CARDIOLOGY 2023

Determining Adequacy and Viability of the Left Ventricle

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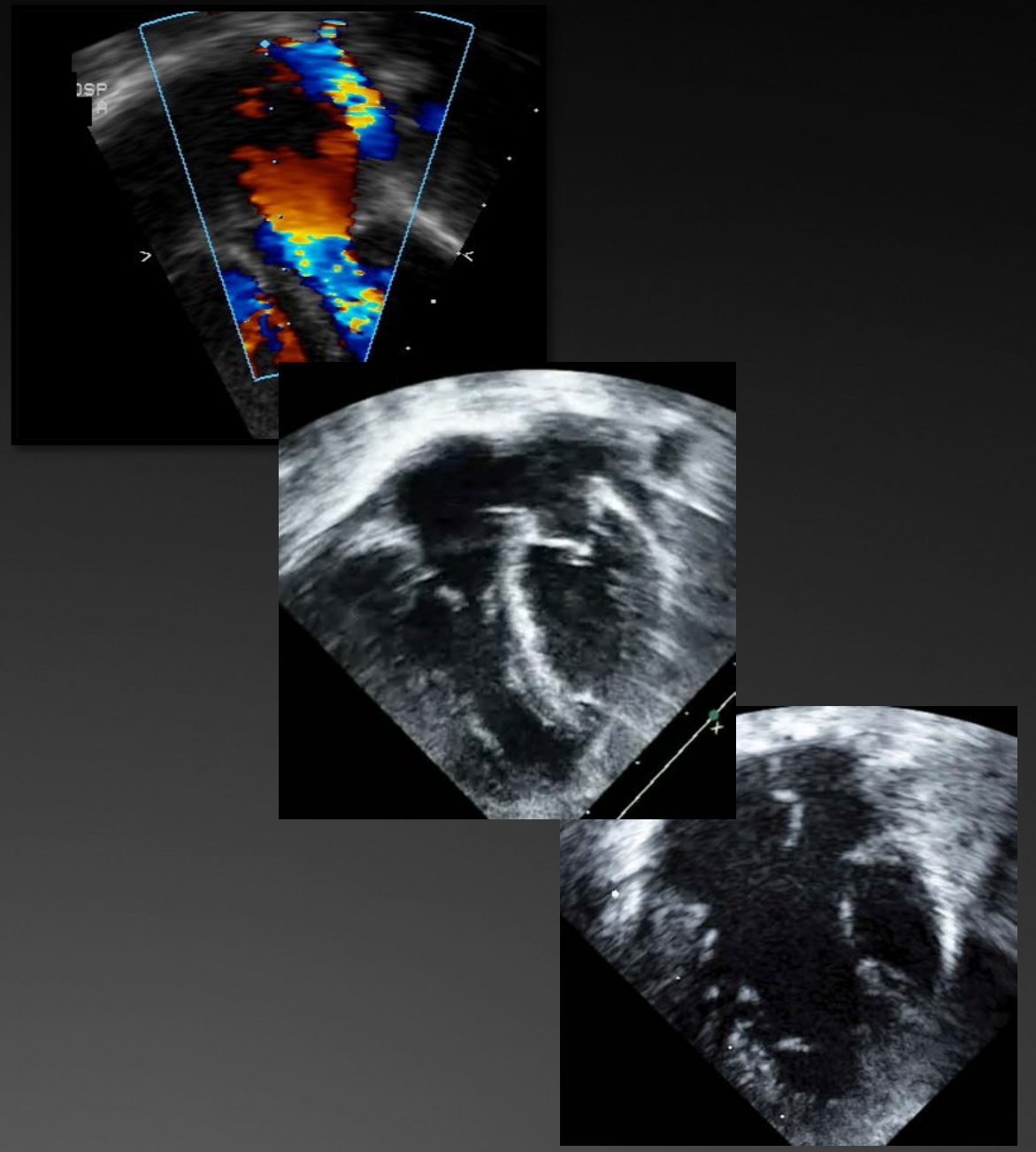

