Pulmonary Hypertension

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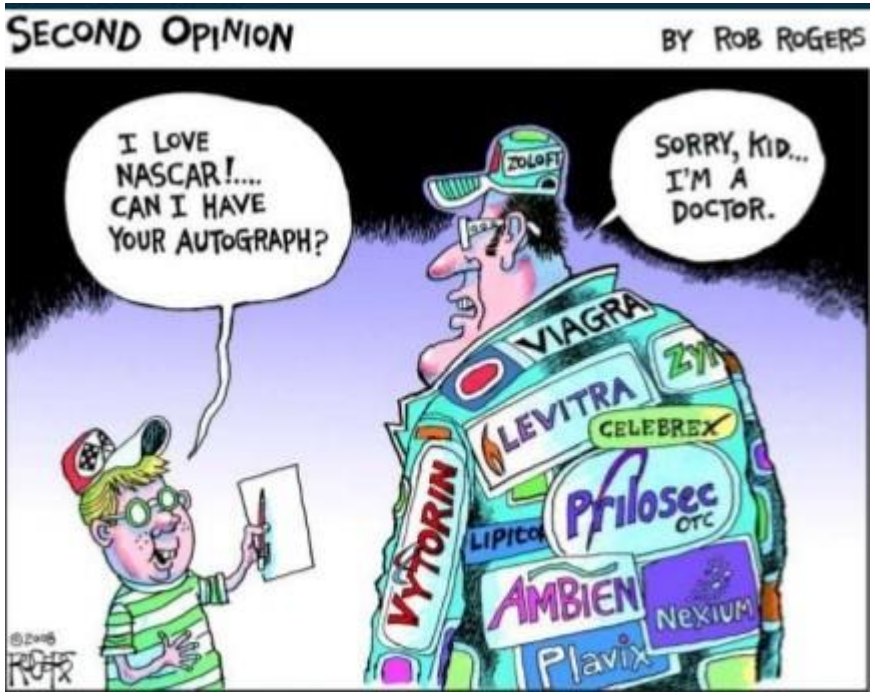
Conflict of interests

- None



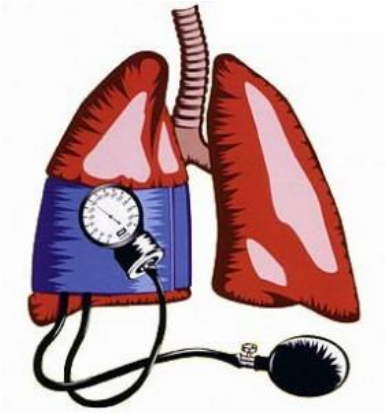
Pulmonary Hypertension

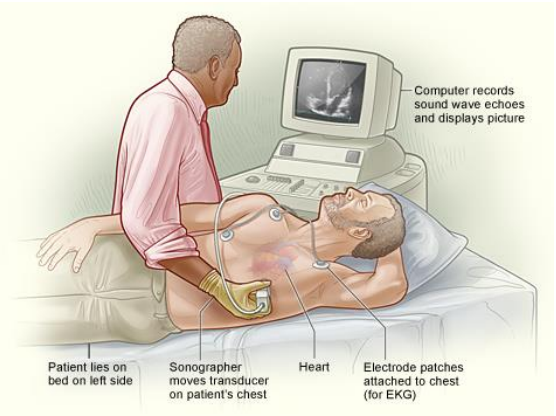
- **Talk outlines**:
  - What is pulmonary Hypertension (PH)
  - Causes and classification of PH
  - Symptoms of PH
  - Diagnosing PH
  - Treatment of PH & PAH
  - Living with PAH

What is Pulmonary Hypertension ?

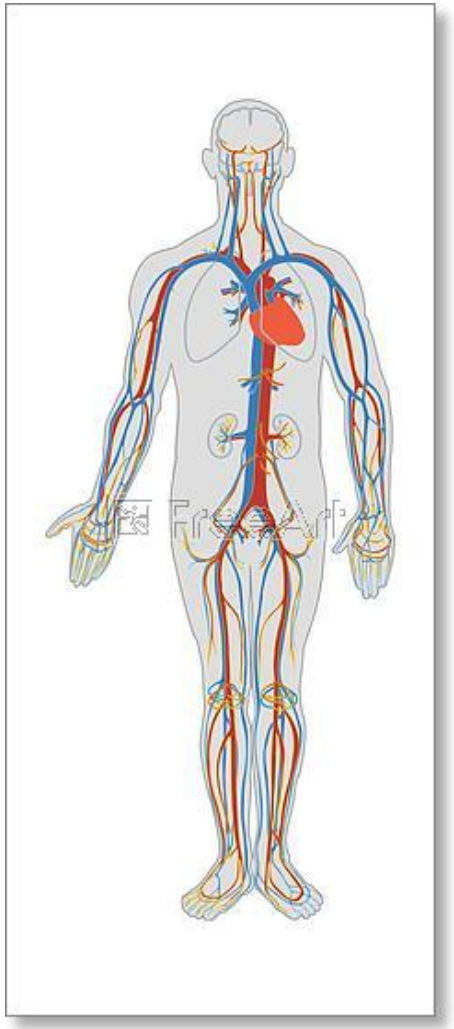
- “Regular” **Hypertension**
  - High blood pressure in the artery throughout body

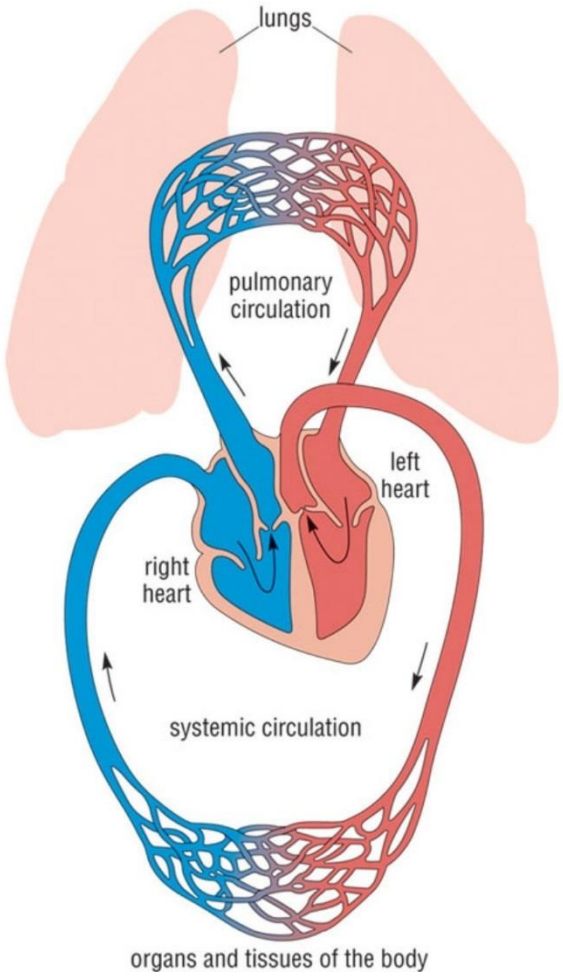
- **Pulmonary Hypertension**
  - High blood pressure in the lungs

