HEALTH & SAFETY TIPS

Preparedness is your first line of defense to minimize potential problems and avoid emergencies. Use these tips to guide you.

Infusion Tips
- 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 and all supplies in your medicine kit.
- If you have a central IV line (catheter), keep all of your site cleaning and dressing supplies with you. This includes gloves, a mask, alcohol pads, dressings and tape.
- A small portable cooler box or bag and some large ice packs will keep epoprostenol (Flolan®) cool for several hours. Make sure there is room in the cooler for several ice packs and your medicine.

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 have my pump or inhalation device, as well as batteries and appropriate power sources.
- I keep my specialty pharmacy’s 24/7 emergency number with me so emergency personnel can call if needed.
- 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 my medications.
- I let my PH center know if I have a planned vacation so they can identify the best hospital for me to go to in case of an emergency.
- I have a three to seven day supply of all my medications and supplies, depending on how soon I can refill my prescriptions based the rules of my insurance plan.
- I keep a current list of emergency contact information including a health care provider and personal caregiver.
- I have discussed an emergency plan with my PH provider.

For additional tips, visit www.PHAssociation.org/Patients/EmergencySituations
What is pulmonary arterial 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 right side of the heart and may become life threatening. PAH may be either associated with other disease states or exist alone for no known reason.

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

- Epoprostenol (Flolan® and Veletri®) intravenous (discussed below)
- Treprostinil (Remodulin®) subcutaneous or intravenous (discussed below)
- Treprostinil (Tyvaso®) inhaled
- Iloprost (Ventavis®) inhaled
- Treprostinil (Orenitram®) oral
- Selexipag (Uptravi®) oral
- Ambrisentan (Letairis®) oral
- Bosentan (Tracleer®) oral
- Macitentan (Opsumit®) oral
- Sildenafil (Revatio®) oral
- Tadalafil (Adcirca®) oral
- Riociguat (Adempas®) oral

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

Flolan®, Veletri® and Remodulin® Emergencies

Intravenous Flolan® and Veletri® are given by continuous infusion through a central venous catheter. The half-life of these medications is expected to be no greater than six minutes. They are delivered by a CADD Legacy pump. Intravenous Remodulin® is given by continuous infusion through a central venous catheter. A CADD Legacy, CRONO 5 or CADD MS-3 pump is used to deliver this medication.

The half-life of Remodulin® is approximately 4 hours, although withdrawal symptoms such as shortness of breath can occur in less time.

Subcutaneous Remodulin® is given by continuous subcutaneous infusion delivered via the CADD MS-3 pump. The infusion site may be reddened. This is normal due to vasodilation.

When providing emergency care to a patient on Flolan®, Veletri® or Remodulin®:

DO NOT TURN OFF THE PUMP. This action could be fatal.
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 PRIME OR FLUSH THE IV LINE. A large volume (bolus) of too much medication is as dangerous as too little and can be fatal.
DO NOT INFUSE ANY OTHER MEDICATION WHERE THE PAH MEDICATION IS INFUSING. A second peripheral IV is required if additional medications or intravenous fluids are needed.
DO NOT GIVE A LARGE VOLUME (BOLUS) OF IV FLUIDS (this may cause/worsen heart failure).
DO NOT CHANGE TO A DIFFERENT INFUSION PUMP without first talking to the patient’s PAH specialist.

DO NOT DRAW BLOOD 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.
INVOKE A PAH SPECIALIST as soon as time allows.

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

Visit the Pulmonary Hypertension Association website at www.PHAssociation.org or call 301-565-3004 for more information.