Congenital Heart Diseases and the Lung

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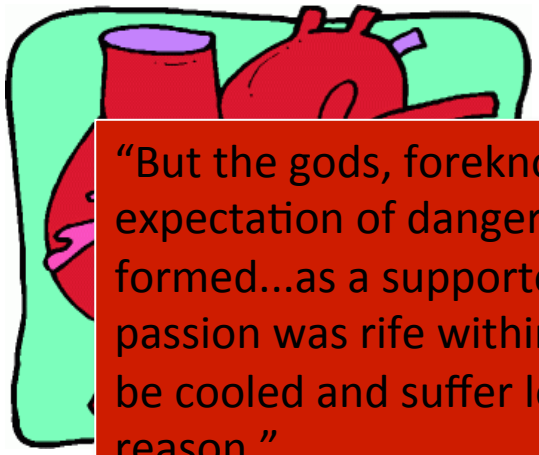
Learning Objectives

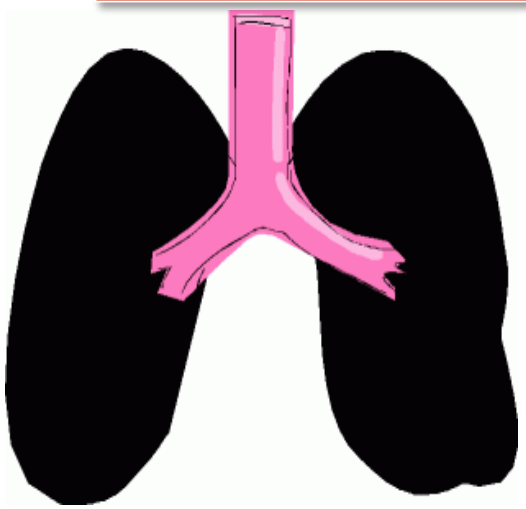
Upon completion of this activity, participants should be able to:

- Describe the impact of congenital heart disease on pulmonary function
- Recognize and treat abnormal congenital heart disease-associate pulmonary dysfunction encountered in the neonatal period
- Discuss advances in neonatal care of patients with congenital heart disease-associated lung pathology

The Heart and The Lungs



"But the gods, foreknowing that the palpitation of the heart in the expectation of danger and the swelling...of passion was caused by fire, formed...as a supporter to the heart the lung...as a soft spring, that, when passion was rife within, the heart, beating against a yielding body, might be cooled and suffer less, and might...join with passion in the service of reason."
—Timaeus, Plato



Congenital heart disease and the lung

- CHD is common (7-9 infants out of 1000 births)
- CHD profoundly influences airway, lung and thoracic anatomy
- CHD profoundly influences airway, lung and thoracic function

