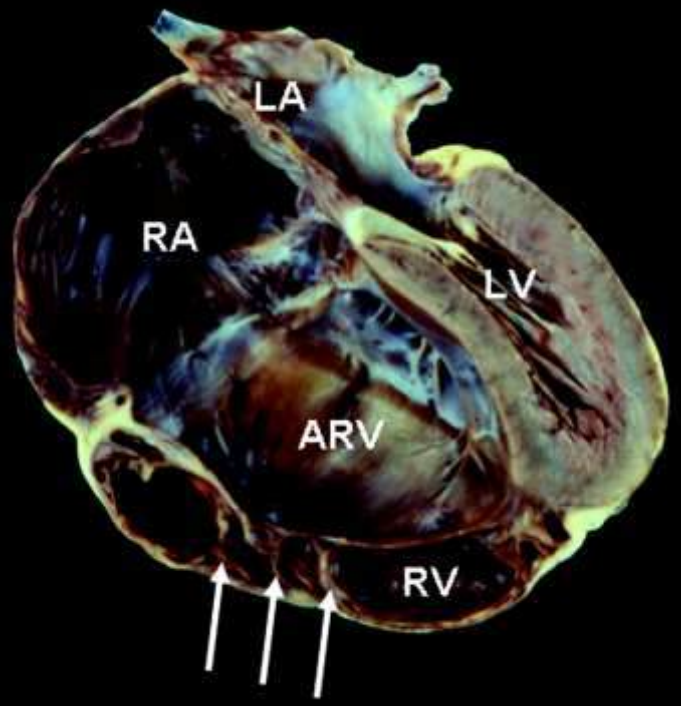
RV electromechanical dyssynchrony and interventricular interactions in Ebstein's anomaly

Mark K. Friedberg, MD, PhD

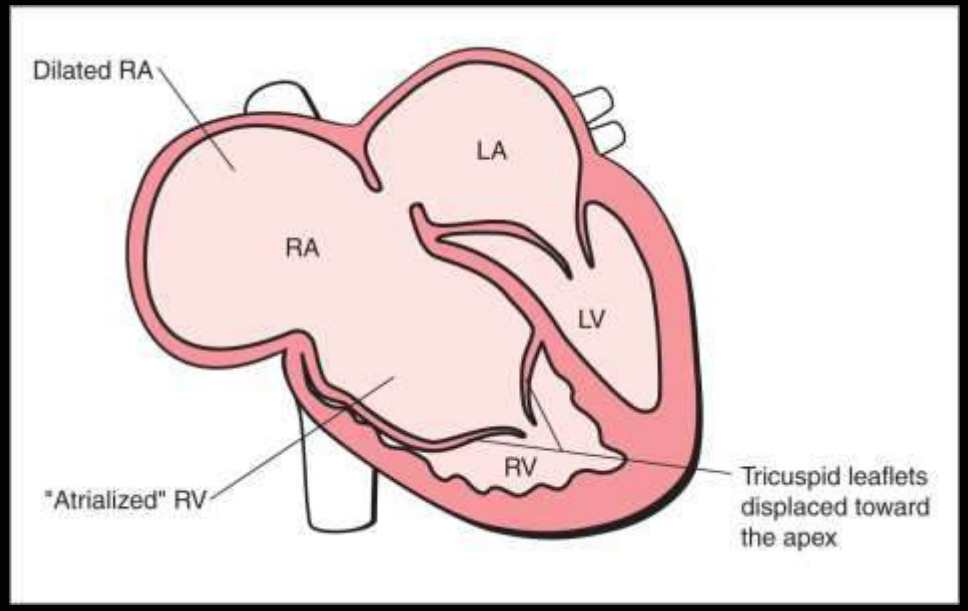


SickKids®
THE HOSPITAL FOR SICK CHILDREN
Echocardiography

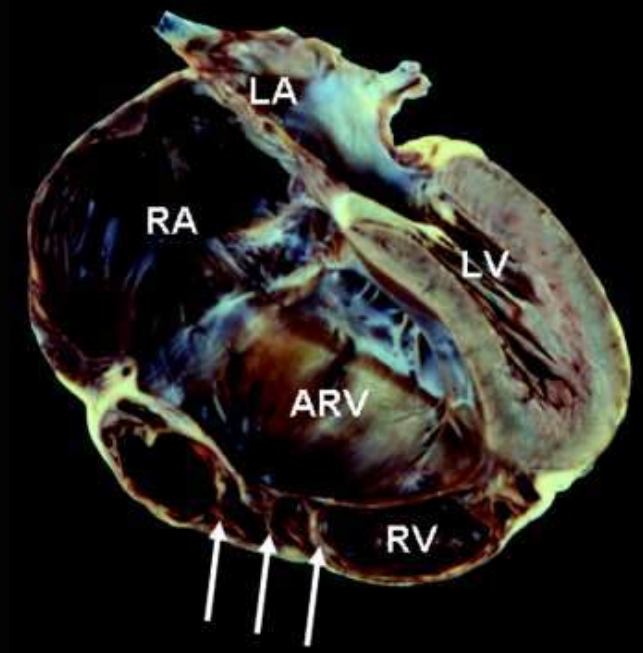
NO DISCLOSURES

Objectives

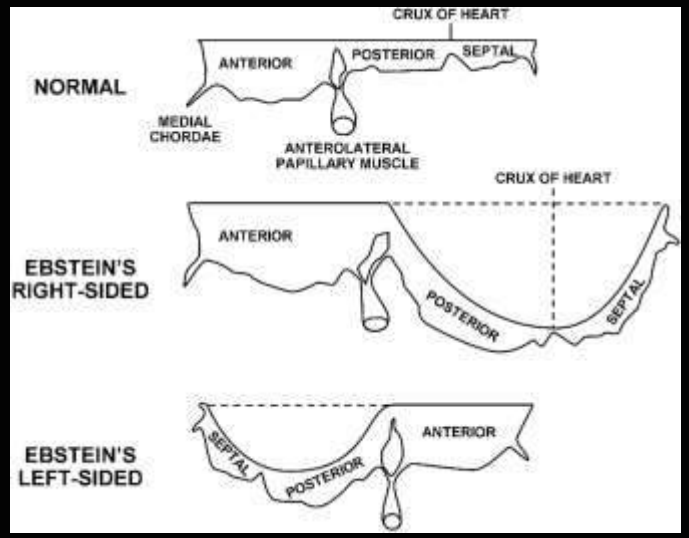
- Briefly review electromechanical dyssynchrony in EA
- Previous insights on RV-LV interactions in EA
- Present our current work using ML
- Show how surgical intervention changes R-L interactions

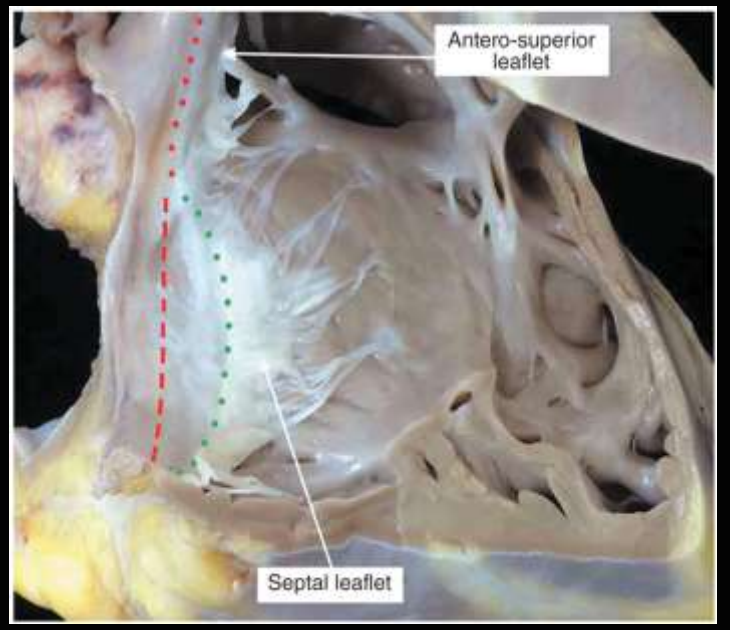


Anderson, Mayo Clin Proc. 1979; 54: 174–180



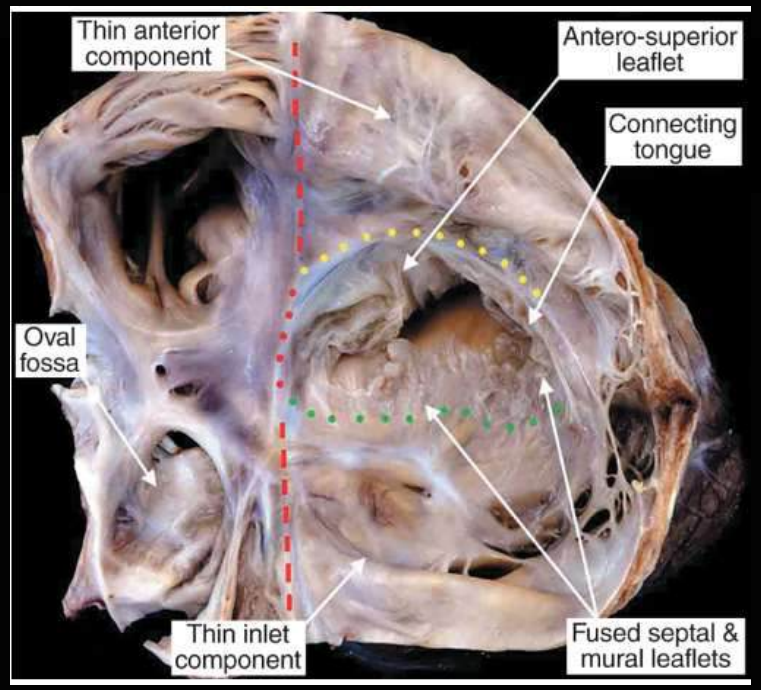
Holst and Dearani

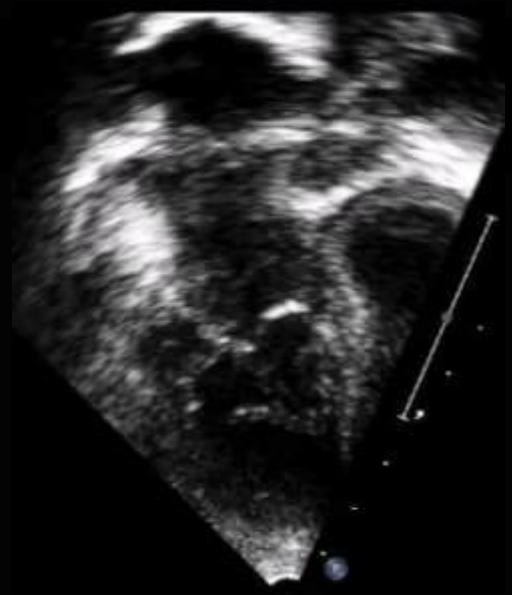


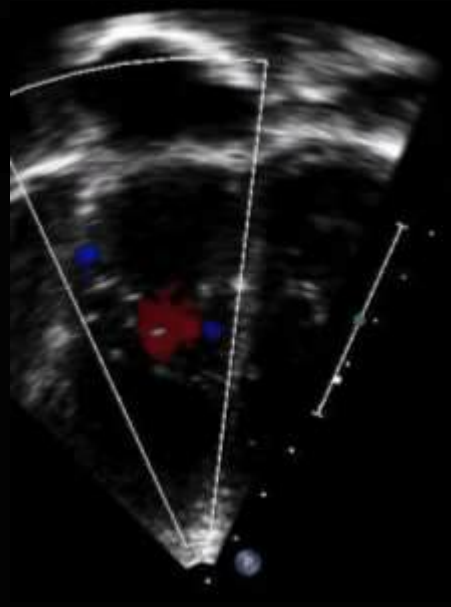


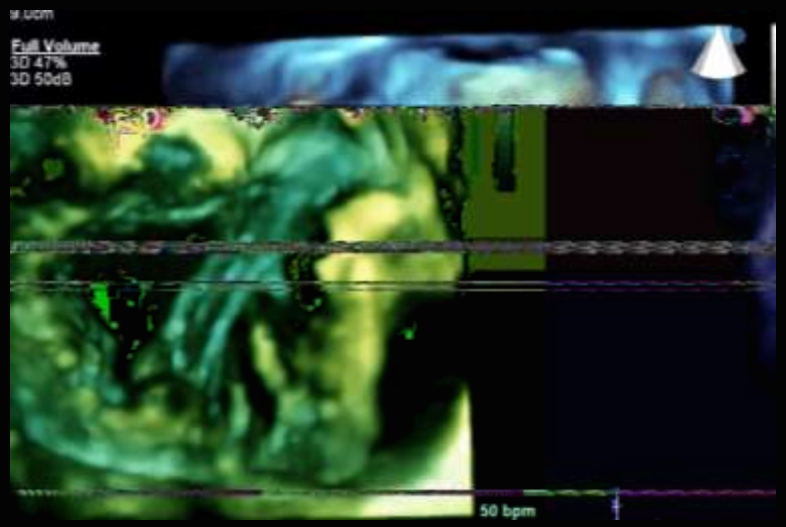
Attenhofer, Circulation. 115, 2007, 277-285

