Presentation: Demystifying Pulmonary Hypertension
Speaker: Barry Cabuay, MD
Minneapolis Heart Institute® at Abbott Northwestern Hospital
Date: Monday, October 12, 2015, 7:00 – 8:00 AM
Location: ANW Education Building, Watson Room

OBJECTIVES
At the completion of this activity, the participants should be able to:
1. Review the basic definition and contemporary classification system of pulmonary hypertension (PH) encompassing the various etiologies.
2. Discuss the comprehensive diagnostic approach based on physiologic principles and the importance of particular findings from key diagnostics such as echocardiogram and right heart catheterization.
3. Recognize the target pathways for therapy and management strategies.

ACCREDITATION
Physicians: This activity has been planned and implemented in accordance with the Essential Areas and policies of the Accreditation Council for Continuing Medical Education (ACCME) through the joint sponsorship of Allina Health and Minneapolis Heart Institute Foundation. Allina Health is accredited by the ACCME to provide continuing medical education for physicians.

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Nurses: This activity has been designed to meet the Minnesota Board of Nursing continuing education requirements for 1.2 hours of credit. However, the nurse is responsible for determining whether this activity meets the requirements for acceptable continuing education.

Others: Individuals representing other professional disciplines may submit course materials to their respective professional associations for 1.0 hours of continuing education credit.

DISCLOSURE STATEMENTS
Speaker: Dr. Cabuay has declared he does not have any conflicts of interest to disclose.

Planning Committee: Dr. Michael Miedema, Eva Zewdie and Jolene Bell Makowesky have declared that they do not have any conflicts of interest associated with the planning of this activity. Dr. Robert Schwartz declared the following relationships - stockholder: Cardiomind, Interface Biologics, Aritech, DSI/Transoma, InstyMeds, Intervalve, Medtronic, Osprey Medical, Stout Medical, Tricardia LLC, CoAptus Inc, Augustine Biomedical; scientific advisory board: Abbott Laboratories, Boston Scientific, MEDRAD Inc, Thomas, McNerney & Partners, Cardiomind, Interface Biologics; options: BackBeat Medical, BioHeart, CHF Solutions; speakers bureau: Vital Images; consultant: Edwards LifeSciences.
Objectives

- Review the basic definition and contemporary classification system of PH encompassing the various etiologies
- Discuss the comprehensive diagnostic approach based on physiologic principles and the importance of particular findings from key diagnostics such as echocardiogram and right heart catheterization
- Understand the target pathways for therapy and management strategies, especially discerning PAH vs PVH
Case 1: Does this pt have PAH?

25yo WF, previously athletic, nonsmoker
3mo h/o progressive DOE, abdominal bloating, fatigue
PEX: Normal BP, 10cm JVP@90°, prominent S2, RV heave, abdominal fullness, no LEE

- Enlarged RA and RV, mild TR
- estimated PA pressure of 80mm Hg
- Sent PH Clinic for evaluation
What is Pulmonary Arterial Hypertension?

**PAH definition:**
- mPAP ≥25 mmHg at rest
- PCWP ≤ 15
- PVR > 3

_Hoepfer M. JACC. 2013;62:D42-D50._

**Epidemiology**

- Rare, increasing recognition
- PAH in multicenter registries (France, USA)
  - prevalence 10.6-15.0/million adults
  - incidence 2-2.4/million adults/yr
  - mean age 36 yrs
  - Female:Male = 2:1 - 4:1


_Ann Intern Med 1987; 107:216-223._
Prognosis of iPAH is Poor

![Graph showing survival rates over years of follow-up.](image)

- Median survival: 2.8 years
- Survival rates:
  - 0 years: 100%
  - 0.5 years: 68%
  - 1 year: 48%
  - 2 years: 34%


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WHO Classification, 5 Groups

Nice, 2013

1. Pulmonary arterial hypertension (PAH)
2. PH secondary to left heart disease
3. PH secondary to lung diseases and/or hypoxia
4. PH secondary to chronic thromboembolism (CTEPH)
5. PH with unclear multifactorial mechanisms

