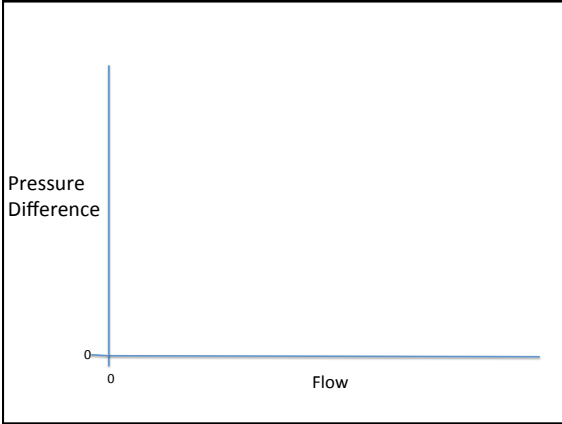
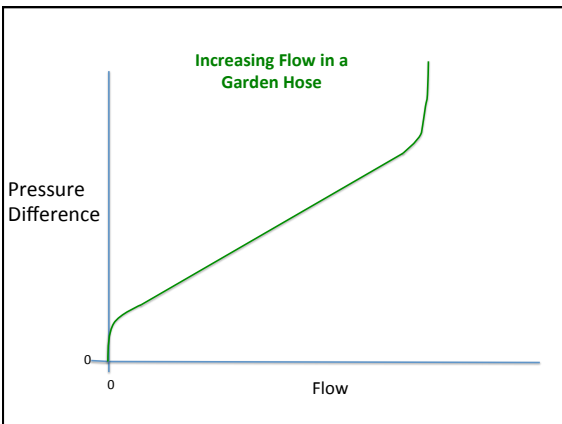
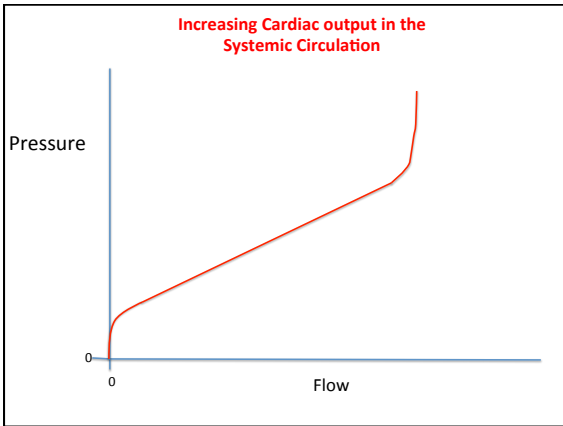


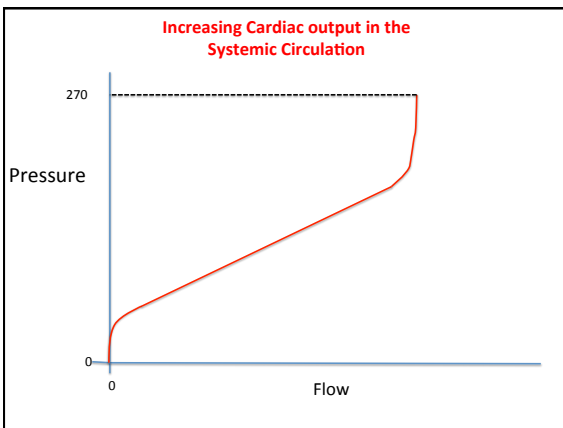
Pulmonary Hypertension: Not (by any stretch of the imagination) Just a Pediatric Problem

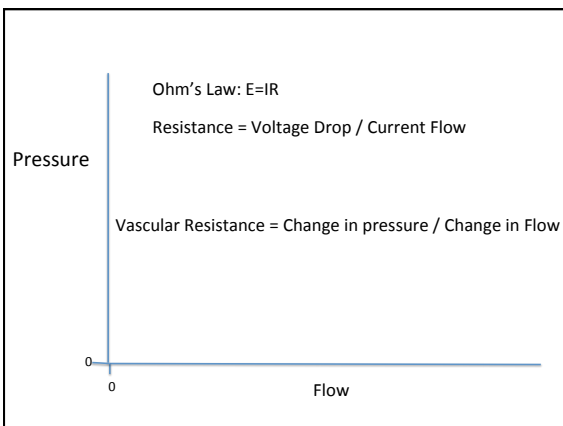
William Clarke, MD, MSc, FAAP
Associate Professor of Pediatrics and Anesthesiology
Medical College of Wisconsin

