



# Emergency 101

## Preparedness Information for Patients

Because pulmonary hypertension is a rare disease many hospitals, physicians, nurses and emergency medical technicians (EMTs) may not fully understand your condition. Preparedness is your first line of defense in minimizing potential problems and avoiding emergencies. Use this guide to help yourself get prepared.

Visit [www.PHAssociation.org](http://www.PHAssociation.org) and learn more about PH and your health.

### Health & Safety Tips

Keep this important information in mind to stay a step ahead of an emergency.

#### Infusion Tips

- » Never leave home without a thermometer. Take your temperature if you think you have a fever, since it could be a symptom of infection.
- » If you are taking medication through an infusion pump, keep your back-up pump in your kit. Be prepared to change pumps if necessary.
- » If you have a central IV line (catheter) keep a catheter repair kit with you, make sure it matches your specific catheter and be aware of the expiration date. A physician will need to repair the catheter.
- » A blood pressure monitor is invaluable when you need to provide vital information to your doctor or clinic over the phone.
- » A small portable cooler box or bag and some large ice packs will keep your medicine cool for several hours. Make sure there is room in the cooler for several ice packs and your medicine.

#### Oxygen Basics

- » Ask your doctor to write an oxygen prescription on a prescription pad and keep a photocopy in your medicine bag.
- » A Portable E-tank (other sizes too) can be a lifesaver during a power outage. You will need a regulator for portable tanks to control the flow.
- » Keep a spare nasal cannula in your kit, since they can wear out quickly and are easy to damage. Some portable tanks require special cannulas.
- » Have an extra section of oxygen tubing and extra connectors with you, even when you go to the hospital.
- » If you suffer from sleep apnea, you must take your CPAP or bipap machine, mask, tubing, humidifier, etc. with you to the hospital. Remember your oxygen enrichment (bleed) connector if you use one.

Share the information on the back of this flier  
with your local EMS!

### Readiness Checklist

If you can check off each of these items, you're well on your way to being Emergency Ready!

I wear a medic alert identifier to convey critical health information. I've included "Pulmonary Arterial Hypertension," and such applicable information as "Do Not Stop Pump," "On Sildenafil - No Nitrates" and/or "Takes Coumadin"

I always carry a cell phone and/or let people know where I am going if I am traveling alone. I have signed up for a roadside assistance program.

I maintain a current list of all of my medications. I've included concentrations, doses and rates for infusion and/or oxygen therapy.

I have a three day supply of all my medications and supplies. I keep it with me at all times.

I keep a current list of emergency contact information.

I am involved with my PH center in teaching local EMT/hospital personnel about PH, my therapy and how to respond in an emergency.

I have consulted my PH-treating medical professional and s/he has assured me that I have all the items I need.



Join PHA and become part of our community.  
Call 301-565-3004 x767 or e-mail  
[membership@PHAssociation.org](mailto:membership@PHAssociation.org), or log on to  
[www.PHAssociation.org/Join](http://www.PHAssociation.org/Join)

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## Information for Emergency Medical Professionals



**What Is Pulmonary Hypertension?** Pulmonary Arterial Hypertension (PAH) is a rare disorder of the blood vessels in the lungs. The pulmonary arteries become narrowed and the pressure in the arteries rises above normal limits. This causes strain on the heart and may become life threatening. PH may be associated with other disease states or may be idiopathic.

**What can I expect from PH Patients?** As with any population, PH patients vary greatly; some may not look sick at all while others are in wheel chairs, require oxygen or have medicine continuously delivered by a pump (more on medications below). PH patients may need emergency assistance because of complications associated with their pulmonary hypertension, an interruption of medicine delivery for those on IV therapy, or because of an accident or natural disaster.

**PH patients may be on one or more of the following medications to treat PH\*:**

- » Epoprostenol (Flolan®) intravenous (discussed below)
- » Treprostinil (Remodulin®) subcutaneous or intravenous (discussed below)
- » Iloprost (Ventavis®) inhaled
- » Bosentan (Tracleer®) oral
- » Letairis (Ambrisentan®) oral
- » Sildenafil (Revatio®) oral
- » Tadalafil (Adcirca®) oral (available fall 2009)

\* **No PH medication should ever be stopped or dosage changed during a hospitalization without first consulting the PH-treating physician.**

### Flolan® and Remodulin® Emergencies

**Flolan®** is a PH drug given by continuous intravenous infusion through a permanent catheter placed in one of the large veins going to the heart. It is given this way because epoprostenol only lasts in the bloodstream for 3-5 minutes. A pump (the CADD Legacy pump or the CRONO Five) is used to deliver the drug.

**Subcutaneous Remodulin®** is delivered via the Mini-Med portable infusion pump or the CADD MS-3. Each infusion site is generally used for up to three days, although some patients use the same site for a longer period of time. **Intravenous Remodulin®** must be administered via a surgically placed central venous catheter. Intravenous Remodulin® is delivered via the CADD Legacy pump or the CRONO Five pump. The half-life of Remodulin® is about 4.5 hours, although symptoms due to drug interruption can occur in less than one hour.

**If a patient is on Flolan® or Remodulin® via intravenous delivery,  
and the catheter comes out or is damaged, or the pump stops working, it is an emergency.**

### If you are providing emergency care to a patient on Flolan® or Remodulin®:

- » **IF THERE IS A PROBLEM WITH THE LINE OR PUMP, YOU MUST START AN IV IN THE PATIENT'S ARM.** When the IV line is in place, screw the pump tubing directly to the IV and make sure the pump is running. Pump tubing should be connected to as little IV tubing as possible.
- » **DO NOT TURN OFF THE PUMP.** This action could be fatal.
- » **DO NOT PRIME THE IV LINE.** A bolus of too much medicine is as dangerous as too little and can be fatal.
- » **DO NOT SWITCH PUMPS,** the calibration (infusion rate) is not correct on a "standard" pump. If you have questions or concerns, call the patient's PH specialist or center before changing the pump.
- » **DO NOT TAKE A BLOOD DRAW FROM THE IV** unless there is a dual lumen and the second lumen is used to draw blood.
- » **NOTE THAT** fever and/or drainage from the catheter in patients on IV therapies may indicate a sudden and serious onset of a line infection.
- » **NO MEDICATIONS CAN BE INFUSED WITH FLOLAN OR REMODULIN.** A second peripheral IV is required if additional medications or intravenous fluids are needed.
- » **DO NOT** give a large bolus of IV fluids (this may worsen heart failure).
- » **NOTIFY THE PH SPECIALIST** of the patient's problem and condition.

Visit the Pulmonary Hypertension Association website at [www.PHAssociation.org](http://www.PHAssociation.org) or call 301-565-3004 for more information.