Echocardiographic Assessment of PPHN

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The ESN is a project by the European Society for Paediatric Research

Faculty Disclosure

Nothing to disclose

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Learning goals

1. PPHN physiology

2. Echocardiographic evaluation of underlying physiology

3. Practical approach to echo in PPHN and integrating findings with treatment

Definition and Epidemiology

Persistant Pulmonary Hypertension of the Newborn (PPHN)

A syndrome of failed circulatory adaptation at birth due to delay or impairment of normal postnatal fall in pulmonary vascular resistance (PVR)

- 1-2:1000
- 90% 5-year survival
- Neurologic impairment in 15-25%

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Etiology

Secondary PPHN (80-90%)		
Maladaptation (Reactive vasoconstriction)	Maldevelopment (Remodelling)	Underdevelopment
Parenchymal lung disease • Pneumonia, MAS, RDS, TTN Stressfull stimuli • Perinatal asphyxia, hypothermia, sepsis Medication • SSRI	Alveolar capillary dysplasia Trisomy 21 CHD AV-malformations	CDH Lung hypoplasia • Oligohydramnion • PPROM Other lung anomaly

Idiopathic PPHN (10-20%)
Abnormal pulmonary vasculature (without known cause)

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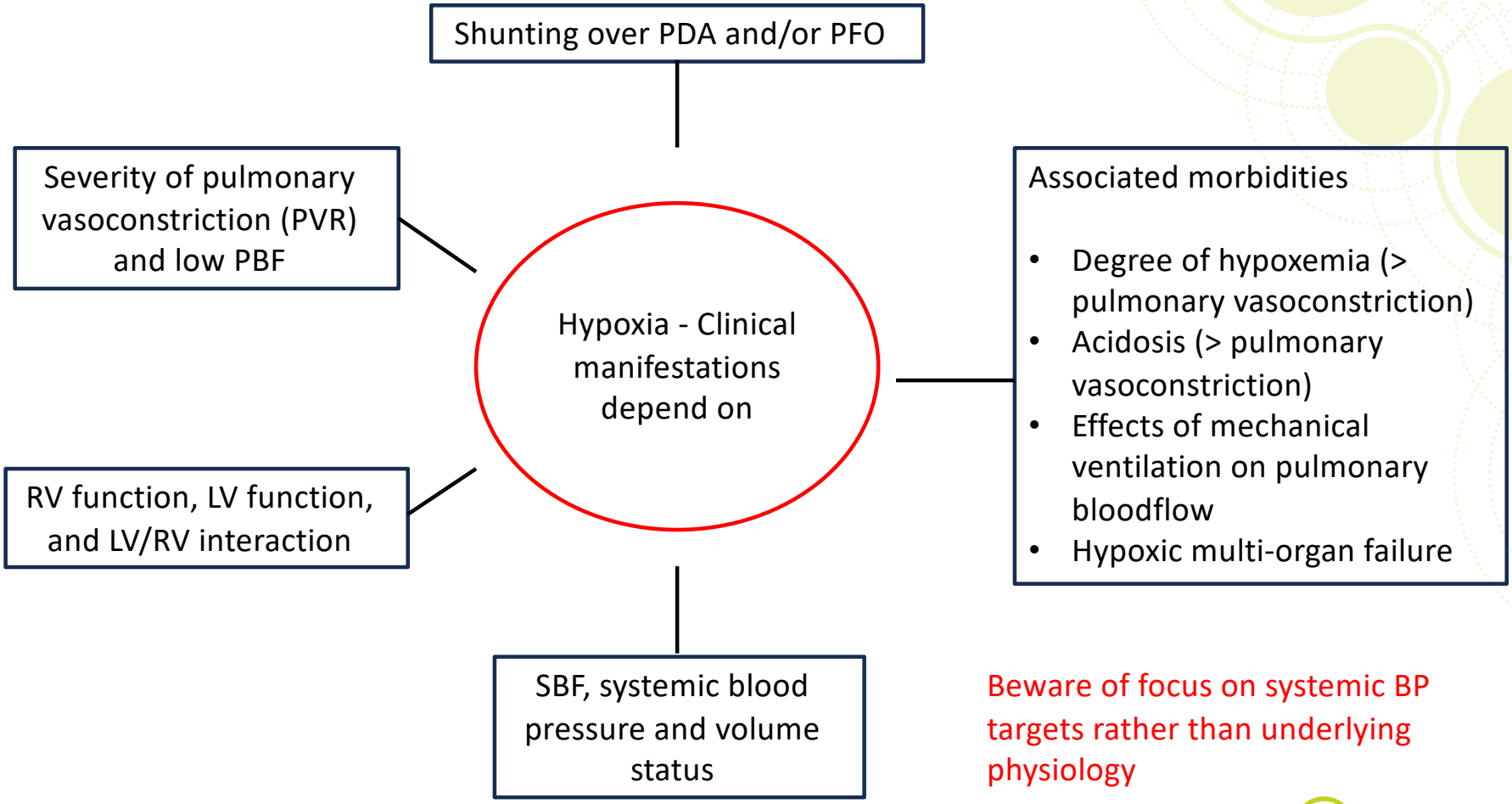

Beware of focus on systemic BP targets rather than underlying physiology

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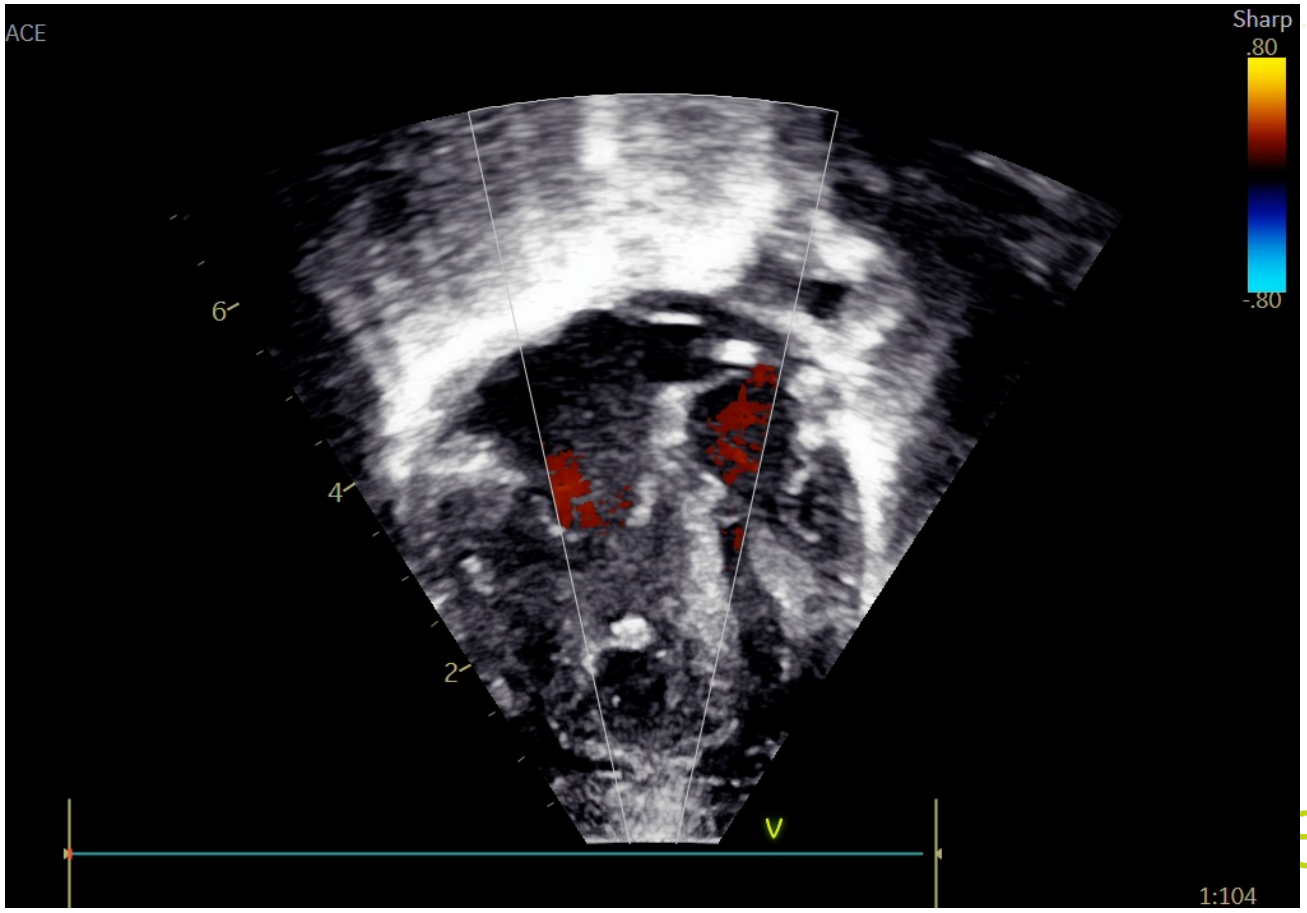
Term baby, MAS, intubated, iNO 20, Dopamine 10. DA closed