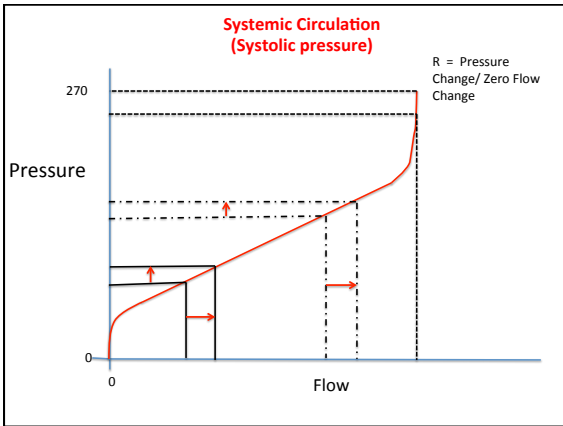


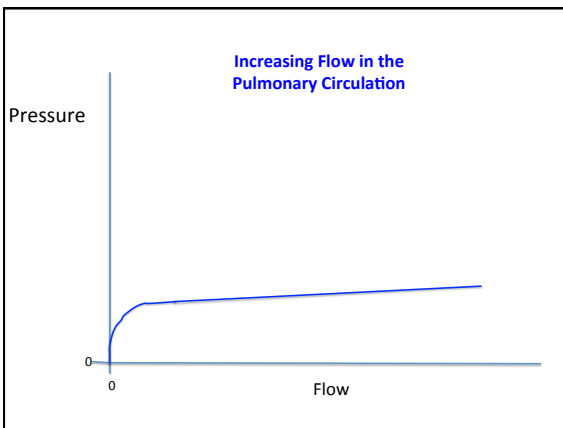


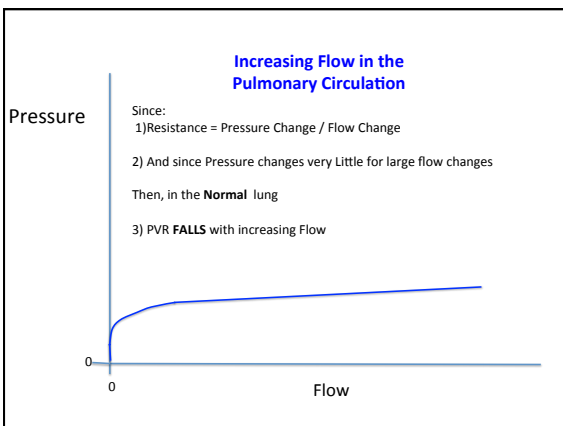


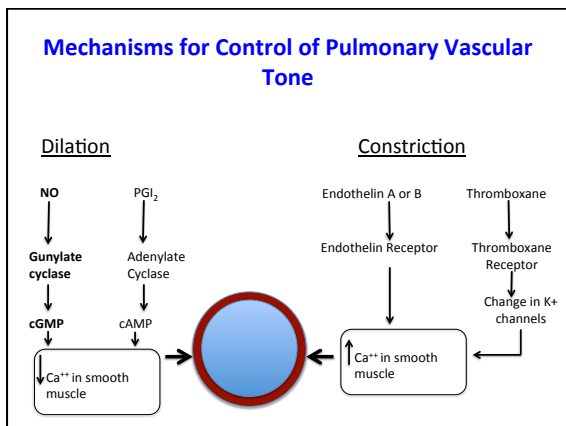






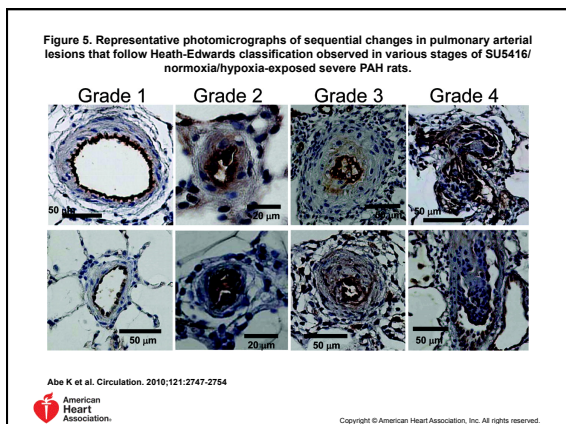


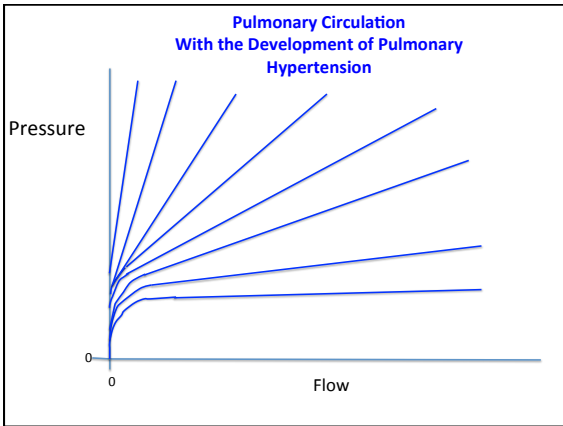


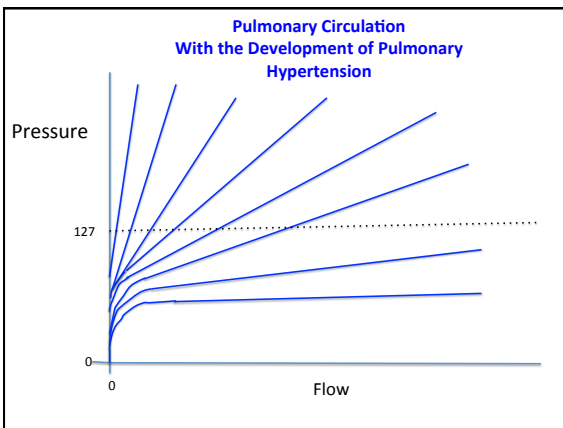


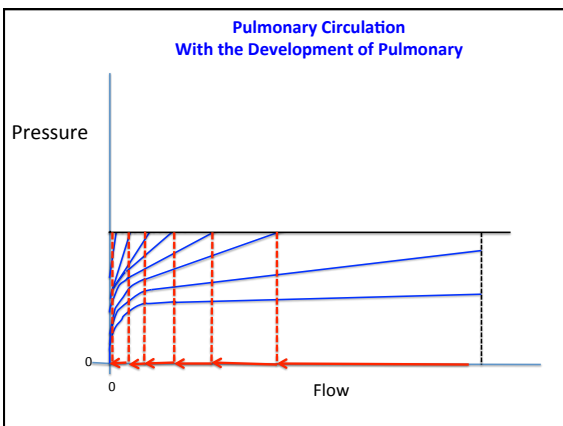
The Pathology of Pulmonary Hypertension

- PH is categorized as "Primary" -- which means we don't know what caused it --and "Secondary" -- which means we do know what caused it.....
- Irrespective of etiology most PH:
 - starts as vaso-spastic where the endothelium emit agents which constrict the vascular smooth muscle and then
 - progresses into vaso-occlusive where the endothelium changes phenotype, and begins to grow and then occludes the vessel
- Thrombotic PH is the most feared as it leads to relatively rapid occlusion of the vessels and rapid RV load





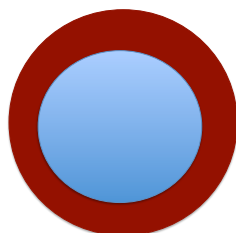




The Key Points in Understanding the Pathophysiology of PH

- 1) People with moderate to advanced PH show symptoms of low cardiac output and eventually die of insufficient cardiac output
- 2) The pulmonary artery pressure tells you next to *nothing* about the state of a patient with moderate to advanced PH

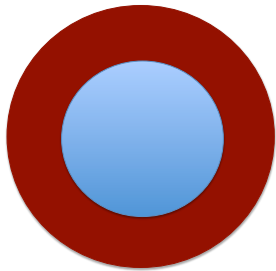
The Shape of Ventricles Means they Handle pressure Loads Very Differently: Circular LV Distributes Wall Stress



