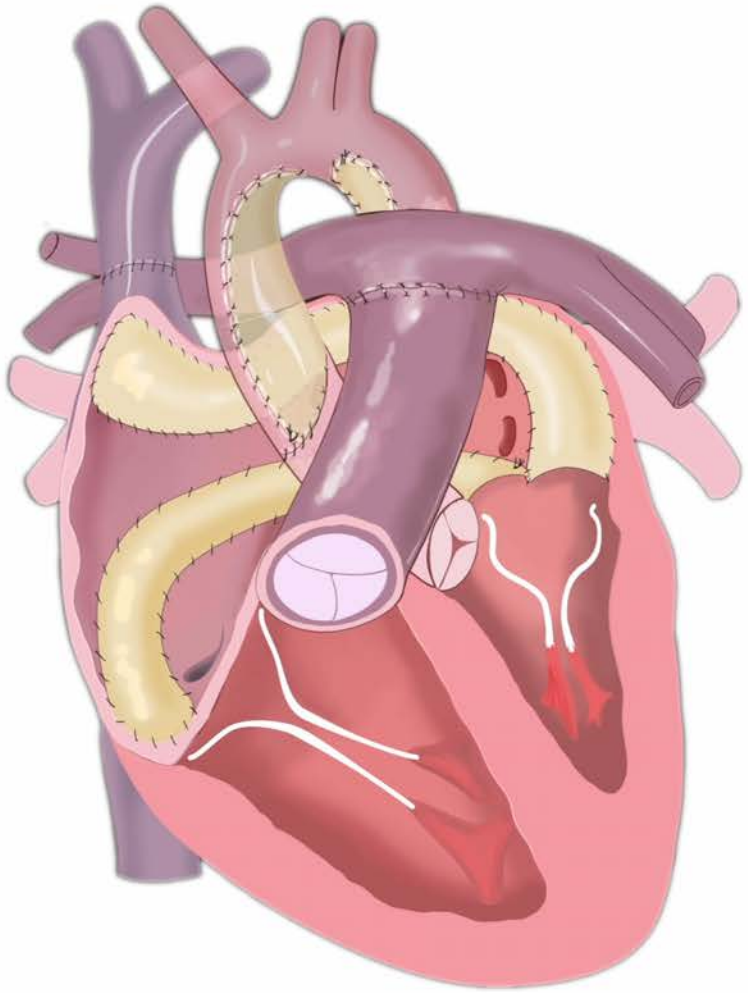
“Reverse Double Switch”



Boston Children's Hospital
Heart Center

Scully et al. Semin Thor Cardiov Surg, 2023,

“Reverse Double Switch”



Boston Children’s Hospital
Heart Center

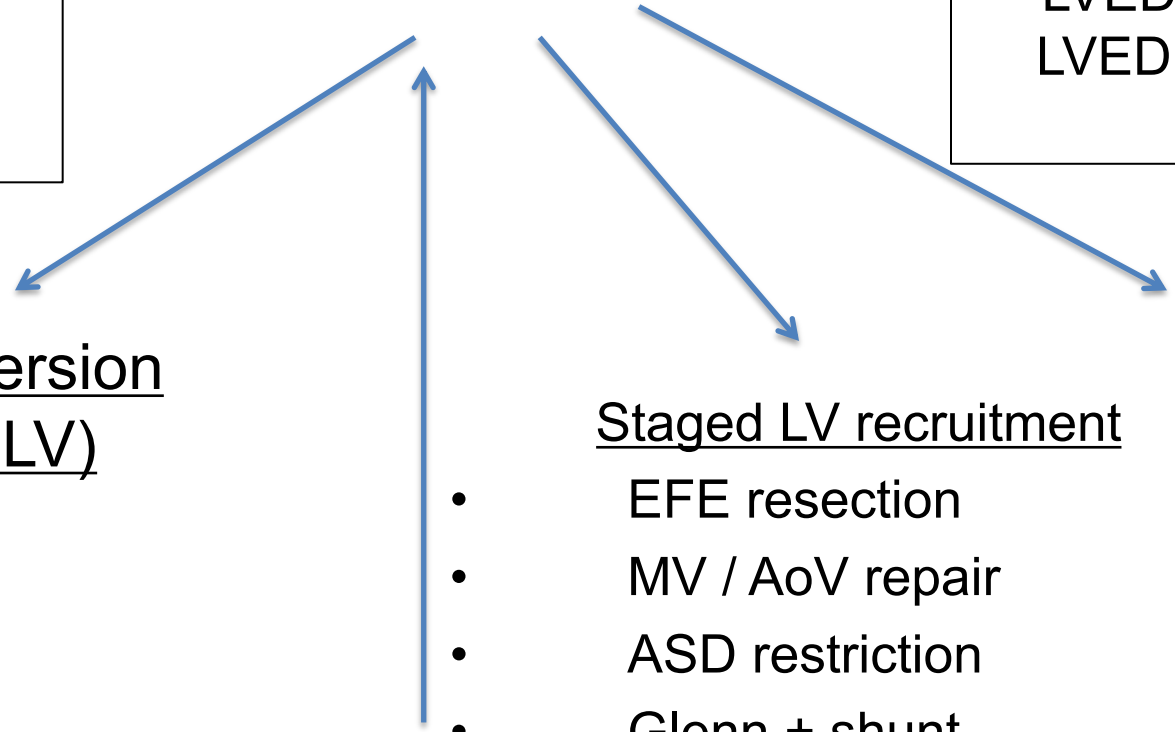
Scully et al. Semin Thor Cardiov Surg, 2023,

Initial Single Ventricle Palliation

MRI + Cath

LVEDV > 30 ml/m2
LVEDP < 13 mmHg
No EFE

LVEDV < 30 ml/m2
LVEDP > 13 mmHg
EFE



Biventricular Conversion (systemic LV)

Staged LV recruitment

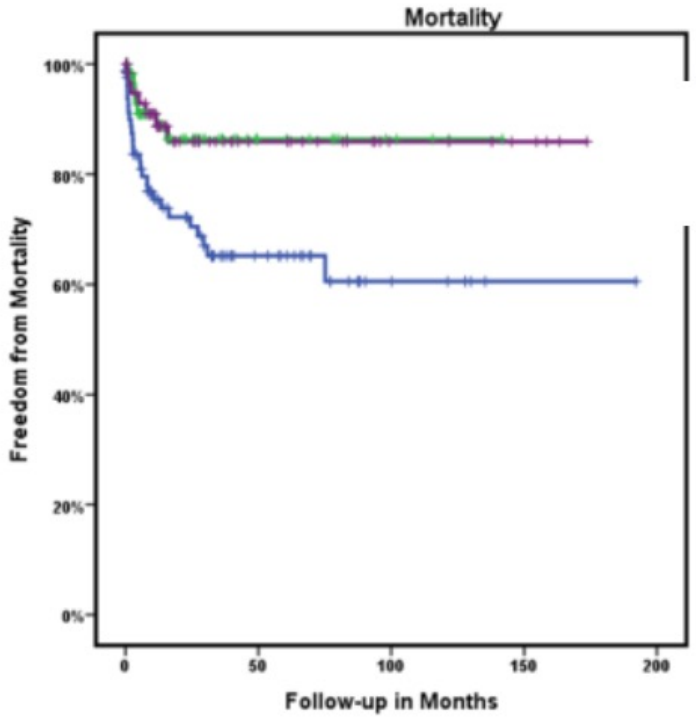
- EFE resection
- MV / AoV repair
- ASD restriction
- Glenn + shunt

