PERSISTENT PULMONARY HYPERTENSION OF NEWBORN

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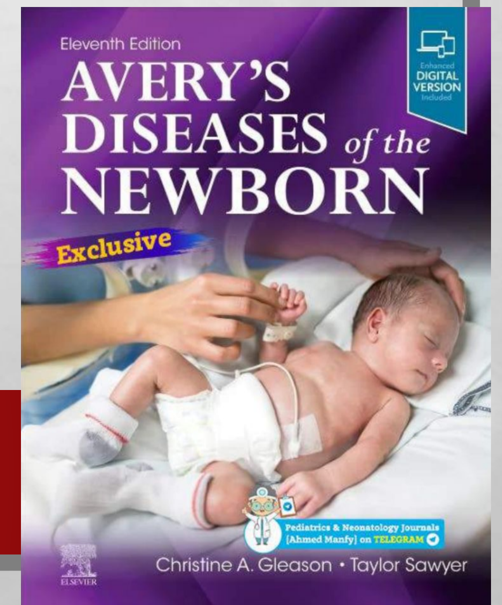
PPHN
PERSISTENT PULMONARY HYPERTENSION OF NEWBORN

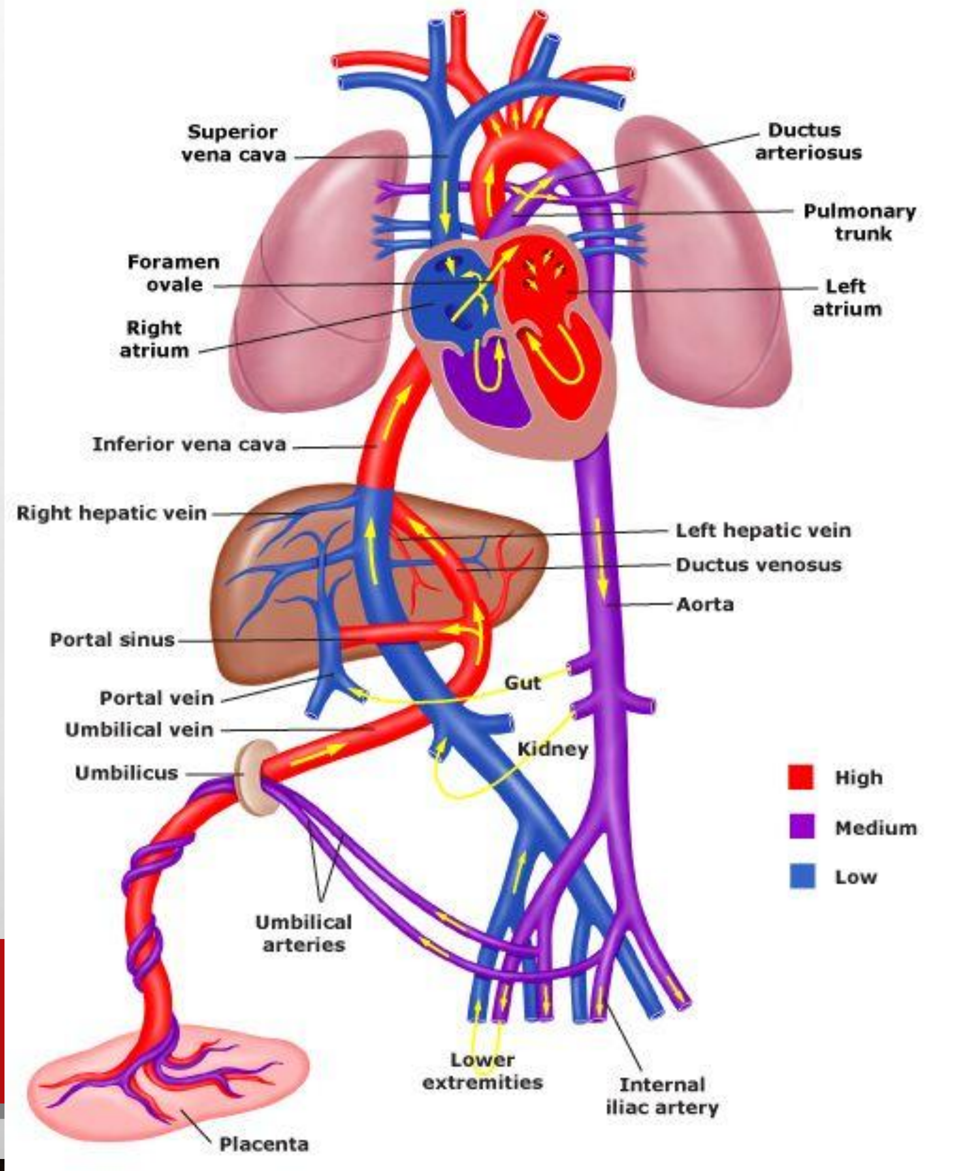
- PPHN IS DEFINED AS THE FAILURE TO ACHIEVE OR SUSTAIN THE NORMAL DECREASE IN PULMONARY VASCULAR RESISTANCE (PVR) AT BIRTH