The Shape of Ventricles Means they Handle pressure Loads Very Differently: Asymmetric RV leads to different stress in free wall vs septum



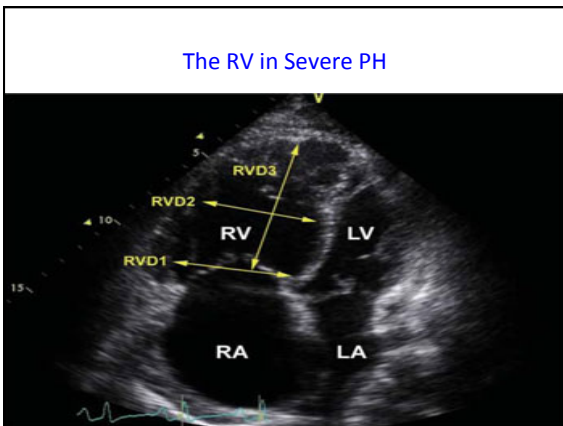
LV Concentric Hypertrophy

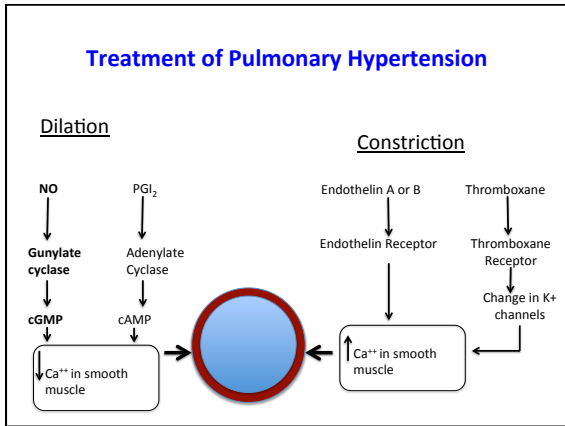


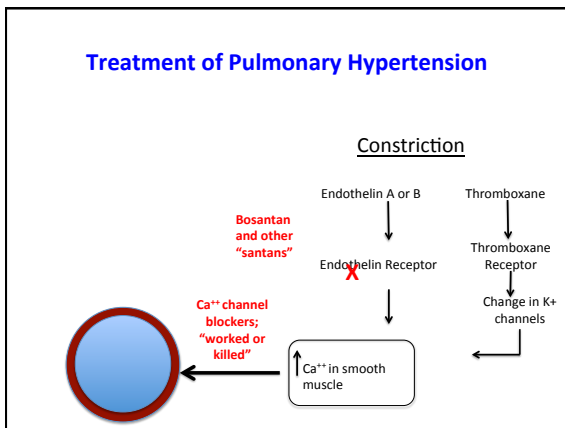
RV Asymmetric Hypertrophy Leads to Huge Free Wall Stress and Dilation

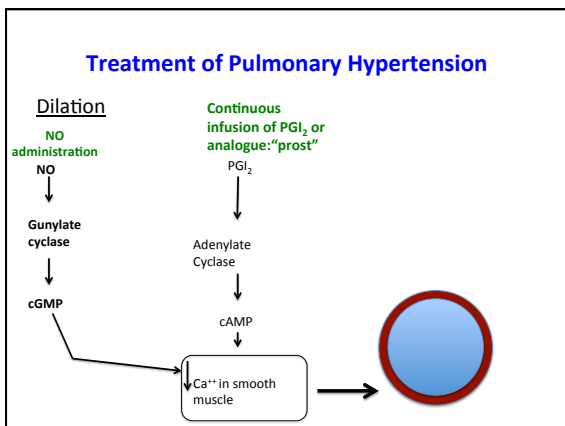


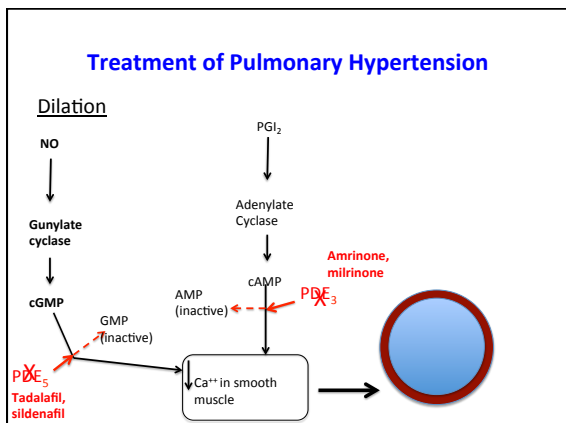
The RV in Severe PH











- ### The Developing Understanding of Treating PH
- We started using these drugs as they were "vasodilators"
 - We found out that more importantly, these drugs can reverse the phenotypic changes of PH after years of therapy
 - PH is a curable disease in some patients

