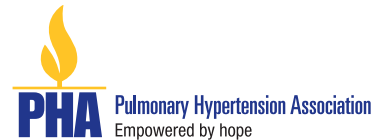


# Emergency 101

## 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. PAH may be either idiopathic or associated with other disease states.

### What can I expect from PAH patients?

As with any population, PAH patients vary greatly; some may not look sick at all, while others are in wheelchairs, require oxygen or have medicine continuously delivered by a pump (more on medications below). PAH patients may need emergency assistance because of complications associated with their PAH, an interruption of medicine delivery for those on IV therapy, or because of an accident or natural disaster.

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

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

\* No PAH medication should ever be stopped or dosage changed during a hospitalization without first consulting the PAH-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 about three to five 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 Five or CADD MS-3 pump is used to deliver this medication. The half-life of Remodulin® is about 4.5 hours, although withdrawal symptoms can occur in less than one hour.
- » **Subcutaneous Remodulin®** is given by continuous subcutaneous infusion delivered via the CADD MS-3 pump. The infusion site may be reddened. This is normal.

**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.**

### 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 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 BOLUS OF IV FLUIDS** (this may 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.
- » **NOTIFY THE PAH 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.