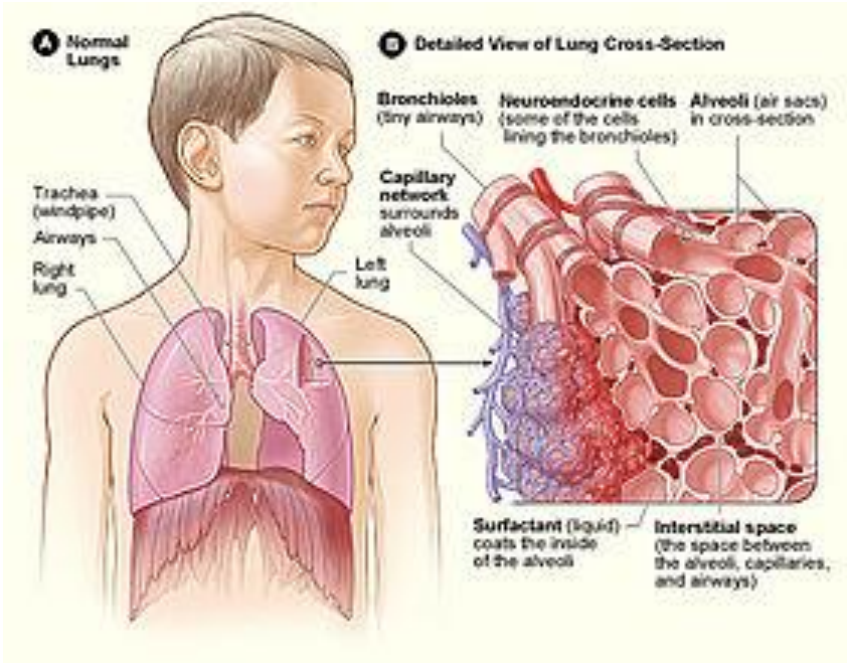




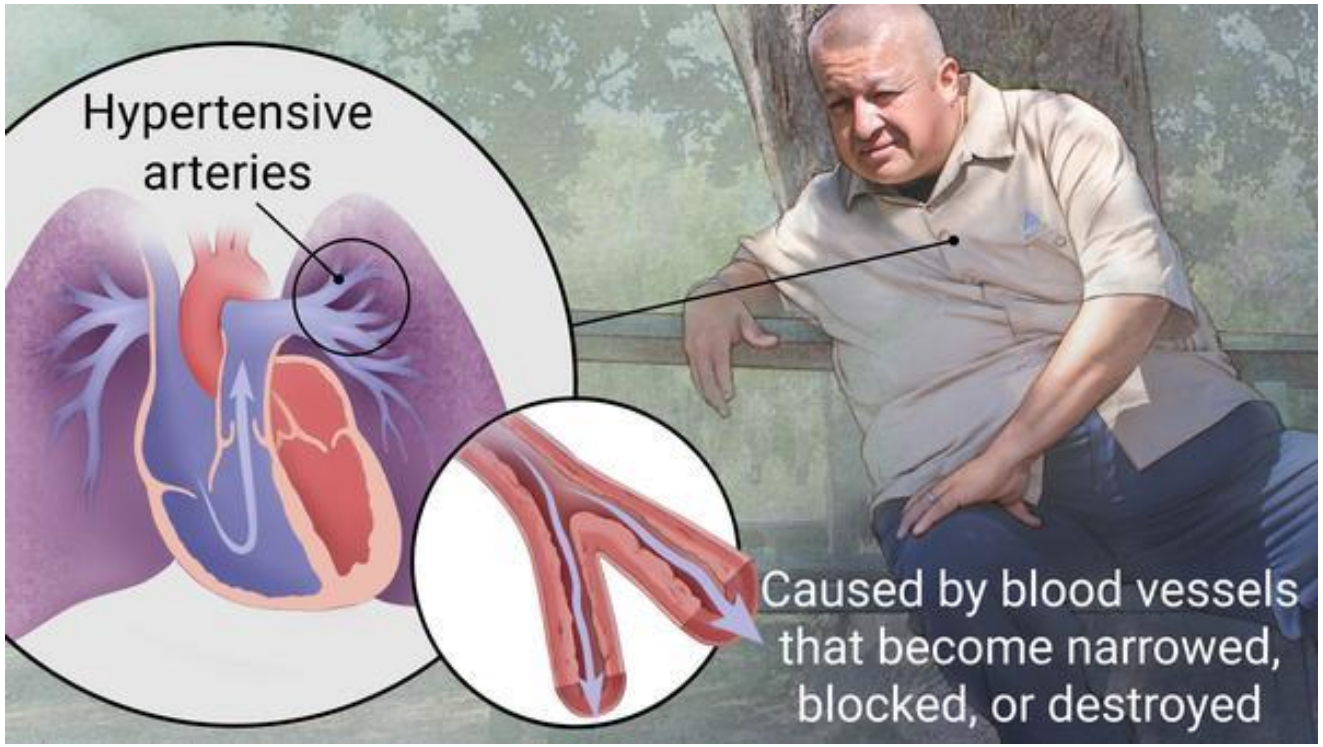
Pulmonary Circulation





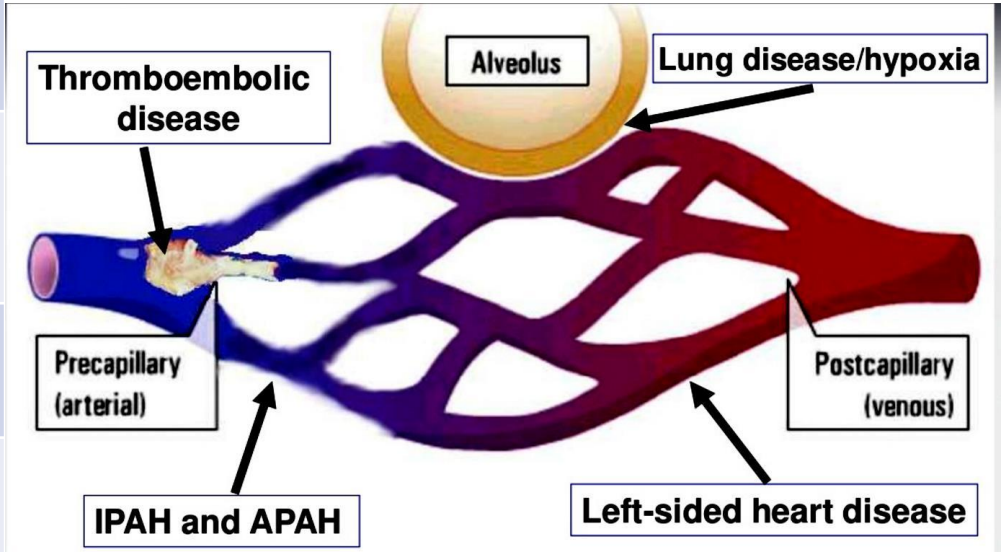


What causes Pulmonary Hypertension?



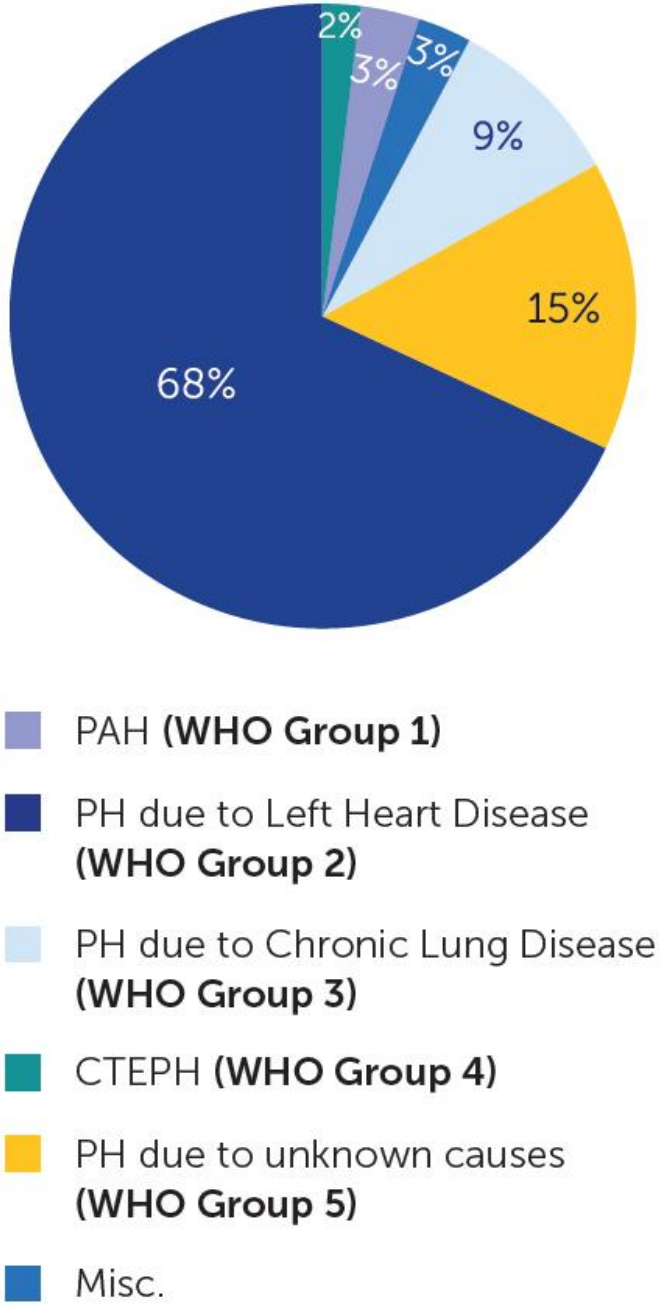
Types of Pulmonary Hypertension

Group	Disease state
1. Pulmonary artery hypertension (PAH)	Idiopathic, familial, associated PAH
2. PH from left sided heart disease	Systolic and diastolic heart failure, valvular heart disease
