



Follow-up Visits: How Clinicians Make Decisions for Your Care

2018 PHA on the Road – Milwaukee

Session Description

Understanding why the frequent follow-up visits requested by your health care team are routine for PH patients can help alleviate stress and anxiety. Information collected during these appointments is crucial to ensuring that PH patients are receiving quality care and treatment. In this session, panelists will explain why some tests you might have already had to diagnose your PH are repeated. Information gathered from testing helps clinicians determine if changing, adding-on or removing PH medications can be considered when your treatment goals are not being met or when medications cause side effects too difficult to manage. Panelists will also discuss the concept of “goal-oriented therapy” and how patient goals, preferences and possible limitations factor into the selection of which therapy is considered, as well as the balance between risk and benefits of taking multiple medications.

Learning Objectives

At the end of the session, attendees will be able to:

- Understand which PH diagnostic tests are repeated in follow-up appointments, how often and why.
- Understand what information clinicians gather from these tests and how this may influence the need for a change or adjustment in PH therapy.
- Know what questions to ask before changing therapy and common goals of therapy that influence decision-making in PH therapy selection.

Most Commonly Repeated Exams During Follow-up Appointments

- Functional class assessment.
- Lab tests: BNP or NT-proBNP.
- 6-minute walk test.
- Echocardiogram.
- ECG/EKG.
- Cardiopulmonary exercise test.

Determining Whether and When to Repeat Tests

- Physicians differ in when repeat tests and their decisions on how often to repeat testing are determined on a case-by-case method and other factors, such as:
 - WHO Group classification.
 - WHO functional class.
 - Your body’s response to a change in or addition of therapy(ies).

Goals of Therapy and Care

- Improve your quality of life.
- Decrease your symptoms.
- Improve your six-minute walk distance.
- Improve your heart function.
- Ability to communicate treatment needs openly and honestly with your health care team as this must be a two-way discussion.

Considering Changing or Combining Therapies

Early Combination Therapy

- A new approach to treating some patients with newly-diagnosed PH is to consider early combination therapy with two (or more) medications started either at the same time or one shortly after the other.
- Early combination therapy results from a large clinical trial evaluating this approach and analysis of patients treated with combination therapy in other clinical trials; it typically involves use of two oral drugs; early combination therapy may be appropriate for patients with more than mild symptoms related to PH, but benefits must be weighed with potential side effects of multiple drug therapy.

PH Specialists' Considerations in Choosing Appropriate Treatment

- Signs of right heart failure.
- Rate symptoms are worsening.
- Patient is passing out.
- World Health Organization (WHO) functional class (system created to define the severity of an individual's symptoms).
- Distance patient can walk on the six-minute walk test (6MWT).
- Significant change in patient's 6MWT between visits.
- Results from a cardiopulmonary exercise test.
- Levels of the NT-proBNP hormone produced by the heart and can be released in response to pressure changes.
- Look of the heart; for example, images taken with an echocardiogram or cardiac MRI.
- Pressure measurements and cardiac output during right heart catheterization.

Addressing Intolerable Side Effects

- Difficult side effects from PH-specific medications are common, including: headaches, nausea, diarrhea, vomiting, lightheadedness, jaw pain, flushing and acid reflux.
- Your PH care team may recommend strategies or medications to reduce side effects from PH-specific medications. Not all medications and strategies will work for all patients; it may be time to re-evaluate the treatment if efforts to reduce side effects have been unsuccessful.
- Infection risk for patients on continuous infusions given intravenously (IV) into the vein or subcutaneously (SQ) under the skin; the risk may be higher for patients taking other medications that reduce the activity of their immune system. Interruption of the IV or SQ PH drug can be quite dangerous and may require hospitalization for patients who are having a pump issue or who suspect an infection; some patients report feeling stressed with these concerns, which can be a barrier in treatment with prostacyclin therapy .

Key Questions to Consider When Clinicians and Patients Consider Changing Therapies

- Understanding potential side effects of a new PH therapy:
 - Review potential side effect(s) for the medications you are taking or might soon be taking with your health care team.
 - Prepare questions in advance to discuss with your PH team related to side effect management; not all patients will experience reported side effects.
 - Stay in close contact with your PH team once you have started a medication; use the nurse educators from the specialty pharmacies or PH pharmacists.
 - Adhere to required monitoring, such as liver function tests (LFTs), pregnancy tests, other labs.
 - Your PH team will review tests that are required for new therapies, as well as possible scheduled interval testing to monitor response to treatment.
 - Pregnancy should be avoided in patients with PH; women of childbearing potential should discuss these risks with their team, have a contraception management plan in



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place, obtain monthly pregnancy tests and notify their PH team immediately if there is a possibility of pregnancy.

- One endothelin receptor antagonist (Bosentan [Tracleer®]) requires monthly monitoring of LFTs and blood counts; changes to the lab values possibly attributed to the medication may require more frequent blood testing or stopping the medication.
- Route of delivery
 - Maintain sterile techniques when preparing medications for IV or SQ prostacyclin therapy to reduce infection risk.
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Takeaway Messages

- Compliance with follow-up appointments and repeat testing for your PH is crucial to better quality of care and life. Make sure to be your own advocate and ask why you are having tests repeated if you are uncertain. The more you know, the more comfortable you'll be with these procedures, no matter how tedious they can be.
- Changing and combining therapies can be an important part of optimizing your overall treatment plan; this usually can be accomplished safely, but it requires an organized and careful plan with you and your PH care team.