AAVBC

AMERICAN ACADEMY OF VALUE BASED CARE

Pulmonary Hypertension

Quick Reference Guide

2026

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1 CLINICAL SNAPSHOT

Definition: Pulmonary hypertension (PH) is a hemodynamic condition defined by a **mean pulmonary artery pressure (mPAP) >20 mm Hg** measured by right heart catheterization (RHC) at rest, reflecting elevated pressures in the pulmonary vasculature that progressively overload the right ventricle. PH encompasses five distinct WHO clinical groups — pulmonary arterial hypertension (PAH, Group 1), PH due to left heart disease (Group 2), PH due to lung diseases and hypoxia (Group 3), chronic thromboembolic PH (CTEPH, Group 4), and PH with unclear or multifactorial mechanisms (Group 5).

Accurate WHO group classification is a clinical and documentation requirement, as distinguishing between these groups is necessary for patient safety. PAH-specific therapies that are first-line for **Group 1** carry **Class III (Harm)** recommendations for **Groups 2 and 3**, which constitute the majority of pulmonary hypertension cases in the Medicare population (age >65).[1,2]

WHO PH Classification

WHO GROUP	NAME	KEY ETIOLOGIES	TREATMENT APPROACH	PAH-SPECIFIC THERAPY?
Group 1	Pulmonary Arterial Hypertension (PAH)	Idiopathic, heritable, drug-induced, CTD-associated, HIV, portal hypertension, congenital heart disease	PAH-specific therapies: endothelin receptor antagonists, PDE5 inhibitors, prostacyclin pathway agents	**Yes** — first-line indication
Group 2	PH due to Left Heart Disease	LV systolic/diastolic dysfunction, valvular heart disease, congenital/acquired cardiovascular conditions	Treat underlying cardiac disease; diuretics; valve intervention if indicated	No — Class III (harm)
Group 3	PH due to Lung Diseases and/or Hypoxia	COPD, ILD (including IPF), sleep-disordered breathing, chronic high-altitude exposure	Treat underlying lung disease; supplemental oxygen; inhaled treprostinil (FDA-approved for PH-ILD)	No — Class III (harm); exception: inhaled treprostinil for PH-ILD
Group 4	Chronic Thromboembolic PH (CTEPH)	Organized thromboembolic obstruction of pulmonary arteries	Pulmonary thromboendarterectomy (potentially curative); balloon pulmonary angioplasty; riociguat for inoperable/residual disease	Riociguat only (not standard PAH therapies)

WHO GROUP	NAME	KEY ETIOLOGIES	TREATMENT APPROACH	PAH-SPECIFIC THERAPY?
Group 5	PH with Unclear/ Multifactorial Mechanisms	Hematologic disorders (sickle cell, myeloproliferative), sarcoidosis, glycogen storage disease, other systemic/metabolic conditions	Treat underlying condition; individualized approach	No established role

ICD-10 Codes:

PH is classified by underlying etiology through a series of specific codes. **I27.20 (pulmonary hypertension, unspecified)** should never be used as a default — it undercodes severity of illness (SOI) and risk of mortality (ROM) and fails to direct treatment. The definitive classification depends on hemodynamic data, specifically the **pulmonary artery wedge pressure (PAWP)**: PAWP ≤15 mm Hg indicates pre-capillary PH (Groups 1, 3, 4, 5), while PAWP >15 mm Hg indicates post-capillary PH (Group 2) or combined pre-and post-capillary PH. Always document: RHC values (mPAP, PAWP, PVR), the WHO group, and the underlying etiology.[3,4]

HCC/RAF V28 Mapping

DIAGNOSIS CATEGORY	ICD-10 SERIES	V28 HCC	RAF VALUE	CLINICAL DOCUMENTATION REQUIREMENT
Primary pulmonary hypertension (idiopathic PAH) Group 1: Idiopathic or Heritable	I27.0	HCC 226	0.36	**Hemodynamics:** Right heart catheterization (RHC) shows mean pulmonary artery pressure (mPAP) **>20 mmHg**, pulmonary artery wedge pressure (PAWP) **≤15 mmHg**, and pulmonary vascular resistance (PVR) **>2 Wood units**. Specify **idiopathic pulmonary arterial hypertension (PAH)** or **heritable PAH**. Document the clinical evaluation for secondary or associated systemic disease and record the **World Health Organization Functional Class (WHO-FC I–IV)**
Secondary Pulmonary Arterial Hypertension Group 1: Associated PAH	I27.21	HCC 226	0.36	**Hemodynamics:** mPAP **>20 mmHg**, PAWP **≤15 mmHg**, and PVR **>2 Wood units**, with an identified associated etiology. Pair with the documented underlying cause, such as **systemic sclerosis (M34.-), systemic lupus erythematosus (M32.-), portal hypertension (K76.6), HIV disease (B20), or congenital heart disease (Q20–Q28)**