3. PH from chronic hypoxic lung disease	Severe COPD, pulmonary fibrosis, severe OSA
4. PH from chronic blood clots in lung	CTEPH
5. PH from unclear multifactorial mechanisms	Sarcoidosis, chronic metabolic and hemolytic disorder, chronic renal failure



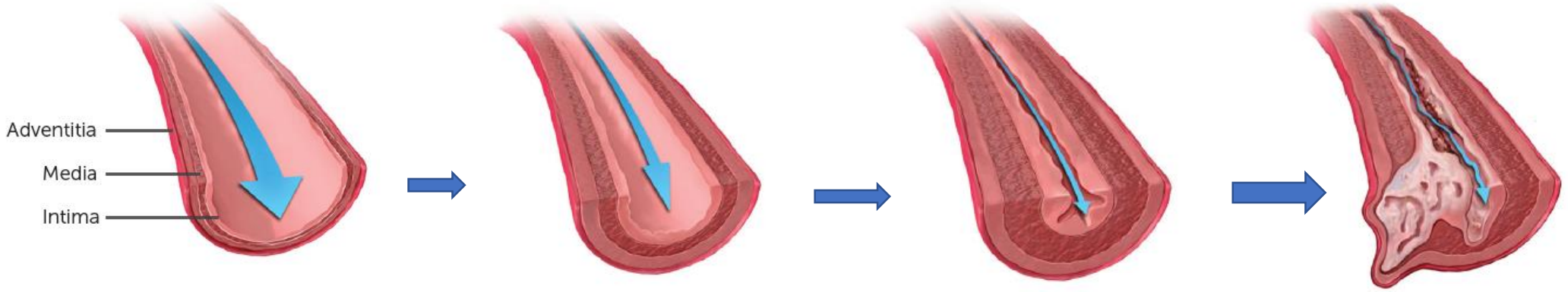
IPAH = idiopathic pulmonary artery hypertension; APAH = associated pulmonary artery hypertension.

