Drug Name: Pulmonary Arterial Hypertension (PAH) Medications
Date: 09-2017

Drug Name:	Pulmonary Arterial Hypertension (PAH) Medications
Prescriber Restrictions:	• Prescriber must be a **pulmonologist** or a **cardiologist**.
Age Restrictions:	
Exclusion Criteria:	

Required Medical Information:	• The medication is being used for an FDA-approved functional class; *and*
	• The medication is being recommended and prescribed by a pulmonologist or a cardiologist at a dose that is within FDA approved guidelines; *and*
	• Documentation of a confirmed diagnosis of PAH World Health Group (WHO Group I); *and*
	• Documentation of the patient's current weight (if drug is dosed by weight); *and*
	• Documentation that the patient has undergone acute vasoreactivity testing and whether or not the results were favorable;
	  o For those patients who demonstrated a favorable response to the acute vasoreactivity testing (defined as a fall in mean pulmonary arterial pressure [mPAP] of at least 10 mmHg to less than 40 mmHg with an increased or unchanged cardiac output), documentation must be submitted that his/her pulmonary hypertension has progressed despite maximal medical treatment with a calcium channel blocker; *and*
	• Requests for Phosphodiesterase Type 5 (PDE5) Inhibitors (e.g. Adcirca) **OR** Guanylate Cyclase Stimulants (e.g. Adempas) require a documented failure or intolerance of a trial with sildenafil; *and*
	• Requests for Endothelin Receptor Antagonists (ERAs; e.g. Opsumit and Tracleer) require a documented failure of intolerance of a trial with Letaris; *and*
	• Requests for Prostanoids (e.g. Remodulin and Tyvaso) require a documented failure or intolerance of a trial with generic epoprostenol (Flolan) or Ventavis.
	  o Note: Ventavis is approvable for treatment of pulmonary artery hypertension (WHO Group 1) in patients with NYHA **Class III or IV** symptoms.

Required Medical Information (continued):	• **If the provider is requesting combination therapy with two agents:** o Documentation must be submitted of an adequate trial (defined by at least 3 months of adherent therapy) with monotherapy; *and* o Documentation must be submitted that the patient has been compliant with monotherapy; *and* o Documentation must be submitted that the patient has clinically deteriorated (e.g. worsening of the symptoms of dyspnea or fatigue, decline in functional class by at least one class, or an increase in 6-minute walk test (6MWD) by greater than 30 minutes) while on monotherapy. • **Requests for Uptravi**: o Patient must have a documented failure or intolerance of an adequate dose and for an appropriate duration; *or* o Patient is currently taking; *or* o Prescriber has provided a clinical rationale documenting why the member is unable to use other PAH therapies. • **Requests for dosage increases**: o Documentation must be submitted of the medical necessity to increase the dosage; *and* • **PA criteria for reauthorization for the following agents – <u>Phosphodiesterase Type 5 (PDE5) Inhibitors, Guanylate Cyclase Stimulants, Prostanoids</u>**: o Continued renewal is based on patient-improved outcomes and tolerance of drug.
Coverage Duration:	• **Initial**: o All orally administered agents: 3 months o Medically administered agents: 4 months • **Renewals**: 6 months