Great Ormond Street Hospital for Children NHS Foundation Trust

Great Ormond Street Hospital

UCL

Borderline Left Ventricle: Does the Size Matter?

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No disclosures

Aim of treatment:

best possible quality of life at longest lifetime

Critical AS: main considerations:

- **Biventricular circulation**:
  - Balloon valvuloplasty, Ross(Konno)
  - Mitral valve repair/ replacement (Melody/Prosthetic)
  - AO valve repair/replacement
  - Homograft replacements $^{(n-1)}$
  - EFE resection/LV rehabilitation
  - Postcapillary hypertension (LVAD/HTx / H+LTx)

- **Uni-ventricular circulation (Fontan)**:
  - Norwood I, BCPA, TCPC

- **Heart transplant**:
  - Later HTx (10y-survival 85% at GOSH), re-HTx?
  - Availability (no neonatal donors in Europe)

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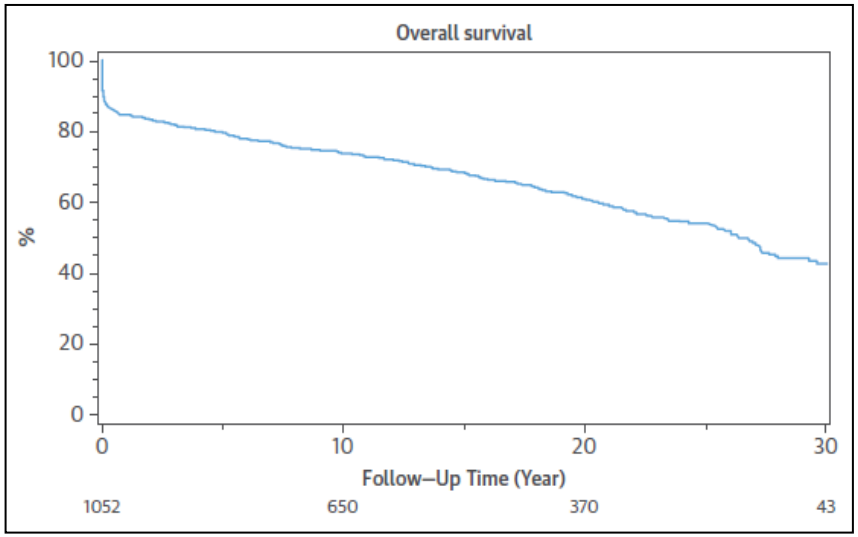
40-Year Follow-Up After the Fontan Operation

Long-Term Outcomes of 1,052 Patients (Mayo experience)

TABLE 1 Patient Demographics (N = 1,052)

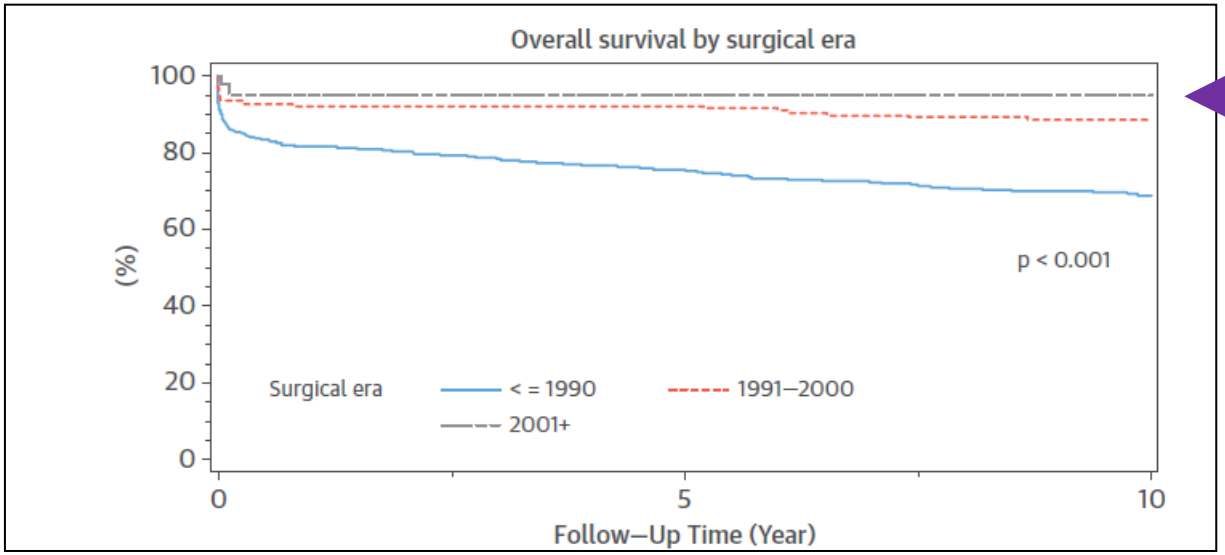
Male	637 (61)
Type of Fontan operation	
Atriopulmonary connection	616 (59)
Lateral tunnel	262 (25)
Extracardiac conduit	120 (11)
Other	54 (5)



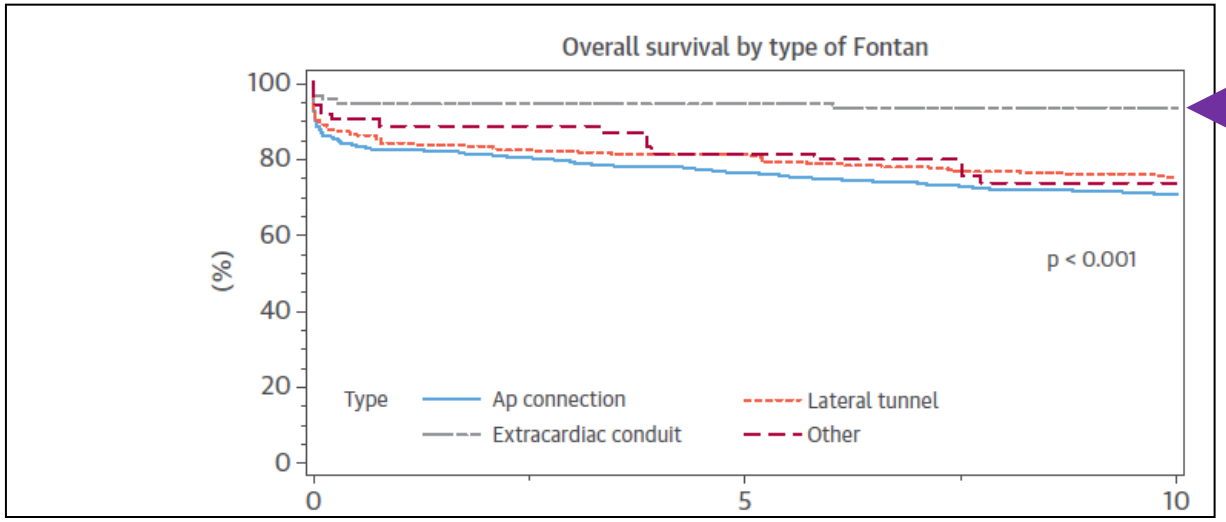
Pundi KN, JACC 2015

40-Year Follow-Up After the Fontan Operation

Long-Term Outcomes of 1,052 Patients (Mayo experience)



