Pulmonary hypertension

Documentation and coding: Individual & Family Plans

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For coding education questions, email CignaHealthcareHCPEducation@CignaHealthcare.com.

It's important to accurately document and code diagnoses when submitting claims for your patients with Cigna Healthcare®-administered coverage. This helps ensure your diagnosis and coding practices comply with all applicable legal requirements[1], while enabling us to provide our customers with the benefits and resources they need. For additional information and resources, visit the Cigna Healthcare Individual & Family Plans page at CignaforHCP.com/IFP.

The information that follows is designed to provide guidance for the documentation and coding of claims for your patients with pulmonary hypertension. It is not meant to replace your judgment when caring for your patients.

Definition

Pulmonary hypertension (PH) is a "frequently identified, highly morbid condition" associated with increased mortality, hospitalizations, and significant financial burden.

The Pulmonary Hypertension Association (PHA) defines PH as a general term used to describe high blood pressure in the lungs from any cause. This elevated pressure thickens the lung's arteries, forcing the right side of the heart to work harder to pump blood into the lungs, ultimately leading to right-side heart failure.

The World Health Organization (WHO) classifies PH into five different groups, known as the PH WHO Groups. Knowing the groups is important, as not only do they help you understand the cause of the condition, but they also help you determine the correct International Classification of Diseases, 10th Revision, Clinical Modification (ICD-10-CM) code.

Group 1: This group is reserved for primary PH, or pulmonary arterial hypertension (PAH), as is also known. PAH is further broken down into three different types: Idiopathic PAH, which has no clear cause; heritable PAH, which is a genetic condition; and other PAH, which results from drug use and other conditions, such as liver disease, and connective tissue diseases, such as lupus and human immunodeficiency virus. Both idiopathic PH and heritable PH are listed as synonyms in this ICD-10 category.

Associated ICD-10-CM code: *I27.0 Primary pulmonary hypertension*

Group 2: Groups 2-5 refer to secondary PH, beginning with "the most common form of PH," according to the PHA. Group 2 PH occurs when problems with the left side of the heart cause blood to back up, raising pressure in the lung.

Associated ICD-10-CM code: *I27.22 Pulmonary hypertension due to left heart disease*

Group 3: PH in this group is a result of numerous other obstructive and restrictive lung diseases. These include chronic lung disease, chronic obstructive pulmonary disease (COPD), emphysema, interstitial lung disease, sleep apnea, and hypoxia (low oxygen levels).

Associated ICD-10-CM code: *I27.23 Pulmonary hypertension due to lung diseases and hypoxia*

Group 4: Known as chronic thromboembolic pulmonary hypertension (CTEPH), this PH group is caused by blood clots in the lungs. The clots, in turn, cause scarring, which restricts blood flow in the lungs, causing the right side of the heart to work harder.

Associated ICD-10-CM code: *I27.24 Chronic thromboembolic pulmonary hypertension*

Group 5: As its associated ICD-10 code suggests, this is a catch-all category of PH caused by other conditions. In this group, however, the causal connection between the conditions is not well understood. The conditions, according to the PHA, include "sarcoidosis, sickle cell anemia, chronic hemolytic anemia, splenectomy (spleen removal), and certain metabolic disorders."

Associated ICD-10 code: *I27.29 Other secondary pulmonary hypertension*

1 Diagnosis inaccuracies that are not addressed can result in administrative sanctions and potential financial penalties.

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ICD-10 code	Description	Additional codes and characters
I27.0	Primary pulmonary hypertension	Excludes1: • Persistent pulmonary hypertension of newborn (P29.30) • Pulmonary hypertension NOS (I27.20) • Secondary pulmonary arterial hypertension (I27.21) • Secondary pulmonary hypertension (I27.29)
I27.1	Kyphoscoliotic heart disease	
I27.20	Pulmonary hypertension, unspecified	Pulmonary hypertension NOS
I27.21	Secondary pulmonary arterial hypertension	• (Associated) (drug-induced) (toxin-induced) pulmonary arterial hypertension NOS • (Associated) (drug-induced) (toxin-induced) (secondary) group 1 pulmonary hypertension code also associated conditions, if applicable, or adverse effects of drugs or toxins, such as: ○ Adverse effect of appetite depressants (T50.5X5) ○ Congenital heart disease (Q20-Q28) ○ Human immunodeficiency virus [HIV] disease (B20) ○ Polymyositis (M33.2-) ○ Portal hypertension (K76.6) ○ Rheumatoid arthritis (M05.-) ○ Schistosomiasis (B65.-) ○ Sjögren syndrome (M35.0-) ○ Systemic Sclerosis (M34.-)
I27.22	Pulmonary hypertension due to left heart disease	Group 2 pulmonary hypertension code also associated left heart disease, if known, such as: • Multiple valve disease (I08.-) • Rheumatic mitral valve diseases (I05.-) • Rheumatic aortic valve diseases (I06.-)
I27.23	Pulmonary hypertension due to lung diseases and hypoxia	Group 3 pulmonary hypertension 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-)
I27.24	Chronic thromboembolic pulmonary hypertension	Group 4 pulmonary hypertension code also associated pulmonary embolism, if applicable (I26.-, I27.82)
I27.29	Other secondary pulmonary hypertension	Group 5 pulmonary hypertension • Pulmonary hypertension with unclear multifactorial mechanisms • Pulmonary hypertension due to hematologic disorders • Pulmonary hypertension due to metabolic disorders • Pulmonary hypertension due to other systemic disorders code also other associated disorders, if known, such as: ○ Chronic myeloid leukemia (C92.10-C92.22) ○ Essential thrombocythemia (D47.3) ○ Gaucher disease (E75.22) ○ Hypertensive chronic kidney disease with end stage renal disease (I12.0, I13.11, I13.2) ○ Hyperthyroidism (E05.-) ○ Hypothyroidism (E00-E03) ○ Polycythemia vera (D45) ○ Sarcoidosis (D86.-) I27.8 Other specified pulmonary heart diseases
I27.81	Cor pulmonale (chronic)	• Cor pulmonale NOS code also, if applicable, right heart failure (I50.81-) • Excludes1: Acute cor pulmonale (I26.0-)
I27.83	Eisenmenger's syndrome	• Eisenmenger's complex • (Irreversible) Eisenmenger's disease • Pulmonary hypertension with right to left shunt related to congenital heart disease code also underlying heart defect, if known, such as: ○ Atrial septal defect (Q21.1-) ○ Eisenmenger's defect (Q21.8) ○ Patent ductus arteriosus (Q25.0) ○ Ventricular septal defect (Q21.0)

Pulmonary hypertension *(continued)*

ICD-10 code	Description	Additional codes and characters
27.84-	**Fontan related circulation**	(-) Add 5th character: 0 – Fontan-associated liver disease [FALD] 1 – Fontan-associated lymphatic dysfunction 8 – Other Fontan-associated condition 9 – Fontan related circulation, unspecified
I27.89	Other specified pulmonary heart diseases	
I27.9	Pulmonary heart disease, unspecified	