Fontan Kreutzer

Reverse double Switch

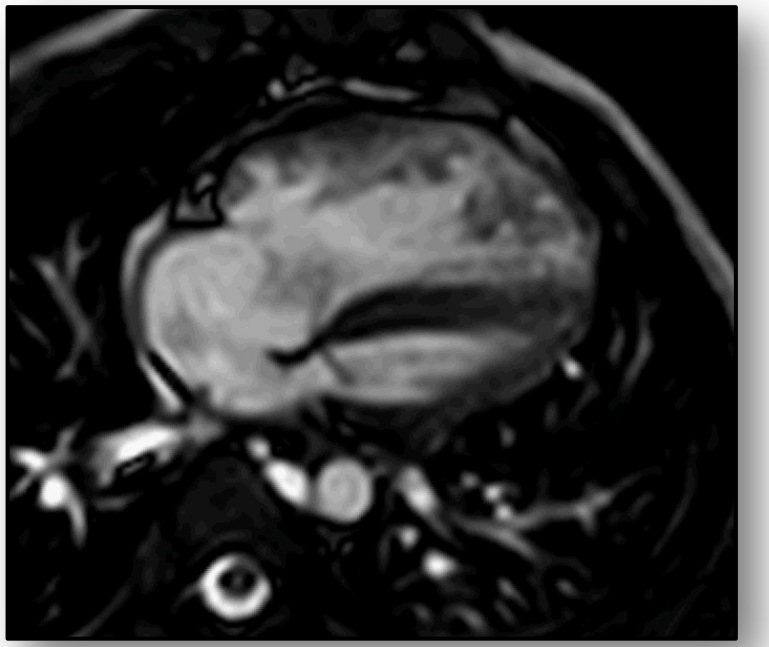
Repeat MRI + Cath in 1 year

Unbalanced AV canal

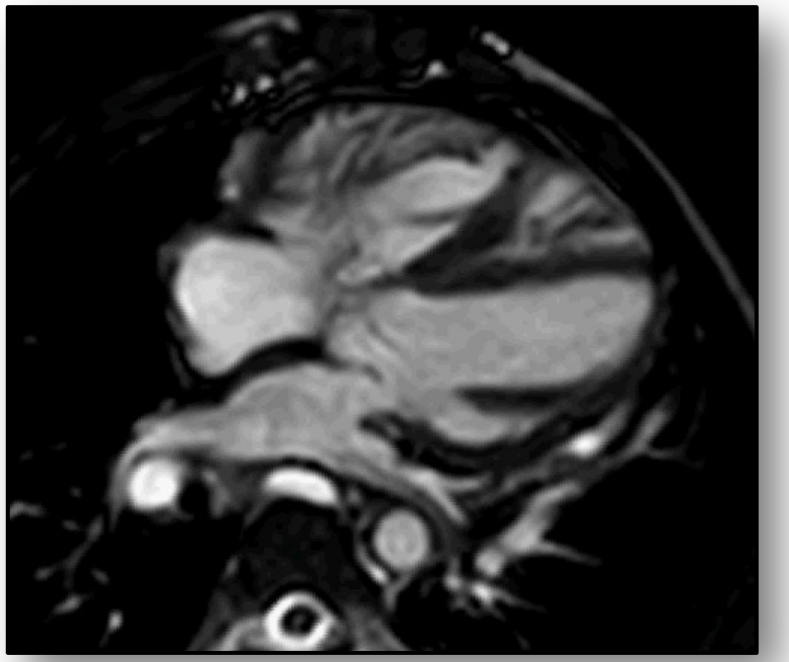


Mortality:	0m	12m	24m	36m	48m	60m	72m	84m	96m	108m	120m
SV	82	49	42	30	24	20	14	10	6	5	5
BIV	67	37	28	22	17	17	14	11	8	7	7
BIVR/BIVC	63	41	29	19	14	1	9	5	5	3	2