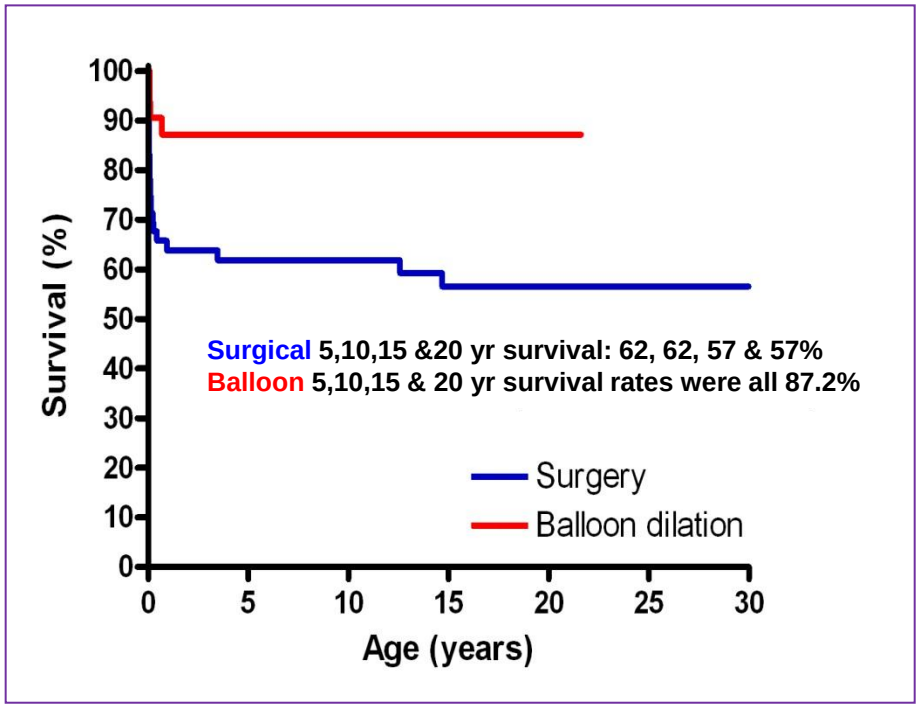
TCPC Extracardiac Conduit

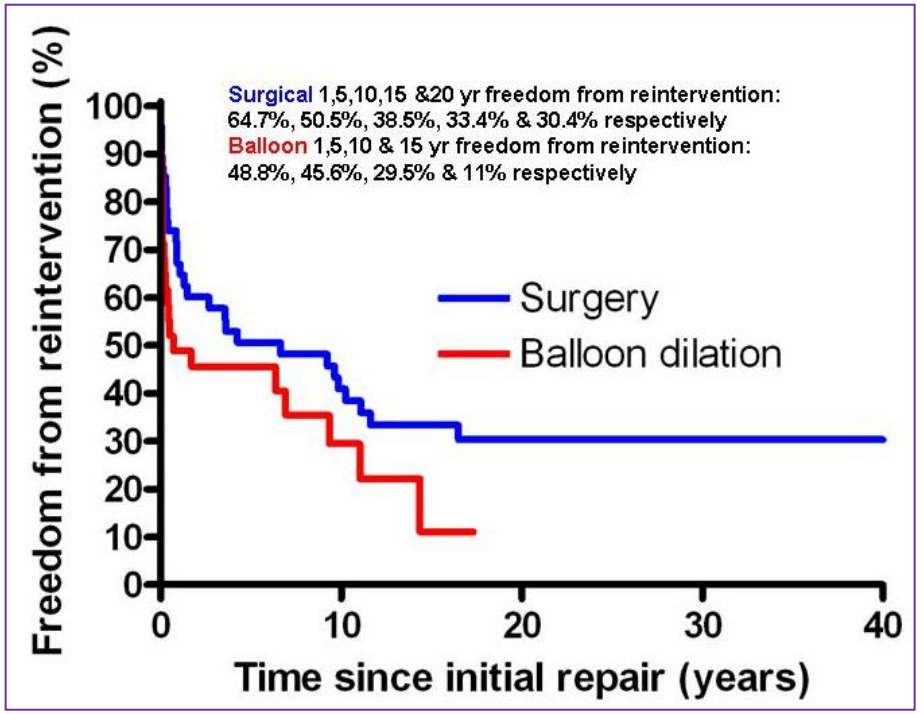


Pundi KN, JACC 2015

Critical Aortic Stenosis: 40y follow up

(GOSH, N=96, 1970-2010)





Carr M, Bonello B, Marek J. 2018

Fetal Aortic Valvuloplasty for Evolving Hypoplastic Left Heart Syndrome: Postnatal Outcomes of the First 100 Patients

Lindsay R. Freud, MD[1], Doff B. McElhinney, MD[1], Audrey C. Marshall, MD[1], Gerald R. Marx, MD[1], Kevin G. Friedman, MD[1], Pedro J. del Nido, MD[2], Sitaram M. Emani, MD[2], Terra Lafranchi, NP-C[1], Virginia Silva, RN[3], Louise E. Wilkins-Haug, MD, PhD[3], Carol B. Benson, MD[4], James E. Lock, MD[1], and Wayne Tworetzky, MD[1]

