

## Monitoring treatment success: Is your treatment working?

Providence, R.I. 2017 PHA *on the Road*



### Session Description:

In this session, we will discuss the medication options that are available for PH and how medications are adjusted or changed based on improvements in symptoms, testing, side effects, and patient preference. Join us, as we discuss how health care professionals monitor effectiveness of therapies chosen, and how decisions to alter or advance therapies are made. This session will discuss the balance between the benefits and risks of multiple agents and how they affect the treatment process for a specific patient. The focus will be to highlight what your PH care team looks for in measuring your treatment goals and the rationale for the timing and adding of PH medications, if needed.

### Learning Objectives

Attendees will be able to:

- Discuss the medical decision-making behind drug therapies
- Understand your role in medication choices
- Review presently available treatment options
- Identify the goals of therapy
- Understand how medication effectiveness is monitored

### Medications

- There are presently 5 FDA-approved classes of medication for the treatment of WHO Group 1 PH (PAH, pulmonary arterial hypertension)<sup>1</sup>.
- There is currently 1 FDA-approved classes of medication for the treatment of WHO Group 4 PH (CTEPH, chronic thromboembolic pulmonary hypertension)
- Synthetic prostacyclins, prostacyclin analogues, and prostacyclin receptor agonists (PGI)
  - Epoprostenol (Flolan® and Veletri®) – Intravenous
  - Iloprost (Ventavis®) – Inhaled 6 to 9 times a day
  - Selexipag (Uptravi®) – Oral twice daily
  - Treprostinil (Remodulin®) – Intravenous or subcutaneous
  - Treprostinil (Tyvaso®) – Inhaled 4 times a day
  - Treprostinil (Orenitram®) – Oral 2-3 times a day
- Endothelin Receptor Antagonists (ERA)
  - Ambrisentan (Letairis®) – Oral 1 time a day
  - Bosentan (Tracleer®) – Oral 2 times a day
  - Macitentan (Opsumit®) - Oral 1 time a day
- Phosphodiesterase-5 Inhibitors (PDE5)
  - Sildenafil (Revatio®) – Oral 3 times a day
  - Tadalafil (Adcirca®) – Oral two pills 1 time a day
- Soluble Guanylate Cyclase Stimulators (sGC)
  - Riociguat (Adempas®) - oral three times a day
    - *Riociguat is an FDA-approved therapy for PAH as well as chronic thromboembolic pulmonary hypertension (CTEPH) that is persistent/recurrent after surgery or deemed inoperable by an expert center. CTEPH is a specific type of PH hypertension caused by chronic blood clots in the more central or larger pulmonary arteries.*

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<sup>1</sup> Calcium channel blockers are oral medications that are also used for a specific group of patients with WHO Group 1 PH who have shown a clinical response in the cardiac catheterization laboratory or ICU, called “vasoreactivity testing.”



**Important Considerations**

- PH Group Classification: The therapies above are indicated for those who have Group 1 PH (PAH).
- There is only one medication approved for WHO Group 4 (CTEPH) who are inoperable or who have PH remaining after PTE surgery.
- There are no FDA-approved targeted therapies for the treatment of WHO Groups 2, 3, or 5. Presently, we treat the underlying causes of these types of PH.
- The cause of PH also plays a role in decision-making, as certain associative disorders tend to be more aggressive, therefore, may warrant a more aggressive treatment plan.
- WHO Functional Classification: How your symptoms impact your daily activities and quality of life is a major factor in determining what medications are right for you.

See this chart below to understand how we determine functional class

WHO Functional Class	Definition
I	Physical activity is not limited. Ordinary physical activity does not cause shortness of breath, feeling tired, chest pain or discomfort, or feeling faint.
II	Comfortable at rest, but ordinary physical activity causes shortness of breath, feeling tired, chest pain or discomfort, or feeling faint.
III	Comfortable at rest, but less than ordinary physical activity causes shortness of breath, feeling tired, chest pain or discomfort, or feeling faint.
IV	Unable to perform any physical activity. Shortness of breath and/or feeling tired may be present at rest, and symptoms increase with almost any physical activity.

Barst RJ, McGoon M, Torbicki A, et al. Diagnosis and differential assessment of pulmonary arterial hypertension. J Am Coll Cardiol 2004; 43: S40–S47.

**PH-targeted therapy options in relationship to WHO Functional Classification**

- Functional Class I means you have no symptoms. Therefore, patients may or may not be treated with PAH medications.
- Functional Class II means you have some symptoms with activities such as shopping or climbing up 2 flights of stairs. You may be treated with either: 1 oral medication, a combination of 1 or more oral medications, or with an inhaled medication. You may also be considered for an infused therapy or in combination with an oral medication.
- Functional Class III means you have symptoms with doing household chores or climbing up one flight of stairs. Treatment plan is similar to that of WHO Functional Class II; but is more likely to be combination therapy.
- Functional Class IV means you are experiencing symptoms all the time, even at rest. The guidelines recommend infusion therapy for those who are Functional Class IV. However, PH specialist recognize that there may be other factors that limit one’s ability to manage infusion therapy.
- Many patients are on combination therapy and maybe on 3 medications to treat the PAH.
  - For example - Infused PGI + sGC + ERA or Infused PGI + PDE5 + ERA

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- **Hemodynamics:** The information obtained on your right heart catheterization and echocardiogram weigh heavily in the discussion regarding treatment options. We also pay attention to how your right ventricle is functioning as determined by a cardiac echo and the right heart cath.
- **Physical limitations are important:** Some medications require the use of devices, such as pumps and inhalers. Discuss any concerns you have regarding your ability, or inability, to manage devices with your healthcare team. The specialty pharmacy nurses can meet with you so you can practice with the device prior to starting therapy. This will allow you and your healthcare team to identify any barriers, or obstacles, in your treatment regimen in order to develop solutions.
- **Social Considerations:** In certain circumstances you may need the assistance of a social worker to help you identify community resources to assist you. Other things to discuss with your team include:
  - Emergency planning: Mapping the distance to nearest hospital or to your PH Center, and developing a plan with friends/family during an emergency.
  - Affordability: There are programs available to aid in the cost of medications. Talk with your specialty pharmacy and/or healthcare team to learn more about these options.

### Additional supportive therapies

- Oxygen: No medication can replace your bodies need for oxygen therapy.
- Fluid restriction: Limiting how much fluid you take in can help you control edema (fluid retention).
- Diuretics (i.e., “water pills”) may be very important in controlling fluid balance too.
- Rehabilitation is also important in helping you regain and maintain a better level of functioning.

### Goals of therapy

- Improve your quality of life
- Decrease your symptoms
- Improve your 6-minute walk distance
- Improve your heart function
- Being able to communicate treatment needs openly and honestly with your healthcare team, as this is a two-way discussion.

### Take-away messages

- Choosing the drug that is right for you is an art and should be done in collaboration with your healthcare team
- Weighing benefits and side effects of each drug is important for both you and your healthcare team.
- Discuss your social situation with your healthcare team as this will also help guide them as they make personalize a treatment plan that is in accordance with your wishes and comfort.

### Additional Resources

- [www.PHAssociation.org/Patients/SurvivalGuide](http://www.PHAssociation.org/Patients/SurvivalGuide)
- <http://www.phassociation.org/Treatments>