From the neonatologist's viewpoint…

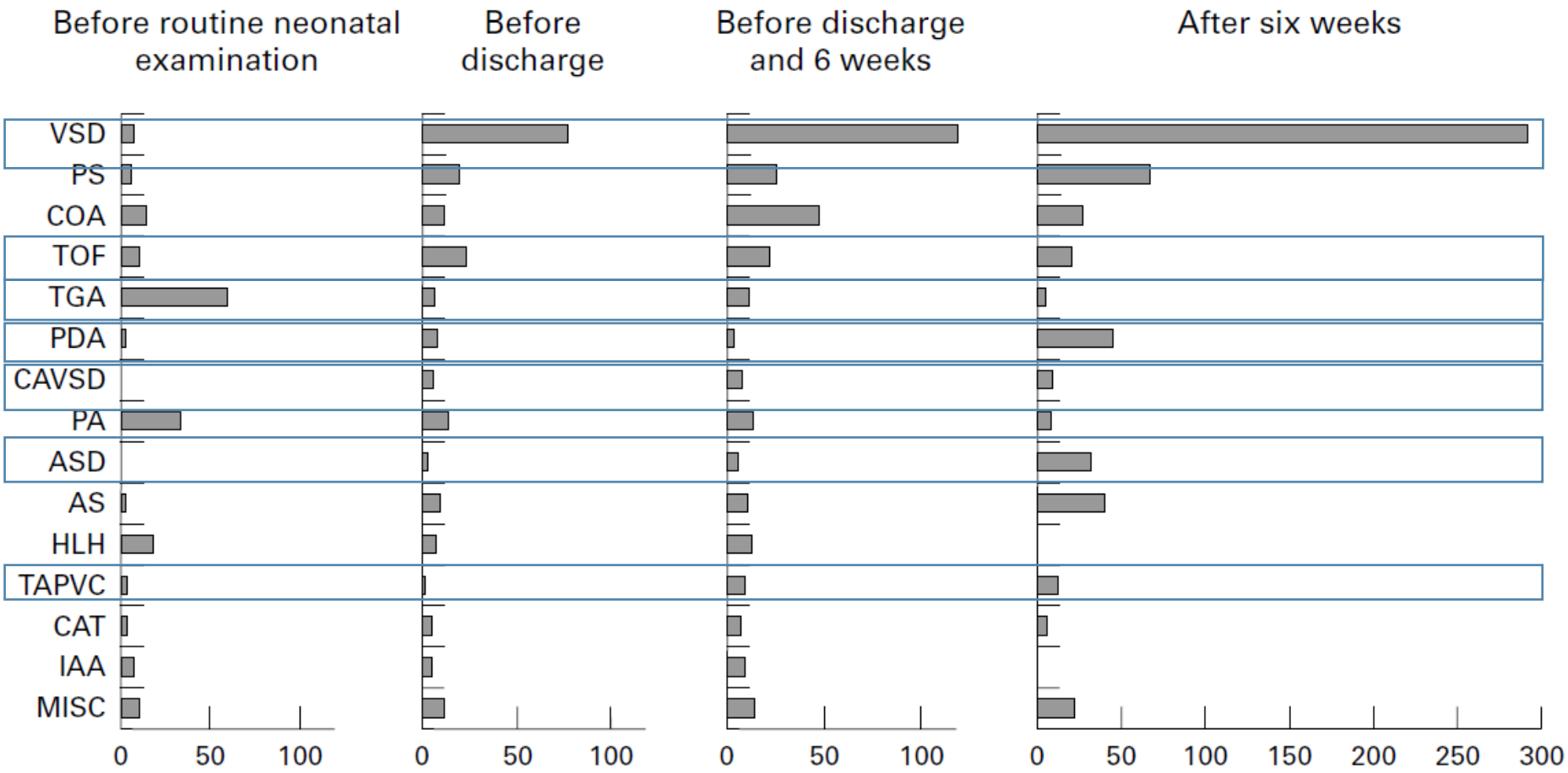
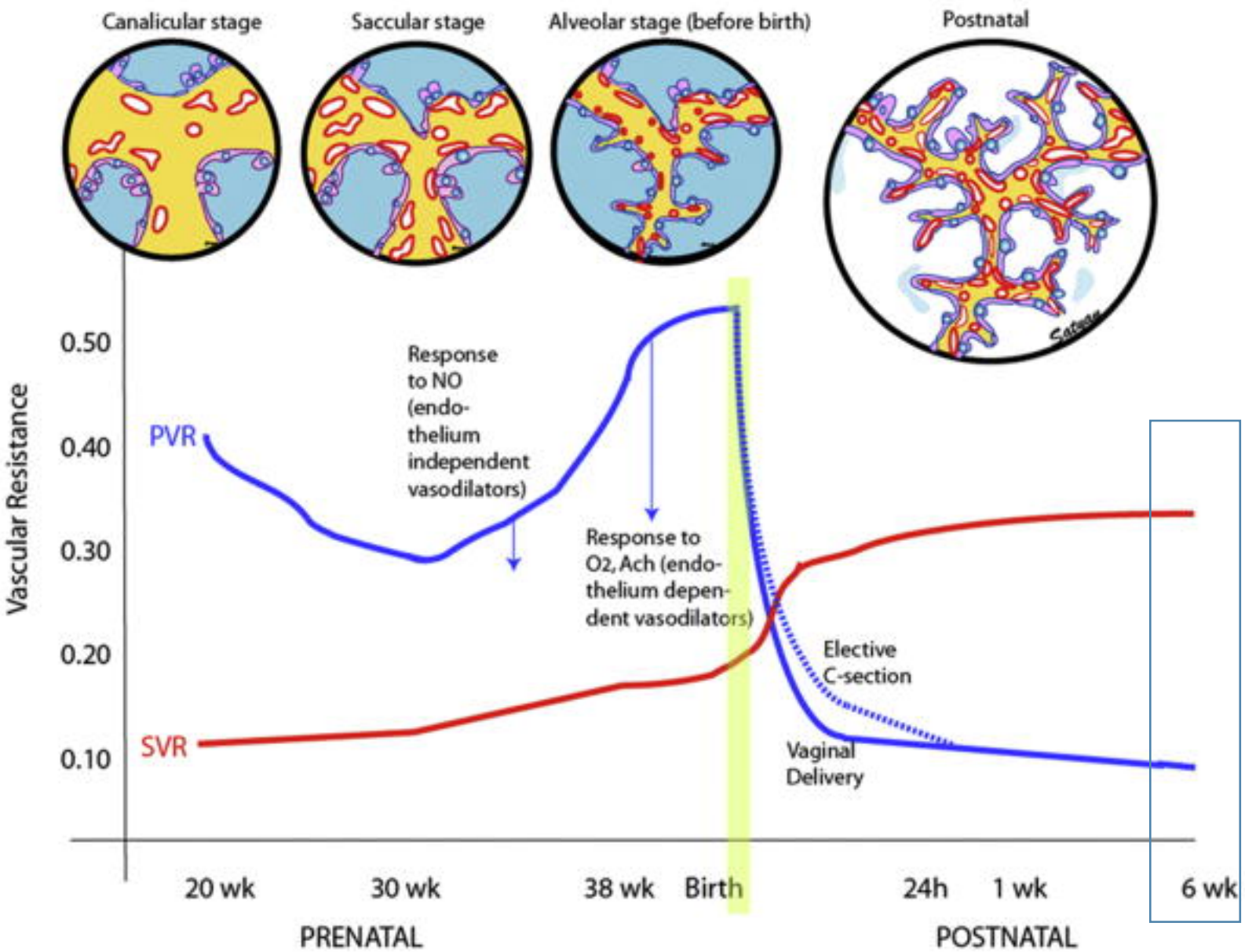


Figure 3 Timing of diagnosis and numbers of cases for each diagnostic group in 1277 babies eligible for routine examination. Displayed in this way, the spectrum of heart disease recognised at each stage is clearly shown.

Wren et al (1999) *Arch Dis Child Fetal Neonatal Ed*



Lakshminrusimha (2012) *Clin Perinatol*

From LUNGS' viewpoint…

CHD be associated with:

- Airway Problems
  - Cardiac compression
  - Vascular compression
  - Abnormal anatomy
  - Abnormal reactivity
- Abnormal Lung Anatomy
  - Orientation
  - Segmentation
  - Parenchyma
  - Ciliation
- Δ Pulmonary Blood Flow
  - ↑PBF
  - ↓PBF
- Δ Pulmonary Vasculature
  - Arterial disease
  - Venous disease
  - Abnormal lymphatic drainage
- Dysregulation of Ventilation
- Infections and Bleeding
- Postoperative Problems`

Often, several pathophysiological paradigms coexist

Airway and lung compression by the heart

Effects of L →R shunt

- Pulmonary
  - Increased total PBF
  - Normal or decreased effective PBF
  - Edema
- Cardiac
  - LA enlargement
  - LV diastolic dysfunction (pulmonary venous hypertension)
  - LA hypertension (pulmonary venous hypertension)
  - Cardiomegaly (direct parenchymal compression)
- Airway
  - Bronchial compression (by heart and vessels)
  - Atelectasis (especially LLL)

VSD

Arterial Trunk

Tetralogy of Fallot and other RVOTO lesions
Effects of congenital abnormal pulmonary valve

- Abnormal pulmonary vessels (most common in ToF APV)
  - Abnormal lung lobulation
  - **Abnormal – OBSTRUCTED – airways**
    - **May need rescue maneuvers to ventilate EVEN WHEN INTUBATED**

NB1: Valve (or RVOT) always stenotic BUT ALSO often regurgitant

NB2: If you cannot ventilate infant with ToF APV – PUT HIM ON HIS BELLY!