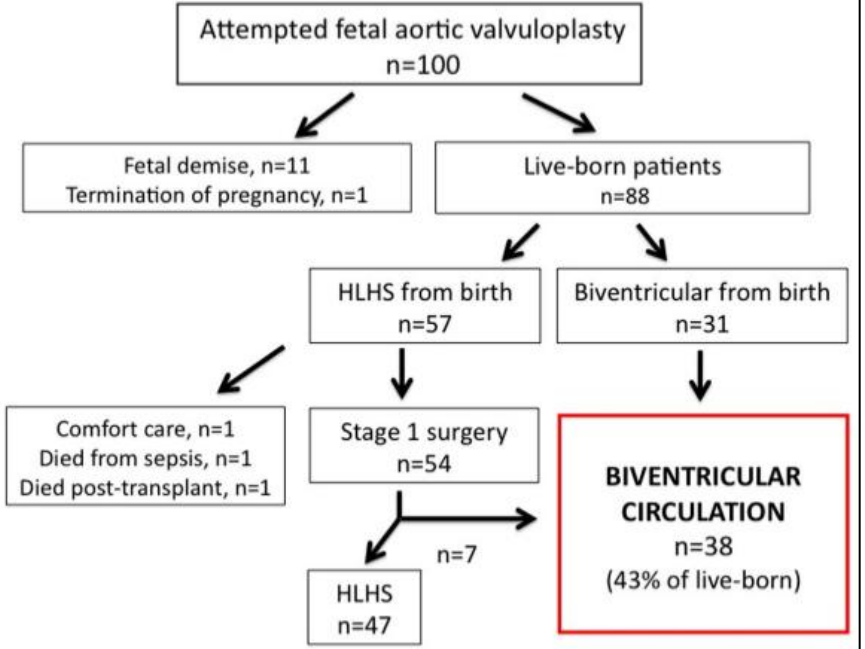
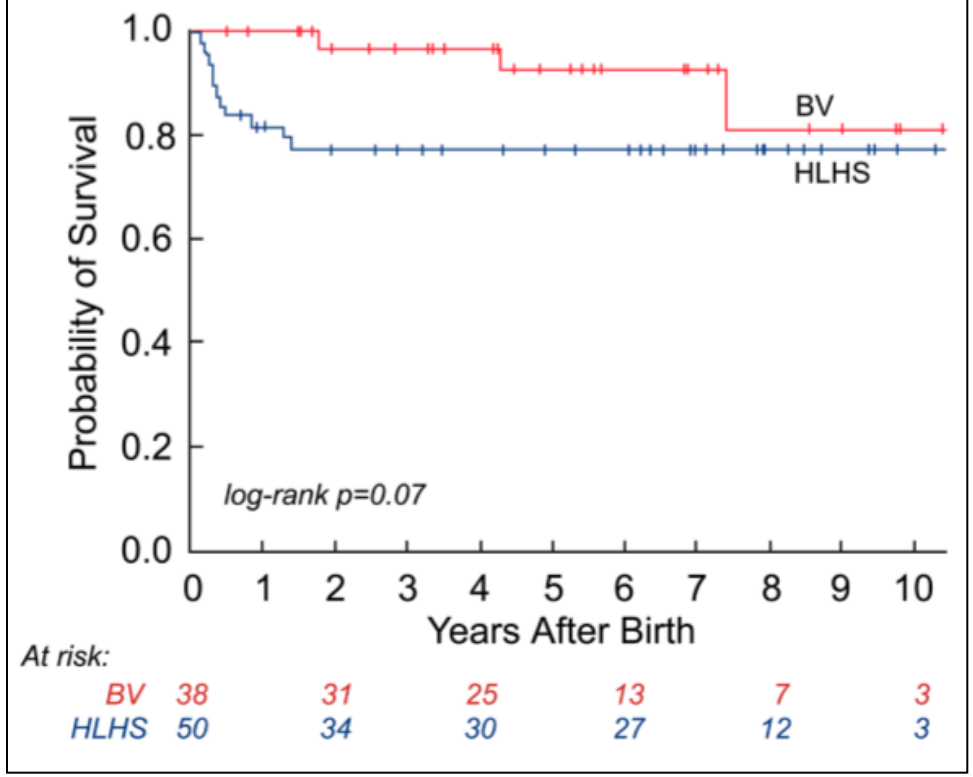
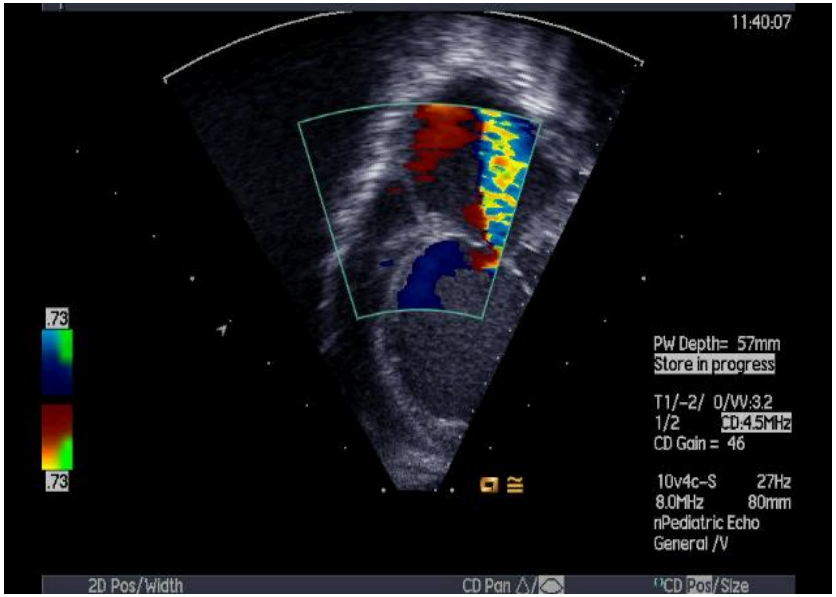


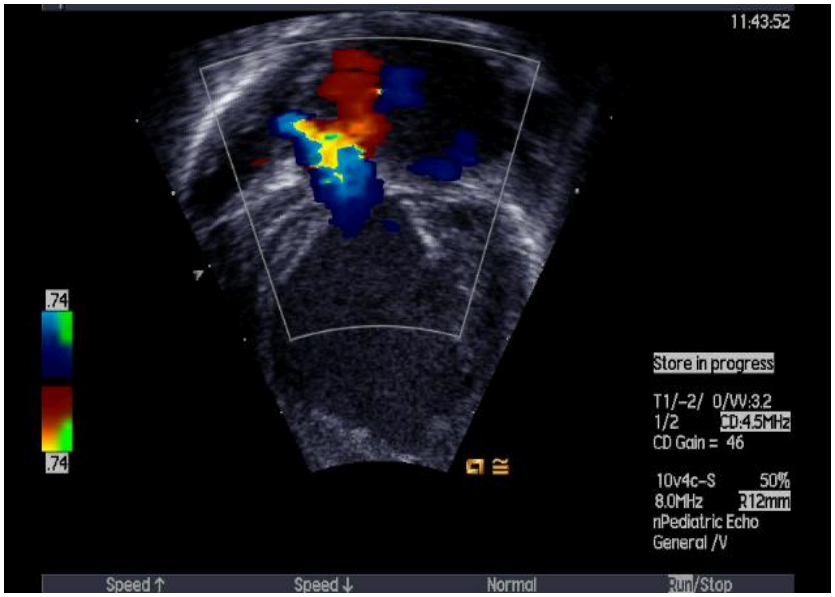
Figure 2.
Flow diagram summarizing postnatal management and outcomes for the entire 100 patient cohort. (HLHS=hypoplastic left heart syndrome)

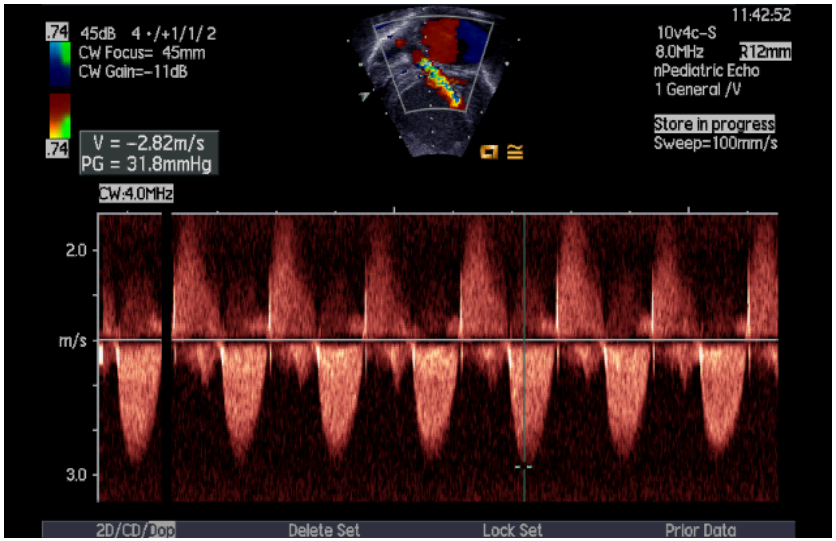


Freud LR, Circulation 2014

Fetal intrauterine balloon aortic valvuloplasty:
Postnatal neonatal outcome







- Postnatal cardiogenic shock
- Unable to stabilise circulation
- Not for Ross / Konno