Simonneau G. JACC 2013
Group 1 PAH

1.1. Idiopathic PAH

1.2. Heritable
   1.2.1. BMPR2
   1.2.2. ALK1, ENG, SMAD9, CAV1KCNK3
   1.2.3. Unknown

1.3. Drug and toxin-induced

1.4. Associated with
   1.4.1. Connective tissue diseases (systemic sclerosis, SLE, Mixed CTD)
   1.4.2. HIV infection
   1.4.3. Portal hypertension
   1.4.4. Congenital heart disease (PVR<2.3 fix, >4.6 not correctable)
   1.4.5. Schistosomiasis
     • 1’ Pulmonary veno-occlusive disease (PVOD) and/or pulmonary capillary hemangiomatosis (PCH)
     • 1” Persistent pulmonary hypertension of the newborn (PPHN)
Group 2
Left heart disease-MCC

2.1. Systolic dysfunction
2.2. Diastolic dysfunction
2.3. Valvular disease
2.4. Congenital/acquired left heart inflow/outflow tract obstruction and congenital cardiomyopathies

Group 3
Lung disease and/or hypoxia

3.1. Chronic obstructive pulmonary disease
3.2. Interstitial lung disease
3.3. Other pulmonary diseases with mixed restrictive and obstructive pattern
3.4. Sleep-disordered breathing
3.5. Alveolar hypoventilation disorders
3.6. Chronic exposure to high altitude
3.7. Developmental abnormalities, musculoskeletal
Group 4
Chronic thromboembolism

4. Chronic thromboembolic pulmonary hypertension (CTEPH)

Group 5
Miscellaneous/Multifactorial causes

5.1. Hematologic disorders: chronic hemolytic anemia, myeloproliferative disorders, splenectomy

5.2. Systemic disorders: sarcoidosis, pulmonary Langerhans cell histiocytosis, lymphangioleiomyomatosis,

5.3. Metabolic disorders: glycogen storage disease, Gaucher disease, thyroid disorders

5.4. Others: tumoral obstruction, fibrosing mediastinitis, chronic renal failure on dialysis
<table>
<thead>
<tr>
<th>Symptom</th>
<th>Initial</th>
<th>Eventual</th>
</tr>
</thead>
<tbody>
<tr>
<td>Dyspnea</td>
<td>60</td>
<td>98</td>
</tr>
<tr>
<td>Fatigue</td>
<td>19</td>
<td>73</td>
</tr>
<tr>
<td>Chest pain</td>
<td>7</td>
<td>47</td>
</tr>
<tr>
<td>Near syncope</td>
<td>5</td>
<td>41</td>
</tr>
<tr>
<td>Syncope</td>
<td>8</td>
<td>36</td>
</tr>
<tr>
<td>Edema</td>
<td>3</td>
<td>37</td>
</tr>
</tbody>
</table>
WHO Functional Class of PAH

<table>
<thead>
<tr>
<th>Class</th>
<th>Description</th>
</tr>
</thead>
<tbody>
<tr>
<td>I</td>
<td><em>No limitation</em> of physical activity.</td>
</tr>
<tr>
<td>II</td>
<td><em>Slight limitation</em> of physical activity. Ordinary physical activity causes undue dyspnea or fatigue, chest pain, or near syncope.</td>
</tr>
<tr>
<td>III</td>
<td><em>Marked limitation</em> of physical activity. Less than ordinary activity causes undue dyspnea or fatigue, chest pain, or near syncope.</td>
</tr>
<tr>
<td>IV</td>
<td>Symptoms with <em>any physical activity</em>. These patients manifest signs of right-heart failure or <em>syncope</em>. Dyspnea and/or fatigue may even be <em>present at rest</em>.</td>
</tr>
</tbody>
</table>

Rubin LJ. Chest. 2004;2004:7S-10S.

Diagnostic Approach

- H&P not sensitive or specific
- CXR
- ECG
- PFTs
- V/Q scan, CT chest, Pulm Angio
- Echocardiogram
- Right heart catheterization
- 6MWT
- BNP
- Ancillary testing: TSH, LFTs, ANA, HIV, Drug tox
Signs of PAH on Chest X-ray

- Prominent proximal PA
- "Pruning" of distal PA
- Loss of retrosternal airspace
- Cardiac enlargement, right-sided
- No evidence of pulmonary edema (sign of left heart disease)
- Lungs look normal


Right Ventricular Hypertrophy

Right Atrial Enlargement  RV strain
Pulmonary Function tests

- Mandatory to screen for significant restrictive or obstructive lung disease
  - FEV1 <60% predicted (COPD)
  - FVC <70% predicted (IPF)
- Diffusing capacity often significantly reduced in patients with scleroderma (<50%)

Seeger W. JACC 2013. D112

Evaluation for Chronic Thromboembolic PH (CTEPH)

