“What is the Definition of Small Systemic Ventricle”

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Contents

- Introduction

✓ Aortic valve stenosis

✓ Aortic coarctation ± hypoplastic aortic arch

✓ Hypoplastic left heart complex

✓ RV pressure and/or volume overload

- Conclusion

Hypoplastic left heart physiology

- Inability of LV to sustain adequate CO following birth because of underdevelopment of one or more left heart structure despite surgical or medical intervention
- Physiologic , not morphologic
- Present within a relatively narrow band of broad continuum of hypoplastic lesion of left heart

Kirklin et al, Cardiac surgery 3rd, 2003

Pre-operative determination can be extremely difficult, particularly in the presence of a Borderline (small) ventricle !!

- Whether LV is adequate to sustain the systemic circulation
- Whether it may became adequate with the available surgical approaches, and therefore a bi-ventricular type of repair is feasible

1-1. Morphometric parameters

- MV diameter and indexed MV area
- LV inflow dimension and cross-sectional area
- Ratio between LV and RV dimension
- LV long axis to heart long axis ratio
- LVEDV, LV mass index
- ratio of the right/left ventricular wall thickness

1-2. Morphometric parameters

- Endocardial fibroelastosis (EFE)
- Cardiac apex not formed by LV
- Diameter of LVOT
- Diameter of AV annulus
- Indexed aortic root

2. Functional parameters

- LV ejection fraction
- LVEDP
- mean PAP
- Direction of the blood flow

  in the ascending Aorta & the level of PDA

Quantitative evaluation allow Scientific approach to the problem

- Tables with normal left ventricular dimensions
- *z*-values, with SD from the normal values

➢ Inadequate to sustain a bi-ventricular type of repair

- indexed MV area < 4.75 cm^2/m^2
- LV inflow dimension <25 mm
- Ratio between the apex-to-base LV & RV <0.8
- Aortic annulus <6 mm

Kirklin et al, Cardiac surgery 3rd, 2003

4 different groups of CHD with VA concordance

- Aortic valve stenosis
- Aortic coarctation, with or without hypoplastic aortic arch
- Hypoplastic left heart complex
- RV pressure and/or volume overload

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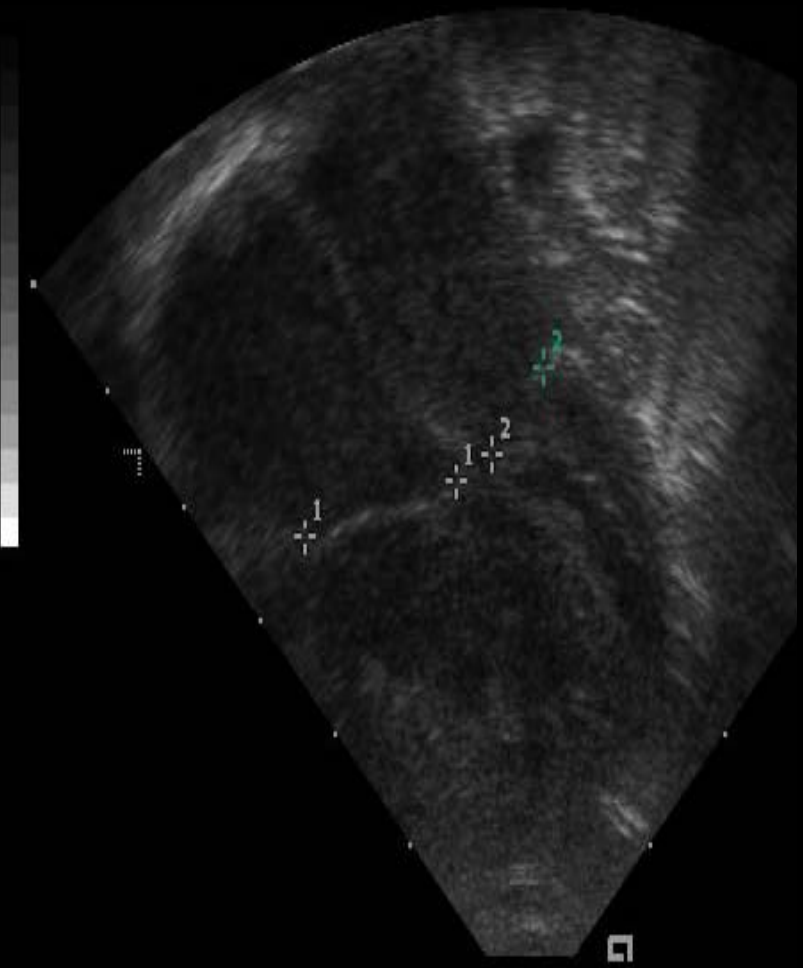
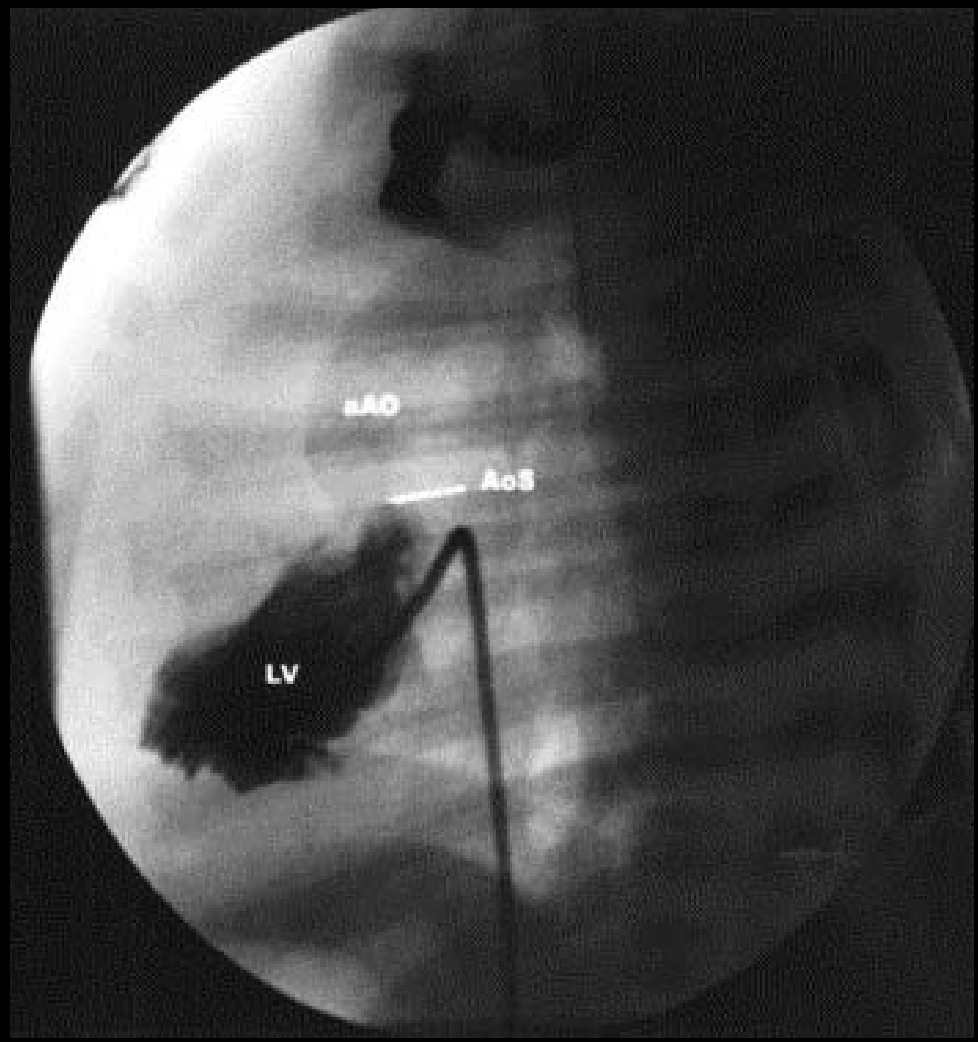
Neonate with AS & small systemic ventricle