TGA borderline LV



Age 3

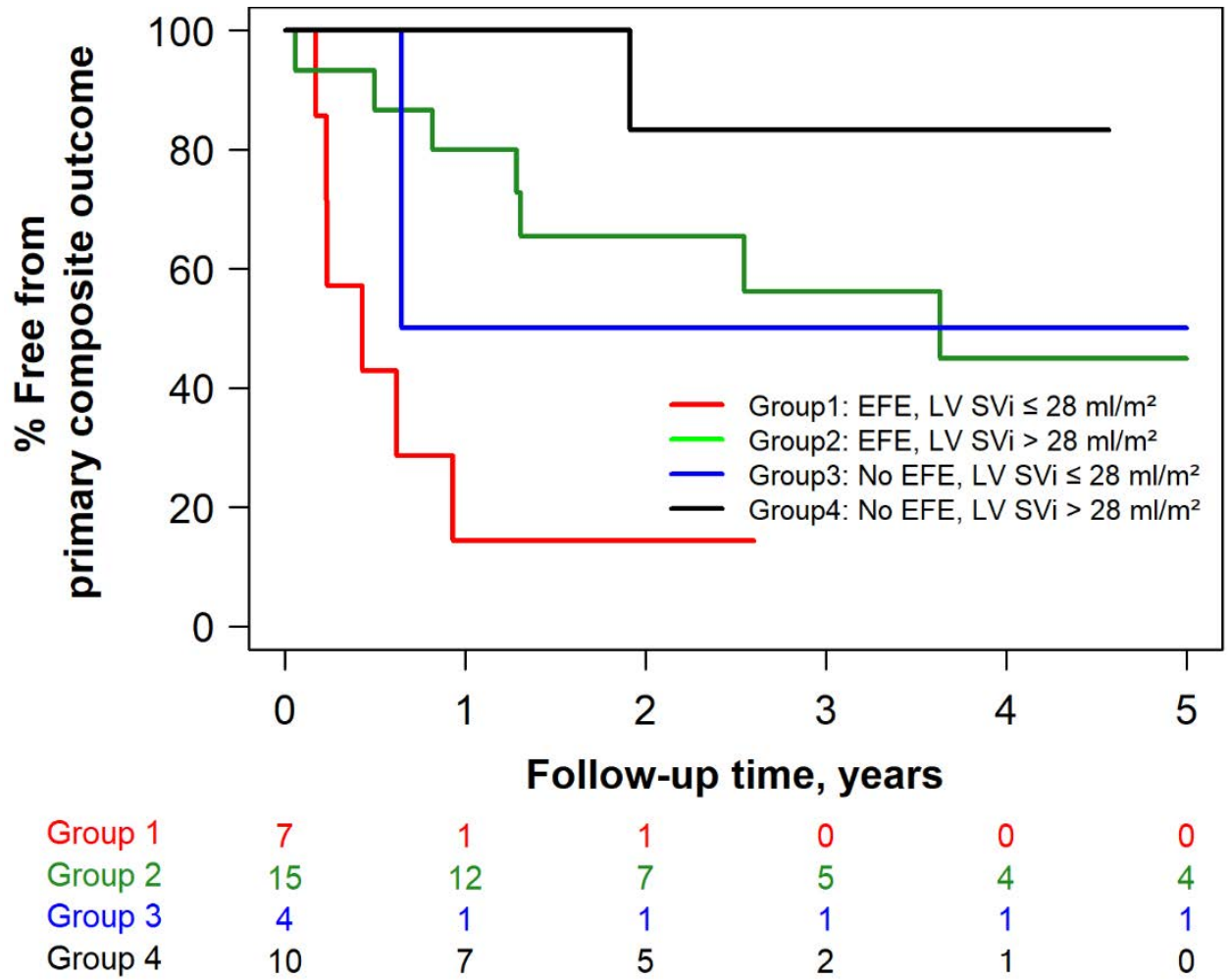


Age 4

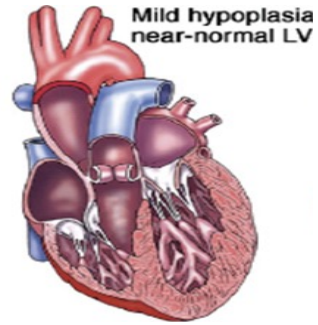
Conclusions

- Borderline LV - don’t force risky Biventricular repair…
- Single ventricle palliation
- Consider staged recruitment – Assess response
- Close monitoring to avoid LA hypertension
- Maintain candidacy for Fontan
- Patient selection for biventricular conversion
- Systemic RV + subpulmonary LV to avoid Fontan

Thank You


% Free from primary composite outcome
100
80
60
40
20
0
0
1
2
3
4
5
Follow-up time, years
Group1: EFE, LV SVi ≤ 28 ml/m²
Group2: EFE, LV SVi > 28 ml/m²
Group3: No EFE, LV SVi ≤ 28 ml/m²
Group4: No EFE, LV SVi > 28 ml/m²
Group 1 7 1 1 0 0 0
Group 2 15 12 7 5 4 4
Group 3 4 1 1 1 1 1
Group 4 10 7 5 2 1 0

What is Borderline Left Heart?



Biventricular repair

100%

Probability of Long-term Successful Biventricular Repair

0%

Z > -2.0 for:
- Ventricle size
- Aortic Valve
- Mitral Valve

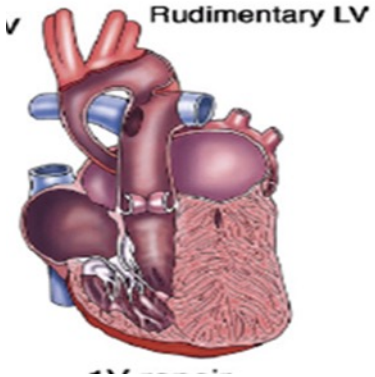
Normal systolic function

Normal diastolic function

What is Borderline Left Heart?

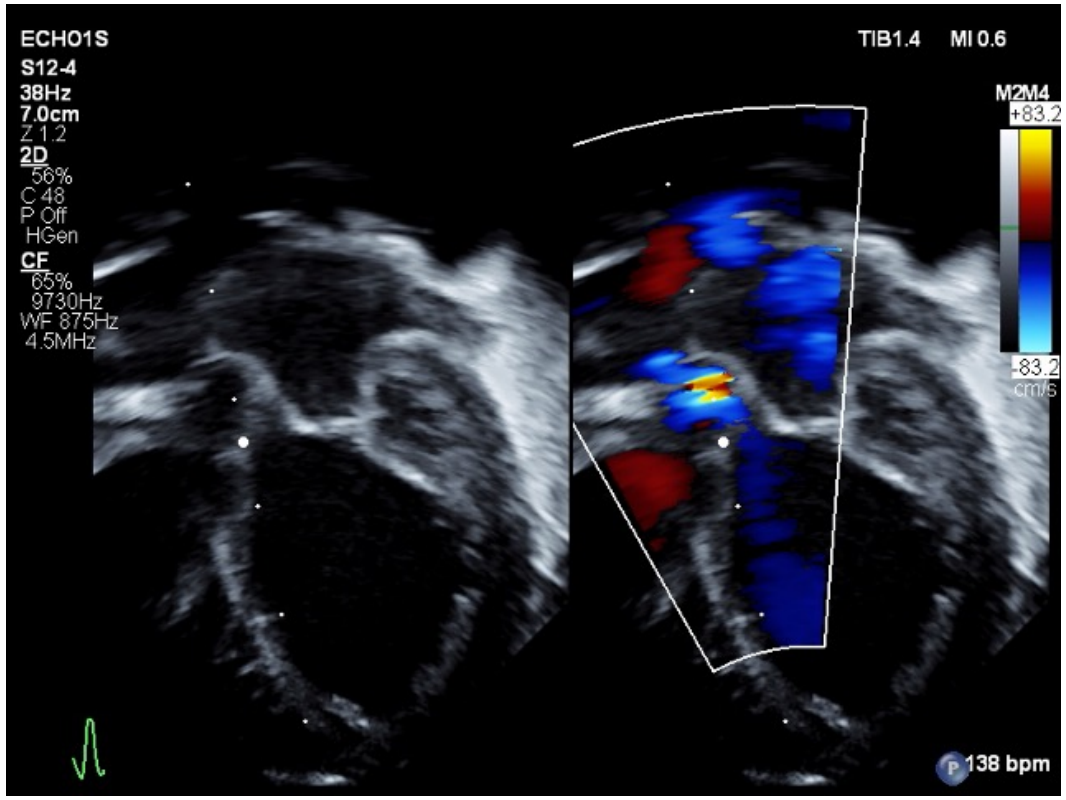
Z < -5.0 or atresia of:
- Ventricle
- AV Valves

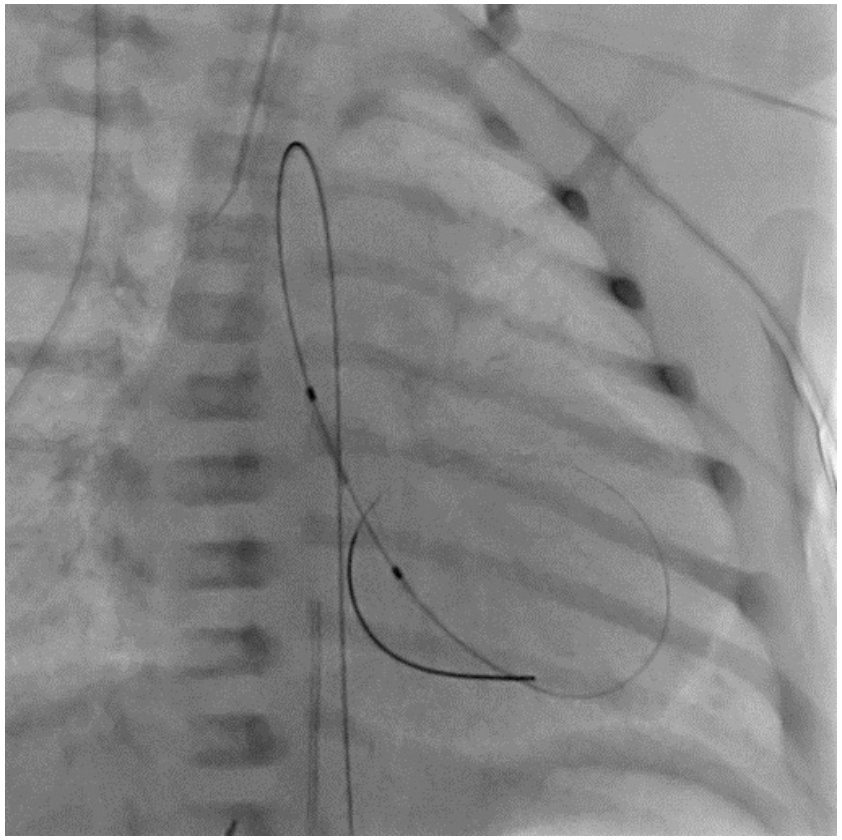
Severe Ventricular dysfunction
Multiple complex lesions

