

# Neonatal Pulmonary Hypertension

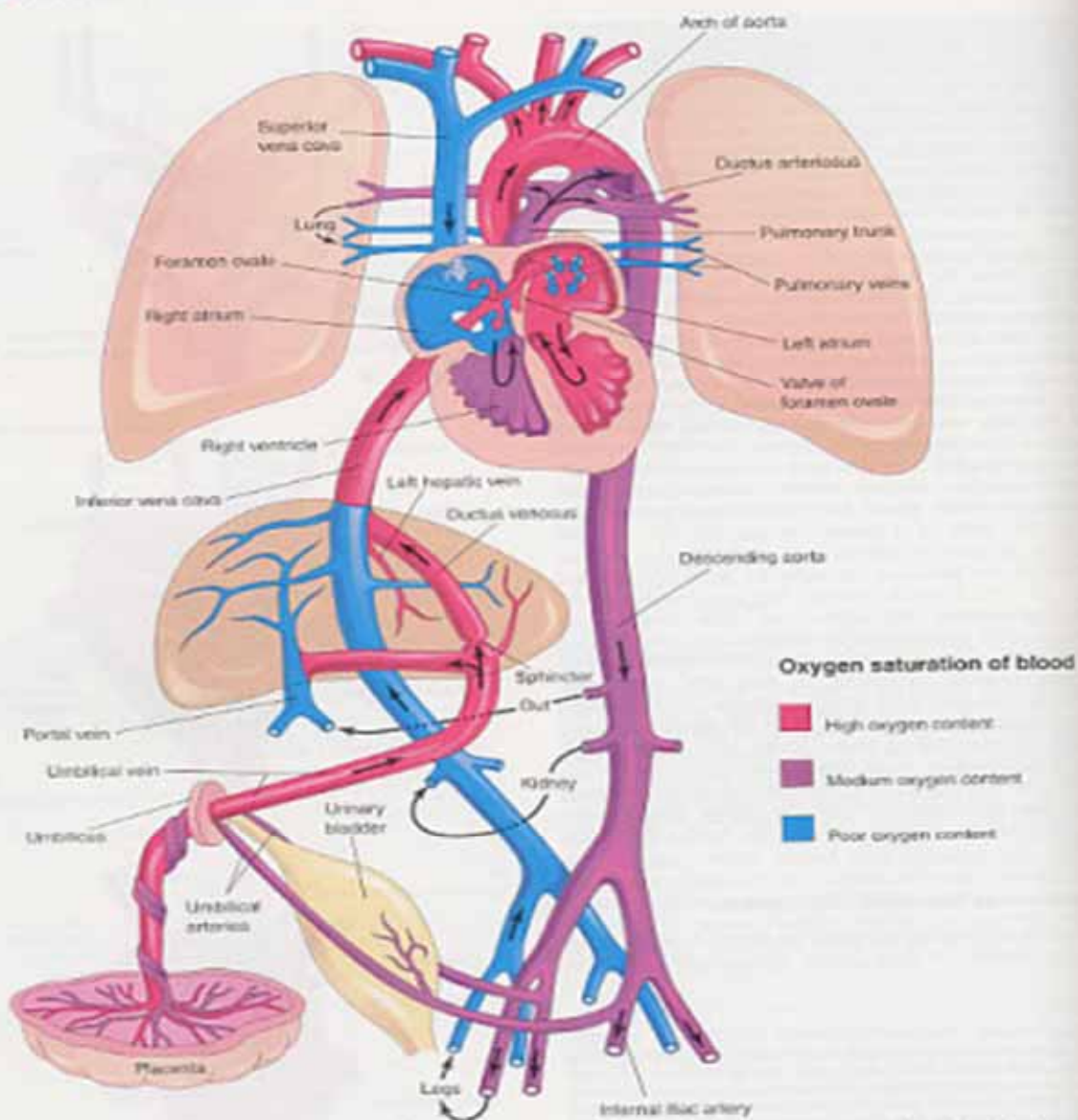
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# Definition

- ▶ PA pressure outside neonatal period =  $14 \pm 3$  mmHg
- ▶ Children/adults PH = PAP  $\geq 25$  mmHg
- ▶ Neonate relatively pulmonary hypertensive
- ▶ PA pressure = PVR X PBF
- ▶ Brisk fall of PVR at birth; 6 weeks to reach adult values





■ **Figure 15-37.** Schematic illustration of the fetal circulation. The colors indicate the oxygen saturation of the blood, and the arrows show the course of the blood from the placenta to the heart. The organs are not drawn to scale. Observe that three shunts permit most of the blood to bypass the liver and lungs: (1) ductus venosus, (2) foramen ovale, and (3) ductus arteriosus. The poorly oxygenated blood returns to the placenta for oxygen and nutrients through the umbilical arteries.