DIAGNOSIS CATEGORY	ICD-10 SERIES	V28 HCC	RAF VALUE	CLINICAL DOCUMENTATION REQUIREMENT
Group 2: Pulmonary Hypertension Due to Left Heart Disease	I27.22	HCC 226	0.36	**Hemodynamics:** mPAP **>20 mmHg** and PAWP **>15 mmHg**. Differentiate **isolated postcapillary pulmonary hypertension** (PVR **≤2 Wood units**) from **combined pre- and postcapillary pulmonary hypertension** (PVR **>2 Wood units**); Pair with the documented left heart condition, such as **heart failure with reduced or preserved ejection fraction (I50.-), mitral valve disease (I05.-), or aortic valve disease (I35.-)**
Group 3: Pulmonary Hypertension Due to Lung Disease or Hypoxia	I27.23	HCC 226	0.36	**Hemodynamics:** mPAP **>20 mmHg**, PAWP **≤15 mmHg**, and PVR **>2 Wood units**, secondary to parenchymal lung disease or hypoxia. Pair with the documented pulmonary condition, such as **chronic obstructive pulmonary disease (J44.-), interstitial lung disease (J84.-), or obstructive sleep apnea (G47.33)**
Group 4: Chronic Thromboembolic Pulmonary Hypertension (CTEPH)	I27.24	HCC 226	0.36	**Hemodynamics:** mPAP **>20 mmHg**, PAWP **≤15 mmHg**, and PVR **>2 Wood units**. Pair with **chronic pulmonary embolism (I27.82)**. Document confirmation by **ventilation/perfusion scan or CT pulmonary angiography**, as applicable, and the formal assessment of surgical operability
Group 5: Other secondary PH (multifactorial)	I27.29	HCC 226	0.36	**Clinical findings:** Elevated mPAP **>20 mmHg** associated with unclear, complex, or multifactorial mechanisms. Pair with the documented systemic cause, such as **sarcoidosis (D86.-), chronic kidney disease (N18.-), or thyroid disease (E03.- or E07.-)**
PH, unspecified	I27.20	HCC 226	0.36	**Avoid.** Always specify the underlying cause. Undercodes SOI/ROM

DIAGNOSIS CATEGORY	ICD-10 SERIES	V28 HCC	RAF VALUE	CLINICAL DOCUMENTATION REQUIREMENT

ABBREVIATIONS: CTPA: CT pulmonary angiography; CTEPH: chronic thromboembolic pulmonary hypertension; eGFR: estimated glomerular filtration rate; HCC: Hierarchical Condition Category; HFpEF: heart failure with preserved ejection fraction; HFrEF: heart failure with reduced ejection fraction; HIV: human immunodeficiency virus; HTN: hypertension; LHD: left heart disease; mPAP: mean pulmonary artery pressure; NOS: not otherwise specified; PAH: pulmonary arterial hypertension; PAWP: pulmonary artery wedge pressure; PH: pulmonary hypertension; PVR: pulmonary vascular resistance; RAF: Risk Adjustment Factor; RHC: right heart catheterization; ROM: risk of mortality; SOI: severity of illness; V/Q: ventilation/perfusion; WHO-FC: World Health Organization Functional Class; WU: Wood units

***RAF values represent the Community Non-Dual Eligible Aged (CNA) coefficient from the 2026 CMS-HCC model; values vary across patient populations based on eligibility and care setting**

Risk-Adjusted Care Resources per Patient/Year

Risk-adjusted care resource allocation — MA base rate ($10,402.34) × RAF coefficient; HCC 226

GROUPs 1-4

$3,745

HCC 226 · RAF 0.36

NOTE: Code both the PH code and the underlying condition (e.g., I27.22 + I50.x for Group 2; I27.23 + J44.x for Group 3 with COPD); Note: All PH codes map to a single HCC 226 with uniform RAF weight regardless of WHO group. Clinical complexity and cost burden vary enormously — Group 1 PAH on combination vasodilator therapy may exceed $100,000/year in drug costs alone, while Groups 2/3 managed through underlying disease optimization cost far less. Accurate group classification drives appropriate resource allocation even though RAF is identical. HCC 226 (Pulmonary Heart Disease Including Cor Pulmonale): RAF 0.36 × $10,402.34 = ~$3,745/year per patient]. RAF values represent the Community Non-Dual Eligible Aged (CNA) coefficient from the 2026 CMS-HCC model; values vary across patient populations based on eligibility and care setting

Prevalence: PH affects approximately **1% of the global population**, with prevalence rising to approximately **10% of adults aged >65 years**. Left heart disease (Group 2) is the most common cause globally, followed by lung diseases (Group 3). Group 1 PAH is rare (**2.0–7.6 cases per million adults per year**), but Groups 2 and 3 account for the overwhelming majority in the Medicare Advantage population. Group 2 is the most prevalent form of PH, accounting for **65–80%** of all cases, and Group 3 PH is the second most common form of pulmonary hypertension globally.[1] PH-related hospitalizations in the MA population carry a **mean length of stay of 12.8 days**, making hospitalization the primary economic driver. Registry data show distinct elderly phenotypes among idiopathic PAH patients: **35.8% left heart phenotype** and **51.6% cardiopulmonary phenotype**, each requiring different management approaches.