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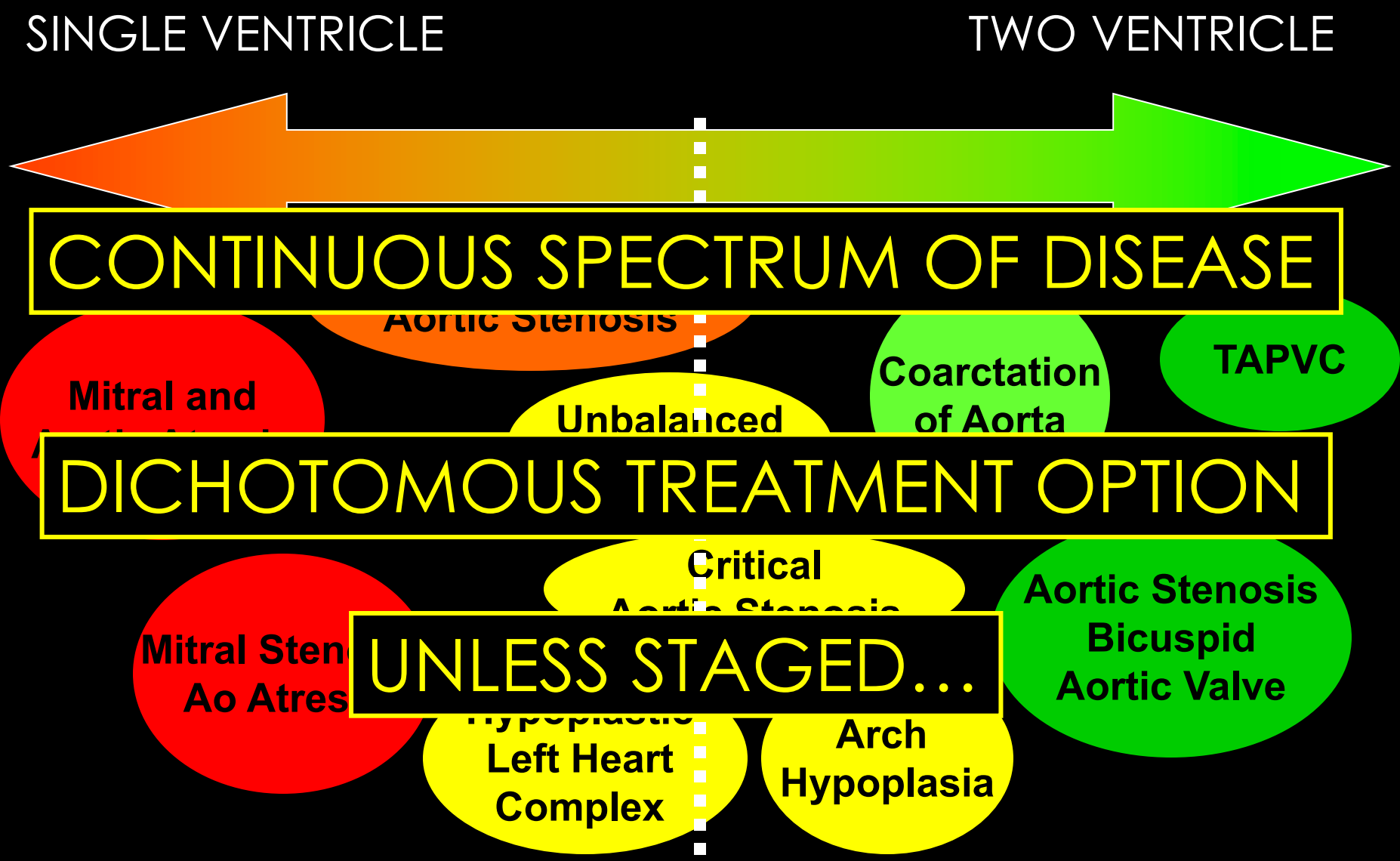
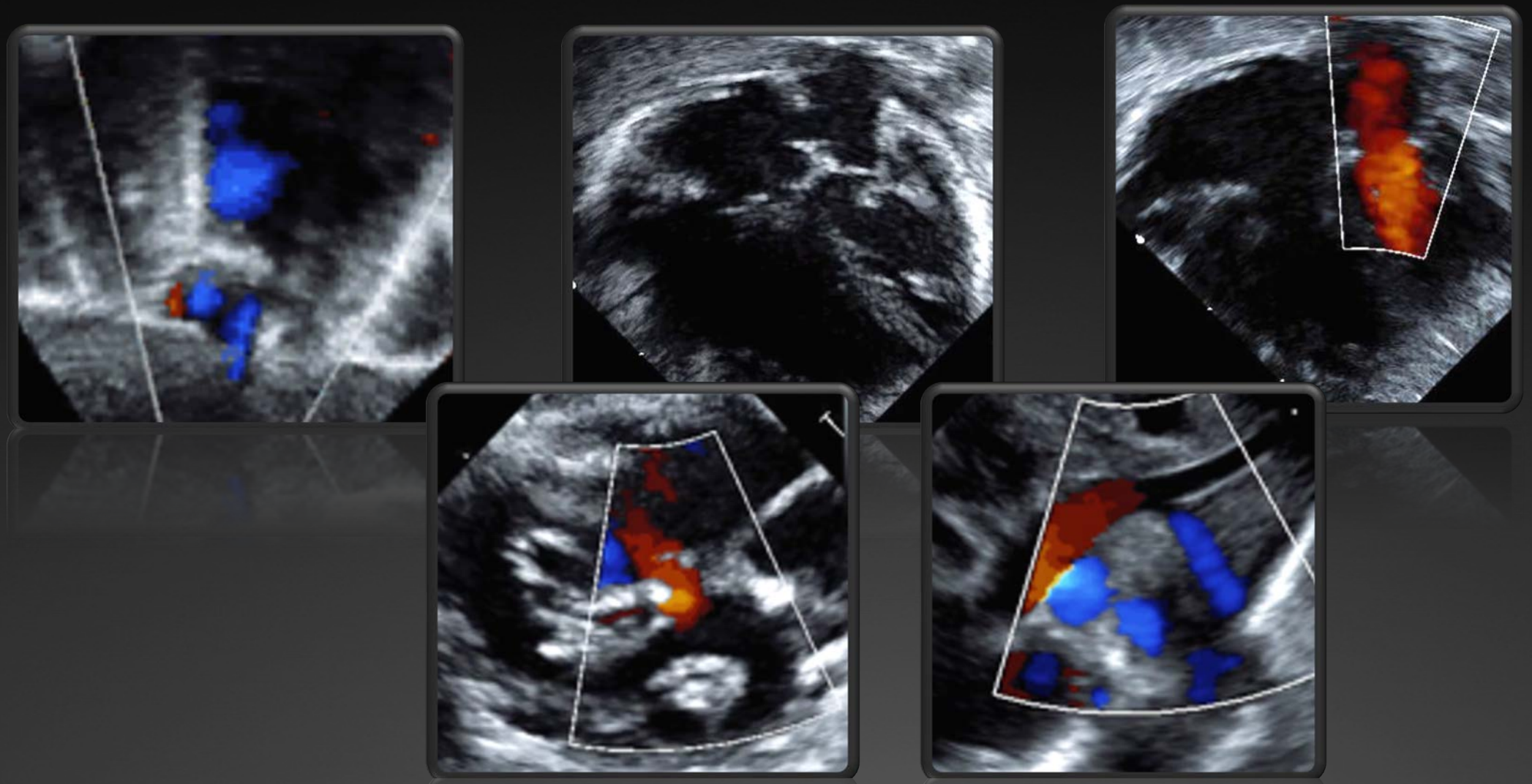
Disclosures

- I have no answers about how to determine adequacy of the left ventricle for biventricular repair
- Sorry…