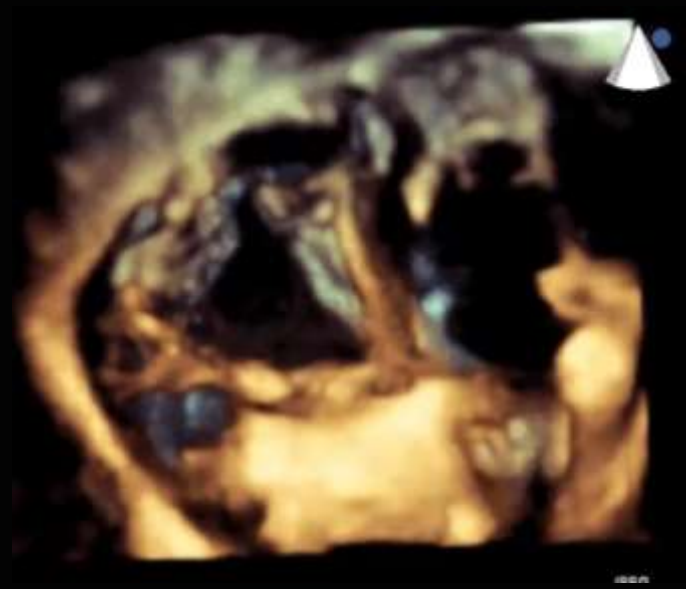
Ebstein anomaly

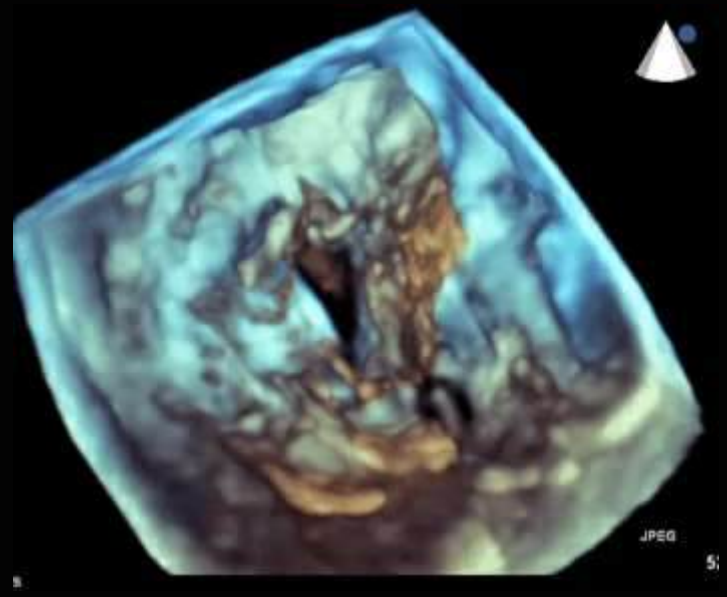




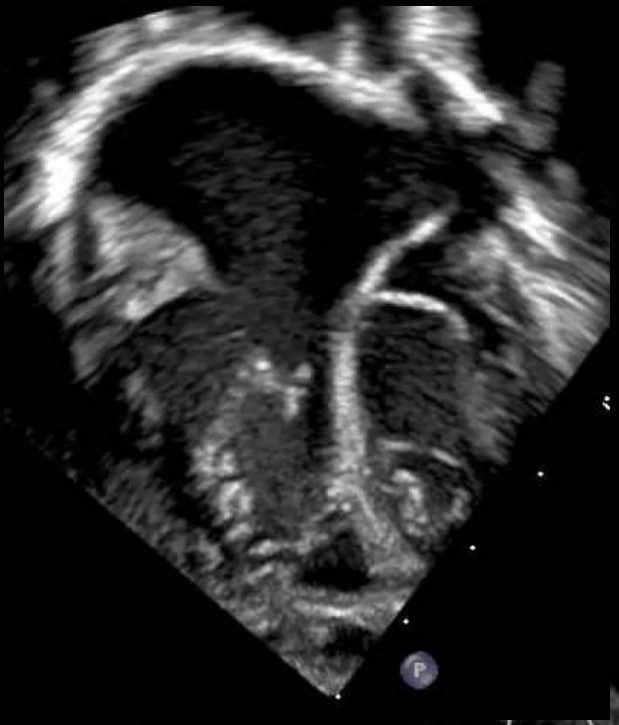


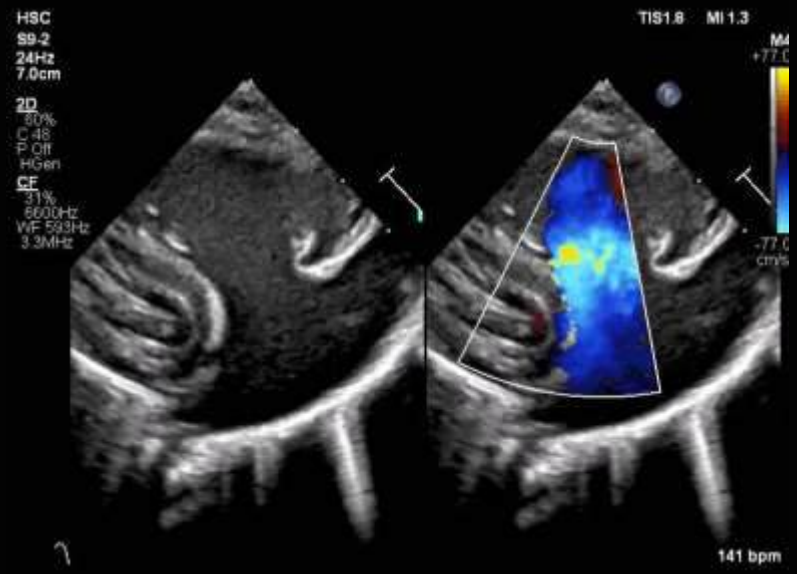


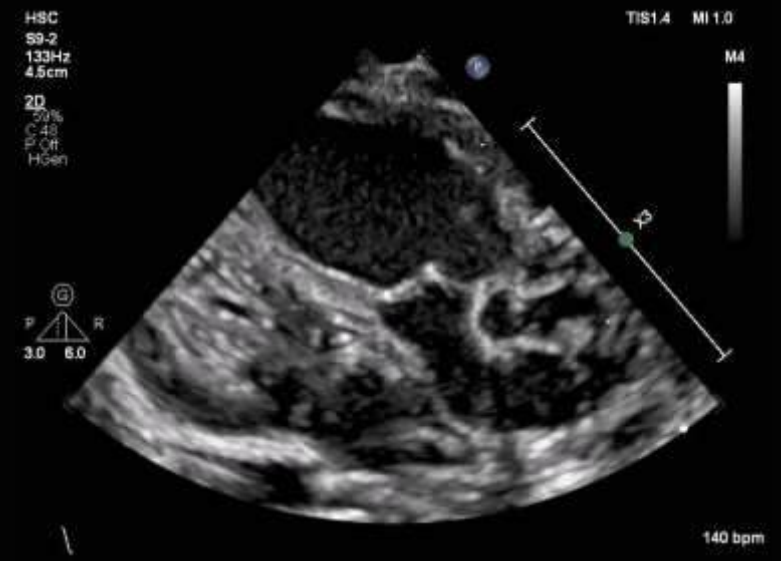


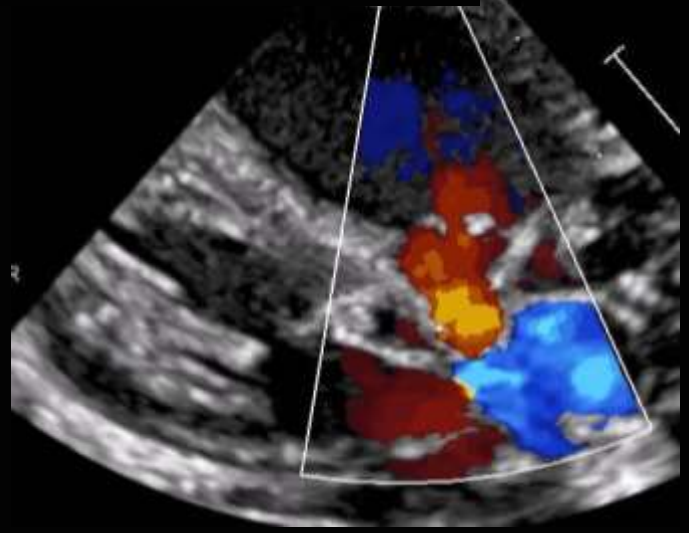


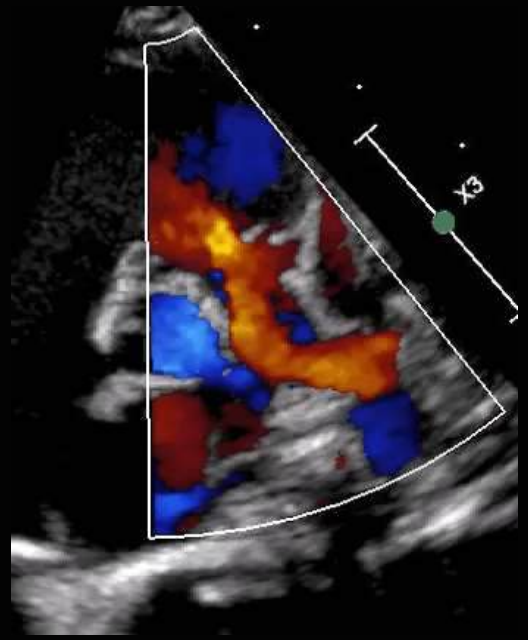
Neonatal Ebstein circle of death

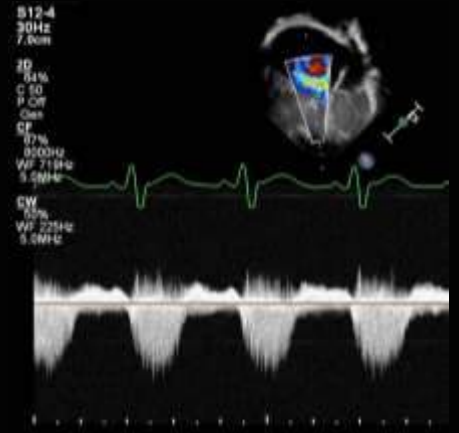




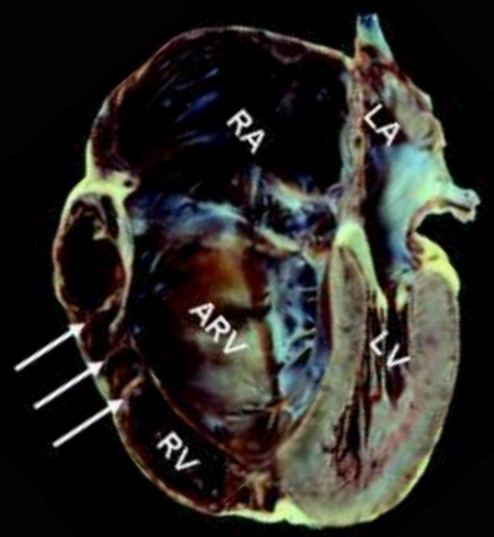


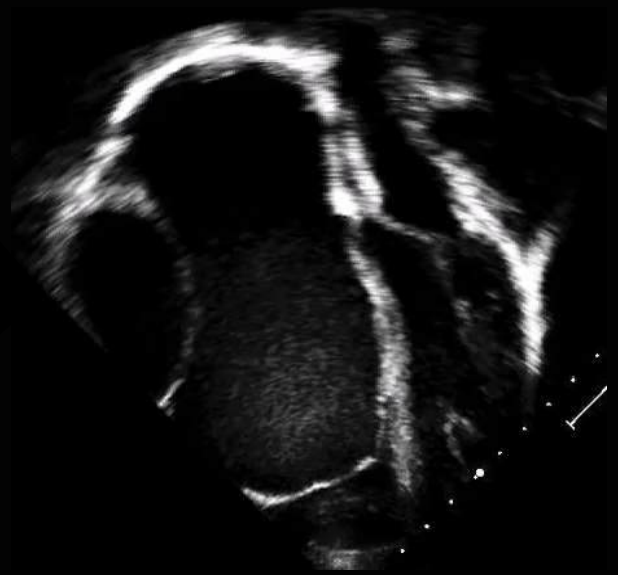


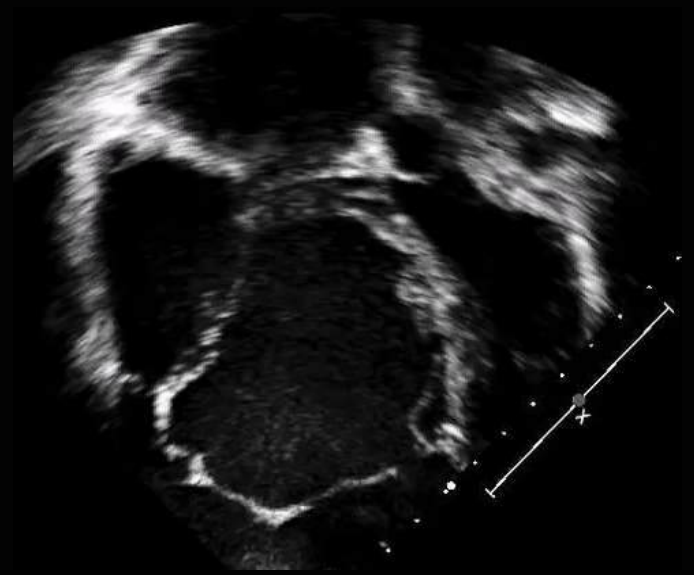


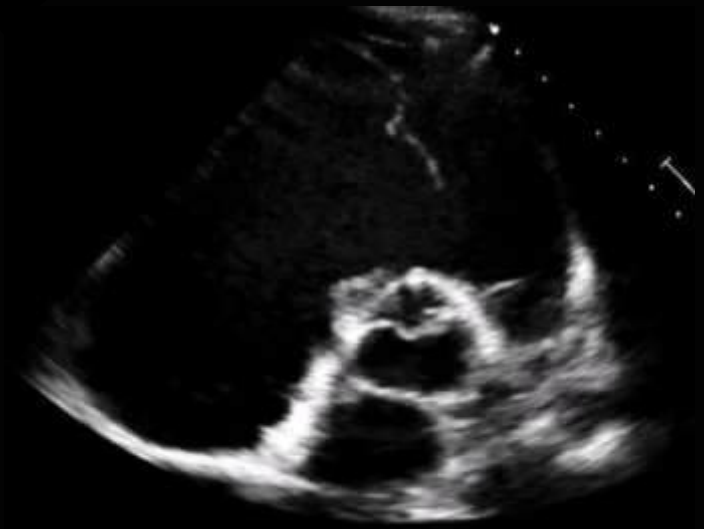


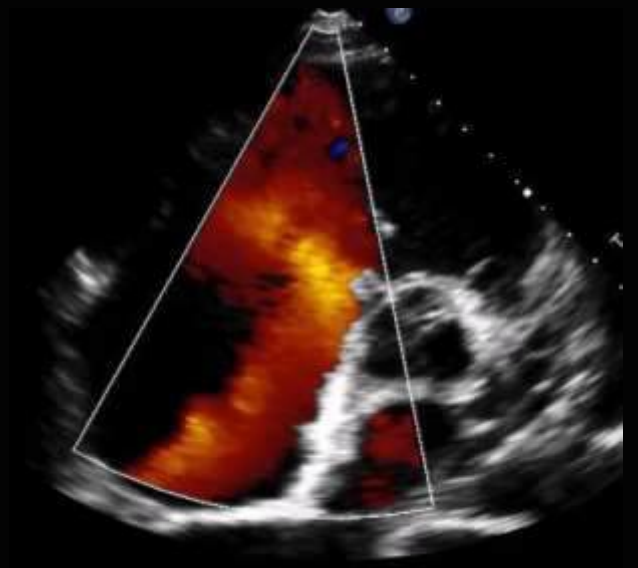
Severe Ebstein anomaly



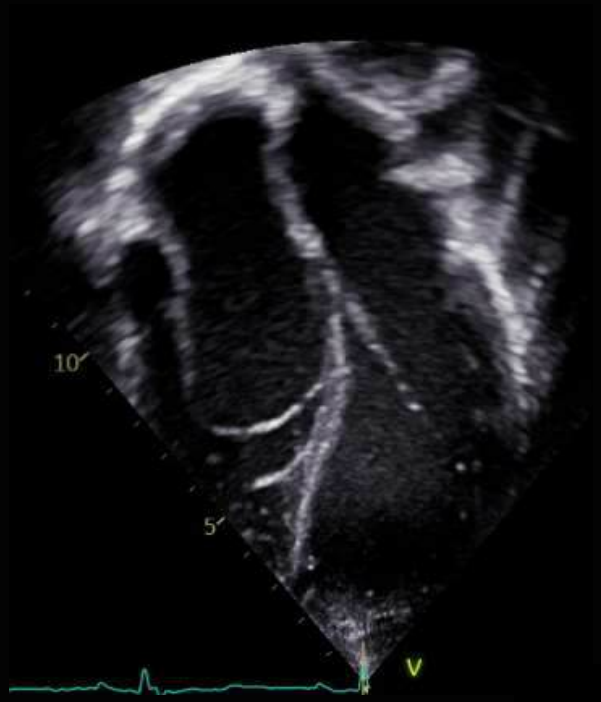


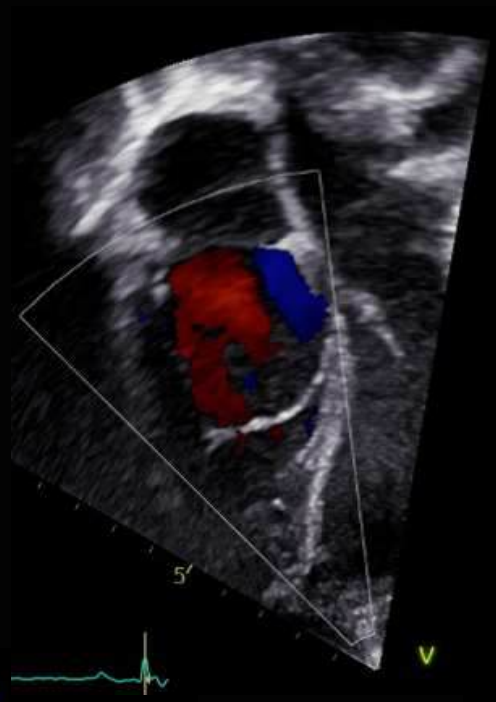


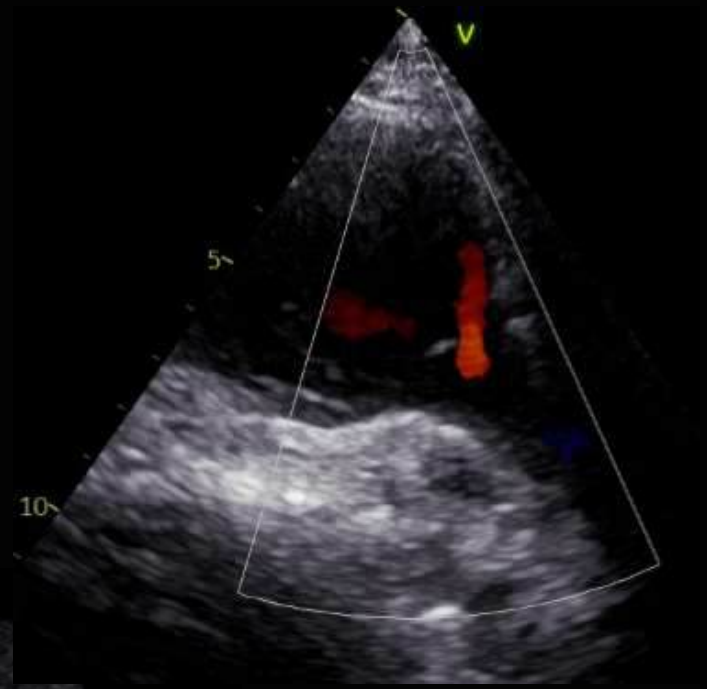


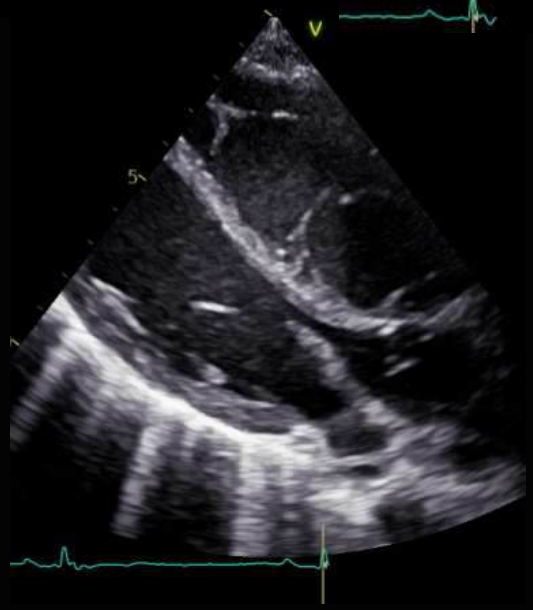


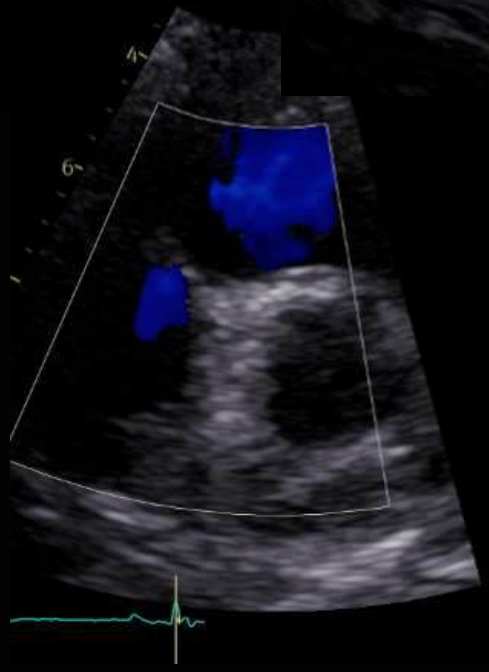
11-year-old girl, pre-natal dx, doing well never had intervention





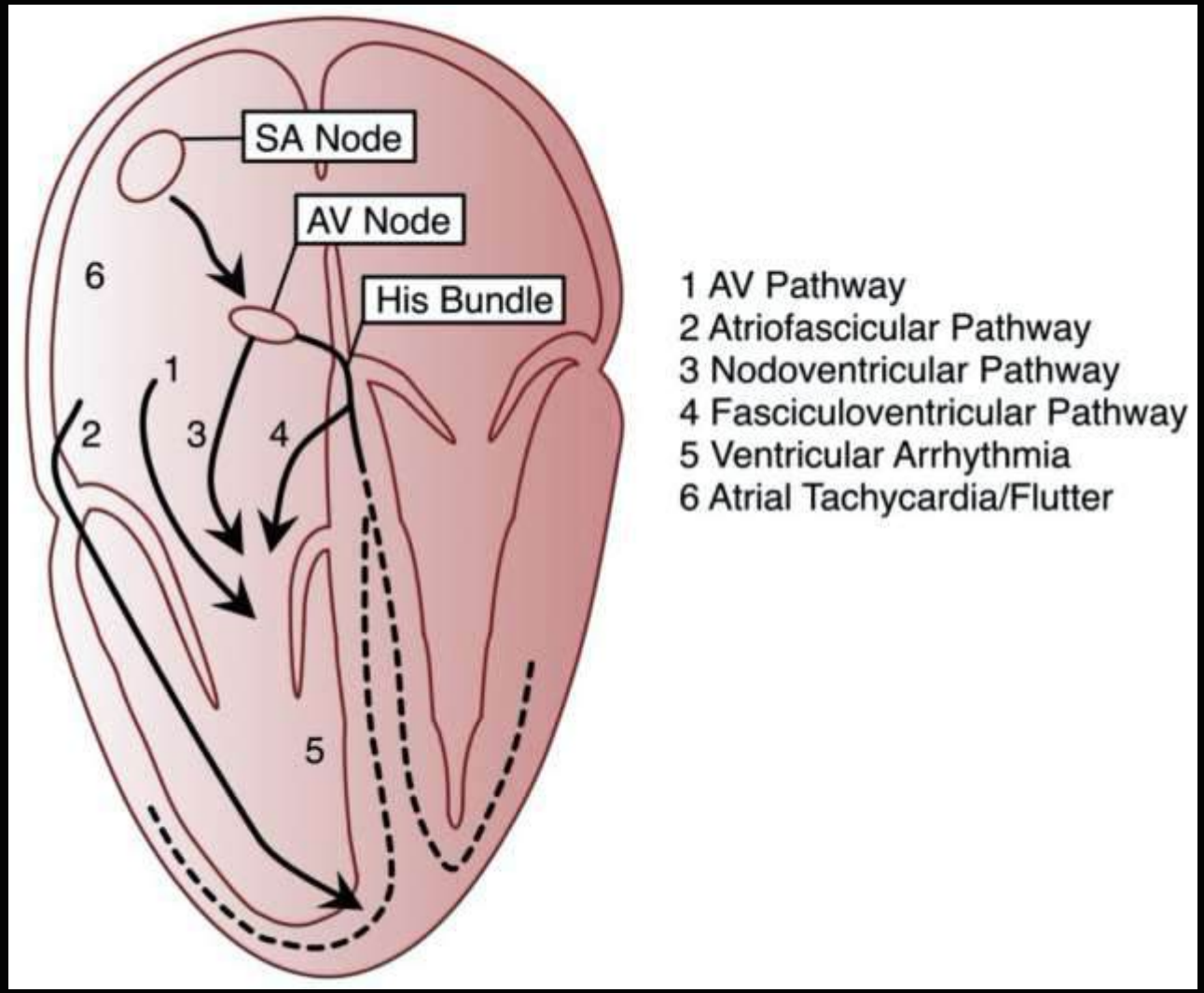






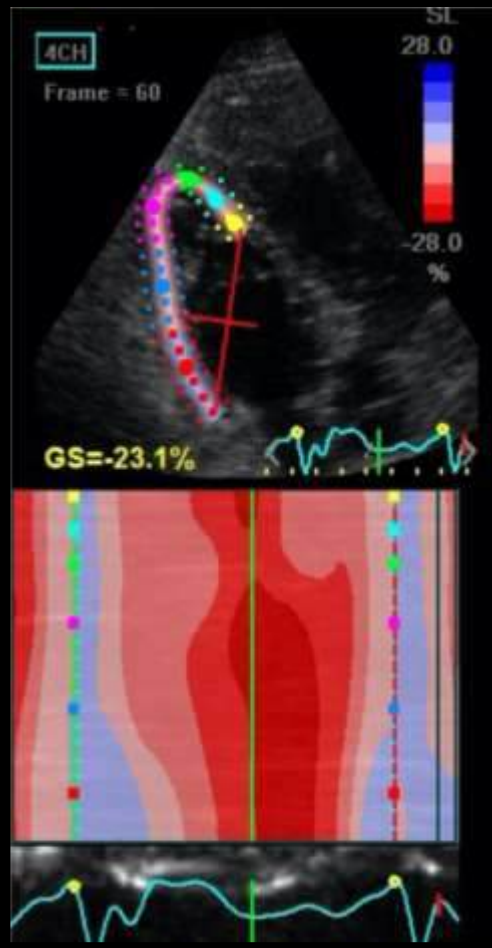
Conduction abnormalities in Ebstein anomaly

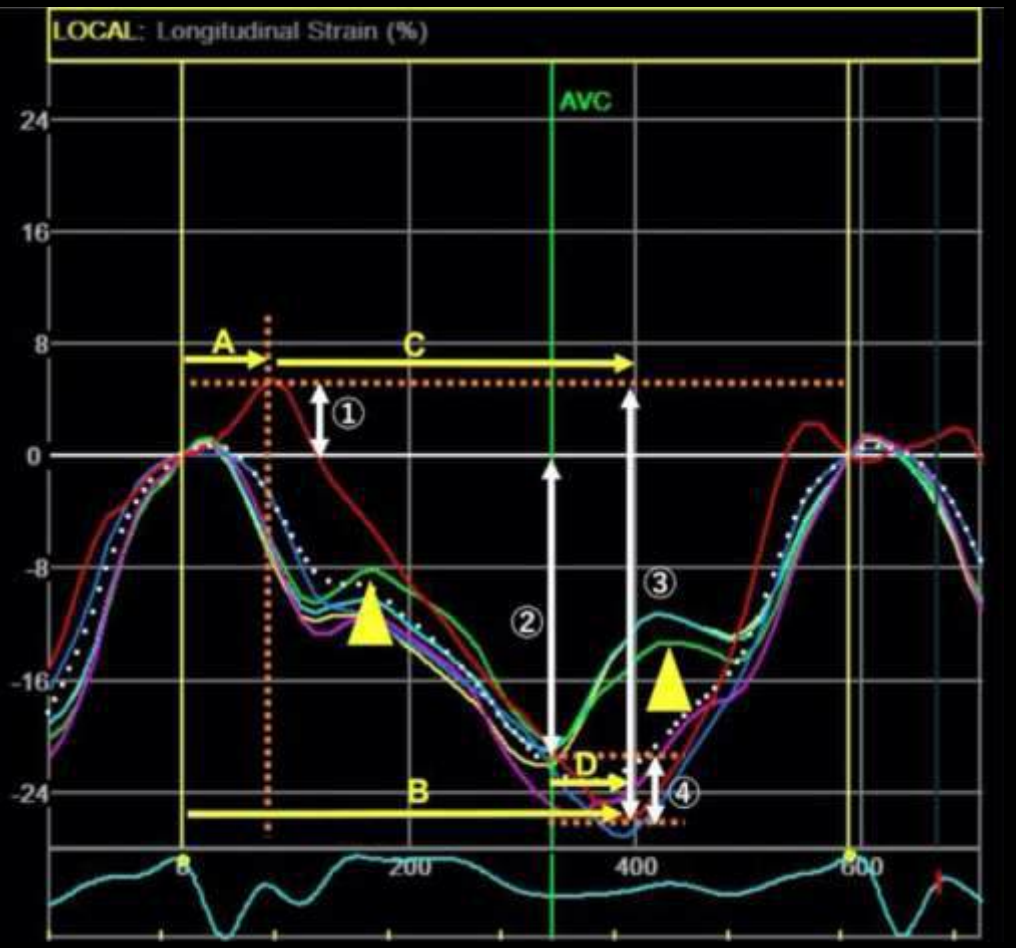
- Delayed intraatrial conduction
- Accessory pathways, ventricular preexcitation, SVT
- RBBB and variable QRS morphologies
- Electromechanical dyssynchrony
- Atrial and ventricular tachycardias

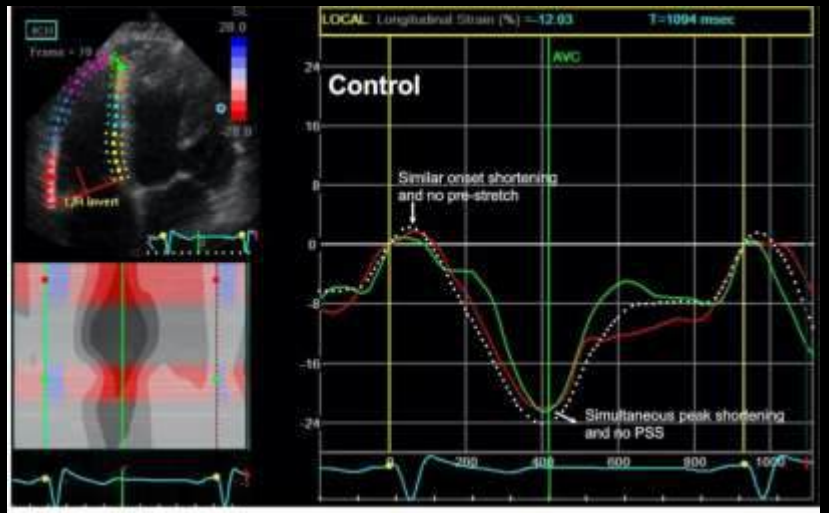


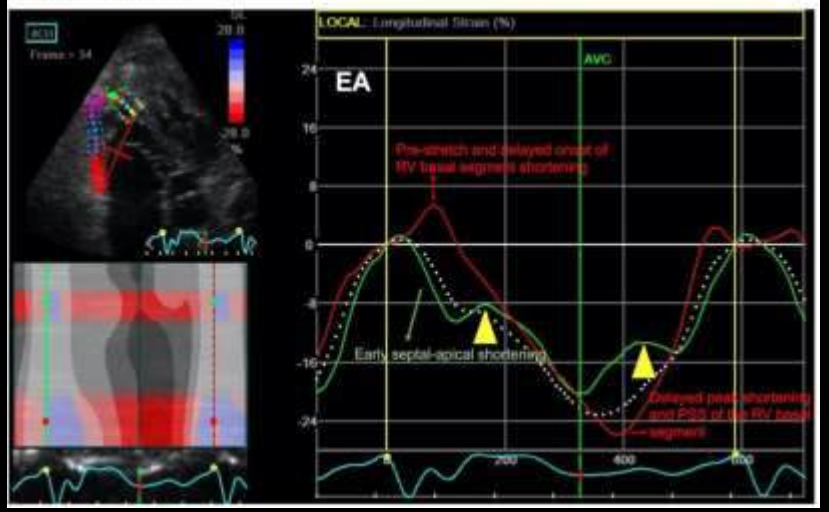
Right Ventricular Electromechanical Dyssynchrony and Its Relation to Right Ventricular Remodeling, Dysfunction, and Exercise Capacity in Ebstein Anomaly

Yohei Akazawa, MD, PhD, Tao Fujioka, MD, PhD, Kana Yazaki, MD, PhD, Martina Strbad, MSc, Jürgen Hörer, MD, PhD, Andreas Kühn, MD, Wei Hui, MD, Cameron Slorach, RDCS, Christoph Roehlig, MD, Luc Mertens, MD, PhD, Bart H. Bijnens, PhD, Manfred Vogt, MD, PhD, and Mark K. Friedberg, MD, *Toronto, Ontario, Canada; Munich, Germany; and Barcelona, Spain*