Cardiogenic shock in infancy

Cardiac aetiology

- Reduced diastolic filling:

  PFC / PPH, TAPVC

- Reduced ejection fraction:

  AS, COA

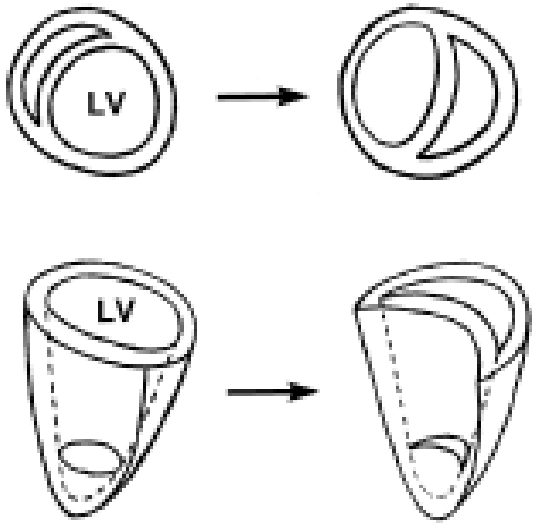
- Combination of both:

  HLHS, AS, HLV, COA, EFE

SMALL LEFT VENTRICLE
Primarily or secondarily small?

Small left ventricle in overloaded (pressure, volume) right ventricle is primarily not only due to compression; rather is due to underfilling

- **TAPVC**
- **Isolated COA**
- **PPH/PFC**



LV can stretch more than anticipated (elastic stocking vs. plastic bag)

Even "hypoplastic" LV cavity < 15-20 ml/m$_2$ can expand to adequate volume

Phoon CK,Am Coll Cardiol, 1997

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

Initially intended BIV (116) - 5yr survival rate 70%

Initially UNIV (179) - 5yr survival rate 60%

Independent risk factors associated with greater survival benefit for BIV versus UNIV:

- Age
- Aortic valve Z-score
- LV length
- Endocardial fibroelastosis
- Tricuspid regurgitation
- Ascendent aorta

Multivariable hazard model

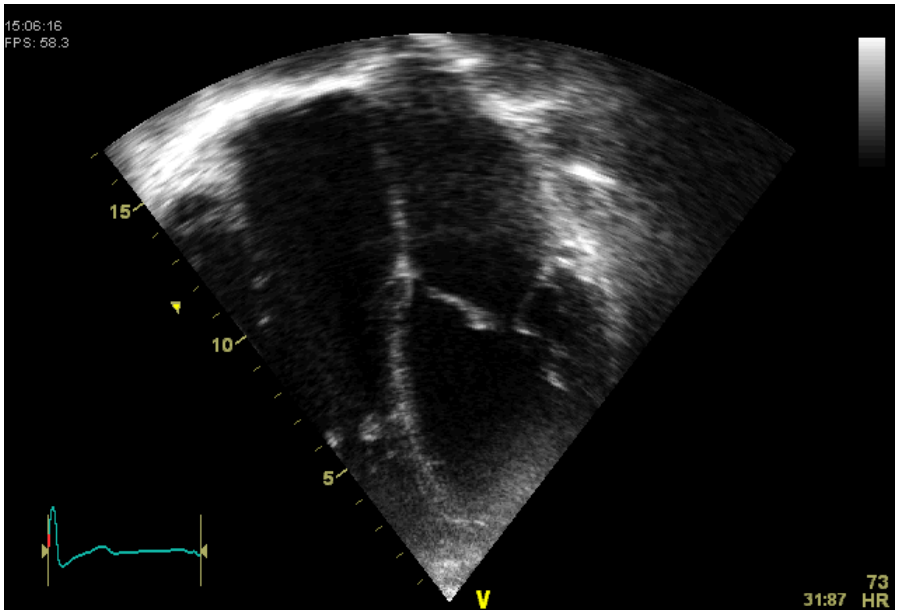
Lofland GK, JTCVS 2001, Hickey EJ, JTCVS 2007

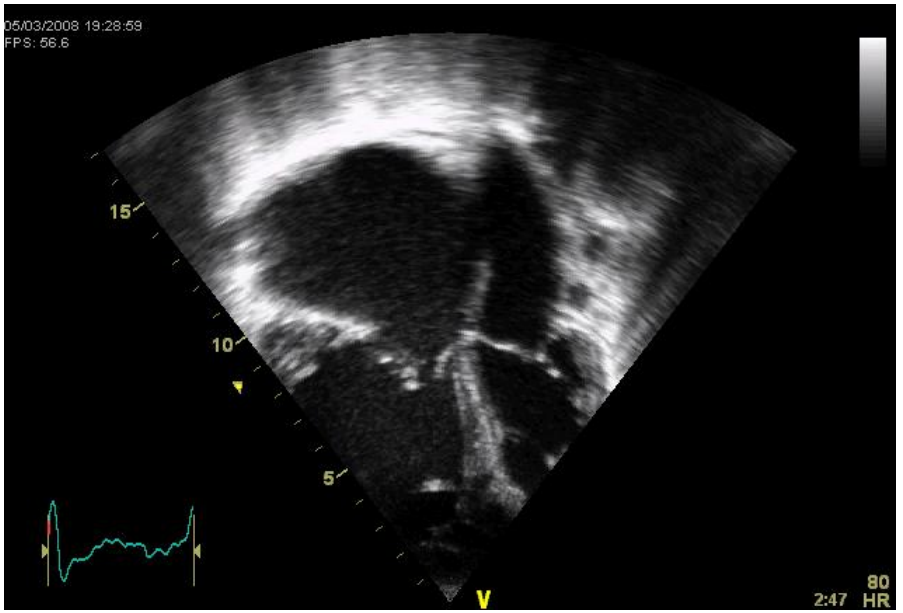
http://www.ctsnet.org/aortic_stenosis_calc/ ← **Calculator**

Critical AS: Late outcome

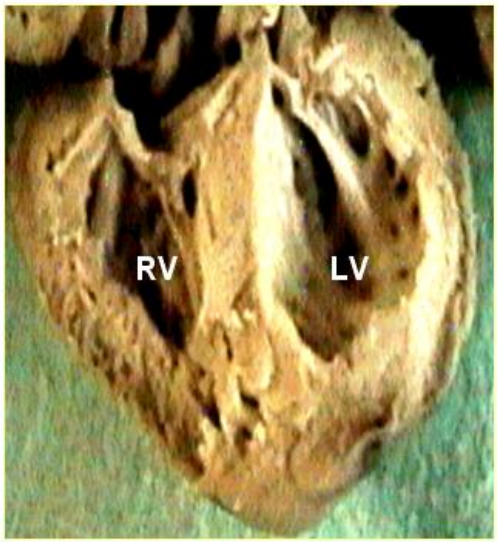
TT, 15 yrs
Critical AS
S/P valvotomy (2d)+ECMO
S/P Balloon VPL (6yrs)
Ross-Konno (at 9yrs)
PPVI (15yrs) – post: PA wedge 23 mmHg

