AETNA BETTER HEALTH <sup>®</sup> Coverage Policy/Guideline					
Name:	Epoprostenol		Page:	1 of 4	
Effective Date: 12/21/2023		Last Review Date:	11/2023		
Applies	⊠Illinois	□Florida	□Florida Kids		
Applies to:	□New Jersey	□Maryland	□Michigan		
10.	🗆 Pennsylvania Kids	□Virginia	□Kentucky PRMD		

## Intent:

The intent of this policy/guideline is to provide information to the prescribing practitioner outlining the coverage criteria for epoprostenol under the patient's prescription drug benefit.

## **Description:**

## FDA-Approved Indication

Epoprostenol/Flolan/Veletri is indicated for the treatment of pulmonary arterial hypertension (PAH) (WHO Group I) to improve exercise capacity. Studies establishing effectiveness included predominantly patients with NYHA Functional Class III-IV symptoms and etiologies of idiopathic or heritable PAH or PAH associated with connective tissue diseases.

All other indications are considered experimental/investigational and not medically necessary.

# Applicable Drug List:

Epoprostenol Flolan Veletri

## **Policy/Guideline:**

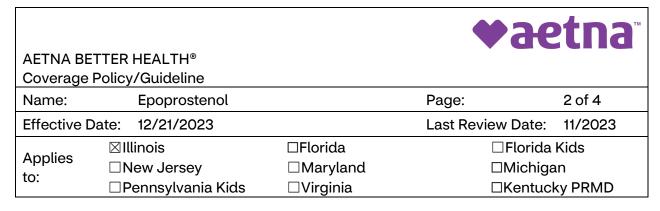
## **Prescriber Specialty:**

This medication must be prescribed by or in consultation with a pulmonologist or cardiologist.

# Criteria for Initial Approval: Pulmonary Arterial Hypertension (PAH)

Authorization of 12 months may be granted for treatment of PAH when ALL of the following criteria are met:

- A. Member has PAH defined as WHO Group 1 class of pulmonary hypertension (refer to Appendix).
- B. PAH was confirmed by either criterion (1) or criterion (2) below:
  - 1. Pretreatment right heart catheterization with all of the following results:
    - i. mPAP > 20 mmHg
    - ii. PCWP ≤ 15 mmHg



- iii. Pulmonary vascular resistance (PVR)  $\geq$  3 Wood units in adult patients or pulmonary vascular resistance index (PVRI)  $\geq$  3 Wood units x m<sup>2</sup> in pediatric patients
- 2. For infants less than one year of age, PAH was confirmed by Doppler echocardiogram if right heart catheterization cannot be performed.
- C. Requests for Veletri: Patient is unable to take Flolan and epoprostenol for the given diagnosis due to a trial and inadequate treatment response or intolerance, or a contraindication.

# Criteria for Continuation of Therapy:

Authorization of 12 months may be granted for members with an indication listed in criteria for initial approval who are currently receiving the requested medication through a paid pharmacy or medical benefit, and who are experiencing benefit from therapy as evidenced by disease stability or disease improvement.

# Appendix

# WHO Classification of Pulmonary Hypertension 1 PAH

- 1.1 Idiopathic (PAH)
- 1.2 Heritable PAH
- 1.3 Drug- and toxin-induced PAH
- 1.4. PAH associated with:
  - 1.4.1 Connective tissue diseases
  - 1.4.2 HIV infection
  - 1.4.3 Portal hypertension
  - 1.4.4 Congenital heart diseases
  - 1.4.5 Schistosomiasis
- 1.5 PAH long-term responders to calcium channel blockers
- 1.6 PAH with overt features of venous/capillaries (PVOD/PCH) involvement
- 1.7 Persistent PH of the newborn syndrome

## 2 PH due to left heart disease

- 2.1 PH due to heart failure with preserved LVEF
- 2.2 PH due to heart failure with reduced LVEF
- 2.3 Valvular heart disease
- 2.4 Congenital/acquired cardiovascular conditions leading to post-capillary PH



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### 3 PH due to lung diseases and/or hypoxia

- 3.1 Obstructive lung disease
- 3.2 Restrictive lung disease
- 3.3 Other lung disease with mixed restrictive/obstructive pattern
- 3.4 Hypoxia without lung disease
- 3.5 Developmental lung disorders

### 4 PH due to pulmonary artery obstruction

- 4.1 Chronic thromboembolic PH
- 4.2 Other pulmonary artery obstructions
  - 4.2.1 Sarcoma (high or intermediate grade) or angiosarcoma
  - 4.2.2 Other malignant tumors Renal carcinoma Uterine carcinoma Germ cell tumours of the testis Other tumours
  - 4.2.3 Non-malignant tumours Uterine leiomyoma
  - 4.2.4 Arteritis without connective tissue disease
  - 4.2.5 Congenital pulmonary artery stenosis
  - 4.2.6 Parasites
    - Hydatidosis

### 5 PH with unclear and/or multifactorial mechanisms

5.1 Hematologic disorders: Chronic hemolytic anemia, myeloproliferative disorders 5.2 Systemic and metabolic disorders: Pulmonary Langerhans cell histiocytosis, Gaucher disease, glycogen storage disease, neurofibromatosis, sarcoidosis

5.3 Others: chronic renal failure with or without hemodialysis, fibrosing mediastinitis 5.4 Complex congenital heart disease

### Approval Duration and Quantity Restrictions: Approval: 12 months

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