- V/Q scan remains the best screening tool
- CT angiogram is excellent for acute, proximal PE but can miss CTEPH
- Gold standard for diagnosis is pulmonary angiogram
- Safe procedure even in patients with marked PH
  - Mortality in 2 large studies: 0/547 and 2/202*


*Hofman LV, et al, AJR, 2004
Pulmonary Endarterectomy

Signs of PAH on Echocardiogram With Doppler

- Increased sPAP or TR jet
- RA and RV hypertrophy
- Flattening of IVS
- Small LV dimension
- Dilated PA
- Pericardial Effusion
- *Bubble Study

Echocardiography in PAH

Synt PAH= Right Ventricular Systolic Pressure (in absence of pulmonary outflow obstruction)

RVSP= 4x LA + RAP*
Echocardiogram Pitfalls

Underestimation of PAP
- suboptimal TR jet-cursor alignment
- weak TR jet

Overestimation of PAP
- artifactually enhanced TR jet
- TV closure motion
- unrecognized RVOT obstruction or PS
- overestimation of RAP

Incorrect interpretation

PH by Echo ≠ PAH.
PASP ≠ mPA

Lung disease 5%
CTEPH 0.6%
PAH 2.3%
Sleep-related hypoventilation 9.7%
Congenital heart disease 1.9%
Unknown 6.8%
Left heart disease 78.7%

Single echo lab / Australian community of 160,000
- N=483 of 4579 patients with echo PASP >40 mm Hg.
- ~10% of patients had est. sPAP>40 mm Hg
- Only 2.3% with PAH after full evaluation

Gabby E. Am J Respir Crit Care Med. 2007;175:A713.

Right Heart Catheterization

- Establishes the diagnosis of PH
- Discriminates PAH from PVH
- Vasodilator testing
- Gold standard

PAH definition:
- mPAP ≥25 mmHg at rest
- PCWP ≤ 15
- PVR >3 WUs
Acute Vasodilator Testing in Pulmonary Arterial Hypertension

- To determine if treatment with a CCB is warranted\(^1\)
- Only ~ 10% to 20% of patients will benefit from long-term CCB therapy\(^2,3\)
- Patients who have a positive long-term response to CCBs have a favorable survival prognosis
- Unfortunately, ~ 10% of patients will have a long-term response
- A negative VDC does not signify a “nonresponder” to PAH therapies

\(^{CCB} = \text{Calcium channel blocker, IV = Intravenous.}\)


Acute Vasodilator Testing

- \(\text{NO at 40 ppm + 100% FIO2 x 5min}\)
  - Adenosine, Epoprostenol, sildenafil
  - Nipride, NTG

- Positive response
  - \(mPAP\) decreased by \(\geq 10\ \text{mmHg}\)
  - Absolute level of \(mPAP\) < 40 mmHg
  - No decrease in cardiac output or increase in PCWP
Correlation of Baseline 6MWT With Survival in iPAH

- <332 m: 20% 3-year survival
- >332 m: 92% 3-year survival

Plasma BNP as a Prognostic Indicator of Mortality in Patients With iPAH

By multivariate analysis, higher BNP at baseline (RR=11.971, P=0.0348) and at follow-up (RR = 25.880, P=0.0243) were independent predictors of mortality.
Echo and CMR Evaluation of the RV in PH

A. RV fractional area change
B. TAPSE
C. RV S’ velocity
D. RV Strain
E. CMR late gadolinium
F. CMR LV ‘D’ w Inspiration

JACC. 2015;65 p1985
Phosphodiesterase-5 Inhibitors

• Sildenafil
  - improves morbidity
• Tadalafil
  - improves morbidity

Phosphodiesterase-5 Inhibitors Side Effects

• Headache
• Hypotension
• Flushing
• Visual changes
• Epistaxis
• Rarely
  - priapism
  - visual loss
Soluble Guanylate Cyclase Stimulant

• **Riociguat**
  - novel medication class with dual mode of action
    1. Increases sGC sensitivity to NO
    2. Directly stimulates the receptor to mimic the action of NO
  - improves morbidity
  - approved for CTEPH
  - hypotension, dizziness, headache, anemia, teratogenic

Endothelin-Receptor Antagonists

• **Bosentan**
  - improves morbidity

• **Ambrisentan**
  - improves morbidity

• **Macitentan**
  - improves morbidity
  - Improves mortality as TTCW
  - Duration to 100wks
Endothelin-Receptor Antagonists