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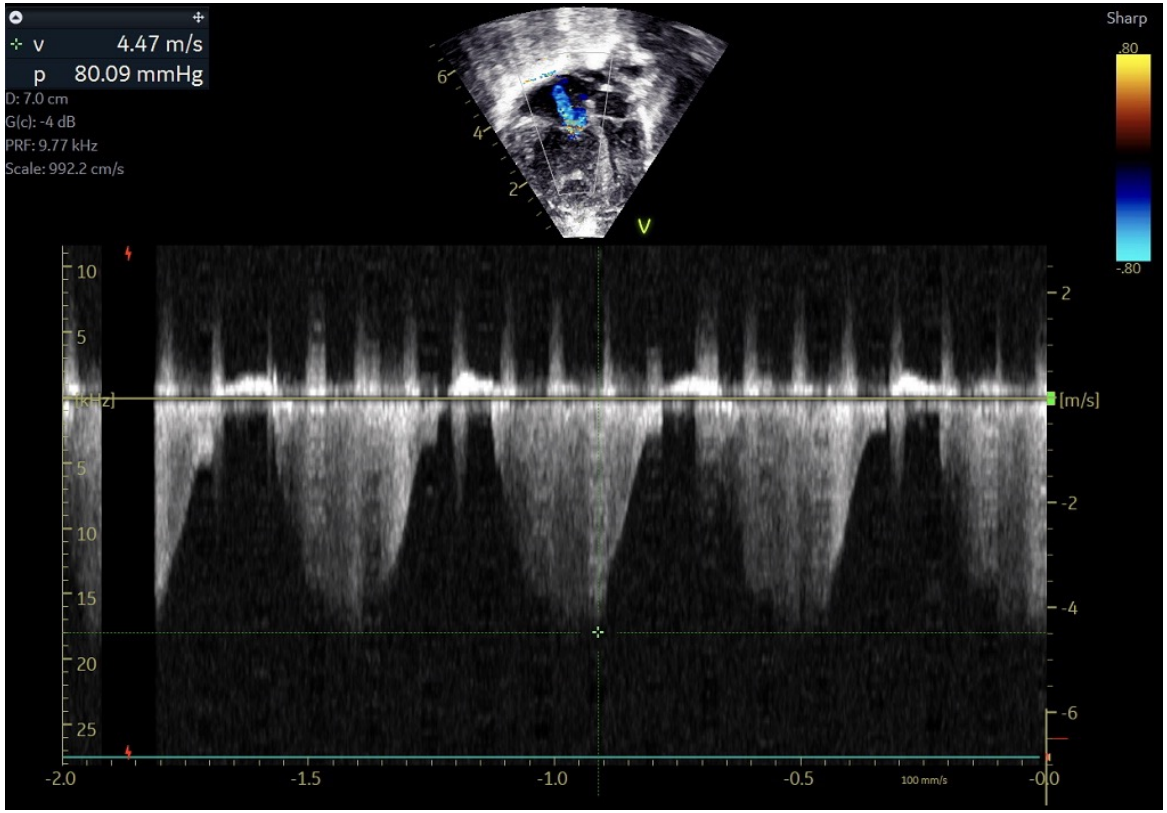

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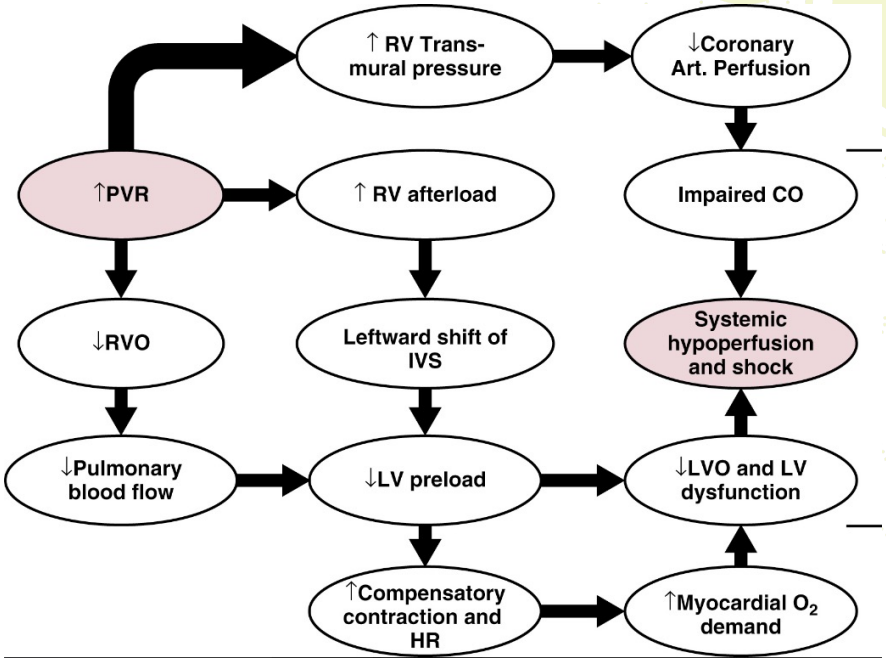
RV: High PVR > Increased RV afterload >

- RV dysfunction
  Worsened by reduced RV preload due to the right-left shunt over the PFO
- Decreased RV SV and RVO (PBF)
  > Ventilation-perfusion mismatch
  > Increased RV end-systolic and end-diastolic pressures > RV dilatation (to preserve stroke volume)
- Decreased coronary artery perfusion

(Offloading via PDA may help)



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Before PGD (closed DA)

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After PGD (open DA)

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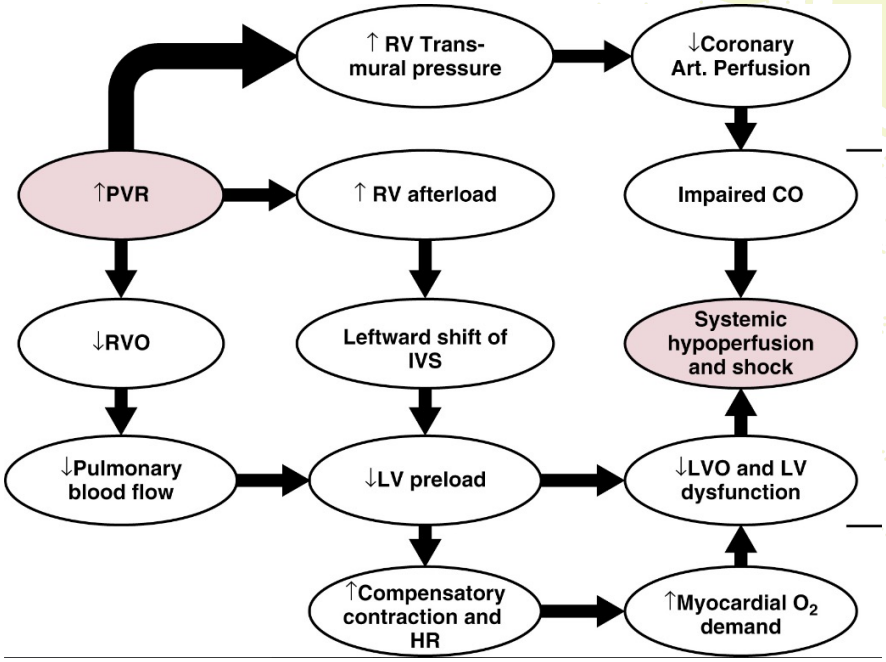
LV: Function may be compromised by

- Hypoxia
- Reduced LV preload (reduced pulmonary blood flow to the left atrium)
- Compression of the ventricular septum from the RV
- If LV afterload is increased (vasopressors) this may further compromise LV function



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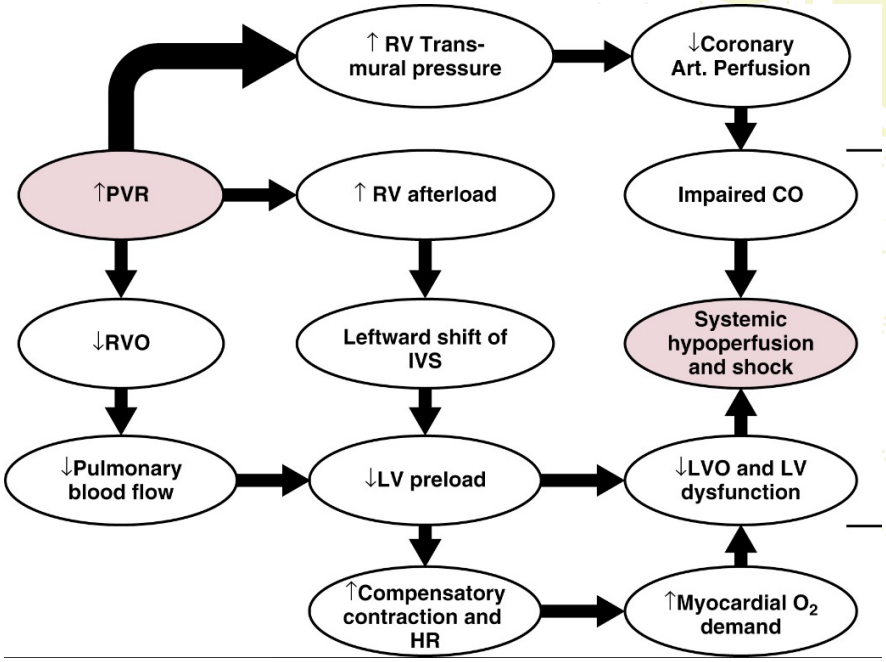
LV dysfunction >

