Pulmonary Hypertension: MHI Rural Experience & Guideline Update

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Disclosures

- Speakers bureau
  - Actelion
  - Bayer Pharmaceuticals
  - Gilead Sciences

“The Art of Medicine” – potential off label use of meds
Objectives

- Recognize when to suspect PH
- Review the classification/etiologies of PH
- Recognize common medications used to treat PAH (Group 1 PH)

“The Heart & Vascular Tree”

By Emily C. Battle

Question 1

- 50 yof presents for evaluation of shortness of breath. Exam demonstrates 2+ pitting lower extremity edema, loud P2, no murmurs. What is the best screening evaluation for pulmonary hypertension?

1. Chest x-ray
2. Transthoracic echocardiogram
3. NT-Pro BNP
4. Pulmonary function tests

Case 1: 55 yof presents DOE x 1 year

- History of mild ILD & Raynaud’s. Mechanic’s hands w/ telangiectasias, velcro crackles & loud P2. BNP 3300.

- Echo: RVSP 100 mmHg with moderate RVE & moderate RV dysfunction

- RHC: RAP 17, mPAP 61, PAWP 7, PVR 27 WU, -VD
### Case 2: 52 yof presents DOE x 1.5 yrs

- History of PE 1.5 yrs ago. Distant heart sounds, +1 LEE. BNP 94
- Echo: RVSP 104 mmHg with severe RVE & moderate RV dysfunction
- RHC: RAP 12, mPAP 62,
  PAWP 11, PVR 10 WU, -VD

### Case 3: 65 yom presents DOE x 10 yrs

- History of cad s/p CABG (1990’s) & constriction s/p pericardiectomy (2012). +3 LEE, loud P2. BNP 89
- Echo: RVSP 92 mmHg with mild RVE & normal RV fxn
- RHC: RAP 15, mPAP 56, PAWP 25, PVR 5.1 WU, TPG 31, -VD
Rural MHI PH Experience - Methods

- Consecutive patients referred to/or within MHI Baxter/Crosby/Aitkin with RVSP > 50mmHg on echo OR with suspected PH OR previous diagnosis of PAH on meds (N=2)

- Study period: 8/1/2015-2/1/2016

- All had an echo and right heart cath

Rural MHI PH Experience - Methods

- PAH = mPAP ≥ 25 mmHg with PAWP ≤ 15 or high transpulmonary gradient (≥ 17)

- Study period: 8/1/2015-2/1/2016

- All had an echo & right heart cath
Rural MHI PH Experience - Results

- N = 29 → 20 met criteria for PAH; 9 with non-group 1 PH
  - Non-group 1 – 8 due to diastolic heart failure (1-COPD)
- 2 pts had prior PAH diagnosis & on medication
- 1 pt with CTEPH

Rural MHI PH Experience – Echo Results

<table>
<thead>
<tr>
<th></th>
<th>RAP (mmHg)</th>
<th>RVSP (mmHg)</th>
<th>Mod/Severe RVE</th>
<th>Mod/Severe RV Dysfxn</th>
<th>Pericardial Effusion (n)</th>
<th>LVEF (%)</th>
</tr>
</thead>
<tbody>
<tr>
<td>PAH</td>
<td>12±5</td>
<td>76±25</td>
<td>9</td>
<td>8</td>
<td>4</td>
<td>63±11</td>
</tr>
<tr>
<td>Group 2-3 PH</td>
<td>11±7</td>
<td>54±15</td>
<td>4</td>
<td>0</td>
<td>0</td>
<td>56±12</td>
</tr>
</tbody>
</table>
## Rural MHI PH Experience

<table>
<thead>
<tr>
<th></th>
<th>RAP (mmHg)</th>
<th>RVSP (mmHg)</th>
<th>mPAP (mmHg)</th>
<th>PAWP (mmHg)</th>
<th>TPG (mmHg)</th>
<th>PVR (WU)</th>
<th>CI (L/min/m2)</th>
<th>VD (n=+)</th>
</tr>
</thead>
<tbody>
<tr>
<td>PAH</td>
<td>11±3</td>
<td>73±18</td>
<td>45±9</td>
<td>16±5</td>
<td>29±10</td>
<td>7 (2.2-11.1)</td>
<td>2.8±0.9</td>
<td>4</td>
</tr>
<tr>
<td>Group 2-3 PH</td>
<td>11±5</td>
<td>52±8</td>
<td>34±8</td>
<td>20±7</td>
<td>13±3</td>
<td>2 (1.5-4.24)</td>
<td>2.99±1.0</td>
<td>0</td>
</tr>
</tbody>
</table>