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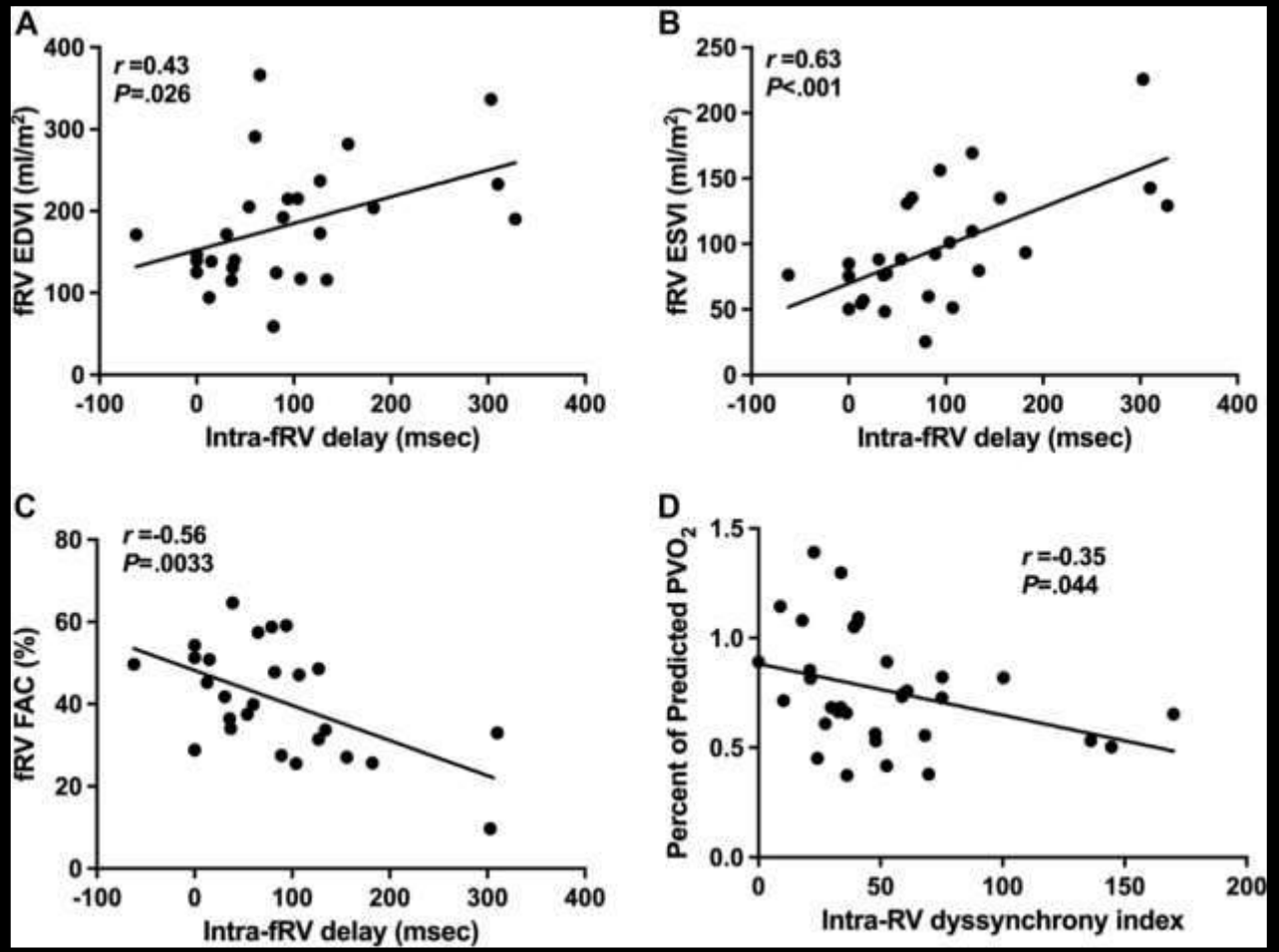
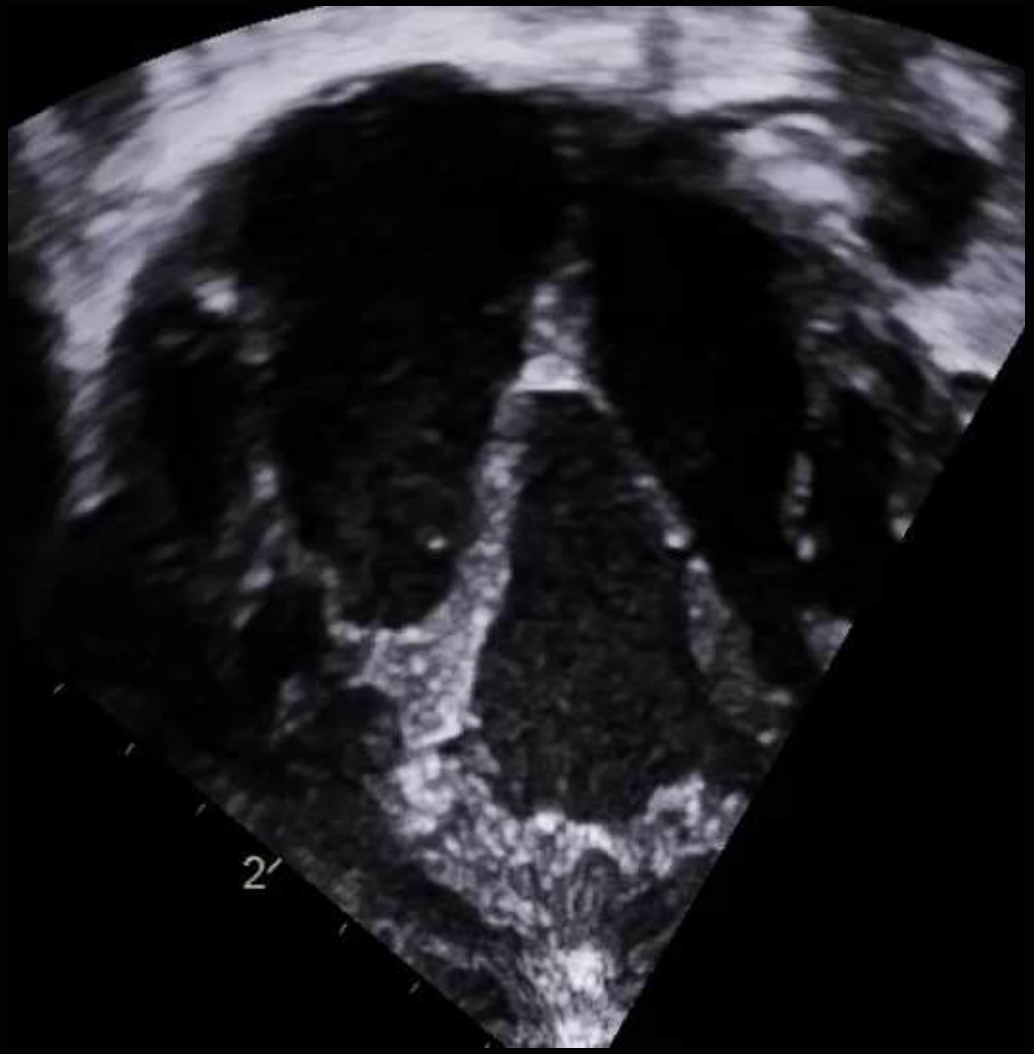
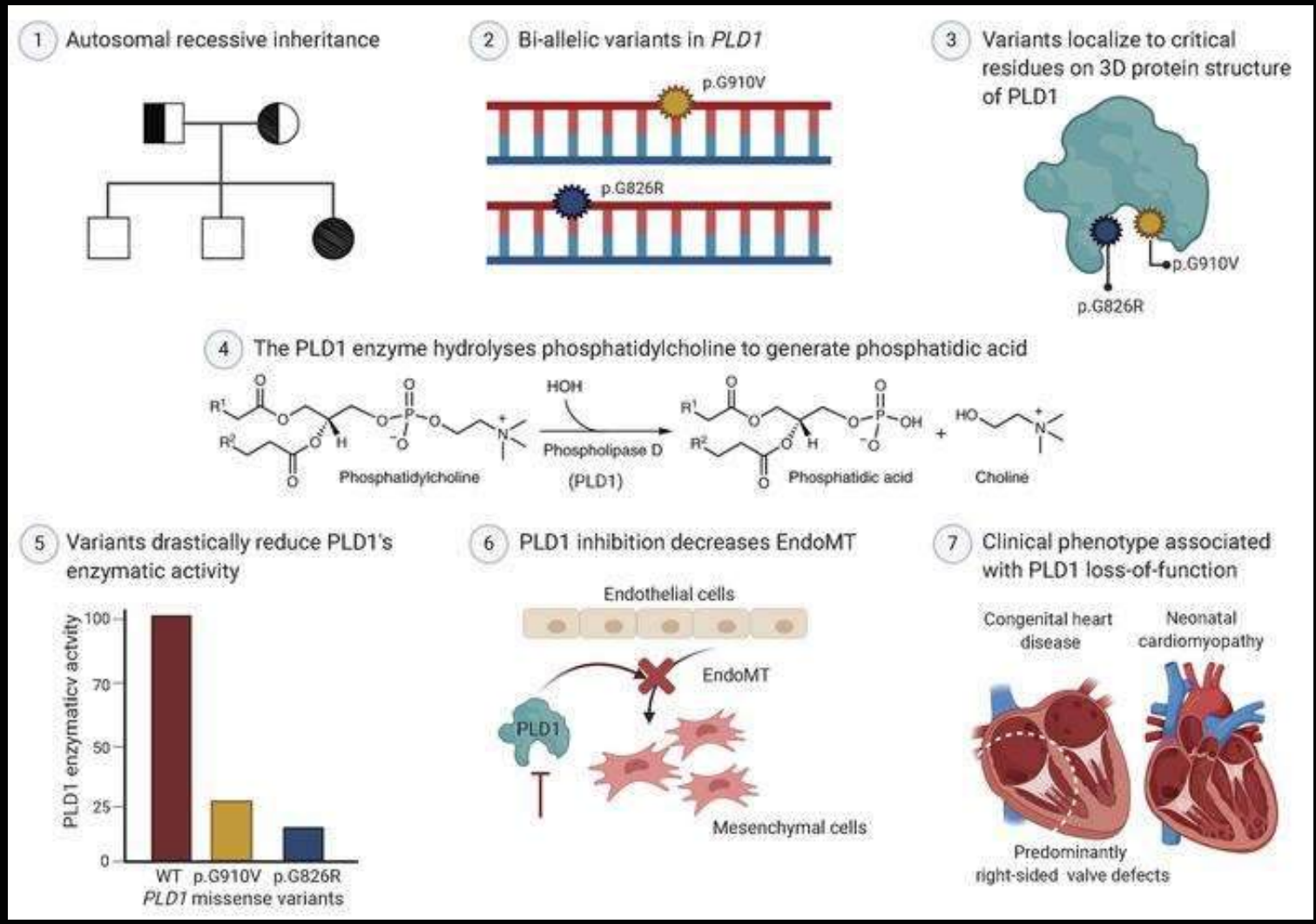


Ebstein anomaly is a myopathy: RV dysfunction and LV non-compaction

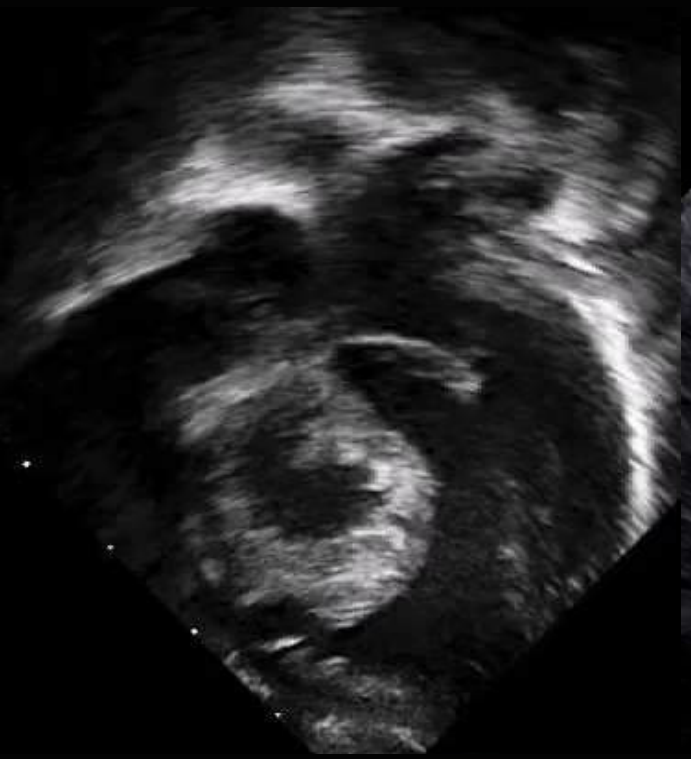


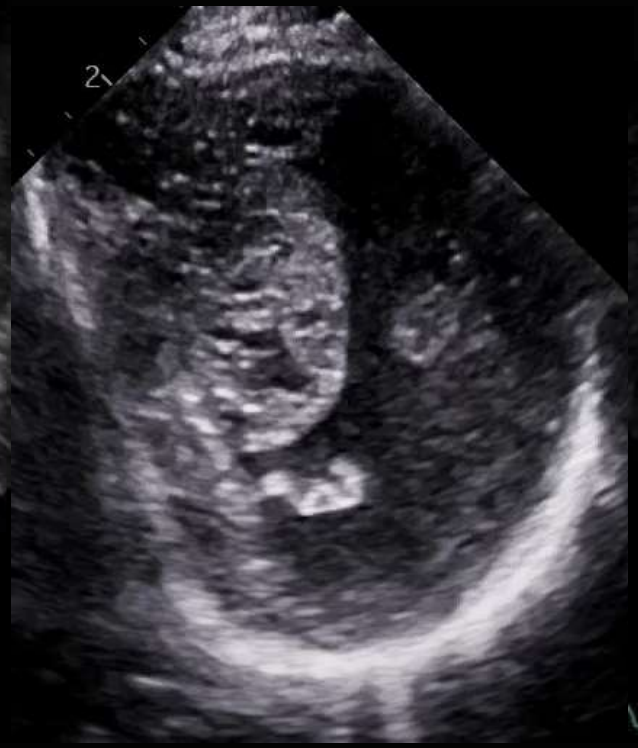
Biallelic loss-of-function variants in PLD1 cause congenital right-sided cardiac valve defects and neonatal cardiomyopathy

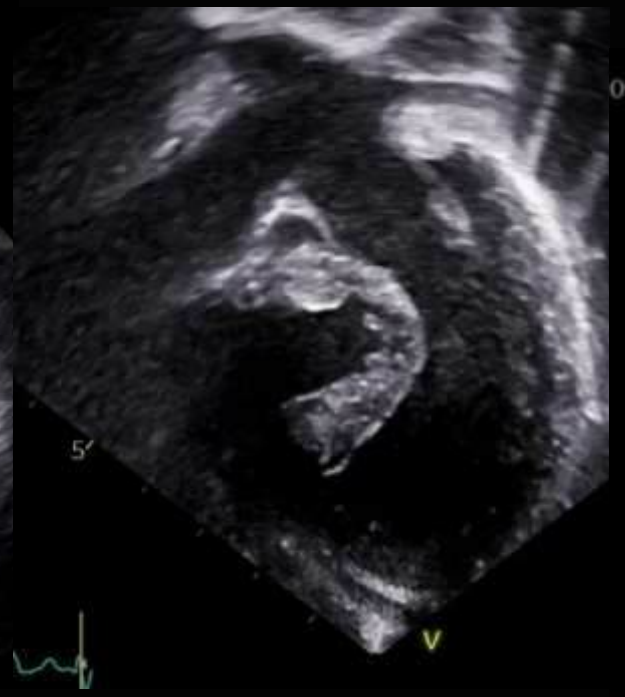


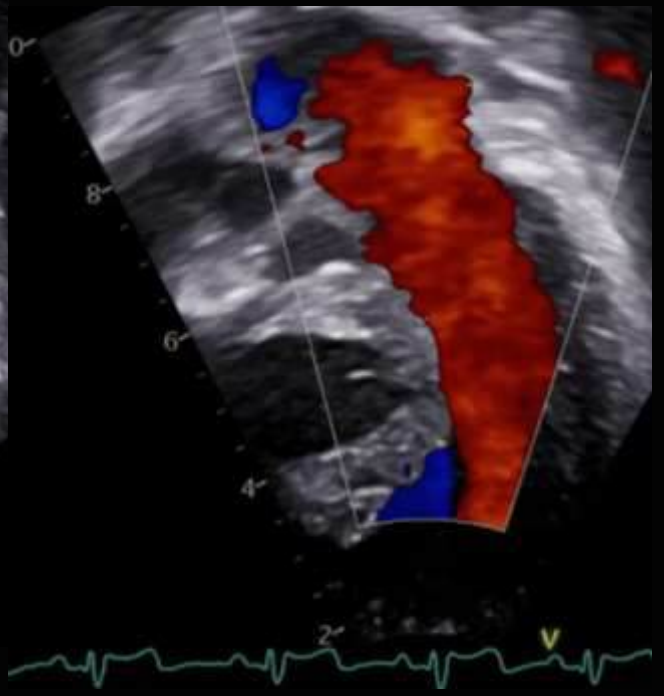
J Clin Invest. 2021;131(5):e142148. https://doi.org/10.1172/JCI142148

PLD1 and Ebstein anomaly with RV-LV interaction

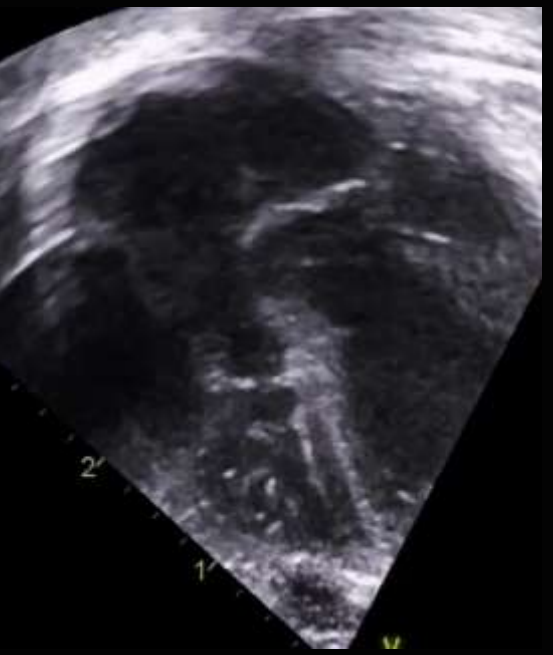


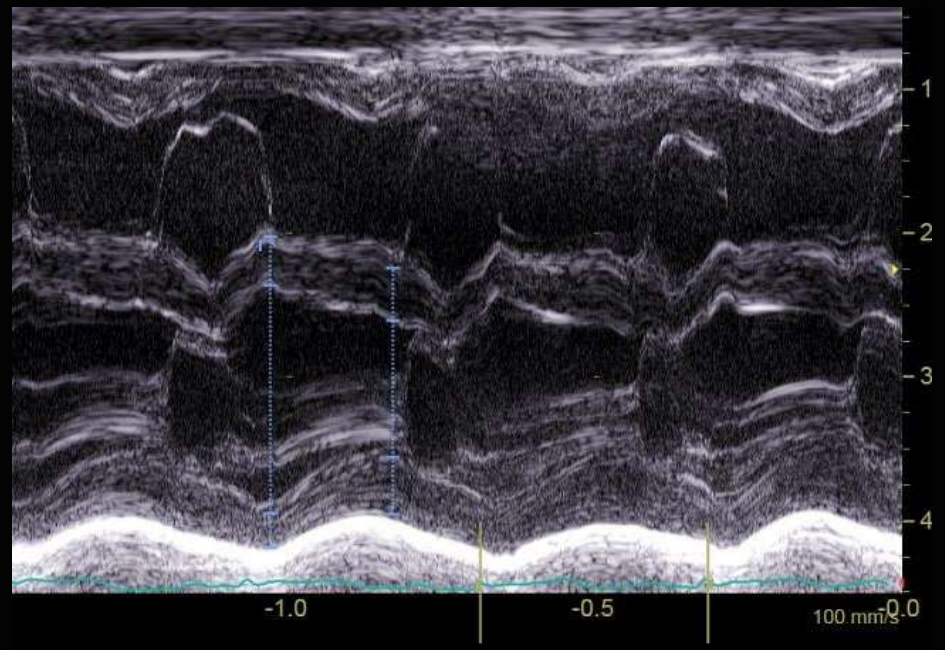


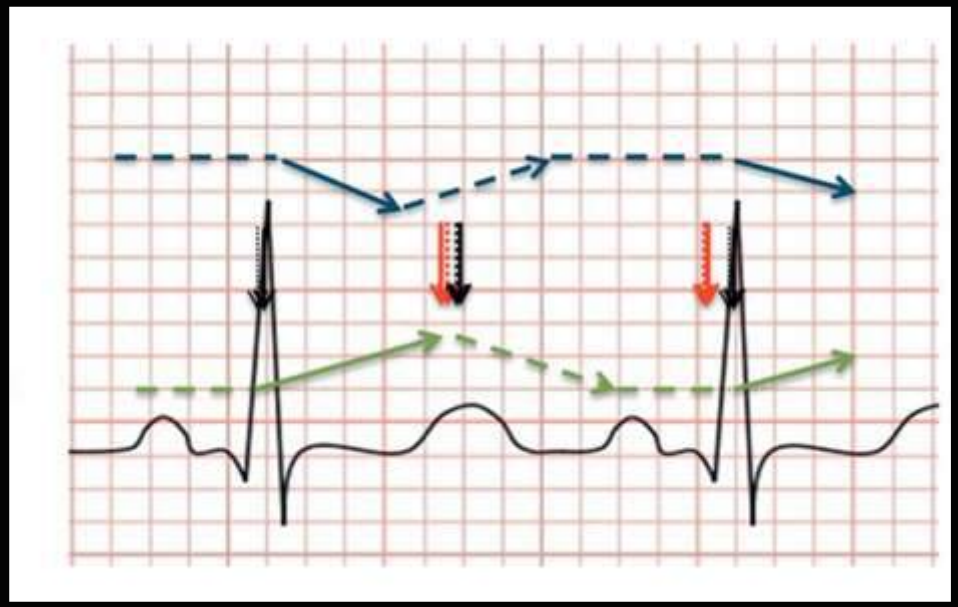


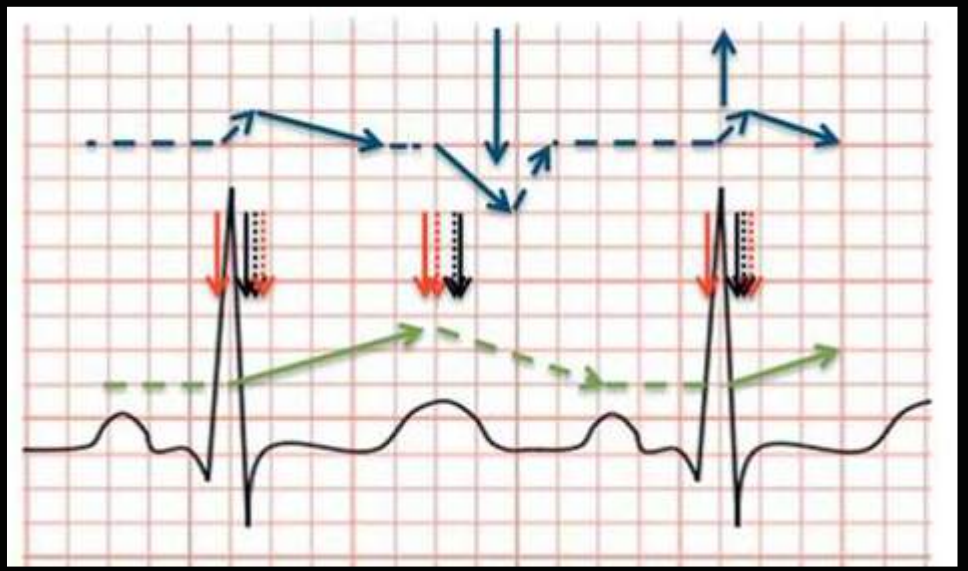


Good RV function, but adverse RV-LV interactions









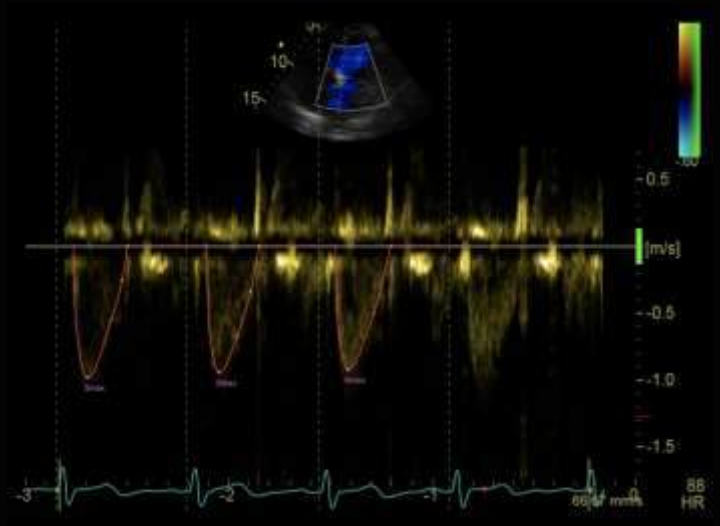
Adverse ventricular–ventricular interactions mediated by abnormal septal kinetics and decreased RV output impede LV filling and function in association with reduced exercise capacity.