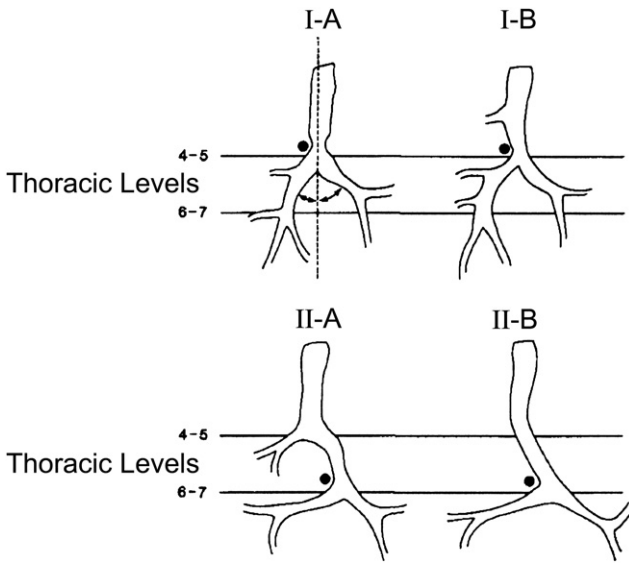
Abnormal, engorged pulmonary vessels move off airway due to gravity

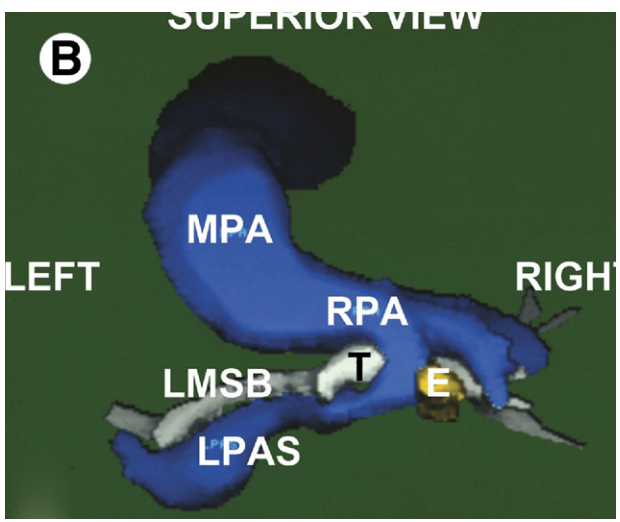
Bronchoscopy in children with congenital heart disease and respiratory symptoms
Extremely valuable

- 50-92% of children with CDH who underwent bronchoscopy due to respiratory symptoms had definitive airway anomalies
- **6-10% of children had upper and lower airway anomalies**

Chapotte (1998) *J Cardiothorac Vasc Anesth*
Lee (2002) *Pediatr Pulmon*

Aberrant left pulmonary artery





Type 1 – "simple"
- Right-sided tracheobronchomalacia
- Right lung hyperinflation (*vs.* CLE)
- No association with complete tracheal rings or CHD
- Easy repair with very low mortality

Type 2 – "not simple"
- Tracheal stenosis and complete tracheal rings – awful intubation
- Right lung hypoplasia
- Right pulmonary artery hypoplasia
- Pulmonary sequestrations
- VACTERAL spectrum disorders
- CHD: ToF, TAPVR (20-30% incidence)
- Difficult repair with 8-20% mortality

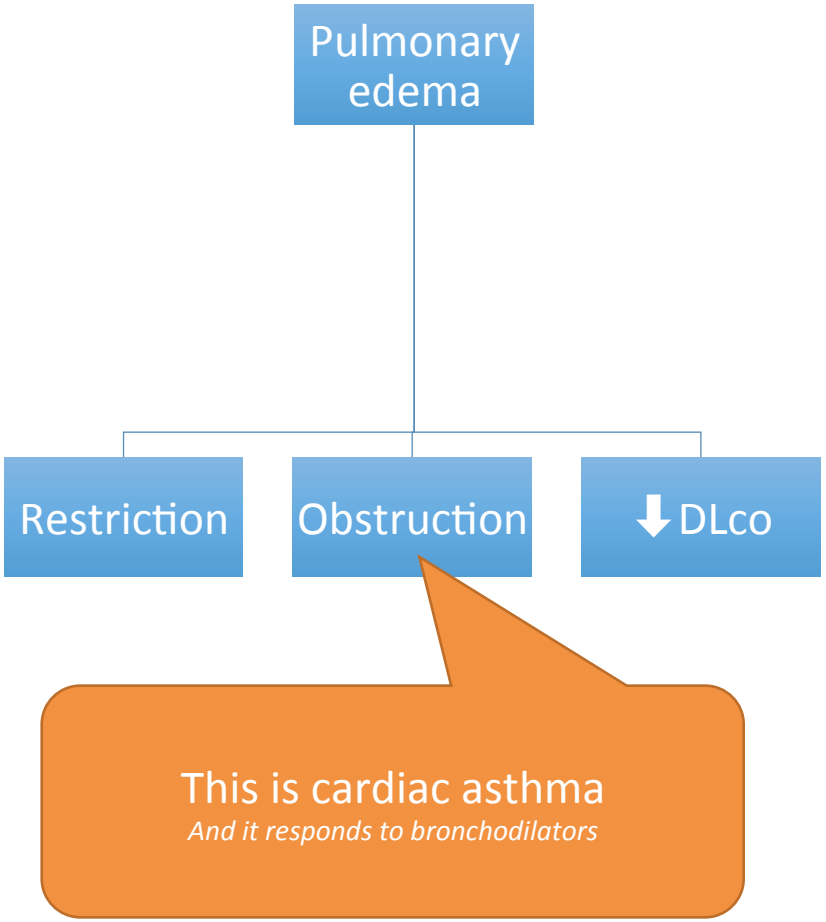
Newman, Cho (2010) *Semin Ultrasound CT MRI*
Hraska et al (2009) *Multimed Man Cardiothorac Surg*

Pulmonary edema
...more complex than it may seem

1. Pulmonary venous hypertension due to:
- Pulmonary veno-occlusive disease or pulmonary vein stenosis
- Cor triatriatum
- Supra mitral ring
- Left ventricular dysfunction
- Transposition of the great arteries or hypoplastic left heart with intact atrial septum

2. Decreased lymphatic flow:
- Lymphangiectasia
- Superior venacaval syndrome
- Single ventricle physiology
- Tricuspid valve stenosis
- Failing or stiff right ventricle
- Right ventricular outflow tract obstruction

