Pulmonary Hypertension Diagnosis and Therapy

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37 year old female
RQ: Presented January 2007

- 37-yr-old woman, previously healthy
- Delivered second child 14 mo previously
- Limited exercise tolerance since delivery, attributed to weight gain
- Dyspnea 8/06 while playing with older child; syncope while walking up an incline
RQ: Initial Symptoms

• Currently has dyspnea with raking, walking about 1 block, walks slowly in store
• Exertional light-headedness
• Atypical chest pain
• Occasional palpitations
• Lower extremity edema
RQ: Additional History

- PMH: 2 children 4 yr and 14 mo
  - IBS: diet-controlled
- Meds: none
- All: contrast
- FH: PPH in a paternal aunt, CAD, DM, HTN
- SH: rare ETOH, o/w unremarkable
RQ: Physical Exam

- HR 90; BP 130/68; Wt 190; Ht 5'4"
- JVP ~15 cm, reduced carotid upstrokes
- Clear lungs
- Palpable RV heave, RRR, nl S, loud P₂, III/VI, TR m
- 2+ LE edema
A Disease of Decline and Deterioration: IPAH Survival if Untreated

- Poor prognosis in an era lacking therapy
- Therapeutic options and research efforts now offer more hope

PAH: Hemodynamic and Clinical Course

Adapted from Gaine S. JAMA. 2000;284:3160-3168.
PAH: Hemodynamic and Clinical Course

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Adapted from Gaine S. JAMA. 2000;284:3160-3168.
Mechanisms of Action of Therapies for PH

Clinical Classification of Pulmonary Hypertension (Dana Point)

1. PAH
   - Idiopathic PAH
   - Heritable
   - Drug- and toxin-induced
   - Persistent PH of newborn
   - Associated with:
     - CTD
     - HIV infection
     - portal hypertension
     - CHD
     - schistosomiasis
     - chronic hemolytic anemia

1’. PVOD and/or PCH

2. PH Owing to Left Heart Disease
   - Systolic dysfunction
   - Diastolic dysfunction
   - Valvular disease

3. PH Owing to Lung Diseases and/or Hypoxia
   - COPD
   - ILD
   - Other pulmonary diseases with mixed restrictive and obstructive pattern
   - Sleep-disordered breathing
   - Alveolar hypoventilation disorders
   - Chronic exposure to high altitude
   - Developmental abnormalities

4. CTEPH

5. PH With Unclear Multifactorial Mechanisms
   - Hematologic disorders
   - Systemic disorders
   - Metabolic disorders
   - Others

Is There a Reason to Suspect PAH?

Clinical Presentation

<table>
<thead>
<tr>
<th>Common Initial Symptoms (N=187)</th>
<th>Patients (%)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Dyspnea</td>
<td>60</td>
</tr>
<tr>
<td>Fatigue</td>
<td>19</td>
</tr>
<tr>
<td>Syncope or near syncope</td>
<td>13</td>
</tr>
<tr>
<td>Chest pain</td>
<td>7</td>
</tr>
<tr>
<td>Palpitations</td>
<td>5</td>
</tr>
<tr>
<td>Leg edema</td>
<td>3</td>
</tr>
</tbody>
</table>

Is There a Reason to Suspect PAH?

*Physical Exam*

**Presence of PH**
- Loud P2
- RV lift
- Systolic murmur (TR)
- Diastolic murmur (PR)
- RV S4

**Presence of RV Failure**
- JVD with V wave
- RV S3
- Hepatomegaly
- Edema
- Ascites
Is There a Reason to Suspect PAH?

**Risk Factors**

- Family history
- Connective tissue disease
- Congenital heart disease
- Portal hypertension—OLT candidate
- Environmental/drug factors
- HIV
Normal sinus rhythm
Incomplete right bundle branch block
Right ventricular hypertrophy
Prolonged QT
Abnormal ECG
RQ: Labs

- ANA-negative
- Echo: nl LV Fn, RAE, RVE, RVSP 60, TEE—no shunt
- Spiral CT: no PE
- PFTs: nl volumes and flows, D2CO 81%
- 6MWD: 222 m, 99-96%
Diagnostic Approach