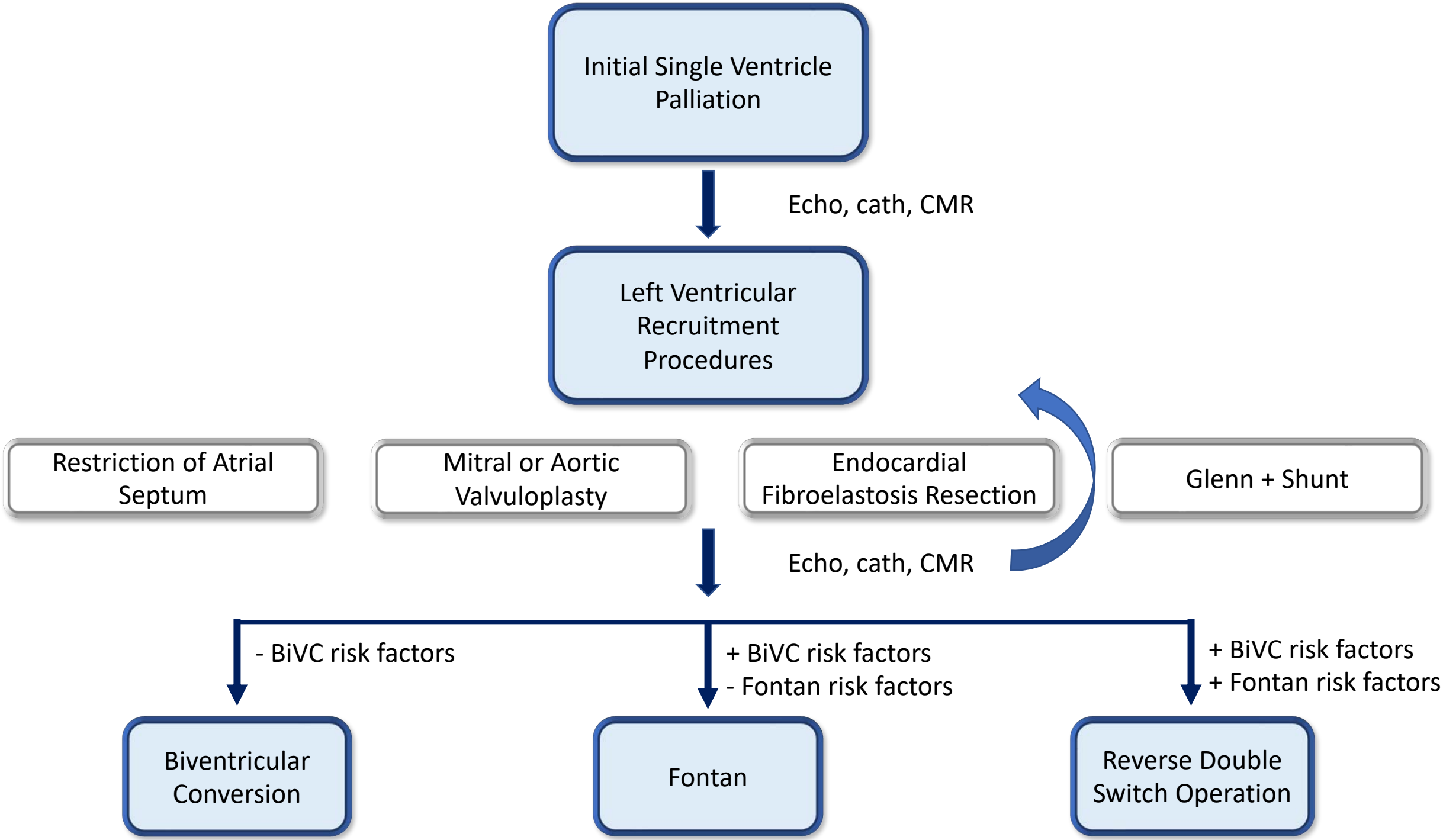


Single Ventricle

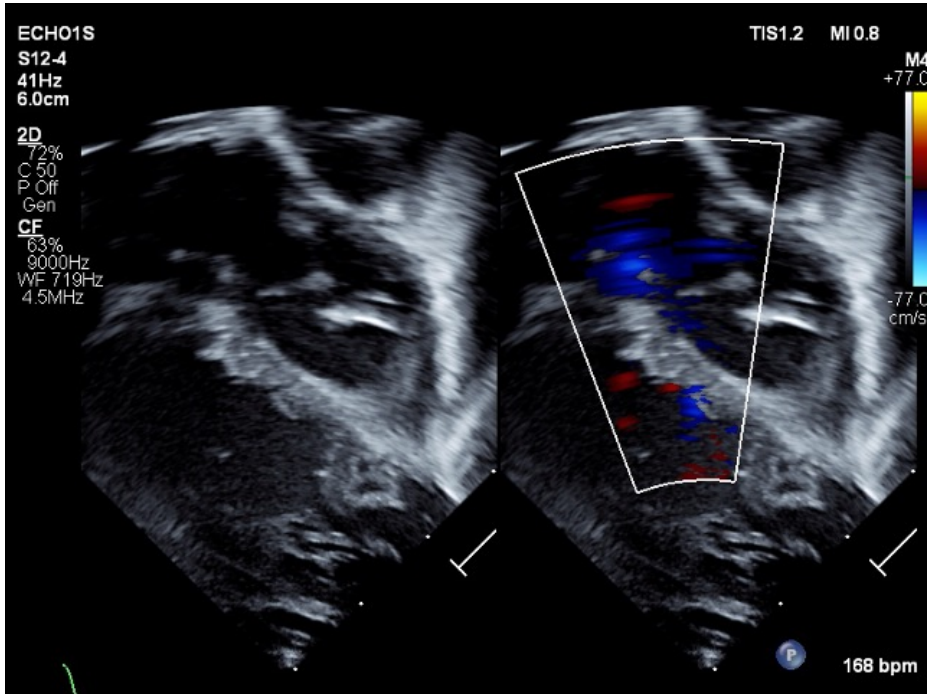
HLHS / borderline LV

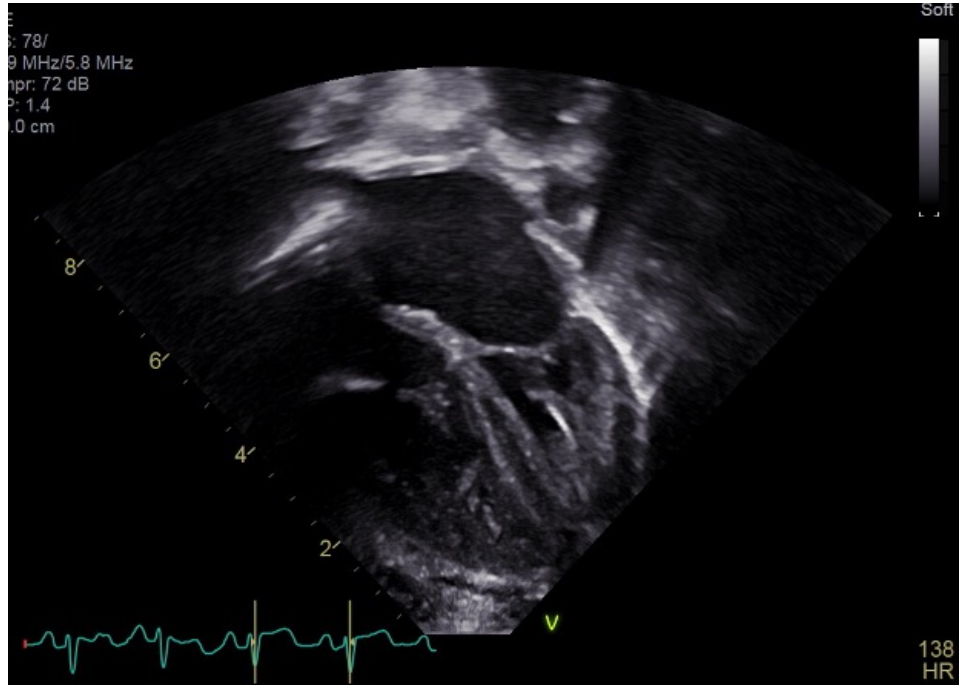





Initial Single Ventricle Palliation
Echo, cath, CMR
Left Ventricular Recruitment Procedures
Restriction of Atrial Septum
Mitral or Aortic Valvuloplasty
Endocardial Fibroelastosis Resection
Glenn + Shunt
Echo, cath, CMR
- BiVC risk factors
+ BiVC risk factors
- Fontan risk factors
+ BiVC risk factors
+ Fontan risk factors
Biventricular Conversion
Fontan
Reverse Double Switch Operation

4 months later





Ventricular Recruitment and Biventricular Conversion after Single Ventricle Palliation in Hypoplastic Left Heart Syndrome

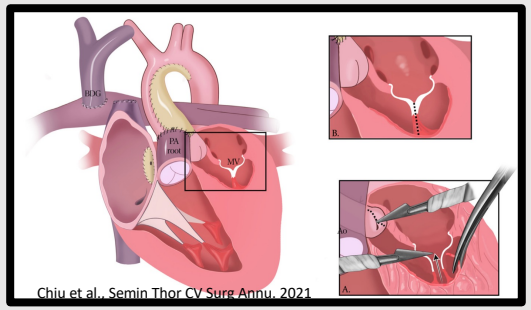
Nicholas Oh, MD, Ilias Doulamis, MD, Mossab Saeed, MD, Alvise Guariento, MD, Breanna Piekarski, RN, Pedro del Nido, MD, Sitaram Emani, MD

Boston Children's Hospital
Where the world comes for answers

HARVARD MEDICAL SCHOOL

Background

Current management of hypoplastic left heart syndrome (HLHS) predominantly involves single ventricle palliation (SVP). However, SVP is associated with morbidity and mortality due to the deranged physiology. An alternative management strategy may be converting HLHS patients from a single ventricle into a biventricular circulation. Recruitment procedures may be performed as an adjunct to promote left ventricular growth prior to biventricular conversion (BVC).



Chiu et al., Semin Thor CV Surg Annu, 2021

Objectives

Our objectives were to:

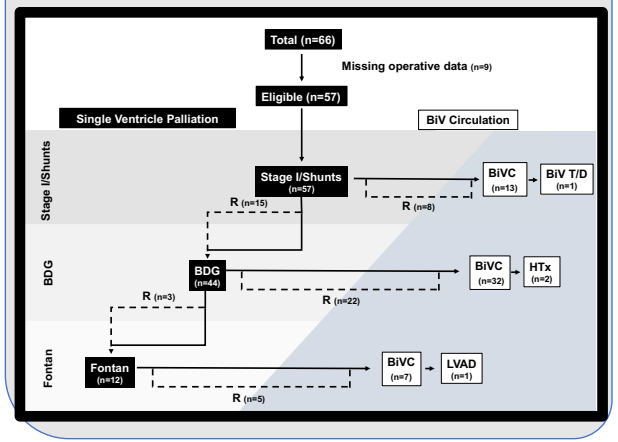
1) Review institutional experience with BVC after SVP in HLHS
