

## Pulmonary Hypertension

S. Clark Bergard, MD  
Boulder Valley Pulmonary  
303-835-9260



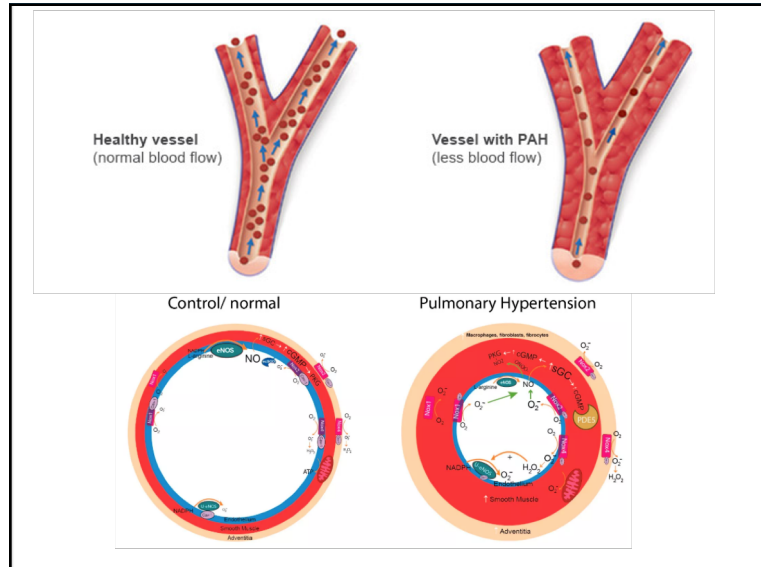
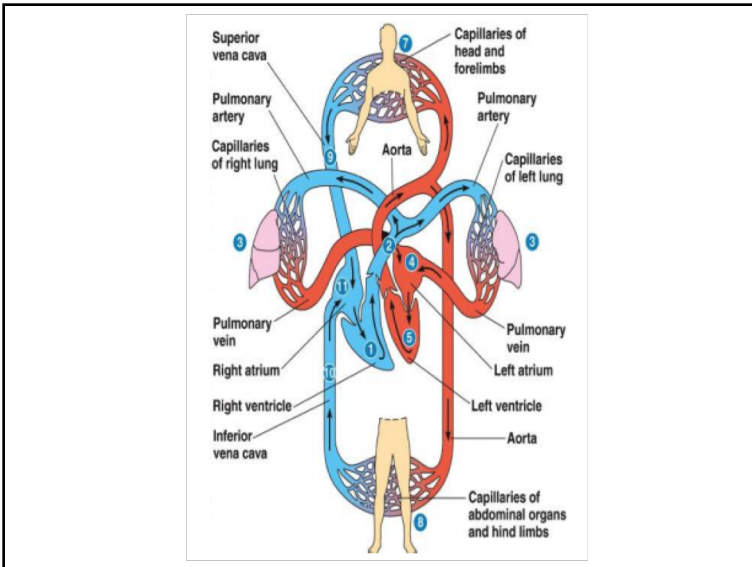
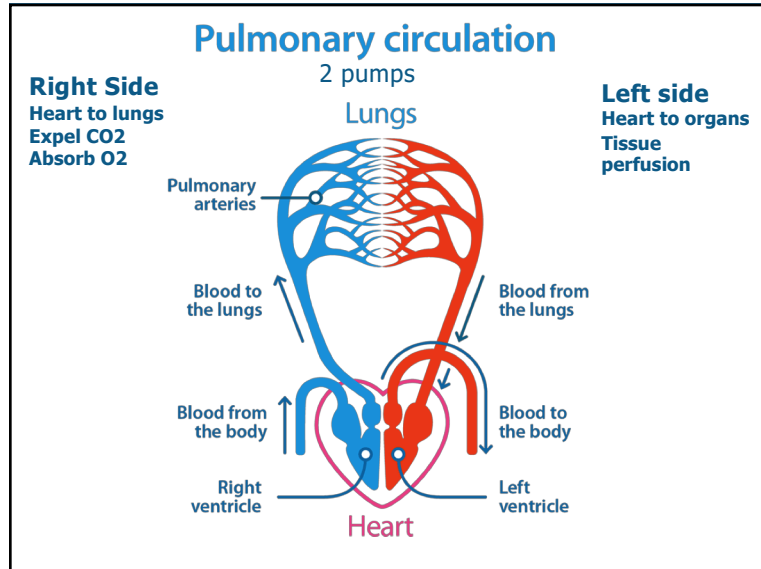
## Pulmonary Hypertension

