Echo in Pulmonary Hypertension

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Outline

1. What is PH?
2. Strategies to estimate RV or PA pressure by echo
3. RV systolic function
4. RV diastolic function

Part 1: What is PH?

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What is PH?

- Elevated pressure in the pulmonary arteries (mean PA pressure > 20 mmHg).
- Can be due to many causes
  - Type 1: Pulmonary arterial disease
  - Type 2: Backup of pressure due to left heart disease
  - Type 3: Lung disease
  - Type 4: Blood clotting disease
  - Type 5: Other

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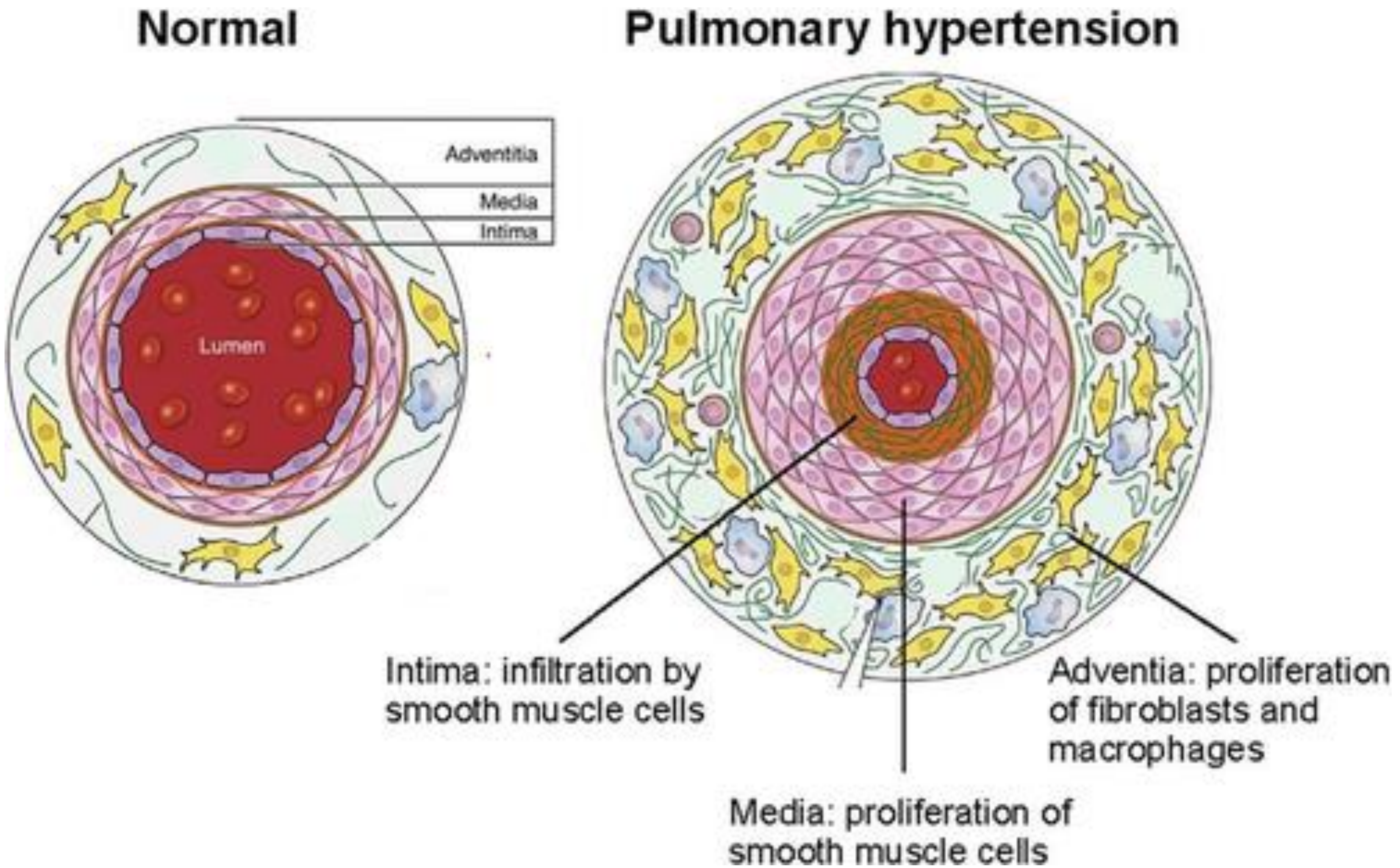
Pathophysiology of PAH (Type 1)

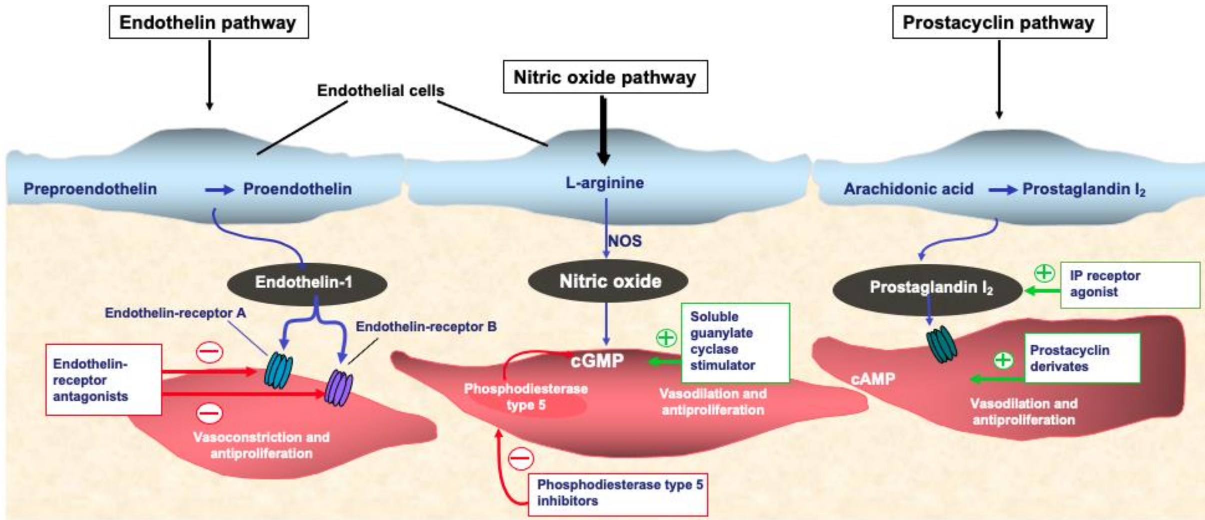


Sysol et al, Cardiology Continuing Education, June 2018 Vol 4 Issue 1. Pp 2-12

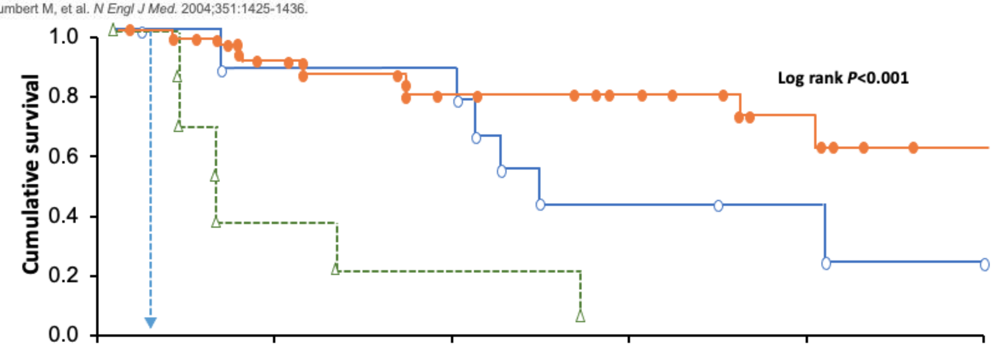
Pathophysiology of PAH (Type 1)