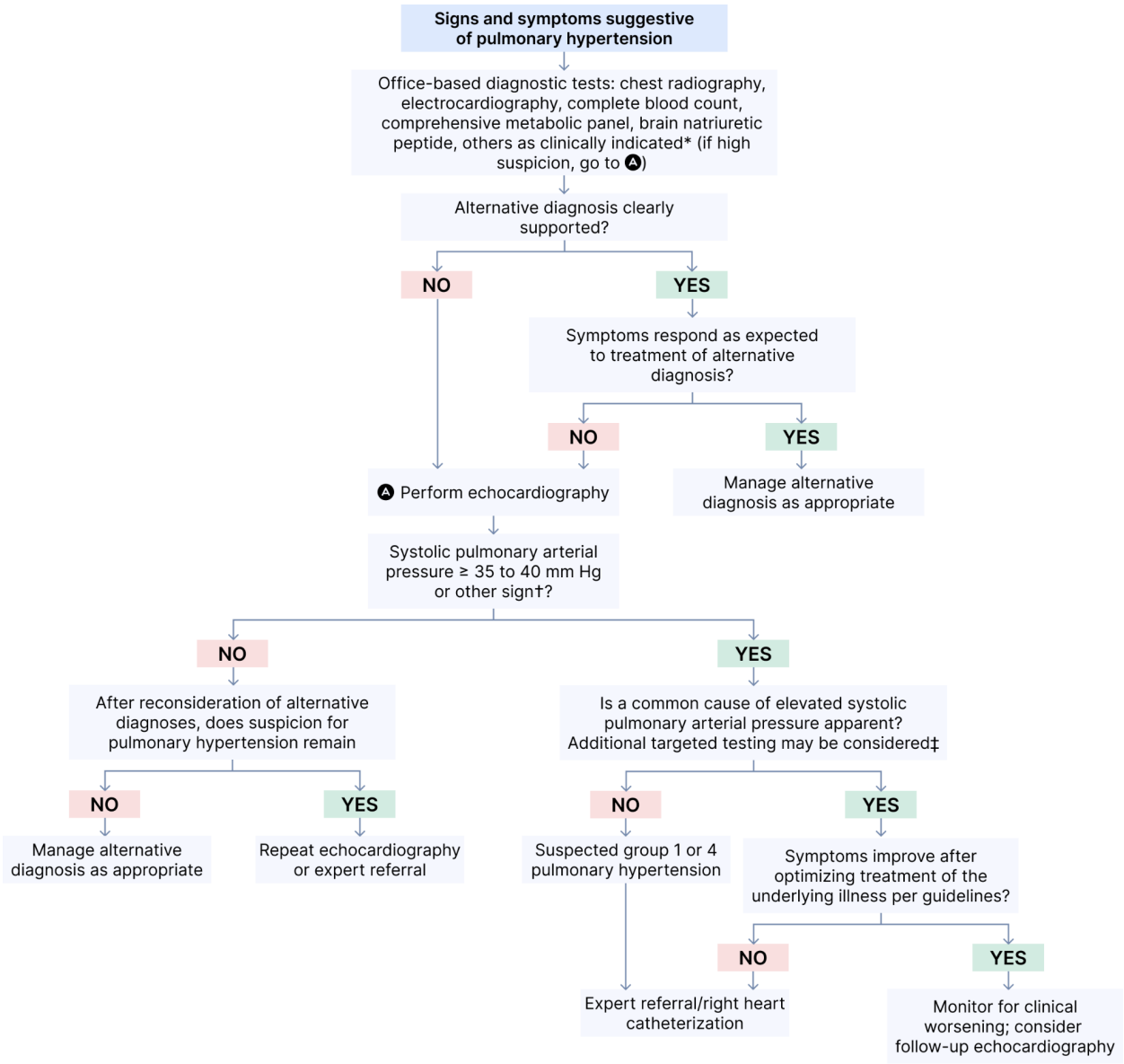
2 RECOGNITION AND DIAGNOSIS

Medicare Diagnostics

The complete diagnostic workup for PH, from initial echocardiographic screening through definitive right heart catheterization and ongoing risk stratification, is covered by Medicare when clinically indicated, with the following table outlining each test, its recommended frequency, applicable CPT code, and clinical role in WHO group classification and disease monitoring.[2]

TEST	COVERAGE	FREQUENCY	CPT CODE	WHAT IT MEASURES
Transthoracic echocardiography	Covered when clinically indicated	At diagnosis; repeat if clinical change	93306/93307	Estimated sPAP (TRV >2.8 m/s suggests PH); RV size and function; TAPSE
Right heart catheterization	Covered when clinically indicated	At diagnosis (mandatory); repeat for uncertainty	93451–93453	Definitive: mPAP, PAWP, PVR — classifies WHO group
V/Q scan (perfusion + ventilation)	Covered when clinically indicated	At diagnosis to exclude CTEPH	78580 + 78582	Perfusion defects indicating chronic thromboembolic disease
Pulmonary function tests with DLCO	Covered	At diagnosis; serial per clinical need	94729	Airflow obstruction, restriction, gas transfer — identifies Group 3 etiology
BNP/NT-proBNP	Covered	At diagnosis; q3-6 months for monitoring	83880/83883	Prognostic biomarker; risk stratification; normal BNP + normal ECG suggests PH unlikely
6-Minute Walk Test	Covered	At diagnosis; q3-6 months for monitoring	94618	Exercise capacity; key component of risk stratification

Pulmonary Hypertension: Diagnosis Pathway



* Human immunodeficiency virus or rheumatologic evaluation.
† Other signs suggestive of pulmonary hypertension include right ventricular or right atrial enlargement, right ventricular hypertrophy, high-velocity regurgitant jet, or flattening of the interventricular septum.
‡ Pulmonary function test, sleep study, ambulatory pulse oximetry, chest computed tomography, or other testing based on clinical suspicion.
§ Consistent with, but not diagnostic of, group 2, 3, or 5 pulmonary hypertension.

