

# Pulmonology Coding Alert

## ICD-10: 5-Group System Helps You Classify Pulmonary Hypertension

**Turn to I27.22 for left heart disease.**

Selecting the right code to describe a patient's pulmonary hypertension will depend on several key factors including whether the hypertension is arterial or is caused by lung disease. Choose the right code from the ICD-10-CM I27.0 (Primary pulmonary hypertension) and I27.2- (Other secondary pulmonary hypertension) code range, using the recommendations that follow.

### Pin Down the Dx

It's not always obvious that the reason for a patient's shortness of breath, swollen ankles, and liver tenderness-to name a few common symptoms-is pulmonary hypertension (PH).

That's why extensive diagnostic testing is necessary to confirm that PH is actually the problem and, equally importantly, to identify its underlying cause, according to research from an April 23 study posted on the clinical decision support site, [www.uptodate.com](http://www.uptodate.com).

**Key indicators:** "Initially, some test results, along with the patient's signs/symptoms may cause the physician to suspect PH," says **Carol Pohlig, BSN, RN, CPC, ACS**, senior coding and education specialist at the Hospital of the University of Pennsylvania. "Increased jugular venous pressure, abnormal heart sounds, peripheral edema, a chest x-ray showing enlarged pulmonary arteries can raise suspicion," she adds.

In order to confirm a diagnosis, the physician may order a transthoracic echo, says Pohlig. This will also identify the severity and potential cause; once confirmed, the underlying cause can be explored, she adds.

**Further tests:** While cardiac etiologies can be considered through heart catheterization, the pulmonologist may focus on the underlying lung disease and order pulmonary function tests (PFTs), a six-minute walk test (6MWT), cardio-pulmonary exercise test (CPET) and a high-resolution chest CT scan, and sometimes a V/Q scan, adds Pohlig.

**Grim reminder:** If left untreated, PH can lead to right heart failure and death.

### Review the 5 PH Groups

When you're assessing patients with suspected PH, having an understanding of the 5 PH classification groups, determined by the World Health Organization (WHO), helps clarify your decision-making. The WHO groups are as follows, as outlined in a Pulmonary Hypertension Association article:

**Group 1: Pulmonary Arterial Hypertension (PAH)** - caused by arterial plaque that narrows arteries and puts extra stress on the heart, resulting in the heart losing its ability to pump sufficient blood to the lungs.

**Code it:** ICD-10 code I27.0 (Primary pulmonary hypertension) is what you'll use for your Group 1 PAH patients, including the 3 types below, which are included in the ICD-10 official long descriptor for I27.0:

### Types of PAH:

- Ideopathic PAH-which means there's no clear cause.
- Heritable PAH-which means the condition's genetic.
- Other PAH-less clearly defined but resulting from drug use, liver disease, connective tissue diseases such as lupus, and HIV.

**Group 2: PH due to left heart disease**-caused by weakness in the left side of the heart, which causes a back-up of blood returning to the lungs, raising the pressure in the lungs. This is the most common form of PH, the PH Association site indicates.

**Code it:** For your patients with this condition, you'll report I27.22 (... due to left heart disease).

**Group 3: PH due to lung disease**-results from obstructive and restrictive lung diseases, including COPD, emphysema, interstitial lung disease, and sleep apnea. The tightening of the lungs from these conditions increases pressure in the lungs.

**Code it:** If patients are in Group 3, you'll report I27.23 (... due to lung diseases and hypoxia).

**Group 4: PH due to chronic blood clots**-otherwise known as "chronic thromboembolic pulmonary hypertension" (CTEPH). This occurs, says the PA Association article, when the body can't dissolve a blood clot in the lungs, which causes flow-impeding scar tissue, making the heart work harder.

**Code it:** You'll use I27.24 (Chronic thromboembolic pulmonary hypertension) for patients with blood clots in the lungs.

**Group 5: PH due to unknown causes**-caused by secondary diseases that "are not well understood," as the article puts it. These associated conditions include sickle cell anemia, splenectomy, and certain metabolic disorders.

**Code it:** To describe the PH for patients in this group, use I27.29 (Other secondary pulmonary hypertension).

### PH Prognosis

If your patient has PAH (Group 1), he or she may receive therapy that targets the pulmonary arteries to alleviate symptoms, improve quality of life and slow down the disease progression, says the PH Association article.

Patients in Groups 2 and 3 can benefit from treating the underlying left heart and lung diseases, respectively.

If your patient's got PH from blood clots (Group 4), the physician may recommend surgery to remove the clots or a PH-targeted therapy if they are unable to have the surgery or have PH remaining after the surgery.

**Resources:** To see the WHO's PH Group classifications, go to <https://phassociation.org/types-pulmonary-hypertension-groups/>.

To review PH material on the uptodate.com site, see <https://www.uptodate.com/contents/clinical-features-and-diagnosis-of-pulmonary-hypertension-of-unclear-etiology-in-adults>.