- Elevated resistance in the small arteries within the lungs.
- Increased RV afterload.
- Clinical effects can include:
  - Growth faltering
  - Low cardiac output
  - Hypoxemia
  - Tachypnea
  - Right heart failure
  - Cardiac arrest

Survival in PH



Adapted from: Humbert M, et al. *N Engl J Med.* 2004;351:1425-1436.



Patients at risk n	0	24	48	72	96	120
	39	26	20	12	7	2
	9	7	6	3	2	0
	1	0	0	0	0	0
	6	2	1	0	0	0

N=66 consecutive, treatment-naïve pediatric patients in Dutch National Network for Pediatric Pulmonary Hypertension

Ploegstra MJ, et al. *Eur Respir J.* 2014;44:1616-1626.

Part 2: Estimating Right Heart Pressures

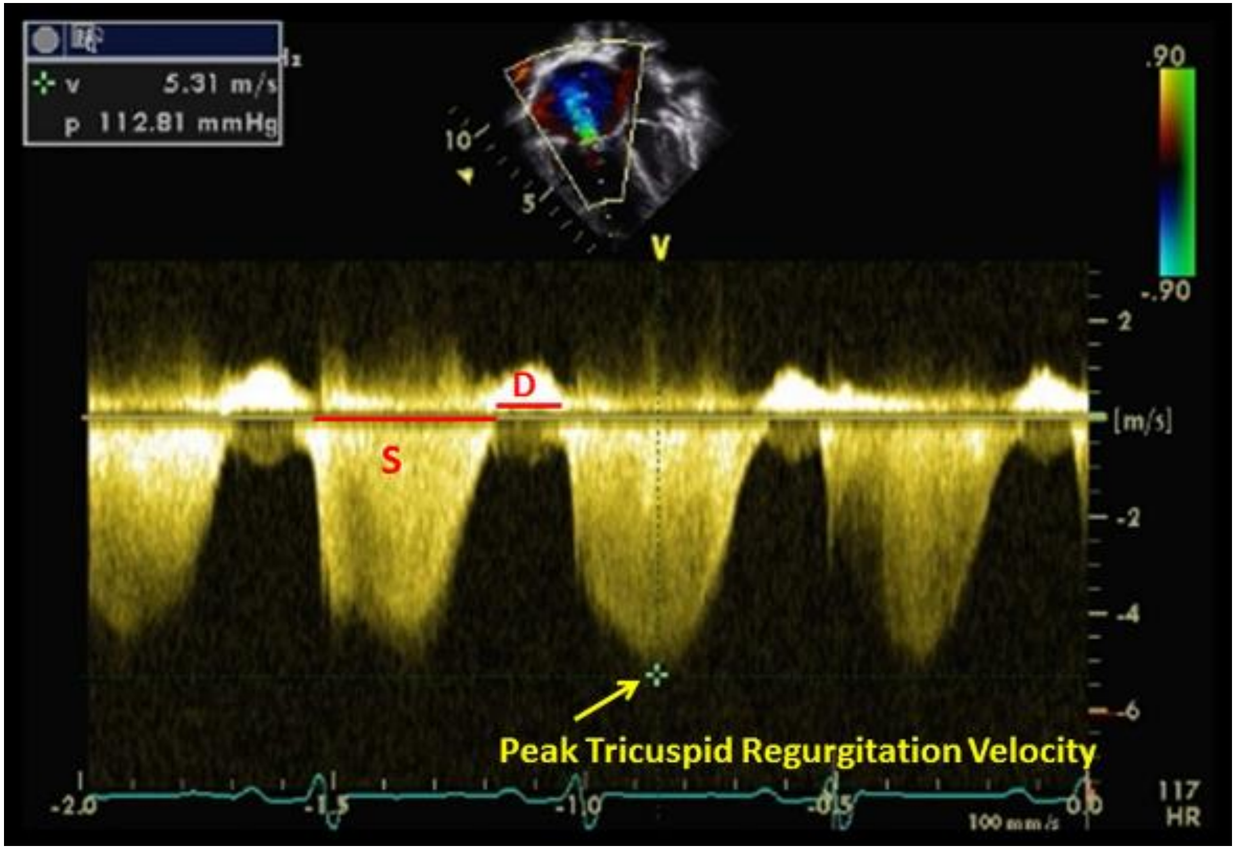
Estimating Right Heart Pressures

- First described in *Hydrodynamica* in 1738 by Daniel Bernoulli, later converted to the form we use today by Leonhard Euler in 1752.
- Simplified for our purposes:
  - deltaP = $4V^2$

Credit: Wikipedia

Estimating Right Heart Pressures

- Tricuspid regurgitation velocity reflects RV pressure above right atrium.



Credit: Jone and Ivy, Front in Peds 2014

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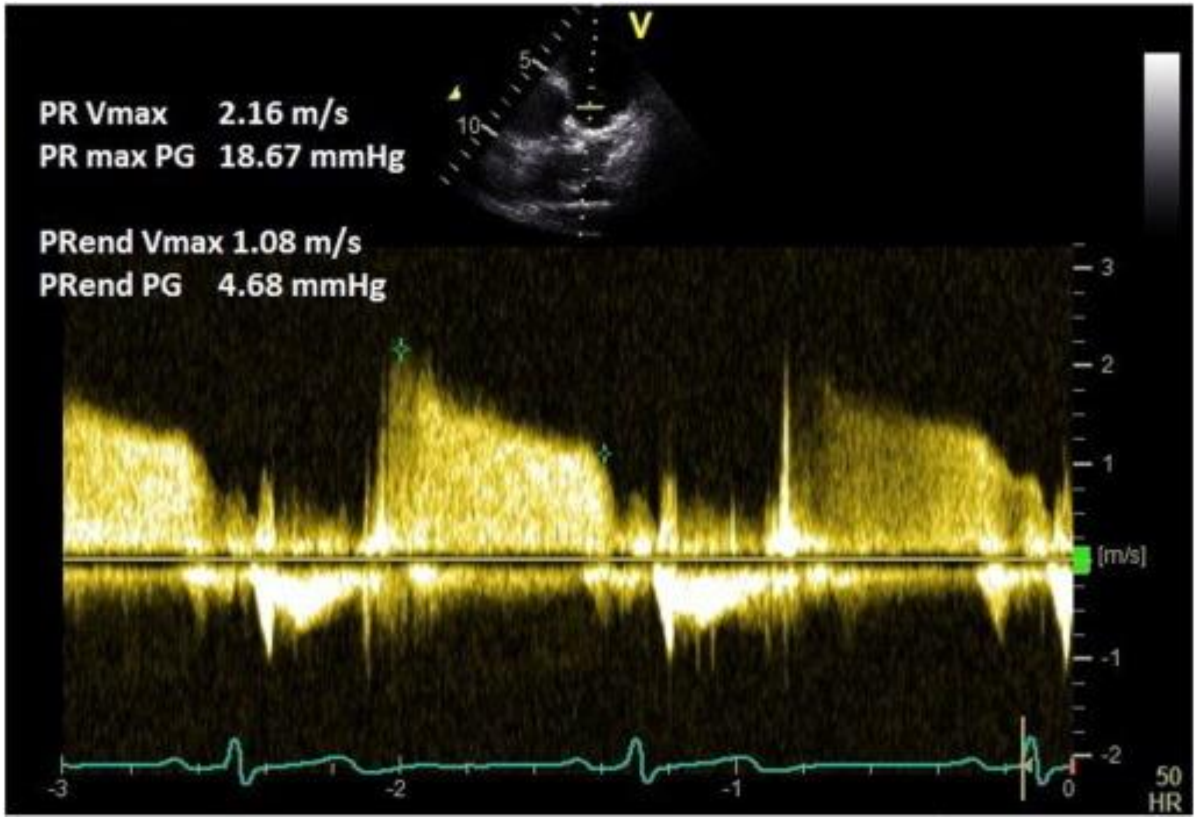
TR continued

