Epoprostenol for Continuous Intravenous Infusion(Flolan®/Veletri®)

IMPORTANT REMINDER

We develop Medical Policies to provide guidance to Members and Providers. This Medical Policy relates only to the services or supplies described in it. The existence of a Medical Policy is not an authorization, certification, explanation of benefits or a contract for the service (or supply) that is referenced in the Medical Policy. For a determination of the benefits that a Member is entitled to receive under his or her health plan, the Member's health plan must be reviewed. If there is a conflict between the medical policy and a health plan or government program (e.g., TennCare), the express terms of the health plan or government program will govern.

The proposal is to add text/statements in red and to delete text/statements with strikethrough:

POLICY

INDICATIONS

The indications below including FDA-approved indications and compendial uses are considered a covered benefit provided that all the approval criteria are met and the member has no exclusions to the prescribed therapy.

FDA-Approved Indications

Flolan/Veletri/epoprostenol is indicated for the treatment of pulmonary arterial hypertension (PAH) (World Health Organization [WHO] Group I) to improve exercise capacity. Studies establishing effectiveness included predominantly patients with New York Heart Association (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.

PRESCRIBER SPECIALTIES

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

COVERAGE 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:

- Member has PAH defined as WHO Group 1 class of pulmonary hypertension (refer to Appendix).
- PAH was confirmed by either of the following criteria: ~~criterion (1) or criterion (2) below:~~
  - Pretreatment right heart catheterization with all of the following results:
    - Mean pulmonary arterial pressure (mPAP) > 20 mmHg
    - Pulmonary capillary wedge pressure (PCWP) ≤ 15 mmHg
    - Pulmonary vascular resistance (PVR) >2 ~~≥ 3~~ Wood units. ~~in adult patients or~~ For pediatric members, pulmonary vascular resistance index (PVRI) > ~~≥~~ 3 Wood units x m^2 is also acceptable. ~~in pediatric patients~~
  - For infants less than one year of age, PAH was confirmed by Doppler echocardiogram if right heart catheterization cannot be performed.

CONTINUATION OF THERAPY

Authorization of 12 months may be granted for members with an indication listed in the coverage criteria section ~~III~~ 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 (PH)

Note: Patients with heritable PAH or PAH associated with drugs and toxins might be long-term responders to calcium channel blockers.

Group 1: Pulmonary Arterial Hypertension PAH

- Idiopathic ~~(PAH)~~
  - Long-term responders to calcium channel blockers
- Heritable ~~PAH~~
- Associated with drugs and toxins ~~-induced PAH~~
- Associated with:
  - Connective tissue disease~~s~~
  - Human immunodeficiency virus (HIV) infection
  - Portal hypertension
  - Congenital heart disease
  - Schistosomiasis

  ~~1.5 PAH long-term responders to calcium channel blockers~~
- PAH with ~~overt~~ features of venous/capillary (pulmonary veno-occlusive disease [PVOD]/pulmonary capillary hemangiomatosis [PCH]) involvement
- Persistent PH of the newborn ~~syndrome~~

Group 2:PH associated with ~~due to~~ Left Heart Disease

- ~~PH due to~~ Heart failure:
  - With preserved ~~left ventricular~~ ejection fraction ~~(LVEF)~~
  - ~~PH due to heart failure~~ With reduced or mildly reduced ejection fraction ~~LVEF~~
  - Cardiomyopathies with specific etiologies (i.e., hypertrophic, amyloid, Fabry disease, and Chagas disease)
- Valvular heart disease
  - Aortic valve disease
  - Mitral valve disease
  - Mixed valvular disease
- Congenital/acquired cardiovascular conditions leading to post-capillary PH

Group 3: PH associated with ~~due to~~ Lung Diseases and/or Hypoxia

- Chronic obstructive pulmonary disease (COPD) and / or emphysema ~~lung disease~~
- Interstitial ~~Restrictive~~ lung disease
- Combined pulmonary fibrosis and emphysema
- Other parenchymal lung diseases (i.e., parenchymal lung diseases not included in Group 5) ~~with mixed restrictive/obstructive pattern~~
- Nonparenchymal restrictive diseases:
  - Hypoventilation syndromes
  - Pneumonectomy
- Hypoxia without lung disease (e.g., high altitude)
- Developmental lung diseases ~~disorders~~

Group 4: PH associated with ~~due to~~ Pulmonary Artery Obstructions

- Chronic thromboembolic PH
- Other pulmonary artery obstructions:
  - Sarcomas (high-or intermediate-grade~~)~~ or angiosarcoma)
  - Other malignant tumors (e.g., renal carcinoma, uterine carcinoma, germ-cell tumors of the testis)
  - Non-malignant tumors (i.e., uterine leiomyoma)
  - Arteritis without connective tissue disease
  - Congenital pulmonary artery stenoses
  - Hydatidosis
  - ~~Renal carcinoma~~
  - ~~Uterine carcinoma~~
  - ~~Germ cell tumours of the testis~~
  - ~~Other tumours~~
  - ~~Uterine leiomyoma~~

Group 5: PH with Unclear and/or Multifactorial Mechanisms

- Hematologic disorders, including inherited and acquired chronic hemolytic anemia and chronic ~~hemolytic anemia,~~ myeloproliferative disorders
- Systemic ~~and metabolic~~ disorders: Sarcoidosis, pulmonary Langerhans cell histiocytosis, ~~Gaucher disease, glycogen storage disease,~~ and neurofibromatosis type 1
- Metabolic disorders, including glycogen storage diseases and Gaucher disease
- ~~Others:~~ Chronic renal failure with or without hemodialysis ~~fibrosing mediastinitis~~
- Pulmonary tumor thrombotic microangiopathy
- Fibrosing mediastinitis
- Complex congenital heart disease

APPLICABLE TENNESSEE STATE MANDATE REQUIREMENTS

BlueCross BlueShield of Tennessee's Medical Policy complies with Tennessee Code Annotated Section 56-7-2352 regarding coverage of off-label indications of Food and Drug Administration (FDA) approved drugs when the off-label use is recognized in one of the statutorily recognized standard reference compendia or in the published peer-reviewed medical literature.

ADDITIONAL INFORMATION

For appropriate chemotherapy regimens, dosage information, contraindications, precautions, warnings, and monitoring information, please refer to one of the standard reference compendia (e.g., the NCCN Clinical Practice Guidelines in Oncology (NCCN Guidelines®) published by the National Comprehensive Cancer Network®, Drugdex Evaluations of Micromedex Solutions at Truven Health, or The American Hospital Formulary Service Drug Information).

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EFFECTIVE DATE

ID_CHS