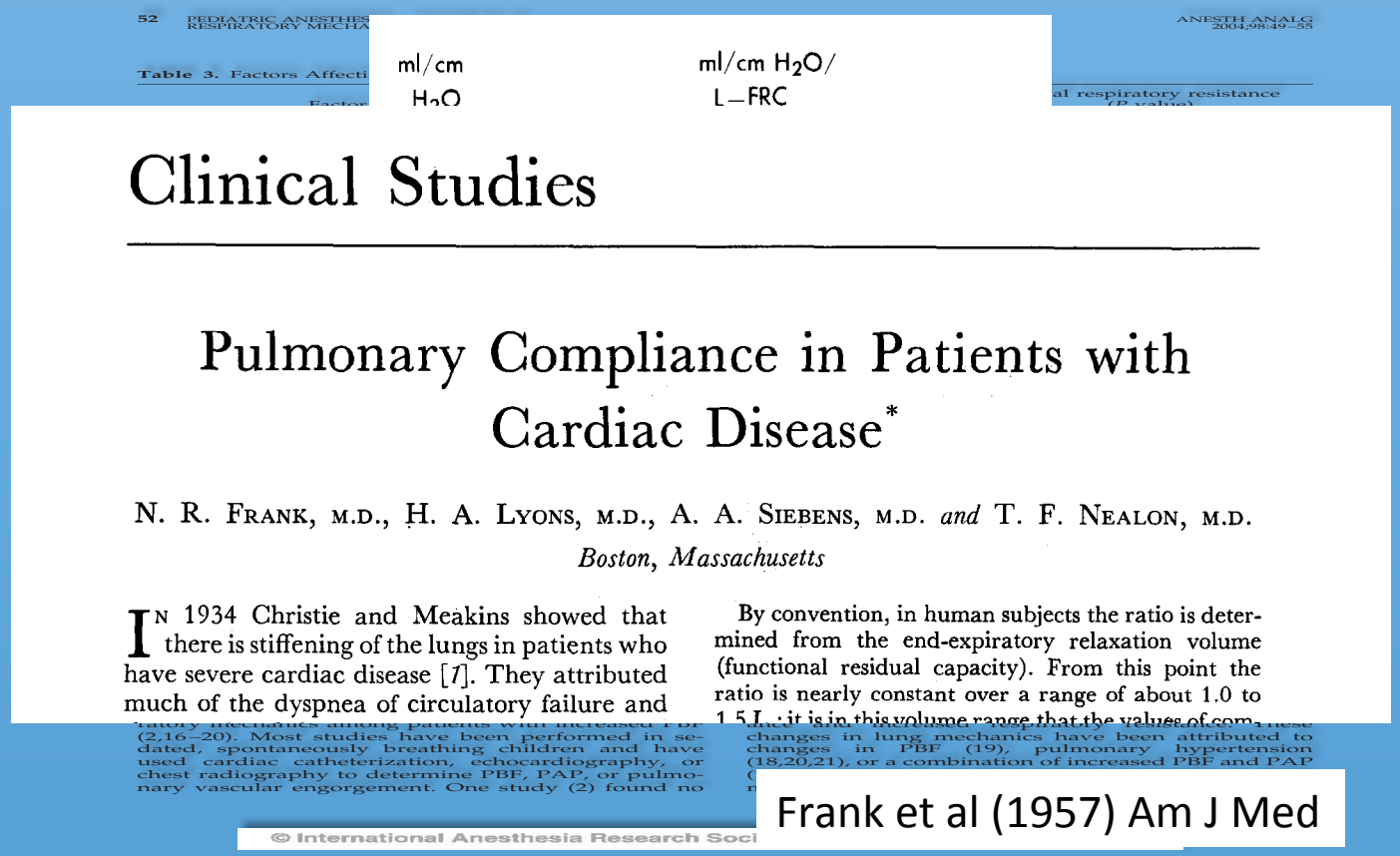
3. Left-to-right heart shunts including:
- VSD
- ASD
- PDA
- Partial anomalous pulmonary venous connection
- Systemic arteriovenous malformations
- Aorto-pulmonary connections including surgical shunts



Healy et al (2012) *Paed Resp Rev*

Effects of increased and decreased PBF on lung mechanics

…there is nothing new under the sun



ml/cm H_2O | ml/cm H_2O/L—FRC

Clinical Studies

Pulmonary Compliance in Patients with Cardiac Disease*

N. R. Frank, M.D., H. A. Lyons, M.D., A. A. Siebens, M.D. *and* T. F. Nealon, M.D.

Boston, Massachusetts

In 1934 Christie and Meakins showed that there is stiffening of the lungs in patients who have severe cardiac disease [*1*]. They attributed much of the dyspnea of circulatory failure and

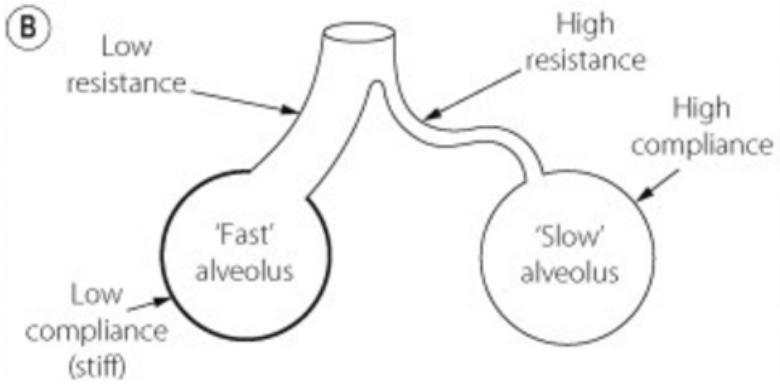
By convention, in human subjects the ratio is determined from the end-expiratory relaxation volume (functional residual capacity). From this point the ratio is nearly constant over a range of about 1.0 to