- Systemic hypoperfusion and low systemic blood pressure
- Increased pulmonary artery pressure
- Pulmonary vein hypertension
- If Left-right shunt over PFO simultaneously with right-left shunt over PDA
  - Systemic circulation may be driven by the right ventricle
  - Increased LV end-diastolic pressure > Pulmonary venous hypertension
  - iNO may worsen pulmonary venous hypertension and/or cause reduced systemic blood flow



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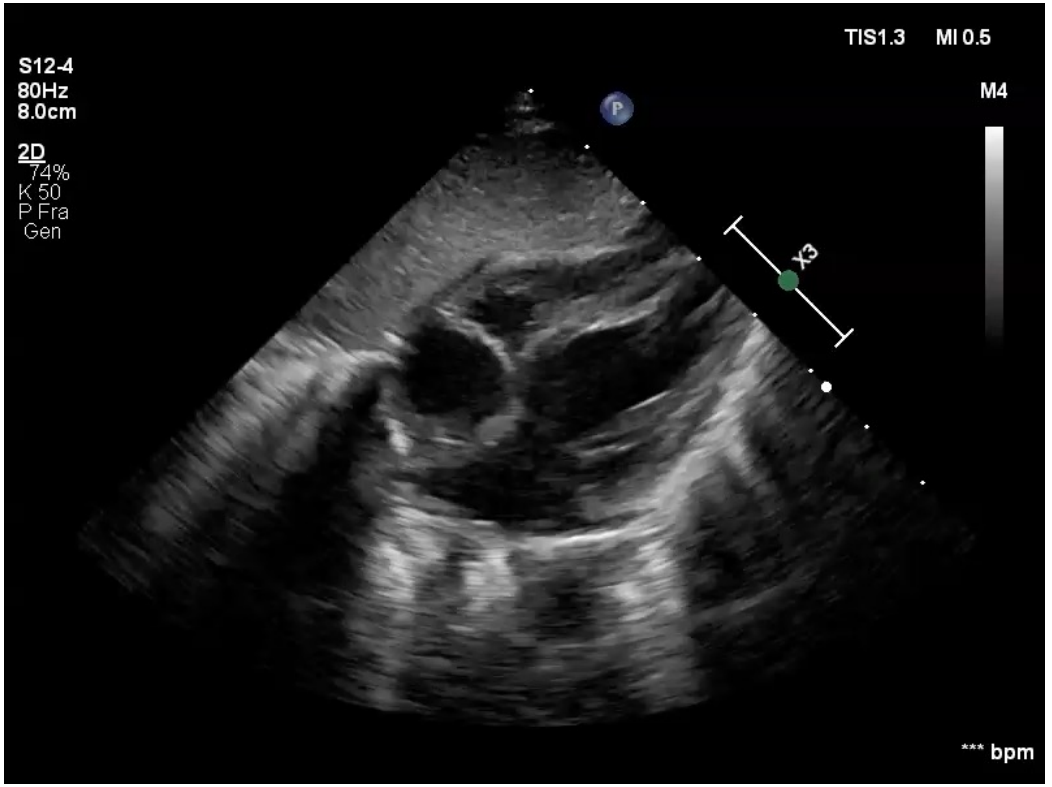
PPHN with LV dysfunction



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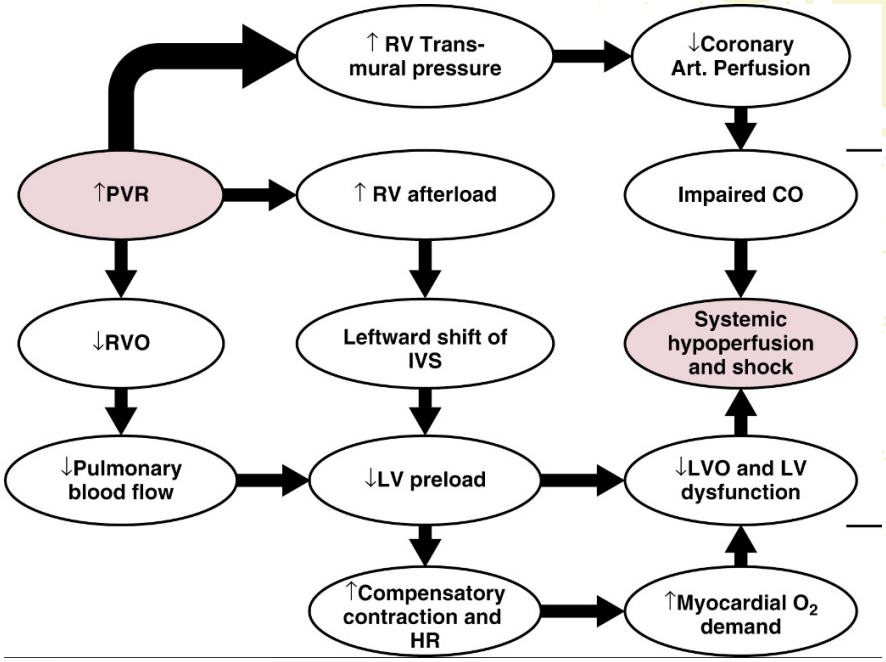
RV/LV interdepence >

- Decreased LV preload
- Large RV affects LV geometry and function
  > contributes to systemic hypotension and low LVCO
- LV is an important contributor to RV systolic function



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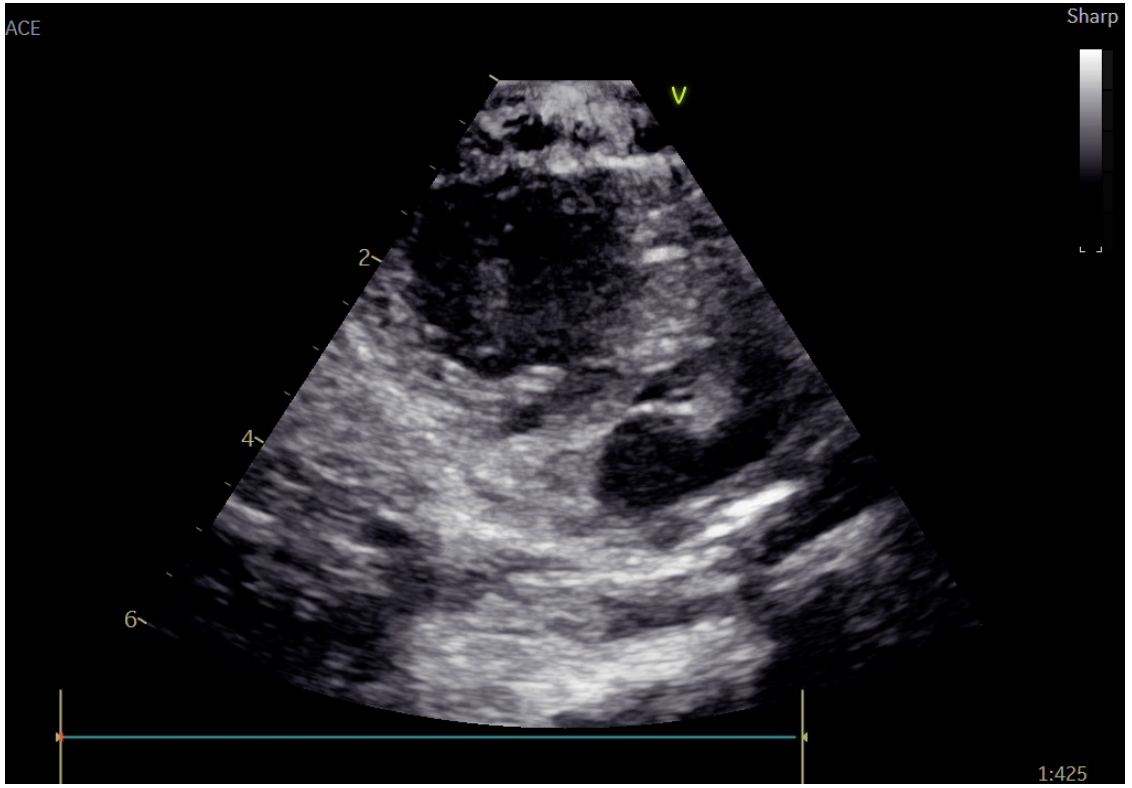

