**FAST FACTS AND CONCEPTS #264**  
**PROSTACYCLIN WITHDRAWAL IN PULMONARY HYPERTENSION**

Christi Bartlett MD and Lindy Landzaat DO in cooperation with the COPE Collaborative*

**Background**  
Pulmonary artery hypertension (PAH) is a disease of the pulmonary vascular system characterized by elevated pulmonary vascular resistance. Patients with advanced PAH are often treated with prostacyclins. This Fast Fact reviews suggested guidelines for withdrawal of prostacyclin therapy in dying patients with PAH.

**Physiology/Symptoms**  
PAH is defined as a mean pulmonary arterial pressure greater than 25 mmHg and may lead to right ventricular failure (1). Symptoms include dyspnea, cough, syncope, fatigue, angina, and peripheral edema, often progressing to respiratory failure and cardiovascular collapse.

**Prostacyclin Treatment**  
Prostacyclin medications, epoprostenol and treprostinil, are effective in extending life in PAH (2). These drugs are administered via continuous parenteral infusion (IV; treprostinil can be given subcutaneously). Epoprostenol's half life is six minutes (3); abrupt withdrawal can lead to significant symptom burden (dyspnea, anxiety, nausea, light-headedness, chest and abdominal pain) and rapid death.

**Discontinuation of Prostacyclin Therapy**  
Despite the use of prostacyclins and other life-prolonging treatments, PAH is usually progressive and fatal without lung transplantation. When the goals of care include discontinuation of prostacyclins, this process can create anxiety among patients and caregivers due to concerns about the rapid development of distressing symptoms. Because of this, it is felt best to taper prostacyclins in a planned, controlled manner. There are no guidelines regarding how to taper off IV prostacyclins, let alone any research data to guide clinicians. The following recommendations, however, have been developed by the COPE Collaborative * and are based on clinical experience and rational pharmacology.

- A benzodiazepine and/or opioid should be given as premedication and be readily available for rapid administration following initiation of withdrawal of prostacyclin therapy. Actual doses however need to be closely tailored to an individual patient’s prior exposure to opioids and benzodiazepines and acceptable level of consciousness. See Fast Fact #34 (regimen A) for some general guidelines.

- The rate of medication taper should account for the medication’s half-life. Because epoprostenol has a short half life, we recommend waiting 4-6 half lives (generally 25-30 minutes) and monitoring for symptoms prior to further titration. Treprostinil, with a half life of approximately 4 hours, can be titrated downward every 4-6 hours.

- Reduce the prostacyclin in 20-25% increments, with close monitoring for symptoms as a new steady state is achieved.

- With each dose reduction, the patient should be closely monitored for increasing symptoms. If symptoms are minimal or otherwise well controlled with opioids or benzodiazepines, the prostacyclin taper may continue. If the patient experiences distressing symptoms (dyspnea, anxiety, chest pain), the taper should be held until symptoms are managed and the taper resumed at a slower rate, with smaller interval dose reductions. Conversely, if the patient is unresponsive and is tolerating the titration well with minimal symptom burden, the rate of titration can be cautiously increased.

- Similar to the planned discontinuation of mechanical ventilation in a dying patient (see Fast Facts #33-35), it is recommended that the de-escalation of IV prostacyclins occur in the inpatient setting under close physician supervision. In the home setting, a physician should be present at the bedside along with the support of a knowledgeable hospice team, with ample medications readily available in-home, as symptoms can develop quickly and patients often need rapid medication adjustments to maintain comfort.

*Care Of Pulmonary hypertension patients at End-of-life (COPE) Collaborative members: Christi Bartlett MD, Lindy Landzaat DO, Karin Porter-Williamson MD, Lewis Satterwhite MD, Leslie Spikes MD, Ryan Westhoff MD, Tim Williamson MD; University of Kansas, Kansas City, KS.
References

Authors' Affiliation: University of Kansas Medical Center, Kansas City, KS.


Fast Facts and Concepts are edited by Sean Marks MD (Medical College of Wisconsin) and associate editor Drew A Rosielle MD (University of Minnesota Medical School), with the generous support of a volunteer peer-review editorial board, and are made available online by the Palliative Care Network of Wisconsin (PCNOW); the authors of each individual Fast Fact are solely responsible for that Fast Fact’s content. The full set of Fast Facts are available at Palliative Care Network of Wisconsin with contact information, and how to reference Fast Facts.

Copyright: All Fast Facts and Concepts are published under a Creative Commons Attribution-NonCommercial 4.0 International Copyright (http://creativecommons.org/licenses/by-nc/4.0/). Fast Facts can only be copied and distributed for non-commercial, educational purposes. If you adapt or distribute a Fast Fact, let us know!

Disclaimer: Fast Facts and Concepts provide educational information for health care professionals. This information is not medical advice. Fast Facts are not continually updated, and new safety information may emerge after a Fast Fact is published. Health care providers should always exercise their own independent clinical judgment and consult other relevant and up-to-date experts and resources. Some Fast Facts cite the use of a product in a dosage, for an indication, or in a manner other than that recommended in the product labeling. Accordingly, the official prescribing information should be consulted before any such product is used.