

Pulmonary Hypertension

At Healthfirst, we are committed to helping providers accurately document and code their patients' health records. This tip sheet is intended to assist providers and coding staff with the documentation and ICD-10-CM selection on services submitted to Healthfirst specifically for common types of **Pulmonary Hypertension (PH)**. It provides information from industry sources about proper coding practice. However, this document does not represent or guarantee that Healthfirst will cover and/or pay for services outlined. Coverage decisions are based on the terms of the applicable subscriber contract/ evidence of coverage and the provider's participation agreement. This includes the determination of any amount that Healthfirst or the member owes the provider.

Pulmonary Hypertension (PH) is high blood pressure in the lungs. PH is different than systemic high blood pressure/hypertension. Pulmonary blood pressure reflects the pressure the heart exerts to pump blood from the heart through the lung arteries. This is pressure that is focused on the blood flow in the lungs. Secondary PH is always caused by another condition.

Diagnosis Codes & Descriptions

ICD-10-CM	Description	PH Group
I27.0	Primary pulmonary hypertension; Heritable; Idiopathic	Group 1
I27.1	Kyphoscoliotic heart disease	Group 1
I27.20	Pulmonary hypertension, unspecified	Group 1
I27.21	Secondary pulmonary arterial hypertension	Group 1
I27.22	Pulmonary hypertension due to left heart disease	Group 2
I27.23	Pulmonary hypertension due to lung diseases and hypoxia	Group 3
I27.24	Chronic thromboembolic pulmonary hypertension	Group 4
I27.29	Other secondary pulmonary hypertension	Group 5

Documentation and Coding

Clinical Documentation Recommendations

Status of Condition	Stable; Improved; Worsening
Link Associated Conditions	• Due to, associated with, secondary to, or with
Signs & Symptoms	• Shortness of breath (dyspnea), • Swelling (edema) in the ankles, legs and eventually in the abdomen (ascites) • Racing pulse or heart palpitations • Fatigue • Dizziness or fainting spells (syncope) • Decreased appetite • Chest pressure or pain • Bluish color to your lips and skin (cyanosis) • Dry cough, sometimes with blood
Diagnostic Test	• Echocardiogram (Echo); Computerized Tomography (CT) scan • Electrocardiogram (ECG/EKG), Chest X-ray; Right Heart Catheterization (RHC) • Other: Blood tests; Ventilation/perfusion V/Q scans, cardiac MRI, exercise tests, lung biopsy
Complications	• Right ventricular dysfunction; Right heart failure. • Arrhythmia; Pericardial Effusion, Pulmonary Artery Aneurysm, Dissection, or Rupture; blood clot
Treatment	• Anticoagulant medicine (blood thinners) • Diuretics (water tablets) • Oxygen treatment • Pulmonary rehab exercise • Balloon Pulmonary Angioplasty (BPA) • Pulmonary Endarterectomy (PEA) • Lung transplant • Medications: Digoxin, Calcium; PDE5 Inhibitors (i.e Sildenafil, Tadalafil) • Vasodilators (i.e. Epoprostenol, Treprostinil, Iloprost, Selexipag)

Documentation and Coding

PH is classified into five groups based on categories that share similar pathological findings, hemodynamic characteristics, and management	
Group 1: Pulmonary Arterial Hypertension	Primary PH: Heritable, due to drugs and toxins, or associated with conditions such as connective tissue diseases, congenital heart disease, portal hypertension, and others
Group 2: PH Caused by Left Heart Disease	Left heart related, valve disorder, diastolic dysfunction, cardiomyopathy, left ventricular systolic dysfunction, left ventricular diastolic dysfunction, congenital cardiomyopathies, inflow and outflow obstruction of left heart not due to valvular disease, and valvular heart disease.
Group 3: PH Due to Lung Disease or Hypoxia	Due to chronic respiratory disorders, COPD that includes emphysema, chronic bronchitis, scarring in the lungs, and pulmonary fibrosis caused by lung disease are common diseases within this classification.
Group 4: Chronic Thromboembolic Pulmonary Hypertension	Chronic thromboembolic PH
Group 5: Other Secondary Pulmonary Hypertension	Unclear multifactorial mechanisms, including Hematologic disorders such as myeloproliferative disorders and splenectomy. Systemic disorders such as sarcoidosis and pulmonary langerhans cell histiocytosis; metabolic disorders such as glycogen storage disease, Gaucher disease and thyroid disorders; and other conditions that lead to ph

Coding Tips

According to CMS, Echocardiograms may be used for diagnoses abstraction as a stand-alone document provided it contains a final interpretation signed by a qualified physician. Reviewers should only submit diagnoses documented in the physician interpretation, not the technical report.

