

Information for EMS and Emergency Department Professionals

Pulmonary arterial hypertension (PAH) is a rare disorder of the blood vessels in the lungs. As pulmonary arteries narrow, blood pressure in the arteries rises above normal limits. This causes strain on the right side of the heart and can be life threatening. As with any population, PAH patients vary greatly. Some might not look sick, while others need wheelchairs, oxygen or medicine continuously delivered by a pump (see below).

- **Listen** to the patient.
- **Don't** administer nitroglycerin for chest pain.
- **Never** remove or stop an infusion line.

PH Medication List

Patients might be on one or more medications to treat PH. Bring the medications listed below to the hospital with the patient:

PAH Medication

Dosage/Infusion Rates

- | | |
|---|-------|
| <input type="checkbox"/> Ambrisentan (Letairis®) oral | _____ |
| <input type="checkbox"/> Bosentan (Tracleer®) oral | _____ |
| <input type="checkbox"/> Tadalafil/ Alyq (Adcirca®) oral | _____ |
| <input type="checkbox"/> (Orenitram®) oral | _____ |
| <input type="checkbox"/> Selexipag (Uptravi®) oral | _____ |
| <input type="checkbox"/> Macitentan (Opsumit®) oral | _____ |
| <input type="checkbox"/> Sildenafil (Revatio®) oral | _____ |
| <input type="checkbox"/> Riociguat (Adempas®) oral | _____ |
| <input type="checkbox"/> Treprostinil (Tyvaso®) inhaled | _____ |
| <input type="checkbox"/> Iloprost (Ventavis®) inhaled | _____ |
| <input type="checkbox"/> Treprostinil (Remodulin®) subcutaneous or intravenous* (discussed below) | |
| <input type="checkbox"/> Epoprostenol (Flolan® and Veletri®) intravenous* (discussed below) | |

If a patient is on intravenous (IV) Flolan, Veletri or Remodulin, and the catheter is dislodged or damaged, or if the pump stops working, **it is an emergency.**

About PH Medication

IV epoprostenol (also known as Flolan and Veletri) are given by continuous infusion through a central venous catheter and pump. The half-life of these medications is three to five minutes.

IV Remodulin is delivered by continuous infusion through a central venous catheter and pump. The half-life of Remodulin is about four hours, although symptoms such as shortness of breath can occur in less time.

Subcutaneous Remodulin is delivered through continuous infusion with a 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

- Don't turn off the pump. It could be fatal.
- Involve a PAH specialist as soon as possible.
- If there is a problem with the line or pump, start an IV in the patient's arm. When the IV line is in place, attach the pump tubing directly to the IV, and make sure the pump is running. Pump tubing should be connected to as short as possible IV tubing.
- Don't prime or flush the IV line. Don't give a large volume (bolus) of IV fluids. It can worsen heart failure.
- Don't infuse other medication into the PH medication line. Use a second peripheral IV for additional medications or IV fluids.
- Don't change the infusion pump without first talking to the patient's PAH specialist (unless patient is capable).
- Don't draw blood from the IV.
- Note: Fever and/or drainage from the catheter can indicate a sudden, serious line infection onset.

Allergies or known drug reactions: _____

Normal oxygen saturation range for this patient: _____

Oxygen needs and tank settings: _____

Additional Medications

Medical Condition	Prescription Name	Dosage

Emergency Contacts

	Name	Address	Phone
PAH Specialist			
Specialty Pharmacy			
Emergency Contact 1			
Emergency Contact 2			