# Causes of Neonatal PH

- ▶ A: PPHN

Pulmonary vasoconstriction: MAS, RDS, pneumonia

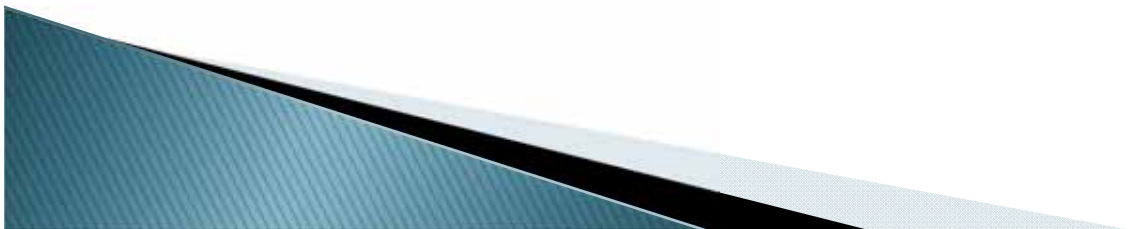
Reduced pulmonary vascular bed: pulm hypoplasia  
(CDH; Oligohydramnios)

Abnormal pulmonary vascular remodelling:  
idiopathic (IDM, maternal NSAID)

- ▶ B: Pulmonary vein stenosis

- ▶ C: Secondary to left heart disease – MV stenosis

- ▶ D: Secondary to lung disease or hypoxia – BPD



# Case Presentation

»» Baby DC



# Maternal Hx

- ▶ 31 years old G3P2

- ▶ POH:

2006: NVD at 37 wks, Clinique Darne – male infant – NCPAP and medical closure of PDA

Subsequent NVD term female infant – no problem

- ▶ Elective CS at 37 wks for breech



# Birth History

- ▶ Elective LSCS breech on 26.01.12 at Clinique Darne
- ▶ Gestation 37 wks; Bwt 3.16kg
- ▶ Cyanosed at birth and needed bag/mask ventilation <1 min. Apgar 6<sup>1</sup> 9<sup>5</sup>
- ▶ Nasal cannula oxygen
- ▶ Severe desaturation on crying – “normal” cardiac echo
- ▶ Transferred to ABH 27.01.12 – age 28 hrs – D2 – NP CPAP



# Respiratory System

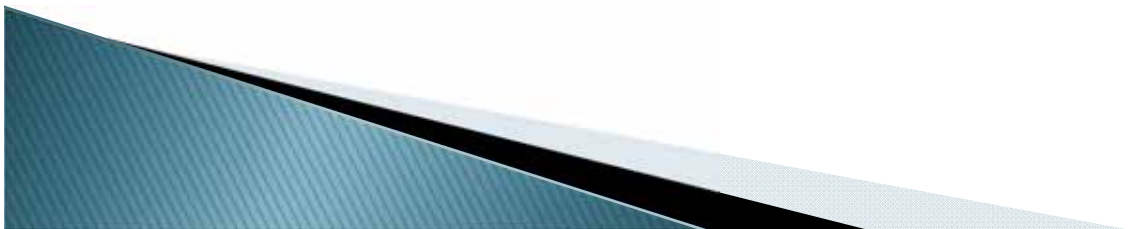
- ▶ High FiO<sub>2</sub> on NCPAP
- ▶ CXR – RDS
- ▶ InSurE – 1<sup>st</sup> dose of survanta
- ▶ Ventilated 5 hrs after admission
- ▶ 2<sup>nd</sup> dose survanta 12 hrs after 1<sup>st</sup> dose – D3
- ▶ Remained unstable with frequent desaturation – Max PIP 24, max FiO<sub>2</sub> 90%





# Cardiovascular System

- ▶ Cardiac echo – moderate PDA and severe pulmonary hypertension (PPHN)
- ▶ Ventilation management for PPHN; Inotropes
- ▶ Ibuprofen for PDA
- ▶ 29.01.12 – D4 – Sildenafil started
- ▶ Gradual improvement
- ▶ Extubated on 31.01.12 – D6
- ▶ NCPAP until D7
- ▶ Nasal oxygen until 05.02.12 – D11
- ▶ Sildenafil stopped D11
- ▶ Cardiac echo 06.02.12 – D12 – no PDA or PH