- References disagree on the true upper limit of normal – probably it is in the 2.8-3.2 m/s range for adults and older kids. Less clear for babies.
- Although imperfect because of several assumptions, one can approximate the mean pulmonary artery pressure from TR via:
  - mPAP = 0.61xRVSP + 2 mmHg
    - [Chemla, *Chest*, 2009]

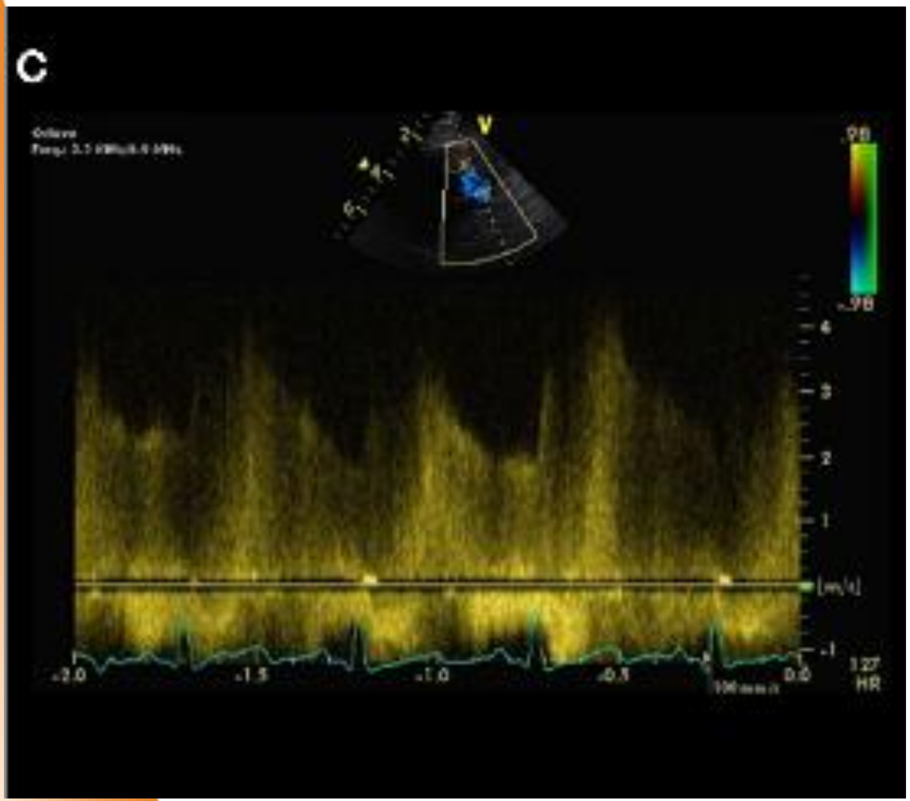
Estimating Right Heart Pressures

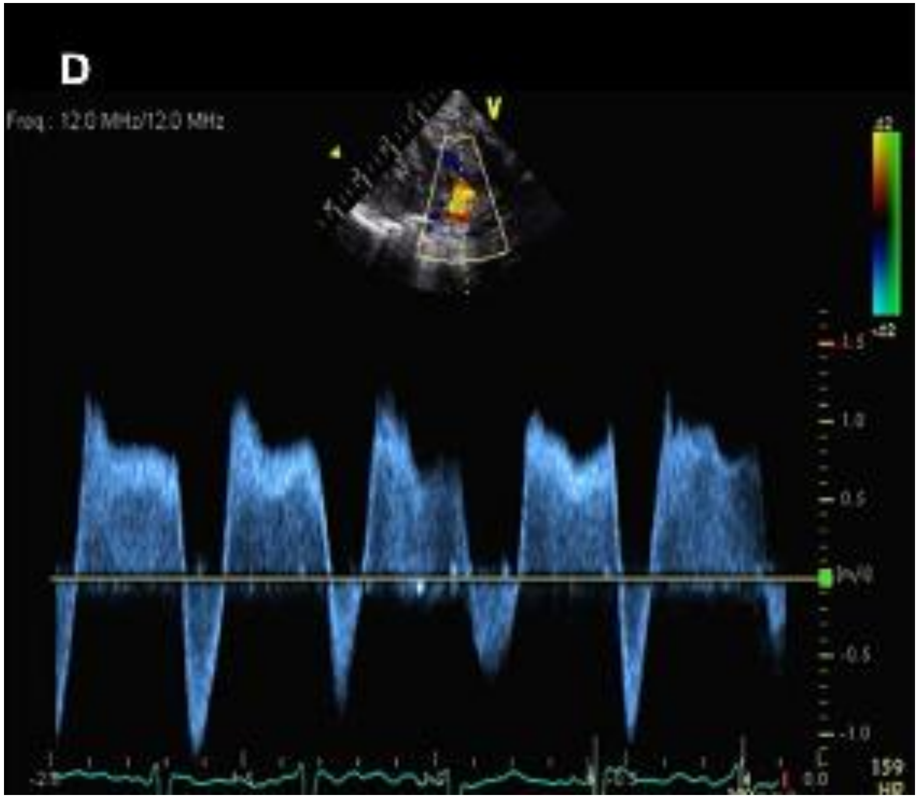
- Pulmonary insufficiency velocity can tell you about both the mean and diastolic PA pressures.