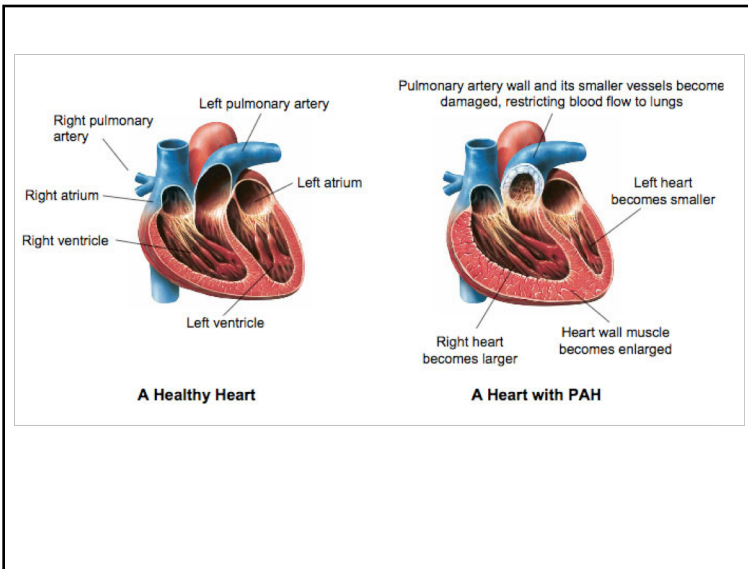
- Definition and types
- Epidemiology
- Pathophysiology
- Treatment Options
  - Medications
  - Pulmonary rehabilitation
- Future directions

## A little about myself

- Chemical Engineering, Biochemistry and Spanish
  - University of Colorado, Magna Cum Lade
- CU School of Medicine
  - Internal Medicine
  - International research year in Guatemala
- Pulmonary, Critical Care and Sleep Medicine
  - UCSD
  - Pulmonary vascular disease
  - Peer reviewed publications

# Physiology Overview...



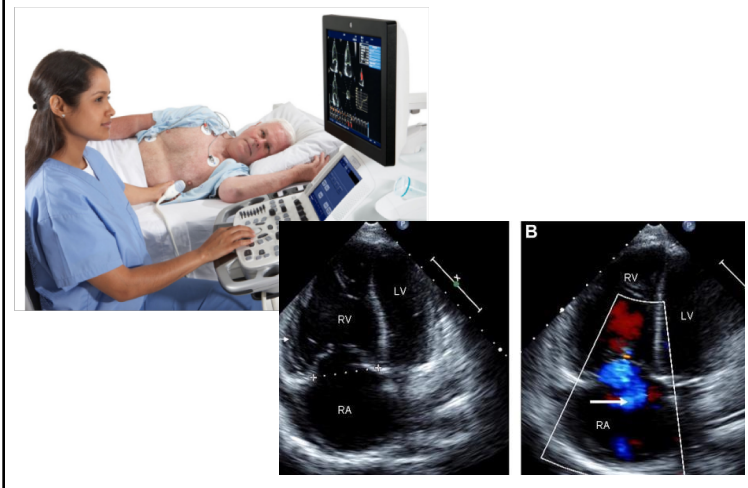


## Signs and Symptoms

- No to minimal symptoms early in the disease
- Shortness of breath
- Fatigue
- Decreased exercise tolerance
- Leg swelling
- Lightheadedness
- Abnormal heart sounds
- Low oxygen levels