### Rural MHI PH Experience - Results

- 17 PAH pts treatment-naïve → 11 started PH-medications
- 7 started combo therapy – 2 on triple therapy since
- 4 pts started CCB only (+VD)
- 6 pts declined PH-medications
- Update (5/1/16): N = 55, 20 pts on meds
Rural MHI PH Experience - Conclusion

- PH may be **underdiagnosed in non-urban areas**

- **Internal echo database** provides an opportunity to identify patients who may benefit from PH-directed treatment

- Will this **improve patient outcomes**? It has significantly improved quality of life in a subset of patients.

Special Thanks

- Dr. Tim Dirks
- Dr. Peter Stokman
- Dr. Jim Furda
- Shawna Reed, RN
- Chrissie Soxman, RN
- Baxter/Crosby RNs & techs & support staff
- Right Heart Cath Team at ANW
What is Pulmonary Hypertension?

- ↑ pressure in pulmonary vasculature
- Results in progressive RV failure & subsequent death
- Why does it matter?
  - 85% - 91% 1 yr survival for PAH
  - 57% 5-yr survival for PAH
- Median 2.7 years from symptoms to diagnosis!

<table>
<thead>
<tr>
<th>VHD</th>
<th>Condition</th>
<th>Overall prevalence of PH (%)</th>
<th>Impact of PH on Outcome</th>
</tr>
</thead>
<tbody>
<tr>
<td>Aortic stenosis</td>
<td>Rest</td>
<td>15%-30%</td>
<td>Controversial; ≈2-fold increase of 1 year in mortality after intervention</td>
</tr>
<tr>
<td></td>
<td>Exercise</td>
<td>55%</td>
<td>≈2-fold increase in risk of cardiac event in asymptomatic patients</td>
</tr>
<tr>
<td>Mitral stenosis</td>
<td>Rest</td>
<td>&gt;40%</td>
<td>Event-free survival: 77% at 10 years and 41% at 15 yrs</td>
</tr>
<tr>
<td></td>
<td>Exercise</td>
<td>79%</td>
<td></td>
</tr>
<tr>
<td>Aortic regurgitation</td>
<td>Rest</td>
<td>&gt;25%</td>
<td>Controversial</td>
</tr>
<tr>
<td></td>
<td>Exercise</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Primary MR</td>
<td>Rest</td>
<td>20%-30% and 6%-30% in asymptomatic patients; &lt;20% in asymptomatic patients with preserved LVEF</td>
<td>&gt;2-fold increase in risk of post-operative death</td>
</tr>
<tr>
<td></td>
<td>Exercise</td>
<td>≈50%</td>
<td>&gt;3-fold increase in risk of occurrence of symptoms</td>
</tr>
<tr>
<td>Secondary MR</td>
<td>Rest</td>
<td>37%-62%</td>
<td>≈1.4-fold increase in risk of death</td>
</tr>
<tr>
<td></td>
<td>Exercise</td>
<td>40%</td>
<td>&gt;3-fold increase of cardiac events; &gt;3-fold increase in risk of death; involved in pathogenesis of acute pulmonary edema</td>
</tr>
</tbody>
</table>

PH as a Comorbidity = Increased Mortality in Aortic Stenosis

PH $= mPAP \geq 25$ mmHg on cath
PH as a Comorbidity = Increased Mortality in Systolic/Diastolic Heart Failure

Guazzi M. Circ, 2012; 126:975-90.