# Others

- ▶ CNS: Subtle convulsion – phenobarbitone.
- ▶ Cranial USS 06.02.12 – D12 – Normal
- ▶ Sepsis: ABs for 9 days. Cultures negative.
- ▶ GIT: Trophic feeds D5; oral feeds D7; full enteral feeds D9. Breast feeding well on discharge.
- ▶ Discharged on 09.02.12 – D15.
- ▶ Now age 7m – normal devt/growth



# Persistent Pulmonary Hypertension of the Newborn

» PPHN



# Presentation

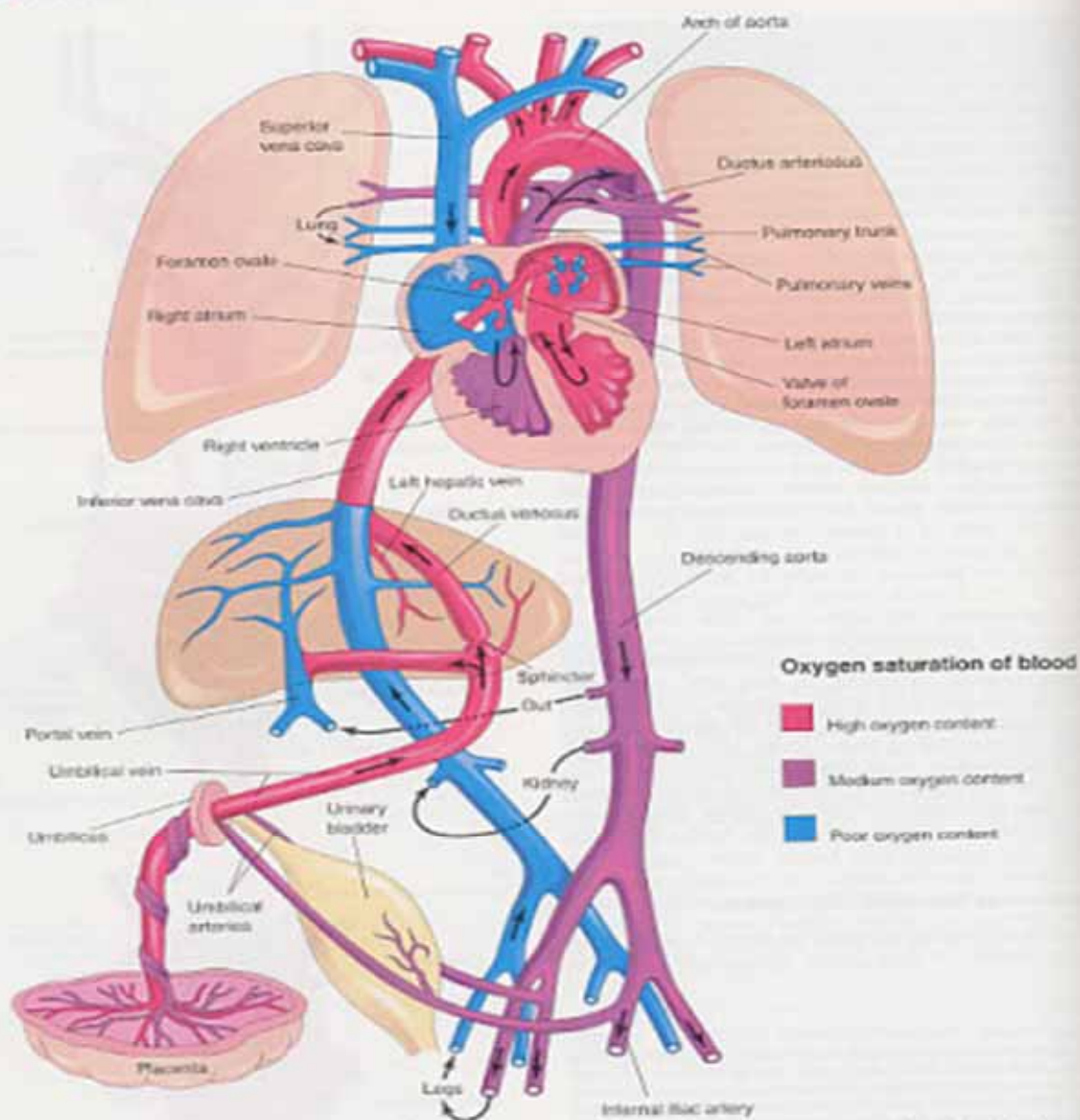
Near-term, term, post-term infants; LSCS;  
2/1000 livebirths