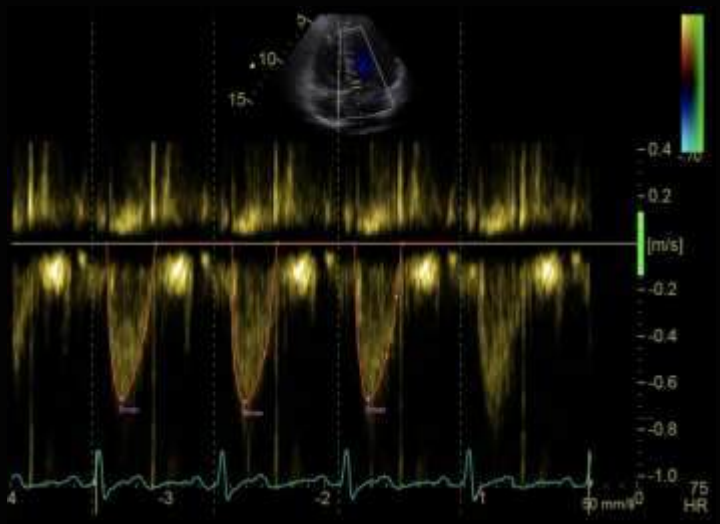
JACC Cardiovasc Imaging. 2019;12:925-927

Aortic and pulmonary outflows

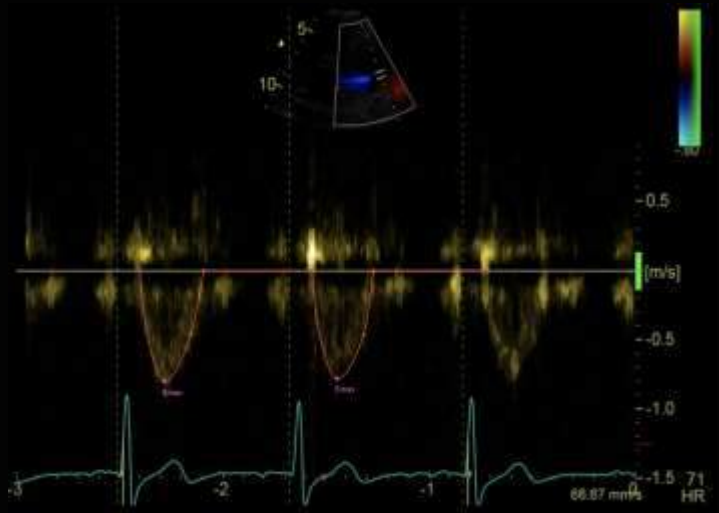
$N_{Control} = 63$
$N_{Ebstein} = 61$

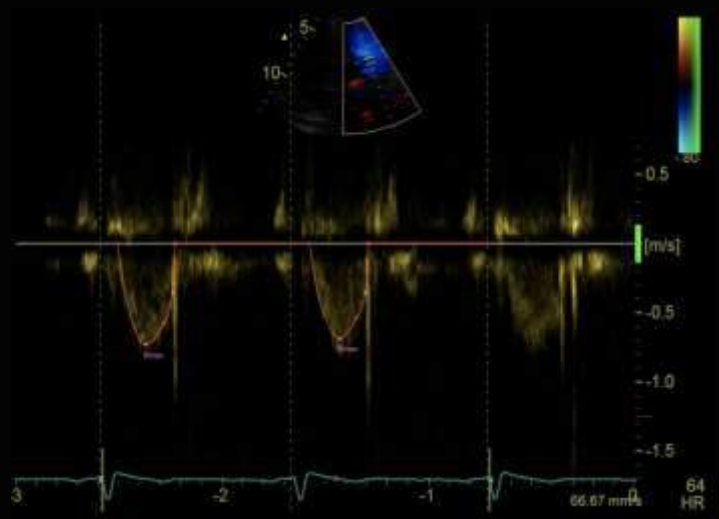
Aortic outflow





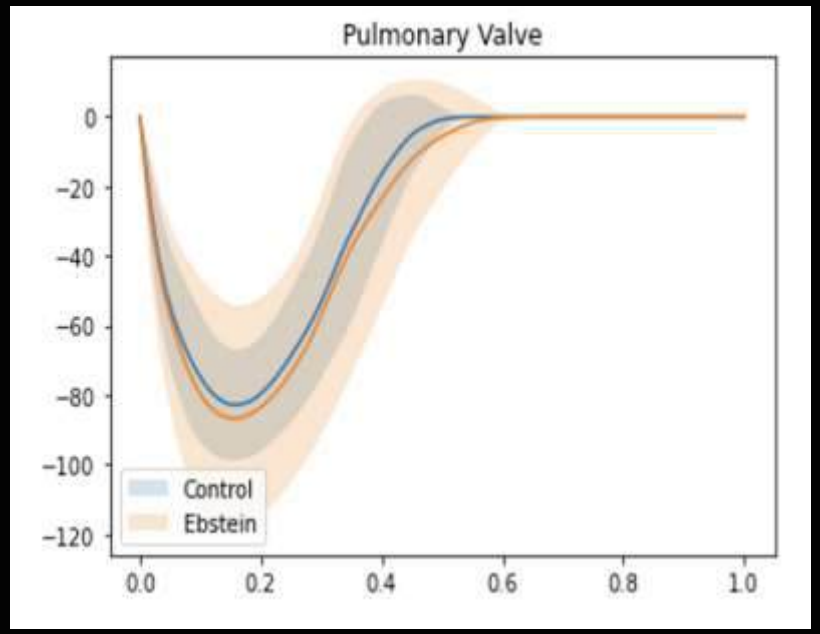
Pulmonary outflow

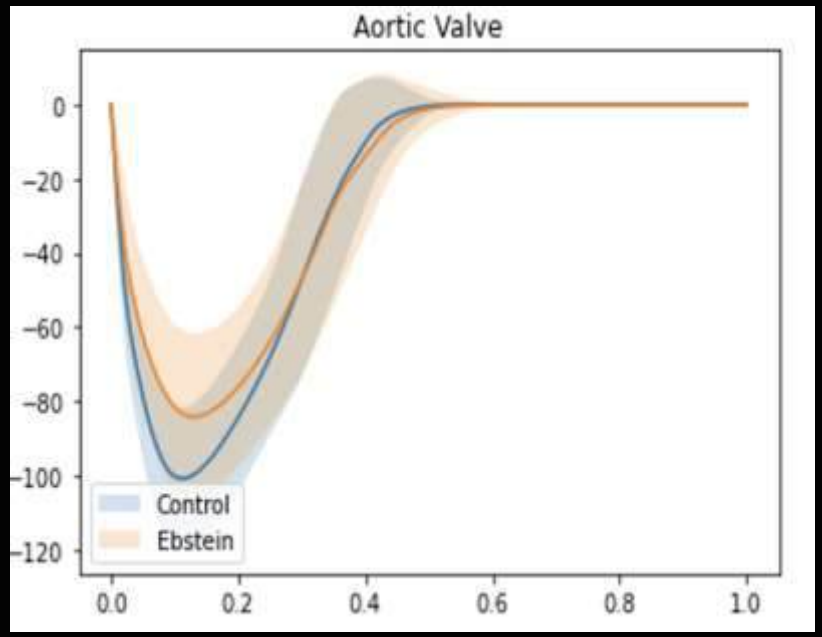




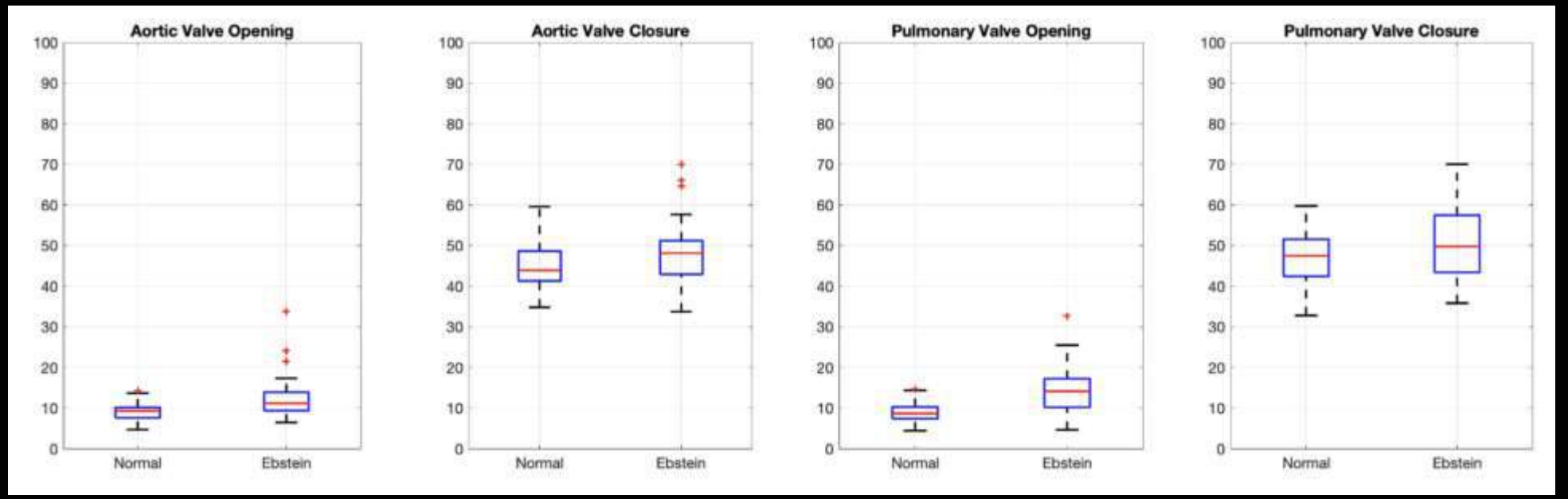
Outflow Doppler summary

$N_{Control} = 63$
$N_{Ebstein} = 61$



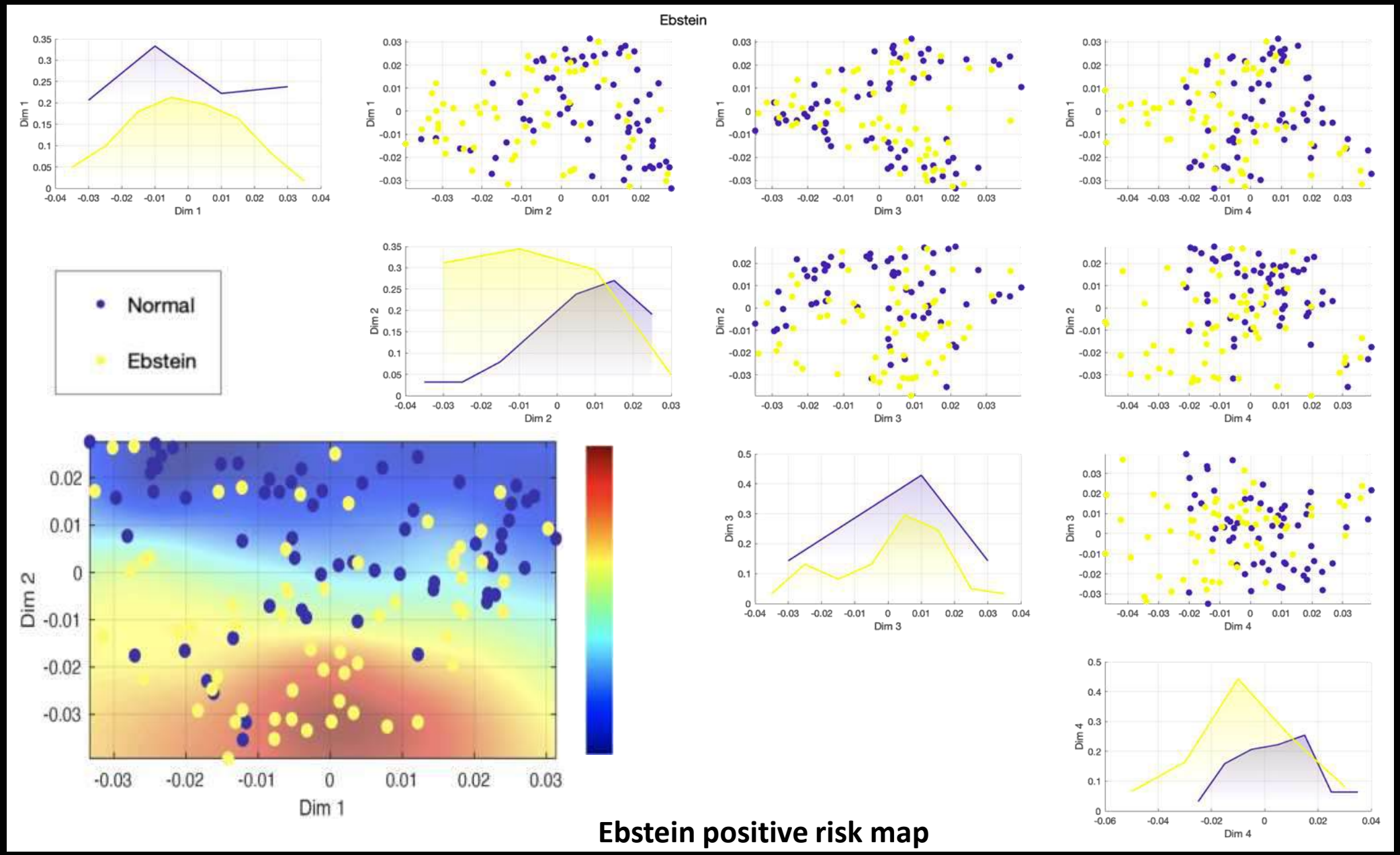


Timing



MKL on raw traces + timing information

Distribution of Ebstein and controls along different dimensions of the simplified data representation

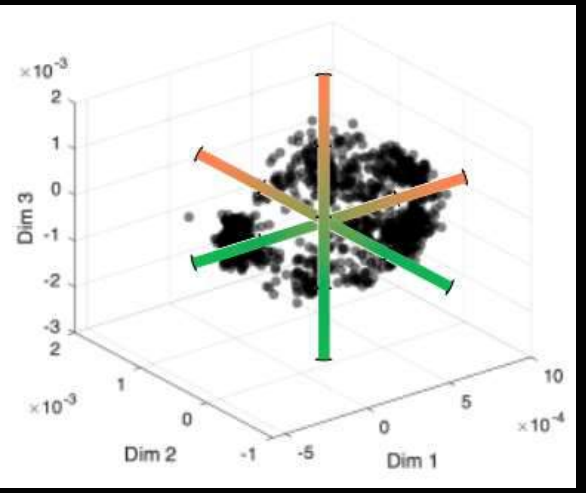


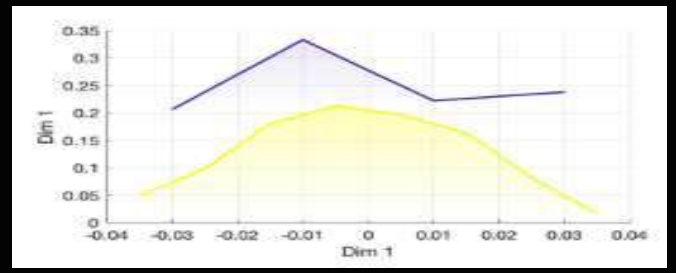
Ebstein positive risk map

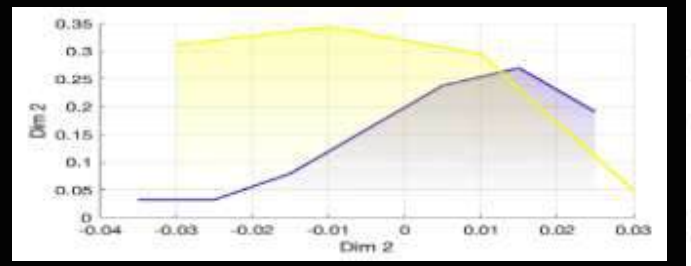
Interpretation of the simplified data representation

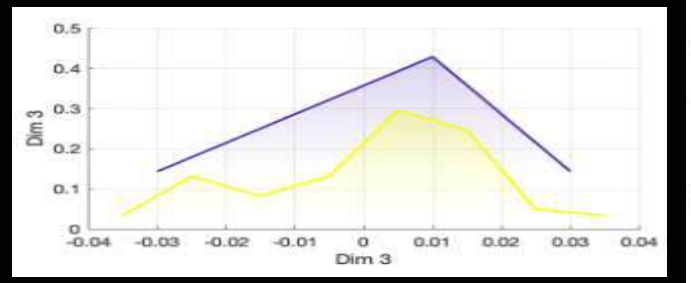
Normal

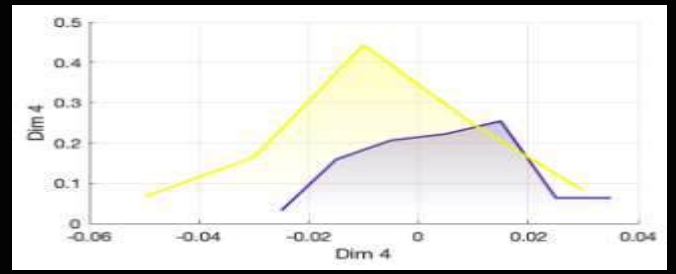
Ebstein

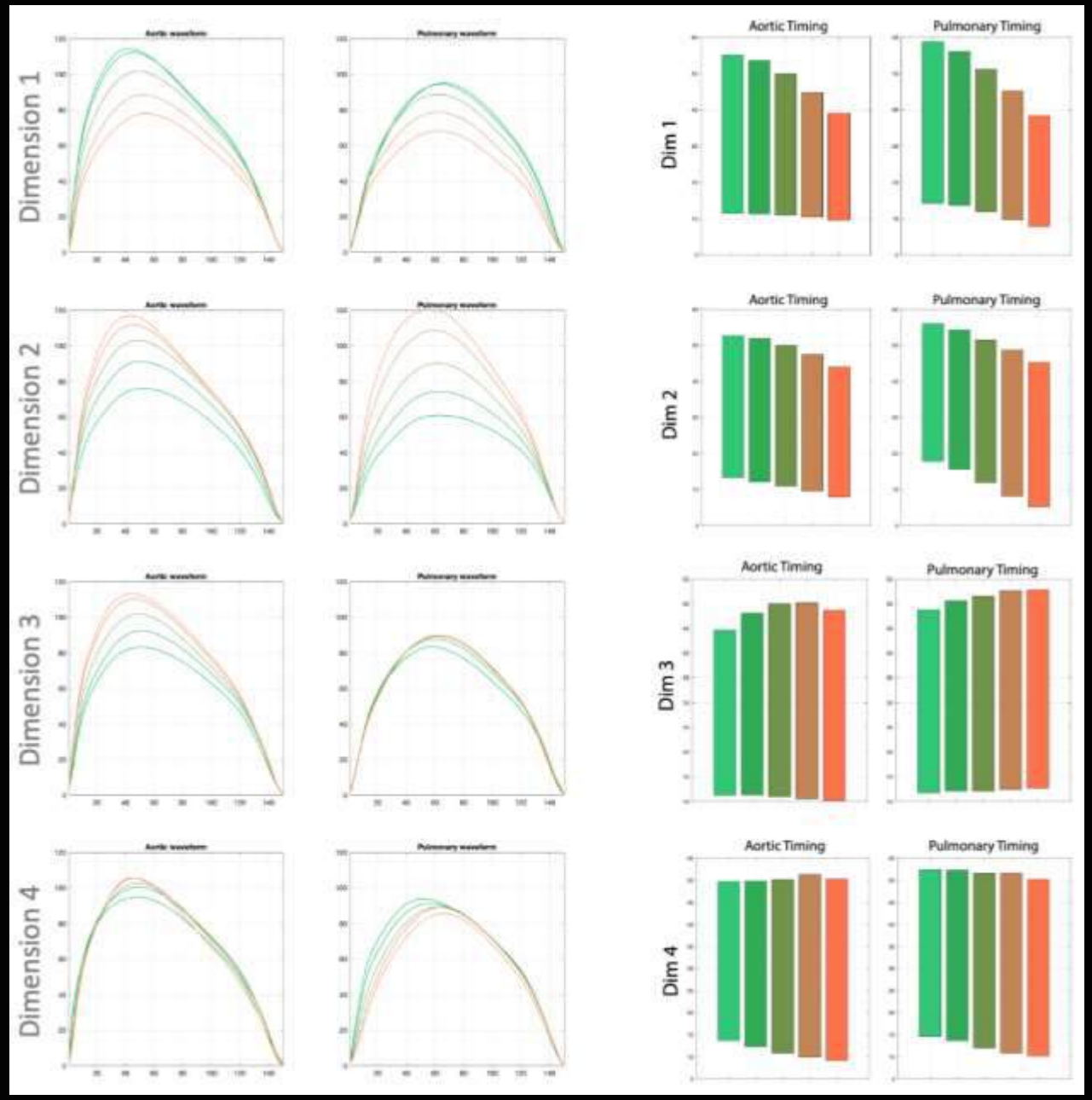








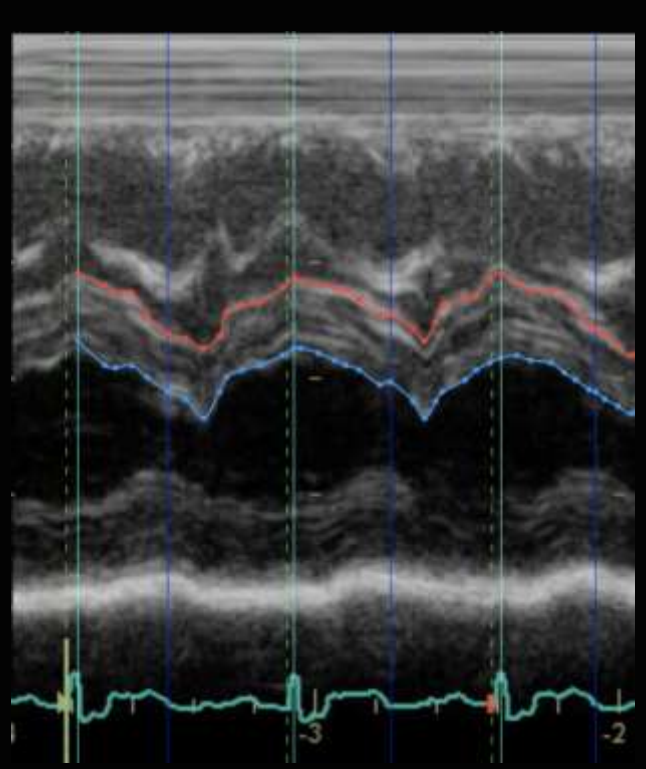


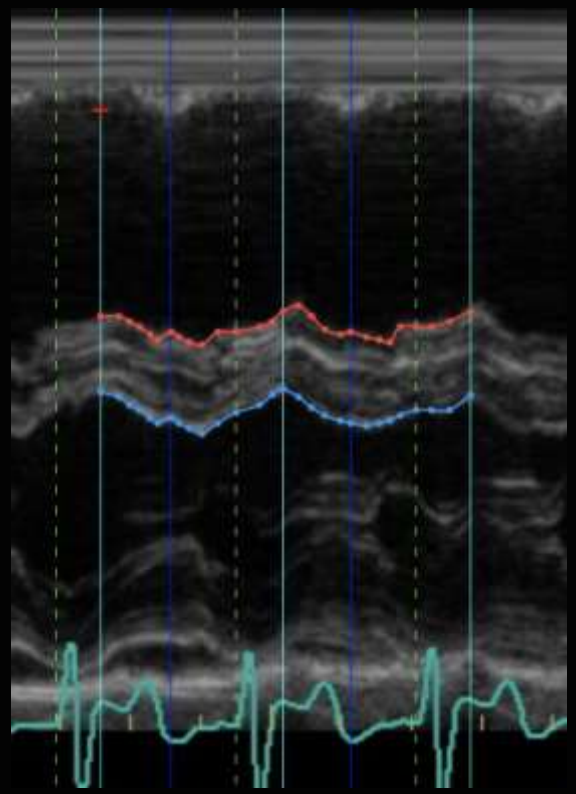


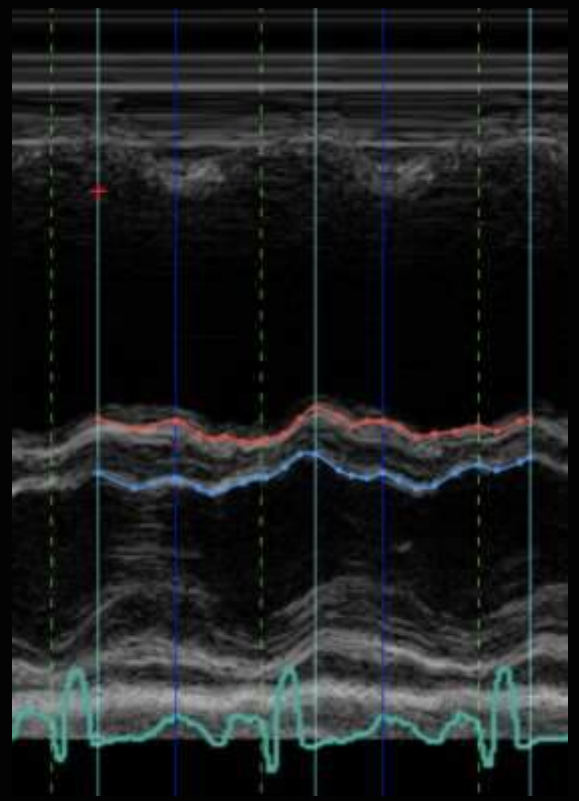
Interventricular interactions

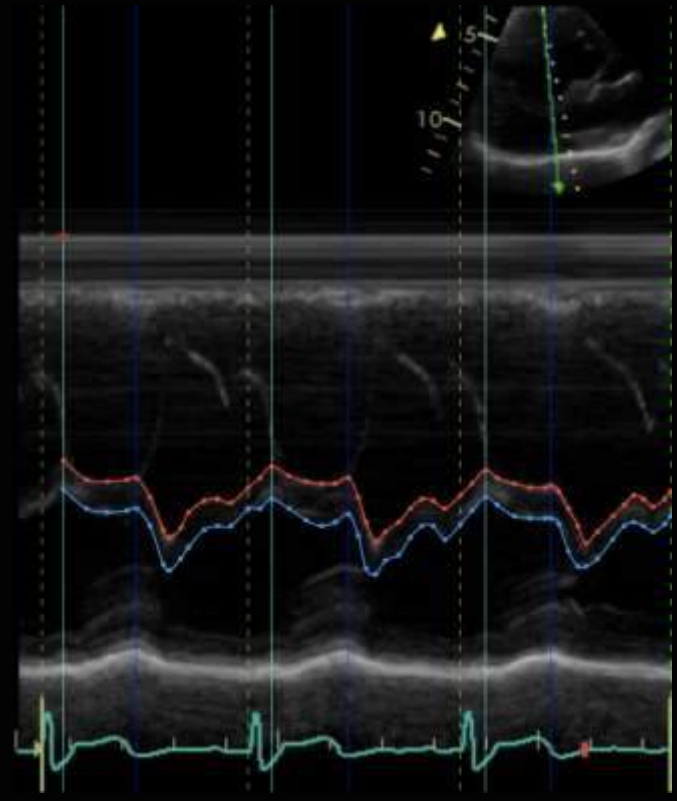
Only Ebstein cases – N = 48

- Aortic and pulmonary waveforms + timing
- LV / RV Septum M-mode waveform + timing (HR, end-diastole, end-systole)