LV Hypoplasia



Day of Life 0

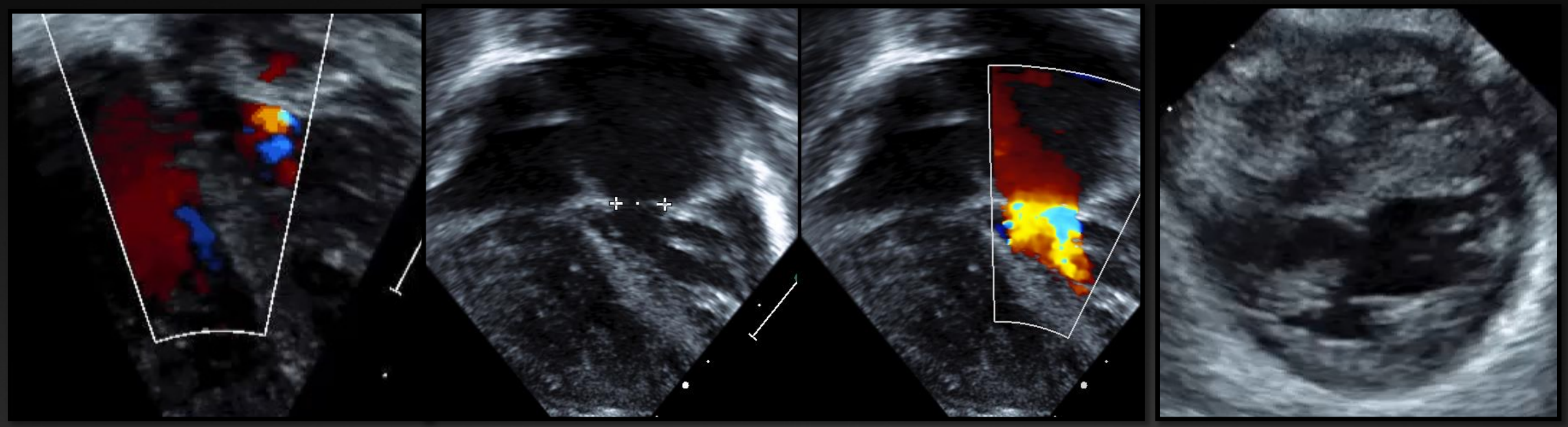


CICU Course

- Repeated echo to re-measure left structures
  - VSD remained bidirectional
  - Arch flow all antegrade
  - Mitral inflow with no gradient
    - Two papillary muscles
    - Probable variant of mitral valve arcade

	DOL 1 cm, z-score	DOL 5 cm, z-score
Mitral Valve	0.5, - 3.0	0.6, -2.5
Aortic Valve	0.4, - 3.1	0.4, -2.8
Transverse Arch	0.2, - 4.2	0.3, -3.1
LVEDD	0.9, - 4.5	1.2, -2.8

Echo 6 Months After Arch Repair



- LVOT ok, VSD right to left in systole
- Mitral inflow mean gradient is 15 mmHg
- Suprasystemic PA pressure; PVR 5.5 WU

Why Haven't We Figured This Out?

- Prenatal physiology ≠ postnatal physiology
- Pre-op physiology ≠ post-op physiology
- Progressive effects of multiple levels of left-sided obstruction
- Intervention may:
  - Leave residual hemodynamic abnormalities
  - Result in "new" disease (MR or AR)
  - Result in diastolic heart failure/pulmonary hypertension

What are we talking about?

- Hypoplastic left heart syndrome
- **Critical aortic stenosis**
- **Hypoplastic left heart complex/arch hypoplasia**
- Unbalanced atrioventricular canal defect
- Double outlet right ventricle
- TAPVC

Important Imaging Features

- LVOT size and anatomy
  - Aortic annulus size
- Aortic arch/CoA
- Presence of atrial and/or ventricular communication
- LV inlet size and anatomy
- LV size and function
- Presence of EFE
- RV function
- Presence of significant TR
- Branch pulmonary arteries
- VSD size and location
  - Re: Baffle to aorta/neoaorta

Important Imaging Features

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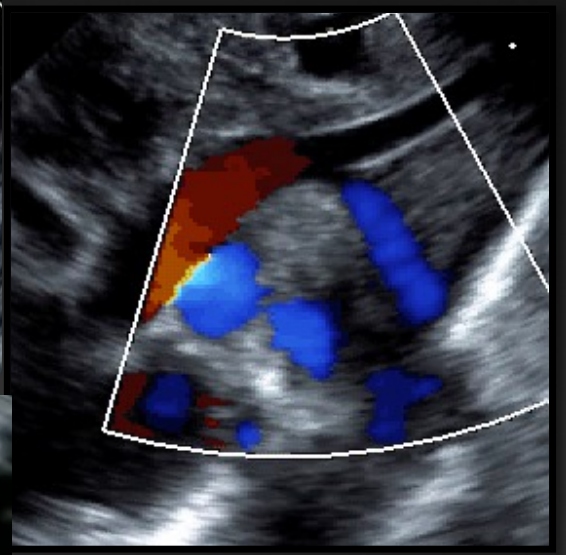

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- **LV inlet size and anatomy**
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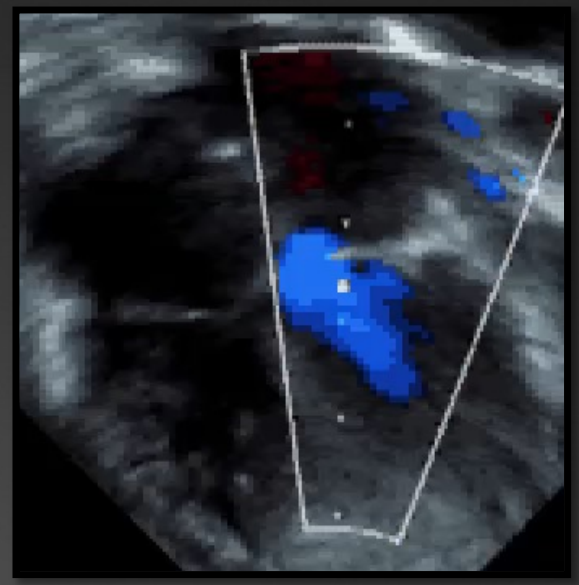
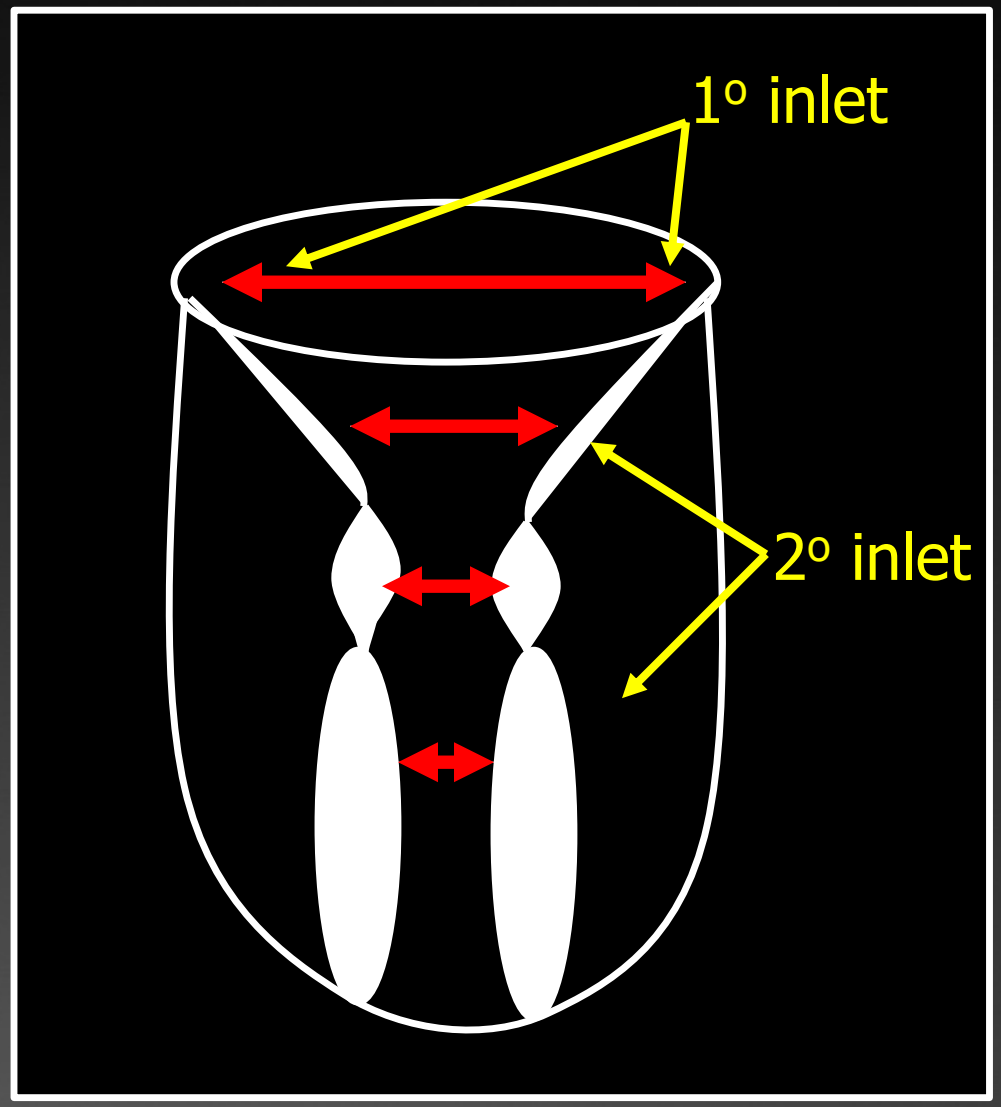