- ### A Caveat
- For reasons we don't understand, in many patients there is an acute, severe vaso-constriction of these drugs are stopped, even in patients who have been on them for years
- Never stop these drugs for your anesthetic for any reason**

**Anesthesia In Patients with PH:
Pre-operative 1**

- You are well aware of the risk of PH and RV overload in patients with advance COPD
- Be suspicious of occult PH in patients with autoimmune diseases such as scleroderma, MCTD and anti-phospholipid antibody syndrome/lupus
- In pediatrics, pulmonary hypertension is most common in kids with high pulmonary flows, usually not present significantly until 2nd decade

**Anesthesia In Patients with PH:
Pre-operative 2**

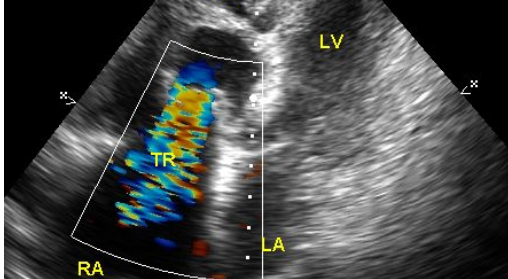
Patients with known PH as usually followed with echocardiograms and these can tell you a LOT:

- 1) Look at the size and shape of the RV; is there LVOT impingement ?
- 2) Look at the description of the RV function; stressed RVs will have areas of asynchronous contraction. This is a VERY bad sign !
- 3) Look at the estimated RV peak pressure; this is calculated from the tricuspid regurgitation *velocity* (in m/sec)

**Anesthesia In Patients with PH:
Preoperative: 3
Tricuspid Regurgitation Velocity**

- 1) As the RV dilates the tricuspid annulus dilates and TR becomes progressively greater
- 2) The TR velocity ("backwards" into the RA) is an estimate of peak RV Pressure

Color Doppler of TR Jet



**Anesthesia In Patients with PH:
Tricuspid Regurgitation Velocity**

Estimated Peak RV pressure = $4 \times (\text{TR velocity in m/sec})^2 + \text{est RA pressure}$

Example: "measured TR velocity is 4.2 m/sec, CVP estimated as 15"

$$RV_{\text{peak}} = 4 \times (4.2)^2 + 15 \text{ or } 85 \text{ mmHg}$$

**Anesthesia In Patients with PH:
Pre-operative: 4**

But, like so many things in Anesthesia, clinical signs and symptoms and your patient's history is the best assessment of functional status....

"The Patsy Questions":

- 1) Can you walk from the car without stopping?
- 2) Can you carry a bag of groceries from the car?
- 3) Can you walk up steps to the house from the car carrying the groceries?

**Anesthesia In Patients with PH:
Intraoperative Monitoring**

- You probably are going to want an arterial line
- If you have NIRS as a non-invasive estimate of perfusion that can be very helpful
- Do NOT use a PA catheter:
 - you probably won't get it in or have it stay due to TR
 - the more you want it the less likely you'll get it in
 - the RV is incredibly irritable and intractable V fib is *well* described when trying to put in PA catheters in patients with PH

Intraoperative Anesthetic Management

- There is no absolutely "wrong" or absolutely "right" way to do these anesthetics
- These are sick, stressed RVs but they do respond to modest dose inotropes
- Patients do NOT tolerate RV preload drop or LV afterload drop (septal obstruction of LVOT)
- Ketamine is known to have NO affect on PVR, it is my choice for induction and during case; midazolam doesn't affect PVR
- N₂O raises normal PVR, probably doesn't do much to already high PVR
- All but the very sickest patients will tolerate some volatile agent IF SVR is maintained



Thank You