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Shunting over PDA & PFO

R-L ductal shunting

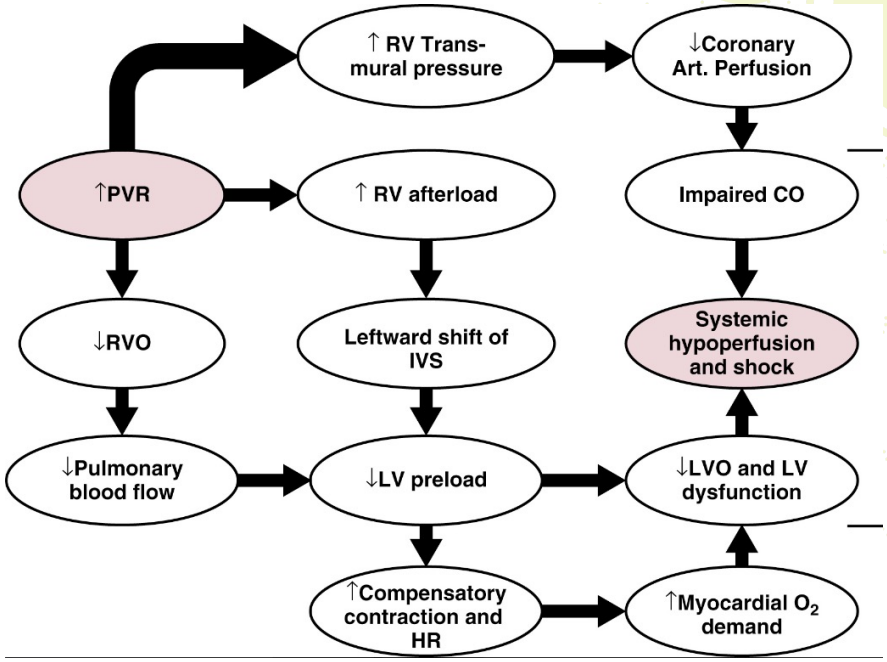
- Due to increased PVR
- May by exacerbated by systemic hypotension

PFO & PDA R-L shunting > worsening of hypoxia

- But may also support cardiac function and maintain cerebral pefusion when preload is severely decreased

PFO shunting

- R-L shunt may be essential to maintaining LV preload
- L-R shunting reduces LV preload



L-R shunting at PDA and PFO with severe hypoxia suggests intrapulmonal shunting

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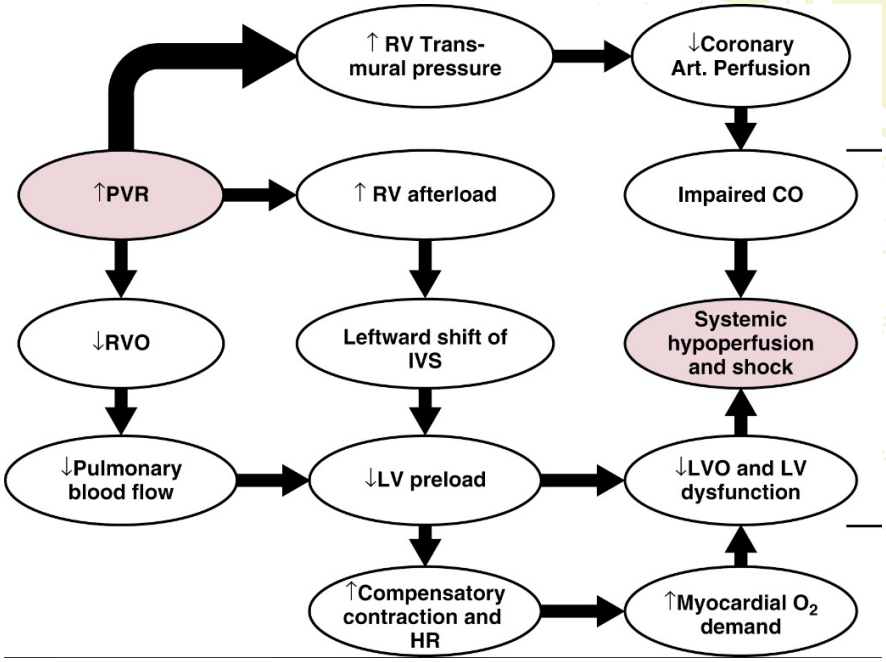
Systemic hypotension may be caused by

- LV dysfunction
  - Decreased preload
    - Reduced PBF
    - Hypovolemia
    - Fluid redistribution (sepsis)
  - Poor myocardial function
- Pulmonary vasodilators (Lusitropes) may also cause systemic vasodilation
- Cardiac output and SBP may not correlate



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Quiz (3 questions)

Goals of Echocardiography assessment

- Rule out CHD (requires expertise)
  - TAPVR, TGA, PA, TOF, Ebstein, CoA and more
- Make the diagnosis and grading the severity
- Identify pathofysiological mechanisms in the individual infant
- Target treatment
- Evaluate treatment response
- Asses onging hemodynamic changes
- When to wean therapy

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Overview of Echocardiographic Parameters

1. Pulmonary hemodynamics (PAP and PVR)
2. RV performance
3. LV performance
4. Shunts
5. Status of peripheral vasoregulation / volume

De Boode, et al. Pediatr Res 2018

Pulmonary hemodynamics (PAP and PVR)

PAP 'Quantitative' measures

TR peak velocity

sPAP ≈ RVSP = 4 x $(VmaxTR)^2$ + RAP

(RAP often assumed to be 5mmHg)

Transductal R-L flow peak velocity

sPAP ≈ RVSP = 4 x $(VmaxDA)^2$ + sSAP

Incorrect estimation with long DA

PR peak velocity

mPAP = 4 x $(VmaxPR)^2$ + RVdP

(RvDP often assumed to be 2-5 mmHg)

Normal systolic pulmonary artery pressure < 25mmHg reached within 2 months after birth

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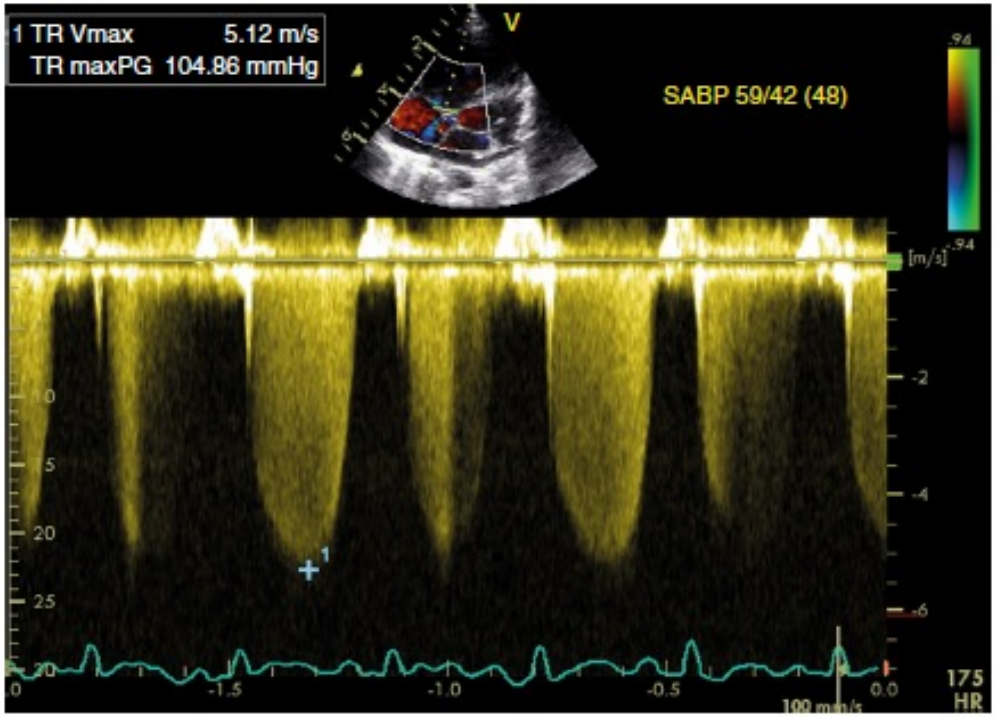