- AN INCIDENCE RANGING FROM
  - 0.4 TO 6.8 PER 1000 LIVE BIRTHS
  - 5.4 PER 1000 LIVE BIRTHS IN LATE PRETERM INFANTS

- MORTALITY OF ALL NEWBORNS WITH PPHN HAS BEEN REPORTED AT 7.6%



FETAL CIRCULATION



PPHN
P.1/5

What is PPHN?

Right hand **Pre-ductal SpO_2**

Relative TR
(TRPG > 25)

TRPG = RVSP-RAP
Tricuspid Regurgitation Peak Gradient

D-shaped RV

Post-ductal SpO_2 Right foot

Elevated PVR

- **Maladaptation** (Lung parenchymal disease)
  - Meconium aspiration syndrome
  - Respiratory distress syndrome
  - Pneumonia
  - Malignant TTN
- **Maldevelopment**
  - NSAIDs / SSRIs
  - In-utero hypoxia / acidosis
- **Underdevelopment**
  - Congenital diaphragmatic hernia
  - Lung hypoplasia
  - Oligohydramnios syndrome
  - Intrinsic obstruction: polycythemia

Respiratory distress, labile hypoxemic newborn

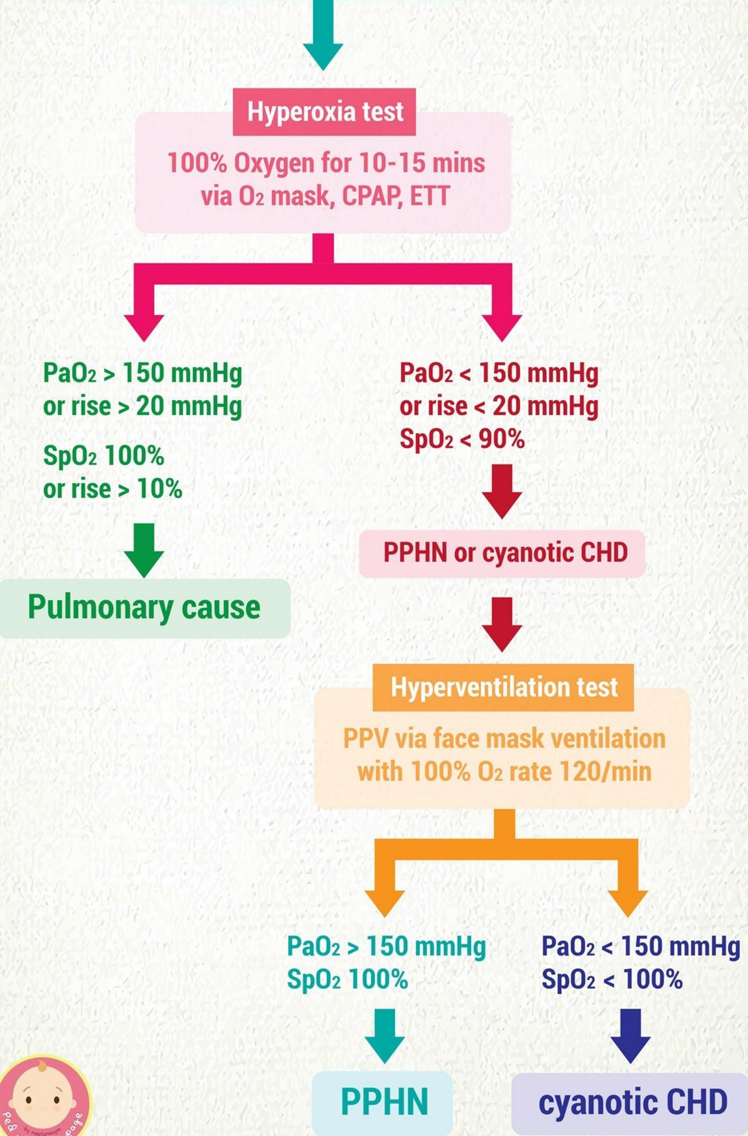
Differential cyanosis -- Different SpO_2 > 5-10% or PaO_2 10-20 mmHg