Epidemiology of Pulmonary Hypertension



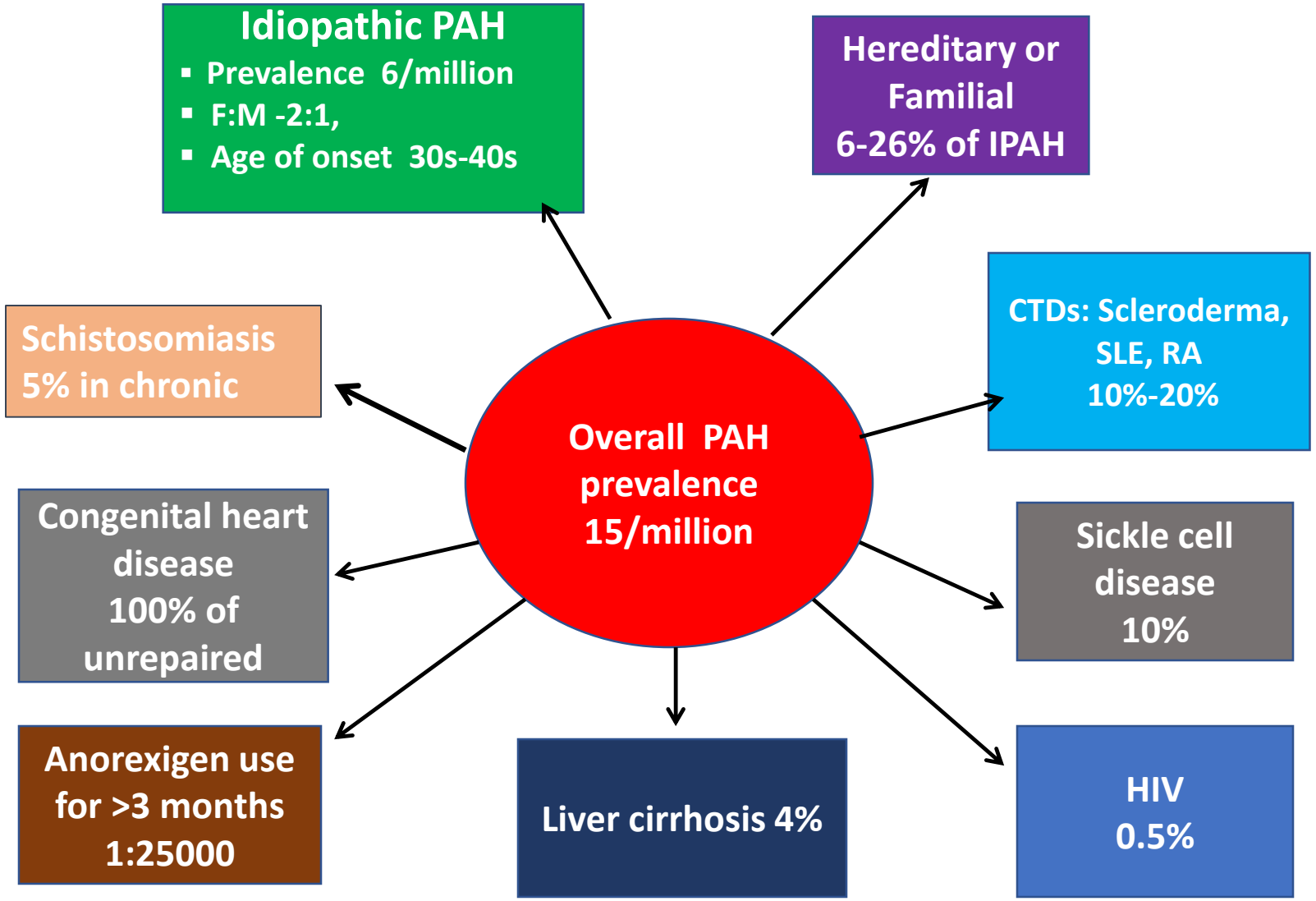
Pulmonary arterial hypertension (Group 1 PH)

- Specific type of pulmonary hypertension where the tiny blood vessels in the lungs become stiff, scarred and narrow.



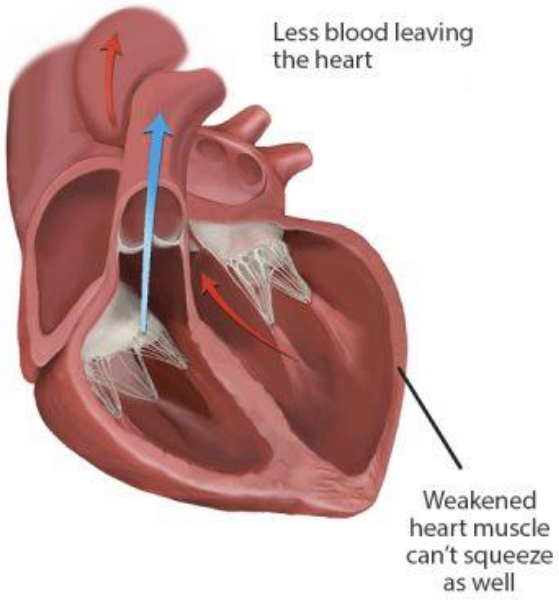
- **Group 1 PAH** is a very serious disease.
- It is not the same as other groups of PH