- If **Pulmonary Hypertension is confirmed on echocardiogram**, PH may be coded
 - Chronic conditions that are documented in an embedded echo report may be abstracted unless there is conflicting documentation within the current encounter.
- For **Secondary pulmonary arterial hypertension (I27.21)** if applicable code also any associated conditions:
 - Congenital heart disease (**Q20-Q28**); human immunodeficiency virus [HIV] disease (**B20**)
 - Polymyositis (**M33.2-**); portal hypertension (**K76.6**); rheumatoid arthritis (**M05.-**)
 - Schistosomiasis (**B65.-**); Sjogren syndrome (**M35.0-**)
 - Systemic sclerosis (**M34.-**); or adverse effects of drugs or toxins (**T50.5X5**)
 - The sequencing is based on the reason for the encounter, except for adverse effects of drugs

Pulmonary Hypertension

Documentation and Coding

- For **Pulmonary hypertension due to left heart disease (I27.22)** Code also associated left heart disease, if known, such as:
 - Multiple valve disease (**I08.-**); rheumatic aortic valve diseases (**I06.-**)
 - Rheumatic mitral valve diseases (**I05.-**)
- For **Pulmonary hypertension due to lung diseases and hypoxia (I27.23)** Code also associated lung disease, if known, such as:
 - Bronchiectasis (**J47.-**); cystic fibrosis with pulmonary manifestations (**E84.0**)
 - Interstitial lung disease (**J84.-**); pleural effusion (**J90**); sleep apnea (**G47.3-**)
- For **Chronic thromboembolic pulmonary hypertension (I27.24)** Code also associated
 - Pulmonary embolism, if applicable (**I26.-, I27.82**)
- For **Other secondary pulmonary hypertension (I27.29)** Code also other associated disorders, if known, such as:
 - Chronic myeloid leukemia (**C92.10-C92.22**); essential thrombocythemia (**D47.3**)
 - Gaucher disease (**E75.22**); hyperthyroidism (**E05.-**); hypothyroidism (**E00-E03**)
 - Hypertensive chronic kidney disease with end stage renal disease (**I12.0, I13.11, I13.2**)
 - Polycythemia vera (**D45**); sarcoidosis (**D86.-**)
- For **Personal history of other diseases of the circulatory system (Z86.79)** - when general hypertension is resolved.
- For **Personal history of other diseases of the respiratory system (Z87.09)** for resolved respiratory-related PH issues.

Coding Example

Case 1	A 67-year-old female patient presents with congestive heart failure and persistent pulmonary hypertension despite normalization of the left ventricular (LV) filling pressure.
ICD-10-CM Diagnosis Codes	I50.9 - Heart failure, unspecified I27.22 - Pulmonary hypertension due to left heart disease

Documentation and Coding

References

- [Lung.org/pulmonary hypertension](https://www.lung.org/pulmonary-hypertension)
- [CMS.medical-record-reviewer-guidance-january-2020](https://www.cms.gov/medicare/coverage/guidance/medicalrecordreviewerguidancejanuary2020)
- [Mayoclinic.org/pulmonary-hypertension/diagnosis-treatment](https://www.mayoclinic.org/pulmonary-hypertension/diagnosis-treatment)
- [2026-icd-10-cm-coding-guidelines](https://www.cms.gov/medicare/coverage/guidance/2026-icd-10-cm-coding-guidelines)

Questions?

Contact us at [#Risk Adjustments and clinical Documentation@healthfirst.org](mailto:#Risk_Adjustments_and_clinical_Documentation@healthfirst.org).

For additional documentation and coding guidance, please visit the coding section on [HFproviders.org](https://www.healthfirst.org/HFproviders.org)

Coverage is provided by Healthfirst Health Plan, Inc., Healthfirst PHSP, Inc., and/or Healthfirst Insurance Company, Inc. (together, "Healthfirst").

<26-5602> <PRX26_44>