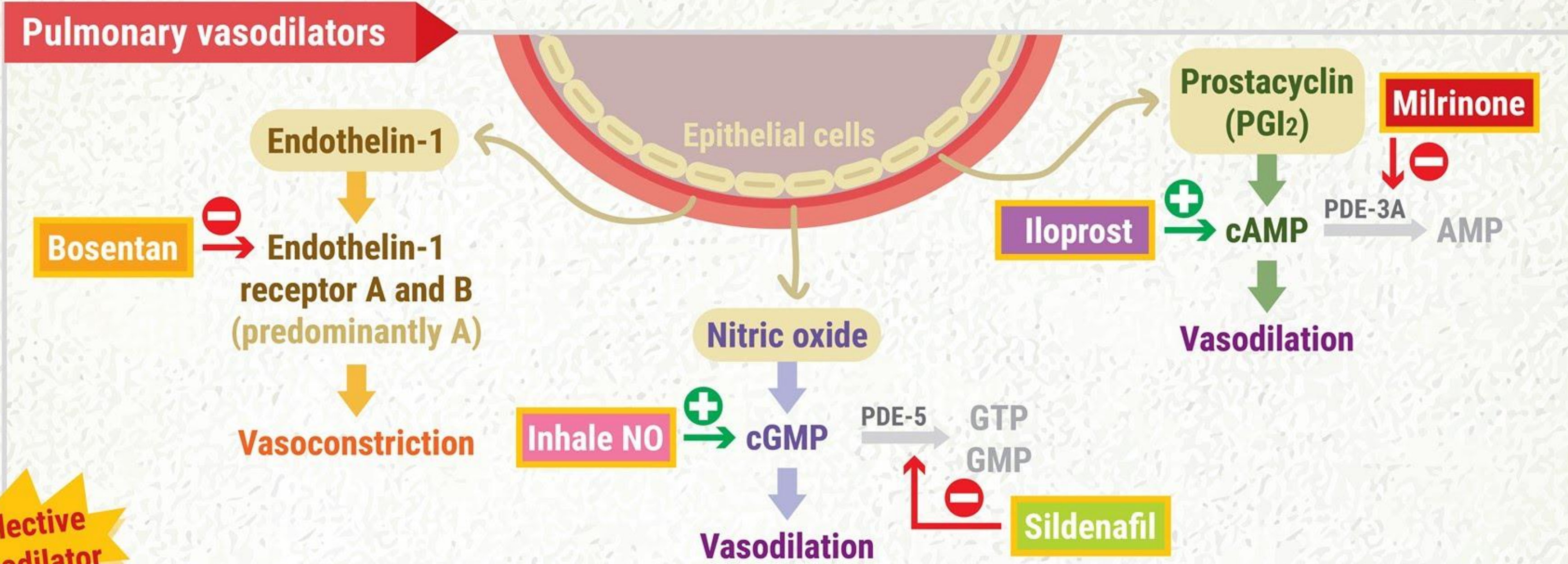
TABLE 47.1 Etiology of Persistent Pulmonary Hypertension of the Newborn by System

Pulmonary	Genetic/Rare Lethal Lung Developmental Disorders
• Meconium aspiration syndrome • Respiratory distress syndrome • Pulmonary hypoplasia (oligohydramnios) • Congenital diaphragmatic hernia • Pneumonia/sepsis • Idiopathic • Pulmonary interstitial glycogenosis • Congenital pulmonary lymphangiectasia	• Congenital surfactant deficiencies (SP-B/C, ABCA3) • TTF-1/Nkx 2.1 • FOXF1 mutation (ACD) • Mutation of CRH receptor-1 • TBX-4 mutation • Inborn error of metabolism • Trisomy 21
Cardiovascular	**Other**

• Myocardial dysfunction
• Structural cardiac disease
    • Mitral stenosis
    • Pompe disease
    • Aortic atresia
    • Coarctation of the aorta
    • Interrupted aortic arch
    • Transposition of great vessels
    • Ebstein anomaly
• Hepatic arteriovenous malformations (AVM)
• Cerebral AVMs
• Total anomalous pulmonary venous return
	• Neuromuscular disease
	• Polycythemia
	• Thrombocytopenia
	• Maternal drug use or smoking

PPHN, Persistent pulmonary hypertension of the newborn.