Important Imaging Features

- LV inlet size and anatomy
- LV size and function
- Presence of EFE
- LVOT size and anatomy
  - Aortic annulus size
- Aortic arch/CoA
- Presence of atrial and/or ventricular communication
- **RV function**
- **Presence of significant TR**
- **Functioning pulmonary valve**
- **Branch pulmonary arteries**

Analysis of LV Inlet

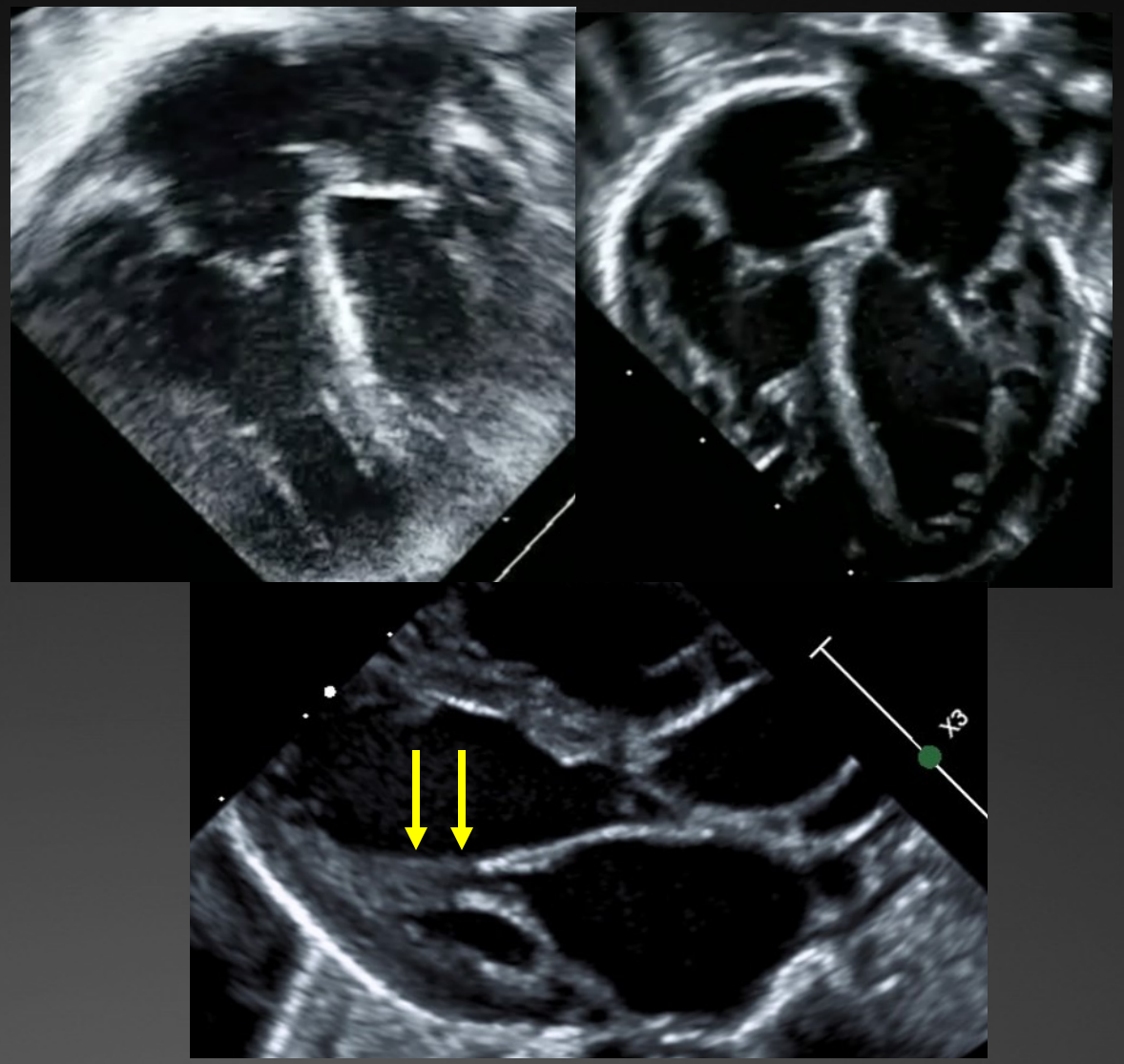
- MV diameter
  - Z-score < ~-2.0
  - Leaflet and chordal anatomy
  - Papillary muscle number and location
  - Is the 2° inlet diameter narrowed?
- If the inlet is inadequate, the LV is likely to be inadequate as well…
  - If flow leads to LV growth, lack of flow leads to hypoplasia



Critical Aortic Stenosis

Risk Factors for BiV Repair

- Small Ao annulus
- LV dysfunction
  - EFE
  - Short, fat LV
- MV abnormalities
- Scores for decision-making
  - Rhodes/Colan
  - CHSS (Lofland)