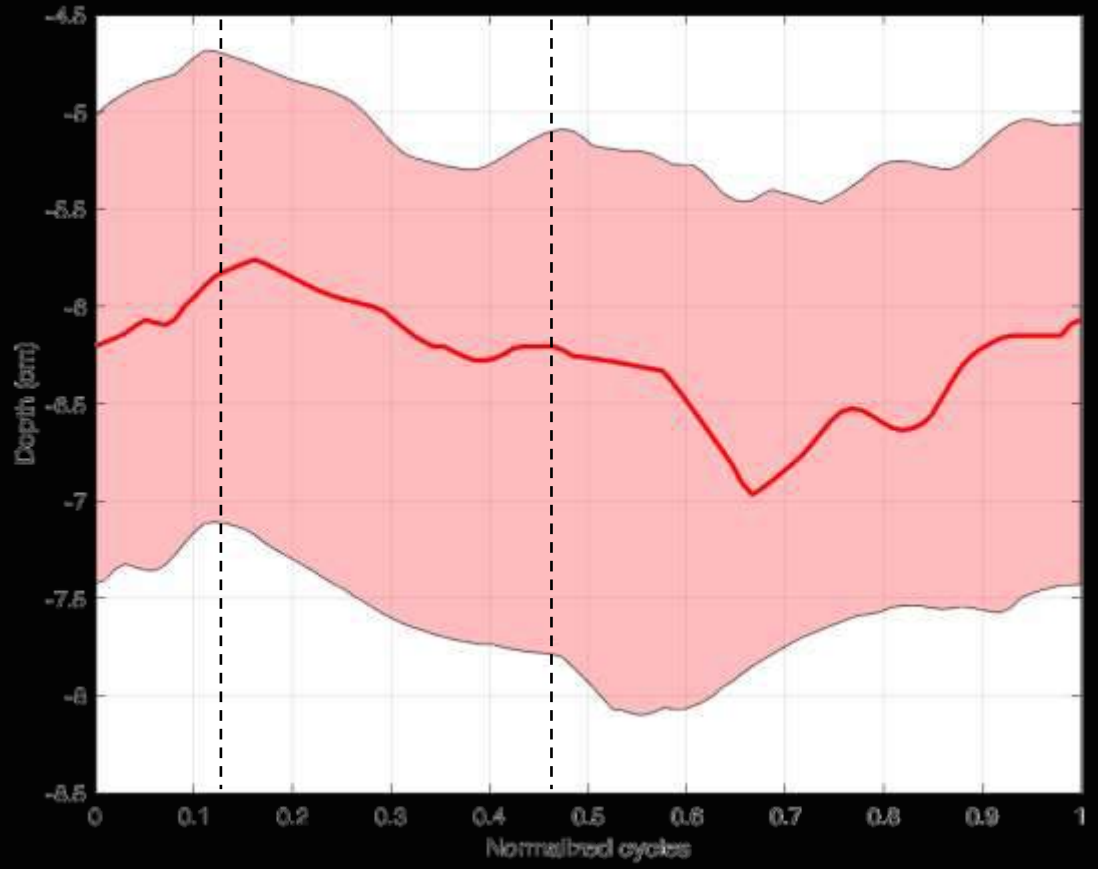




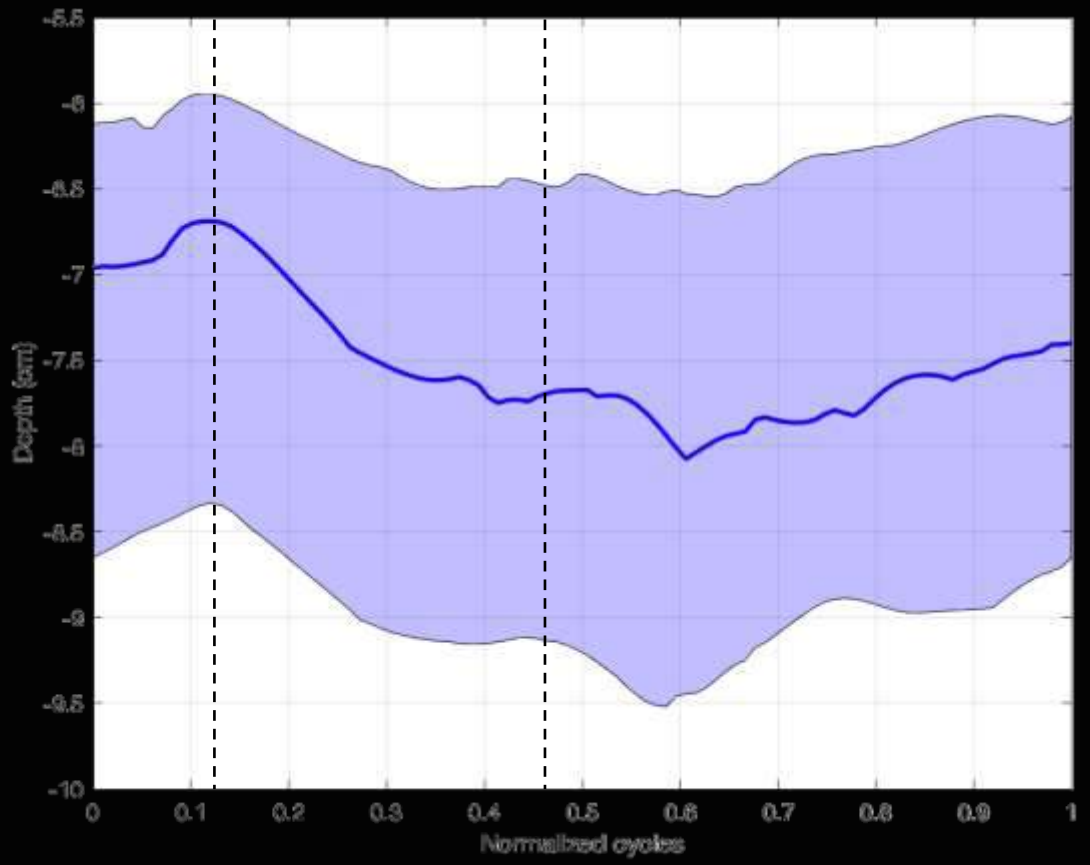


Summary of septal motion

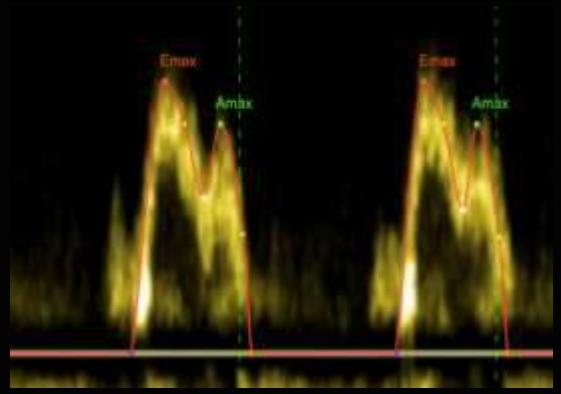
RV Septal M-Mode

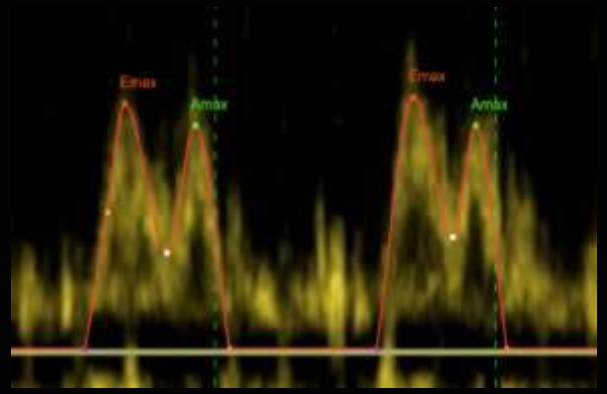


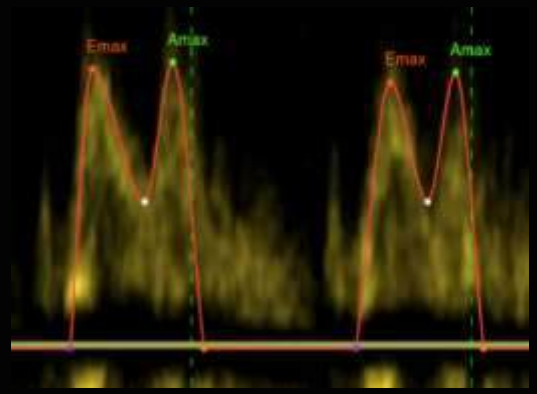
LV Septal M-Mode

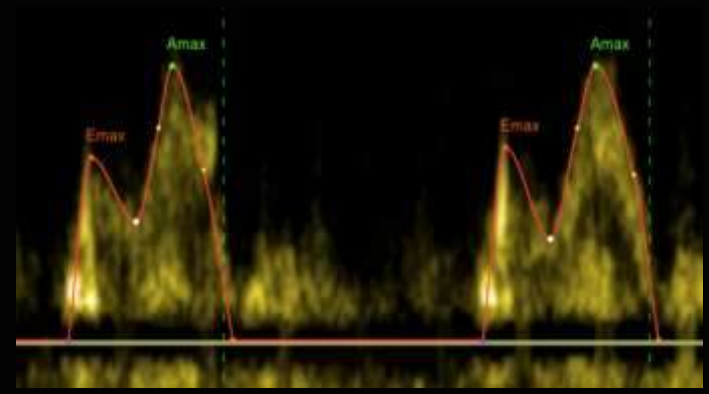


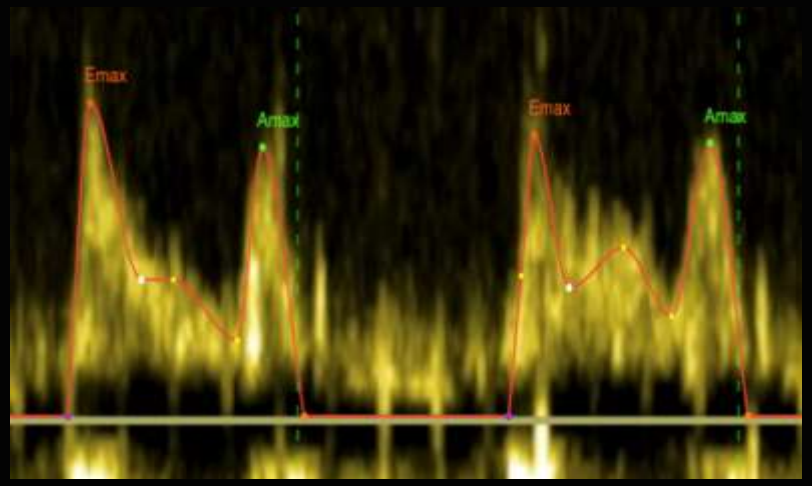
Mitral inflow in Ebstein Anomaly

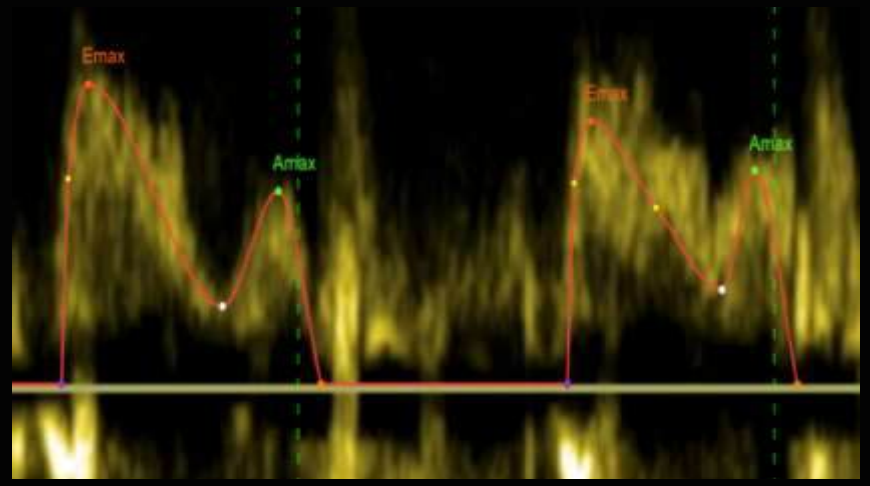


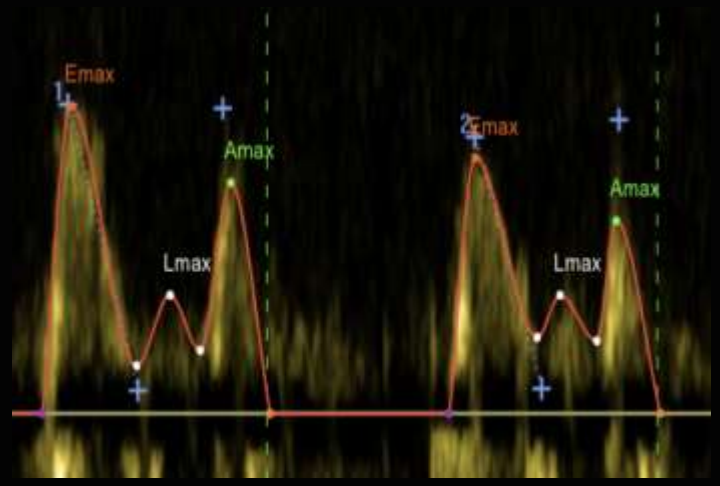






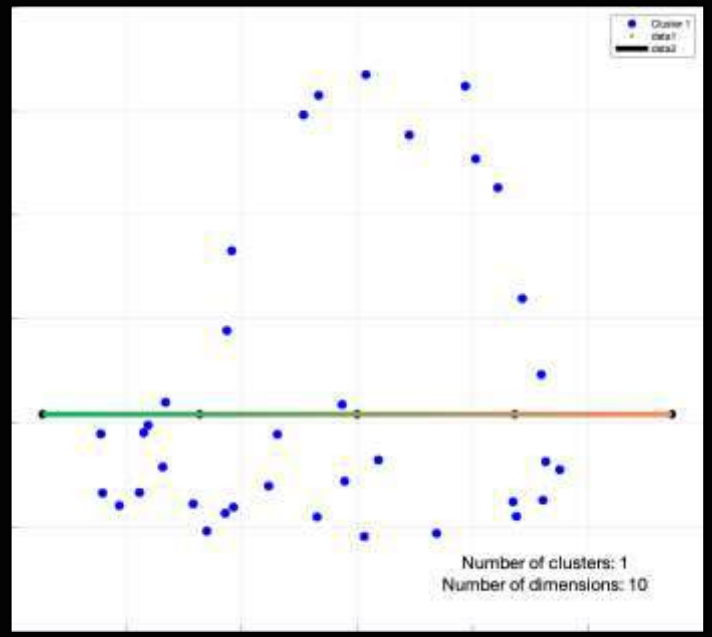




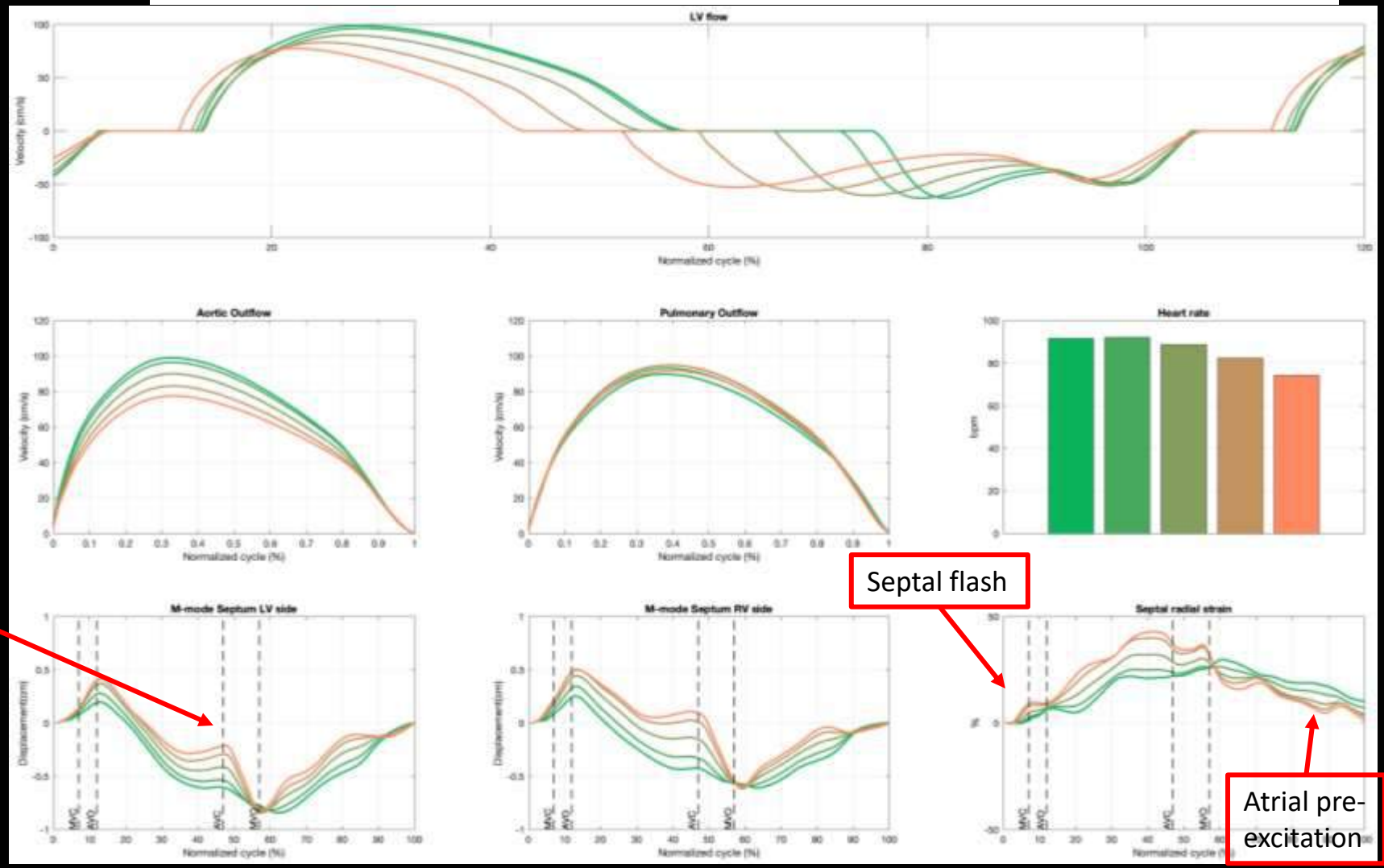


Machine learning Dimension 1

	-2 STD	-1 STD	mean	1 STD	2 STD
VO2 (mL/(kg·min))	30.7	31.4	29.5	25.8	22.5
LVEF (%)	61.4	63.2	63.3	62.3	60.0
QRS width (ms)	144	151	153	150	142
Age (years)	27.7	28.4	29.0	30.2	31.7