Risk factors for Group 1 PAH

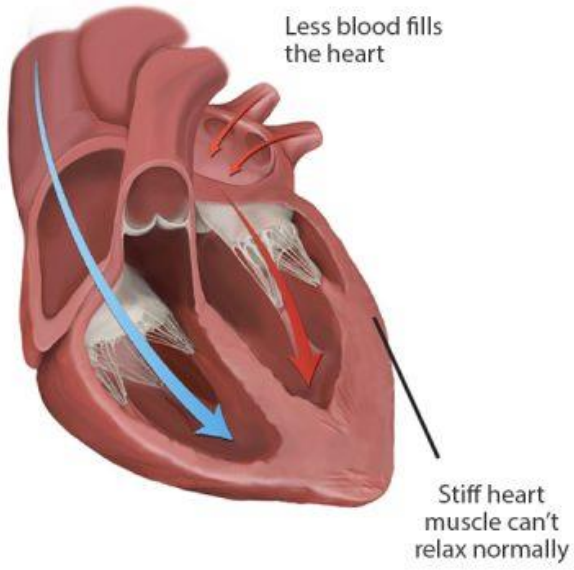


ACCF/AHA 2009 Expert Consensus Document on Pulmonary Hypertension

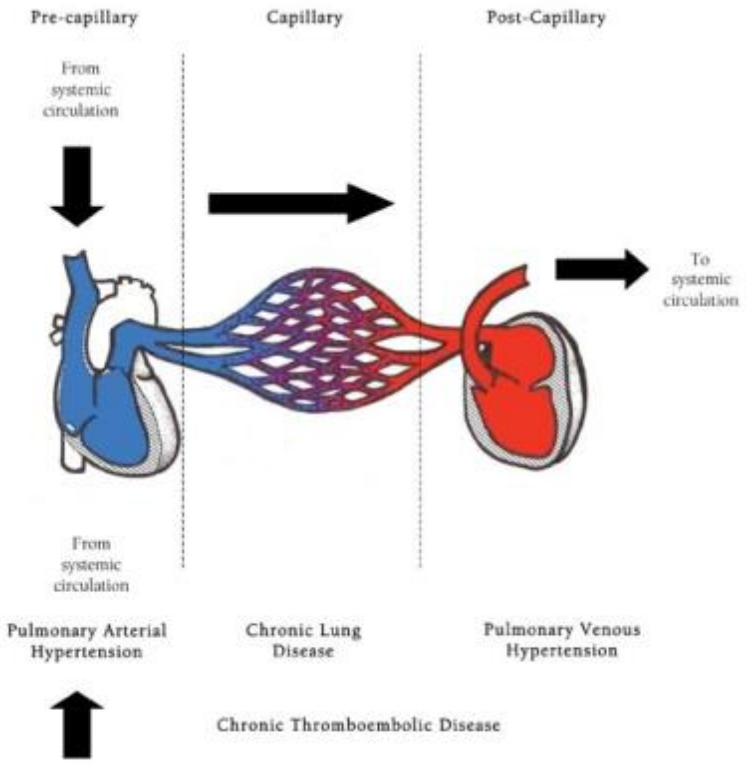
Group 2 PH: due to left heart disease



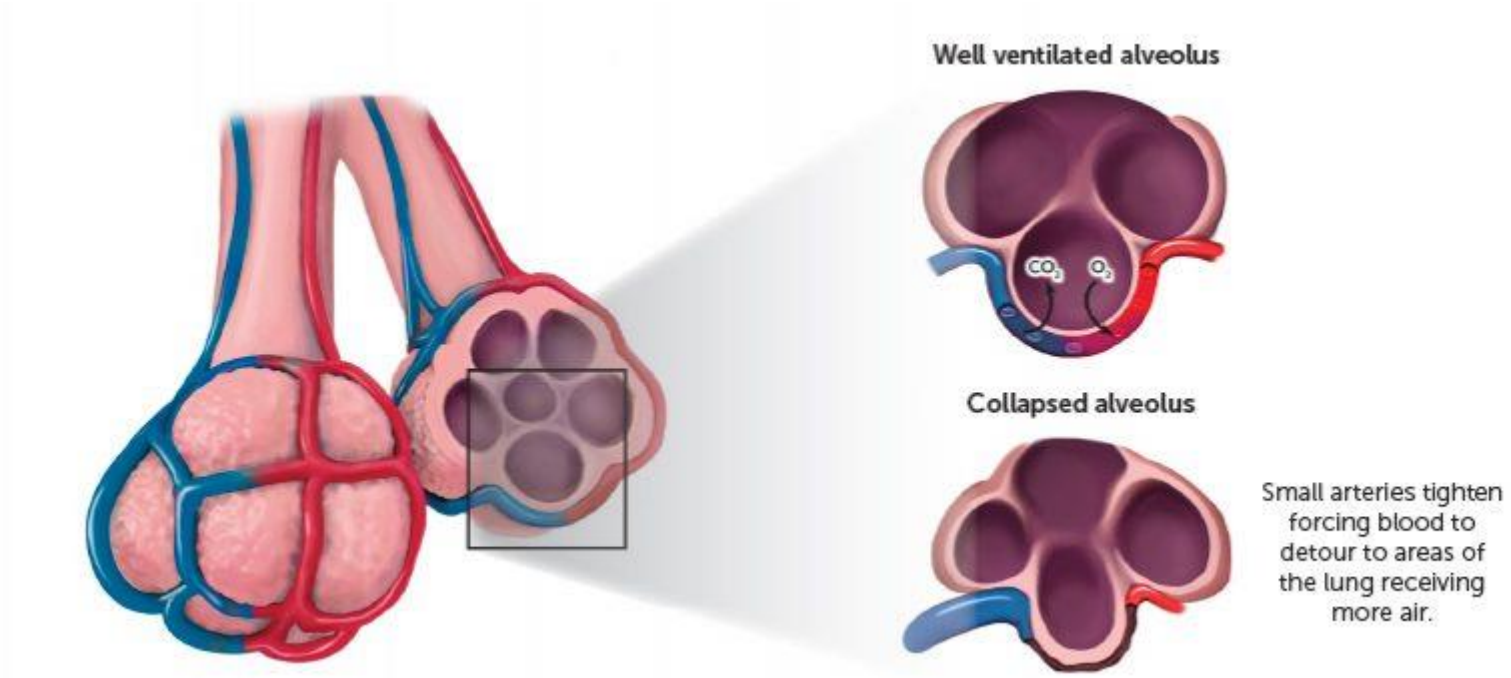
Systolic heart failure



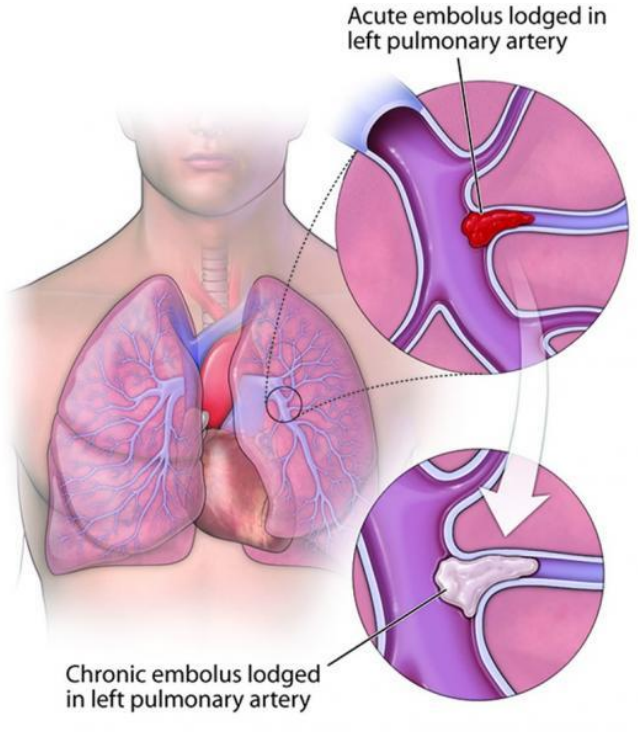
Diastolic heart failure

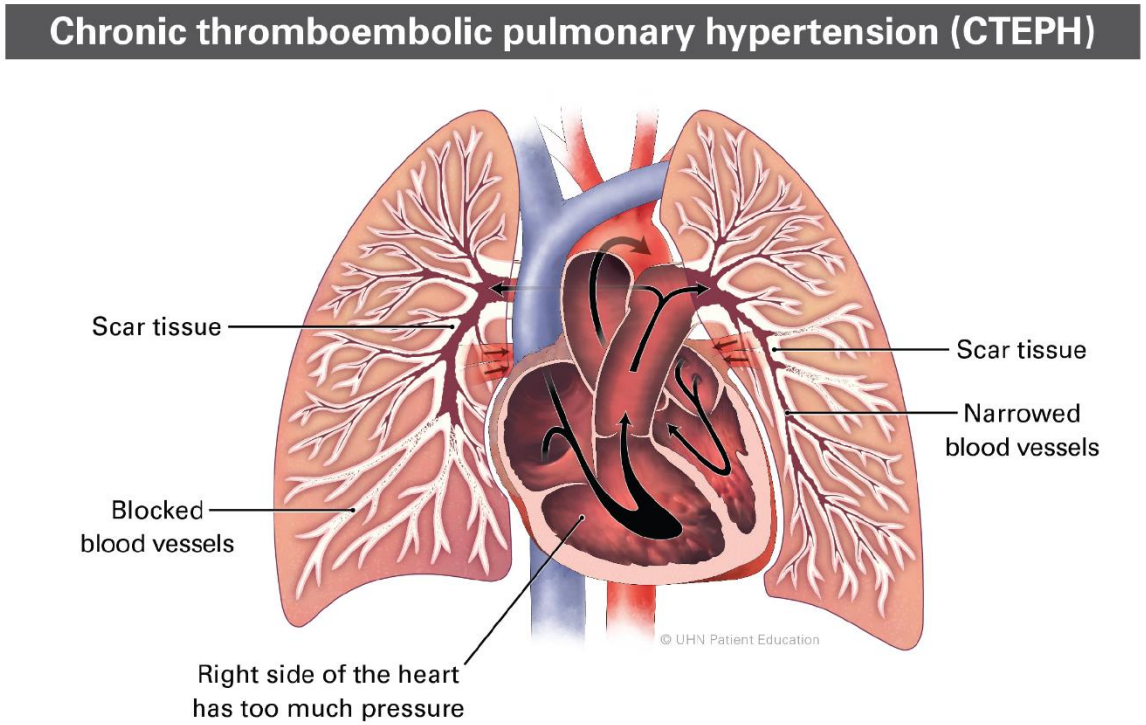


Group 3 PH : due to chronic hypoxic lung disease



Group 4 PH: due to chronic blood clot in the lungs





Symptoms of Pulmonary Artery Hypertension

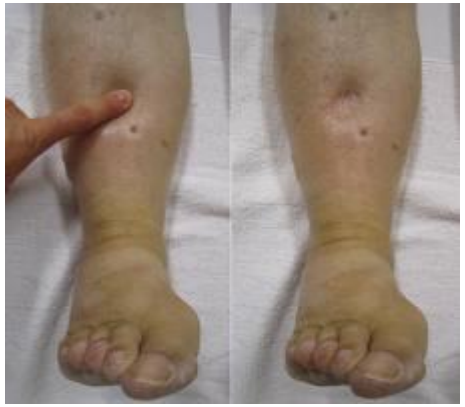
Shortness of breath in 98%

Fatigue in 73%

Chest pain in 47%

Palpitations in 33%

Syncope in 36%



Leg swelling 21%

NIH Registry

