

# Pulmonary Hypertension

## NMHS Pulmonary Symposium

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# Pulmonary hypertension

# Learning objectives

- By the end of this talk, you will be able to:
  - 1. What Pulmonary Hypertension actually is (and is NOT)
  - 2. When to **worry** about it
  - 3. What tests really matter (and why right-heart caths are not as scary as they sound)
  - 4. How each of YOU change outcomes
  - 5. When to escalate care (i.e. let's get some professionals involved)
  
- 6. Have fun! (Maybe)

# What is pulmonary hypertension?

- High blood pressures in the pulmonary arteries
  - Not your typical hypertension
- 2022 ESC/ERS definition: mean pulmonary artery pressure (mPAP) > 20 mmHg at rest (measured by right-heart catheterization)
- Pre-capillary PH (the dangerous kind): mPAP >20 mmHg + pulmonary artery wedge pressure (PAWP) <15 mmHg + Pulmonary Vascular Resistance (PVR) >2 WOOD Units (recently updated from older cut-off 3 WOOD Units)

# WHO GROUPS

## The 5 clinical categories



- Groups 2 & 3 are the most common in our daily practice
- You do NOT need to memorize these... just recognize patterns

# Why should we care?

## Epidemiology and impact

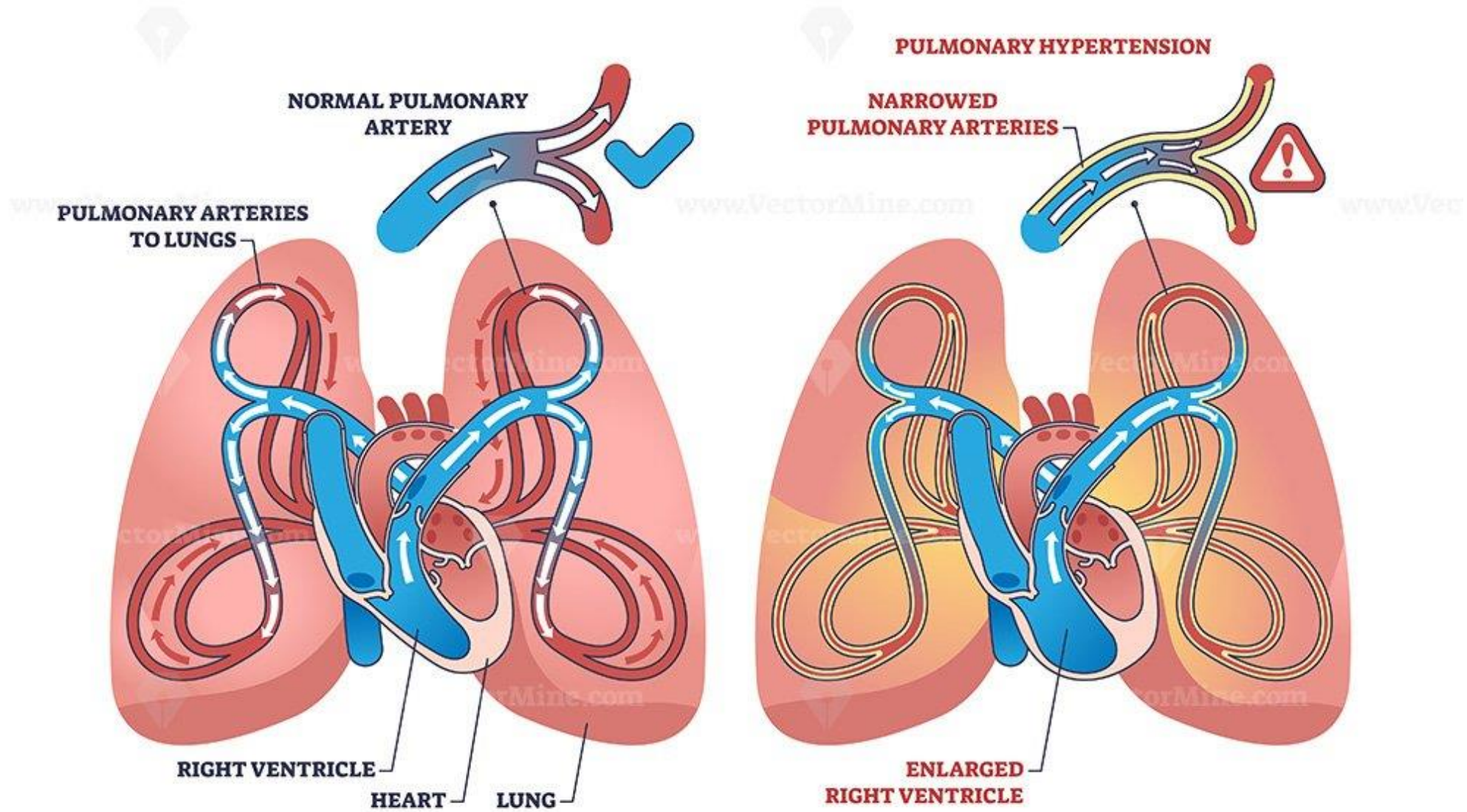
- This affects ~1 in 10-20 heart-failure or COPD patients (Group 2 and 3 are common; Group 1 PAH is rarer but worse prognosis)
- 5-year survival without treatment historically <50% in severe cases
  - Now markedly improved with better care delivery; team-based care
- High hospital readmission rates
  - Right-heart failure, reduced functional status
- This is the patient who keeps coming back
- This is the COPD that doesn't get better