Anatomy - Pulmonary Hypertension

Increased flow
High output
Shunt: ASD/VSD/pulm

Irreversible Plexiform lesions

Protective vasoconstriction
PVOD

Elevated left sided pressures
Aortic valve disease
Mitral valve disease
LV diastolic dysfunction
LV systolic dysfunction

Slide courtesy of Rick Nishimura, MD
PH Incidence/Background

- **Incidence**: Group I = 15-26 cases per million

- 12% of patients with Systemic Sclerosis

- 2% - 6% in portopulmonary HTN

- 15% - 20% incidence in OSA

- 2% - 5% incidence after acute PE
PH Symptoms

- Nonspecific
- Dyspnea, DOE, chest pain
- Syncope or presyncope
- Lower extremity edema, abdominal bloating, early satiety

Figure 1. Symptoms encountered most often in patients with pulmonary arterial hypertension. Y-axis shows percent of respondents. SOB: shortness of breath.

Physical Exam Findings

- RV lift/heave
- Accentuated P2
- Holosystolic murmur with inspiratory ↑ (TR)
- Rhonchi/crackles
- Pulsatile liver & ascites, hepatojugular reflux

WHO Functional Classification

- **Class I** – no limitation
- **Class II** – slight limitation but not at rest
  - Ordinary physical activity causes sx’s
- **Class III** – marked limitation w/ activity
  - OK at rest
  - Less than ordinary activity = sx’s
- **Class IV** – severe limitation – at rest
Pivotal Tests

- History
- Exam
- CXR
- ECG

- Echocardiogram
- VQ Scan
- PFTs
- Overnight Oximetry
- HIV
- ANA
- LFTs

- Functional Test
- And Biomarkers

- RH Cath

Contingent Tests

- TEE
- Exercise Echo
- Pulmonary Angiography
- Chest CT Angiogram
- Coagulopathy Profile

- ABGs
- Polysomnography
- Other CTD Serologies

- Vasodilator Test
- Exercise RH Cath
- Volume Loading
- Left Heart, Coronary

Contribute to Assessment of:

- Index of Suspicion of PH
- RVE, RAE, TRVSP, RV Function
- Left Heart Disease
- VHD, CHD
- Chronic PE
- Ventilatory Function
- Gas Exchange
- Sleep Disorder
- HIV Infection
- Scleroderma, SLE, RA
- Portopulmonary Htn
- Establish Baseline
- Prognosis
- Confirmation of PH
- Hemodynamic Profile
- Vasodilator Response
- r/o LMCA Compression

Approach to PH Evaluation

Table 12: Recommendations for diagnostic strategy

<table>
<thead>
<tr>
<th>Recommendations</th>
<th>Class</th>
<th>Level</th>
<th>Ref</th>
</tr>
</thead>
<tbody>
<tr>
<td>Echocardiography is recommended as a first-line non-invasive diagnostic investigation in case of suspicion of PH</td>
<td>I</td>
<td>C</td>
<td>47</td>
</tr>
<tr>
<td>Ventilation/perfusion or perfusion lung scan is recommended in patients with unexplained PH to exclude CTEPH</td>
<td>I</td>
<td>C</td>
<td>93</td>
</tr>
<tr>
<td>Contrast CT angiography of the PA is recommended in the workup of patients with CTEPH</td>
<td>I</td>
<td>C</td>
<td></td>
</tr>
<tr>
<td>Routine biochemistry, haematology, immunology, HIV testing and thyroid function tests are recommended in all patients with PAH to identify the specific associated condition</td>
<td>I</td>
<td>C</td>
<td>67</td>
</tr>
<tr>
<td>Abdominal ultrasound is recommended for the screening of portal hypertension</td>
<td>I</td>
<td>C</td>
<td>36</td>
</tr>
<tr>
<td>Lung function test with DLCO is recommended in the initial evaluation of patients with PH</td>
<td>I</td>
<td>C</td>
<td></td>
</tr>
<tr>
<td>High-resolution CT should be considered in all patients with PH</td>
<td>IIA</td>
<td>C</td>
<td>94</td>
</tr>
<tr>
<td>Pulmonary angiography should be considered in the workup of patients with CTEPH</td>
<td>IIA</td>
<td>C</td>
<td></td>
</tr>
<tr>
<td>Open or thoracoscopic lung biopsy is not recommended in patients with PAH</td>
<td>III</td>
<td>C</td>
<td></td>
</tr>
</tbody>
</table>