Diagnosing Pulmonary Artery Hypertension

- Very often overlooked, and patients typically experience 2-3 years of frustrating physician visits before being correctly diagnosed.

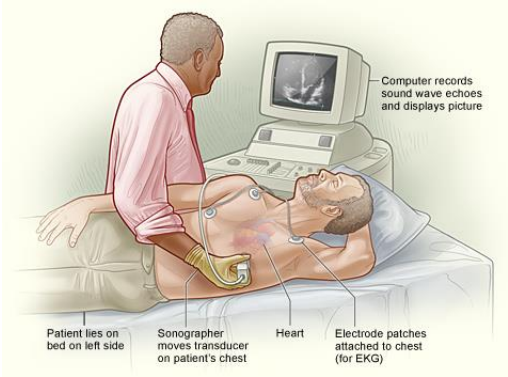
Why?

- Because most of the symptoms are common with other common lung disease like asthma, COPD

- High degree of suspicion is required (Risk factors)

Diagnosing Pulmonary Artery Hypertension

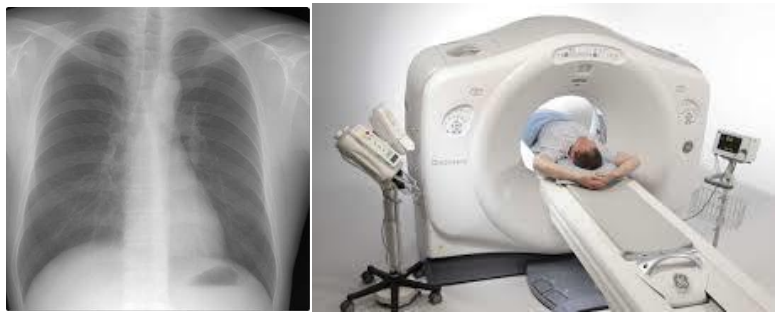
- Once PH is suspected, first test to order is ?
  - Echocardiogram



  - Can overdiagnoses or miss pulmonary hypertension

Diagnosing Pulmonary Artery Hypertension

- Once PH is diagnosed on Echocardiogram, further tests are ordered to find out the causes.