Frank et al (1957) Am J Med

Stayer et al (2004) Anaesth Analg

Effects of increased PBF on infant breathing patterns

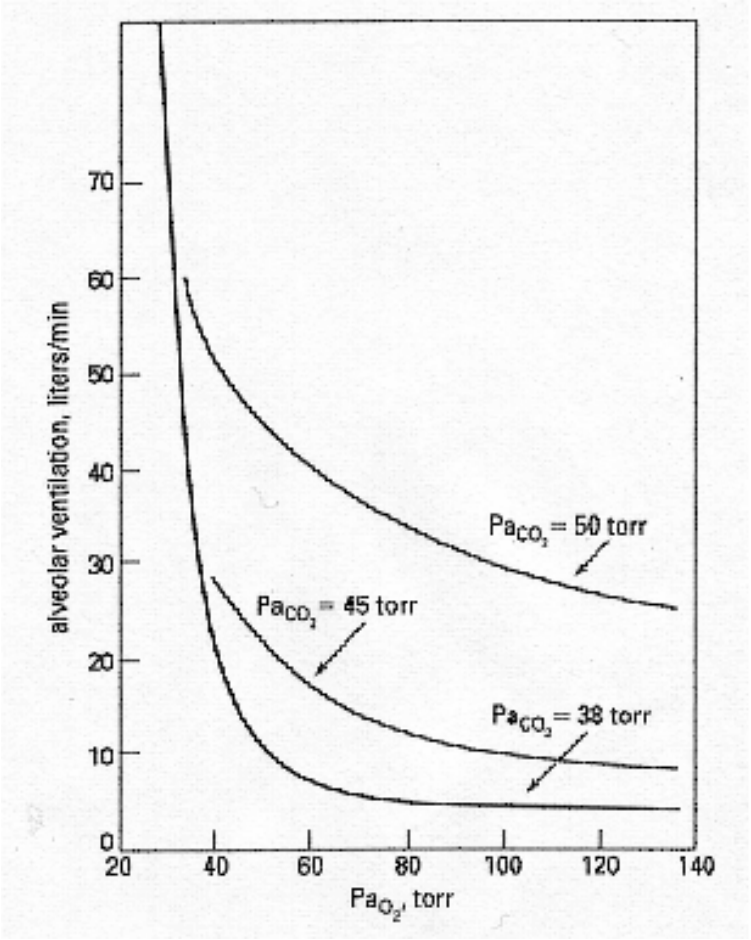
- Due to decreased compliance, these infants should favor <u>decreased VT and increased RR</u>
- Due to increased resistance, these infants should favor <u>exactly the opposite</u>
- Breathing in "overcirculating" infants <u>extremely inefficient</u>
  - and systemic cardiac output is abnormal
- Low threshold for intubation
- Careful attention to QP:QS



Their paradigm is not one of these

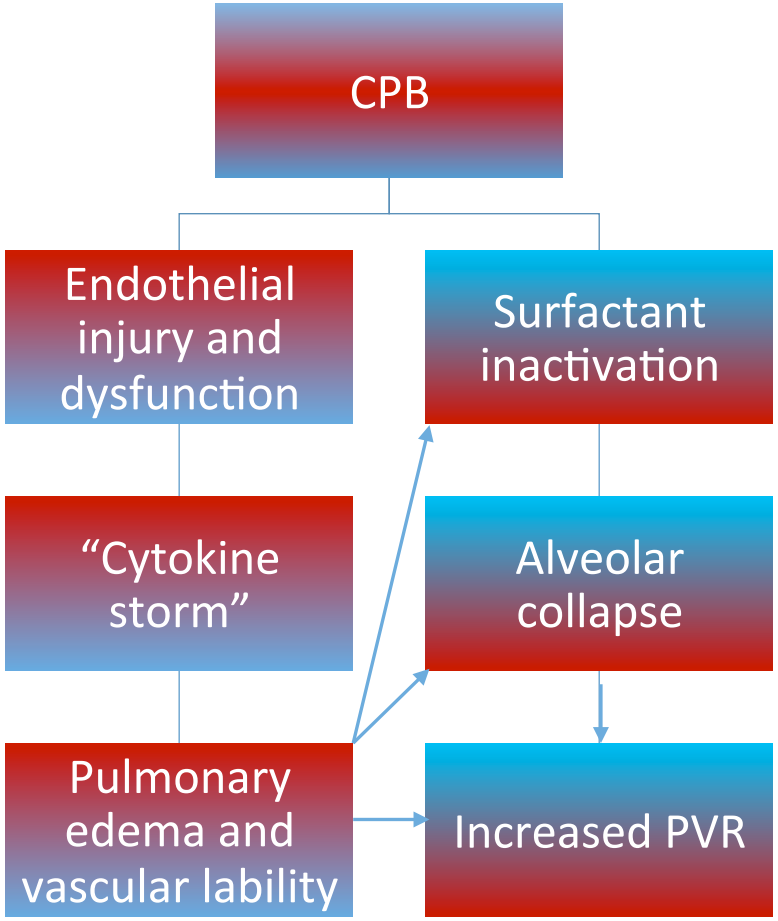
Abnormal ventilatory drive

- Edelman et al (1970) NEJM: Cyanotic patients have blunted ventilatory response to hypoxemia
- Blesa et al (1997) NEJM: This response normalizes soon after repair
- Georgiadou et al (2004) Heart: Periodic breathing in ADULTS after ToF repair is associated with RV failure



Postoperative problems
Pulmonary hypertension

- Common problem
- Affects evaluation and perioperative management
- Affected by type of CHD, CBP, etc
- Influences long-term morbidity and mortality