- Echocardiogram
  - RVE, RAE, ↑RVSP
  - Left heart disease
  - VHD
  - CHD

- Exam
  - CXR
  - ECG

- PFTs
- Sleep study
- Sleep disorder
- Ventilation-perfusion scan, Contrast CT, Angiography

- LFTs and clinical evidence of cirrhosis and portal htn

- HIV test
- HIV
- Autoantibody tests
  - Scleroderma
  - SLE
  - RA
  - Vasculitis

- Portopulmonary hypertension
- • Functional test
  - BNP
  - RH cath
  - Vasodilator test

- Chronic thromboembolism

Echocardiogram

- Chamber size
- LV and RV systolic function
- LV diastolic function
- Valvular function
- TR
- Bubble study

Normal

Pulmonary Hypertension
### Dana Point Definition of PH/PAH

<table>
<thead>
<tr>
<th>PH</th>
<th>Mean PAP ≥25 mm Hg</th>
</tr>
</thead>
<tbody>
<tr>
<td>PAH</td>
<td>Mean PAP ≥25 mm Hg plus PCWP/LVEDP ≤15 mm Hg</td>
</tr>
</tbody>
</table>

PH: The Importance of Hemodynamics

- Pulmonary venous hypertension
  - Elevated PCWP, normal PVR

- PAH
- PH with respiratory disease
- CTEPH
  - Normal PCWP, elevated PVR

Other: High CO
## RQ: Right Heart Cath

<table>
<thead>
<tr>
<th>Metric</th>
<th>1/29/07 Baseline</th>
<th>Nitric Oxide 20 ppm</th>
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<tbody>
<tr>
<td>RAP (mm Hg)</td>
<td>19</td>
<td>20</td>
</tr>
<tr>
<td>PAP (mm Hg)</td>
<td>93/40, mean 63</td>
<td>93/46, mean 64</td>
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<tr>
<td>Left ventricular EDP (mm Hg)</td>
<td>10</td>
<td></td>
</tr>
<tr>
<td>Oxygen saturation (%)</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Pulmonary artery</td>
<td>52.9</td>
<td>58.3</td>
</tr>
<tr>
<td>Femoral artery</td>
<td>91.4</td>
<td>91.7</td>
</tr>
<tr>
<td>Cardiac output / Cardiac index (L/min) Fick</td>
<td>2.5/1.3</td>
<td>2.88/1.52</td>
</tr>
<tr>
<td>PVR (Wood units) Fick</td>
<td>21.2</td>
<td>15.2</td>
</tr>
</tbody>
</table>
Importance of Right Heart Cath

- PH
- PAH (Group 1)
- Hypoxic/Lung CTEPH
- PVH
- PAWP
- LVEDP
- LAP
- LH Disease PV Obstruction

- PVR
- TPG
- CO
- Fever
- Thyrotoxicosis
- Anemia
- Pregnancy
- Some PoPH
Cardiac Catheterization

- Exclude congenital heart disease
- Measure wedge pressure or LVEDP
- Establish severity and prognosis
- Test vasodilator therapy

Catheterization is required when pulmonary hypertension is suspected.
PH by Echo ≠ PAH

- Single echo lab/Australian community of 160,000
- Etiology of PH noted on echocardiogram

N=483 of 4579 patients with echo PASP >40 mm Hg.
Gabbay E. *Am J Respir Crit Care Med.* 2007;175:A713.
Summary

- High index of suspicion
- Thorough diagnostic evaluation
- Exclude thromboembolic disease
- Evaluate potential causes/contributing issues
- Right heart catheterization required prior to initiating PAH therapy
- Baseline functional evaluation
RQ: Initial Management

• Admitted to U of M following cath
• IV diuresis
• IV epoprostenol initiation
RQ: Return Visit May 2007

• Significantly improved
• No limitations
• Functional Class I
• Meds
  – epoprostenol 30ng/kg/min
  – warfarin
  – furosemide 20 mg
  – KCL 10 mEq qd
RQ: Follow-up Physical Exam