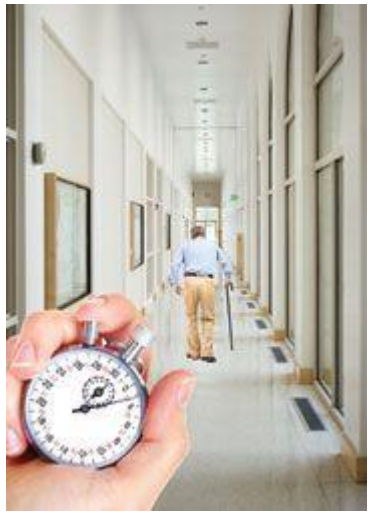
Chest X-ray and CT scan

PFT (breathing test)

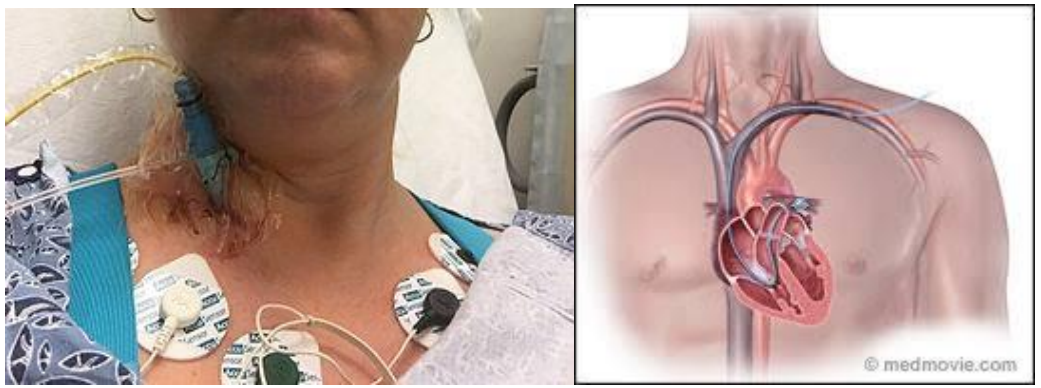
Nuclear (V/Q) scan

Sleep apnea test

Blood tests



6 minutes walk test



Right heart catheterization (Gold standard)

Treatment of PAH & PH

- PAH is a progressive condition with a high mortality if left untreated
  - Survival rates in IPAH at 1, 3, & 5 years were 68%, 48% and 34% before discovery of meds
- **Overall survival improves with "EARLY DIAGNOSIS and start of proper therapy in time"**
- Major advancements have been made in the PAH specific drugs in the past two decades
- Survival rate in IPAH at 5 years has improved to **60% from 34% with current therapy**
- There is no cure for PH

Treatment of PH

- **Supportive medical therapies** **(For all groups of PH)**
  - **Oxygen** inhalation via a nasal cannula or face mask
  - **Diuretics** – Get rids of excess fluid that puts pressure on the heart
  - **Digoxin** – Assists the pumping of the heart
  - **Blood thinner pills** to prevent blood clot formation
  - **Physical therapy to** improve DOE

- **PAH specific drugs or Pulmonary vasodilators** **(For only group 1 PAH)**
- **Lung Transplant if medical therapy fails**

PAH specific drugs:

- **Targets pulmonary arteries and decrease blood pressure by dilating them**
- **Used only in group 1 PAH**
- **Major advancement in last two decades**
- **Improves :**
  - **symptoms,**
  - **exercise capacity,**
  - **quality of life and survival**
- **& Prevents clinical worsening**

- **Available in pills, Inhalational, subcutaneous and Intravenous forms**

PAH specific drugs:

- **Epoprostenol (Flolan®) -IV**
- **Treprostinil (Remodulin, Tyvaso, Orenitram)**
- **Iloprost (Ventavis®)**
- **Selexipag (IP receptor agonist)**

Prostaglandins

- **Bosentan (Tracleer®)**
- **Ambrisentan (Letairis®)**
- **Macitentan (Opsumit)**

Endothelin receptor antagonists

- **Tadalifil (Adcirca®)**
- **Sildenafil (Revatio®)**

Phosphodiesterase 5 inhibitors

- **Riociguat (Uptravi)**

Soluble Guanylate cyclase activators

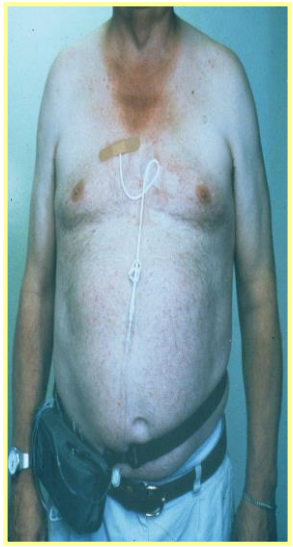
Comparison of PAH specific therapy

Drugs	Cost $ (annual)	Route	Frequency	Ease of Use	Side effects	
Epoprostenol	~100,000	IV	Continuous	+	+++	
Treprostinil	>175,000	Oral, SQ, IV, Inhaled	Continuous	++	+++	
Iloprost	~175,000	Inhaled	6-9x per day	++	++	
Sildenafil	~15,000	Oral	TID	+++	+	
Tadalafil	~12,000	Oral	Daily	+++	+	
Bosentan	~75,000	Oral	BID	++++	+	
Ambrisentan Macitentan	~75,000	Oral Oral	Once a day Once a day	++++	+	

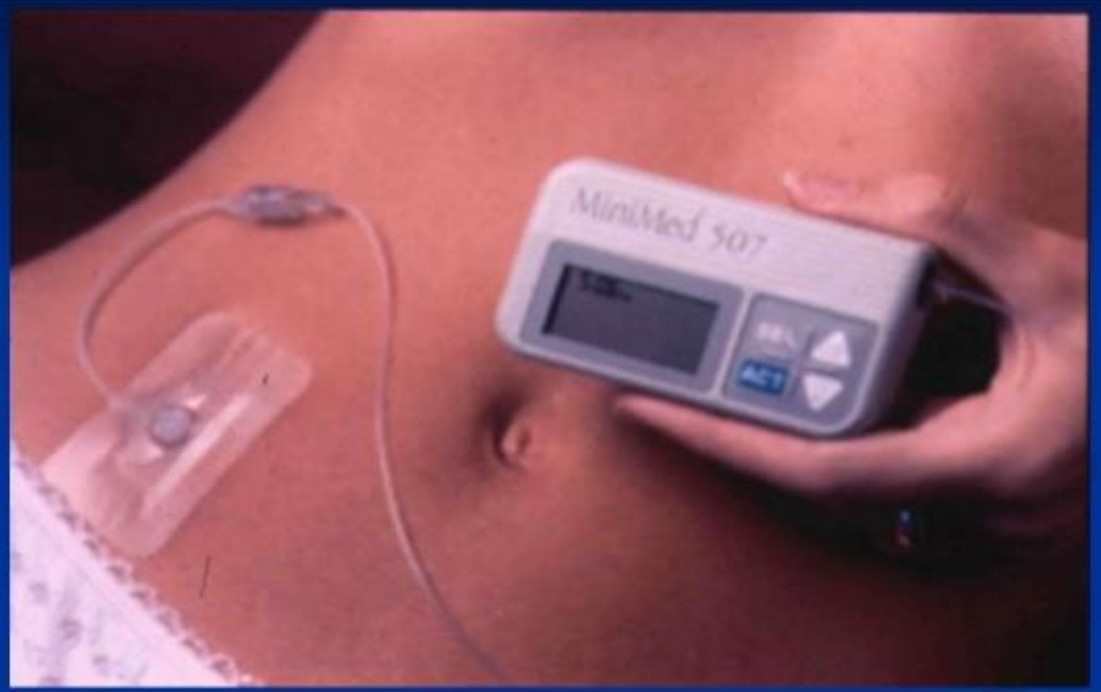
IV Epoprostenol (Flolan®) & Treprostinil (Remodulin)