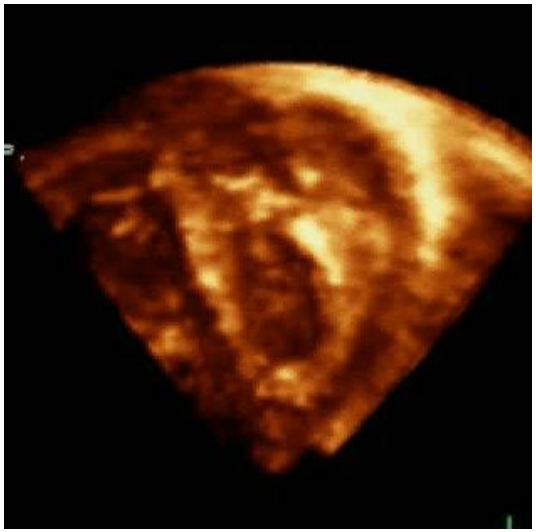
YM, 15 yrs
Critical AS
S/P valvotomy (3d)
S/P Ross (at 3yrs)
Progressive PH, Stroke (12yrs)
PPVI (13yrs) – post:
MPA 75/34/36 Ao 80/42/56
Waiting list H+LTx
Died (15yrs)





Endocardial fibroelastosis: ECHO





Neither endocardial echo-brightness nor geometric variables of the left ventricle allow for accurate prediction of the presence of endocardial fibroelastosis.

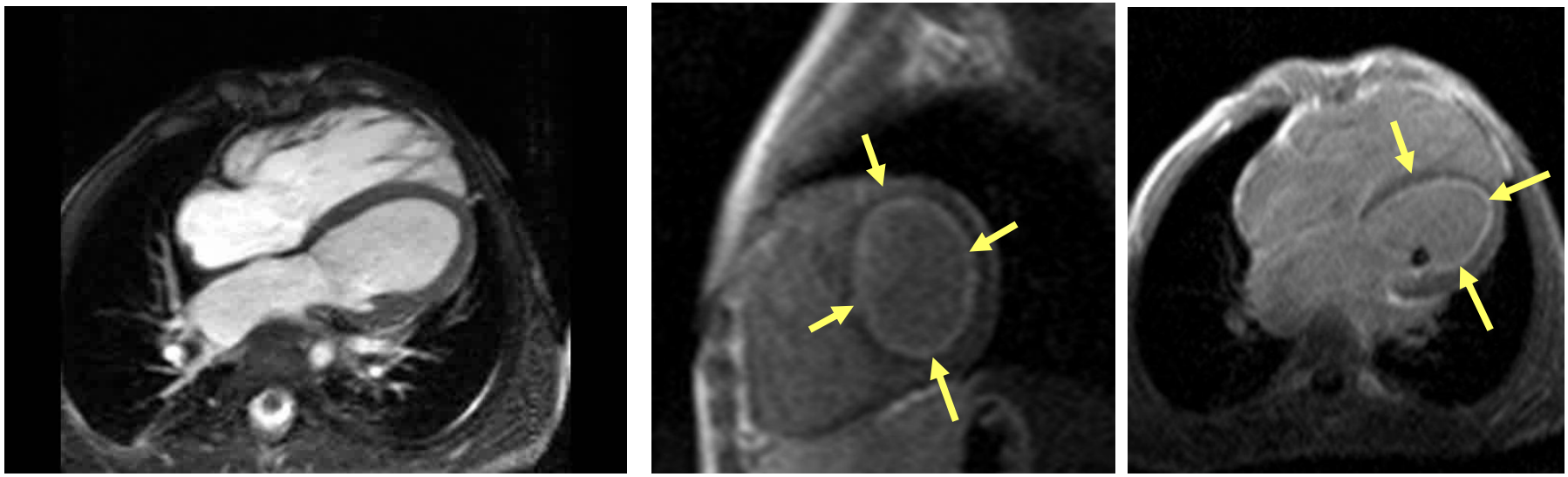
Tworetzky W, Am J Cardiol 2005

Mahle WT, Am J Cardiol 1998

Colan SD, JACC 2006

McElhinney DB, Circulation 2009

Endocardial fibroelastosis: MRI

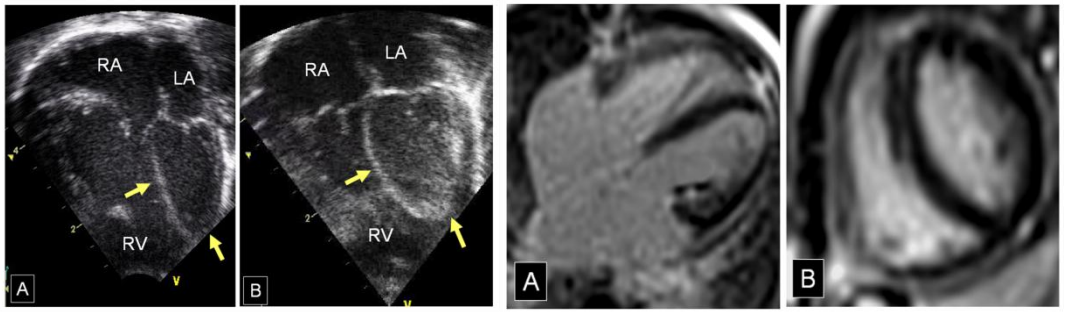


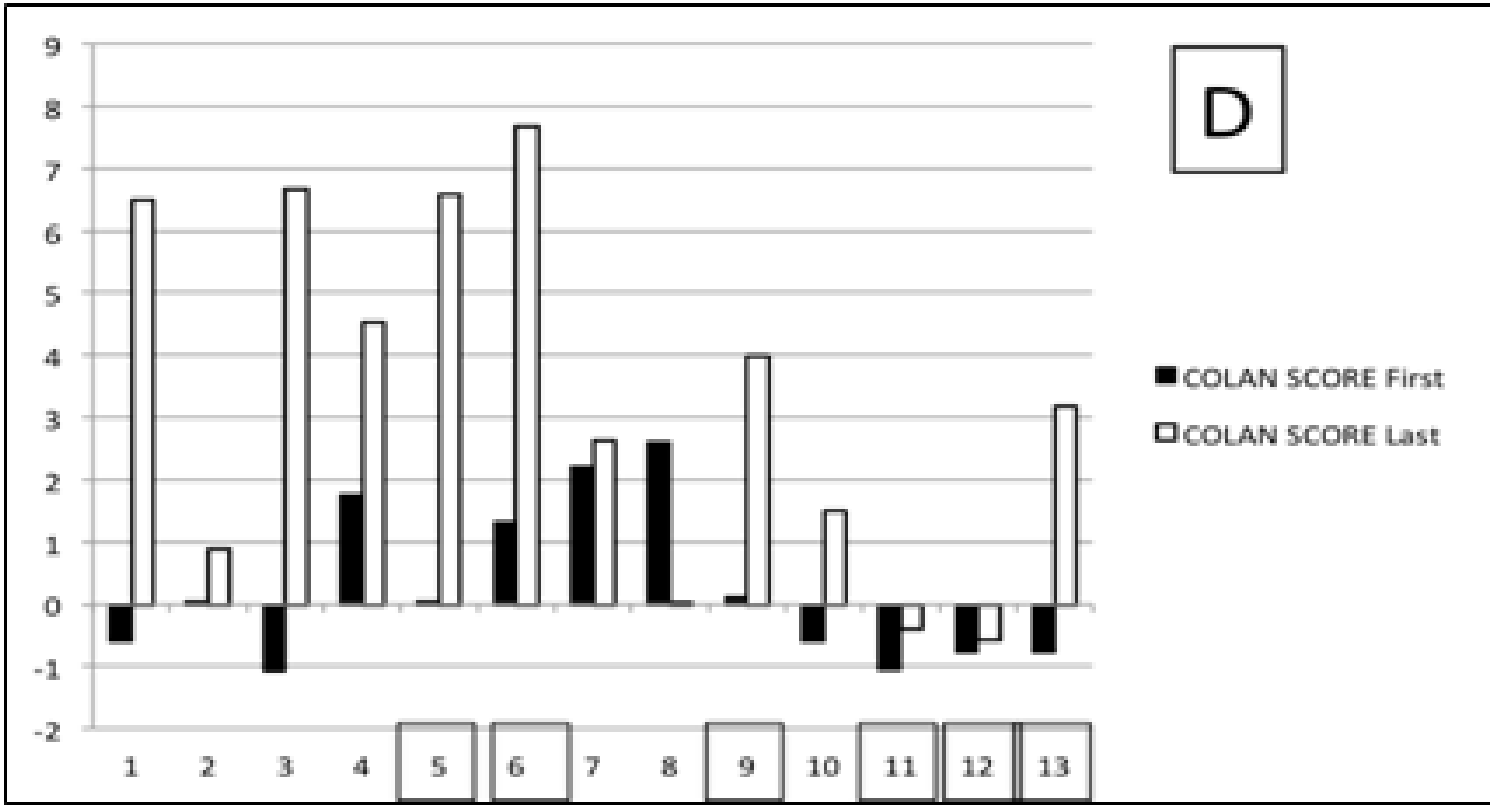
Late gadolinium enhancement:

- Inversely related to HR
- Contraindicated in LCO (nephrotoxicity)

Obstructive Left Heart Disease in Neonates With a "Borderline" Left Ventricle: Diagnostic Challenges to Choosing the Best Outcome

Giulia Tuo · Sachin Khambadkone · Oliver Tann · Martin Kostolny · Graham Derrick · Victor Tsang · Ian Sullivan · Jan Marek





Ped Cardiol 2013

Endocardial fibroelastosis resection

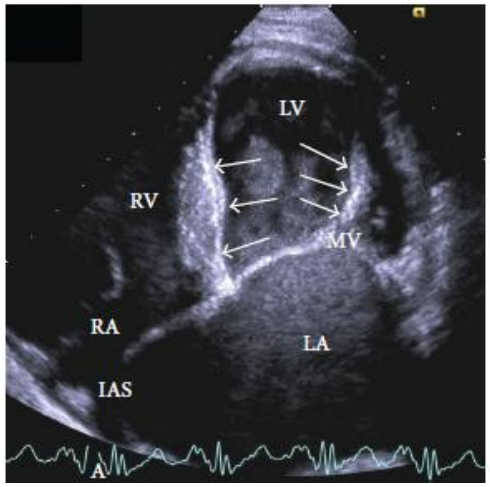
- No operative mortality, median follow-up
  25 months (6m - 10y)
- 1 death from noncardiac causes, 2 reoperations
  (1 x MVR, 1 x AO + MV repair)
- 3 pts. recurrent EFE

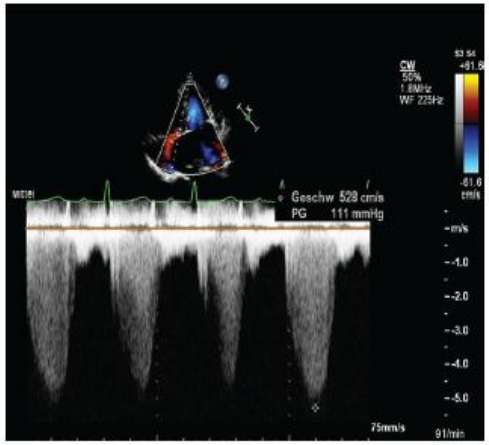
	Preop	Recent	p
LVEDV Z-score	-0.2±1.7	2.7 ±1.8	<0.01
RV:LV SP ratio	0.78 ±0.36	0.32 ±0.11	<0.05
LAp (mmHg)	28 ±6.3	11 ±2.4	<0.01
EF (%)	36 ±12	58 ±10	<0.01
AO grad (mmHg)	39 ±22	28 ±19	NS

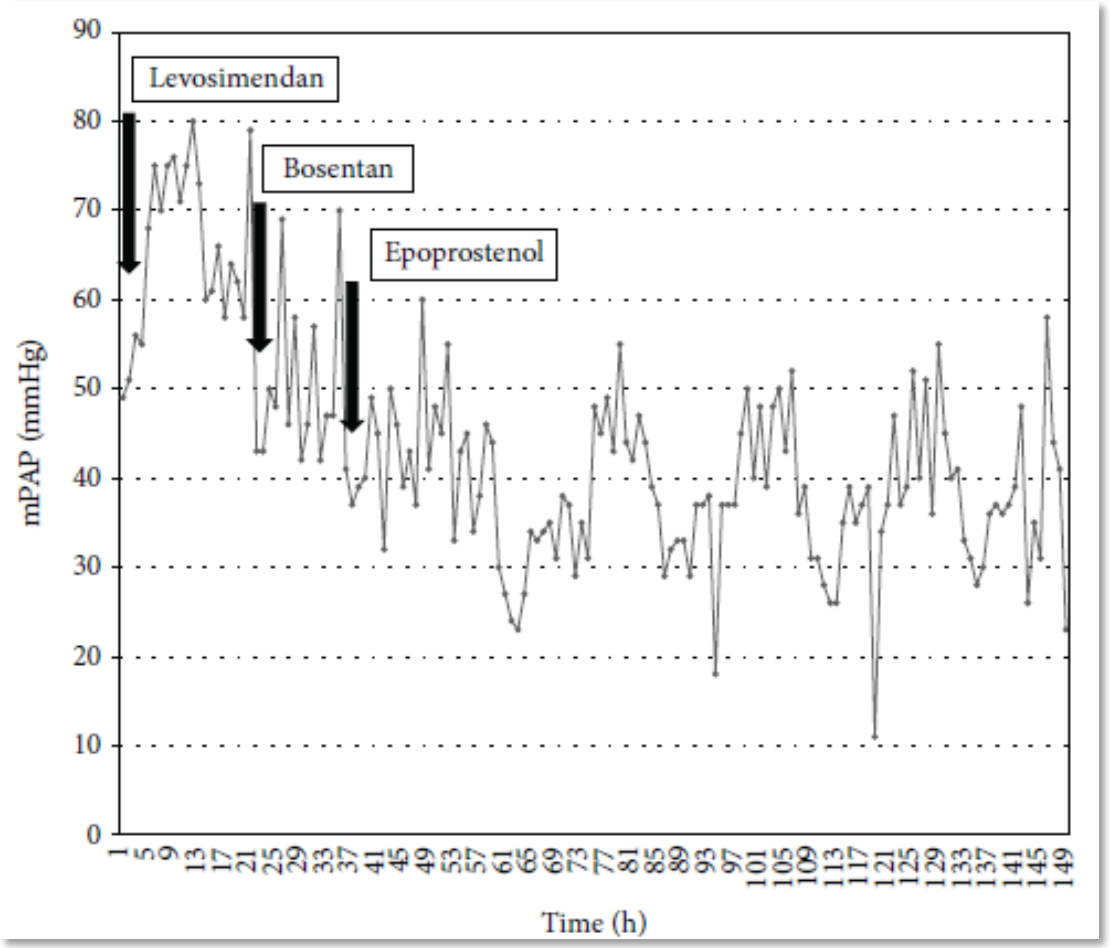
Emani SM, JTCS (in print)