Critical Aortic Stenosis

RHODES CRITERIA, CIRC 1991

- Score = 14.0 (BSA) + 0.943 (Aortic root dimension$_i$ + 4.78 long-axis dimension of heart + 0.157 (mitral valve area$_i$ - 12.03
- If score < -0.35, predicts failure of two ventricle repair

or 2 or more of following risk factors

- LV long-axis to heart long-axis ratio ≤ 0.8
- Aortic root diameter$_i$ ≤ 3.5 cm/m^2
- Mitral valve area$_i$ ≤ 4.75 cm^2/m^2
- LV mass index ≤ 35 g/m^2

Published Series on Critical Aortic Stenosis

Author	Predictors of Mortality
Latson, 1981	LV X-sect area < 1.6 cm^2
Mocellin, 1983	EFE, LVEVV < 80% normal
Gundry, 1986	EFE, Low EF, High LVEDP
Pelech, 1987	MV < 11mm, AoV < 6.5 mm
Hammon, 1988	LV X-sect area < 1.6 cm^2
Karl, 1990	Non-Apex forming LV
Parsons, 1991	LVEDD <13mm, vol <20ml/M^2
Rhodes, 1991	MV, AoV, Score < -0.35
Kovalchin, 1998	Retrograde flow in Asc Aorta
Lofland, 2001	EFE, smaller Ao V, older age
Colan, 2006	MV, New Score < -0.65, EFE

Mitral Valve

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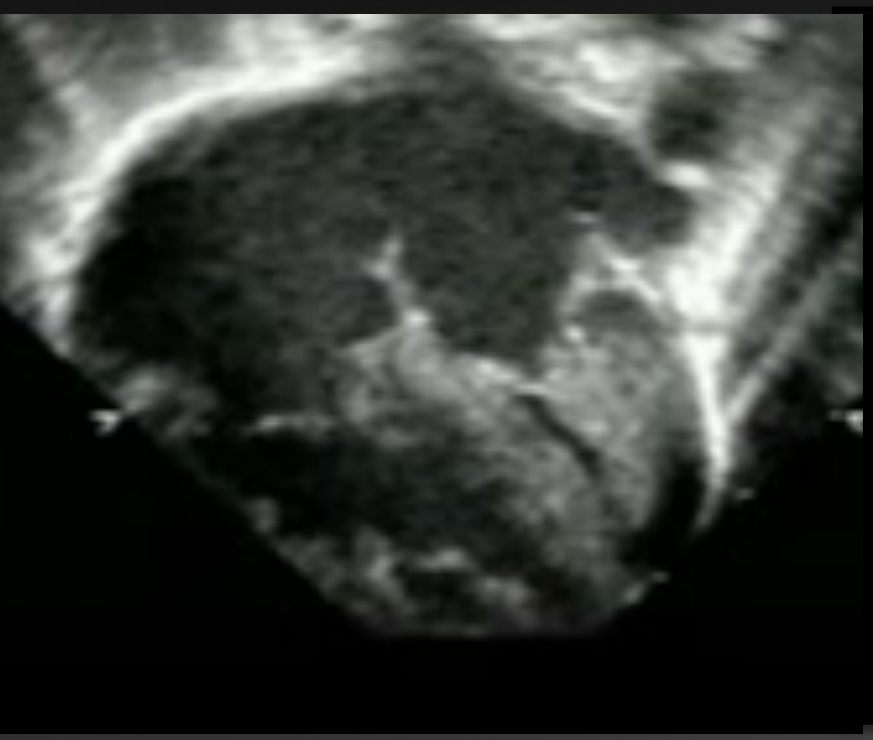
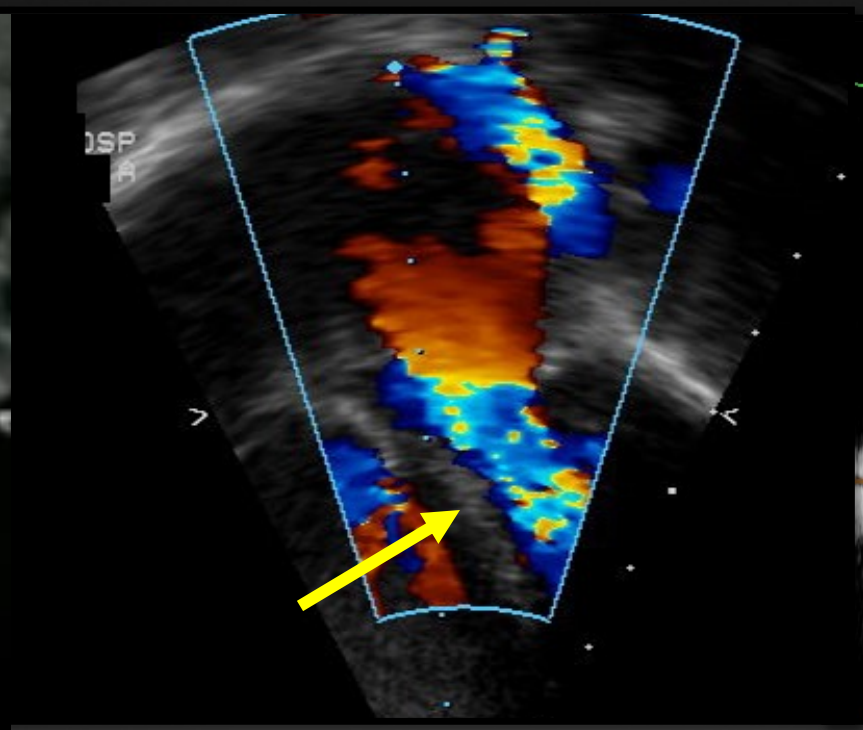
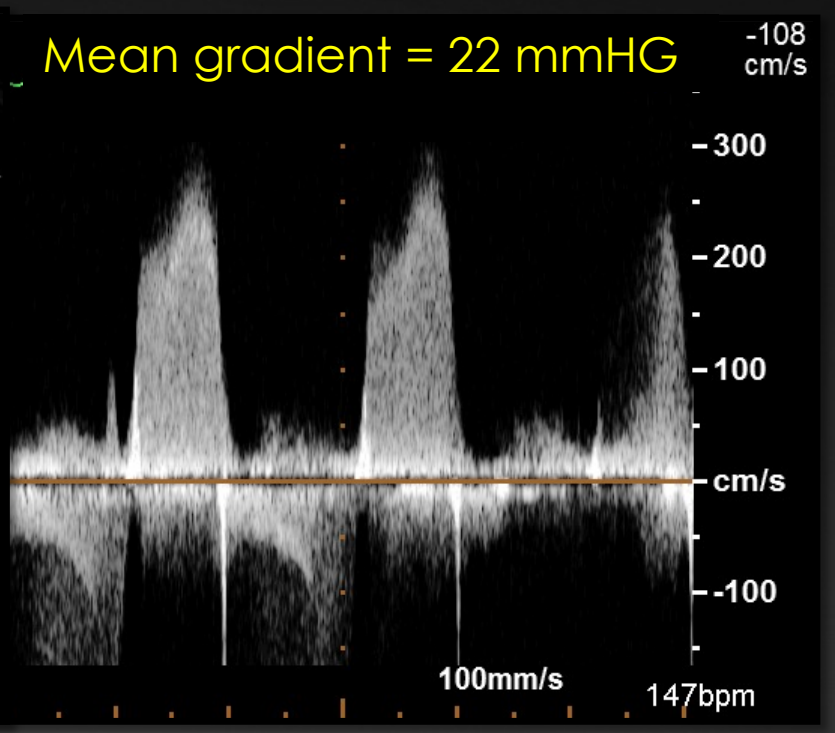
Mitral Arcade with Severe Progressive Stenosis