Credit: Parasuraman et al, IJC:HV 2016

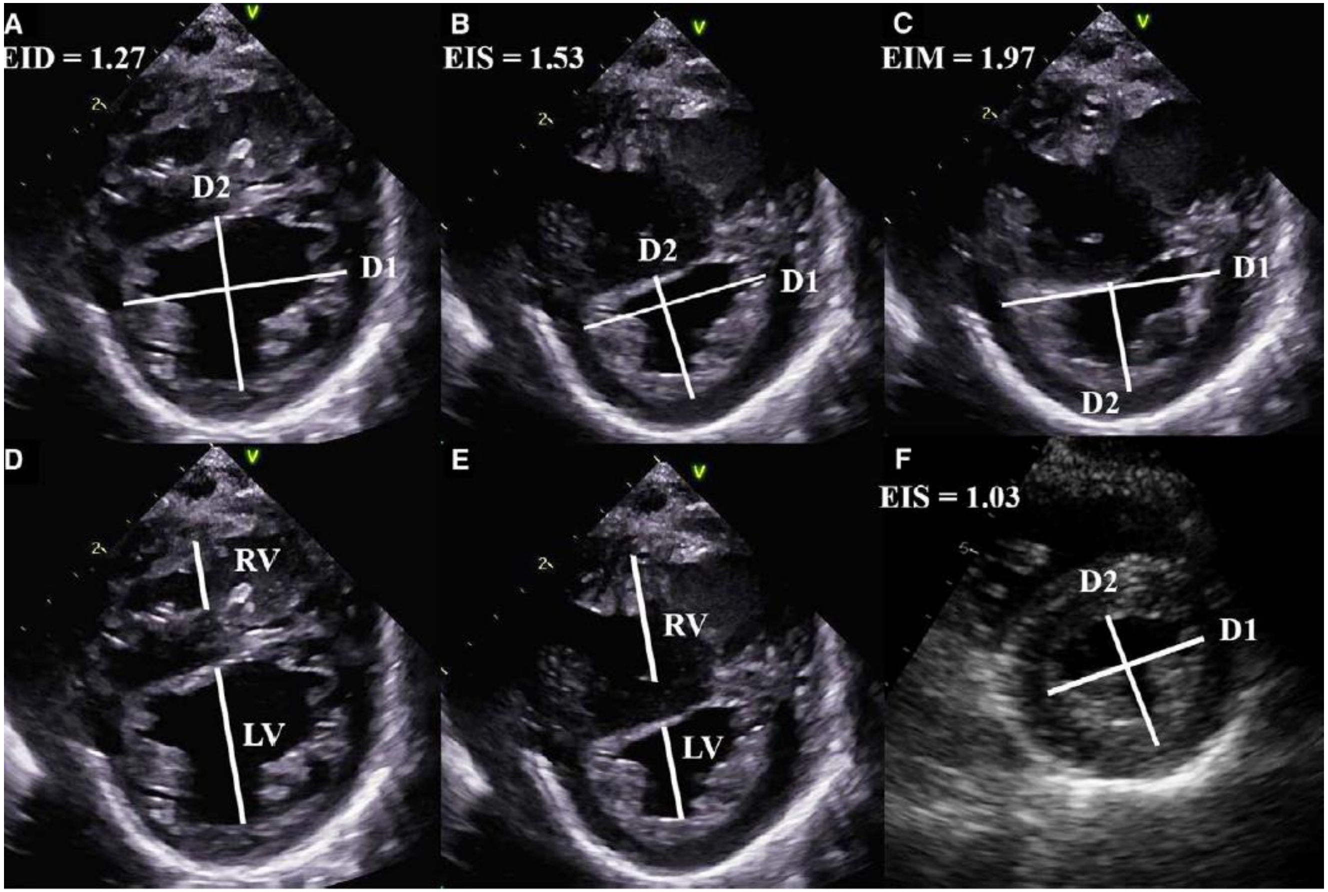
PDA flow patterns





Eccentricity Index

- Well published strategy to quantify septal flattening.



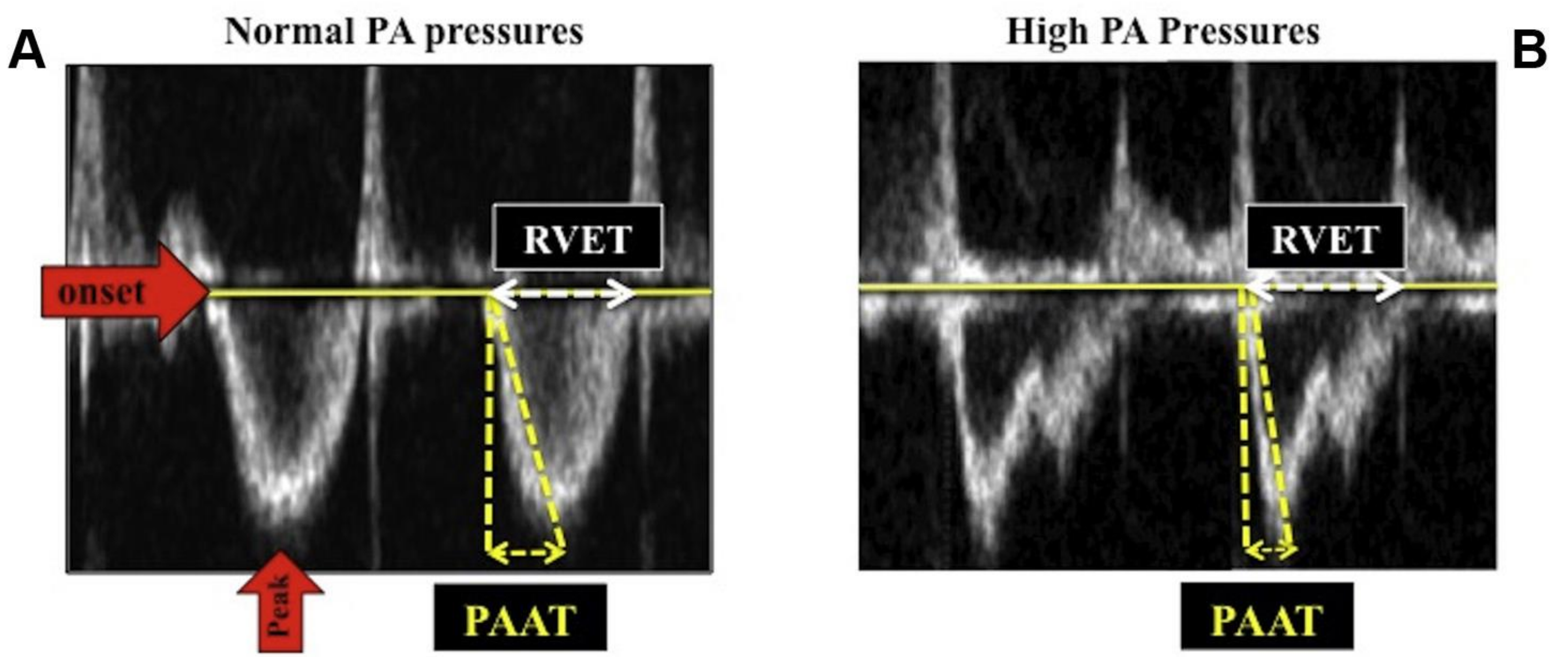
Credit: Burkett et al Circ: CVI, 2020

Eccentricity Index - Systolic

- Normal patients reliably had EI < 1.1, mostly close to 1.0.
- Mild septal flattening EIs ~1.2-1.5
- Moderate septal flattening EIs ~1.5-2
- Severe septal flattening EIs >2

Pulmonary Artery Acceleration Time (PAAT)

- As PVR rises, the PAAT shortens (think of this as quantification of the flying W).
- PAAT/RVET < 0.31 identified as cutoff to identify abnormal.