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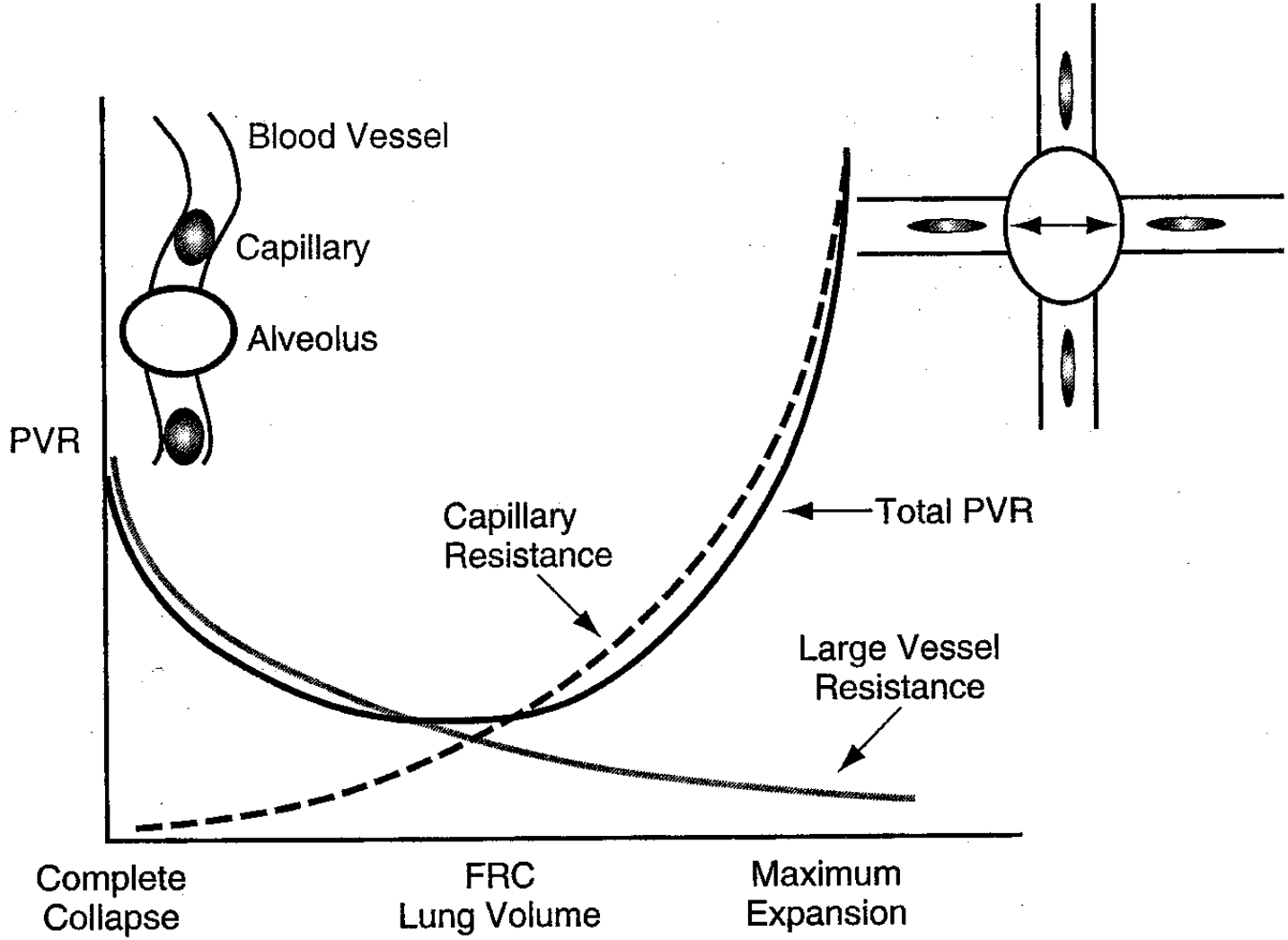


Whittenberger et al. (1960) *J Appl Phys*

Postoperative problems
Surgical trauma

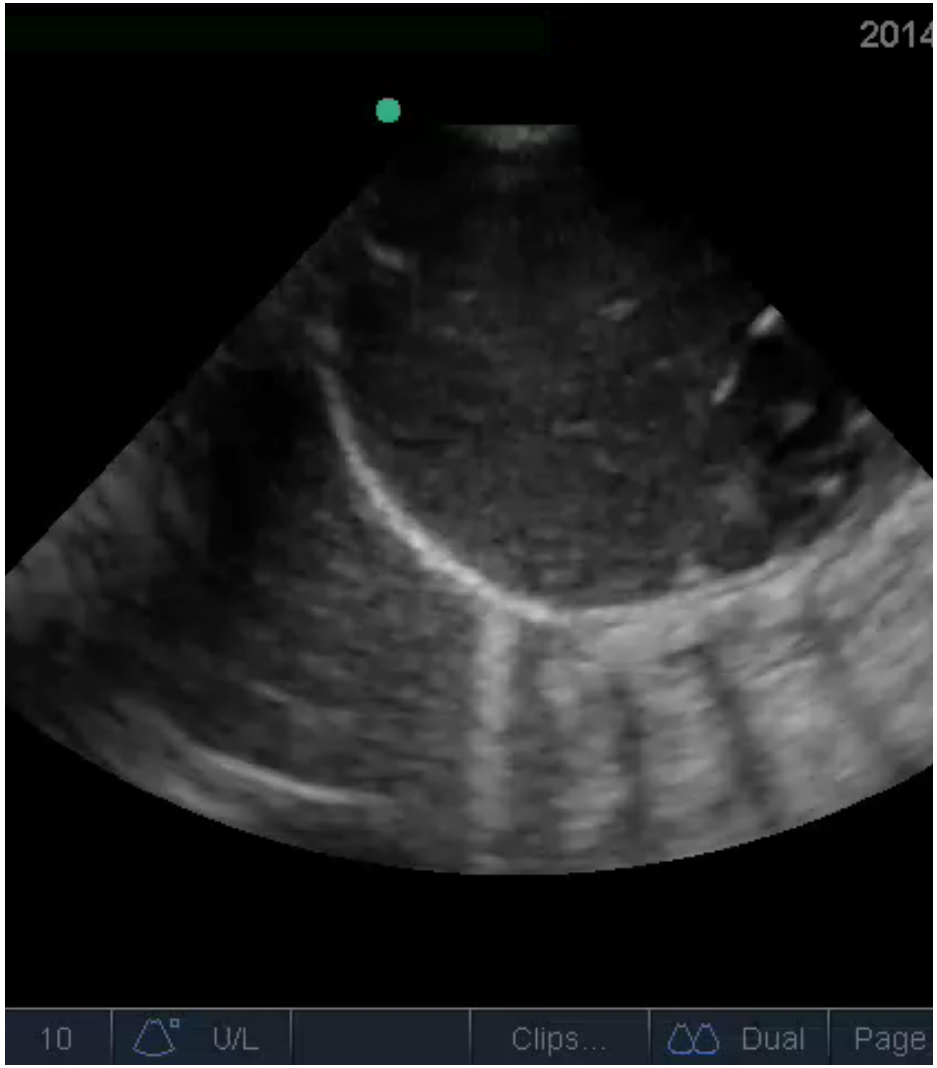
Surgical Trauma to the Respiratory System