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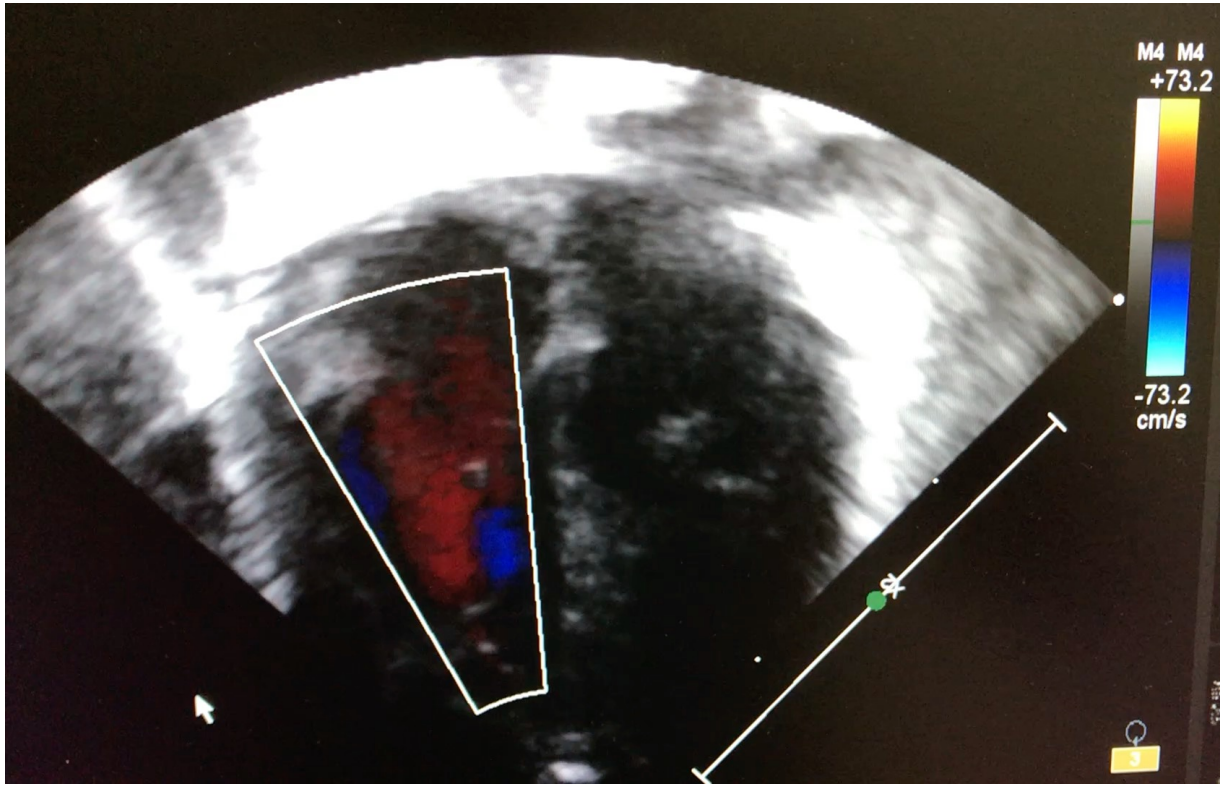
TR



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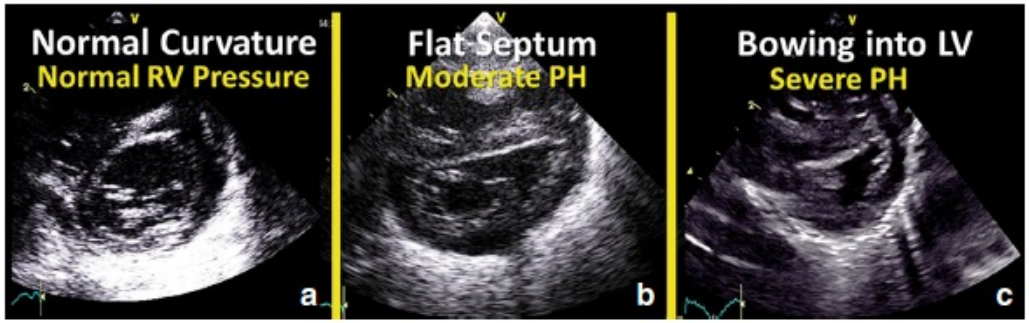
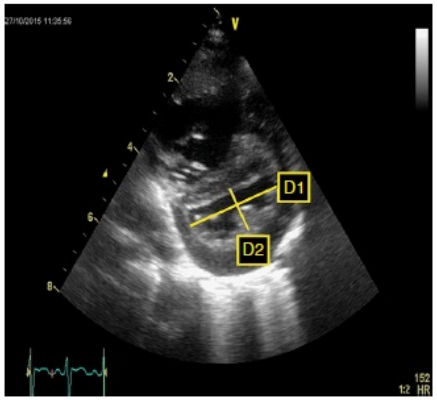
Pulmonary hemodynamics (PAP and PVR)

PAP Qualitative measures
IVS configuration
LV-sEI=D1/D2





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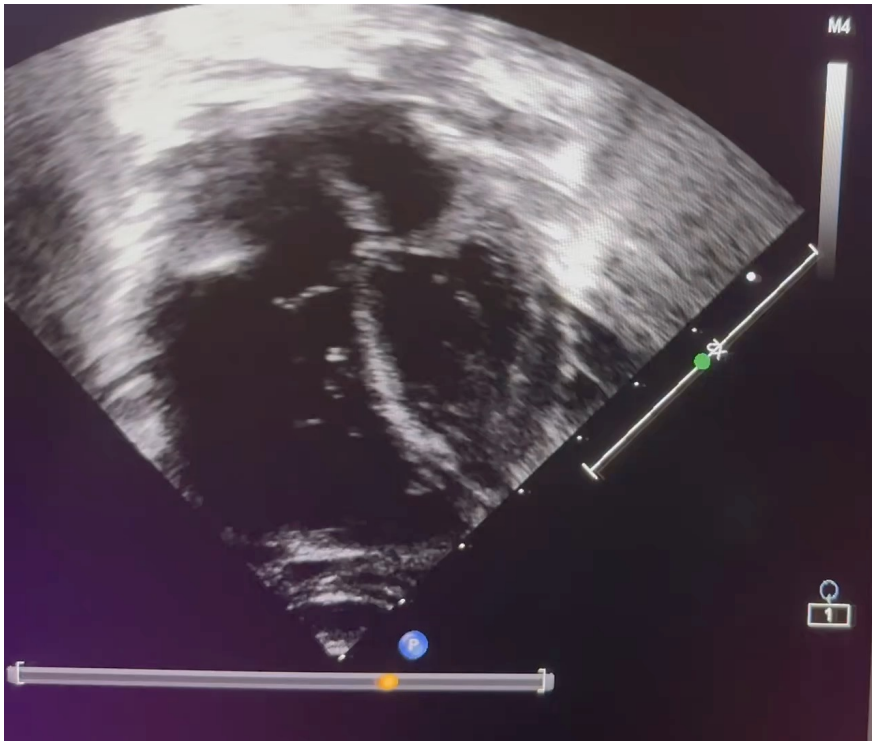
Pulmonary hemodynamics (PAP and PVR)



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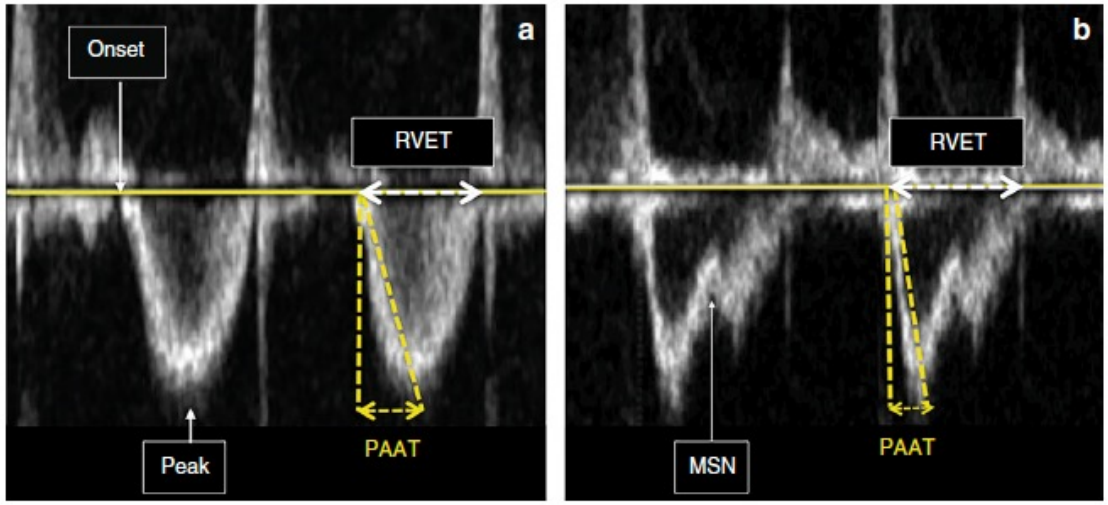
Pulmonary hemodynamics (PAP and PVR)

PVR

RV systolic time intervals

- PAAT
  - < 90ms abnormal
  - < 40ms severe PH
- PAAT/RVET ratio
  - < 0.23 indicates PH
- Altered RV output waveform
  - Midsystolic notch indicates PH