Risk factors of Aortic valve stenosis correlated with poor results

- LV cross-sectional area <1.6 cm2, Small (60–80% of normal) and hypoplastic (<60% of normal) LVEDV, indexed LVEDV <20 mL/m2, extensive EFE, LV inflow dimensions <25 mm, diameter of MV <11 mm, small MV orifice <9 mm and diameter of AV <6 mm, LVOT <5 mm, cardiac apex not formed by the left ventricle
- Preop. clinical conditions - severe CHF, cardiomegaly on X-ray and RVH (as possible indication of diminished LV volume or mass)
- LVEDP >20 mmHg and LVEF <40%, Pre-operative mean PAP >50 mmHg

A very extensive retrospective analysis by the Boston group on echo data

- left ventricular long axis to heart long axis ratio <0.8
- indexed aortic root diameter <3.5 cm/m^2
- indexed mitral valve area <4.75 cm^2/m^2
- LV mass index <35 g/m^2

Rhodes, JACC, 1991

Score of Rhodes

- Based on multivariate analysis, the ‘Score of Rhodes’

  =14.0 (BSA)+0.943 (iROOT)+4.78 (LAR)+ 0.157 (iMVA)−12.03

- allowed prediction of death after bi-ventricular type of treatment in the presence of a discriminating Score

  < - 0.35

Rhodes, JACC, 1991

A prospective multi-institutional clinical study: risk factors for hospital mortality by the CHSS

- Predict the best chance for survival
- Bi-ventricular type of repair - younger age, presence and degree of EFE, LV length and lower *z*-value of the aortic valve diameter (only aortic valvotomy ?)
- Uni-ventricular type of repair - presence of moderate or severe TV regurgitation, lower diameter of the ascending aorta

Lofland et al, J Thorc Cardiovasc Surg, 2001

http://www.chssdc.org/

Hemodynamic and morphologic predictors,
John Kovalchin, 1998, JACC

Table 2. Comparison of Hemodynamic Variables for Survival and Nonsurvival in Patients With Two-Ventricle Repair