Poorly perfused/cyanosed within few hours of  
birth

Cyanosis refractory to supplementary O<sub>2</sub>;  
Escalation of Rx → IPPV

Pulmonary hypertensive crisis

Mortality 10–20%; Handicap 12–25%



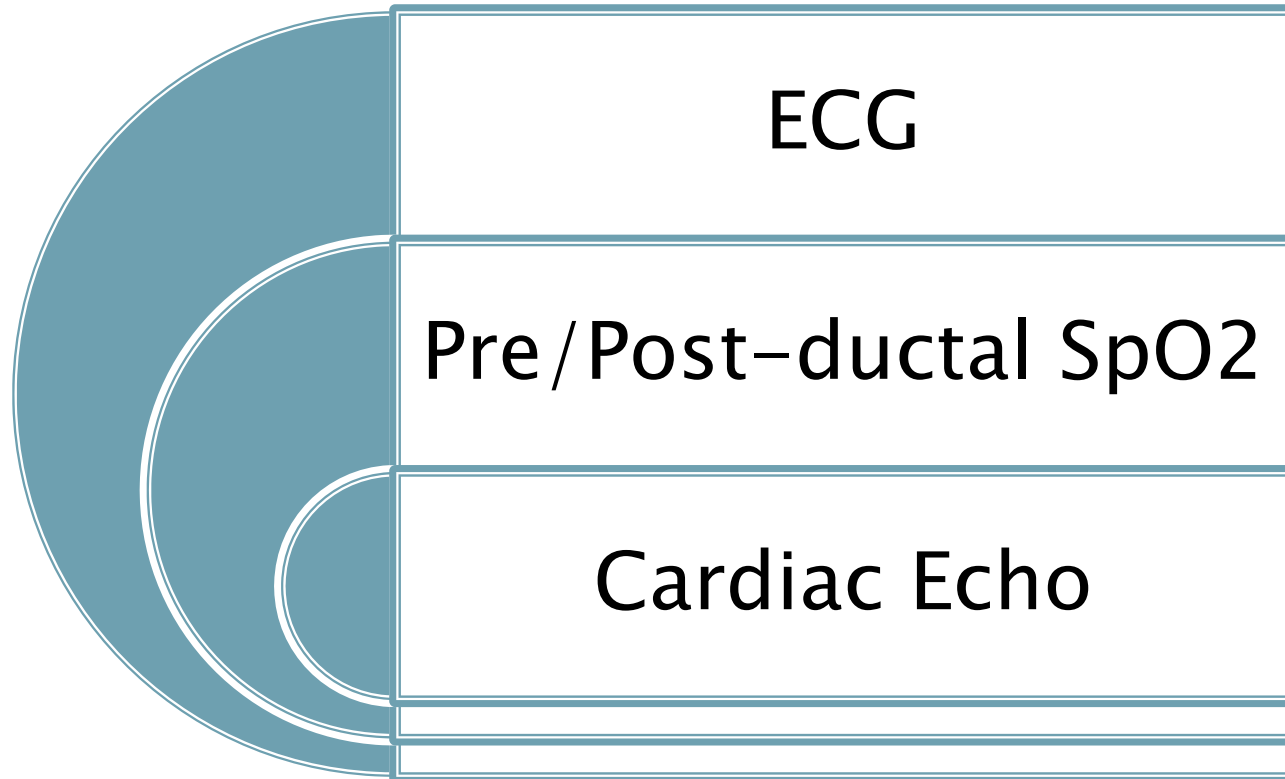
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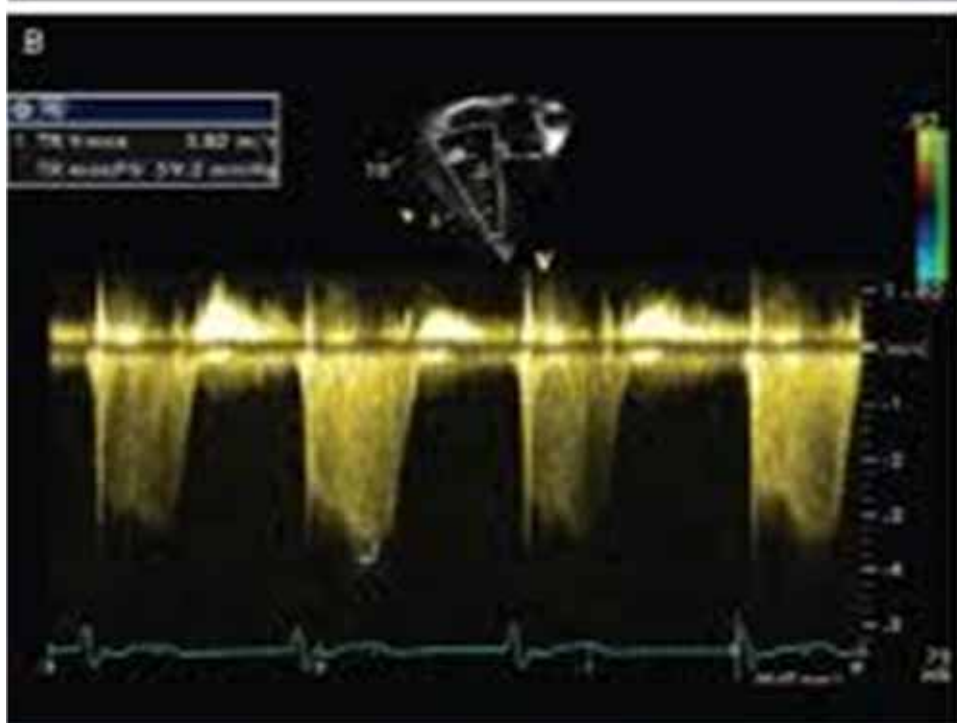
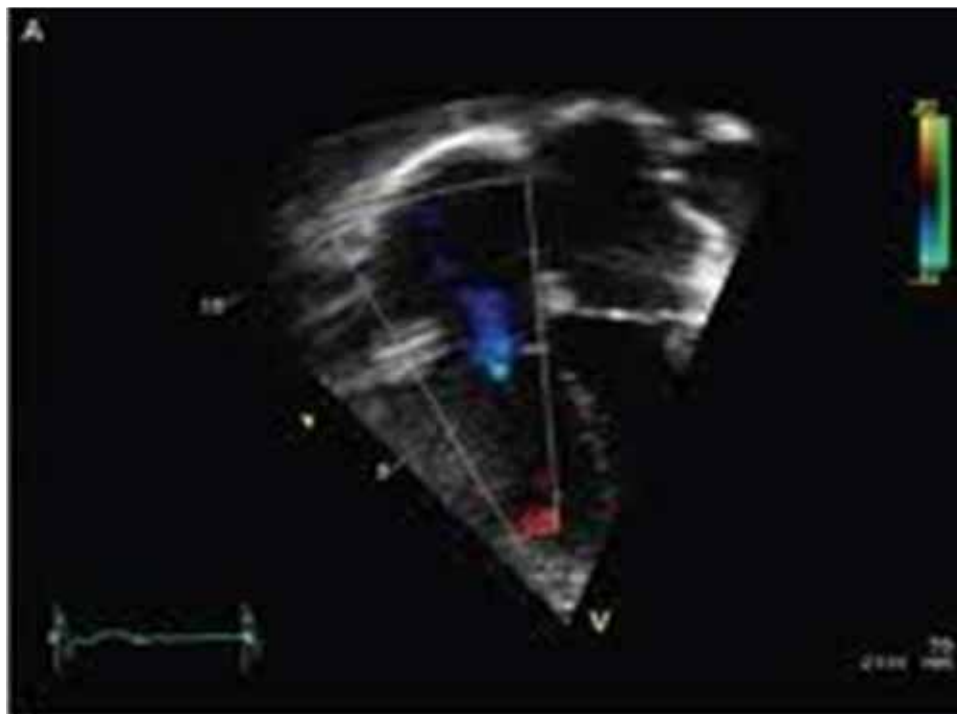
# Differential Diagnosis

- ▶ Septicaemia
- ▶ Cyanotic CHD
  - Pulmonary atresia
  - Hypoplastic LHS
  - TGA
  - TAPVD
  - Ebstein anomaly



# Diagnosis







## Tricuspid Incompetence.

A proportion of babies have physiological tricuspid incompetence (TI) and it is more likely to be present if pulmonary artery pressure is high. When present, this is probably the most accurate method. The problem with this method is that, even when there is pulmonary hypertension, many babies don't have it.