Credit: Levy et al 2016

Part 3: Right Ventricle Systolic Function

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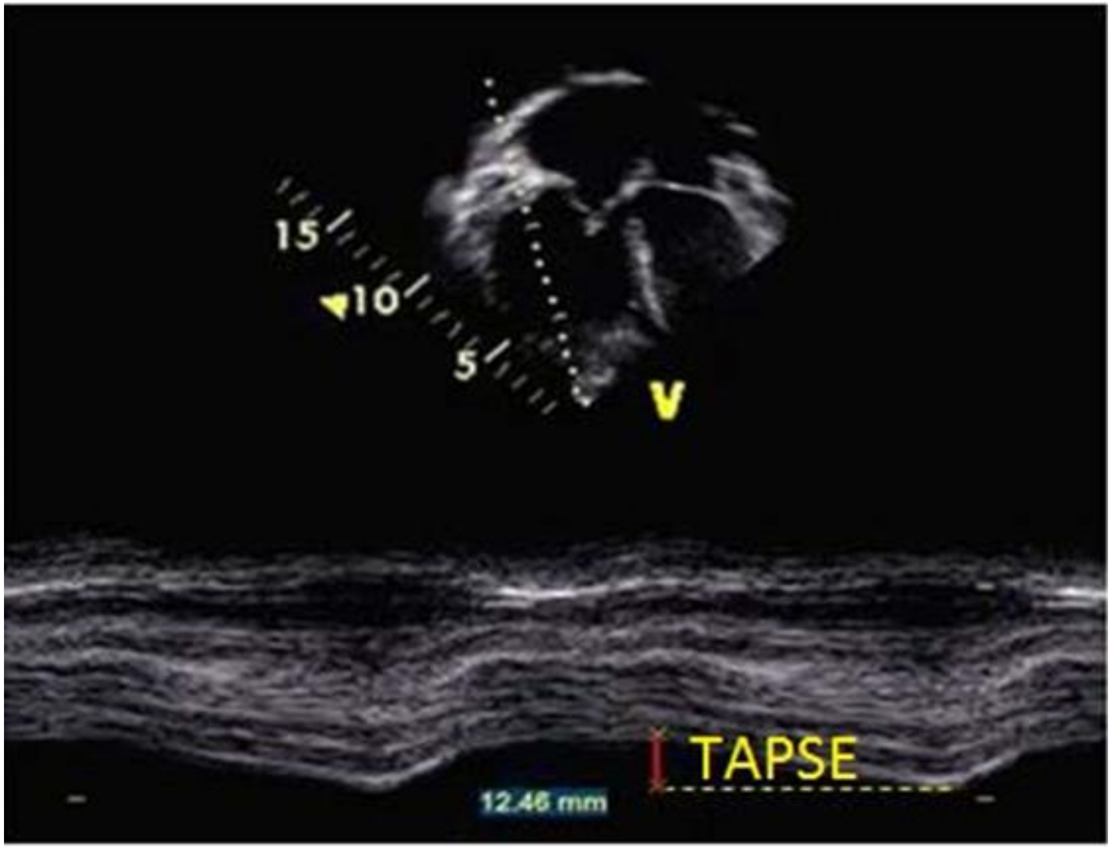
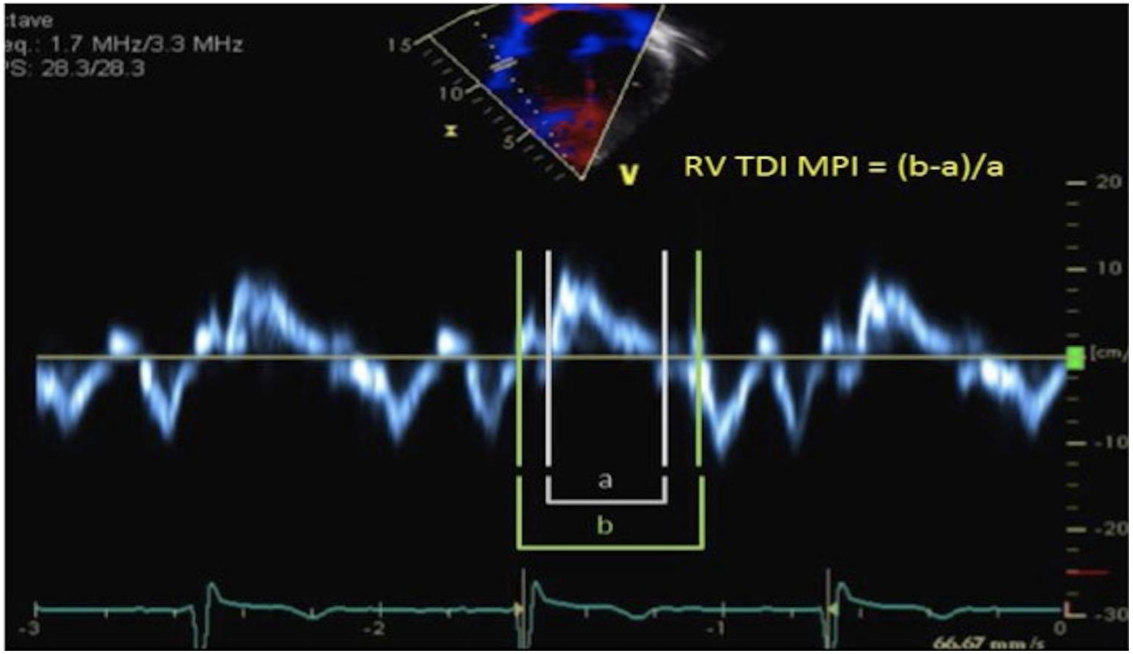
TAPSE

- One dimensional measurement
- Specific but not sensitive for RV dysfunction.
- Normal varies by age.



