

## Pulmonary Arterial Hypertension Emergency Complications and Evaluation: Practical Guide for the Advanced Practice Registered Nurses in the Emergency Department

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**Background:** Pulmonary hypertension (PH) complicates common disease and can lead to worsening symptoms and increased mortality. A specific group of PH, pulmonary arterial hypertension (PAH), World Health Organization Group 1, may present to the emergency department (ED).

**Methods:** We review common ED presentations of patients with PAH such as cardiac arrest/sudden death, right ventricular failure, syncope, hypoxemic respiratory failure, arrhythmias, hemoptysis, pulmonary embolism, chest pain/left main compression syndrome, infection, and considerations for PAH medication management.

**Results:** We include a case study to illustrate a real example with a positive outcome, and an algorithm for evaluation and triaging patients with PAH in the ED.

**Conclusions:** The ability to recognize, triage, and communicate changes in PAH disease status in a multidisciplinary team approach between the patient, family, specialty pharmacy, and specialized health care providers such as the PH team, is essential for ED providers who are evaluating and treating patients with PAH.


Figure 1. Table 1. PAH-specific medications

Target Pathway	Drug	Route	Typical Dose	Half-life	Available at Local vs Specialty Pharmacy	Risks of Interruption, Special considerations	Resources	Pump Name	Photo of Pump
Nitric Oxide	Sildenafil* (Revatio®) <sup>31</sup>	oral	20mg TID	4 hours	Insurance dependent	All: Do not administer nitrates	Pfizer 1-800-879- 3477		
	Tadalafil (Adcirca®) <sup>32</sup>	oral	20mg or 40mg daily	35 hours	Insurance dependent	Do not hold doses unless speaking with PH treatment team	Adcirca: 1-800- 545-5979		
	Riociquat (Adempas®)** <sup>33</sup>	oral	0.5mg to 2.5mg TID	12 hours	Specialty	Missed doses of riociquat may require re-titration	Bayer: 1-888- 842-2937		

						to lower dose. Riociquat requires monthly pregnancy test for females of CBP	
Endothelin	Bosentan (Tracleer®) <sup>34</sup>	oral	62.5mg or 125mg BID	5 hours	Female: Specialty Male: Insurance dependent	All require monthly pregnancy test for females of CBP. Monitor for signs and symptoms of edema and anemia Tracleer requires monthly LFTs	Actelion PH Pathways: 1-866-228-3546
	Ambrisentan (Letairis®) <sup>35</sup>	oral	5mg or 10mg daily	9 hours	Specialty		Gilead LEAP Program: 1-866-664-5327
	Macitentan (Opsumit®) <sup>36</sup>	oral	10mg daily	16 hours	Specialty		Actelion PH Pathways: 866-228-3546
Prostacyclin	Selexipag (Uptravi®) <sup>37</sup>	oral	200-1600mcg BID	6.2-13.5 hours of the active metabolite	Specialty	Initiation by dose titration	Actelion PH Pathways: 1-866-228-3546
	Treprostinil (Orenitram®) <sup>25</sup>	oral	Variable, tablets supplied in 0.125mg, 0.25mg, 1mg, 2.5mg, and 5mg per label	4 hours	Specialty		United Therapeutics: 1-877-864-8437



Iloprost (Ventavis®) <sup>38</sup>	Inhaled 6-9 times daily	2.5 or 5mcg	20-30 minutes	Specialty	Must be administered via pump I-neb® AAD® device	Actelion 1-866- 228-3546	I-neb® AAD®	
Treprostinil (Tyvaso®) <sup>39</sup>	Inhaled 4 times day	1-9 breaths (6 mcg per breath) in 4 separate treatment daily, 4 hours apart	4 hours	Specialty	Must be administered via pump Tyvaso Inhalation System® device	United Therapeutics: 1- 877-864-8437	TD-100®-ir Tyvaso Inhalation System	
Treprostinil (Remodulin®) <sup>40</sup>	Continuous IV or SQ infusion	Dose is ng\kg\min. Highly variable and patient specific.	4 hours	Specialty	For IV prostacyclins: do not abruptly stop infusions and maintain as dedicated line. Do not push or mix anything without proper protocol (see section <i>PAH Infusion Medication Compromise</i> )		CADD-Legacy® 1 CRONO five CADD MS™ 3	
Epoprostenol (Flolan®) <sup>41</sup>	Continuous IV Infusion	Dose is ng\kg\min. Highly variable and patient specific.	6 minutes	Specialty		GlaxoSmithKline 1-888-825-5249	CADD-Legacy® 1	

	Epoprostenol (Veletri®) <sup>42</sup>	Continuous IV infusion	Dose is ng\kg\min. Highly variable and patient specific.	6 minutes	Specialty		Actelion 1-866-228-3546	CADD-Legacy® 1	
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\*IV formulation of sildenafil for use when tablets cannot be given. Dose conversion is half the normal oral dose.

\*\*FDA approval for both Group 1 PAH and Group 4 CTEPH

Figure 2. Figure 1. Algorithm for evaluating and triaging the patient with pulmonary arterial hypertension in the emergency department