LV Mechanical assist device as **Bridge - to - HTx**

Heart Transplantation in a 14-Year-Old Boy in the Presence of Severe Out-of-Proportion Pulmonary Hypertension due to Restrictive Left Heart Disease: A Case Report







Schwienbacher M, Case Reports in Cardiology 2013

Critical AS: Late outcome

LV Mechanical assist device as **Bridge - to - HTx**

- **O2**
- **Epoprostenol (22 ng/kg/min)**
- **Sildenafil (0.5-2.0 mg/kg/4h)**
- **Bosentan (2-4 mg/kg)**
- **Diuretics and anticoagulants**

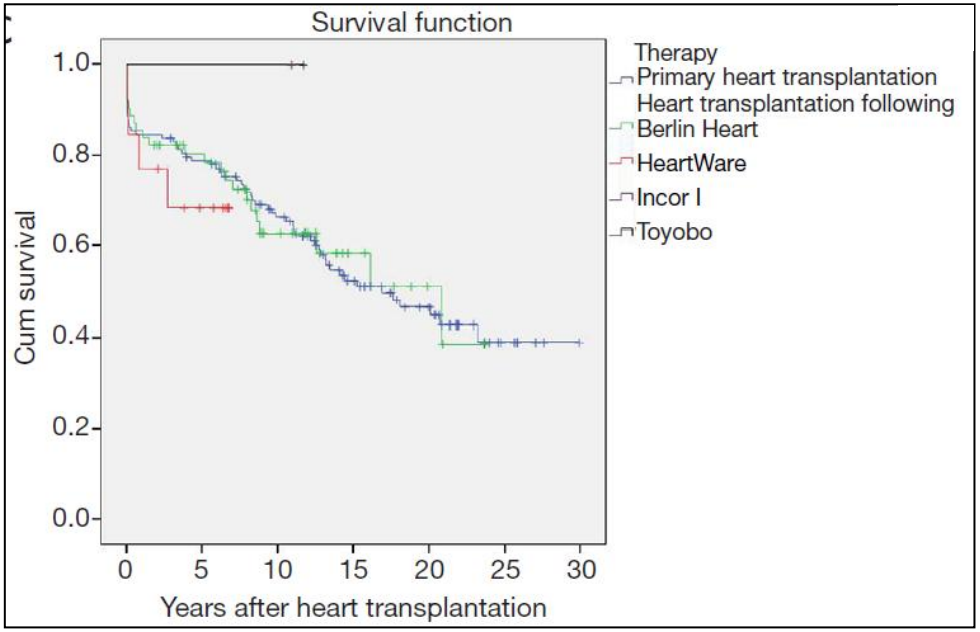
- Unloading the LV + LA hypertension
- Decreases postcapillary wedge pressure
- Induces a decline in the PAP
- Less hypoxia at the tissue level
  - reduces an important stimulant of pulmonary vasoconstriction

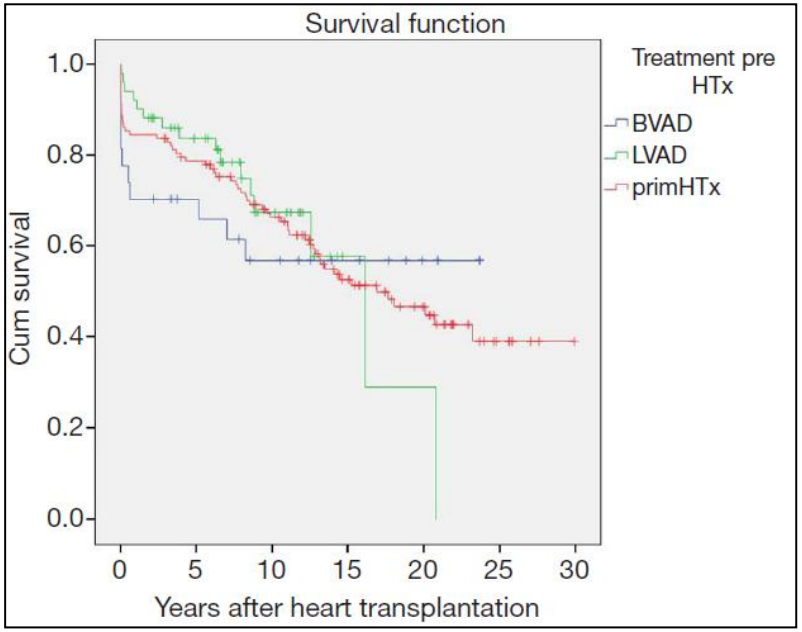
Role of paediatric assist device in bridge to transplant
Berlin, N=78

Diagnosis	
DCMP	56
Myocarditis	14
End-stage CHD	5
Ischemic heart failure	2
Tumour	1
Cardiopulmonary resuscitation	15
ECMO prior to VAD	7

Hetzer R, Ann Cardiothorac Surg 2018

Role of paediatric assist device in bridge to transplant
Berlin, N=78





- VAD implantation in “small” LV?
- Donor availability

Hetzer R, Ann Cardiothorac Surg 2018

Critical Aortic Stenosis

- Myocardial function rather than the size!?
- Role of endocardial fibroelastosis (EFE) and myocardial fibrosis

Critical Aortic Stenosis

Fetal atrial stenting rather than fetal balloon valvuloplasty to offer better Fontan outcome ?

Echocardiography & Prenatal Cardiology

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Jo Wolfenden
Nicola Callaghan