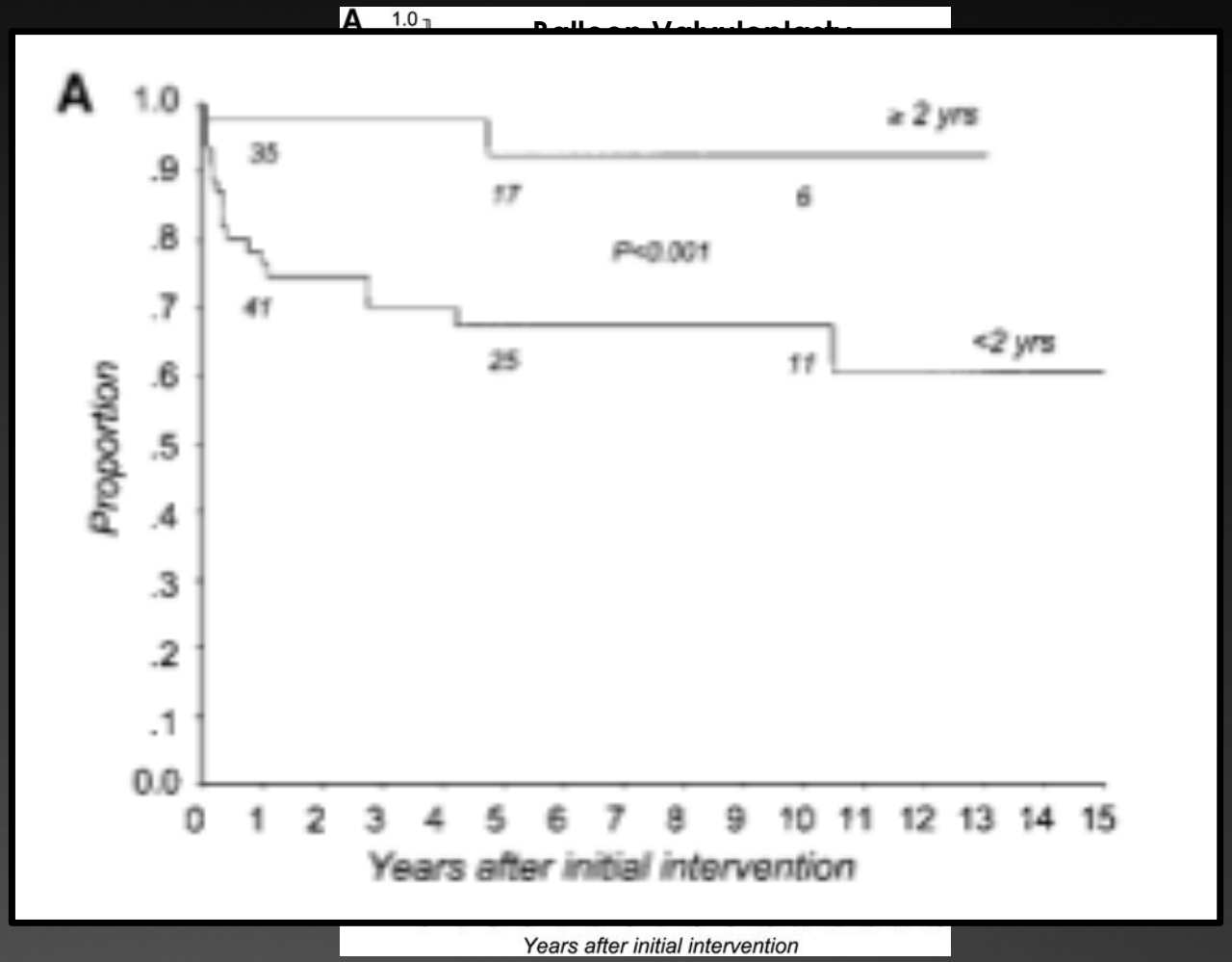






Congenital Mitral Stenosis

- 108 pts (1 mo to 18 yrs)
  - Surgical valvuloplasty (n=33)
  - Mitral valve replaced (n = 11)
  - Balloon MV dilation (n = 64)
- 38 deaths
- 3 Heart transplant
- 30 re-interventions
- 28% developed severe MR