- HR 80; BP 103/59; Wt 144.8 lb
- JVP 6, carotid upstrokes nl
- Clear lungs
- Palpable RV heave, nl S, loud P$_2$, II/VI TR murmur
- No LE edema
RQ: 6MWD

- 222 m: 99-96% in January 2007
- 486 m: 99-97% in May 2007
RQ: Return Visit September 2007

• Continues to do well
• No limitations
• Functional Class I
• Meds
  – epoprostenol 39 ng/kg/min
  – warfarin
  – furosemide 20 mg MWF
  – KCL 10 mEq qd MWF
RQ: Physical Exam

- HR 84; BP 108/67; Wt 140.4 lb
- JVP 6, carotid upstrokes nl
- Clear lungs
- Palpable RV heave, nl S, loud P2, II/VI TR murmur
- No LE edema
RQ: 6MWD

• 222 m: 99-96% in January
• 486 m: 99-97% in May
• 556 m: 99-97% in September
## RQ: Subsequent Right Heart Cath

<table>
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<tr>
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<th>1/29/07 Baseline</th>
<th>1/7/08 Epo 38 ng/kg/min</th>
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<td>2</td>
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<tr>
<td>PAP (mm Hg)</td>
<td>93/40, mean 63</td>
<td>65/24, mean 40</td>
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<tr>
<td>PCWP (mm Hg)</td>
<td>10</td>
<td>7</td>
</tr>
<tr>
<td>Oxygen saturation (%)</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Pulmonary artery</td>
<td>52.9</td>
<td>76.2</td>
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<tr>
<td>Femoral artery</td>
<td>91.4</td>
<td>97</td>
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<tr>
<td>Cardiac Output / Cardiac Index (L/min) Fick</td>
<td>2.5/1.3</td>
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<td>PVR (Wood Units) Fick</td>
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<tr>
<td>6MWD (m)</td>
<td>222</td>
<td>602</td>
</tr>
<tr>
<td>Functional class</td>
<td>IV</td>
<td>I</td>
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</table>
PAH Treatment Goals

- Fewer/less severe symptoms
- Improved exercise capacity
- Improved hemodynamics
- Prevention of clinical worsening
- Improved quality of life
- Improved survival
What Is the Optimal Treatment Strategy?

Anticoagulate ± Diuretics ± Oxygen ± Digoxin

Acute Vasoreactivity Testing

Positive

Oral CCB

Sustained Response

No

Lower Risk
- No
- Gradual
- II, III
- Longer (>400 m)
- Peak VO$_2$ >10.4 mL/kg/min

Continue CCB

Yes

Determinants of Risk
- Clinical evidence of RV failure
- Progression of symptoms
- WHO class
- 6MWD
- CPET

Higher Risk
- Yes
- Rapid
- IV
- Shorter (<300 m)
- Peak VO$_2$ <10.4 mL/kg/min

Pericardial effusion, significant RV enlargement/dysfunction; RA enlargement

Hemodynamics
- RAP >20 mm Hg;
- CI <2.0 L/min/m$^2$

Significantly elevated

What Is the Optimal Treatment Strategy?

Anticoagulate ± Diuretics ± Oxygen ± Digoxin

Acute Vasoreactivity Testing

Positive

Oral CCB

Sustained Response

No

Yes

LOWER RISK
- No
- Gradual
- II, III
- Longer (>400 m)
- Peak VO₂ >10.4 mL/kg/min

DETERMINANTS OF RISK
- Clinical evidence of RV failure
- Progression of symptoms
- WHO class
- 6MWD
- CPET

Minimal RV dysfunction

Echocardiography

HIGHER RISK
- Yes
- Rapid
- IV
- Shorter (<300 m)
- Peak VO₂ <10.4 mL/kg/min

Pericardial effusion, significant RV enlargement/dysfunction; RA enlargement

RAP <20 mm Hg; CI <2.0 L/min/m²

Significantly elevated

RAP <10 mm Hg; CI >2.5 L/min/m²

Minimally elevated

Survival in IPAH

Long-term CCB Responders

- Long-term CCB responders (~50% of acute responders or ≤6% of IPAH patients)
- Long-term CCB failure

Survival,

\[ p = 0.0007 \]

Subjects at risk, n

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<th>4</th>
<th>6</th>
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<td>1</td>
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<tr>
<td>19</td>
<td>12</td>
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<td>4</td>
<td>0</td>
<td></td>
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</table>

Calcium Channel Blockers Only If “Vasodilator Responsive”

“Vasodilator Response”

- Fall in mPAP $\geq 10$ mm Hg
- $+\ PAP_m$ (absolute) $<40$ mm Hg
- $+\$ Normal CO

What Is the Optimal Treatment Strategy?