Echo is a Screening Test

- Normal RV pressure < 35 mmHg
- Estimate RVSP with modified Bernoulli equation
  - $RVSP = 4(TRv)^2 + RAP$
- Can over or underestimate
- RV size and function: TAPSE, $S'$, FAC


Right Heart Catheterization

- Gold standard for diagnosis
- Mean PA pressure ≥ 25 mm Hg
- $PAH = PCWP \leq 15$ mm Hg

Positive Vasodilator Test
1. mPAP by ≥ 10 mmHg
2. mPAP < 40 mmHg
3. Normal or ↑ in CO

~10% of patients have + vasodilator study
**Table 1** Updated Classification of Pulmonary Hypertension*  

<table>
<thead>
<tr>
<th>Classification</th>
<th>PAH</th>
<th>Left Heart</th>
<th>Lungs</th>
<th>VTE</th>
<th>Misc</th>
</tr>
</thead>
<tbody>
<tr>
<td>1. Pulmonary arterial hypertension</td>
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</tr>
<tr>
<td>1.1 Idiopathic PAH</td>
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<tr>
<td>1.2 Heritable PAH</td>
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<tr>
<td>1.2.1 BMP2R</td>
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<tr>
<td>1.2.2 ALK1, ENG, SMA9, CAV1, KCNK3</td>
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<tr>
<td>1.2.3 Unknown</td>
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<tr>
<td>1.3 Drug and toxin induced</td>
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<tr>
<td>1.4 Associated with:</td>
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<tr>
<td>1.4.1 Connective tissue disease</td>
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<td>1.4.2 HIV infection</td>
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<tr>
<td>1.4.3 Portal hypertension</td>
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<tr>
<td>1.4.4 Congenital heart diseases</td>
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<tr>
<td>1.4.5 Scleroderma</td>
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<tr>
<td>1. Persistent pulmonary hypertension of the newborn (PPHN)</td>
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<tr>
<td>2. Pulmonary hypertension due to left heart disease</td>
<td></td>
<td></td>
<td></td>
<td></td>
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</tr>
<tr>
<td>2.1 Left ventricular systolic dysfunction</td>
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<td></td>
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<tr>
<td>2.2 Left ventricular diastolic dysfunction</td>
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<td></td>
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<tr>
<td>2.3 Valvular disease</td>
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<tr>
<td>2.4 Congenital/acquired left heart inflow/outflow tract obstruction and congenital cardiomyopathies</td>
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<td></td>
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<td></td>
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<tr>
<td>3. Pulmonary hypertension due to lung diseases and/or hypoxia</td>
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<td></td>
<td></td>
<td></td>
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<tr>
<td>3.1 Chronic obstructive pulmonary disease</td>
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<tr>
<td>3.2 Interstitial lung disease</td>
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<tr>
<td>3.3 Other pulmonary diseases with mixed restrictive and obstructive pattern</td>
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<td></td>
<td></td>
<td></td>
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<tr>
<td>3.4 Sleep-disordered breathing</td>
<td></td>
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<td></td>
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<tr>
<td>3.5 Atrial hypoplasia disorders</td>
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<td></td>
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<tr>
<td>3.6 Chronic exposure to high altitude</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
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<tr>
<td>3.7 Developmental lung diseases</td>
<td></td>
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</tr>
</tbody>
</table>


*5th WSPH Nice 2013. Main modifications to the previous Dana Point classification are in bold.

BMPR = bone morphogenetic protein receptor type II; CAV1 = caveolin-1; ENG = endoglin; HIV = human immunodeficiency virus; PAH = pulmonary arterial hypertension.

Creation of Accredited PH Centers

- Underdiagnosis & later referral
- Misdiagnosis
- Treatment delays
- Treatment guidelines underutilized within 6 months of death

- 29 in US; 1 in MN
RePHerral Study – 140 Consecutive Pts

- Referred by cardiologist or pulmonologist to a tertiary/quarternary referral center for PH
- 98 pts with definitive diagnosis before referral
  - 32 (33%) pts misdiagnosed
  - 59 new caths – 25/59 (42%) pts received different diagnosis
  - 24/42 (57%) pts on meds contrary to published guidelines
- Conclusion: pts referred to PH centers for Dx & Tx are often referred late, misdiagnosed, & inappropriately prescribed meds

BMPR2 mutation – 80% of familial cases; cell growth & proliferation unchecked

- A) Plexiform lesion
- B) Plex lesion with recanalization
- C & D) Plex lesions in fen-phen
- E-H) Recanalizing thrombi with plexiform lesions


Mean Survival in PAH Based on Etiology
What are the prognostic indicators in PAH?