Variable	All Pts With Two-Ventricle Repair: Survival (n = 14)	All Pts With Two-Ventricle Repair: Nonsurvival (n = 5)	p Value	Norwood Group: All Pts (n = 9)
Ascending aortic flow				
Antegrade	13	2		4
Retrograde	1	3	0.01	5
Transverse arch flow				
Antegrade	12	2		1
Retrograde	2	3	0.046	8
Ductus arteriosus flow				
Antegrade	5	1		0
Retrograde	6*	4	0.31	9
Aortic valve Doppler gradient (mm Hg)	52 ± 23	27 ± 17	0.06	44 ± 19
LVEF	0.27 ± 0.12	0.19 ± 0.16	0.28	0.16 ± 0.07

Hemodynamic and morphologic predictors

John Kovalchin, JACC, 1998

Table 3. Comparison of Morphometric Variables for Survival and Nonsurvival in Patients With Two-Ventricle Repair

Variable	All Pts With Two-Ventricle Repair			Norwood Group: All Pts (n = 9)
	Survival (n = 14)	Nonsurvival (n = 5)	p Value	
Age at initial procedure (days)	15.5 ± 22.1	1.4 ± 0.9	0.18	6.6 ± 6.0
Body surface area (m^2)	0.23 ± 0.03	0.24 ± 0.03	0.71	0.22 ± 0.03
Indexed aortic annulus (cm/m^2)	2.9 ± 0.5	1.9 ± 0.3	0.0002	2.0 ± 0.4
Indexed aortic root (cm/m^2)	3.6 ± 0.5	2.6 ± 0.6	0.003	2.9 ± 0.7
Indexed ascending aorta (cm/m^2)	3.5 ± 0.7	2.5 ± 0.3	0.008	2.4 ± 0.3
Indexed LV long axis (cm/m^2)	13.2 ± 2.5	10.0 ± 0.8	0.01	8.7 ± 2.1
Indexed transverse arch (cm/m^2)	2.3 ± 0.3	2.0 ± 0.3	0.06	2.2 ± 0.3
Indexed aortic isthmus (cm/m^2)	1.9 ± 0.4	1.8 ± 0.3	0.78	1.8 ± 0.2
Relative LV long axis	0.9 ± 0.1	0.9 ± 0.1	0.44	0.7 ± 0.2
Indexed LV EDV (cm^3/m^2)	42.6 ± 26.8	21.8 ± 7.2	0.11	14.5 ± 11.0
Indexed LV ESV (cm^3/m^2)	33.1 ± 24.5	18.2 ± 8.6	0.21	12.3 ± 9.8
Indexed LV mass (g/m^2)	85.4 ± 30.1	88.3 ± 41.3	0.88	53.9 ± 18.1
Indexed MV area (cm^2/m^2)	6.3 ± 2.1	4.8 ± 0.8	0.14	4.3 ± 0.7
Relative MV area	0.6 ± 0.2	0.5 ± 0.1	0.14	0.4 ± 0.1

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COA ± hypoplastic aortic arch

- Main difference between critical AS and COA (even indexed LVEDV 10 mL/m2 and cardiac apex not formed by LV)

  " Predictable growth of LV"

- Morphometric parameters like the score of Rhodes should be used with caution in lesions with systemic obstructions

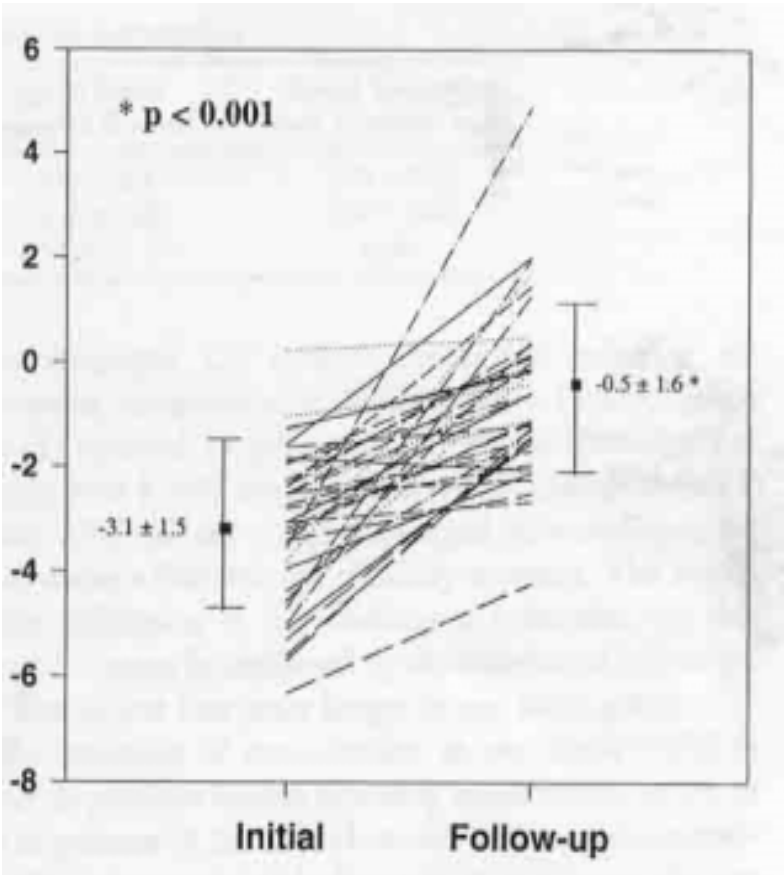
Two considerations : hemodynamic parameters and the presence of ASD