Anticoagulate ± Diuretics ± Oxygen ± Digoxin

Positive

Acute Vasoreactivity Testing

Negative

Oral CCB

Sustained Response

Yes

No

Continue CCB

LOWER RISK

No

Gradual

II, III

Longer (>400 m)

Peak VO₂ >10.4 mL/kg/min

Minimal RV dysfunction

RAP <10 mm Hg; CI >2.5 L/min/m²

Minimally elevated

DETERMINANTS OF RISK

Clinical evidence of RV failure

Progression of symptoms

WHO class

6MWD

CPET

Echocardiography

Hemodynamics

BNP

HIGHER RISK

Yes

Rapid

IV

Shorter (<300 m)

Peak VO₂ <10.4 mL/kg/min

Pericardial effusion, significant RV enlargement/dysfunction; RA enlargement

RAP >20 mm Hg; CI <2.0 L/min/m²

Significantly elevated

Approved Therapeutic Targets

Endothelin Pathway
- Pre-proendothelin → Proendothelin
- Endothelin-1
- Endothelin receptor A
- Endothelin receptor B
- Vasoconstriction and proliferation
- Endothelin receptor antagonists

Nitric Oxide Pathway
- L-arginine → L-citrulline
- Nitric Oxide
- Phosphodiesterase type 5
- cGMP
- Exogenous nitric oxide
- Phosphodiesterase type 5 inhibitor

Prostacyclin Pathway
- Arachidonic acid → Prostaglandin I₂
- Prostacyclin (prostaglandin I₂)
- cAMP
- Prostacyclin derivatives

Prostacyclin Analogues: Intravenous, Subcutaneous, or Inhaled

- Epoprostenol (Flolan®)
- Treprostinil (Remodulin®)
- Iloprost (Ventavis®)
- Treprostinil (Tyvaso®)
Survival Among Patients With IPAH: Epoprostenol vs Conventional Therapy

Week

Epoprostenol (n=41)

Conventional therapy (n=40)

Survival (%)

0 2 4 6 8 10 12

p=0.003*

*Two-sided, by log-rank test.
Prostanoid Side Effects

- Flushing
- Headache
- Diarrhea, nausea, vomiting
- Jaw pain
- Leg pain
- Hypotension
- Dizziness
- Syncope
- Cough (inhaled)
- Delivery site complications

Vary according to drug and route of delivery
Approved Therapeutic Targets

**Bosentan**: 6-MWD (351 and BREATHE-1)

*Tracleer®*

**p<0.05 vs baseline; p=0.021 vs placebo. Values are mean±SEM.


Ambrisentan* in PAH: 6MWD (ARIES)

ARIES-1

- 10 mg ambrisentan
- 5 mg ambrisentan
- Placebo

ARIES-2

- 5 mg ambrisentan
- 2.5 mg ambrisentan
- Placebo

*p-values are vs placebo.
Endothelin Receptor Antagonists: Side Effects

- Nasal congestion
- Abnormal hepatic function
  - reversible transaminase elevations >3X ULN
  - may require dose adjustments or discontinuations
  - monthly LFTs required
- Edema
  - lower extremity edema may require diuretic adjustment
- Use requires dual contraceptive methods (hormonal plus barrier)
Approved Therapeutic Targets

**Endothelin Pathway**
- Pre-proendothelin → Proendothelin
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- cGMP
- Exogenous nitric oxide
- Phosphodiesterase type 5 inhibitor