Years after initial intervention

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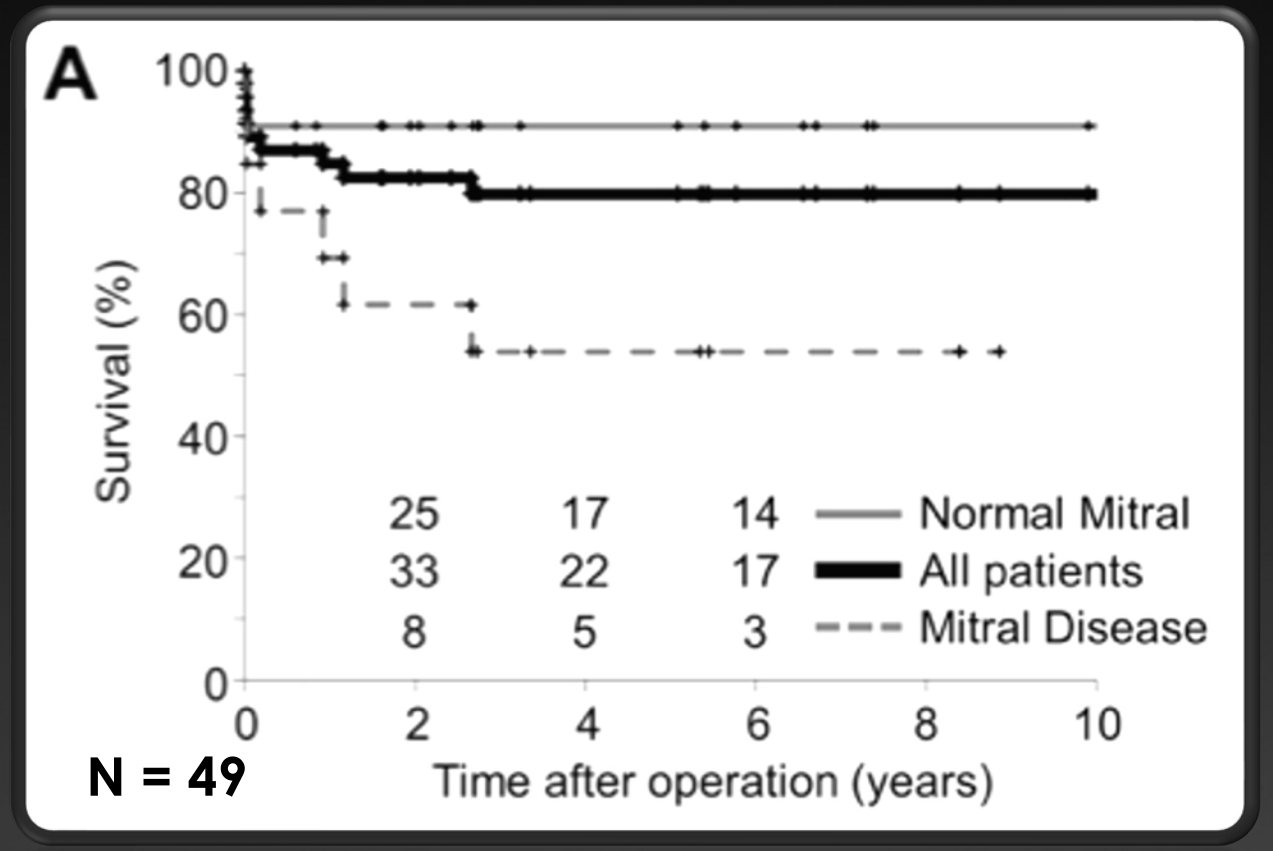
McElhinney D et al. Circulation;2005

Ross/Konno: Mitral Valve



All late deaths (4) in pts with abnormal mitral valve

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Vergnat M et al. Ann Thorac Surg 2014

LV Hypoplasia

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Endocardial Fibroelastosis

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Colan, 2006	New Score < -0.65, **EFE**

Endocardial Fibroelastosis

- Predictor of mortality
- Cessation of LV growth in utero
- Causes diastolic dysfunction/PH

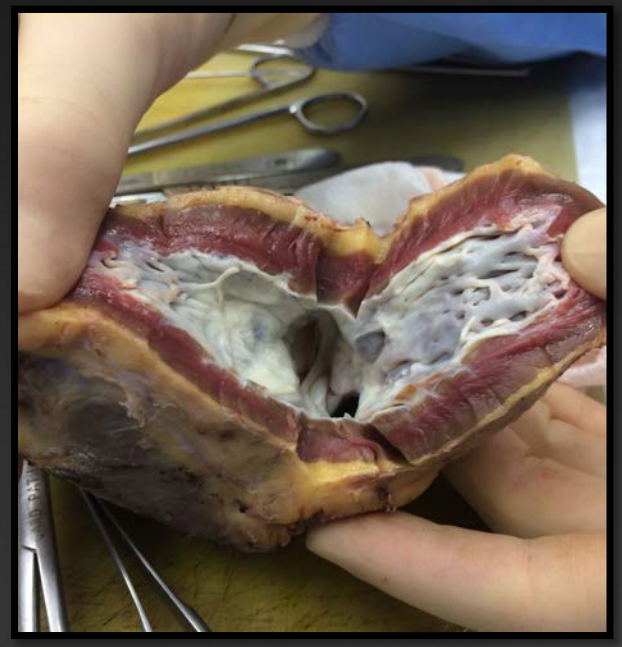
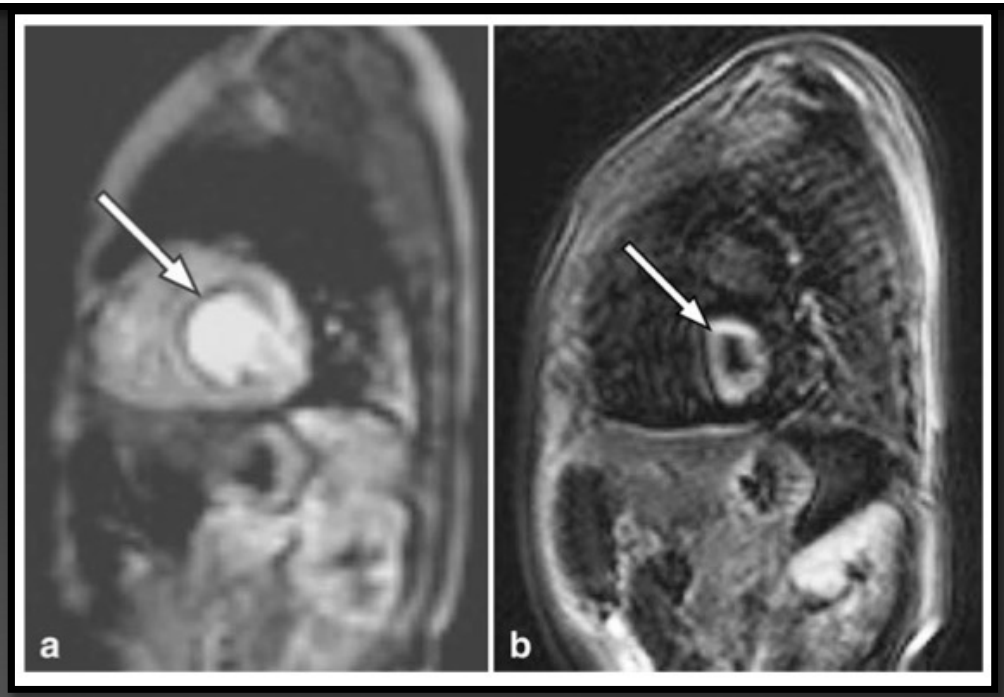


TABLE I Comparison of Echocardiographic and Histopathologic Evaluation for Endocardial Fibroelastosis*

	Biopsy Positive	Biopsy Negative
Echo-bright endocardium	19	3
Normal endocardium	1	9

*Sensitivity = 95%, specificity = 75%.