Myocardial Performance Index (MPI)



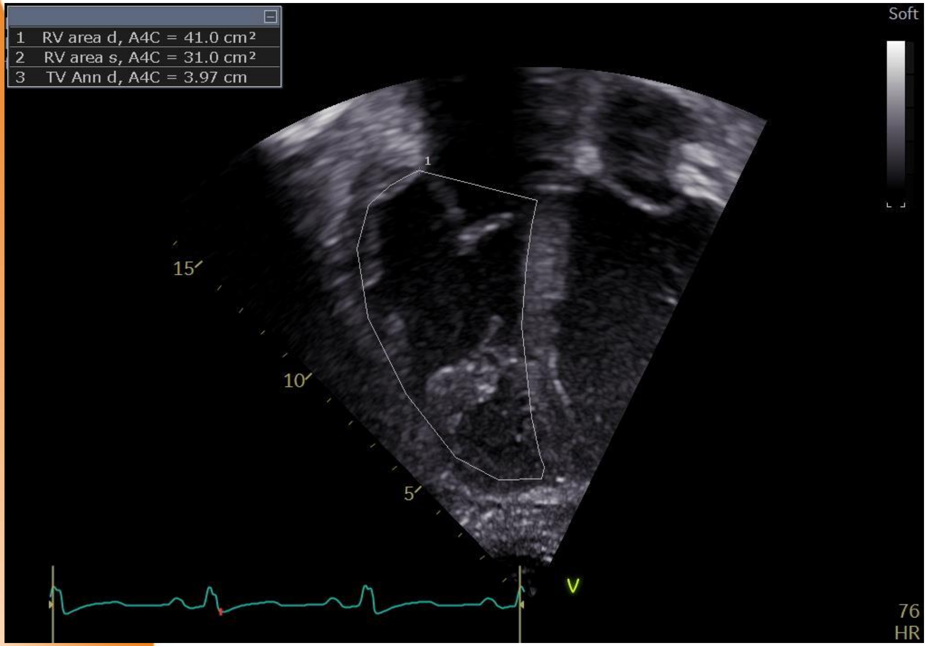
- The theory is that a well performing ventricle will require less time for isovolumic contraction and relaxation than a sick ventricle.
- Larger MPI = worse function

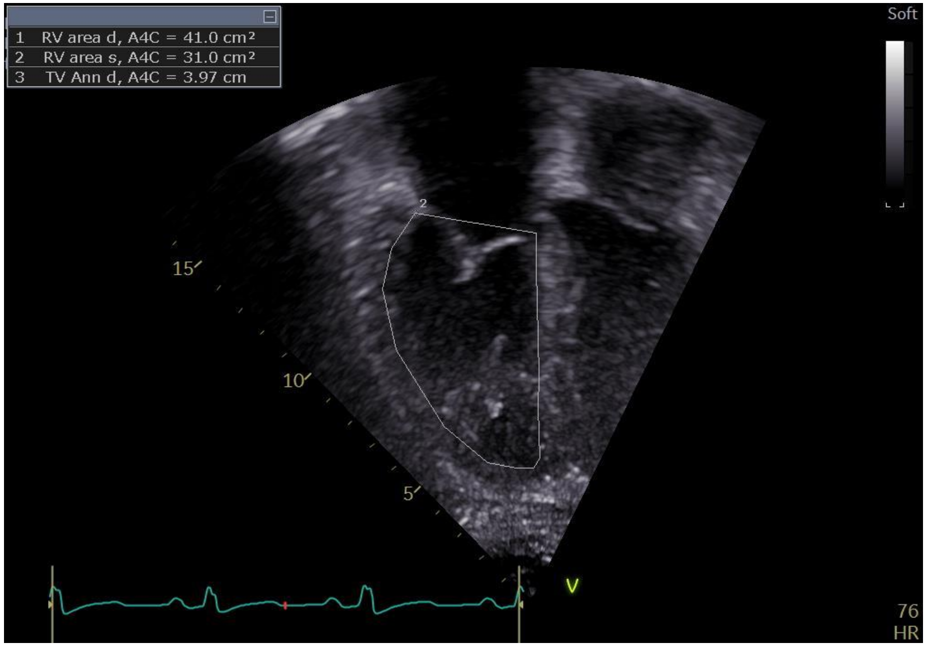
Credit: Friesen et al 2017

Fractional Area Change (FAC)

- Two-dimensional measure
- (RVd - RVs) / RVd * 100
- <u>Pros</u>: takes into account both longitudinal and radial function.
- <u>Cons</u>: reproducibility is a challenge, especially when RV visualization is tricky.
- We use 35% as a cutoff for the lower limit of normal.

RV FAC



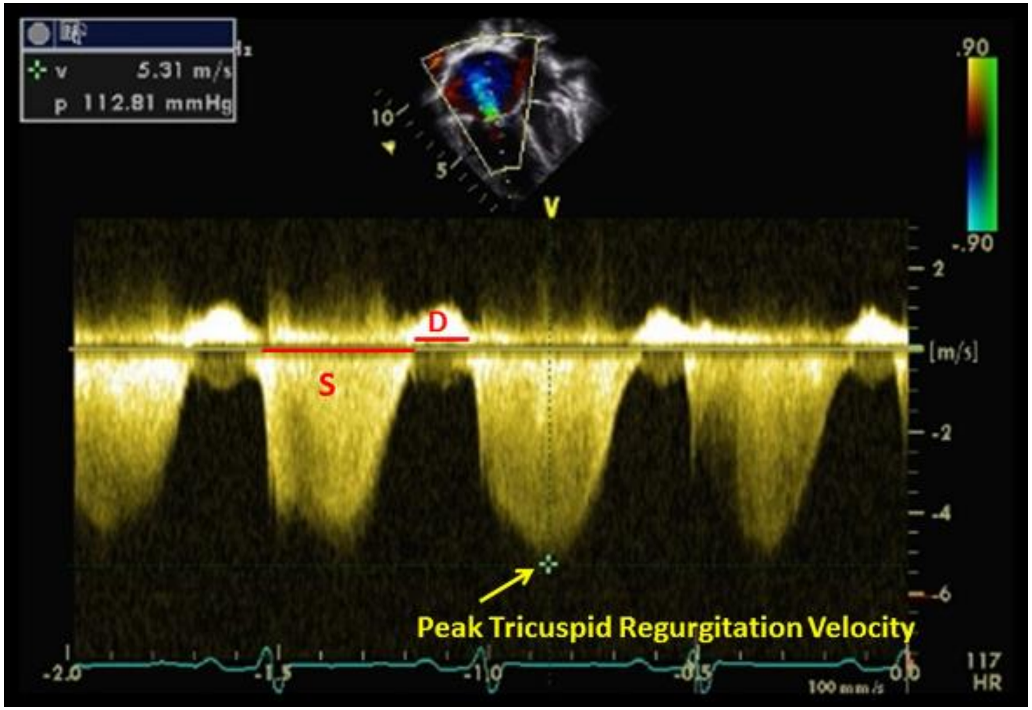


Diastolic area = 41 cm^2
Systolic area = 31 cm^2

RV FAC = (41-31)/41 = 24.4% = moderately decreased

Systole : Diastole ratio

- Sick RVs take longer to eject, prolonging systole (leading to higher S:D ratio).
- S:D > 1.4 associated with worse prognosis. Normal would be <1.



Credit: Jone and Ivy, Front in Peds 2014, Truong 2020.