- 1-year survival: Pre-modern ~70% → Post-modern ~90%
- 3-year survival: Pre-modern ~45% → Post-modern ~75%
- 5-year survival: Pre-modern ~30% → Post-modern ~60%

# Pathophysiology made “easy”

- Endothelial Injury
  - ↓
  - Vasoconstriction + Remodeling
  - ↓
  - Pulmonary Pressure ↑
  - ↓
  - Right Ventricle Strain
  - ↓
  - RV Failure
- ***If you remember one thing: patients do NOT die from the lung pressures, the mortality is due to right heart failure***

# PULMONARY HYPERTENSION



# Clinical presentation

## What we see first

- What symptoms do you hear every day?



- Signs: loud P2, jugular venous distention, RV heave, edema, ascites
- These look like common problems/symptoms... and they are!

# Diagnosis:

## A stepwise approach

- Symptoms



- History & Physical + Basic Testing (EKG, CXR, PFTs)



- ECHO 🩺 (screening tool)



- V/Q Scan (rule out CTEPH)



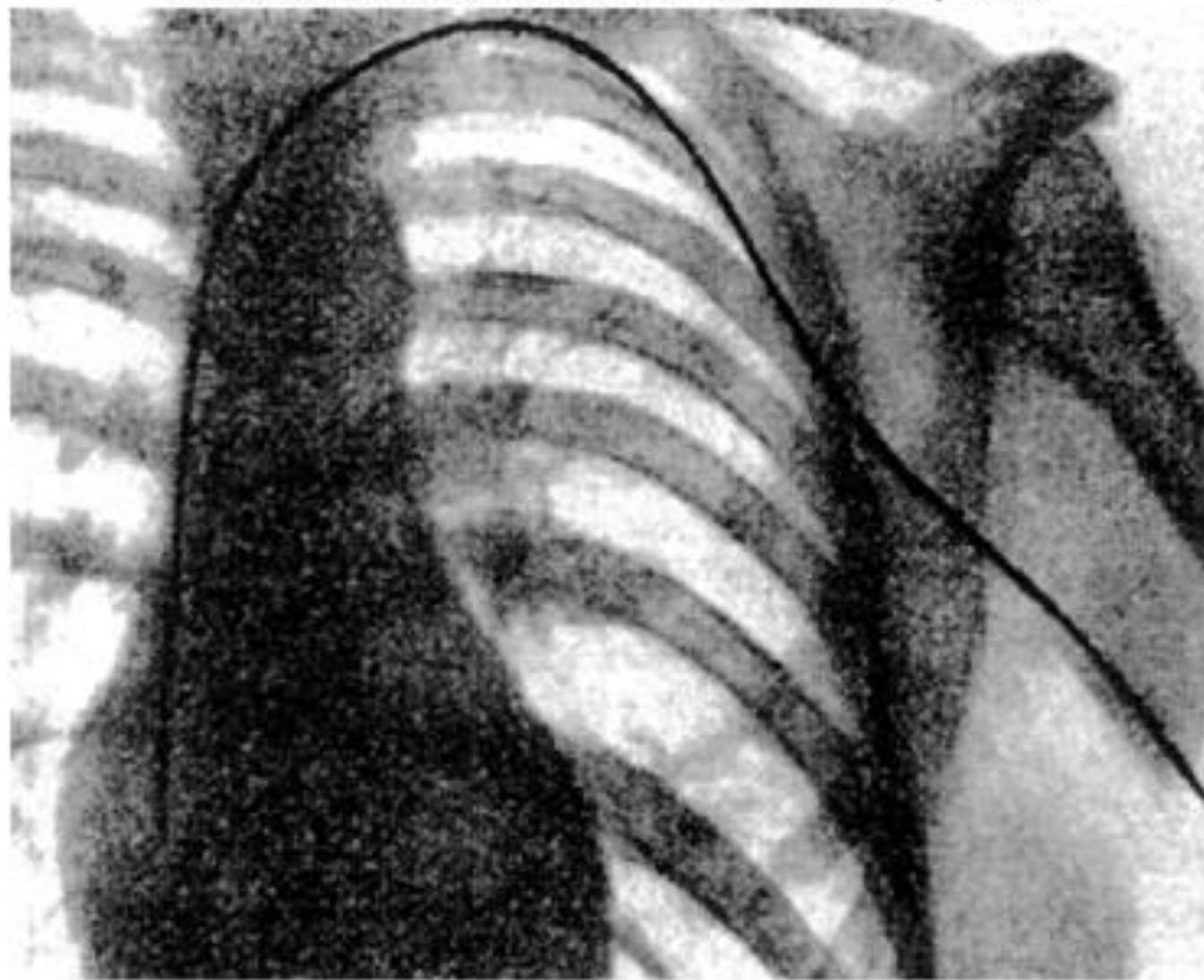
- RIGHT HEART CATH (Gold Standard)