Normal PPHN

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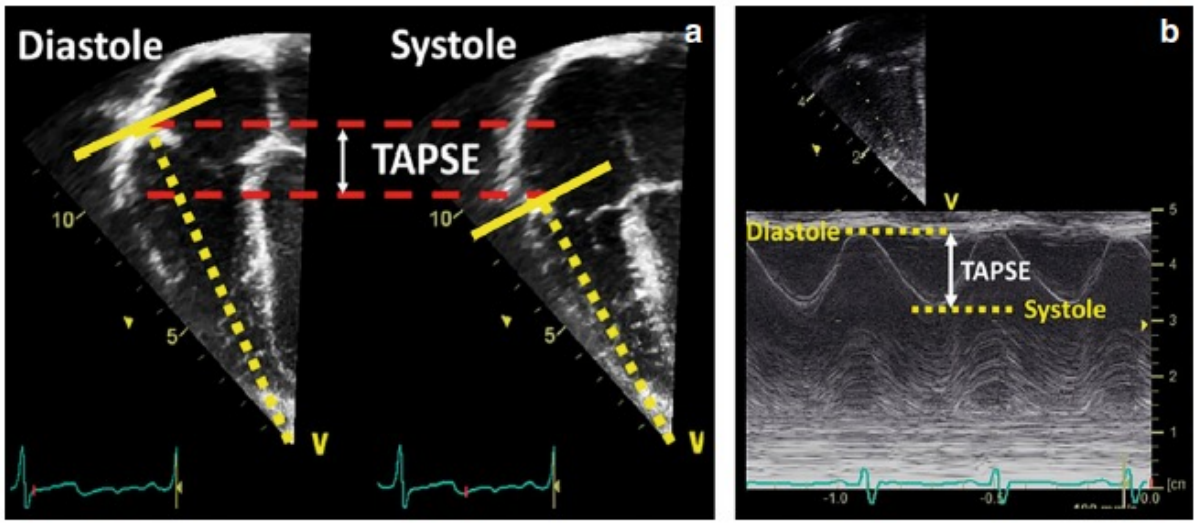
RV Performance

Systolic function

TAPSE

- A4CH
- TAPSE < 4mm associated with increased risk of ECMO and death in PPHN



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RV Performance

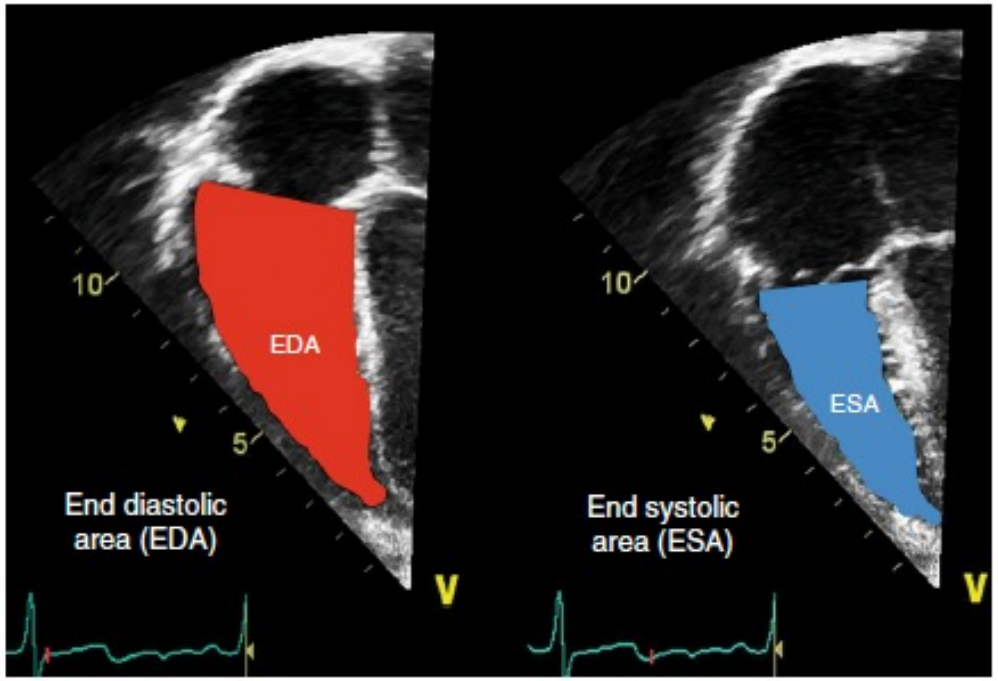
Systolic function

TAPSE

- A4CH
- TAPSE < 4mm associated with increased risk of ECMO and death in PPHN

Fractional area change (FAC)

- A4CH or A3CH
- FAC (%) = (EDA-ESA)/EDA



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RV Performance

Systolic function

TAPSE

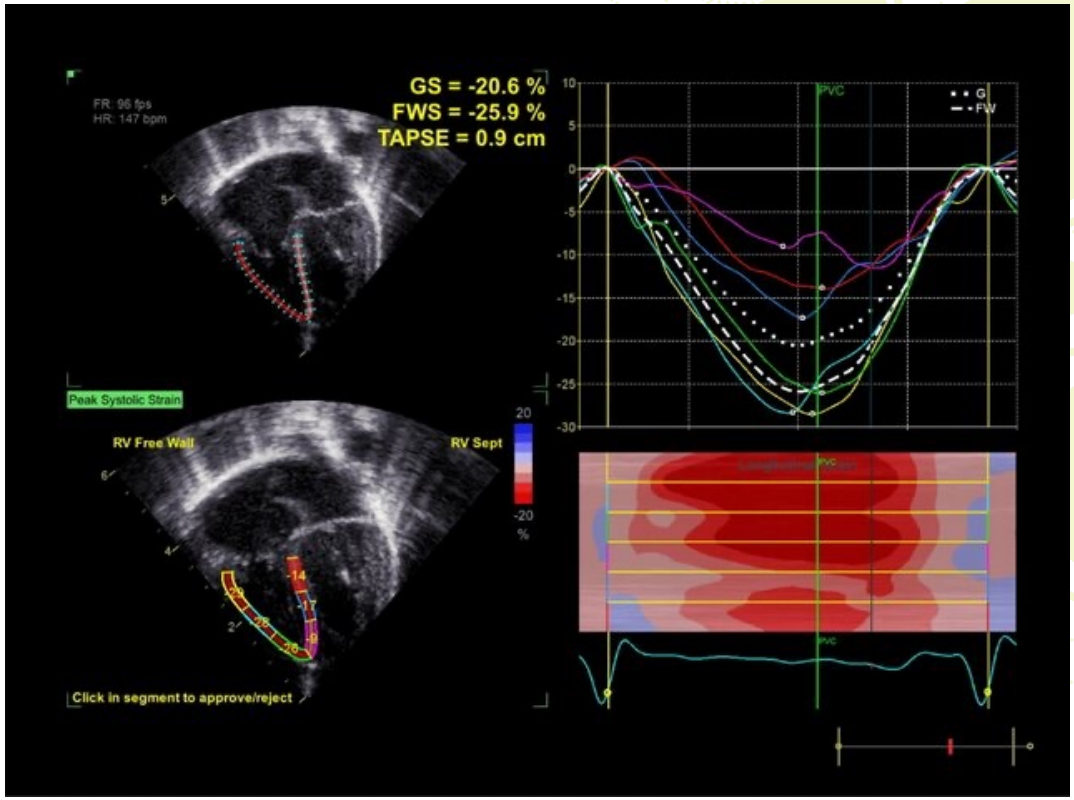
- A4CH
- TAPSE < 4mm associated with increased risk of ECMO and death in PPHN

Fractional area change (FAC)

- A4CH or A3CH
- FAC (%) = (EDA-ESA)/EDA

RV Strain

- 2D speckle tracking
  - RV global systolic peak strain associated with risk of ECMO or death
  - RV global longitudinal strain reduced in PPHN



August 2022. Pediatric Cardiology 43(6):1-11

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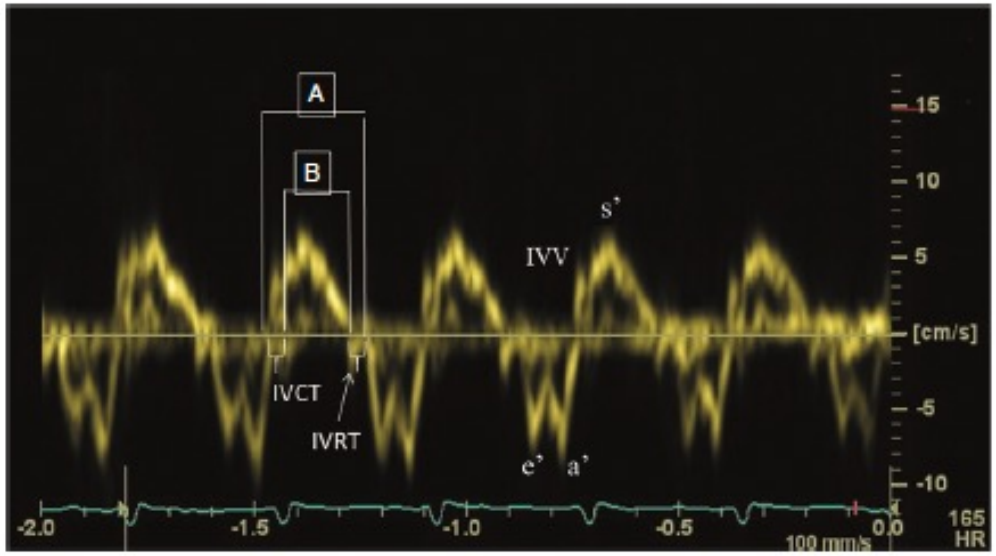
RV Performance