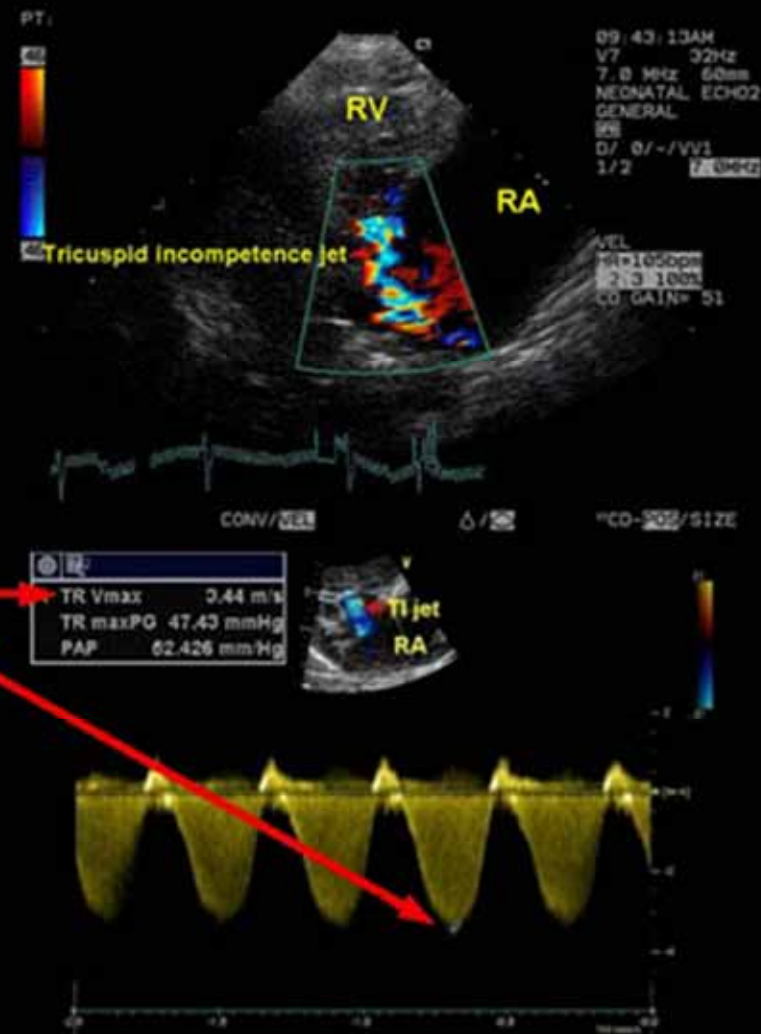
With each ventricular systole, a jet of blood leaks back from the right ventricle into the right atrium.

By measuring the velocity of that jet we can use the modified Bernoulli equation to derive the systolic pressure gradient between the right ventricle and right atrium.

Because the pressure in the right atrium is low (5-10 mmHg), the right ventricular systolic pressure can be derived from the calculated pressure gradient + a fudge factor for the RA pressure (usually 5 or 10). In a normal heart, right ventricular systolic pressure will be the same as pulmonary artery systolic pressure.

For example, if we measure the TI jet velocity as 3.44 m/sec. The pressure gradient will be  $4 \times 11.8 = 47.4$  mmHg. Adding a fudge factor of 5 for the RA pressure gives a pulmonary artery systolic pressure of 52.4 mmHg.

Continue



# Management

- ▶ General measures
  - IPPV
  - Sedation +/- paralysis
  - Surfactant
  - Correction of hypothermia, acidosis
  - Inotropes



# Management

- ▶ High Frequency Oscillatory Ventilation (HFOV)

But no clear benefit over CV

- ▶ Inhaled Nitric Oxide (INO)