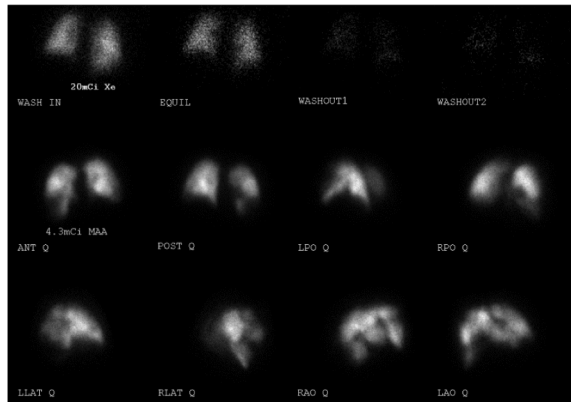
## Echocardiogram



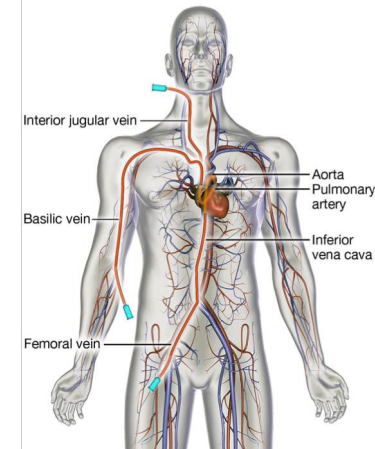
## Lung Function Testing and Overnight Oximetry



## Ventilation Perfusion Scan

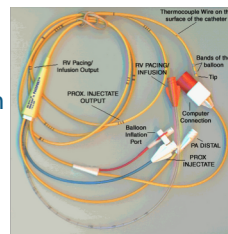


## Right Heart Catheterization



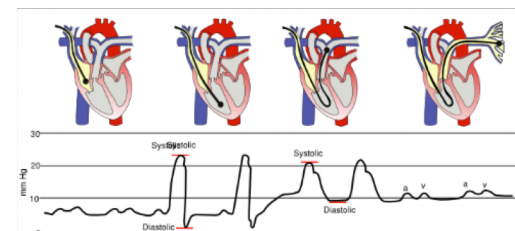
## Basics of Right Heart Catheterization

- Rule of 5's
  - Right Atrium 0-5
  - Right Ventricle 25/5 (mean)
  - Pulmonary Artery 25/10
  - Wedge Pressure <15
- Cardiac Output by thermodilution
- Cardiac Output by the Fick Equation
- Pulmonary Vascular Resistance



## Pulmonary Arterial Hypertension

- Mean pulmonary artery pressure  $\geq 25$  mm Hg
- Pulmonary Arterial Hypertension
  - Mean pulmonary artery pressure  $\geq 25$  mm Hg
  - Pulmonary artery occlusion pressure  $<15$
  - Pulmonary vascular resistance of 3 Wood Units



## World Health Organization (WHO) Classification

- Classification based on etiology
- Proper classification is essential
  - Prognosis
  - Treatment
- Five distinct classes
- Treatment varies based on classification

## Pulmonary Hypertension

1. Pulmonary arterial hypertension
2. Pulmonary venous hypertension (left heart disease)
3. Pulmonary hypertension due to chronic hypoxemia
4. Chronic thromboembolic pulmonary hypertension (CTEPH)
5. Pulmonary Hypertension with unclear, multifactorial mechanisms (will not discuss tonight)

## World Health Organization (WHO) Classification

1. Pulmonary Arterial Hypertension
  - Collagen Vascular Disease (Autoimmune Disease)
  - Congenital Heart Disease (L→R Shunt)
  - HIV
  - Drug and Toxin: Anorexigens (i.e. Fen-Phen) and amphetamines
  - Porto-pulmonary HTN (liver disease)
  - Familial/Heritable PAH (BMPR2, ALK-1, ENG, SMAD, CAV1)
  - Schistosomiasis (parasite)
  - Idiopathic (unknown) PAH
- 1'. Pulmonary veno-occlusive disease
- 1". Persistent pulmonary hypertension of the newborn

## WHO group 2, pulmonary hypertension due to left heart disease

- Congestive heart failure
- Diastolic dysfunction/failure
  - Longstanding essential hypertension
  - Advanced age
- Valvular heart disease
  - Aortic stenosis and regurgitation
  - Mitral stenosis and regurgitation

### WHO group 3, pulmonary hypertension due to lung disease

- Chronic obstructive pulmonary disease (COPD)
- Asthma
- Interstitial lung disease
- Chronic hypoxemia
- Sleep apnea, both central and obstructive
- Chronic exposure to high altitudes

### WHO group 4, pulmonary hypertension due to chronic thromboembolic disease

- 5% of patients with pulmonary embolism do not resolve their clots
- Chronic clot becomes scar tissue
- Both mechanical obstruction and primary vasculopathy
- Often missed and treated at WHO group I PAH
- Surgery can be curative