- Requires <u>continuous infusion via chronic indwelling catheter</u>.
  - Risks of drug interruption
  - Risk of iv catheter infection, blood clot

- **Half-life short**
- Abrupt withdrawal can result in rebound PH (can be fatal)



Subcutaneous Treprostinil (Remodulin®)



- SQ administration
- Longer half-life than epoprostenol
- Pre-mixed
- Stable at room temperature

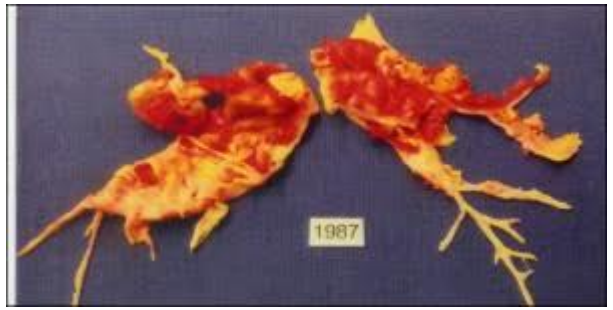
Inhaled Prostanoids

Tyvaso

Ventavis

Treatment of other group of PH

- **Group 4 due to chronic blood clots in the lungs**
  - Surgical removal of blood clots



  - Riociguat (PAH specific drug) in patients not a candidate for surgery
  - Blood thinner and other supportive treatment

- **Group 2, 3 & 5**:
  - Treatment of underlying heart and lung disease plus supportive treatment
  - PAH specific drugs are not used

Living with PAH

- **What to expect regarding treatment**
  - **There is no cure for PAH.**
  - **The goals for treatment is to slow the course of the disease.**
  - **Response to medicine is somewhat unpredictable and depends on how advanced the disease is at diagnosis as well as the underlying cause.**

Living with PAH

- **What else to expect?**
  - **Change in life styles**
    - Medications (continuous IV, SQ, Inhalation 4-6 times/day)
    - Appointments q3-4 months
    - Testing (Echo, 6 minutes walk tests, blood tests)
  - **Financial stress**
  - **Psychological stress**

Living with PAH

- **To best manage your PAH you should**:
  - Take your medication exactly as directed.
  - Do not stop a medication without consulting with your doctor.
  - Take care to never run out of medication.
  - Avoid things that can put a strain on the lungs and heart.
    - Smoking, illicit drugs, OTC decongestants
  - Adhere to a low salt diet and limit fluid intake.
  - Monitor your weight to recognize fluid retention.
  - Develop strategies to help cope with fatigue and shortness of breath.
  - Develop a careful exercise program with your healthcare providers.
  - Get immunized against flu and pneumonia.
  - Talk to your doctor about anxiety and depression
  - Talk to your provider about support group

Living with PAH: Finding Support

CARE, SHARE, BE AWARE

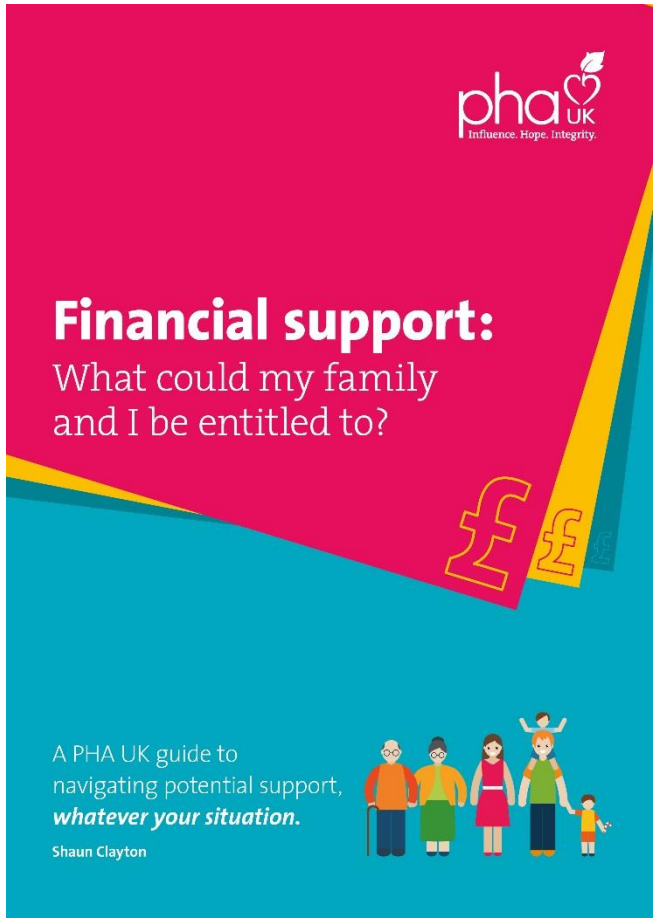
PHA

Pulmonary Hypertension Association

Support Groups

myPHA

Join PHA's free social network exclusively for the pulmonary hypertension community.



Pulmonary Hypertension program at SUNY Upstate

- **Treating Physicians**
  - Birendra Sah, MD
  - James Sexton, MD
  - Anish Desai, MD

- **Clinical Nurse co-ordinator**
  - Naquia Worrell, RN
  - Direct phone line: 315 464 3132

Take home points

- **Pulmonary hypertension is high blood pressure in the lungs**
- **Classified in 5 groups based on etiology (can be difficult)**
- **Classification is important for treatment**
- **Group 1 PH is also called PAH**
- **PAH progressive and fatal if left untreated**
- **PH specific drugs (pulmonary vasodilators) are used only to treat group 1 PAH**
- **Living with PAH can be challenging**