**Prostacyclin Pathway**
- Arachidonic acid → Prostaglandin I₂
- Prostacyclin (prostaglandin I₂)
- Prostacyclin derivatives
- Vasodilation and antiproliferation
- Smooth muscle cells

Effect of Sildenafil* on 6MWD (SUPER)

*Revatio®

Change in 6MWD (m)

-20 -10 10 20 30 40 50 60 70

Week

Placebo
20 mg of sildenafil
40 mg of sildenafil
80 mg of sildenafil

$p<0.001$

*Revatio®
Effect of Tadalafil* on 6MWD (PHIRST)

*Adcirca®
PDE-5 Side Effects

• Nose bleed
• Headache
• Dyspepsia
• Flushing
• Diarrhea
• Visual changes
• Contraindicated with use of nitrates
Treatment of PAH: Evidence-based Approach

- How is initial treatment chosen?
- When and how is treatment response assessed?
- When should treatments be changed or combined?
Combination Therapy

Prostanoids

Endothelin Receptor Antagonists

Phosphodiesterase Inhibitors
## Combination Therapy: Other Ongoing or Recently Completed Clinical Trials

<table>
<thead>
<tr>
<th>Study</th>
<th>Current therapy</th>
<th>Added therapy</th>
<th>Patients (n)</th>
<th>Study duration</th>
<th>Primary end point</th>
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</thead>
<tbody>
<tr>
<td>FREEDOM-C</td>
<td>Bosentan and/ or sildenafil</td>
<td>Treprostinil oral</td>
<td>300</td>
<td>16 weeks</td>
<td>6MWD</td>
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<tr>
<td>AMBITION</td>
<td>Ambrisentan/ tadalafil/combo</td>
<td>Combo vs mono</td>
<td>300</td>
<td>Event-driven</td>
<td>Morbidity/mortality event</td>
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<tr>
<td>Pfizer</td>
<td>Bosentan</td>
<td>Sildenafil</td>
<td>106</td>
<td>12 weeks</td>
<td>6MWD</td>
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<td>COMPASS-1</td>
<td>Bosentan</td>
<td>Sildenafil</td>
<td>45</td>
<td>Single dose</td>
<td>PVR</td>
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<td>COMPASS-2</td>
<td>Sildenafil</td>
<td>Bosentan</td>
<td>250</td>
<td>Event-driven</td>
<td>Morbidity/mortality event</td>
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<tr>
<td>COMPASS-3</td>
<td>Bosentan</td>
<td>Sildenafil</td>
<td>100</td>
<td>16 weeks</td>
<td>6MWD</td>
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<td>ATHENA-1</td>
<td>Sildenafil or tadalafil</td>
<td>Ambrisentan</td>
<td>40</td>
<td>24 weeks</td>
<td>PVR</td>
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<td>SERAPHIN</td>
<td>Naïve/PDE-5/PGI/combo</td>
<td>Macitentan</td>
<td>742</td>
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<tr>
<td>PATENT</td>
<td>Naïve/PGI/ERA</td>
<td>Riociguat</td>
<td>462</td>
<td>12 weeks</td>
<td>6MWD</td>
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<tr>
<td>IMPRES</td>
<td>≥2 current therapies</td>
<td>Imatinib</td>
<td>200</td>
<td>24 weeks</td>
<td>6MWD</td>
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<td>ATPAHSS</td>
<td>Ambrisentan/ tadalafil/combo</td>
<td>Combo vs mono</td>
<td>63</td>
<td>36 weeks</td>
<td>RV mass/PVR</td>
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<td>GRIPHON</td>
<td>ERA, PDE5 or both</td>
<td>Selexipag</td>
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<td>Novartis</td>
<td>Stable PAH therapy</td>
<td>Noilotinib</td>
<td>66</td>
<td>6 months</td>
<td>PVR</td>
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</table>
61-year-old Female
Presentation

• 61-yr-old female with a 3-yr history of progressively worsening dyspnea was referred because of PH noted on an echocardiogram

• She denied CP, LH, syncope, and LE edema

• Her symptoms had progressed to functional class III
  – she denied symptoms c/w OSA