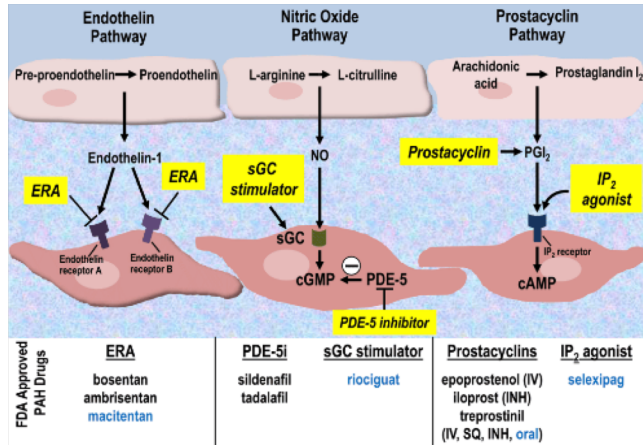
### Treatment

- Who to treat
- Who NOT to treat
- When to intervene
- Why
  - Improve quality of life
  - Improve survival
  - Prevent progression of disease

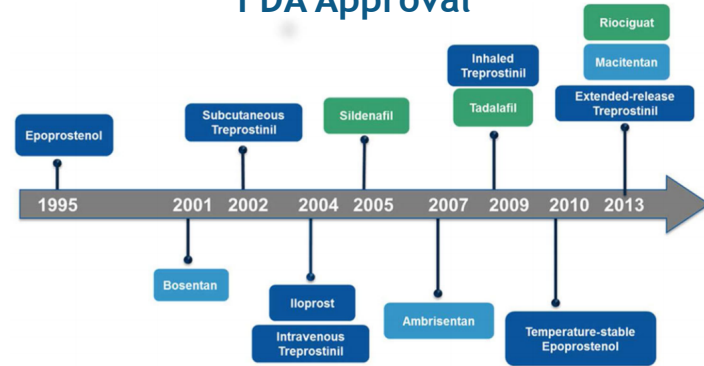
### PH Specific Therapy

- Pulmonary Vasodilators
- Improvements in survival, and quality of life
- Patients with WHO I PAH
- Occasionally used in other types
  - Inappropriate use can be harmful

### 3 Pathways for PAH Medications



### Timeline of PAH Medications Gaining FDA Approval

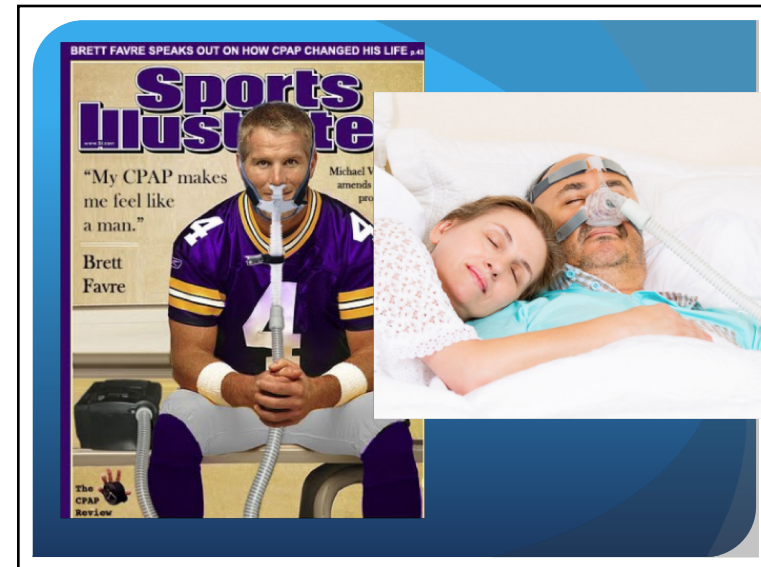


### WHO 2 PH - Left sided Heart and Lung Disease

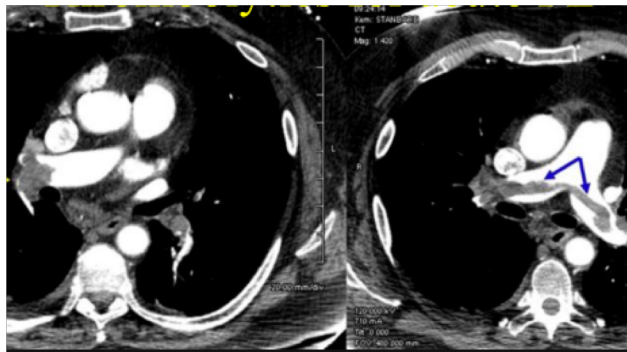
- Pulmonary Venous hypertension
- Pressures build and back flows from the left side
- Correct valve problems - mitral and aortic valve problems
- Optimize blood pressure and heart failure medications
- Adjunctive treatments have proven efficacy