- Antegrade flow in the ascending aorta & presence of bi-directional blood flow at the level of PDA characterize LV with potential for growth

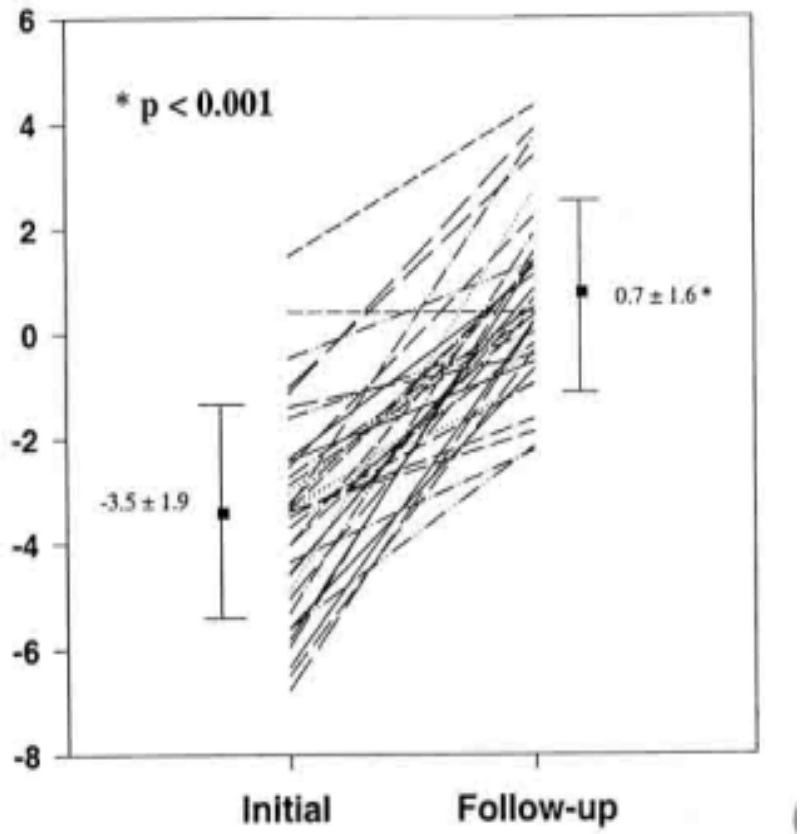
  → suitable for bi-ventricular type of repair
- Pre-operatively a significant left-to-right shunting at the atrial level, largely due to diastolic dysfunction secondary to the increased after-load