2) Review the impact of recruitment procedures prior to BVC on growth of hypoplastic ventricles
3) Compare outcomes of SVP and BVC in HLHS

Methods

This is a single institution, retrospective cohort study of HLHS patients who underwent SVP and subsequent BVC with and without recruitment procedures. Mortality, unplanned reintervention rates, and imaging measurements were analyzed.

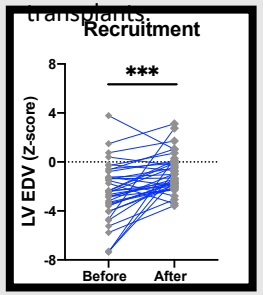
Results

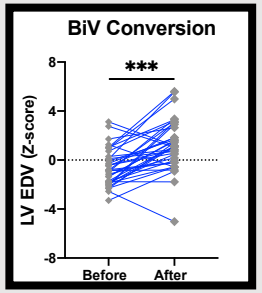
Between 2001 and 2021, 57 patients with HLHS underwent BIVC after SVP. 72% (41/57) underwent recruitment procedures, which includes fenestrated ASD (40%), mitral valve repair (80%), aortic valve repair (72%), and endocardial fibroelastosis (EFE) resection (51%). Median age at BVC was 3.4 years old. BVC led to increased LVEDV z-scores (-0.7 (1.5) vs. 1.2 (2.1)), *p<.001*. After recruitment, LVEDV z-scores improved from -2.5 (2.4) to -0.9 (1.5), *p <.001*.

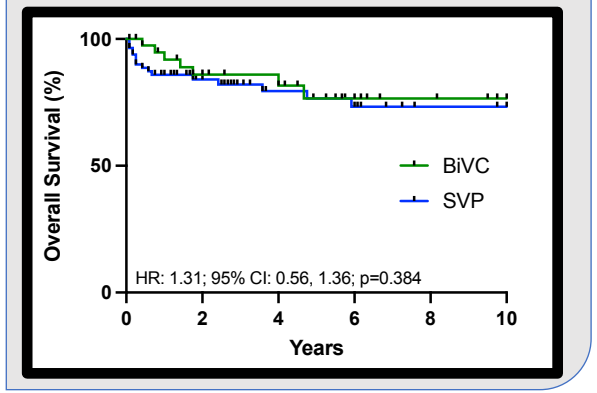


Results (cont.)

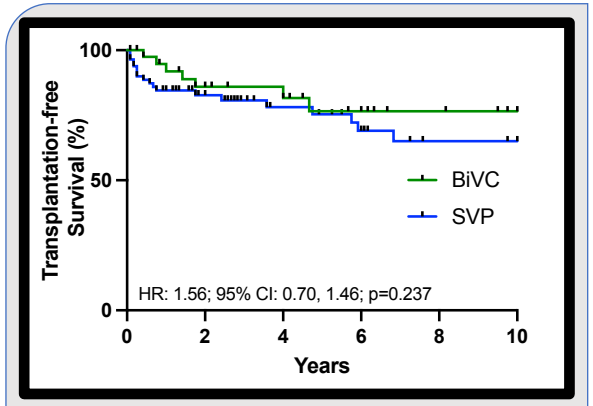
EFE resection with recruitment was associated with LVEDV growth, though statistical significance was not established (LVEDV score 1.2 (1.2) vs. 2.1 (2.07)). Median follow-up time was 4.0 years. Overall, we report no 30-day mortality after biventricular conversion. 10-year survival was 76% (CI: 56-88%). We compared outcomes with 97 HLHS patients who were treated by SVP. We found overall survival and transplant free survival to be similar. Among all BVC patients, 1 patient returned to SVP, 1 proceeded to an LVAD, and 2 had heart transplants.







