

Communicating and advocating in a PH emergency

Portland, Ore. 2017 PHA on the Road



Session Description

In this session, we will focus on providing strategies that PH patients and their families can use to optimize medical care during a “PH emergency.” This session aims to educate attendees on common symptoms of a PH emergency, causes of a PH emergency and when to call your PH doctor. We will discuss how to be proactive and prepared to minimize the impact of emergencies or — better yet — avoid them altogether. We will discuss strategies to improve navigation through the emergency department and develop good communication skills for any PH emergency to ensure that care is consistent and aligned with the goals and wishes of the PH patient.

Learning Objectives

Attendees will be able to:

- Define “PH emergency,” and understand why immediate action is critical
- Know when to contact their PH doctor to prevent a PH emergency
- Know the importance of developing a “PH emergency” plan with their provider to convey important information about PH and medications to emergency responders and hospital physicians, including utilization of their PH doctor
- Advocate for themselves or their family members during the management of a PH emergency

What is a “PH emergency?”

- Sudden worsening of pulmonary hypertension that can lead to rapid decline in heart function, organ failure and even death.
- Common symptoms can include: confusion, low blood pressure or oxygen levels, increased shortness of breath, dizziness or lightheadedness, syncope (“passing out”), chest pain and abnormal heart beat.
- It is never inappropriate to seek urgent emergency department (ED) evaluation for such symptoms, even for patients who do not have PH.
- In some cases, an urgent situation might be handled better outside the ED – for example infusion pump problems can sometimes be addressed by a call to the PH clinic or specialty pharmacy nurses. Make sure you know who to contact for pump problems if you are on continuous infusion prostacyclin therapy and keep the phone numbers where you can easily access them.
- Common causes of a “PH emergency” include:
 - Failure or interruption of PH medications
 - Fluid overload
 - New infection
 - Abnormal heart rhythms
 - Blood clots
 - New anemia or bleeding
 - Syncope or fainting
- In some cases, surgical procedures or pregnancy can lead to a “PH emergency.”
 - **Immediate** medical attention is required, and a PH specialist should be involved.



- **A PH emergency can happen anywhere, always be prepared!**

When do I call my PH doctor?

- Have a “PH emergency plan” talk with your provider. Find out what you should do if an emergent or urgent problem occurs. Know ahead of time what your plan is, especially if you’ll be traveling.
- For the onset of any symptoms described above, your doctor can help to decide the best way to get you medical attention.
- For patients using continuous infusions (IV or subcutaneous) of prostacyclin medicines, you should call your doctor immediately for any problems with the IV, pump or medication.
- Signs/symptoms of an IV port/line or SQ site infection.
- Increase in O₂ usage to get O₂ levels (sats) in desired range?
- If you are developing progressive symptoms, including weight gain (more than 4 pounds in a week or 2 pounds in a day). Note that this may vary provider to provider.
- Well before you run out of your PH medications or if you need a refill
- Well in advance of any planned surgeries
 - It is important that your PH care team talk to the surgeon and anesthesiologist prior to ANY surgical procedure or other procedure requiring sedation.
- As soon as you know that you are pregnant

What should I do if I have to call 911? Or go the emergency department?

Careful preparation lets you do the one thing you probably can’t do very well during an emergency – think clearly. Here are some things that can be done ahead of time:

- Calling 911
 - Have a place in your kitchen where your medications are kept. Any PH-specific medications should be kept together separate from the others – many hospitals do not have these medications, so EMS should bring them with you. Make a sign on the container in which they are kept so the providers know to bring them.
 - Have a frequently updated information list (with copies so the ambulance crew can take it to the hospital with them) including the following:
 - Medication list with the most recent date it was changed. This includes the medication name, dose, and frequency of administration. On this piece of paper should also be contact name and number of your PH specialist(s).
 - Medical history and surgical history. As many general providers are not familiar with pulmonary hypertension but are familiar with hypertension it can be helpful to put “**pulmonary**” in all caps or underlined.
 - Information about what your normal vital sign ranges are, especially blood pressure and pulse ox levels. For example, an ambulance crew knowing that your blood pressure is normally 90/50 and that they should not immediately give you a lot of fluids can keep them from unknowingly giving you too many fluids.

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- If you have difficulty with getting accurate pulse ox levels on your fingers due to diseases such as Raynaud's, put this on the paper as well. Anything you can express to a medical provider of what is normal for you (that would be very abnormal to someone else) helps direct your treatment appropriately.
- Contact name and number of a trusted friend or family member that can help advocate for you.
- It is also helpful to have a similar piece of paper in your wallet by your ID.
- Have information on your fridge printed on a brightly colored piece of paper titled "**Attention: EMS**" – this should tell providers where your medication and information is kept if you are not able to speak for yourself.
- Going to the Emergency Department by private vehicle:
 - Contact your PH provider to find out if they can call the ED prior to your arrival.
 - Bring your PH-specialty medications with you as it is unlikely the hospital will have these available, or at least won't immediately.
 - Bring a copy of your home paperwork mentioned above to give to the emergency department staff.
- Many times, you will have care delivered for non-PH related issues in hospitals without a PH specialist. **You must be equipped and ready to advocate on your own behalf.**
- Have a small stockpile of necessary medications and supplies (ice packs, inhalation devices used to administer medications and pump- if you are on prostacyclin therapy) in a packet ready to go out the door with you in an emergency. **Include package inserts for the PH medications so that they will be readily available to emergency personnel and doctors.**
 - Make sure you know who to contact for pump problems if you are on IV or SQ prostacyclin therapy. In most cases, contact information is immediately available on the side of the pump. Have a back-up pump at all times.
- In an emergency, give the emergency caregivers a clear, concise explanation of your diagnosis and situation. Let them get a handle on the situation. But also give them direction (e.g. Do not interrupt prostacyclin infusion. If necessary, transfer to a peripheral IV and attach your pump). Please have a list of medications in your wallet. Include all PH medications and doses. This is especially important with IV or SC prostacyclin. Write down and know your dose, **not just** your pump rate.
- **Don't be afraid to remind the emergency providers that your IV PH medications cannot be stopped or changed for any reason. Ask them to call your PH specialist to discuss changes to any of your other PH medications.**
- Ask your doctor if it is appropriate for you to be transferred to the hospital where your PH specialist works.
- Don't forget that not every problem in a PH patient is due to PH. A thorough review of all possibilities should not be neglected.



How do I advocate for myself or my family member during a PH emergency, so that my/his/her personal wishes/goals are being respected?

- Discuss your wishes with a trusted family member or friend in advance and know who will be designated to make important decisions for you if you are too sick to make them yourself. Share your physician orders of life sustaining treatment (POLST) and advance directives with your chosen advocate.
- Make this person your appointed health care agent or durable power of attorney (DPOA), which helps your doctors to know that this advocate is speaking with your expressed interests at heart.
- Talk about the status of your PH regularly with your PH doctor. If your disease is worsening despite medications, are there non-medical options for additional treatment that would be appropriate in your case? Would these options be consistent with your overall goals/wishes?
 - Have your POLST and advanced directives on file with your PH doctor's office.
 - Know your "PH emergency" plan and speak up if something does not seem right.
 - You and your family are the best advocates for your health. Be involved with your care.