RV 3D

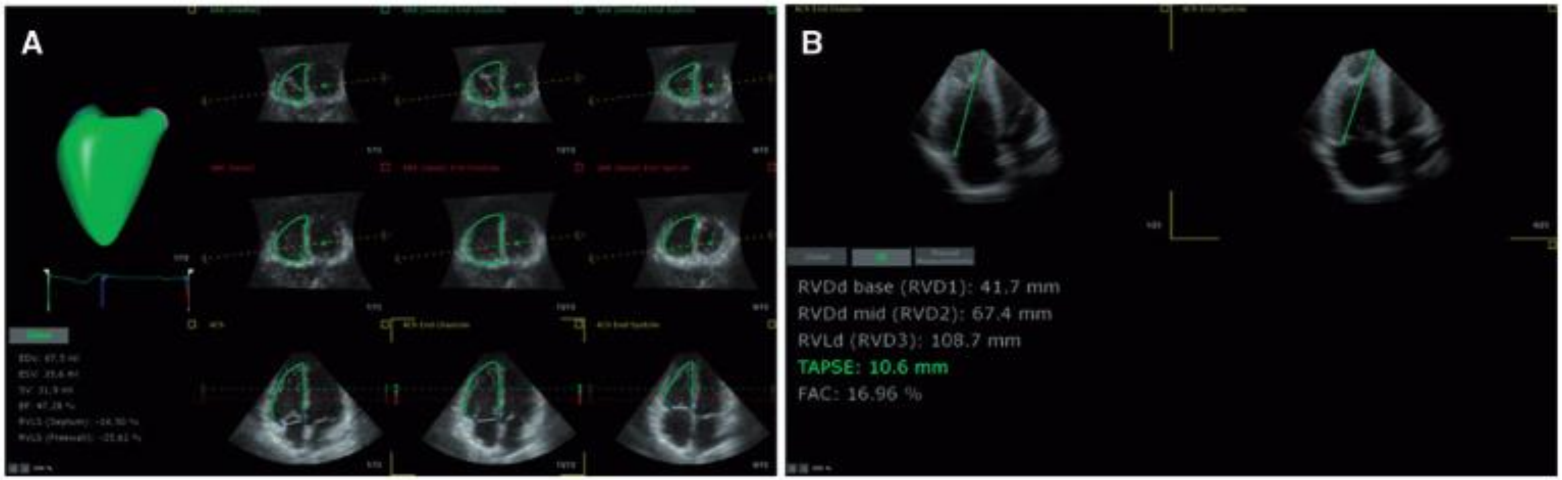
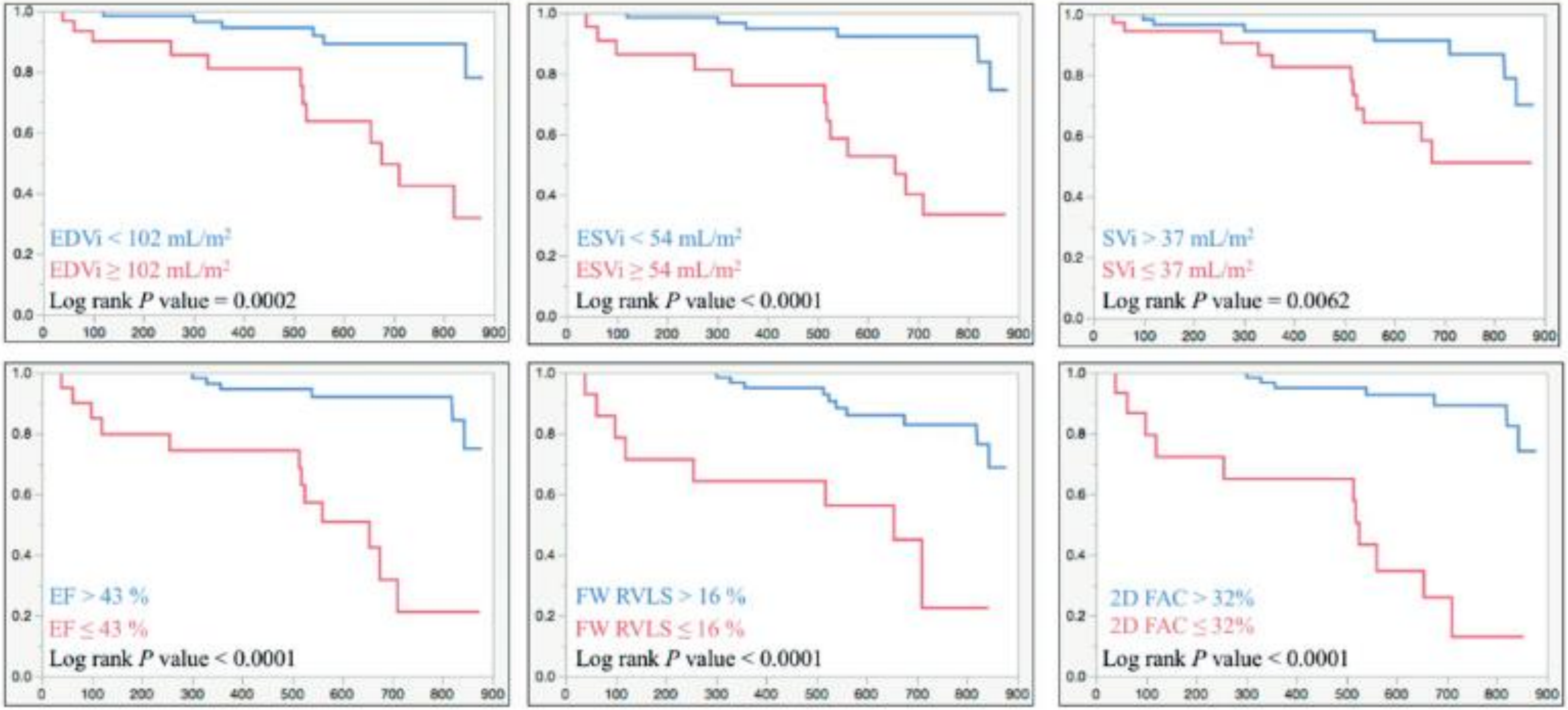


Figure 1 4D right ventricular function 2 software automatically generate 3D volumes, ejection fraction, right ventricular strain, tricuspid annular plane systolic excursion, and fractional area change.

		Adverse clinical events		
	All patients ($n = 96$)	**Event free ($n = 78$)**	**With event ($n = 18$)**	**P-value**
EDVi (mL/m^2)	83 (49–111)	75 (45–105)	101 (69–153)	0.0312
ESVi (mL/m^2)	41 (33–56)	40 (33–53)	44 (33–72)	0.0817
SVi (mL/m^2)	37 ± 13	36 ± 12	42 ± 16	0.1542
EF (%)	46 ± 5	47 ± 4	39 ± 7	0.0002

Credit: Jone 2017

RV 3D continued



RV numbers to remember

- 3D Ejection Fraction: 45%
- Free wall Longitudinal Strain: -16%?
- 2D Fractional Area Change: 35%
- S/D ratio: 1.0
- TAPSE – normal varies by age. Gotta look it up.