Results (cont.)



Univariable analysis demonstrated that BVC after Fontan was a risk factor for mortality (HR 5.53, 95% CI: 1.23 – 24.93, *p=.026*).

Lessons Learned

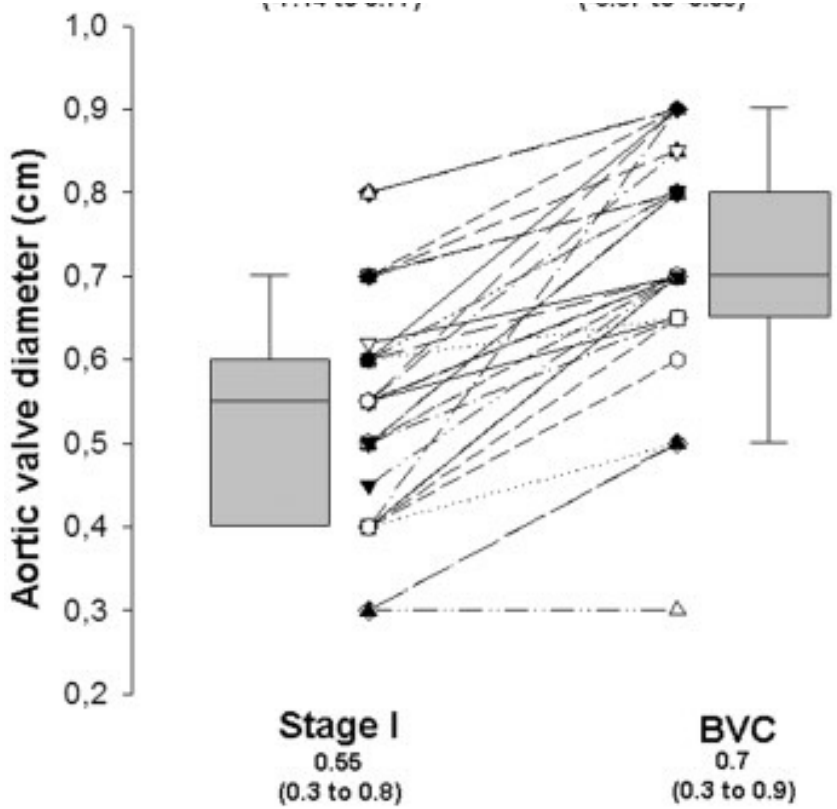
BVC from SVP is an alternative strategy to manage HLHS patients with acceptable mortality.

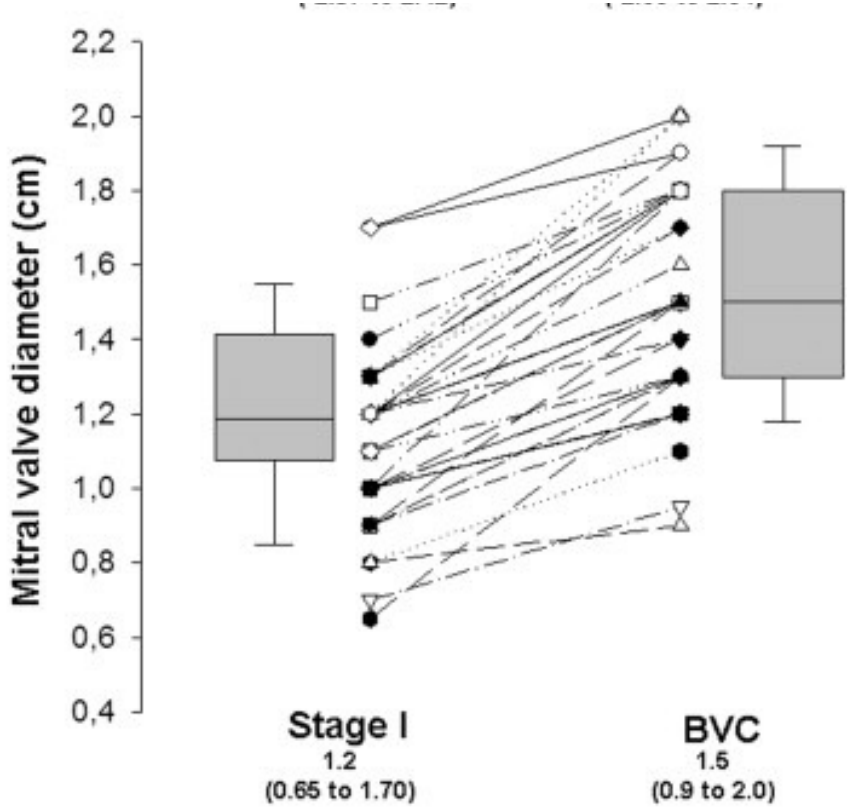
BVC and Ventricular recruitment allows for statistically significant growth of hypoplastic ventricles.

Acknowledgements

Acknowledgement to the Kaplan Endowed Fellowship for funding, and Dimitrios Poutias for administrative responsibilities.