Side Effects

• Anemia
• Edema
• Rhinitis
• Teratogenic
• Testicular atrophy
• Abnormal LFTs (Bosentan)

Prostanoids

• Epoprostenol
  - improves morbidity and mortality
  - continuous IV infusion, inhalation
  - instability at room temperature
• Treprostinil
  - improves morbidity
  - longer T1/2 More stable
  - IV, SC, inhalation, and PO
• Iloprost
  - improves morbidity
  - inhalation
Prostanoids
Side Effects

- Vasodilation/flushing
- Jaw pain
- Headache
- Hypotension
- Cough
- Nausea

Conventional Therapy

<table>
<thead>
<tr>
<th>Therapy</th>
<th>Use</th>
</tr>
</thead>
<tbody>
<tr>
<td>Oxygen</td>
<td>Goal &gt;90% sats</td>
</tr>
<tr>
<td>Diuretics</td>
<td>to maintain near-normal intravascular volume</td>
</tr>
<tr>
<td>Anticoagulants</td>
<td>For in situ microscopic thrombosis in patients with IPAH: Goal INR = 1.5 - 2</td>
</tr>
<tr>
<td></td>
<td>For patients at increased risk of pulmonary thromboembolism due to right ventricular failure and venous stasis</td>
</tr>
<tr>
<td>Digoxin (digitalis)</td>
<td>refractory right ventricular failure and/or atrial dysrhythmias</td>
</tr>
<tr>
<td>Contraception</td>
<td>Hi mortality rates for both patient and fetus</td>
</tr>
<tr>
<td>Patient Education</td>
<td>Daily wts, Fluid and salt restriction, avoid NSAIDS</td>
</tr>
</tbody>
</table>

## Pulmonary Specific Vasodilators

### FDA APPROVED

- **PDE-5 Inh:** po
  - Sildenafil 20mg tid
  - Tadalafil 40mg qd
- **ERAs:** po
  - Bosentan 125mg bid
  - Ambrisentan 5/10 mg qd
  - Macitentan 10 mg
- **Prostanoids**
  - Epoprostenol: IV
  - Treprostinil: IV, SC, inh
  - Iloprost: inh

### COSTS

- **PDE-5**
  - $12,000-$15,000/year
- **ERAs**
  - $75,000/yr
- **Prostanoids**
  - Flolan: $100k/yr
  - Treprostinil IV+SC: >$100k/yr
  - Iloprost: $170k/yr

## Hi Risk Markers

- **Syncope**
- **NYHA/WHO FC**
- **6MWD**
- **CPET**
- **ECHO**
- **RHC**
- **CMR**

- **Yes**
- **IV**
- **<330 m**
- **pVO2 <12 ml/kg/min**
- **Pericardial Effusion**
- **TAPSE <1.5 cm**
- **RAP >15**
- **CI ≤2**
- **RVEF <35%**
PAH Treatment Algorithm

Vasoreactivity testing

Positive

Oral CCB

Lower risk

WHO-FC II-III

ERAs
PDE-5i
Prostacyclins
sGCS

No sustained response

Higher risk

WHO-FC IV

Prostacyclins Combo therapy

Goal directed therapy

Atrial septostomy
Transplant PEA

Negative

Goal directed therapy

Goals of Medical Therapy for PAH:
Targeted Risk-Benefit Considerations

**BENEFIT**

- ↑ Exercise capacity
- ↓ Symptoms
- Improve hemodynamics
- Improve quality of life
- ↑ Survival

**RISKS**

- Side effects
- Risks with continuous IV/SC infusion and inhalation delivery
- Hepatotoxicity
- Drug interactions
- Social issues

Combination Therapy

- N = 500
- 2:1:1 randomization
- 1: First event of clinical failure

PH Therapy

PH Therapy Depends on Group

- WHO(1)PAH - Prostanoids, ERAs, PDE-5 inhibitors, sGCS

- WHO(2)PH with Left Heart Disease – HTN mgmt, heart failure therapy/valve repairs/CAD options/advanced surgical options.