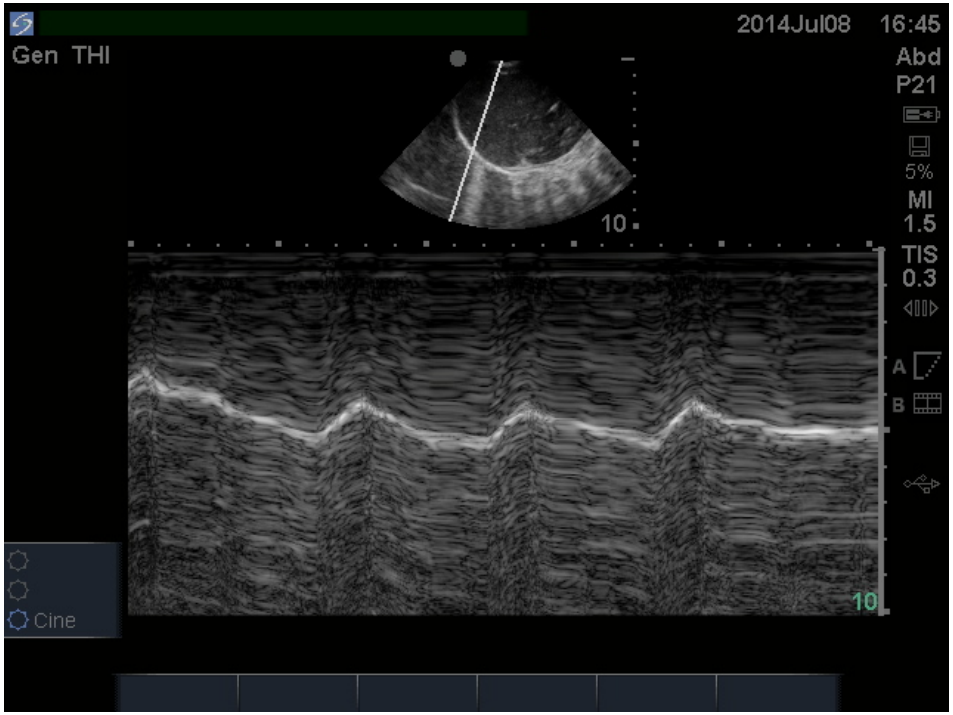
Pulmonary Complication	Aetiology
Chylothorax	Direct injury to the thoracic duct or smaller vessels High central venous pressures Central vein thrombosis
Recurrent laryngeal nerve injury	Surgery involving the ductus arteriosus, descending aorta or left pulmonary artery Manipulation of right common carotid artery or internal jugular vein for ECMO
Diaphragmatic paralysis	Direct trauma (section of the phrenic nerve) Stretching of the phrenic nerve Disruption of blood supply to the phrenic nerve
Subglottic stenosis	Direct compression by the endotracheal tube or cuff causing oedema and ischaemia

- CHD repair leading cause of chylothorax in tertiary hospitals
  - Incidence: 3.8%
- Incidence of phrenic injury and diaphragmatic paralysis up to 10%

Healy et al (2012) *Paed Resp Rev*

Postoperative problems
Diaphragm





US diagnosis of abnormal diaphragmatic motion by PCICU physicians as good as by fluoroscopy – and faster

Sanchez de Toledo et al (2010) *Cong Heart Dis*

Late lung function abnormalities in congenital lung disease

Lung Function Abnormalities in Congenital Heart Disease

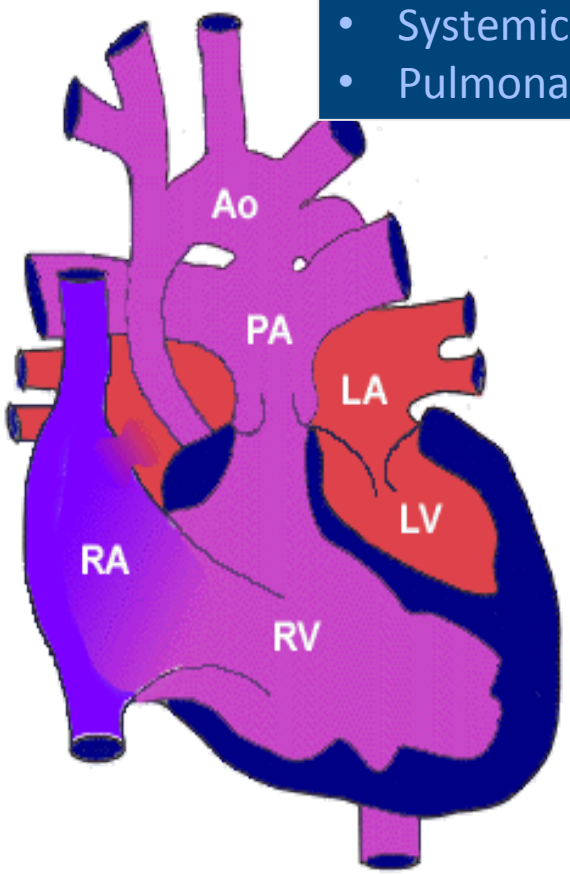
Lung function abnormality	Pathophysiology
Obstructive lung disease	Peribronchial cuffing
	Airway compression
	Vocal cord paralysis
	Infection (bronchiolitis)
Restrictive lung disease	Chest wall deformity
	Impaired lung growth
	Reduced lung compliance
	Diaphragmatic paralysis
Diffusion impairment	Reduced pulmonary vascular bed
	Atelectasis
	Pulmonary oedema

Healy et al (2012) *Paed Resp Rev*

The Single Ventricle and the Lung

Perioperative "single ventricle physiology"
Balancing the circulations

Parallel circulations:
- Systemic (coronary)
- Pulmonary



Control of pulmonary blood flow
- Conduit
- Drainage
- pH
- pCO_2
- pO_2
- SVR (MAP)
- MAWP

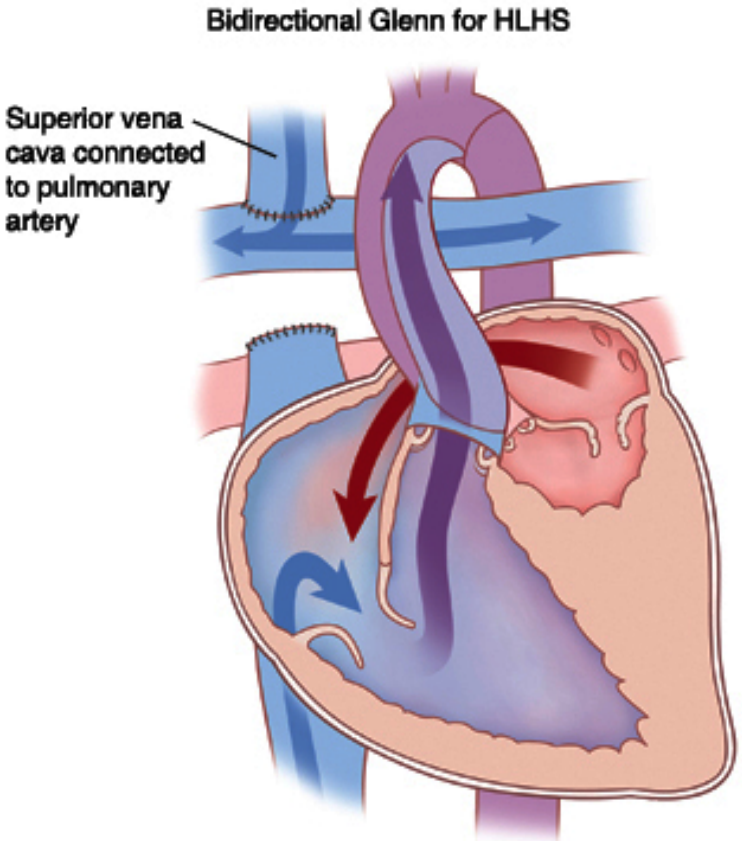
Conduits:
- Arterial duct
- Blalock-Taussig shunt
- Sano (RV-PA) shunt