Follow-up of COA repair in neonate,
Puchalski et al, 2004, JACC

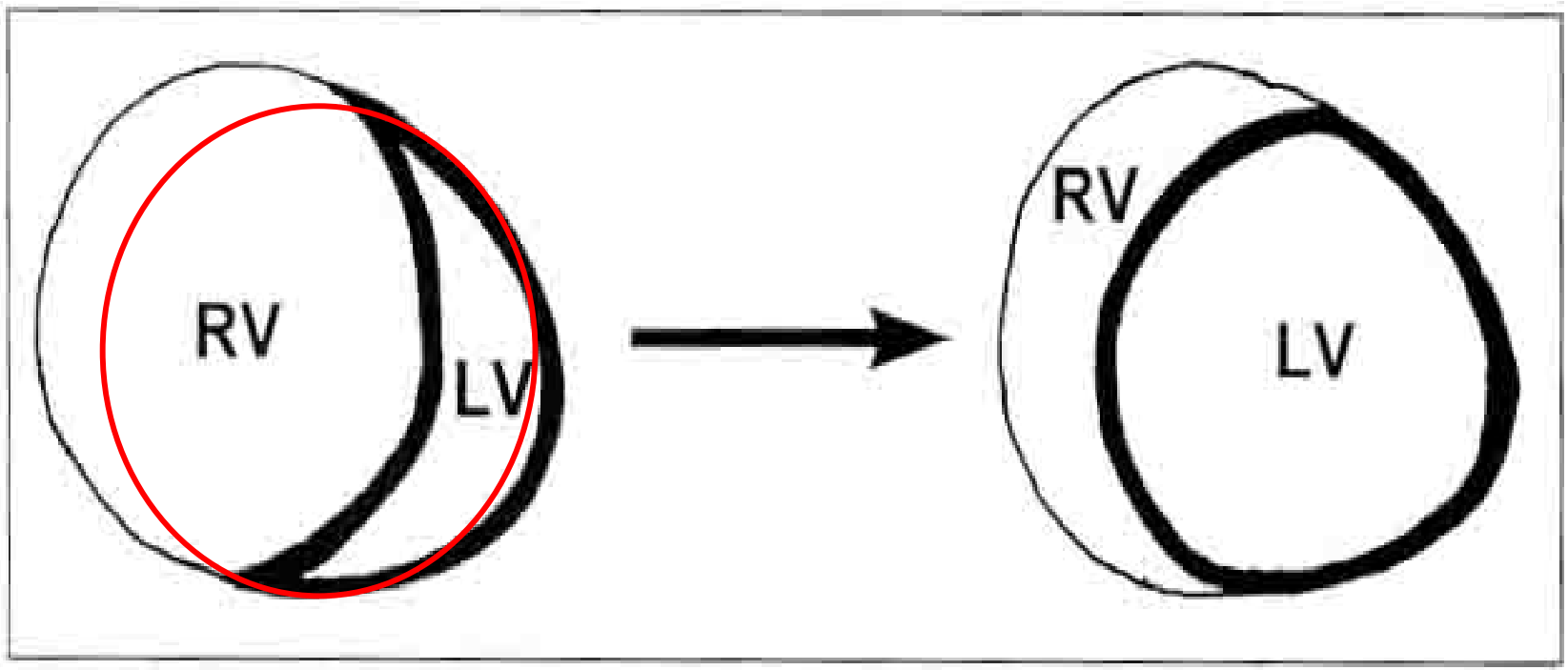
MV



AV



LV volume after correction of isolated COA in neonate, Krauser, 2000, Am J Card



LV volume after correction of islated COA in neonate, Krauser, 2000, Am J Card

TABLE I Patient Characteristics and Echocardiographic Measurements

Pt.	Additional Lesions	BSA	ROOT	LAR	MVA	LV Volume (ml [ml/m²])*			%Δ	Rhodes
						Potential	Preop	Postop		
1	–	0.26	3.90	0.83	2.35	6.5 (25.2)	4.6 (17.8)	7.8 (29.9)	67.6	−0.37
2	VSD, ASD	0.23	3.95	0.87	3.98	8.2 (35.5)	5.9 (25.5)	9.1 (39.7)	55.8	−0.31
3	VSD, PFO	0.23	4.78	0.92	3.69	7.5 (32.5)	6.7 (29.3)	9.3 (40.4)	37.9	0.67
4	PFO	0.24	4.08	0.94	4.14	7.7 (31.9)	7.2 (29.8)	8.5 (35.4)	18.9	0.32
5	ASD	0.23	3.29	0.97	4.31	6.9 (30.2)	5.7 (24.8)	6.6 (28.7)	15.6	−0.39
6	PFO	0.23	4.21	0.92	3.70	7.5 (32.8)	7.4 (32.2)	8.5 (36.9)	14.8	0.14
7	VSD	0.27	3.15	0.85	2.87	13.8 (51.2)	12.2 (45.0)	13.8 (50.9)	13.1	−0.77
8	PFO	0.27	5.52	0.95	6.05	13.0 (48.0)	12.9 (47.6)	14.3 (53.0)	11.3	2.41
9	PFO	0.23	4.30	0.91	3.28	9.9 (43.0)	9.0 (38.9)	9.8 (42.8)	9.9	0.11
10	VSD, ASD	0.24	3.83	0.78	2.57	6.2 (26.0)	5.5 (22.9)	6.0 (24.9)	8.7	−0.93
11	PFO	0.22	3.50	0.79	1.97	4.5 (20.3)	4.0 (18.3)	4.3 (19.5)	7.0	−1.6
12	PFO	0.26	3.96	0.97	4.58	14.3 (54.8)	13.3 (51.1)	13.5 (52.0)	1.7	0.73
13	PFO	0.24	4.46	0.82	3.85	10.1 (42.0)	8.9 (37.1)	8.8 (36.5)	−1.5	0.06
14	VSD, PFO	0.21	4.81	0.95	4.81	8.0 (38.1)	7.5 (35.7)	7.3 (34.8)	−2.6	0.74
Mean		0.24	4.1	0.89	3.7	8.9 (36.5)	7.9 (32.6)	9.1 (37.5)†	18.4	0.06
SD		0.02	0.6	0.07	1.1	3.0 (10.2)	3.0 (10.5)	3.0 (10.0)	21.0	0.95