• PMH: systemic hypertension and diabetes, 3 uncomplicated deliveries

• Current medications were verapamil 240 mg bid and atenolol 25 mg qd

• SH: no tobacco, alcohol, or illicits

• FH: noncontributory
Physical Exam

- Height: 5' 4"
- Weight: 180 lb
- BP: 150/90 mm Hg; normal JVP; normal carotid pulse
- Clear lungs
- Palpable RV tap, loud S2, RS4, II/VI TRM, no LE edema
Test Results

• ECG: NSR, normal axis
• V/Q: normal
• Echo: normal LV systolic function, normal RV size, ↑ PAP
• PFTs: slight restriction
• ANA and HIV-negative
What Is the Next Step?

A. Initiate therapy with an ERA
B. Initiate therapy with a PDE-5 inhibitor
C. Right heart catheterization
D. Repeat echo in 6 months
What Is Highest on Your Differential?

- Idiopathic PAH
- Diastolic dysfunction
- Chronic thromboembolic PH
- An ASD
## Test Results

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<tr>
<th></th>
<th>Baseline</th>
<th>Nipride (2.0)</th>
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<tbody>
<tr>
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<td>80/28</td>
<td>44/15</td>
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<tr>
<td>Ao</td>
<td>205/95</td>
<td>165/72</td>
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<tr>
<td>RA</td>
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<tr>
<td>PCWP</td>
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<td>7</td>
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<td>LVEDP</td>
<td>29</td>
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<td>CO</td>
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<td>3.03</td>
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<td>Ao Sat</td>
<td>95</td>
<td>92</td>
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<td>PA Sat</td>
<td>68</td>
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<tr>
<td>PVR</td>
<td>5.14</td>
<td>3.11</td>
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</table>
LVH/Diastolic Dysfunction

RCM, 4cm, 922, 25
LVH/Diastolic Dysfunction
Mitral Inflow: Grade 3 Diastolic Dysfunction
Follow-up

- Patient was referred back to primary MD with recommendations for BP control and fluid management
- Patient remained dyspneic and primary MD initiated therapy with IV epoprostenol
Hypertensive Heart Disease

• What are clinical clues that etiology of PH is on left side of heart?

• What is “pulmonary hypertension out of proportion to left heart disease”?

• How do you treat it? What if PA pressures are still high despite optimal treatment of BP?

• Is echo adequate for assessing left heart function?

• Is wedge pressure an adequate measure of left heart function?
50-year-old Caucasian Male
Presentation

• 50-yr-old white male with past medical history of hypertension presents with shortness of breath x 3 yr, worsening; outside echo: PH

• Patient had episode of “pneumonia” a few years ago and “never seemed to get over it”

• NYHA Class III

• No history of heart disease, DVT/PE, family history, drug use, CTD, sleep apnea Sx, or lung disease
Physical Examination

- BP 110/68; HR 86; RR 18; O₂ sat 94% on room air
- No JVD
- Lungs: no rales or wheezes
- Heart: loud P2, II/VI syst murmur LLSB
- 1-2+ edema, L > R
Initial Testing

• Echo: moderate RA/RV enlargement, PA syst 64 + CVP, neg bubble, normal LV size/function
• PFTs: mild restriction, DLCO 56% predicted
• CXR: cardiomegaly, no infiltrates
What Next?

V/Q vs CT
Teaching Point
CT Scan in CTEPH

- May be useful in confirming diagnosis
- More useful in ruling out other processes
- Mosaic pattern of perfusion useful finding
CTEPH: Mosaic Perfusion
Differential Diagnosis of CTEPH: What Is It?
Pulmonary Artery Sarcoma
Differential Diagnosis of CTEPH: What Is It?

Calcifications

Fibrosing mediastinitis
Back to our patient...
Next Step?

Right heart cath
PA GRAM

RA 9, PA 86/48(61), PCW 5,
CO 4.6, PVR 12
Zone of accessibility
Case: Post-op

mPAP: 26
PVR: 3
Questions

1. What physical exam finding might have been useful if present?
   *Pulmonary flow murmur*

2. Does the absence of a history of DVT/PE help?
   *No*