**Table 13** Risk assessment in pulmonary arterial hypertension

<table>
<thead>
<tr>
<th>Determinants of prognosis* (estimated 1-year mortality)</th>
<th>Low risk &lt;5%</th>
<th>Intermediate risk 5–10%</th>
<th>High risk &gt;10%</th>
</tr>
</thead>
<tbody>
<tr>
<td>Clinical signs of right heart failure</td>
<td>Absent</td>
<td>Absent</td>
<td>Present</td>
</tr>
<tr>
<td>Progression of symptoms</td>
<td>No</td>
<td>Slow</td>
<td>Rapid</td>
</tr>
<tr>
<td>Syncope</td>
<td>No</td>
<td>Occasional syncope*</td>
<td>Repeated syncope*</td>
</tr>
<tr>
<td>WHO functional class</td>
<td>LII</td>
<td>III</td>
<td>IV</td>
</tr>
<tr>
<td>6MWD</td>
<td>&gt;440 m</td>
<td>165–440 m</td>
<td>&lt;165 m</td>
</tr>
<tr>
<td>Cardiopulmonary exercise testing</td>
<td>Peak VO₂ &gt;15 ml/min/kg (45% predicted)</td>
<td>Peak VO₂ 11–15 ml/min/kg (35–35% predicted)</td>
<td>Peak VO₂ &lt;11 ml/min/kg (&lt;35% predicted)</td>
</tr>
<tr>
<td></td>
<td>VE/VO₂ slope &gt;36</td>
<td>VE/VO₂ slope 36–44</td>
<td>VE/VO₂ slope &gt;44</td>
</tr>
<tr>
<td>NT-proBNP plasma levels</td>
<td>BNP &lt;50 ng/l</td>
<td>BNP &gt;50 ng/l</td>
<td>BNP &gt;100 ng/l</td>
</tr>
<tr>
<td></td>
<td>NT-proBNP &lt;300 ng/l</td>
<td>NT-proBNP 300–1000 ng/l</td>
<td>NT-proBNP &gt;1000 ng/l</td>
</tr>
<tr>
<td>Imaging (echocardiography, CTA imaging)</td>
<td>RA area &lt;18 cm²</td>
<td>No pericardial effusion</td>
<td>RA area &gt;18 cm²</td>
</tr>
<tr>
<td></td>
<td>No or minimal pericardial effusion</td>
<td>Pericardial effusion</td>
<td></td>
</tr>
<tr>
<td>Hemodynamics</td>
<td>RAP &lt;8 mmHg</td>
<td>CI 2.5–3.5 l/min/m²</td>
<td>RAP &gt;14 mmHg</td>
</tr>
<tr>
<td></td>
<td>So₂ &gt;65%</td>
<td>CI 2.0–2.4 l/min/m²</td>
<td>So₂ &lt;60%</td>
</tr>
</tbody>
</table>


Pericardial Effusion Associated with Mortality

![Graph A](image1)

Moderate or greater effusion

Small effusion

No effusion

No. at risk

<table>
<thead>
<tr>
<th>Follow-up, years</th>
<th>No effusion</th>
<th>Effusion</th>
</tr>
</thead>
<tbody>
<tr>
<td>0</td>
<td>427</td>
<td>101</td>
</tr>
<tr>
<td>1</td>
<td>333</td>
<td>101</td>
</tr>
<tr>
<td>2</td>
<td>277</td>
<td>83</td>
</tr>
<tr>
<td>3</td>
<td>229</td>
<td>62</td>
</tr>
<tr>
<td>4</td>
<td>158</td>
<td>50</td>
</tr>
<tr>
<td>5</td>
<td>116</td>
<td>31</td>
</tr>
</tbody>
</table>