### WHO 3 Pulmonary Hypertension Lung Disease and Sleep Apnea

- 20-40% of patients with OSA have mild elevations in pulmonary artery pressures.
- Nocturnal oxygen testing
- Sleep apnea testing
- Treatment with oxygen and CPAP can be curative



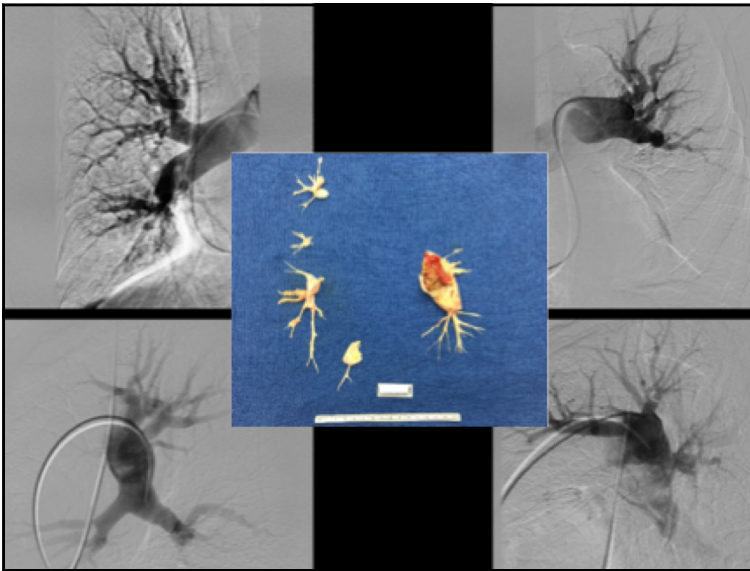
### WHO 4 - Chronic Thromboembolic Pulmonary Hypertension



### Chronic Thromboembolic Pulmonary Hypertension

- 3-5% patients with acute PE do not resolve their thrombus, despite systemic anticoagulation
- Nonspecific symptoms, up to 30% do not have prior PE diagnosis
- High index of suspicion required to make an accurate diagnosis
- Persistent SOB after 3-6 months of anticoagulation warrants further investigation
- Surgery is curative.



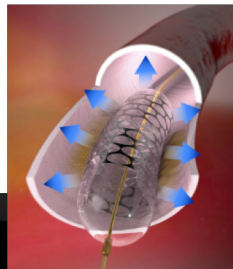


## Adjunctive Treatment Options

- Pulmonary vasodilators in WHO I PAH
- CPAP and supplemental oxygen
- Inhaled beta agonists improve exercise capacitance
- Aspirin can attenuate PA pressures
- Special diuretics can improve RV function
- Pulmonary Rehabilitation

## On the horizon

- Pulmonary artery denervation
- Mounting data for WHO 1 and 2 disease



## Pulmonary Rehabilitation

- Proven benefits in multiple clinical trials
- shown to increase exercise capacity and peak oxygen consumption and resting heart rate
- Improve health related quality of life
- Reduce depression and fatigue



## Pulmonary Rehab at BCH

- Gail Anderson, RN, BSN
- Tricia Huso, RT
- Keri Anderson, RT

- Clinically proven benefits in a variety pulmonary diseases
- Experience less difficulty breathing
- Increase muscle strength and endurance
- Improve their ability to cope with daily activities
- Understand how to use medication and oxygen
- Improve quality of life
- Reduce hospitalizations
- Improve depression

## Phase II Pulmonary Rehab

- PHASE II
- Paid for by insurance
- Monitored exercise sessions
- Individual care plans
- Education sessions on diet, exercise, stress management, medication, risk factor reduction, and goal setting strategy and technology.

## Phase III, or Maintenance

- Some patients need and/or enjoy continued therapy
- Patients must have a prescription from their provider to attend Phase III
- This program is self-pay
- Cost is \$70 for 10 visits, or \$7 per visit
- Much cheaper than a personal trainer

## In Summary

- Unexplained shortness of breath
- Vague symptoms
- Determining type and etiology is essential to guide treatment and prognosticate
- Not all types lead to progressive declines and death
- Many treatment options with more on the horizon

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## Questions?



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