Ventricles can grow…

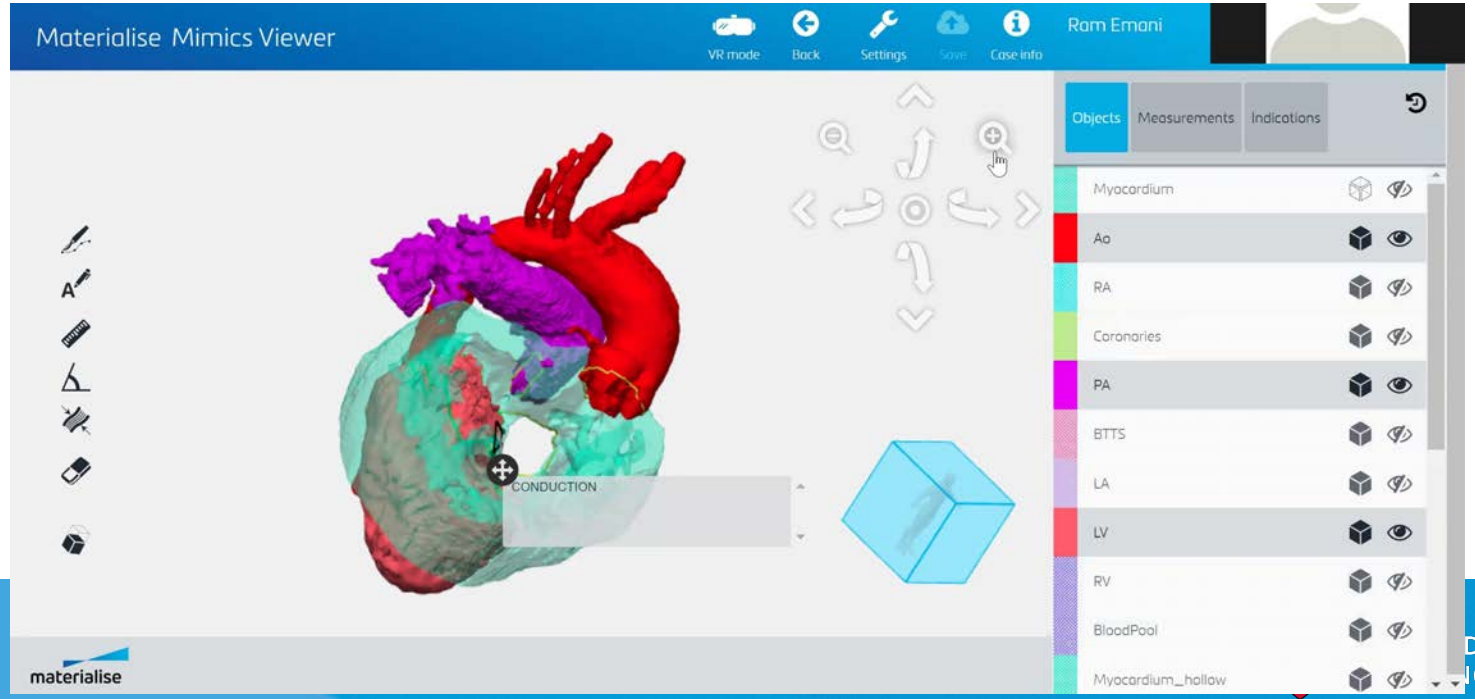




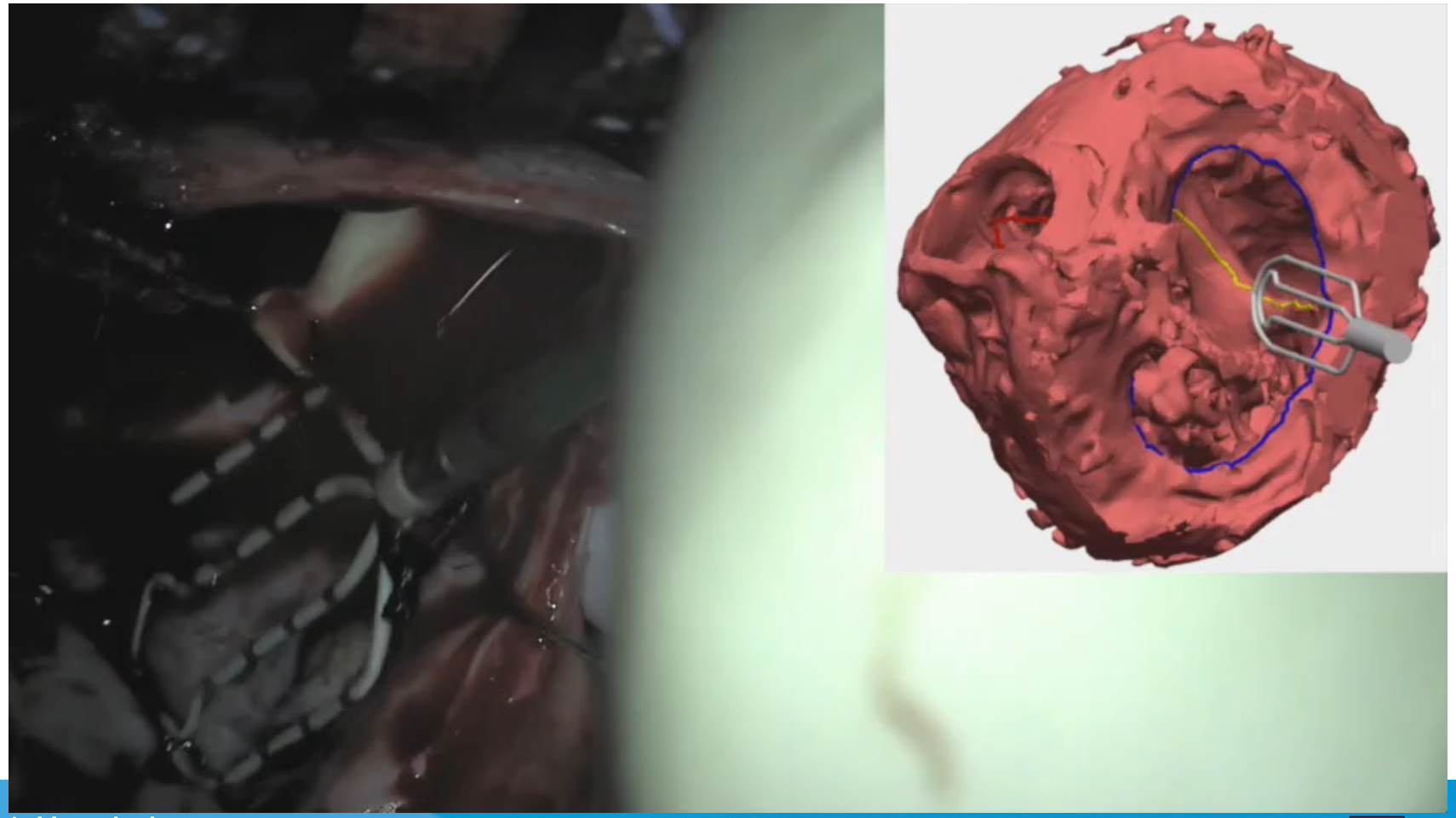
Boston Children's Hospital
Heart Center

Yerebakan et al. JTCVS 149(4)2015, Pages 1112-1122.e2

3D Models for surgical planning



Intraoperative conduction mapping



BiV preferred

Anatomic

Pulm artery hypoplasia

Physiologic

Pulm vein stenosis

Pulm Hypertension

RV dysfunction

AV valve regurg

Airway / Lung abnormalities

Non-sinus rhythm

Non-cardiac

Genetic abnormalities

SVP preferred

Cardiac

Valvular atresia

Severe hypoplasia of LV

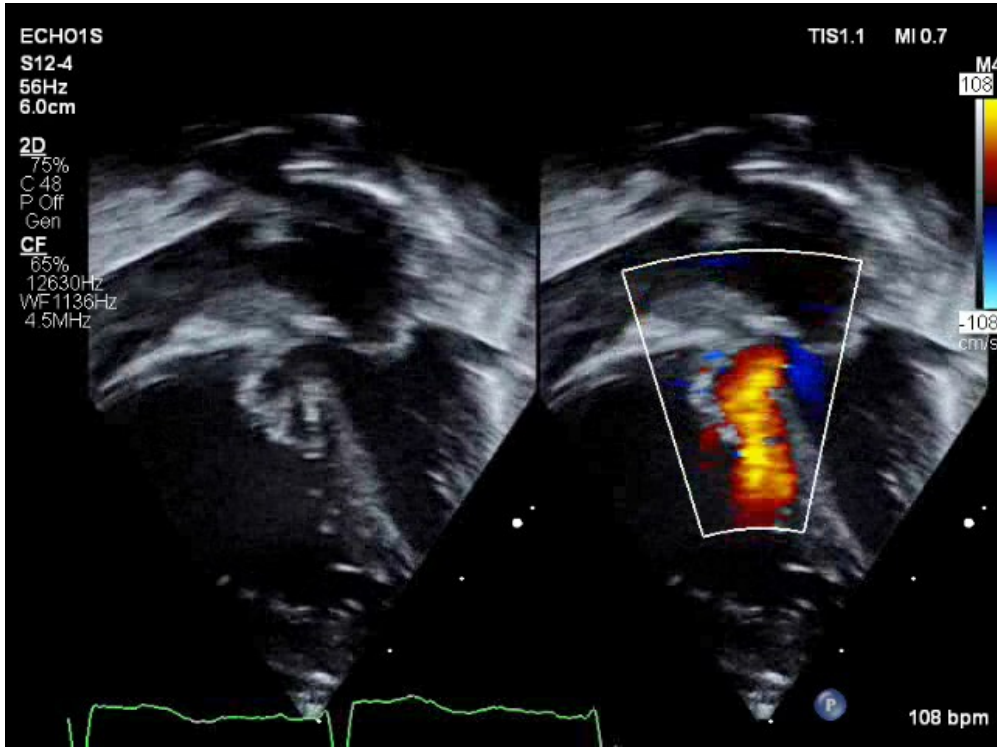
Single LV with no risk factors

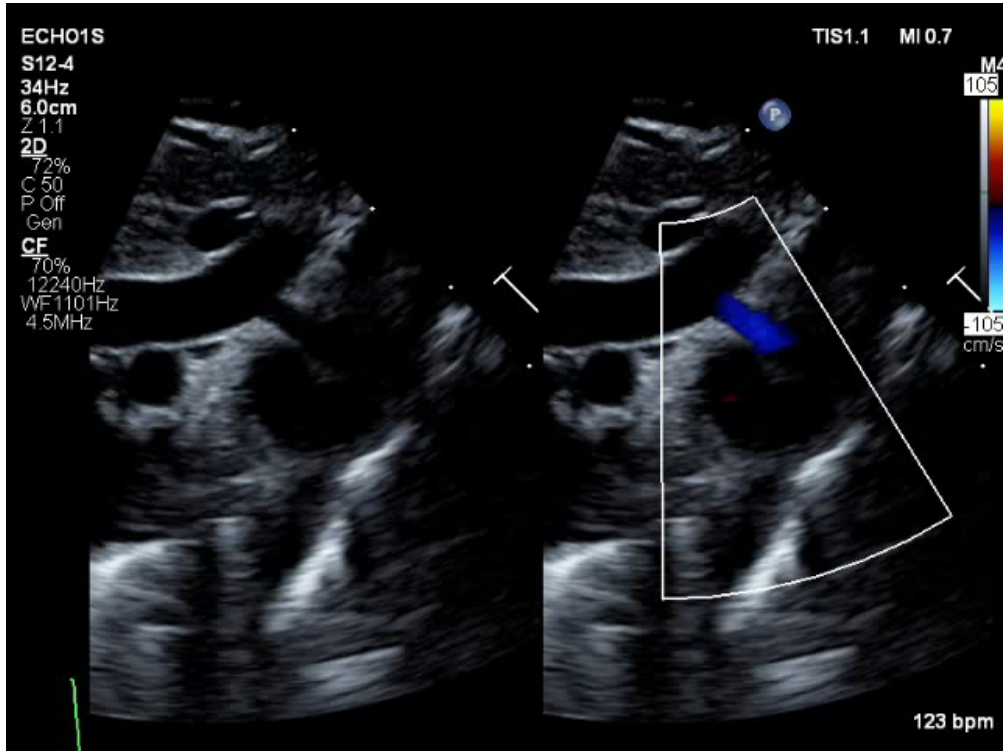
Non-cardiac

Institutional experience with BiV

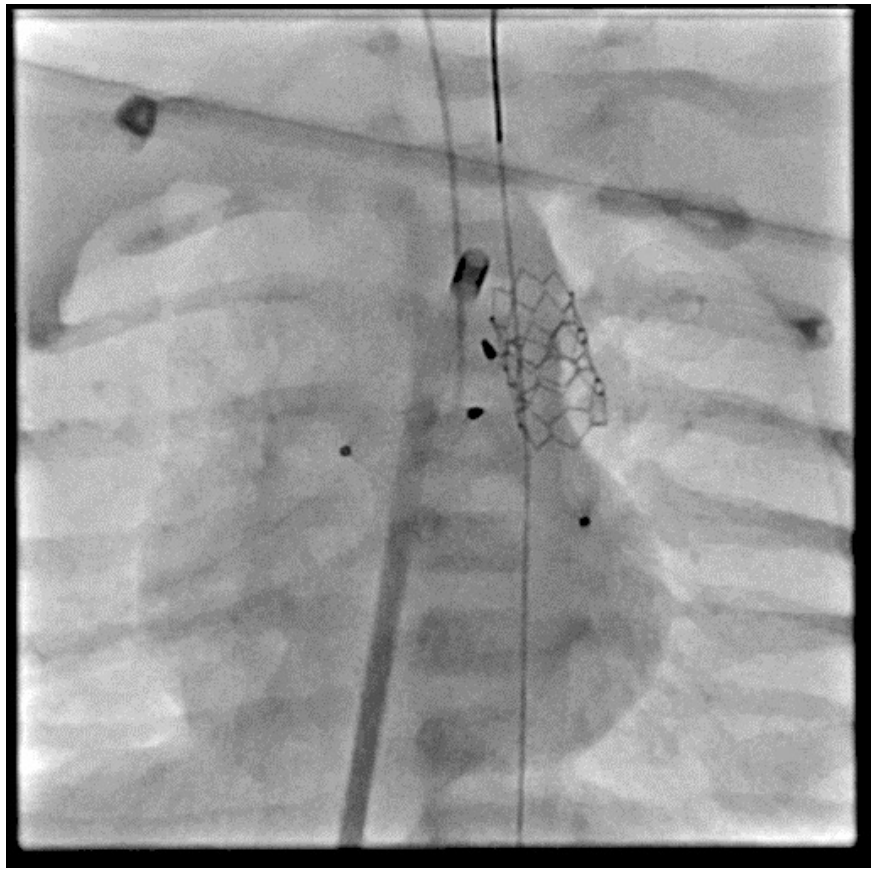
