

Pulmonary Arterial Hypertension (PAH) Criteria

Revised: 06/26/2024

PULMONARY ARTERIAL HYPERTENSION (PAH)

Length of Authorization: Initial: 6 months

Renewal: 1 year

Website:

DIAGNOSIS OF PULMONARY ARTERIAL HYPERTENSION (PAH) (WORLD HEALTH ORGANIZATION [WHO] GROUP 1)

- Minimum age: 18 years
- Prescribed by, or in consultation with, a cardiologist or pulmonologist
- Patient has had a right heart catheterization (documentation required)
- Results of the right heart catheterization confirm the diagnosis of WHO Group 1 PAH
- Approve for one year.

DIAGNOSIS OF PULMONARY HYPERTENSION ASSOCIATED WITH INTERSTITIAL LUNG DISEASE (WORLD HEALTH ORGANIZATION [WHO] GROUP 3)

- **Note:** this involves diagnosis such as idiopathic interstitial pneumonia, combined pulmonary fibrosis and emphysema, WHO Group 3 connective disease, and chronic hypersensitivity pneumonitis.
- Minimum age: 18 years
- Prescribed by, or in consultation with, a cardiologist or pulmonologist
- Baseline forced vital capacity < 70% for patients with connective tissue disease Evidence of diffuse parenchymal lung disease on computed tomography of the chest
- Patient has had a right heart catheterization (documentation required).
- Results of the right heart catheterization confirm the diagnosis of WHO Group 3 interstitial lung disease associated with pulmonary hypertension.

ADDITIONAL CLINICAL INFORMATION TO AID IN THE FINAL DECISION

- The maximum recommended dose for Revatio® is 20 mg TID (no increased efficacy was shown with higher doses).
- **Revatio or Sildenafil** request for children < 18 years of age — document current weight and daily dose (in milligrams) and forward request to Clinical Pharmacist for potential MD review.
- For continuation of therapy requests, enter a 3-day DOS PA so child does not go without medication while request is reviewed.

REVISION HISTORY

Date	Issues/Updates
06/26/2024	<ul style="list-style-type: none">Initial draft creation