Postoperative single ventricle – Glenn

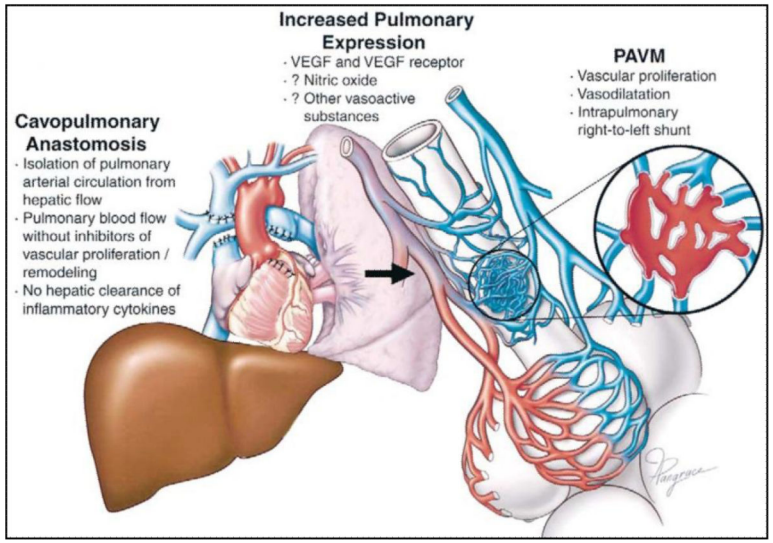
The <u>unusual</u> coupling of ventilation and oxygenation

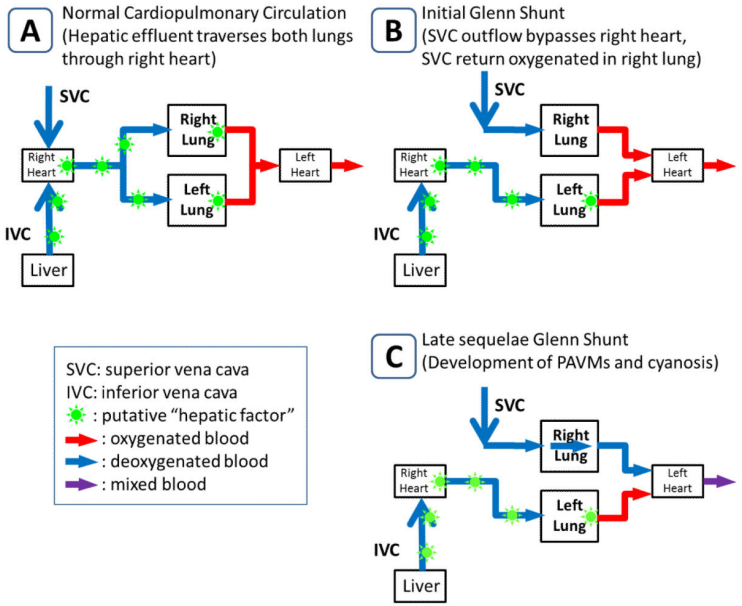
Control of pulmonary blood flow

- Conduit
- pH
- pCO_2
- pO_2
- MAWP



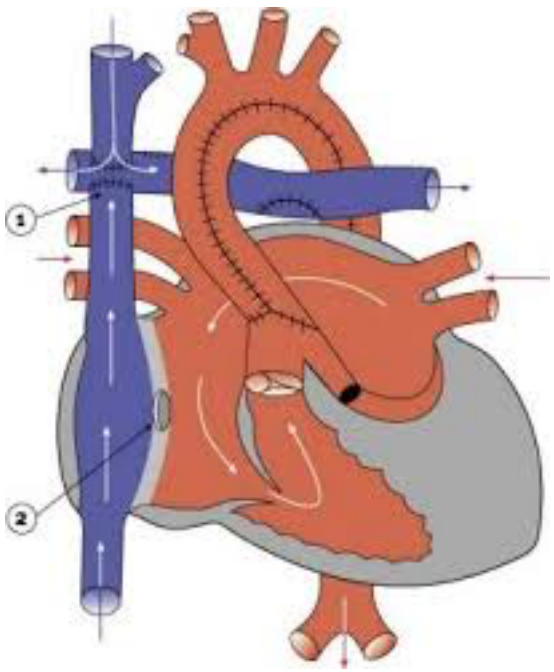
Postoperative single ventricle – Glenn
Roles of "hepatic factor" and pulsatility





Kavarana et al (2015) *Expert Rev Cardiovasc Therap*

Postoperative single ventricle – Fontan
Slow, non-pulsatile, vein-to-artery flow



- Lung hypoplasia – etiology?
- Vascular anomalies
  - AVMs (non-pulsatile, hepatic factor)
  - PE (slow, PLE)
- Pulmonary edema (collaterals, RV systolic, diastolic, or combined failure)
- Insufficient PBF
- Plastic bronchitis
  - Type 1
  - Type 2

Summary

- Heart and lungs are interdependent in structure and function – and congenital heart diseases emphasize that interdependence
- Congenital heart diseases influence the lung in a predictable way
- NB for the neonatologist:
  - Airway anomalies in aberrant LPA
  - Airway anomalies and the value of prone ventilation in ToF APV
  - Effects of cardiopulmonary bypass on pulmonary vasculature and lung mechanics
  - Management of PH
  - Management of PBF in single-ventricle circulations