PPHN
P.2/5
Cyanotic newborn
Hyperoxia test
100% Oxygen for 10-15 mins
via O2 mask, CPAP, ETT
PaO2 > 150 mmHg
or rise > 20 mmHg
SpO2 100%
or rise > 10%
Pulmonary cause
PaO2 < 150 mmHg
or rise < 20 mmHg
SpO2 < 90%
PPHN or cyanotic CHD
Hyperventilation test
PPV via face mask ventilation
with 100% O2 rate 120/min
PaO2 > 150 mmHg
SpO2 100%
PPHN
PaO2 < 150 mmHg
SpO2 < 100%
cyanotic CHD



Inhaled NO	Sildenafil (Viagra)	Iloprost	Milrinone
Start > "Rule of 20" : OI 20 Dose 20 ppm ↑ PaO_2 20 mmHg Wean off > "Rule of 60" : PaO2 60 mmHg or SpO2 > 90% for more than 60 mins	PDE-5 inhibitor IV load: 0.42 mg/kg x 3 hours then: 1.6 mg/kg/day (0.07 mg/kg/hr) PO 1-2 mg/kg/dose q 6 hr	Synthetic PGI_2 NB 1-2.5 mcg/kg q 2-4 hr IV 0.5-3 ng/kg/min (max 10 ng/kg/min)	Increase cAMP in vascular smooth m. & cardiac m. IV load: 50 mcg/kg in 30-60 mins then: 0.33 mcg/kg/min (max 1 mcg/kg/min)

Systemic vasodilator = HYPOTENSION

General management in PPHN

Hemodynamic status

Optimize cardiac output & LV function
Inotropic drugs as needed
If hypotension* > resuscitate with;
NSS bolus 10 ml/kg
Dopamine > NE
Hydrocortisone
(consider adrenal shock)

Indication for ECMO

1. Persistent hypoxemia
2. OI > 40
3. A-a gradient > 600

* despite aggressive medical treatment

Indication for HFOV

pCO_2 persist > 60 mmHg
despite PIP > 25 and TV > 6 ml/kg

Respiratory management

Optimize lung expansion
- Small tidal volume, low PIP
- Optimal PEEP
- Keep permissive hypercapnia: pH 7.30-7.40, pCO_2 < 60

Avoid hyperoxia: lowest FiO_2 for maintain SpO_2 90-95%, PaO_2 55-80 mmHg
Surfactant: RDS, MAS, pneumonia

Sedation
Fentanyl, Midazolam

Minimal stimulation
Eye covers, ear muffs

Correct metabolic status
- Electrolytes
- Avoid acidosis
- Hypo/hyperglycemia
- Temperature control
- Optimal Hct 35-40%

REVIEW CASE

เฝ้าระวังความเสี่ยง

ก่อนคลอด

ขณะคลอด

หลังคลอด

ก่อนคลอด

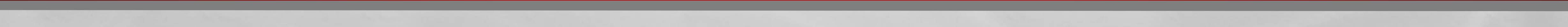
- ประวัติช่วง **ANC >> CASE HIGH RISK**
- โรคของมารดา **OVERT DM >> SURFACTANT DEFICIENCY**
- ยาที่มารดาใช้ระหว่างตั้งครรภ์ **>> NSAIDS , SSRI**
- **OLIGOHYDRAMNIOS , ANHYDRAMNIOS, LUNG HYPOPLASIA**
- **PRENATAL DX > CONGENITAL DIAPHRAGMATIC HERNIA**

ระยะคลอด

- PRETERM >> RDS
- MECONIUM STAINED AF >> MAS
- FETAL DISTRESS (IN UTERO HYPOXIA/ACIDOSIS)

หลังคลอด

- **RD** หลังคลอด จากภาวะต่างๆ
  - >> RDS /MAS/ TTNB/ PNEUMONIA
- SEPSIS
- POLYCYTHEMIA



- ประวัติสำคัญ
- **CLOSE MONITORING** >อุปกรณ์ต้องเพียงพอ
- **APPROPIATE TREATMENT**
- ทีมงานเตือนกันได้