- ***If it is suspected, but not confirmed... it could be managed incorrectly***

# History of the right-heart Catheterization

- In the 18<sup>th</sup> and 19<sup>th</sup> century, physiologists proved catheters could reach the heart in animals and measured pressure waves correlating to heart contractility/cardiac cycle
- In 1929, a young medical student (Werner Forssmann) inserted the first catheter into a human right atria
  - This was dismissed as reckless despite the success and the goal being for emergent drug delivery

FIRST CARDIAC CATHETERIZATION, 1929

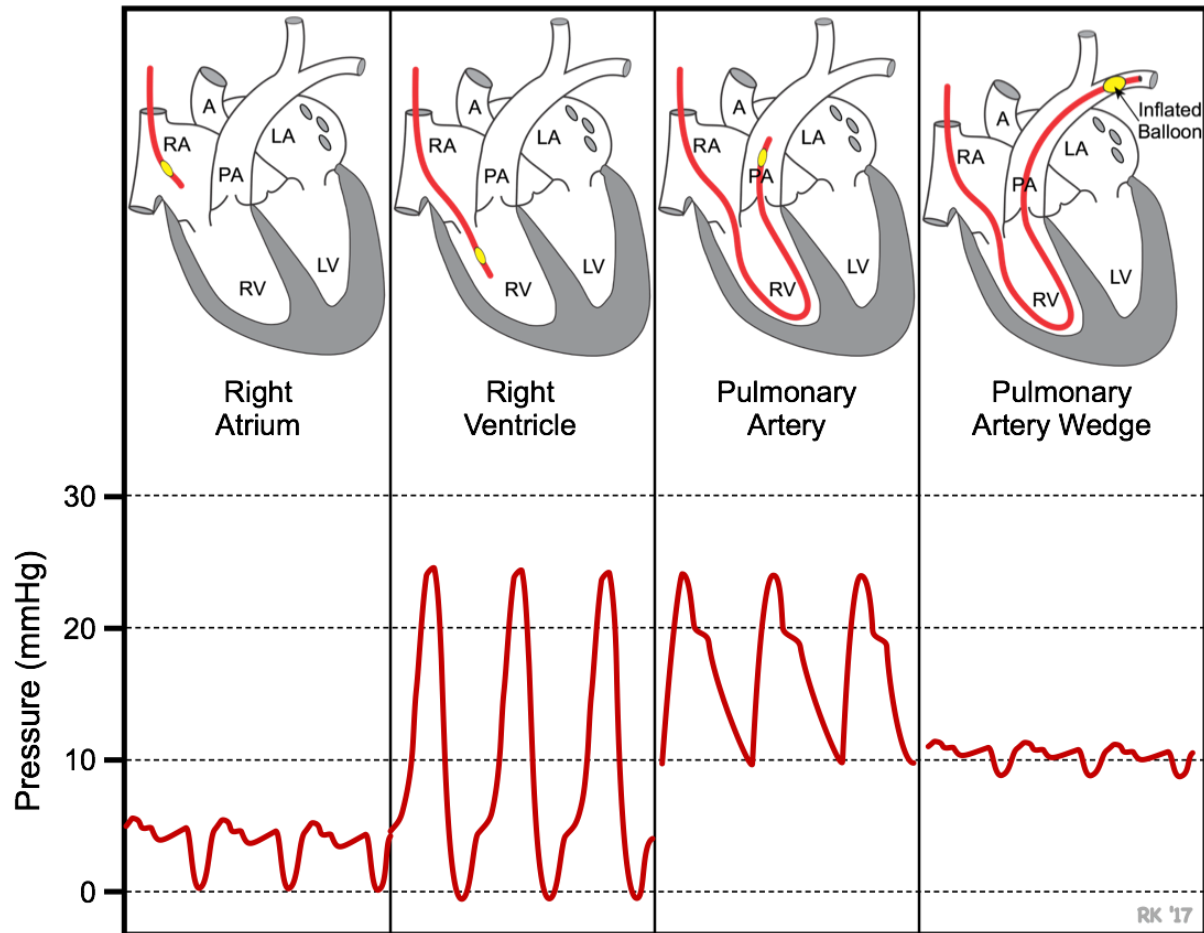


Werner Forssmann

# Modern era and lasting impact (1970 – today)

- In the 1940s, the use was refined for safe and repeat use and standardized measurements
- In the 1970s, Jeremy SWAN and William GANZ introduced the balloon-tipped, flow-directed design still used today
- This allowed repeatability, ease of use (bedside insertion, without fluoroscopy) and continuous monitoring of cardiac/pulmonic pressures and outputs
- Today, it remains essential for pulmonary hypertension, complex shock, critical care