Infrastructure

What is a borderline LV?



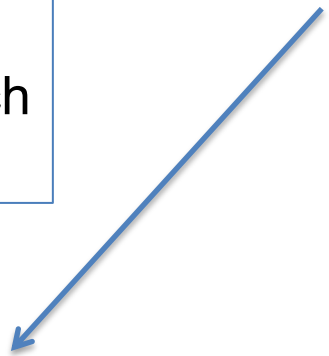


Total transcatheter stage 1



Neonate on PGE

Less than 2/3 apex
Retrograde flow in arch
EFE



Initial Stage 1

Hybrid vs. traditional

Neonate on PGE

Less than 2/3 apex
Retrograde flow in arch
EFE

Apex forming LV
Antegrade flow in arch
No EFE

Initial Stage 1

Hybrid vs. traditional

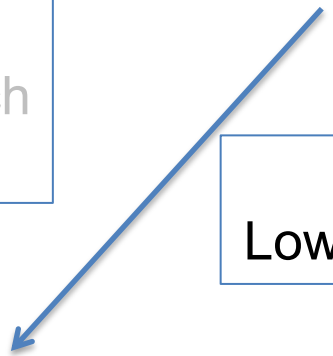
Signs of LA hypertension

Biventricular Repair

- Aortic valve balloon / off PGE
- Arch repair
- Aortic valvotomy
- Limited Mitral papillary splitting

Neonate on PGE

Less than 2/3 apex
Retrograde flow in arch
EFE



"In-between" anatomy
Low threshold for Initial Stage1

Apex forming LV
Antegrade flow in arch
No EFE

Initial Stage 1

Hybrid vs. traditional