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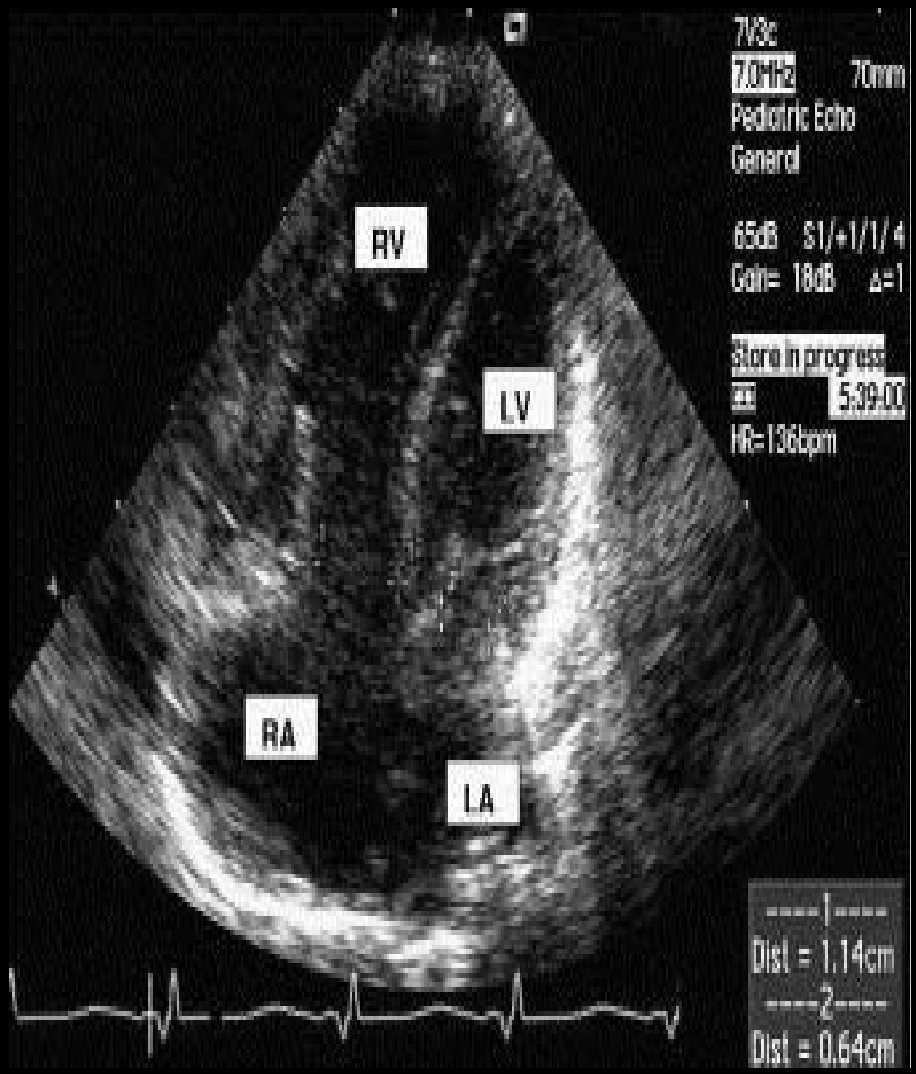
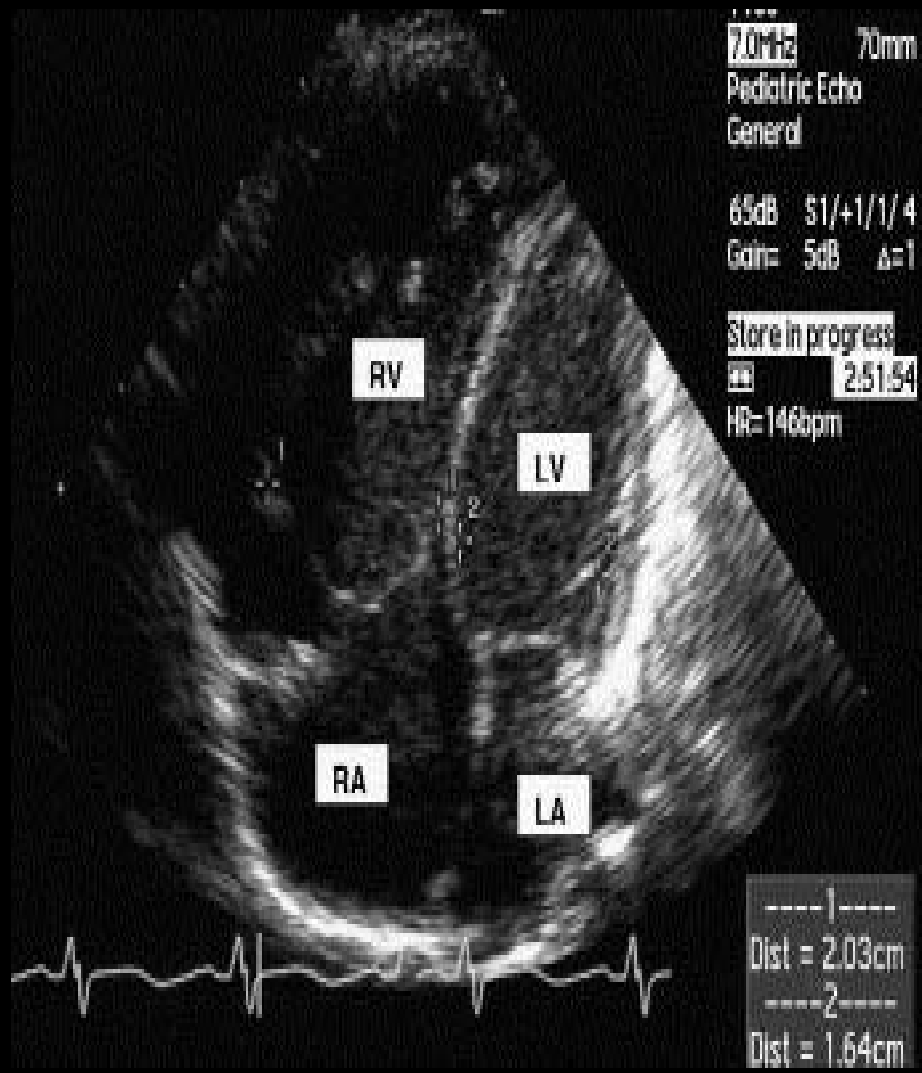
Hypoplastic left heart complex

- Favorable end of the spectrum of HLHS
- Characterized by hypoplasia of the structures of the left heart-aorta complex,
- Aortic and mitral valve hypoplasia without valvular stenosis or atresia, hypoplasia of LV, hypoplasia of LVOT, hypoplasia of the ascending aorta and of the aortic arch ± COA.