Global RV function

Myocardial performance index (MPI)

- = Tei index
- = Isovolumetric time / ejection time
- Global heart function
- RV-MPI = (IVET+IVRT)/RVET



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RV Performance

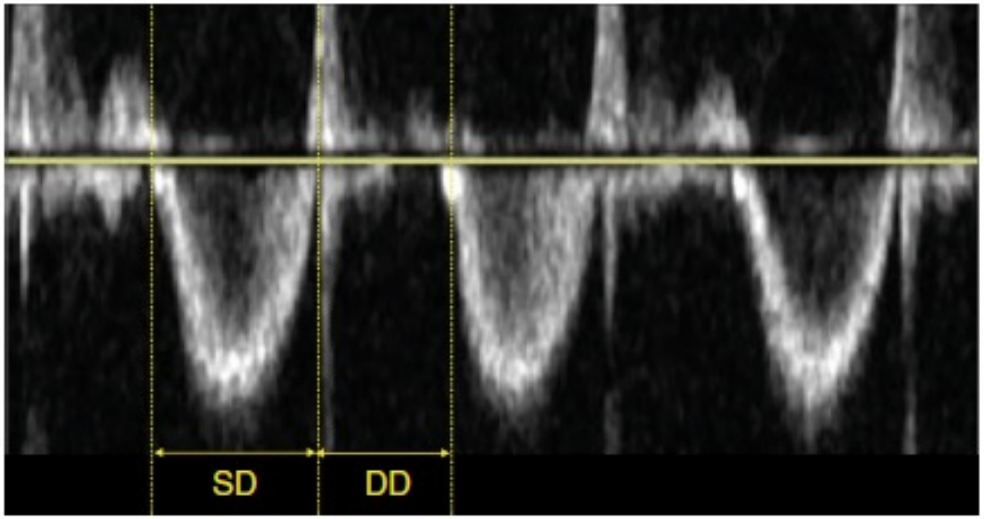
Global RV function

Myocardial performance index (MPI)

- = Tei index
- = Isovolumetric time / ejection time
- Global heart function
- RV-MPI = (IVET+IVRT)/RVET

RV S/D ratio

- Measured from TR wave
- Increased S/D ratio with increased afterload
- Related to sPAP and RV performance
- RV S/D > 1.3 associated with ECMO or death



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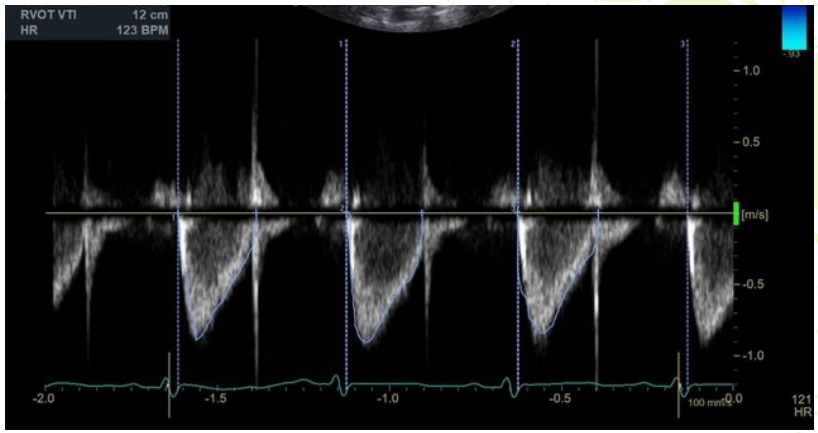
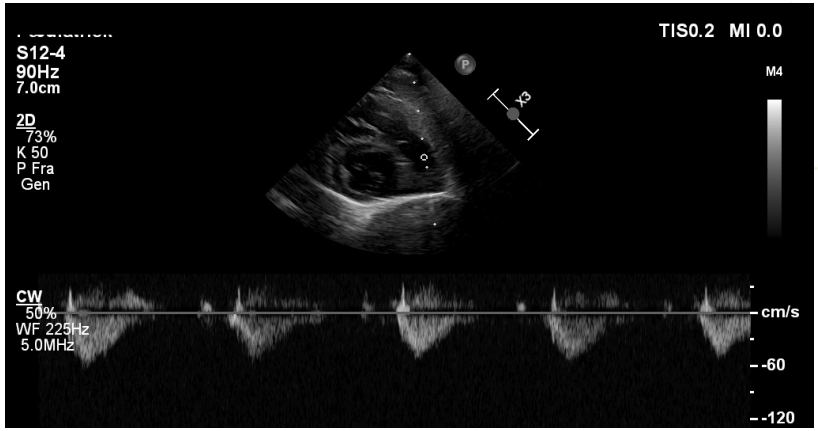
RV Performance

August 2022. Pediatric Cardiology 43(6):1-11

Reduced PBF

- RVOT VTI trend
- RVC0 often 'contaminated' by shunts
- Reduced LPA velocity is predictive of a good responde to iNO





RV Diastolic function (less studied)

Strain

RA dilatation

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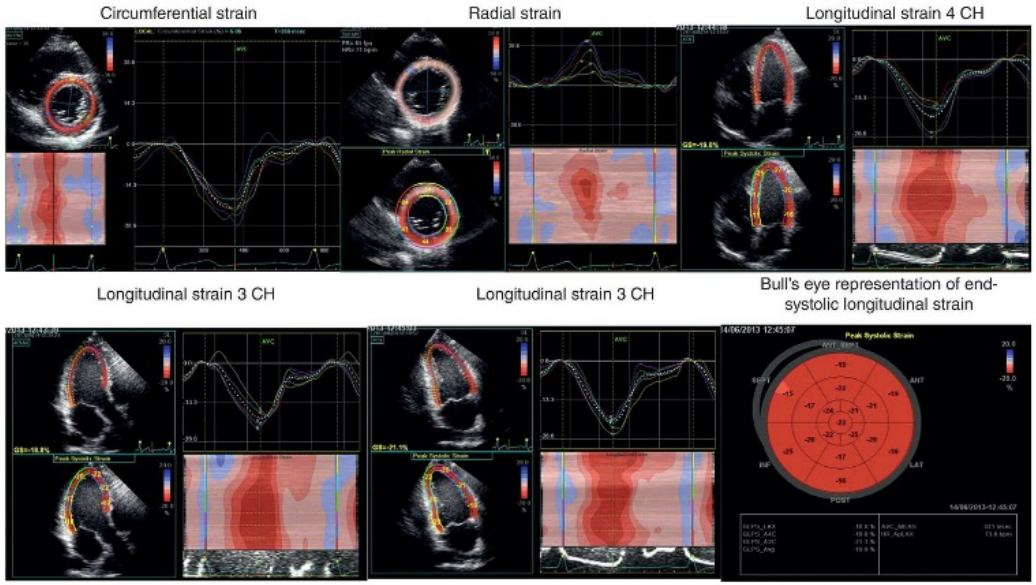
LV Performance

Systolic function
2D ST strain (strain rate)
LV-SV
LVCO

Diastolic function
MV E/A ratio
2D ST strain (strain rate)
Tissue velocities (E', A')

Global function
LV-EF (Biplane Simpson)
LV-MPI
Torsion



Echocardiography in Pediatric and Congenital Heart Disease. Wiley.

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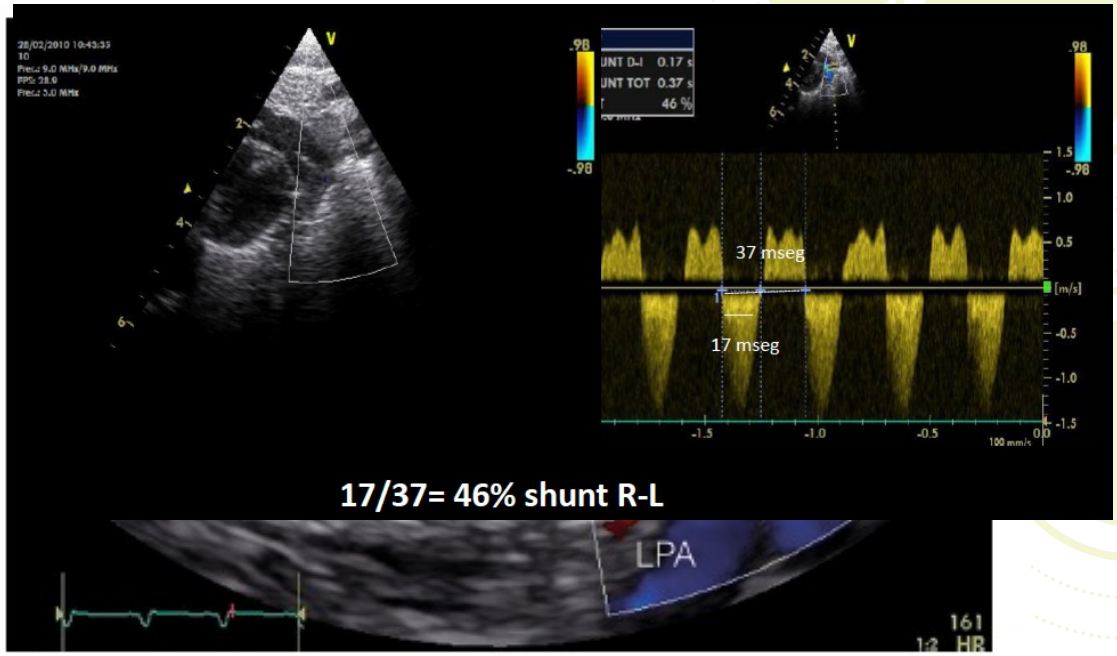
Shunts