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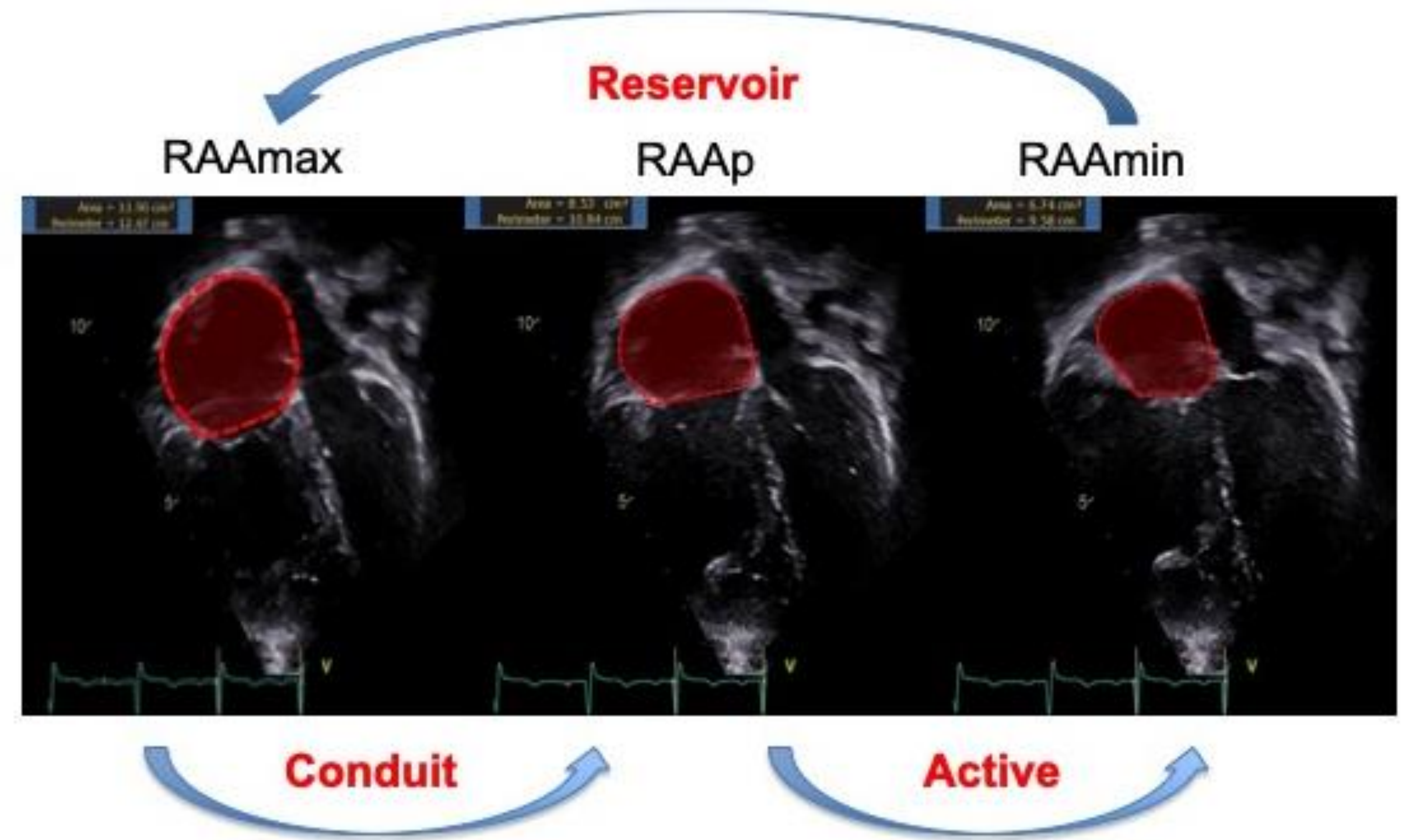
Part 4: RV diastolic function

- Gold standard is end diastolic pressure measured by cath.
- Diastolic measures with some validation in adult LVs are notoriously unreliable both in children and in RVs.
  - For example E/e' is not known to correlate with RVEDP or outcomes.
- Larger RA area (>18 cm^2 in adults) is associated with worse prognosis.
- During diastole (with tricuspid valve open) RA mechanics may give insight into RV diastolic performance.

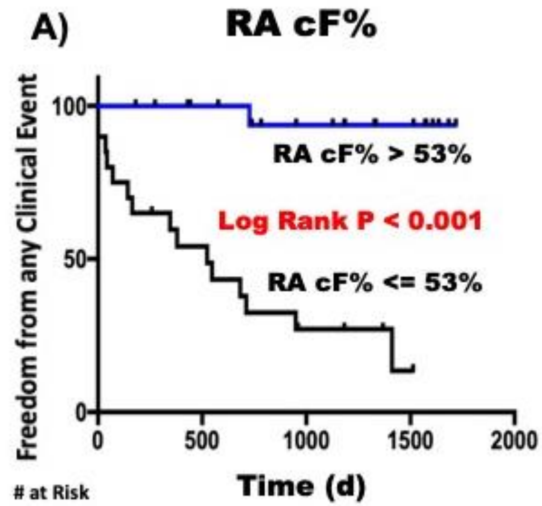
RA Conduit Fraction %



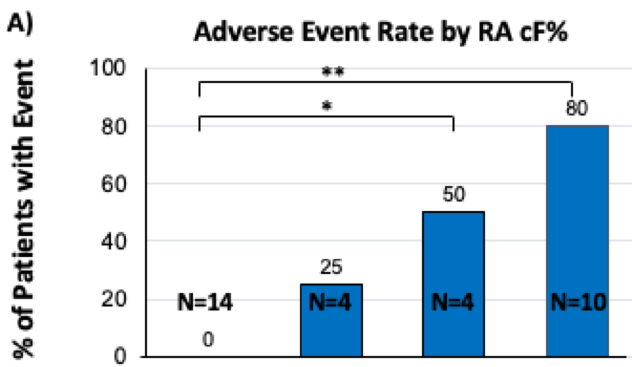
RAAC = RAAmax – RAA min

RA cF% = (RAAmax – RAAp) / RAAC

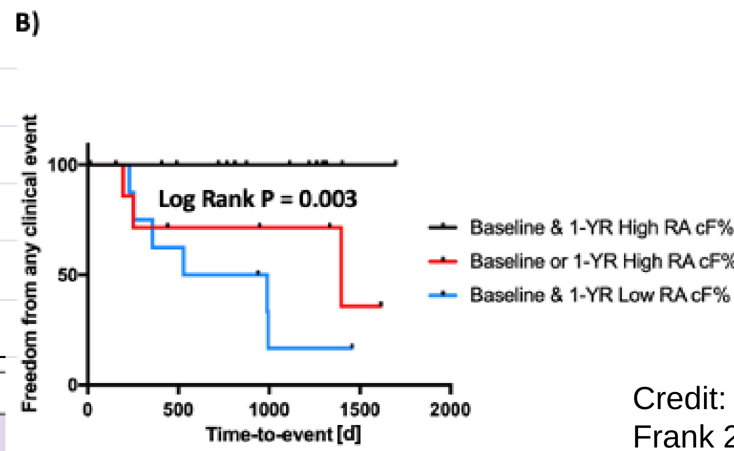
RA Conduit Fraction %



# at Risk				
All	41	29	18	9
>53%	20	17	12	7
<=53%	21	12	6	2



Baseline RA cF%	>60%	<60%	>60%	<60%
1YR RA cF%	>60%	>60%	<60%	<60%



Credit: Frank 2020, Frank 2021

Takeaways

- PH is not all about PA pressures – RV performance is very important too!
- Images to make sure to get:
  - RV-centric apical
  - PSAX with both pap muscles
  - CW of TR and PI anywhere you see it
  - Doppler all shunts!

The End

- Questions? Thoughts? Concerns?