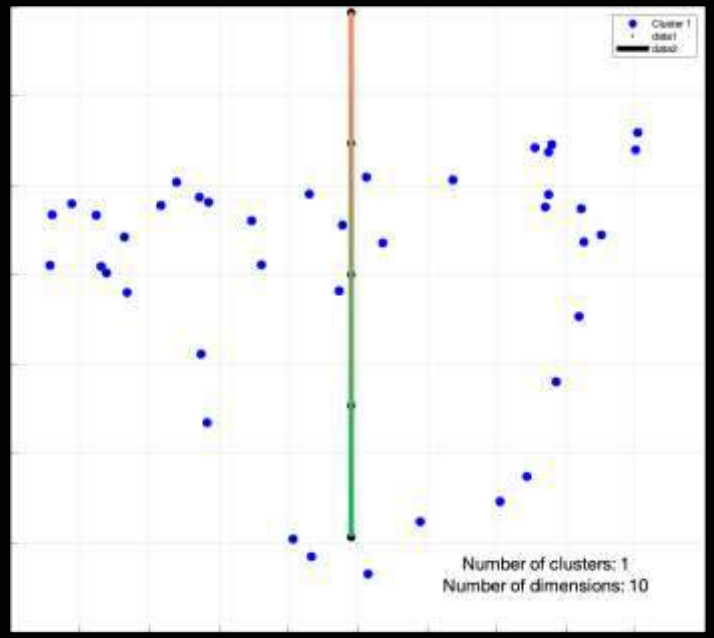


Right – Left Heart Interaction

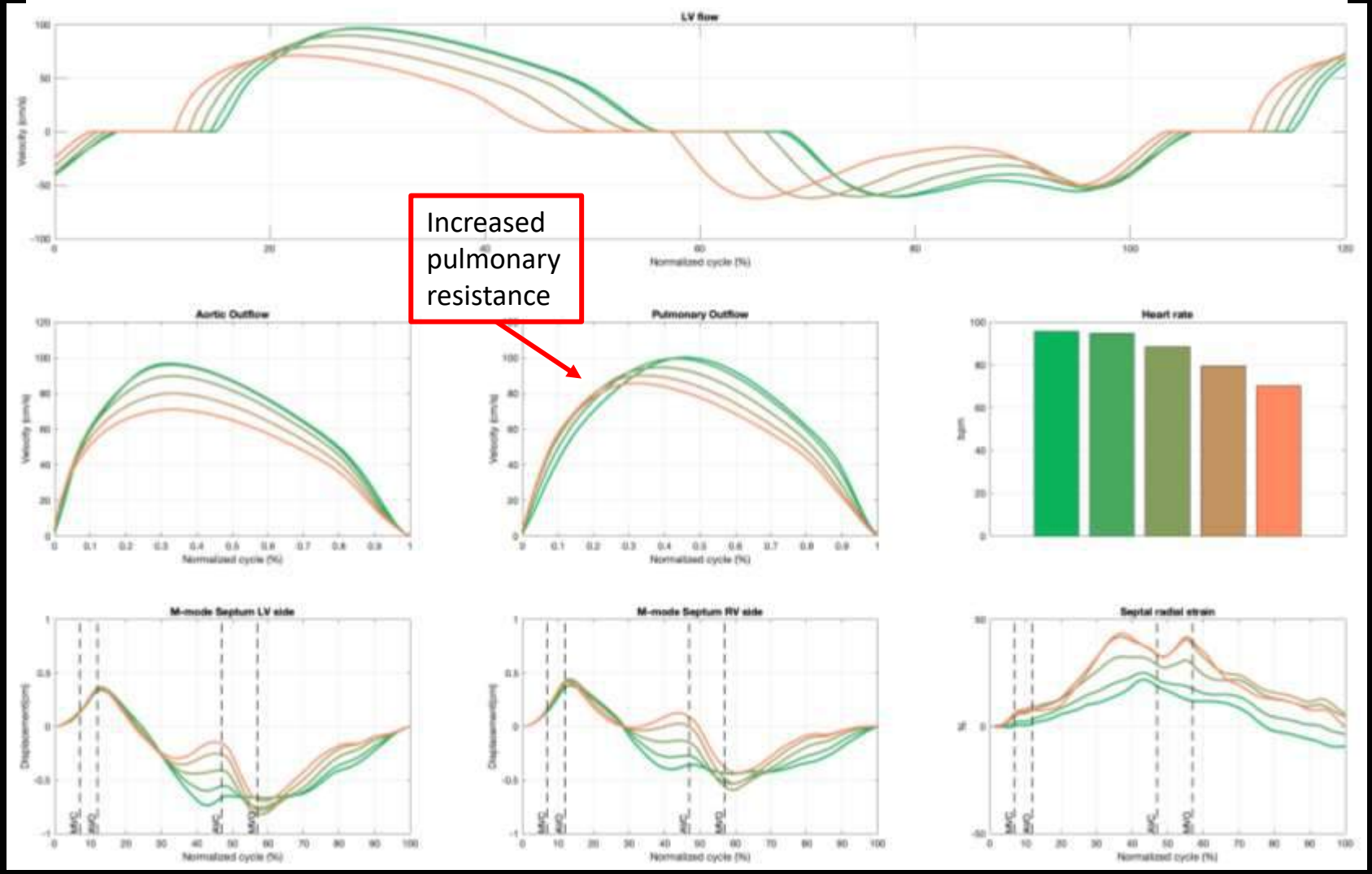


Machine learning Dimension 2

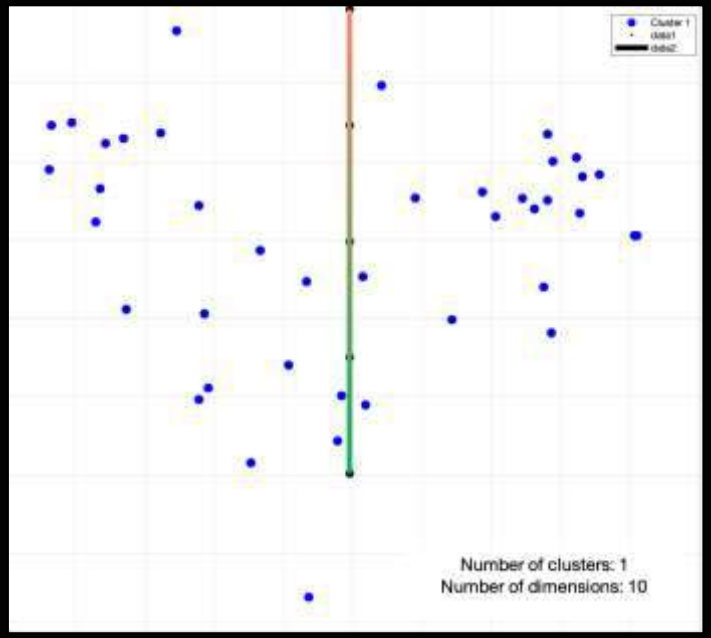
	-2 STD	-1 STD	mean	1 STD	2 STD
VO2 (mL/(kg·min))	31.5	31.0	29.5	28.2	26.2
LVEF (%)	61.7	63.7	63.3	61.5	58.6
QRS width (ms)	131	143	154	155	145
Age (years)	29.5	29.4	29.0	30.0	31.5



Increased RV pressure

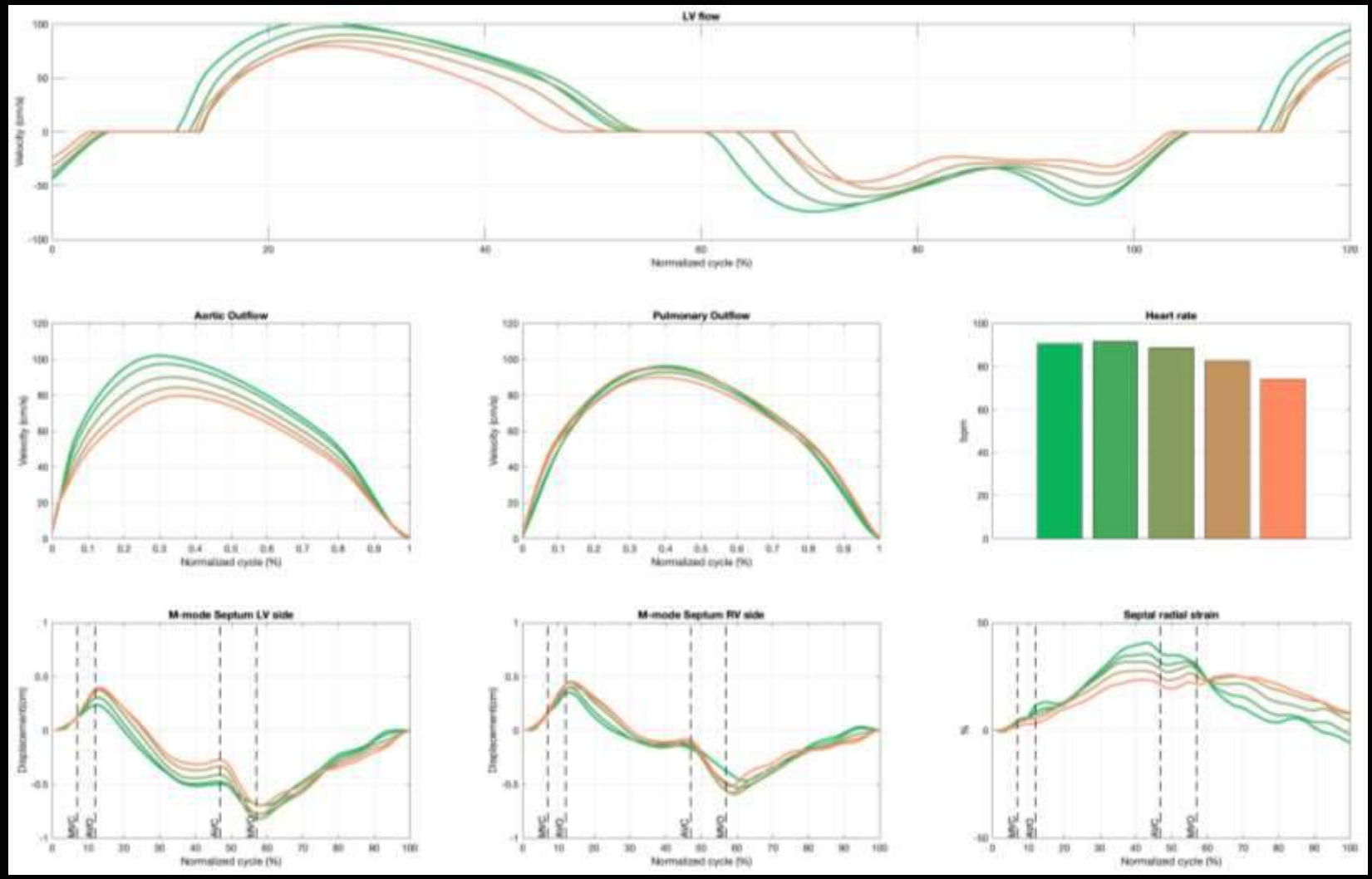


Machine learning Dimension 3

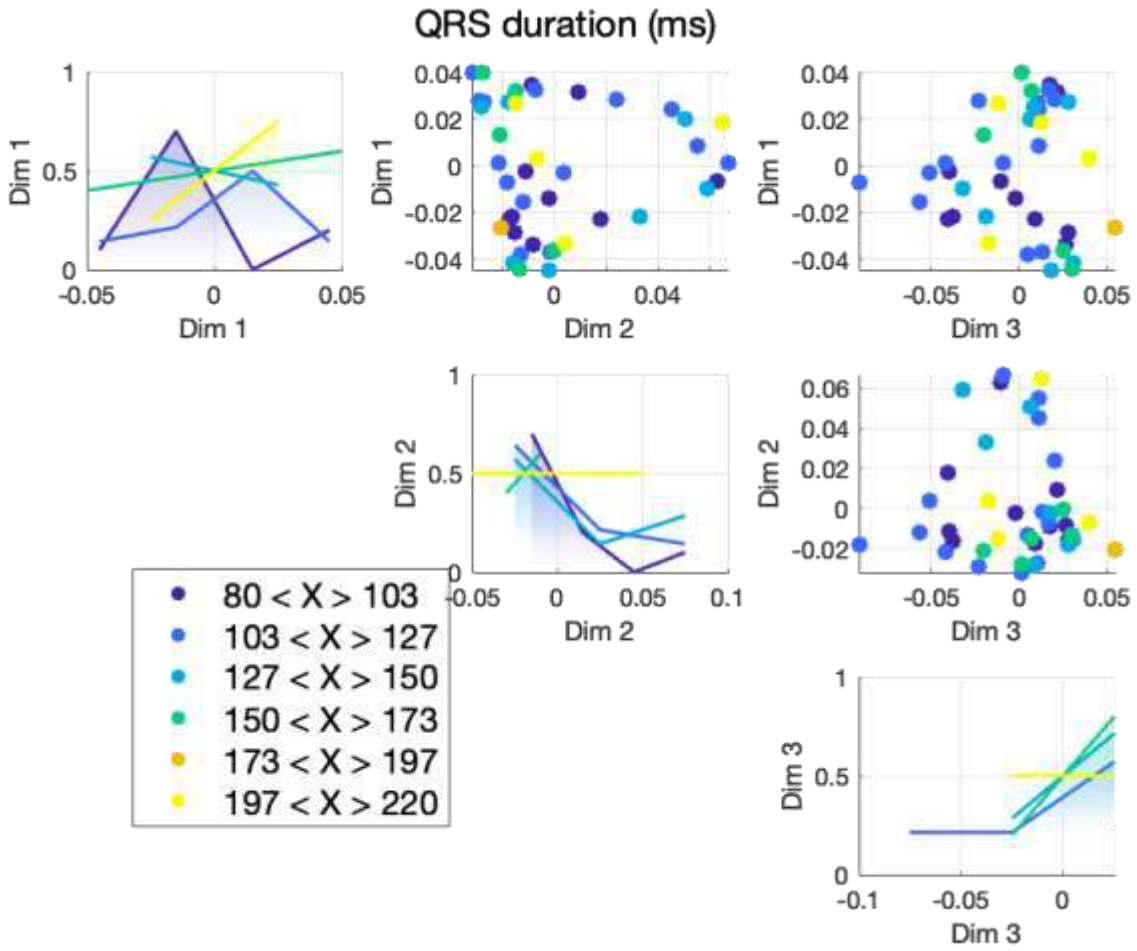


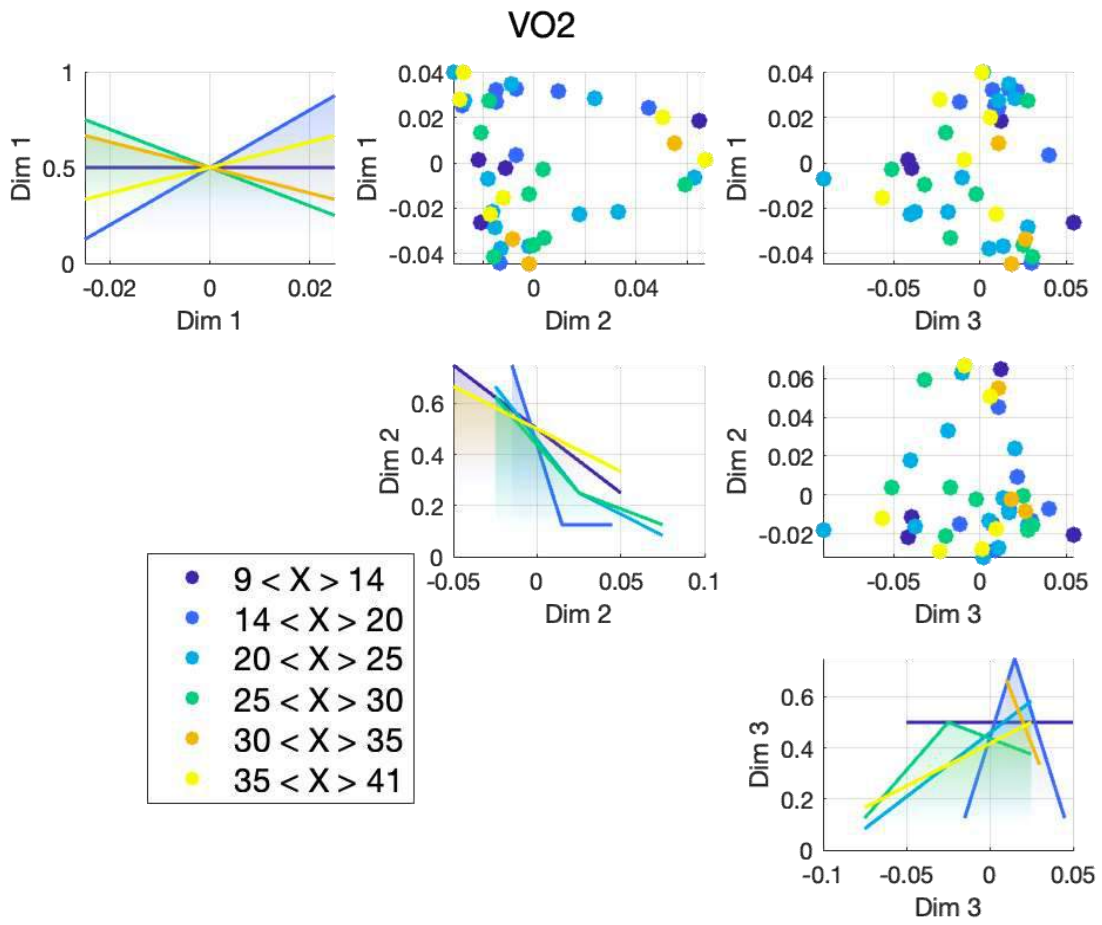
Myocardial dysfunction

	-2 STD	-1 STD	mean	1 STD	2 STD
VO2 (mL/(kg·min))	33.2	32.5	29.5	26.5	23.8
LVEF (%)	65.3	65.6	63.2	59.6	55.6
QRS width (ms)	139	152	153	145	136
Age (years)	25.5	27.2	29.0	30.3	32.0



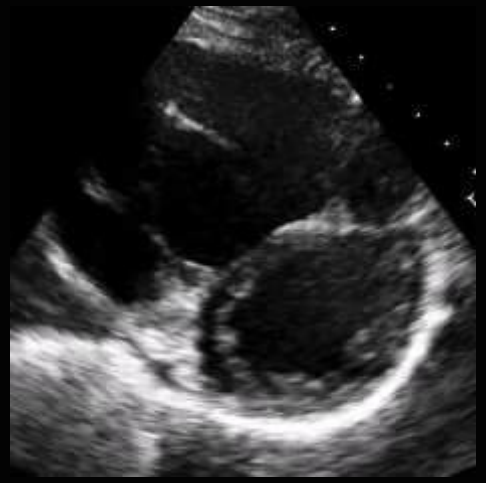
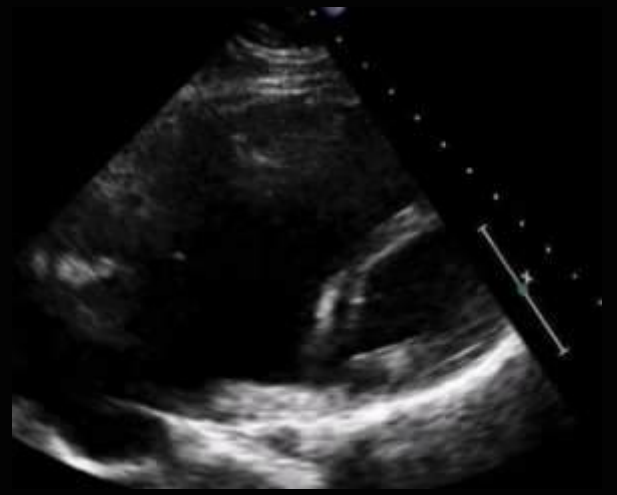
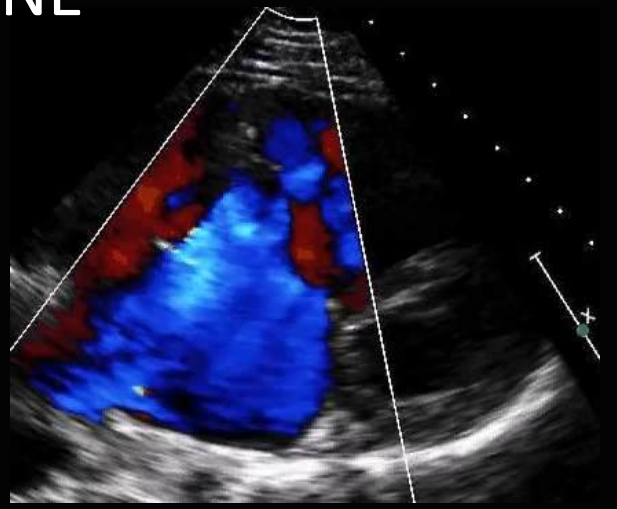
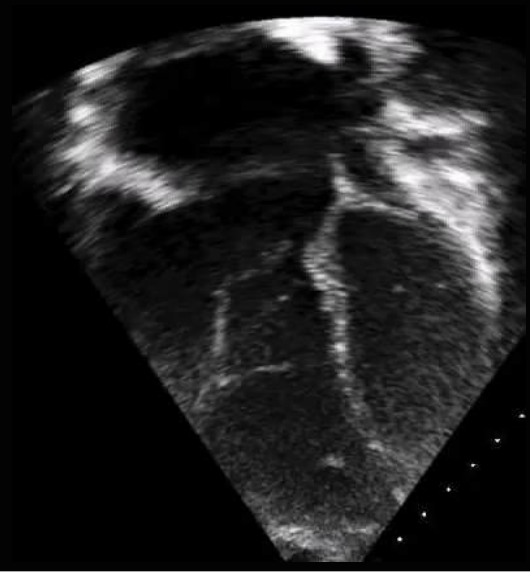
Identifying risk groups



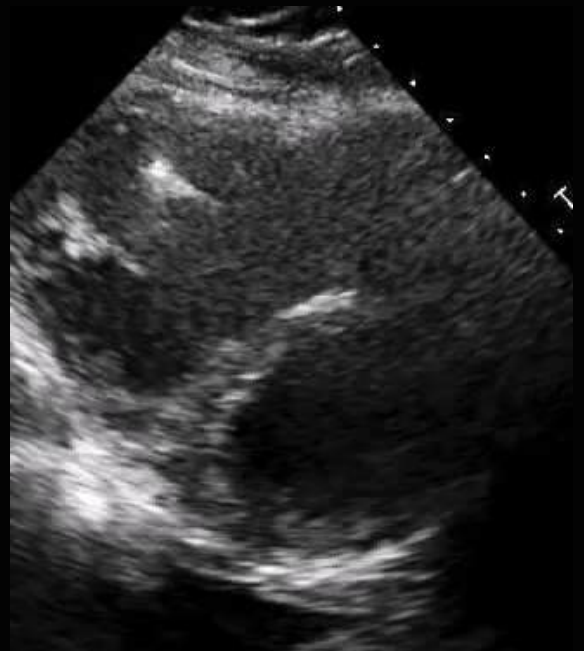
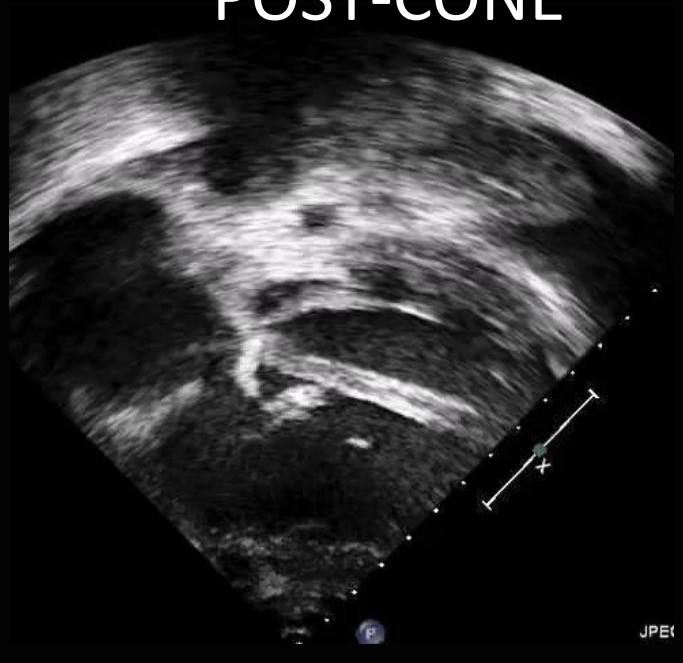
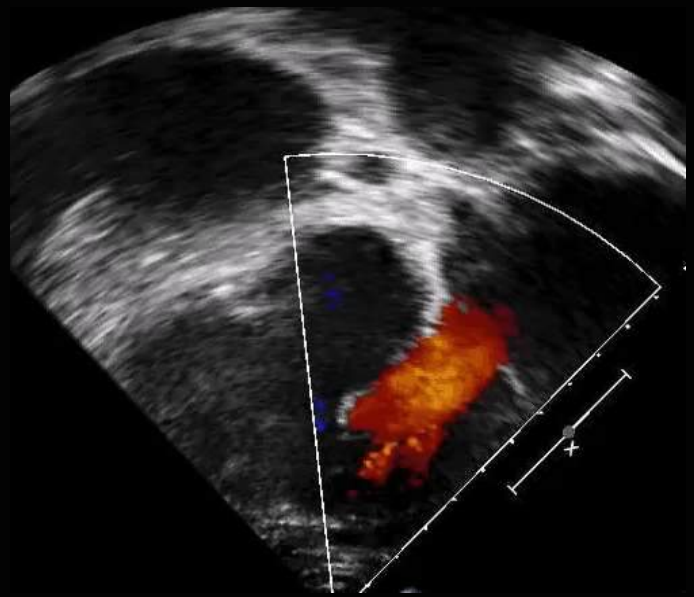


RV dysfunction pre and post-Cone procedure

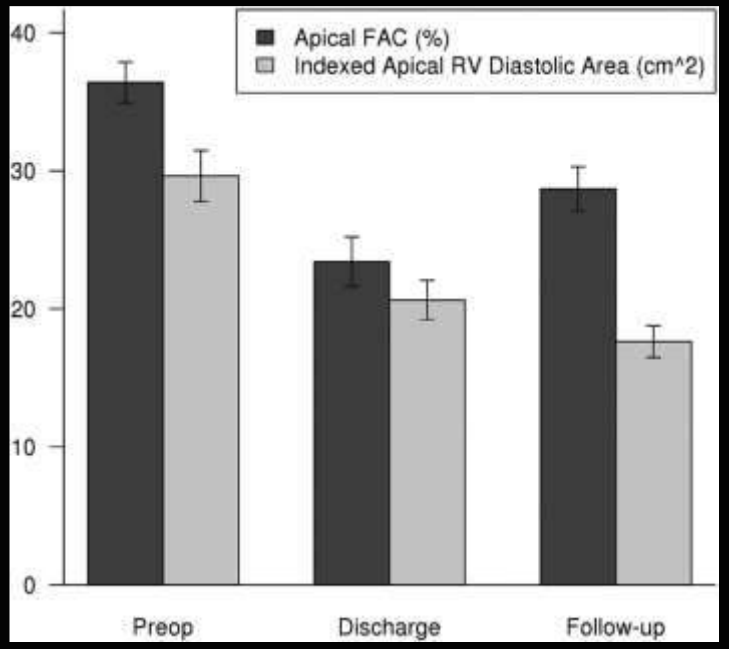
PRE-CONE

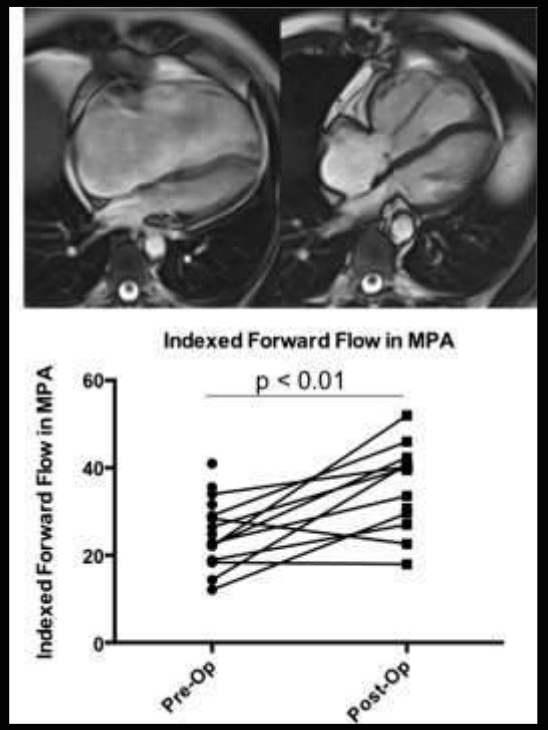
POST-CONE

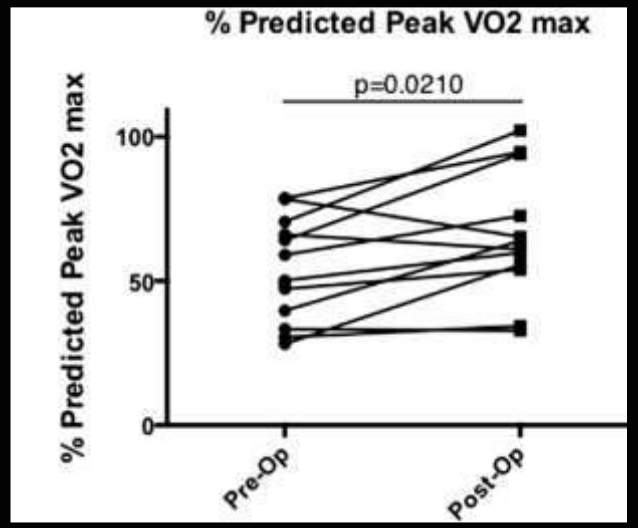




So why does the Cone procedure work?