- WHO(3)PH with Lung Diseases/Hypoxemia - O2, treat underlying disease

- WHO(4)CTEPH - anticoagulation, pulmonary endarterectomy, PAH Rx/Riociguat(inoperable/distal)

- WHO(5) Misc – treat underlying, supportive, ??PAH therapies in selected cases
Clues to causes of PH other than PAH

- Dilated LA on echo or on CT angiogram
- Hx of systemic vascular disease
- LVH, LAE, no evidence of RVH or RAD on ECG
- Orthopnea
- Crackles, clubbing
- Development of pulmonary edema with pulmonary vasodilators
  - Think of disorders affecting pulmonary veins

Cascade of PH development in HFpEF
Distinguishing WHO Group II (Left Heart Disease)

Pulmonary artery
Pulmonary vein

PAH vs PVH
Precapillary and Postcapillary

PAH
PH with respiratory disease
CTEPH
PCWP/LVEDP \( \leq 15 \) mm Hg
PVR > 3 Wood units

PVH: HFrEF, HfP EF, Valvular Mixed etiology
PCWP/LVEDP > 15 mm Hg

PAH vs PVH
Diastolic Pressure Gradient (DPAP-PCWP)

\[\begin{align*}
\text{mPAP} & \geq 25 \text{ mmHg at rest} \\
\text{PCWP} & < 15 \text{ mmHg} \\
\end{align*}\]

\[\begin{align*}
\text{mPAP} & > 25 \\
\text{PCWP/LVEDP} & > 15 \\
\text{TPG} & > 12-15 \\
\text{PVR} & > 2.5-3 \text{ WU} \\
\end{align*}\]

\[\begin{align*}
\text{mPAP} & \geq 25 \text{ mmHg at rest} \\
\text{PCWP} & < 15 \text{ mmHg} \\
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\text{mPAP} & > 25 \\
\text{PCWP/LVEDP} & > 15 \\
\text{TPG} & > 12-15 \\
\text{PVR} & > 2.5-3 \text{ WU} \\
\end{align*}\]

*: TPG = transpulmonary gradient (mPAP-PCWP)
*: DPG = diastolic pressure gradient (diastolic pulm. pressure-PCWP)

JHLT 2015;34. p276
### HFpEF

**Table 2. Clinical Characteristics of Heart Failure With a Preserved Ejection Fraction in Negative Clinical Trials to Date**

<table>
<thead>
<tr>
<th>First Author/Trial</th>
<th>Japanese-HFP</th>
<th>ELADO**</th>
<th>I-PRESERVE***</th>
<th>D-PEEP</th>
<th>ALDO-HFP</th>
<th>RAMAD-FEP**4</th>
<th>RELAX**4</th>
<th>TOPCAT**5</th>
</tr>
</thead>
<tbody>
<tr>
<td>Intervention</td>
<td>Carvedilol</td>
<td>Metoprolol</td>
<td>Isradipine</td>
<td>Digoxin</td>
<td>Spironolactone</td>
<td>Eprosartan</td>
<td>Spiranolactone</td>
<td></td>
</tr>
<tr>
<td>Sample size</td>
<td>124</td>
<td>116</td>
<td>123</td>
<td>86</td>
<td>422</td>
<td>44</td>
<td>216</td>
<td>340</td>
</tr>
<tr>
<td>Exclusion criteria</td>
<td>LVEF &gt; 40%</td>
<td>&lt; 40%</td>
<td>&lt; 40%</td>
<td>&lt; 40%</td>
<td>&lt; 40%</td>
<td>&lt; 40%</td>
<td>&lt; 40%</td>
<td></td>
</tr>
<tr>
<td></td>
<td>Diastolic dysfunction</td>
<td>NYHA class II-III</td>
<td>Deleterious NYHA class II-III</td>
<td>Established HF within past 6 mo</td>
<td>Elevated BNP</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td></td>
<td>Combination of cardiovascular symptoms and HF related hospitalization for HF</td>
<td>Clinical signs/symptoms of HF in past 6 mo</td>
<td>Elevated BNP</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
</tbody>
</table>

**Primary end point**
- Composite cardiovascular death and unplanned hospitalization for HF
- Change in eGFR

**Secondary end points**
- Composite cardiovascular death from any cause or HF mortality for HF in past 6 mo.
- Changes in diastolic function (4%) and combined HF hospitalization for cardiovascular cause.
- Change in LVEF (10%)
- Change in peak VO2

**Outcome**
- Negative
- Negative
- Negative
- Negative
- Negative
- Negative
- Negative
- Negative
- Negative
- Negative

**1-y survival (%)**
- Placebo 90%
- Placebo 90%
- Placebo 77%
- Placebo 100%
- Placebo 100%
- Placebo 90%
- Placebo 77%
- Placebo 100%
- Placebo 90%
- Placebo 77%
- Placebo 100%
- Placebo 90%
- Placebo 77%
- Placebo 100%