# Right heart catheterization: The technical (Fun) part



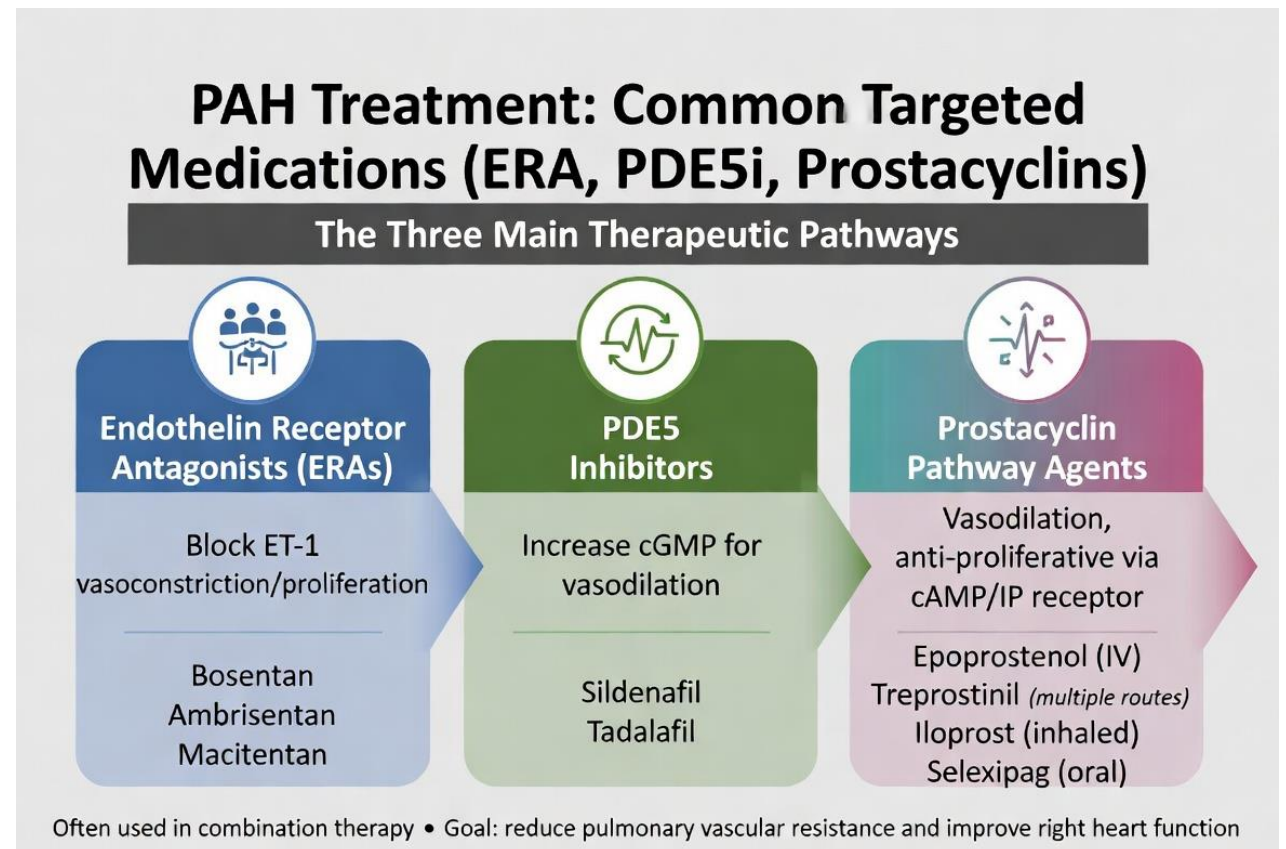
# General Management

## Things we all can do

- This is where YOU make the biggest difference
- Treat underlying causes (optimized heart failure treatment, COPD, OSA)
- Oxygen targets for sat levels >90-92%
- Utilize diuretics, salt restriction, daily weights
- Supervised exercise/cardio-pulmonary rehabilitation (HUGE evidence-based success!!!)
- Infection prevention, primary prevention of other diseases

# Targeted therapies and risk stratification

- PAH (Group 1): Endothelin-Receptor Antagonists, PDE-5 inhibitors, Prostacyclins
  - (inhaled, oral, IV)
- CTEPH (Group 4): lifelong anticoagulation + evaluation for balloon pulmonary angioplasty (BPA) or surgery
- Group 2/3 we treat the underlying disease processes!



# Pulmonary Hypertension Drugs

## Endothelin Receptor Antagonists (ERA)

Effect: vasodilation of pulmonary arteries + anti-proliferative

### Common drugs

Bosentan  
Ambrisentan  
Macitentan

## PDE5 inhibitors

Effect: Pulmonary vasodilation + mild anti-proliferative

### Common drugs:

Sildenafil  
Tadalafil

## Prostacyclin Pathway Agents

Effect: strong vasodilation, anti-platelet, anti-proliferative, and inotropic

### Common drugs:




Epoprostenol, Treprostinil, Iloprost, Selexipag

# Real world case






- 68-year-old gentleman with COPD and worsening dyspnea
  - History of heavy smoking, known COPD on 2-3L home O2
- 6 months ago: mild dyspnea, 3 months ago: worsening; now: 50-100 ft tolerance
- Echo: RVSP 65-70 mmHg, enlarged right ventricle with reduced function
- 6-minute walk test: 280 meters on 4L O2, desaturation to mid 80s
- *What would you do?*
- *Who has seen this exact patient?*

# The multi-disciplinary team

## Your major impact

- YOU catch the early decline
- YOU prevent readmission
- YOU improve outcomes
-  Nursing: medication adherence, symptom monitoring, infusion pumps (if on IV agents)
-  PT/OT: safe exercise programs, cardio-pulmonary rehabilitation, energy conservation, functional assessments
-  Respiratory Therapy: oxygen titration, non-invasive ventilation strategies, airway clearance, assistance in the cath labs
- Team communication = fewer readmissions and better survival!
- Team-based care has markedly improved 5-year survival!

# Key take-aways

-  Think PH in dyspnea
-  Echo suggests diagnosis → Cath confirms
-  RV failure = driver of mortality
-  Rehab works!!!
-  Early referral/intervention matters

Any questions?

# Sources/citations

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Thank you