Subtle Early Signs in Adults

PH diagnosis is delayed by more than two years in over 20% of patients because its earliest symptoms overlap with common conditions seen daily in primary care, making the following clinical signals critical to recognize before irreversible right ventricular remodeling occurs.[1,2,6]

SIGN/SYMPTOM	CLINICAL SIGNIFICANCE
Exertional dyspnea disproportionate to known cardiac or pulmonary disease	**Most common presenting symptom**; often attributed to deconditioning or aging — suspect PH when dyspnea exceeds what underlying disease explains
Progressive exercise intolerance with fatigue	Reflects declining cardiac output; may manifest as inability to complete activities previously tolerated; **track functional decline over months**
Exertional lightheadedness or presyncope	Indicates limited ability to augment cardiac output with exercise; concerning for RV compromise; **not explained by orthostatic hypotension alone**
New or worsening peripheral edema without clear HF escalation	Suggests right ventricular failure and elevated central venous pressure; often misattributed to venous insufficiency or medication side effects
Unexplained loud P2 (pulmonic component of S2) on auscultation	Physical exam clue to elevated pulmonary pressures; often missed in routine exams; **correlates with sPAP elevation on echo**
Declining 6-Minute Walk Distance without obvious musculoskeletal cause	Objective measure of functional decline; **>15%** decline triggers concern for disease progression; part of formal risk stratification
Nonproductive cough, hoarseness (Ortner syndrome)	Dilated pulmonary artery can compress the recurrent laryngeal nerve; rare but pathognomonic when present

ABBREVIATIONS: RV = right ventricle; HF = heart failure; sPAP = systolic pulmonary artery pressure; P2 = pulmonic component of second heart sound sPAP = systolic pulmonary artery pressure; TRV = tricuspid regurgitation velocity; RV = right ventricle; TAPSE = tricuspid annular plane systolic excursion; mPAP = mean pulmonary artery pressure; PAWP = pulmonary artery wedge pressure; PVR = pulmonary vascular resistance; CTEPH = chronic thromboembolic pulmonary hypertension; DLCO = diffusing capacity of the lung for carbon monoxide; BNP = brain natriuretic peptide; NT-proBNP = N-terminal pro-B-type natriuretic peptide; ECG = electrocardiogram; V/Q = ventilation/perfusion

Risk Factors

Pulmonary hypertension rarely occurs in isolation, specific comorbidities and exposures not only increase the likelihood of developing PH but also independently worsen survival, making systematic risk factor identification essential for early detection, accurate WHO group classification, and targeted management.[1,7–10]

FACTOR	RISK SIGNAL	EVIDENCE SUMMARY	CLINICAL IMPLICATION
COPD	**HR 1.59** for **death** (95% CI 1.34–1.90)	Worse 6MWD and functional class; **51.6%** of elderly IPAH have cardiopulmonary phenotype	**Screen** for PH in COPD patients with dyspnea out of proportion to airflow limitation
Type 2 diabetes	**HR 1.73** for **death** (95% CI 1.40–2.13)	Significantly worse 6MWD; metabolic comorbidity burden	Tight glycemic control; assess for contribution to LV diastolic dysfunction