Circ Res. 2014; 115. p89

### HF and PH

**Table 2. Long-term, Placebo-Controlled Studies in Patients With Heart Failure and Pulmonary Hypertension**

<table>
<thead>
<tr>
<th>Study</th>
<th>Drug</th>
<th>Duration</th>
<th>Population</th>
<th>N</th>
<th>Results</th>
</tr>
</thead>
<tbody>
<tr>
<td>Lewis 2007**</td>
<td>Sildenafil (25 to 75 mg TID)</td>
<td>12 wk</td>
<td>NHA class II-IV, LVEF &lt; 40%, mPAP &lt; 25 mmHg</td>
<td>34</td>
<td>Sildenafil increased peak VO2 and cardiac output and reduced PVR with exercise; no effect on PAP, blood pressure, or heart rate; improved 6-MIN walking distance and reduced HF admissions; higher incidence of headache</td>
</tr>
<tr>
<td>Kusko 2008**</td>
<td>Bosentan (6–125 mg BID)</td>
<td>20 wk</td>
<td>NHA class II-IV, LVEF &lt; 40%, mPAP &lt; 25 mmHg</td>
<td>94</td>
<td>No difference from baseline to week 20 in mPAP (0.1 to 11.5 mmHg, P&lt;0.05) or other echocardiographic parameter; more SAEs in the bosentan arm</td>
</tr>
<tr>
<td>Guazzi 2011**</td>
<td>Sildenafil (50 mg TID)</td>
<td>1 yr</td>
<td>LVEF &lt; 40%, mPAP &lt; 25 mmHg (echocardiography)</td>
<td>44</td>
<td>Sildenafil increased mean PAP by 42.0±13.0%, improved right-ventricular function, and reduced right-atrial pressure by 54.0±7.2% and PAP by 15.7±3.1%</td>
</tr>
<tr>
<td>Guazzi 2012†</td>
<td>Sildenafil (50 mg TID)</td>
<td>1 yr</td>
<td>LVEF &lt; 40%, mean mPAP 25–35 mmHg</td>
<td>32</td>
<td>Sildenafil increased peak VO2 and exercise ventilation efficiency, and decreased PAP, mean mPAP, and pulmonary vascular resistance</td>
</tr>
</tbody>
</table>

Circ HF. 2013; 6. p584
Summary

- Pulmonary hypertension is common and multifactorial. PAH is a rare progressive disease with a guarded prognosis.
- PH can NOT be diagnosed by Echo alone. Verify that the patient has PAH before treating. A thorough evaluation for all patients is mandatory.
- Right heart catheterization is necessary in ALL patients. It establishes diagnosis, severity, and prognosis of PH.
- Specific pulmonary vasodilator treatment is indicated in WHO group-1 after a comprehensive evaluation including a RHC with a negative VDC.
- Treatment of non-PAH WHO Group patients may be neutral or harmful with rare exception. It is important to discern differences in patients with PAH/WHO 1 and WHO 2 with left heart disease.
- PH patients should be referred to a PH center to confirm diagnosis and manage treatment given the complexities of underlying disease, with the cost and side effect profile of therapies for timely evaluation and care initiation. Comanagement with local care is paramount.

Case: Is PAH therapy indicated?

68yoF
- 3mo h/o progressive DOE, lightheadedness, fatigue
- HTN, GERD, Raynaud’s, telangiectasias
- TTE: RVE, RAE, RVSP 85, diastolic dysfunction
- RHC: RA 15, PA 80/35x50, PCWP 18, CO 4.3
  - Negative VDC
- 6MWT of 180 m; BNP 540

What labs help establish the DX?
- ANA, SCL70, anticentromere abs
- DPG is 17
- Started ERA
- at 3mo, 6MW 300 m/BNP 300; at 12 mo, 6MW 450 m, BNP 180
Thank you

Case 2: Does this pt have PAH?

48yo AAF

• 3mo h/o progressive DOE, abdominal bloating, fatigue

• PEX: Obese, 10cm JVP@90°, prominent S3, RV heave, HSM along LSB, abdominal fullness, 2+ LEE

• TTE:
  – Mildly enlarged RA and RV, moderate TR,
  – LVH, normal LVSF
  – estimated PA pressure of 60 mm Hg

Dx: OSA

Sent PH clinic for evaluation