Before and after aortic coactectomy in patient with the borderline LV





Hypoplastic left heart complex - pathophysiology

- Constant presence of antegrade blood flow in the ascending aorta and the proximal branches of the aortic arch
- Absence of LV EFE
- Growth of LV, proved after bi-ventricular repair

Controversy for management of the frequently associated inter-atrial communication.

- Benefit
- reduce the risk of low CO
- improve the left ventricular filling and therefore the subsequent left ventricular growth
- Risk

stormy post-operative course with very elevated left atrial pressure particularly during the first 24–48 hr

“Partial closure of ASD”: Biventricular repair approach in ducto-dependant neonate with hypoplastic but morphological normal LV, Serraf A, 1999, JACC

- Neonate with hypoplastic left heart complex
- Severe aortic coarctation
- Hypoplastic aortic arch and ascending aorta
- Large ASD (diameter=10 mm)
- Small MV, 6mm, TV, 11 mm)
- Small LVEDV 14.5 mL/m2

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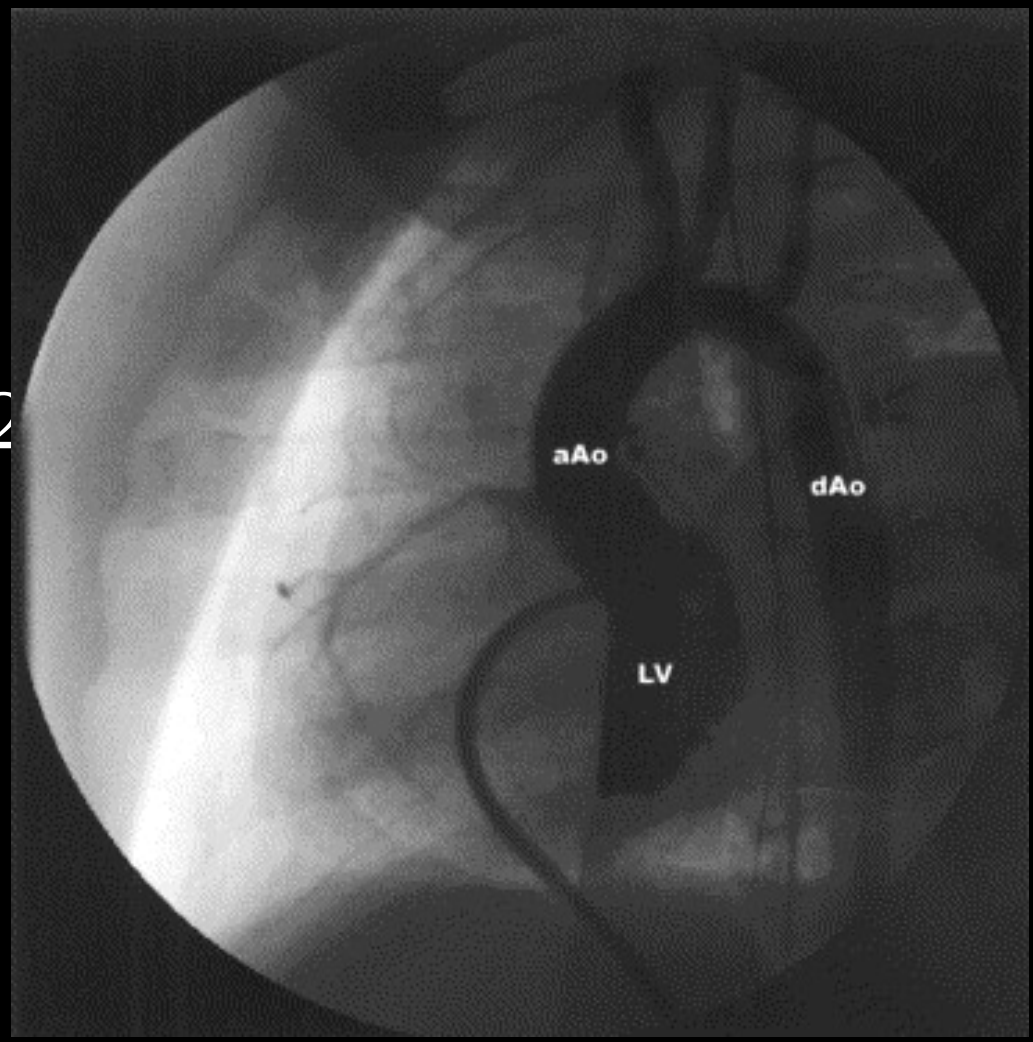
- Neonate with TAPVR

  small LV
- LVEDV=14 mL/m2



RV pressure and/or volume overload

- Total anomalous pulmonary venous connection,
- Unbalanced atrio-ventricular septal defect
- Cor triatriatum