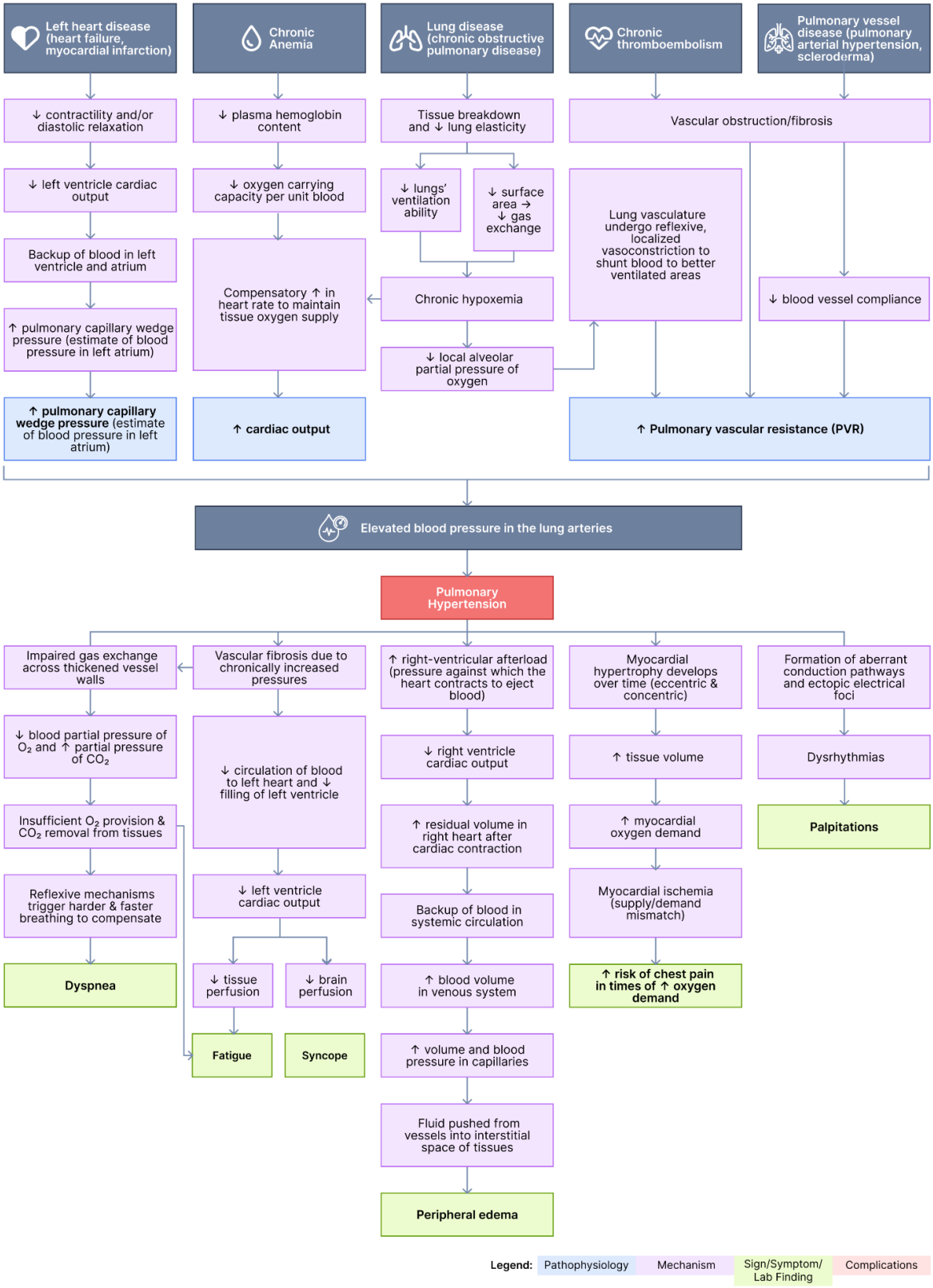
FACTOR	RISK SIGNAL	EVIDENCE SUMMARY	CLINICAL IMPLICATION
Hypertension	Worse 6MWD	Associated with **left heart phenotype** (35.8% of elderly IPAH)	Aggressive BP control; screen for Group 2 PH; avoid assuming PH is solely Group 1
Obesity	Worse functional class; **HR 0.73** for death (**paradoxical**)	Obesity paradox: **worse function but reduced mortality**; contributes to HFpEF phenotype	Screen for sleep apnea; assess for Group 2 contribution; weight management counseling
Combined CV + lung comorbidities	**HR 1.86** for mortality (95% CI 1.20–2.89)	Synergistic risk; common in elderly PH	Comprehensive comorbidity optimization is foundation of treatment
Systemic sclerosis	**5–19% prevalence** of PAH	Annual screening recommended; DETECT algorithm for FVC ≥40% and DLCO <60%	**Annual** echo, DLCO, and biomarkers — most common connective tissue cause of PAH
Prior pulmonary embolism	**~3%** develop **CTEPH**	**Persistent dyspnea** post-PE triggers evaluation at 3 months	V/Q scan + echo + BNP/NT-proBNP for post-PE patients with persistent symptoms

ABBREVIATIONS: HR = hazard ratio; CI = confidence interval; 6MWD = 6-Minute Walk Distance; IPAH = idiopathic pulmonary arterial hypertension; AF = atrial fibrillation; BP = blood pressure; CV = cardiovascular; DLCO = diffusing capacity for carbon monoxide; FVC = forced vital capacity; CTEPH = chronic thromboembolic pulmonary hypertension; PE = pulmonary embolism; V/Q = ventilation/perfusion; BNP = brain natriuretic peptide

AAVBC PERSPECTIVE

*AAVBC recommends that PCPs **prioritize early recognition**, **structured risk stratification**, and **coordinated multidisciplinary care**. **Proactive screening of high-risk patients** including those with unexplained dyspnea, connective tissue disease, chronic liver disease, or congenital heart disease through routine cardiovascular and metabolic assessment enables detection before irreversible right ventricular remodeling occurs. Once PH is suspected, **timely referral** to a PH specialist center and WHO group classification is essential. Ongoing management centers on guideline-directed therapy optimization, serial risk reassessment using the four-stratum model, and structured follow-up intervals designed to maintain or restore low-risk status.*

Pulmonary Hypertension: Pathogenesis and Clinical Findings



⚠ RED FLAG — URGENT ACTION

- **Syncope or presyncope on mild exertio**n → Immediate cardiology/PH specialist evaluation; suggests severely limited cardiac output reserve
- **Clinical signs of right heart failure:** elevated JVP, new or worsening peripheral edema, ascites, hepatomegaly → Urgent assessment for hemodynamic decompensation; consider hospital admission
- **Hemodynamic instability:** hypotension (SBP <90 mm Hg) with tachycardia → Emergency department; potential cardiogenic shock from RV failure
- **Rapid weight gain >5 lbs in 1 week with worsening dyspnea** → Same-day evaluation; aggressive diuresis and medication adjustment
- **New poorly tolerated arrhythmias with hemodynamic compromise** → Emergency evaluation; AF or flutter common in advanced PH
- **Markedly elevated BNP/NT-proBNP (NT-proBNP >1400 ng/L) with clinical deterioration** → High-risk category; urgent treatment escalation and PH center referral