Mahle WT et al. Amer J Cardiol 1998
Stranzinger E et al. Pediatr Radiol 2008

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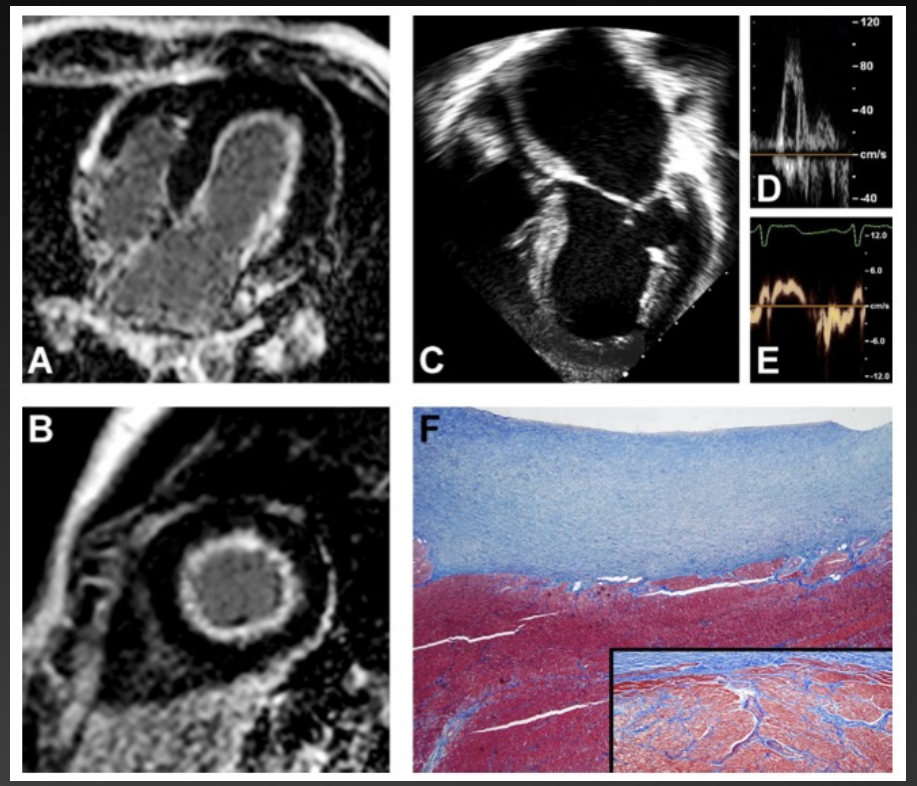
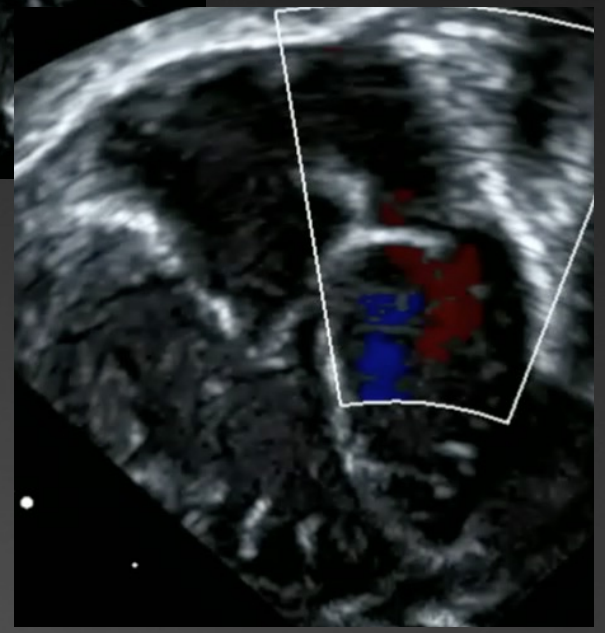
Case Reports of PH



- Late PH in pts with critical AS
- Removing all EFE is not possible
- Is the myocardium underneath the resection normal?

Hypoplastic Left Heart Complex

- Arch hypoplasia with:
  - Small MV, AoV but no obstruction
- Narrow LV with good function
- **No EFE**



- Almost always amenable to 2V repair

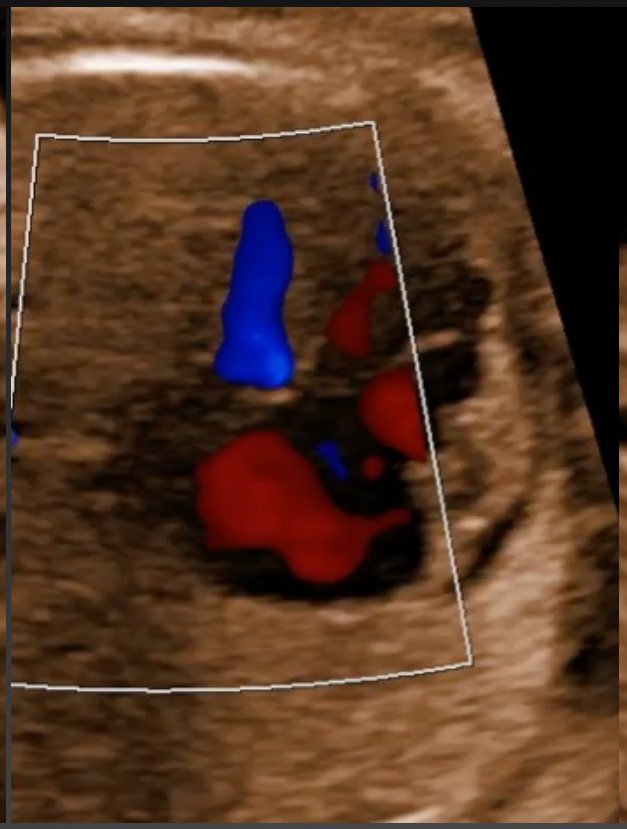
Hypoplastic Left Heart Complex

- Sometimes not straightforward:
  - ASD: Causes significant left to right shunt and systemic RVp
  - VSD: Does it shunt right to left to augment cardiac output?
  - MV abnormalities may be masked in infancy
- Left-sided obstruction may develop

LV Hypoplasia: Imaging

- The ventricular "inlet" is the most important element to success of biventricular repair
- Consider pre-op MRI to
  - Assess LV volume and function
  - Presence of EFE
- Long term concerns:
  - Recurrent left-sided obstruction
  - Effect of EFE resection
  - Diastolic dysfunction and pulmonary hypertension

How to counsel a family with this fetal echo?



Borderline LV

"What are we to do when the need to offer clinical advice meets with the immovable object of flawed evidence?"

"All we can do is our best: give the advice, but alert the advisees to the flaws in the evidence on which it is based."

- Oxford Centre for Evidence Based Medicine

THANK YOU