RAE Associated with Mortality


RVH on ECG Predicts Mortality

Prognosis and Prediction
Composite Indices
REVEAL Risk Calculator

Survival (%)

Risk Scores
1-5
6-7
8-9
10-11
12-15

0 10 20 30 40 50 60 70 80 90 100

0 3 6 9 12

Months from Enrollment
CTEPH Incidence/Background

- **Incidence:** Group 4 = 2-5% after acute PE
- 25% of pts with CTEPH have no history of PE

![CTEPH Types](image)

Figure 1. Cumulative incidence of chronic thromboembolic pulmonary hypertension (CTEPH) after a first episode of pulmonary embolism without prior deep vein thrombosis. Reproduced by permission from Reference 30.

CTEPH Diagnosis

- RHC: mPAP ≥ 25 mmHg & PAWP ≤ 15 mmHg
- Distal or proximal thromboembolic occlusions
  - High probability VQ – perfusion defects
  - + PE on CT after anticoagulation > 3 months

CTEPH Treatment & Prognosis

- Proximal disease = surgery (PTEA)
  - At an expert center
  - 11-35% of pts will have residual PH
- Distal disease = Medication
  - Anticoagulation
  - PH-directed meds
  - Balloon pulmonary angioplasty?

Paradigm shift...

Survival Improvement with Treatment Advances

According to repeated nationwide surveys,

More Doctors Smoke CAMELS than any other cigarette!

Survival (%)

Years

NIH '81-5 (N=194) PPH
China '99-04 (N=72) I/FPAH
French '02-3 (N=56) I/F/TPAH (incident)
REVEAL '01-9 (N=985) I/FPAH (NIH criteria)
French '06-11 (N=281)I/F/TPAH

MultiRx
Epo
CCB only
~10% of patients have + vasodilator study


Everyone gets these!
What about combo therapy?

Table 20  Recommendations for efficacy of initial drug combination therapy for pulmonary arterial hypertension (group 1) according to World Health Organization functional class. Sequence is by rating

<table>
<thead>
<tr>
<th>Measure/treatment</th>
<th>Class* Level#</th>
<th>Ref*</th>
</tr>
</thead>
<tbody>
<tr>
<td>Ambrisentan + tadalafil</td>
<td>I B I B Iib C</td>
<td>247</td>
</tr>
<tr>
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<tr>
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<tr>
<td>Other ERA or PDE-5i + other Iv. prostanoid analogues</td>
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Ongoing symptoms/RV dysfunction = addition of meds

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Balloon atrial septostomy & lung transplant considered if maximal meds & ongoing symptoms


Case 1: 55 yof presents DOE x 1 year

- History of mild ILD & Raynaud’s. Mechanic’s hands w/ telangiectasias, velcro crackles & loud P2. BNP 3300.

- **Diagnosis:** PAH with antisynthetase syndrome

- **Treatment:** Combo therapy x 4 months with mild symptom improvement – starting triple oral therapy
  - Next step – IV prostacyclin

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Case 2: 52 yof presents DOE x 1.5 yrs

- History of PE 1.5 yrs ago.

- **Diagnosis:** CTEPH w/ +VQ & proximal disease on ECG-gated CT

- **Treatment:** Riociguate & Pulmonary thromboendarterectomy – “500% BETTER!”
  - RVSP 40 mmHg – no septal flattening, normal RV size & fxn
Case 3: 65 yom presents DOE x 10 yrs

- History of cad s/p CABG (1990’s) & constriction s/p pericardiectomy (2012).

- **Diagnosis:** PH out of proportion with high transpulmonary gradient & PVR

- **Treatment:** aggressive diuresis, mono then combo, then triple therapy → RVSP 49 mmHg – “I haven’t felt this well in the last 10 years!”

Take Home Messages

- PAH is RELATIVELY rare

- RHC is gold standard; Echo = screening test

- Push for accredited centers of excellence

- VQ scan to rule out chronic thromboembolic PH

- Groups 1-5

- Appears more aggressive up-front treatment may lead to improved outcomes
Thanks!
Email:
Eric.Fenstad@allina.com

References

- Simonneau G. JACC 2013, 62(2S).
References

- Benza et al. REVEAL: ATS 2009

References