Diagnostic Thresholds

TEST	DIAGNOSTIC VALUE	NOTES
RHC — mPAP	>20 mmHg (rest)	**Defines PH**; updated from previous >25 mm Hg criterion in 2022 ESC/ERS; mandatory for diagnosis, noninvasive testing alone cannot establish PH
RHC — PAWP	≤15 mm Hg = pre-capillary >15 mm Hg = post-capillary	**Distinguishes PAH** (Groups 1/3/4/5) **from Group 2 LHD-PH**; frequently absent from documentation
RHC — PVR	>2 WU = pre-capillary P >5 WU = severe PH in Group 3H	Refined threshold (previously >3 WU)
Echo — TRV	≤2.8 m/s= low probability 2.9-3.4 m/s= intermediate probability >3.4 m/s= high probability	Screening only — does not confirm diagnosis; RHC required; false positives in elderly. **Non-invasive.** Interpret TRV with symptoms, risk factors, and additional echocardiographic findings
Echo — estimated sPAP	>35 mm Hg with ≥2 anatomic abnormalities	Intermediate/high probability of PH; additional signs: RV dilation, septal flattening, RA enlargement. **Non-invasive**
Echo — TAPSE	<17 mm	**Indicates RV systolic dysfunction**; prognostic marker; part of echo risk assessment. **Non-invasive**
BNP/NT-proBNP	Normal BNP + normal ECG → PH unlikely	**Prognostic**: NT-proBNP <300 = low risk; 300–1400 = intermediate; >1400 = high risk

TEST	DIAGNOSTIC VALUE	NOTES
6MWD	>440m = low risk <165m = high risk	Key **functional measure;** <440 m with symptoms warrants further evaluation; track serially
V/Q scan	Mismatched perfusion defects	**Excludes or confirms CTEPH** (Group 4); superior to CT for chronic disease detection

ABBREVIATIONS: RHC = right heart catheterization; mPAP = mean pulmonary artery pressure; PAWP = pulmonary artery wedge pressure; PVR = pulmonary vascular resistance; WU = Wood Units; TRV = tricuspid regurgitation velocity; sPAP = systolic pulmonary artery pressure; RV = right ventricle; RA = right atrium; TAPSE = tricuspid annular plane systolic excursion; BNP = brain natriuretic peptide; NT-proBNP = N-terminal pro-B-type natriuretic peptide; ECG = electrocardiogram; 6MWD = 6-Minute Walk Distance; V/Q = ventilation/perfusion; CTEPH = chronic thromboembolic pulmonary hypertension

CLINICAL PEARL: DIAGNOSIS

Echocardiography is the primary, **non-invasive** screening tool used to estimate the probability of PH, while **catheterization** remains the **gold standard for definitive diagnosis**.
Cardiac catheterization is an invasive procedure but is generally safe, with major complications occurring in less than **1%** of diagnostic cases. Risks include bleeding, infection, artery damage, arrhythmias, and rarely, stroke or heart attack. Common, minor side effects include bruising or discomfort at the insertion site, with serious risks more common in complex or emergency procedures.

Clues to Dig Deeper

- **Dyspnea exceeding what underlying cardiac or pulmonary disease explains:** Consider PH as a contributing or independent diagnosis — especially in patients with HFpEF, COPD, or ILD where PH is underdiagnosed[1]
- **Echocardiographic TRV >2.8 m/s without known PH diagnosis:** intermediate-to-high probability of PH; pursue RHC to classify WHO group before initiating any targeted therapy[1]
- **Persistent dyspnea 3+ months after pulmonary embolism treatment:** Screen for CTEPH (Group 4) with V/Q scan and echocardiography; 3% of post-PE patients develop CTEPH[11]
- **Declining DLCO out of proportion to restrictive or obstructive pattern on PFTs:** May indicate pulmonary vascular disease independent of parenchymal lung disease; especially concerning in systemic sclerosis patients[12]
- **Echo showing RV dilation, septal flattening, or pericardial effusion in a patient without known PH:** These structural findings suggest significant hemodynamic burden — obtain RHC for definitive classification[1]

Common Oversights

- **Treating PH based on echocardiographic findings alone without RHC confirmation:** Echo screens but does not diagnose; PAWP (only available from RHC) determines the WHO group and therefore the entire treatment pathway[1]
- **Prescribing PDE5 inhibitors (sildenafil, tadalafil) for Group 2 or Group 3 PH:** Class III harm warning; no benefit and potential harm in left heart disease and non-severe lung disease PH[1]
- **Failing to document PAWP from RHC in the clinical note:** The single most important value for WHO group classification is frequently absent from documentation; without it, Group 2 vs. Group 1 distinction is impossible[1]
- **Using standard "up-titrate to max" approach in elderly patients:** Elderly patients respond differently to vasodilators due to age-related vascular changes; monotherapy preferred over dual combination in patients with comorbidities