Shunting over PDA

- R-L shunt in 70-90% of PPHN
- > 30% R-L shunt indicates PPHN

Shunting over PFO

- R-L shunt in 70-100% of PPHN



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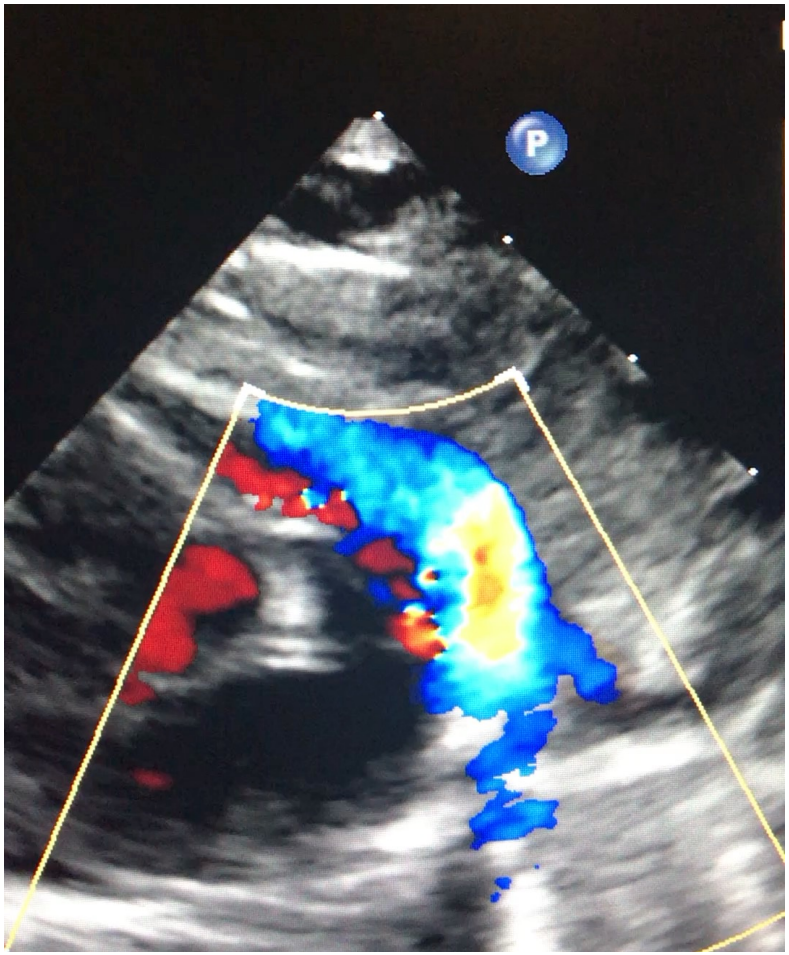
Shunts



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Practical approach to NPE in PPHN

1. Rule out CHD
Cardiologist

2. PAP assessment
TR peak velocity

3. RV performance and geometry
TAPSE
2D ST Strain
PAAT/RVET ratio
RV/SD ratio
RV FAC
RV VTI trend

4. LV performance
LV-MPI
LV-SV
LVCO
LV-EF (Biplane Simpson)
2D ST strain

5. DA Shunt
Shunt direction

5. PFO Shunt
Shunt direction

6. Peripheral vasoregulation / volume
Heart filling (end-diastolic diameter)
IVC diameter and collapsibility

7. Focused serial re-evaluation

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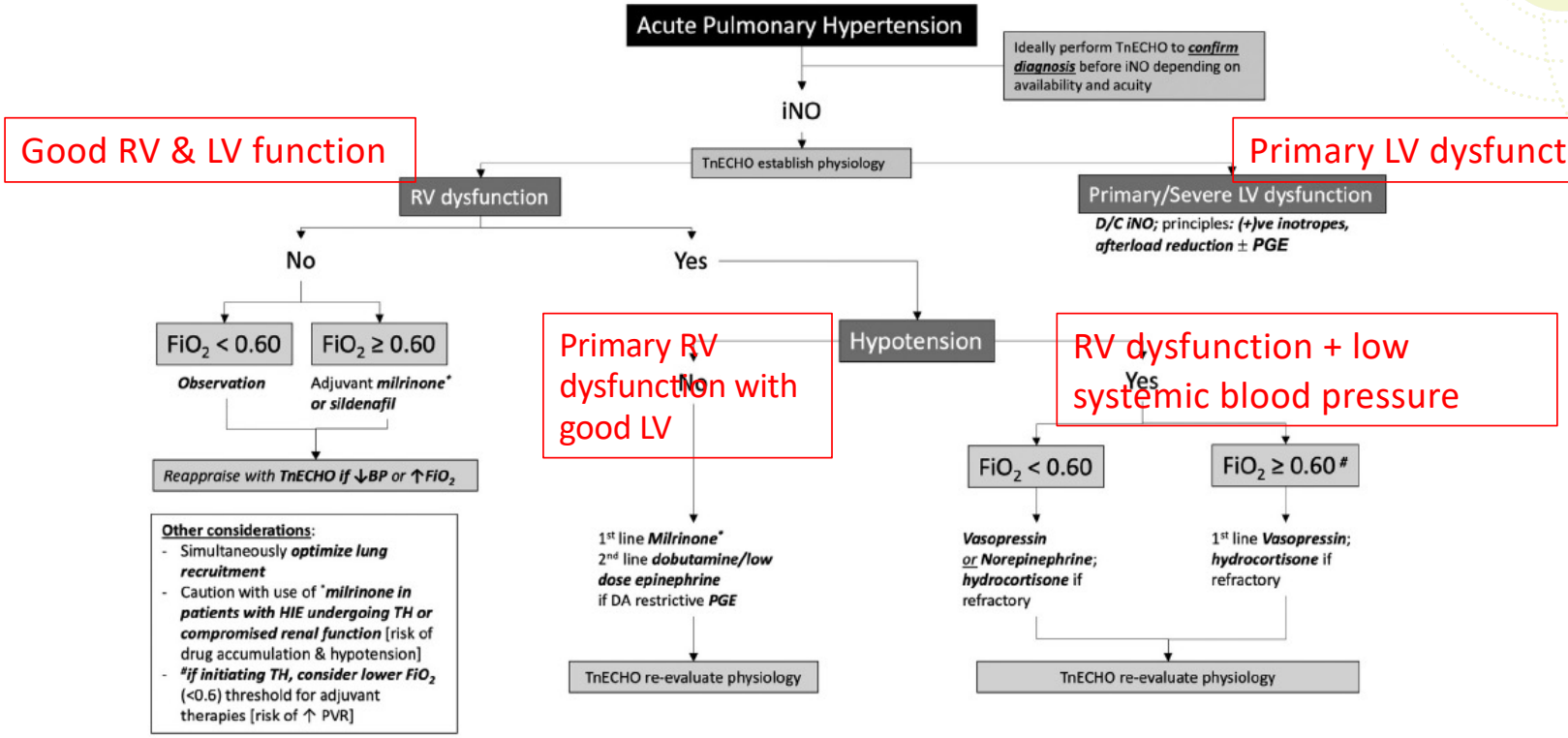
Integrating finding with treatment



McNamara, Giesinger, and Lakshminrusimha. jpeds.2022.03.022

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Some pitfalls

- Pulmonary venous stenosis > pulmonary venous hypertension, may mimick 'Classic' PPHN
- Cardiac hypertrophy (infant of mother with diabetes)
  - Volume (avoid inotropy or tachycardia)
- HIE
  - CAVE reduced renal drug clearance
  - TH may increase PH
- iNO
  - Infants with low systemic blood pressure, myocardial impairment and respiratory component contributing to hypoxia are less likely to respond well
- Clinical deterioration after iNO start
  - Primary LV dysfunction may be worsened by iNO
  - TAPVR
  - CHD

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Two Cases

What have we learnt?

1. The use of iNO in an unselected population without the use of echocardiography, may be dangerous
2. Confirmation of normal LV systolic performance is essential
3. Indiscriminate use of iNO may reverse the transductal shunt, and increasing PBF at the expense of SBF in infants with LV dysfunction
4. Key point: coexistence of R-L PDA shunt and L-R atrial shunt associates with moderate to severe LV dysfunction

Take-home messages

- PH diagnosis is challenging
- Echocardiography is a non-invasive test for the initial diagnosis and serial follow-up
- Echocardiographic assessment of the right and left heart performance
- Exclude CHD!

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Thank you for your attention

Any questions?

Thank you for your attention!

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