Gold-standard but toxic by-products; not readily available

- ▶ Prostaglandins

Disadvantage: systemic hypotension

Advantage: Ductal patency




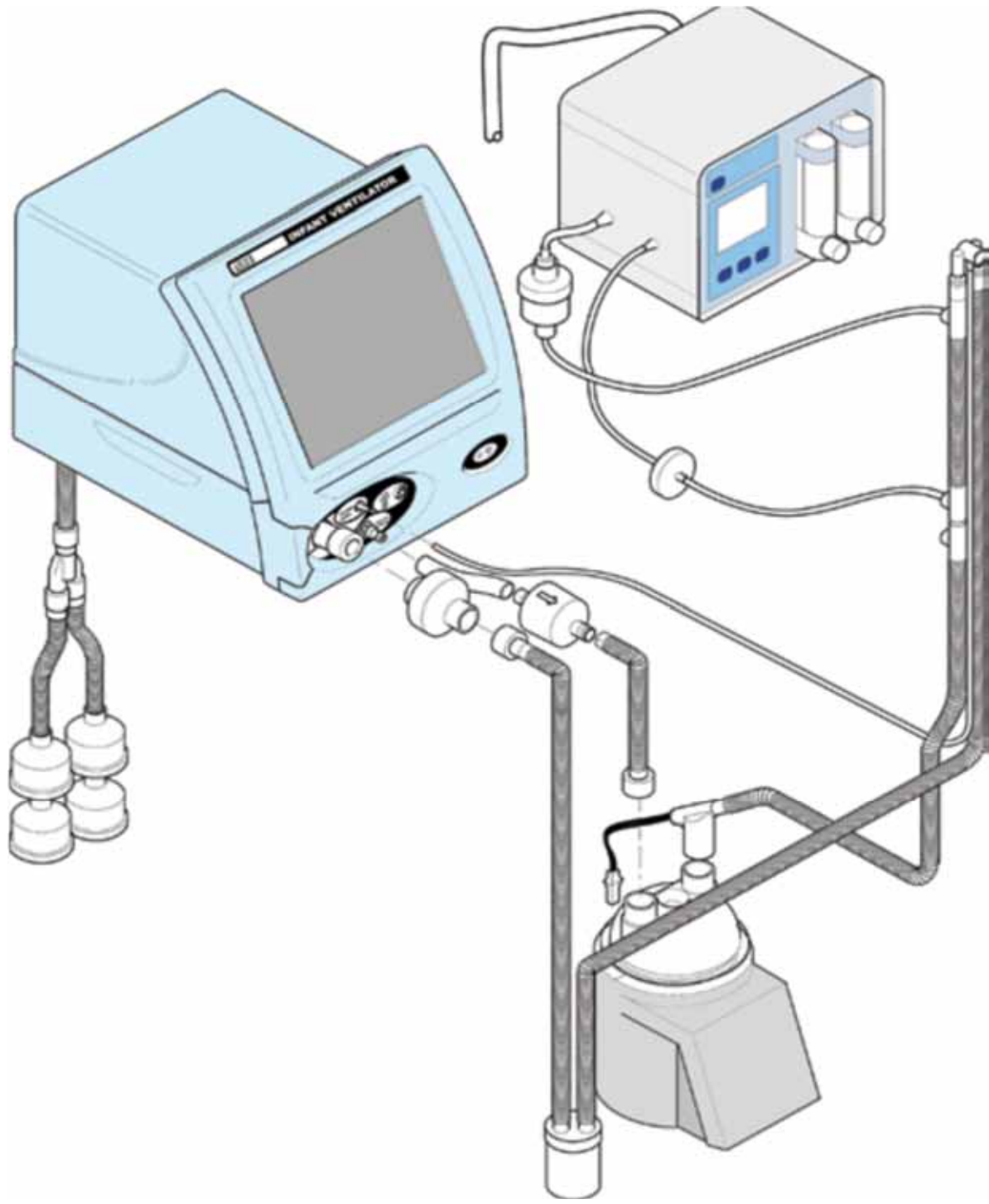
# Management

- ▶ Extracorporeal Membrane Oxygenation (ECMO)
- ▶ Magnesium Sulphate
- ▶ Phosphodiesterase Inhibitors
  - Milrinone
  - Sildenafil

Safe; useful in developing countries where INO  
NA

Dose: start at 0.25–0.5mg/kg/dose. Max  
2mg/kg/dose – 6–8 hrly.







# Sildenafil

- ▶ Used to treat erectile dysfunction & PAH
- ▶ Inhibits PDE-5:  $\uparrow$ cGMP  $\rightarrow$  relaxes arterial wall smooth muscle of penis and lungs
- ▶ Originally developed by British scientists
- ▶ Brought to the market by Pfizer
- ▶ 1<sup>st</sup> became available in 1998: erectile dysfunction
- ▶ Approved for Rx PAH in 2005





"Sildenafil, please... No! It's not for THAT!"