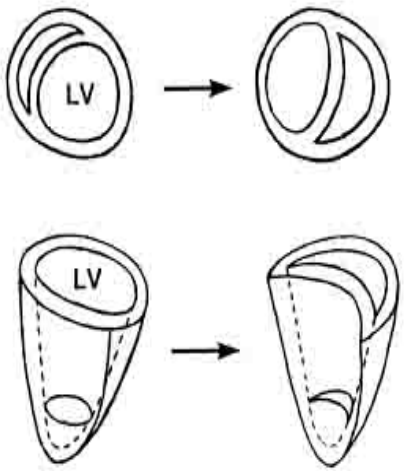
RV pressure and/or volume overload

- LV with normal mass, despite a reduced size of the aortic valve circumference

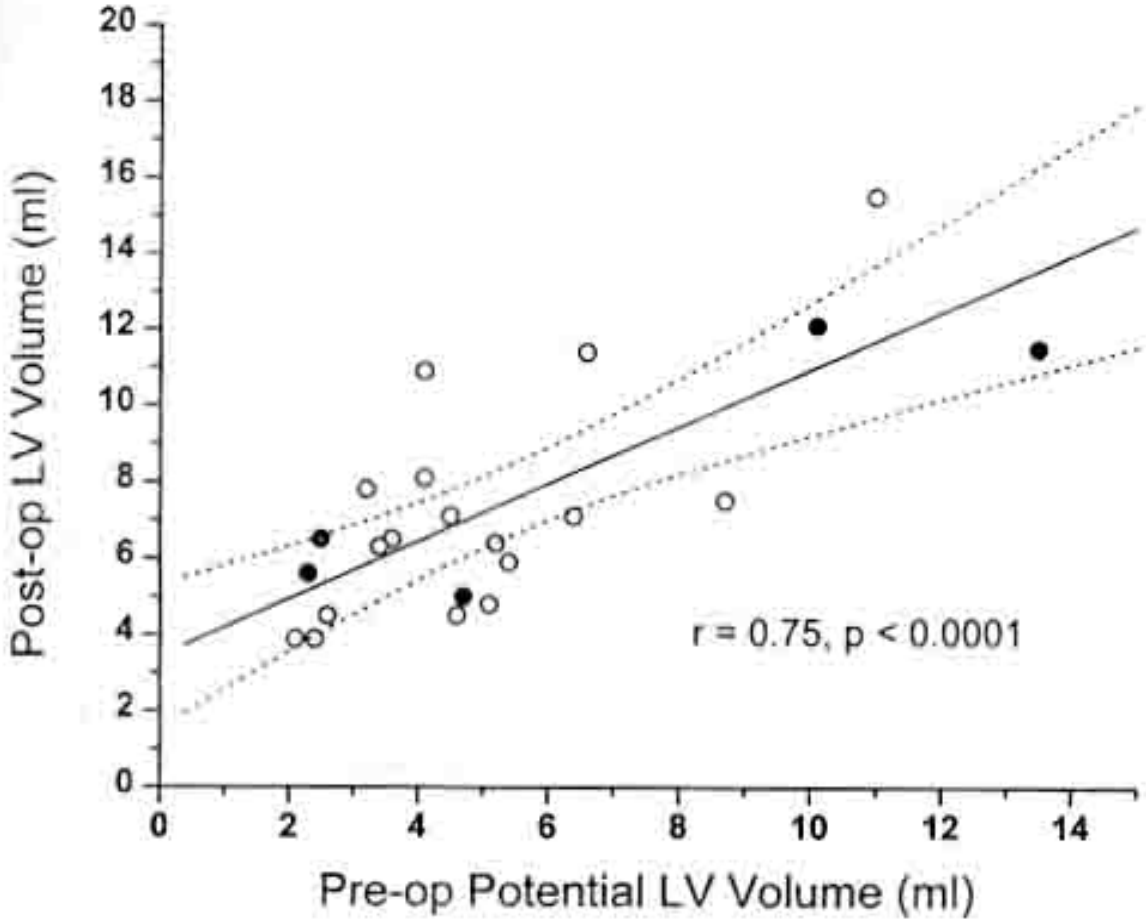
→ caused by low LV output probably compromised by

- the left to right shunt
- the septal displacement

Hypoplastic LV or simply a squashed ventricle? Collin Phoon, 1997, JACC



- Circumference
- Potential area
- Potential LV volume



Hypoplastic LV or simply a squashed ventricle? Collin Phoon, 1997, JACC

- Hypothesized PO LV capacity may be better reflected by Preop. potential LV volume
- calculated as LV volume if the septal position were normal, without the reverse bowing of the inter-ventricular septum
- the right-to-left bowing of the inter-ventricular septum does not induce any change of the endocardial circumference nor of the LV length

Hypoplastic LV or simply a squashed ventricle? Collin Phoon, 1997, JACC

- Small pre-operative LV volume is not primarily due to compression by an overloaded right ventricle, but rather to underfilling of LV
- " LV is not 'hypoplastic', is simply 'squashed"

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Elements to be considered for the decision making process

- Morphometric parameters
- Functional parameters
- Hemodynamic data
- Available surgical options
- Results of the personal and institutional experience

Following considerations may help…

- Fontan after a failed attempt of 2 ventricular repair → increased mortality

  vs. the opposite → better outcome
- the question ‘Is a high-risk bi-ventricular repair always preferable to conversion to a single ventricle repair?’
- the most important consideration ‘every patient is unique’