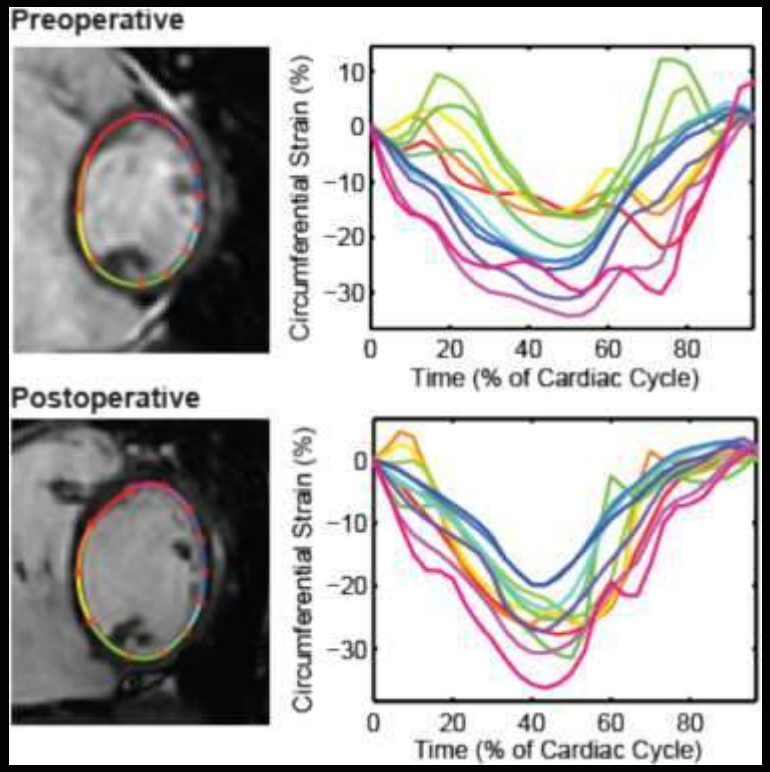


Holst , Ann Thorac Surg. 2018;105:160





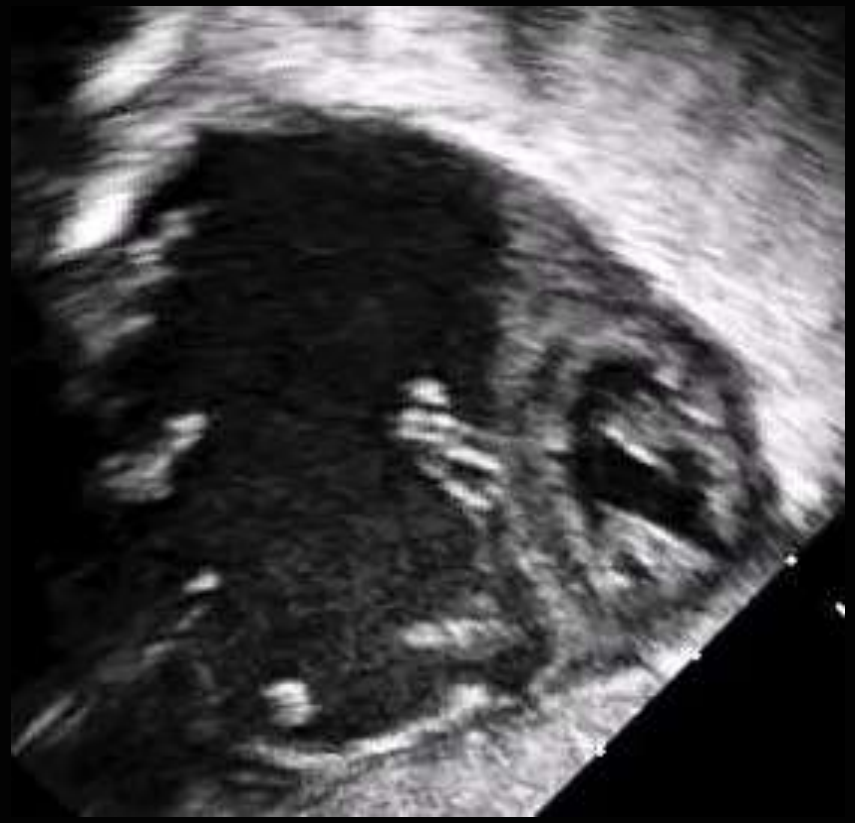
Ibrahim, JTCVS 2015;149:1144-50

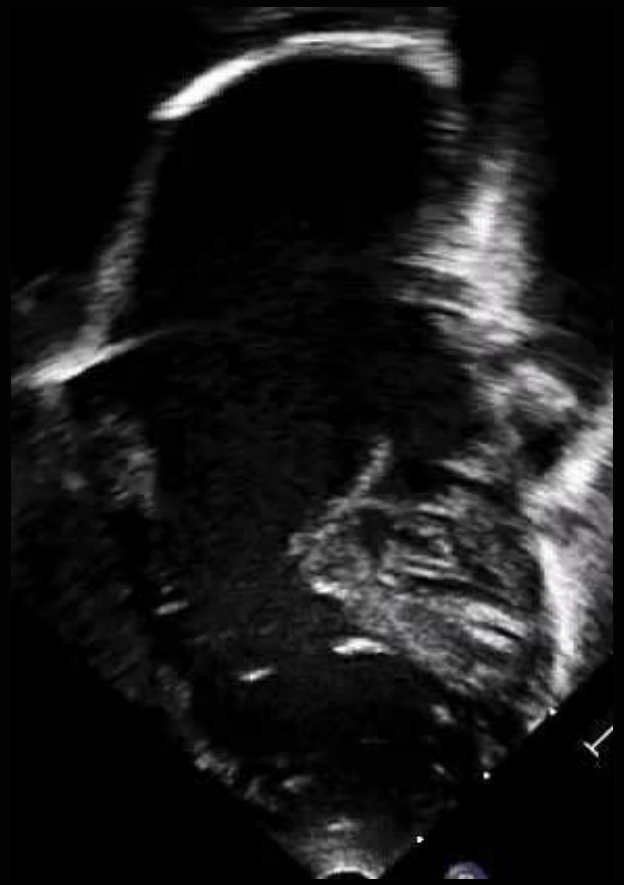


Beroukhim, JCMR, 2018;20:32

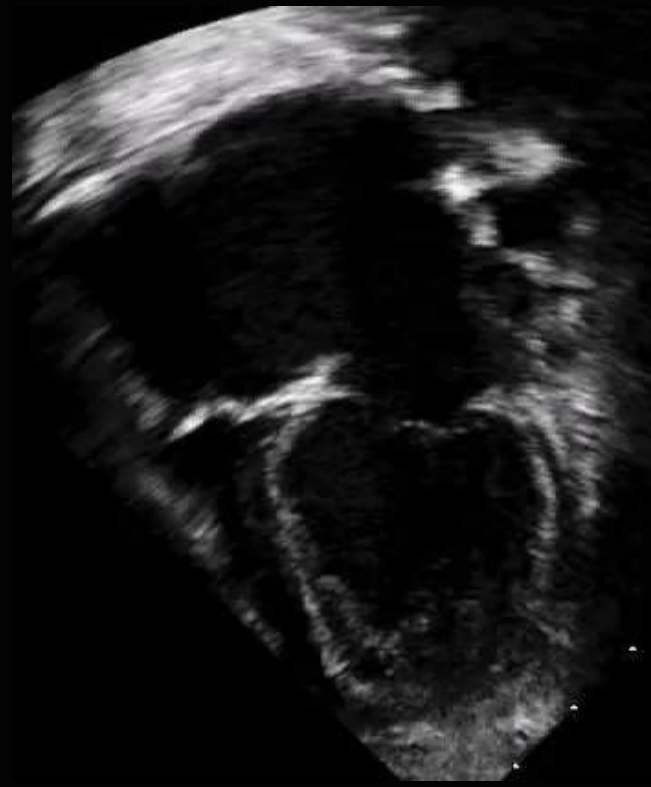
Severe Ebstein –Starnes procedure

Pre-Starnes



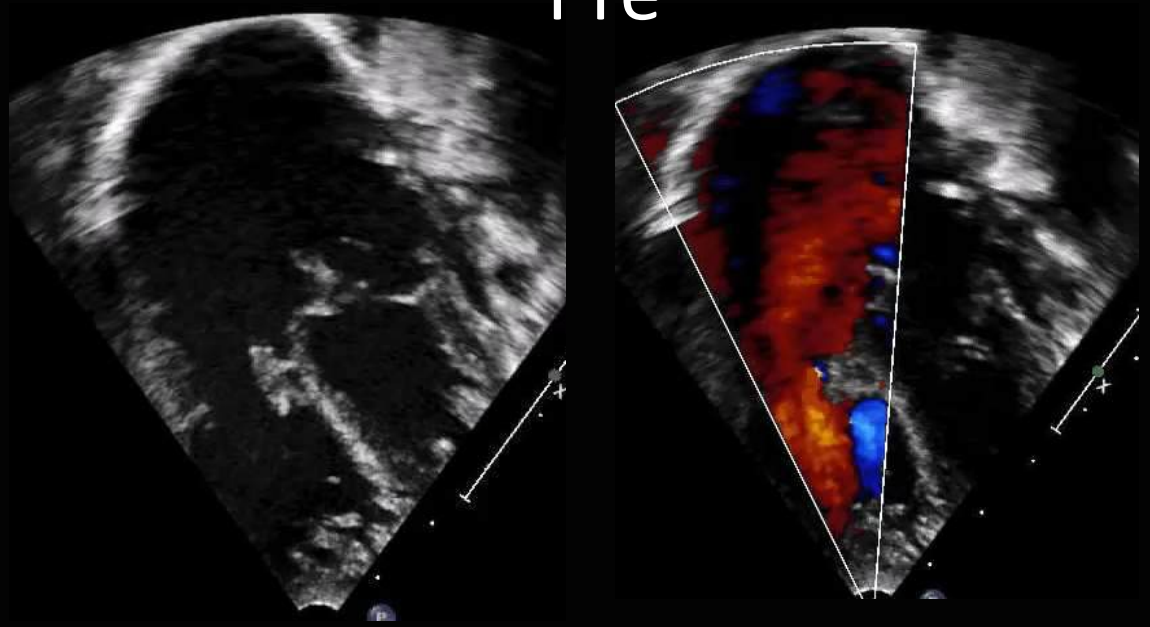


Post-Starnes

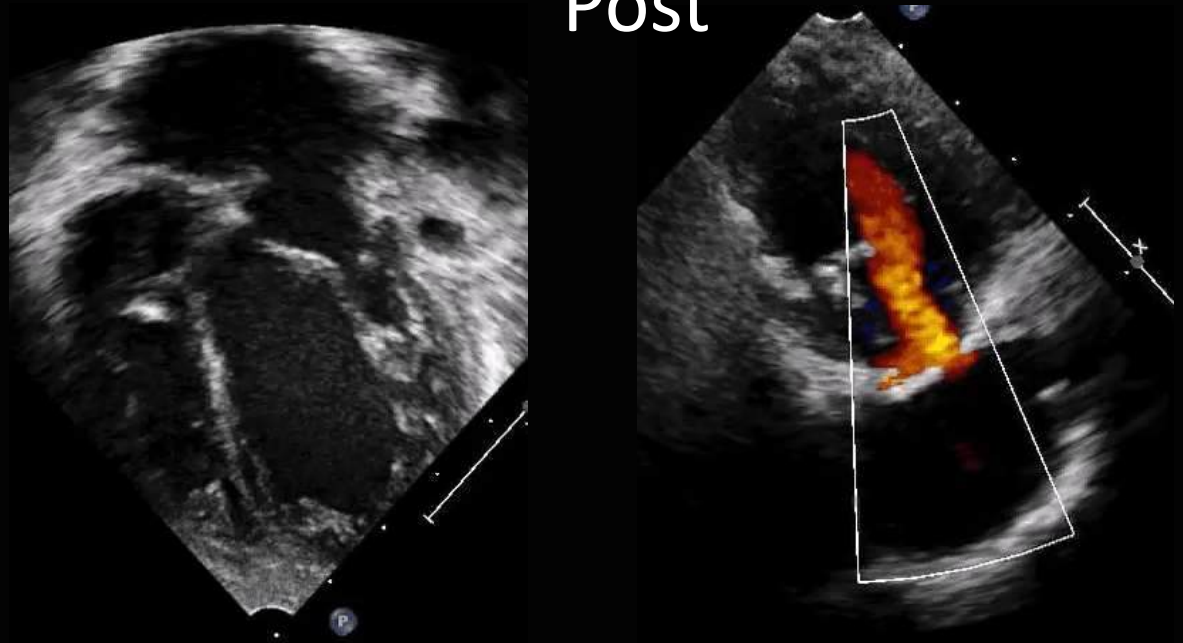


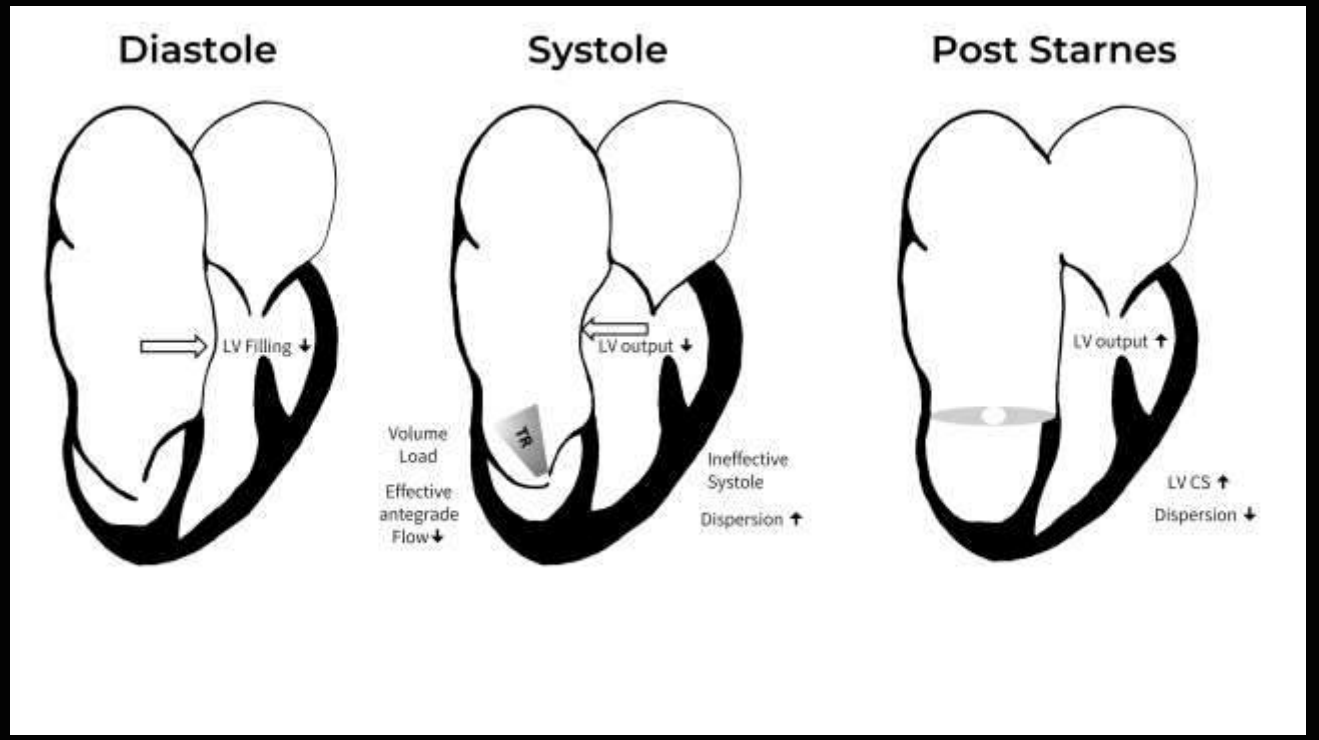
Starnes procedure: Excluding the RV

Pre



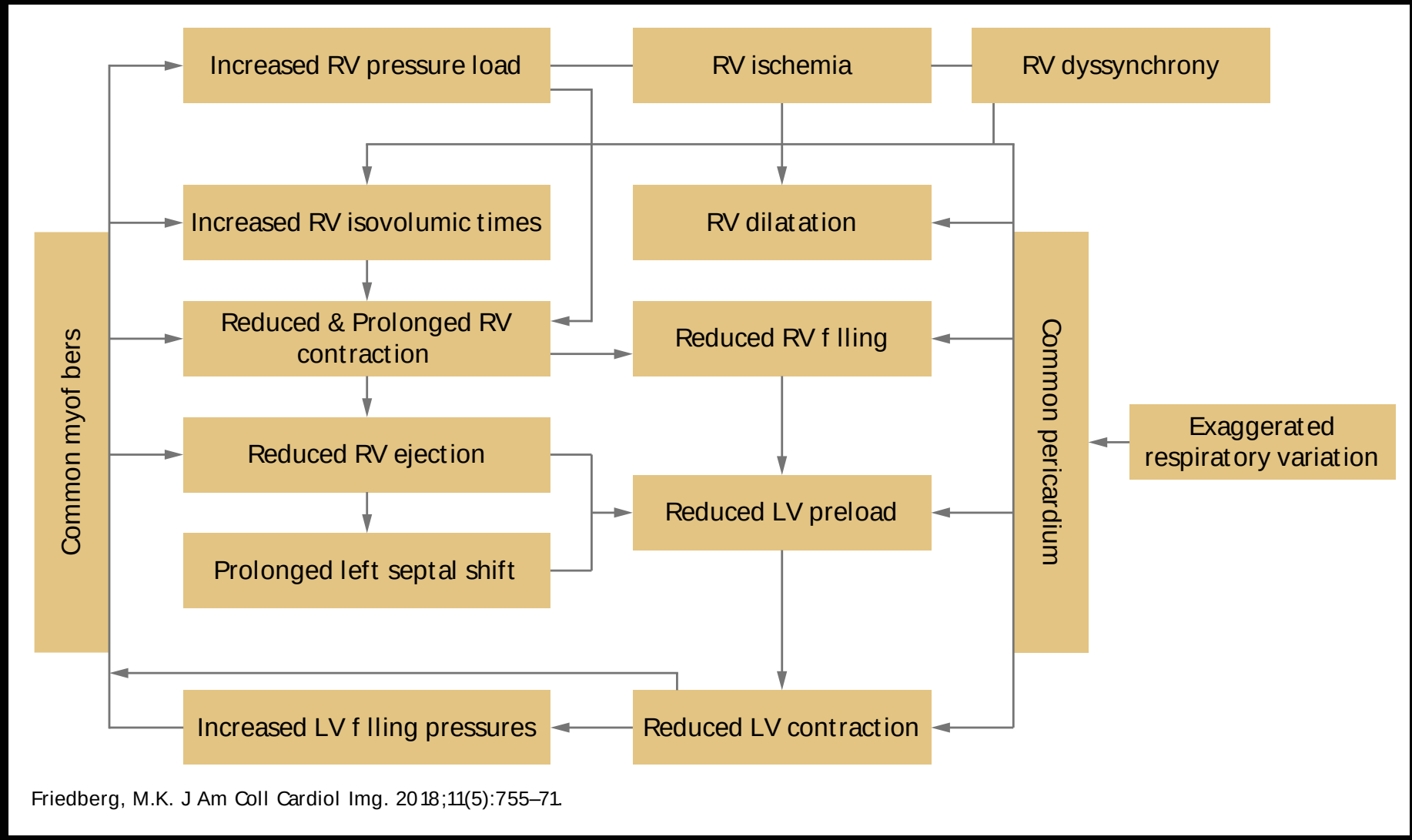
Post





Bitterman Y, Friedberg MK. JASE 2024;37:255-258

Pathophysiology of Ventricular Interdependence



Friedberg, M.K. J Am Coll Cardiol Img. 2018;11(5):755–71.