Cardiac Sonographers

Gill Riley
Ian McLeod
Rebecca Banks
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Kalaiiarasi Janagarajan
Jabeen Bibi
Sally Punzal
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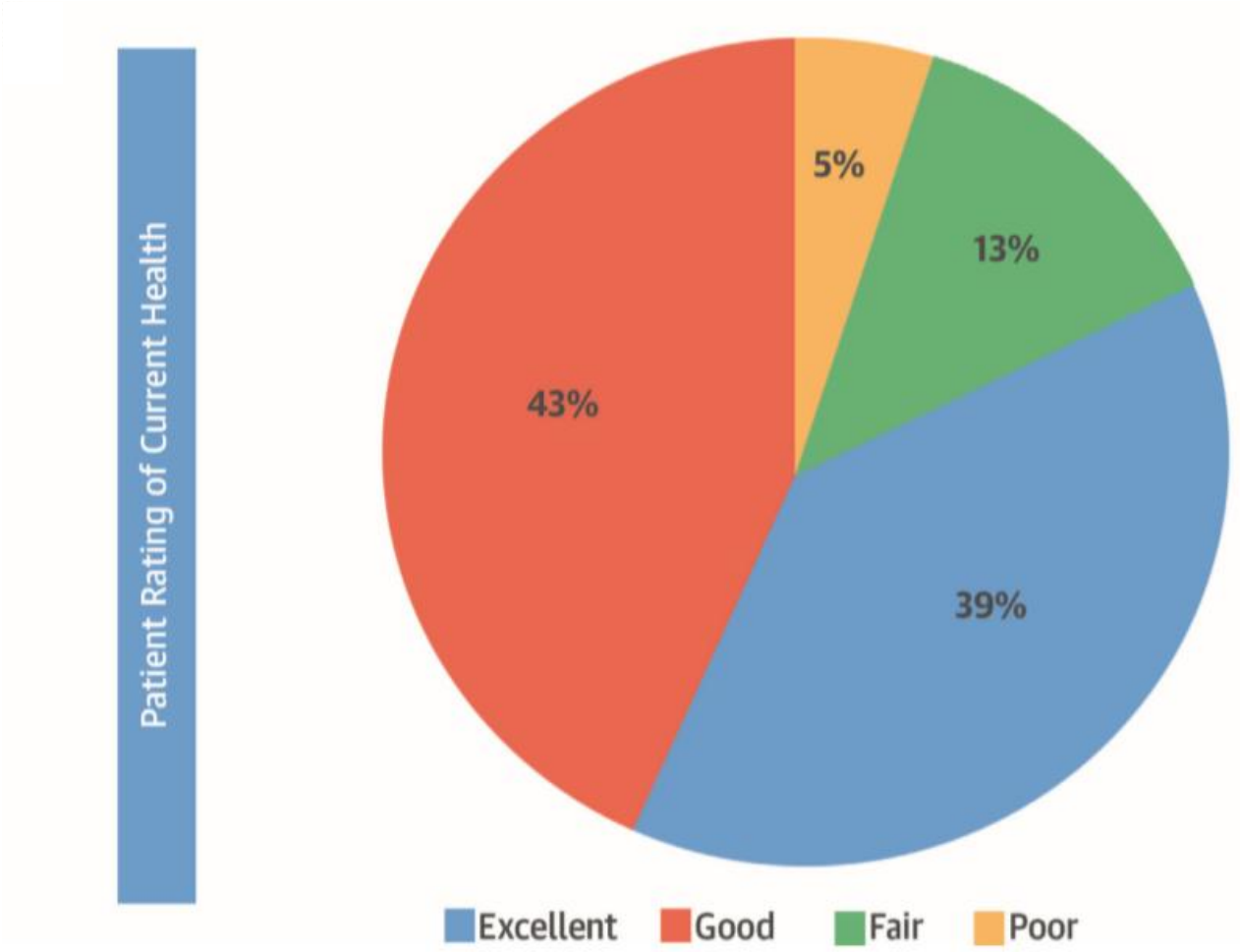
William Regan
Gabrielle Norish

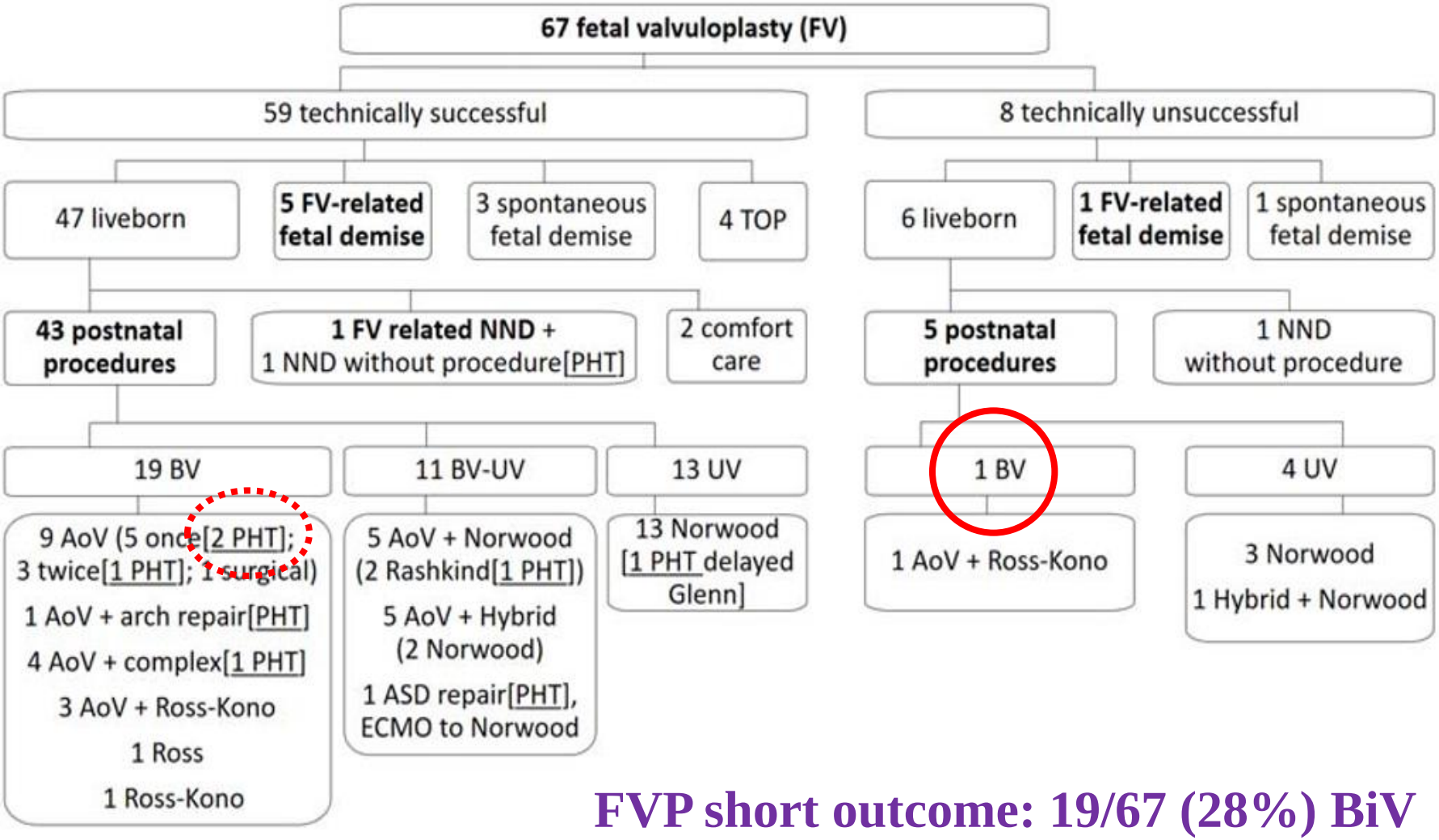
40-Year Follow-Up After the Fontan Operation

Long-Term Outcomes of 1,052 Patients (Mayo experience)



Pundi, K.N. et al. J Am Coll Cardiol. 2015; 66(15):1700-10.

Fetal hemodynamic response to aortic valvuloplasty and postnatal outcome: a European multicenter study



FVP short outcome: 19/67 (28%) BiV

Kovacevic A, Ultrasound Obstet Gynecol 2017