Key Differentials

The following table summarizes the key differentials for pulmonary hypertension:[1]

DIAGNOSIS	DISTINGUISHING FEATURE	DIAGNOSTIC TEST	NEXT STEP
Left heart failure (HFpEF)	**PAWP >15 mmHg** on RHC; echocardiogram demonstrates elevated left ventricular filling pressures	**RHC:** PAWP >15 mmHg; *Note:* **PVR ≤2 WU** indicates Isolated Post-Capillary (IpcPH); **PVR > 2 WU** indicates Combined Pre-/Post-Capillary (CpcPH)	**Code I27.22.** Optimize underlying heart failure management. **Do not prescribe PAH-specific targeted therapies** (can cause fatal pulmonary edema)
Chronic obstructive pulmonary disease	**PVR ≤5 WU** (typical non-severe profile); characteristically displays a purely **obstructive** pattern on PFTs	**RHC:** Normal or mildly elevated mPAP **PFTs:** Airflow obstruction (FEV1/FVC <0.70); hyperinflation	**Code I27.23.** Optimize COPD therapy (bronchodilators, oxygen). Avoid PDE5 inhibitors (e.g., sildenafil) in non-severe cases to prevent worsening V/Q mismatch

DIAGNOSIS	DISTINGUISHING FEATURE	DIAGNOSTIC TEST	NEXT STEP
Interstitial lung disease	**DLCO <40%** predicted; characteristically displays a **restrictive** pattern on PFTs; elevated mPAP driven by chronic hypoxia	**PFTs:** Restriction (low TLC) and severely blunted DLCO **HRCT:** Fibrosis/ honeycombing **RHC:** Confirms pre-capillary PH driven by lung disease	**Code I27.23.** Exclude connective tissue disease (CTD) etiologies. **Inhaled treprostinil** is the only FDA-approved, targeted vasodilator option for PH-ILD
Chronic thromboembolic PH	Documented history of PE or DVT (though absent in ~25% of cases); **mismatched perfusion defects** on V/Q scan	**V/Q Scan:** Wedge-shaped, segment-level perfusion defects **CTPA/Conventional Angiography:** Intraluminal webs/bands **RHC:** Confirms pre-capillary profile	**Code I27.24.** Immediately refer to a specialized center for surgical **pulmonary endarterectomy (PEA) assessment**. If inoperable, initiate riociguat
Pulmonary arterial hypertension (Group 1)	Idiopathic, heritable, or drug-induced; **PAWP ≤15 mmHg** and **PVR >2 WU** with completely clear lung fields/ left heart	**RHC:** Strict pre-capillary pattern (mPAP >20 mmHg, PAWP ≤15 mmHg, PVR >2 WU); Exclude CTD, CHD, HIV, and portal hypertension	**Code I27.0** (Idiopathic) or **I27.21** (Associated). Patient is an immediate candidate for specialized dual oral upfront vasodilator therapy pathways

ABBREVIATIONS: PAWP = pulmonary artery wedge pressure; PVR = pulmonary vascular resistance; RHC = right heart catheterization; HFpEF = heart failure with preserved ejection fraction; PFTs = pulmonary function tests; DLCO = diffusing capacity for carbon monoxide; PE = pulmonary embolism; DVT = deep vein thrombosis; V/Q = ventilation/perfusion; CTPA = CT pulmonary angiography; CTD = connective tissue disease; CHD = congenital heart disease

Comorbidity Screening

Pulmonary hypertension rarely exists in isolation,particularly in the Medicare population, where overlapping cardiovascular, metabolic, and autoimmune conditions both drive PH development and complicate its management, making systematic comorbidity screening essential at diagnosis and throughout follow-up.[1,13,14]

COMORBIDITY	SCREENING QUESTION	KEY FINDING	ACTION
Atrial fibrillation	Any history of palpitations, syncope, or documented arrhythmia?	AF present in **~30%** of elderly PH with left heart phenotype	ECG + Holter if indicated; rate control + anticoagulation per guideline

COMORBIDITY	SCREENING QUESTION	KEY FINDING	ACTION
Sleep apnea	Snoring, witnessed apneas, daytime somnolence, uncontrolled HTN despite meds?	**Central or obstructive apnea increases mPAP**; contributes to Group 2/3 phenotype	Sleep study if suspected; CPAP initiation can improve outcomes
Chronic kidney disease	Baseline creatinine; CKD-EPI eGFR; urinalysis for proteinuria?	CKD impacts diuretic response and drug clearance; **fluid retention worsens PH**	Monitor renal function q3-6 months with diuretics; avoid nephrotoxic agents
Anemia/iron deficiency	Baseline hemoglobin; ferritin; transferrin saturation?	**Iron deficiency common in PAH**; contributes to exercise intolerance; correct before ESA	Ferritin >100, TSAT >20%; IV iron preferred; recheck in 3 months
Connective tissue disease (CTD)	Raynaud, skin tightening, arthralgia, dry eyes/ mouth, or known CTD diagnosis?	Systemic sclerosis most common CTD-PAH; **5–19% prevalence**; requires **annual screening**	Autoimmune panel, ANA, anti-centromere; annual echo + DLCO + BNP
Liver disease/ portal hypertension	Cirrhosis, hepatic encephalopathy, ascites, or variceal bleeding history?	**Portal HTN causes PH** (Group 1 associated); increases bleeding risk on anticoagulation	Liver function panel; assess candidacy for liver transplant or portosystemic shunt
Obesity	BMI >30; sleep apnea; diabetes; HTN constellation?	**Obesity associates with worse function**; HFpEF phenotype; metabolic syndrome	Weight reduction counseling; screen for OSA; SGLT2i for HFpEF
Prior PE/DVT	Any history of provoked or unprovoked VTE?	**~3% develop CTEPH**; persistent dyspnea post-PE is CTEPH red flag	Persistent post-PE dyspnea at **3 months → V/Q scan + RHC for Group 4 evaluation**