Conclusions

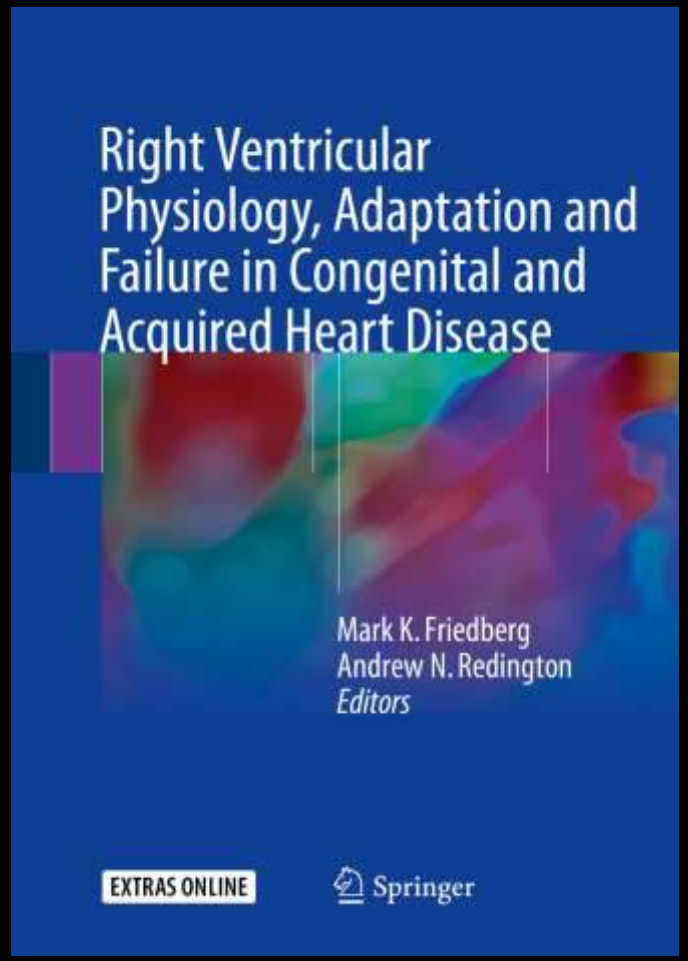
- Ebstein anomaly is a disease of the TV and RV
- Pulmonary antegrade flow, LV filling and RV-LV interactions may be as or more important than RV function per se
- RV-LV interactions are comprised of:
  - prolonged RV contraction
  - Elevated PVR
  - Reduced LV preload
  - Leftward septal motion in early LV diastole
  - RBBB electromechanical dyssynchrony
  - Possible atrial pre-excitation
- Surgical interventions in part act via improving RV-LV interactions

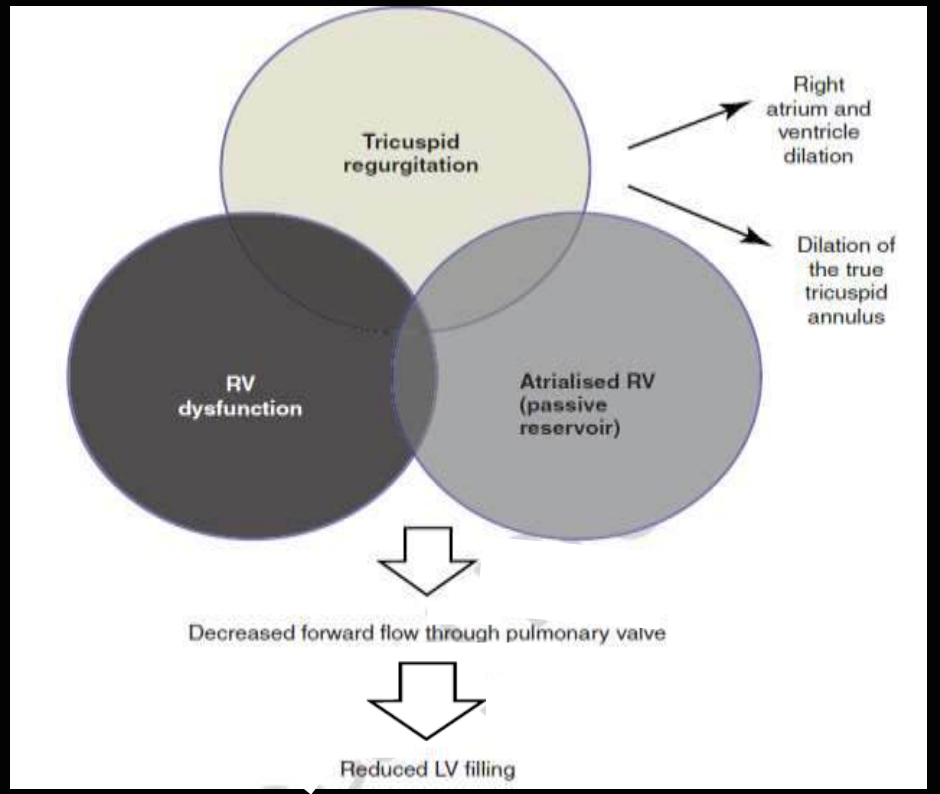
Thank you

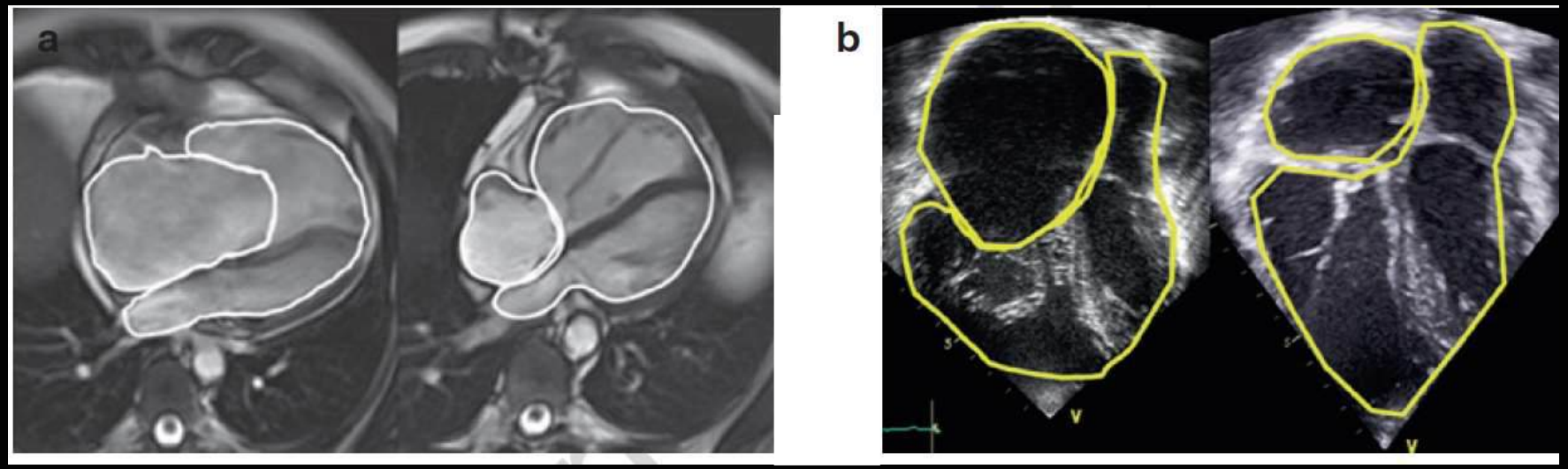
Right Ventricular Function in Ebstein's Anomaly of Tricuspid Valve

7

Jan Marek, Marina L. Hughes, and Victor Tsang





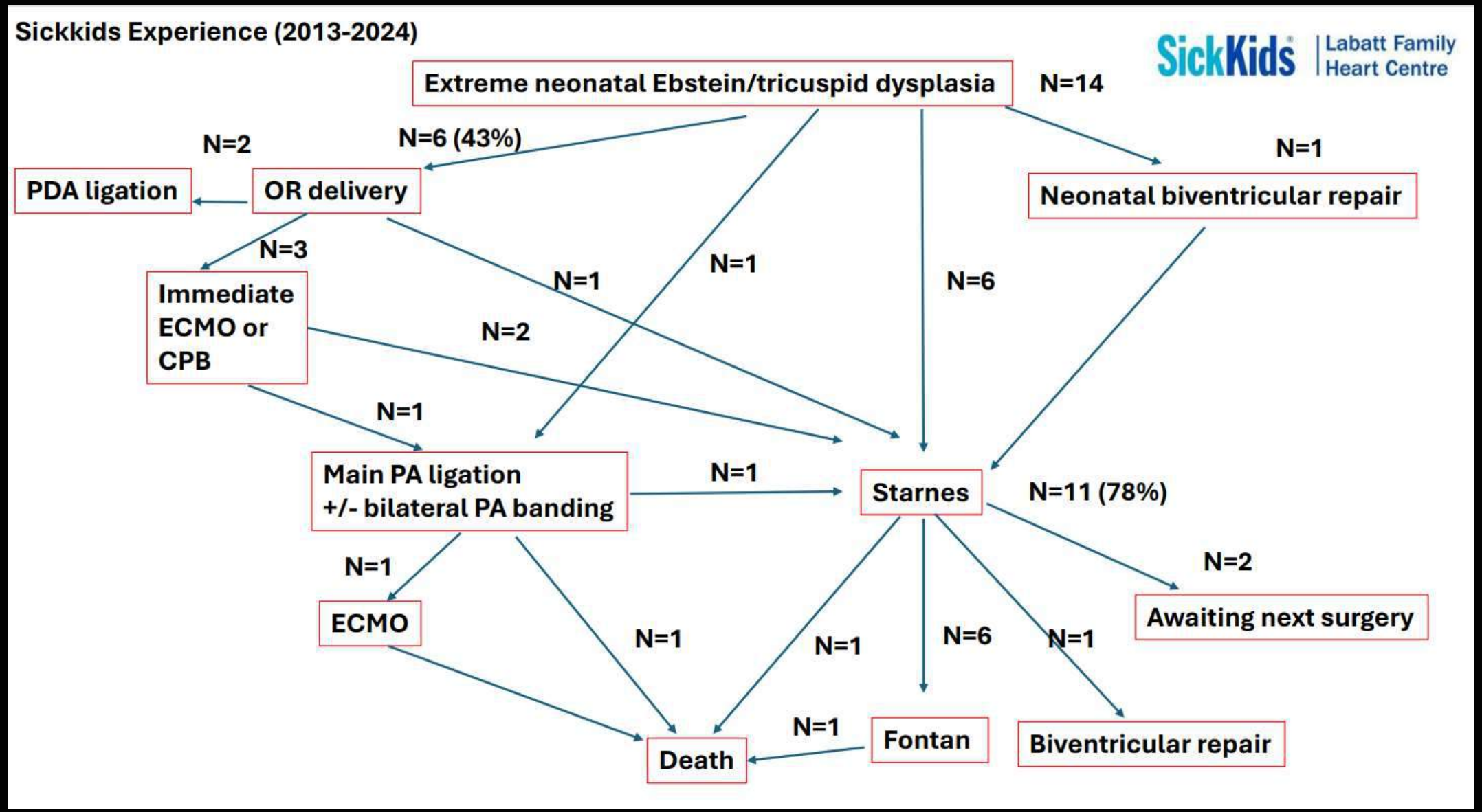


GOS score

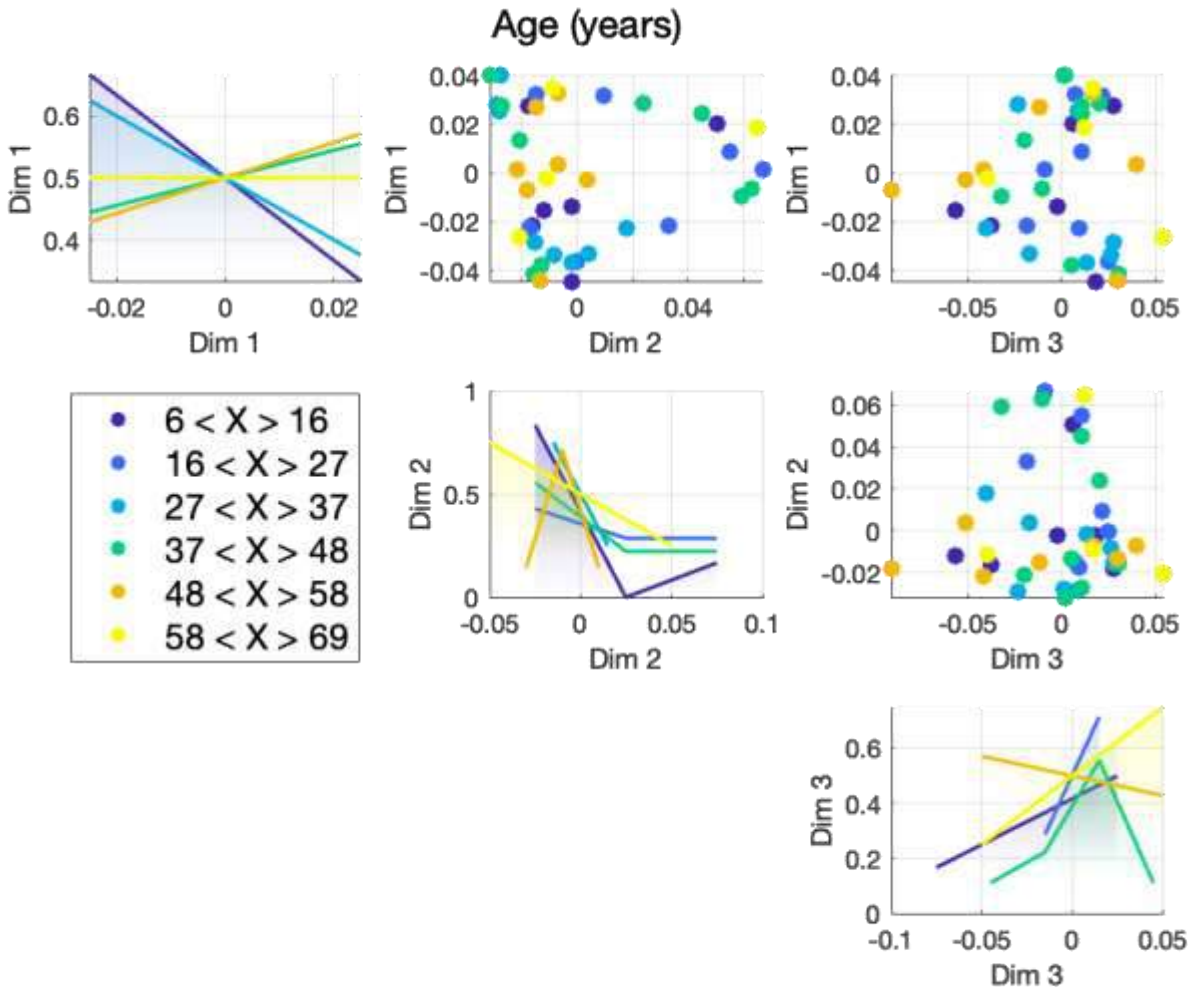
Echo assessment of Ebstein anomaly

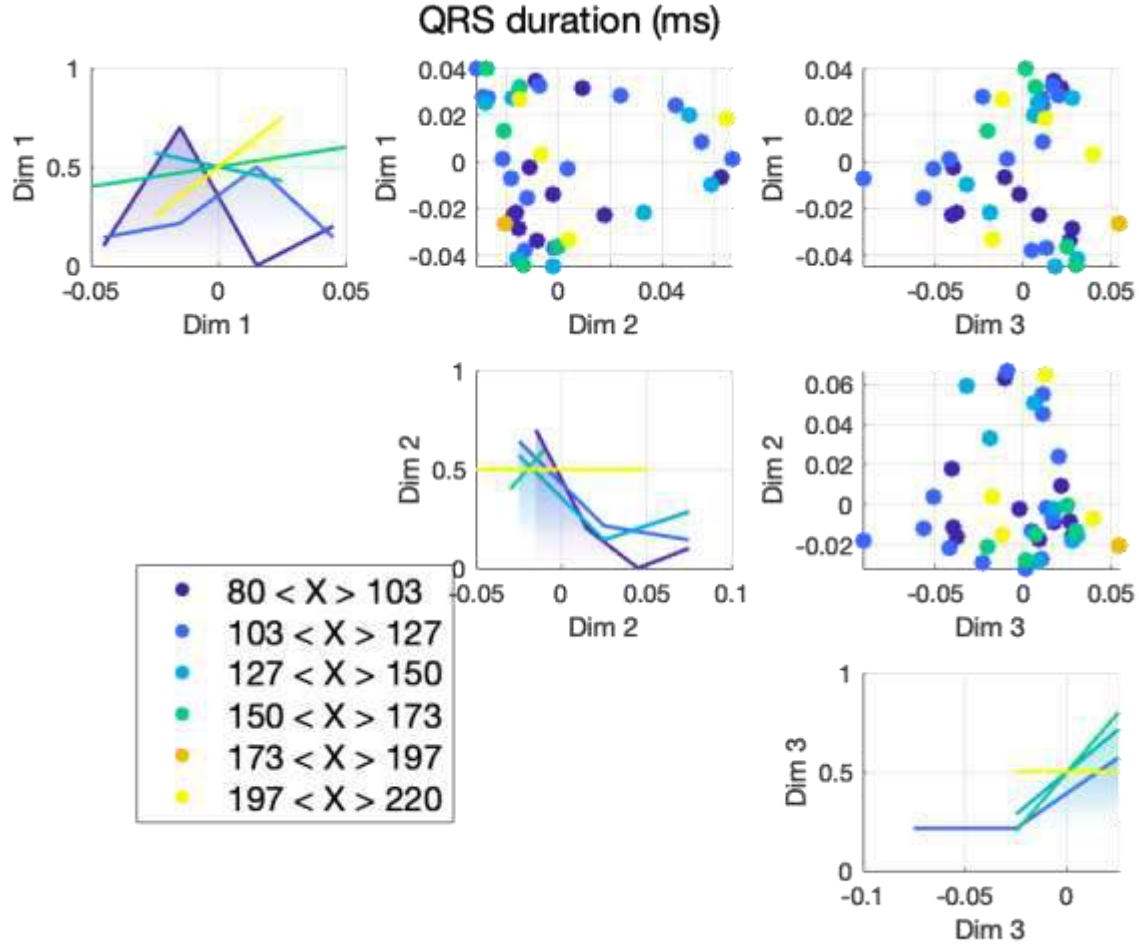
- Apical displacement of the septal leaflet (≥8mm/m2).
- Tricuspid valve size (Z-score) and Morphology
  - dysplastic thickened and rolled leaflets, fenestrations, multiple orifices, shortened chordae, underdeveloped papillary muscles, restricted mobility/tethering, or stenosis.
- Tricuspid regurgitation and stenosis
  - difficult to quantify given compliance of the atrialized RV and multiple regurgitation jets.
- Pulmonary stenosis/ atresia
  - Presence of PR can differentiate “functional” from true pulmonary atresia. Direction of PDA flow can help determine adequacy of pulmonary valve antegrade flow.
- Presence and direction of flow across an atrial septal defect or PFO.
- Circular shunt physiology (severe TR, ASD, PDA and PR).
- RV size and function
  - Including effects on LV size and function.
- Assess anatomic severity:
  - Chamber Area Ratio (end diastole in apical 4 ch-view): (RA + aRV)/(RV + LA + LV); ratio of ≥1 in neonate can indicate a poor prognosis

Sickkids outcomes in extreme neonatal EA

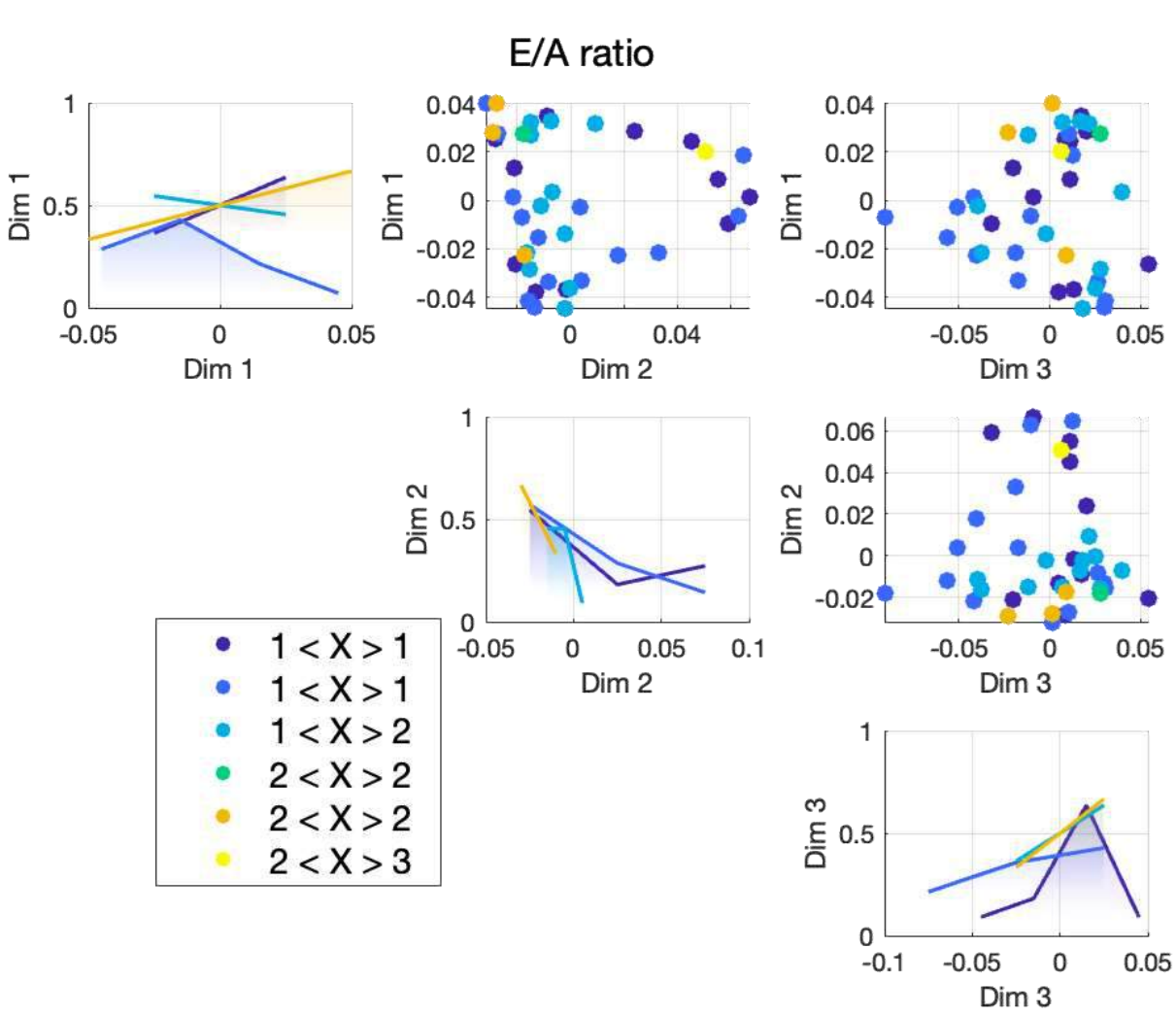


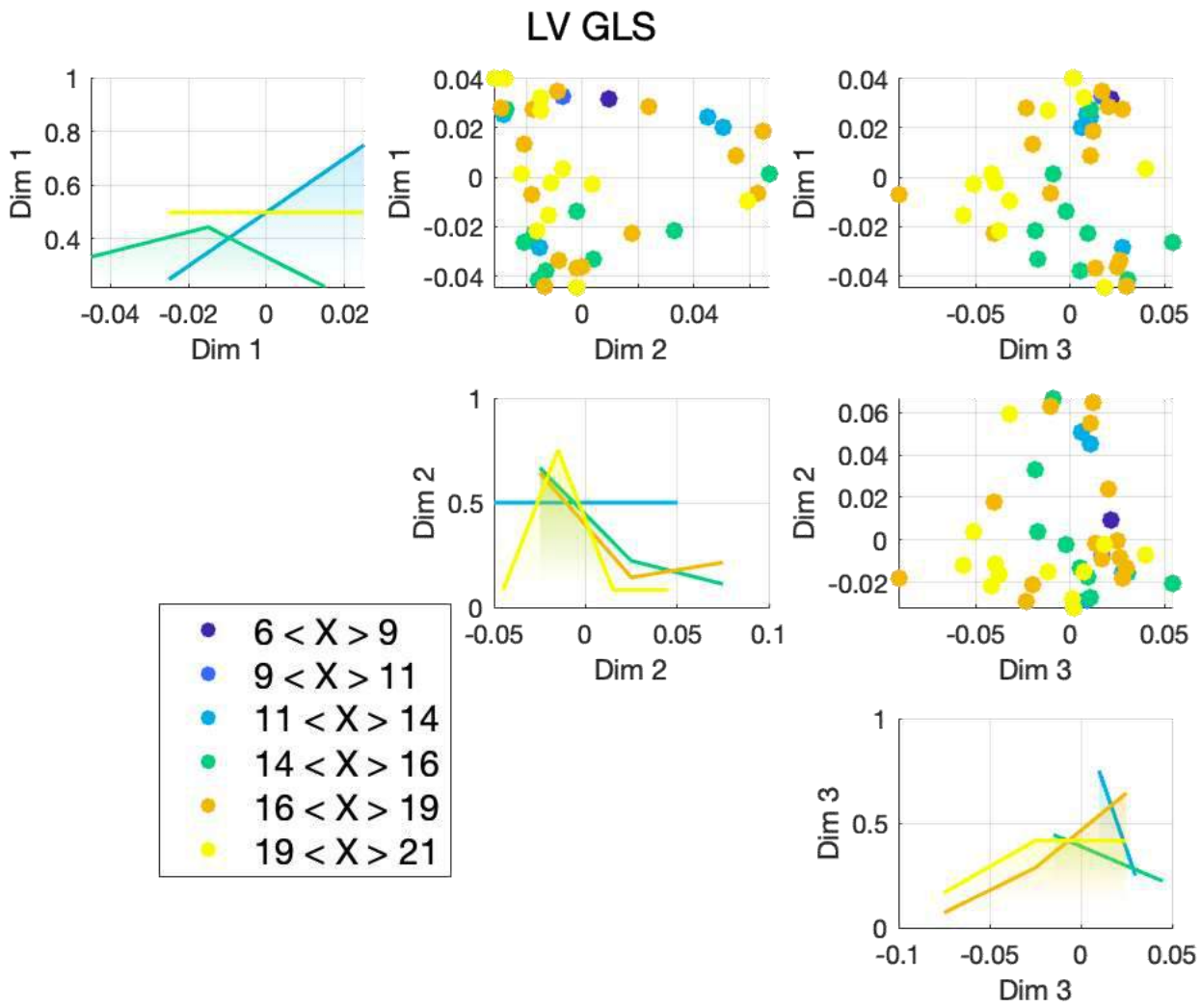
Machine learning risk factors





Machine learning risk factors





Machine learning risk factors