ABBREVIATIONS: AF = atrial fibrillation; ECG = electrocardiogram; HTN = hypertension; eGFR = estimated glomerular filtration rate; ESA = erythropoiesis-stimulating agent; TSAT = transferrin saturation; IV = intravenous; CTD = connective tissue disease; ANA = antinuclear antibody; DLCO = diffusing capacity for carbon monoxide; BNP = brain natriuretic peptide; BMI = body mass index; HFpEF = heart failure with preserved ejection fraction; SGLT2i = SGLT2 inhibitor; OSA = obstructive sleep apnea; VTE = venous thromboembolism; CTEPH = chronic thromboembolic pulmonary hypertension; PE = pulmonary embolism; DVT = deep vein thrombosis; V/Q = ventilation/perfusion; RHC = right heart catheterization

Staging/Severity — Four-Stratum Risk Model (ESC/ERS 2022)

PH uses a multiparameter risk stratification model rather than a single staging system. The four-stratum model below is recommended for follow-up assessment and treatment decision-making.[1]

RISK CATEGORY	WHO-FC	6MWD	NT-PROBNP (NG/L)	1-YEAR MORTALITY	MANAGEMENT IMPLICATION
Low	I–II	>440 m	<300	0–3%	Maintain current therapy; **reassess q6 months**
Intermediate-Low	I–II	165–440 m	300–1400	2–7%	Consider treatment escalation; **reassess q3-6 months**
Intermediate-High	III	165–440 m	300–1400	9–19%	Escalate therapy; add prostacyclin pathway or triple therapy; **reassess q3 months**
High	IV	<165 m	>1400	>20%	**Urgent escalation to triple therapy** including parenteral prostacyclin; PH center referral

ABBREVIATIONS: WHO-FC = WHO Functional Class; 6MWD = 6-Minute Walk Distance; NT-proBNP = N-terminal pro-B-type natriuretic peptide

Calculation: Assign points 1–4 per variable, divide by number of variables, round to nearest integer. Use at least 3 variables: WHO-FC + 6MWD + BNP/NT-proBNP. Goal: achieve and maintain low-risk status.

CLINICAL PEARL: RISK STRATIFICATION

Risk stratification in PH requires the **four-stratum model using at least three variables** — WHO Functional Class alone is insufficient. A patient in WHO-FC II with a 6MWD of 200m and NT-proBNP of 1200 ng/L is intermediate-high risk despite appearing "mildly symptomatic."

3 MEAT DOCUMENTATION ESSENTIALS

A 72-year-old woman with COPD (FEV_1 48%), type 2 diabetes, and obesity (BMI 34) presents with dyspnea beyond what her lung disease explains. RHC confirmed Group 3 pre-capillary PH: mPAP 34, PAWP 12, PVR 4.8 WU. On tiotropium, fluticasone/salmeterol, home O_2 2L, and metformin.

MONITOR: "Group 3 PH (**I27.23**), confirmed RHC. mPAP 34, PAWP 12, PVR 4.8 WU, pre-capillary, non-severe. WHO-FC III. 6MWD 285m (↓16% from 340 m over 6 months). NT-proBNP 890 ng/L (↑ from 620). SpO_2 91% rest on 2L, desaturates to 84% on exertion. Weight stable. Adherent to all medications.

EVALUATE: "RHC 3/25/26: Group 3 confirmed. **V/Q 3/20/26:** no mismatched defects. CTEPH excluded. **Echo 2/15/26:** sPAP 52, TAPSE 16 mm (borderline RV dysfunction), mild RV dilation. **PFTs 1/10/26:** FEV_1 **48%**, DLCO **38%** disproportionately reduced, suggesting pulmonary vascular contribution. **Risk stratification:** WHO-FC III + 6MWD 285m + NT-proBNP 890 = intermediate-high risk."

ASSESS: "PH due to COPD (**I27.23 + J44.1**), Group 3, pre-capillary, non-severe. Intermediate-high risk — declining 6MWD, rising NT-proBNP, borderline RV function. COPD optimized. PH progressing despite underlying disease management. PDE5 inhibitors **NOT** recommended for non-severe Group 3 (Class III). Inhaled treprostinil being considered."

TREAT: "Continue COPD regimen; reinforce technique. Increase O_2 to 3L NC continuous; target SpO_2 >90%. **Refer PH specialist (4–6 weeks):** consider inhaled treprostinil — only approved therapy for PH-ILD; avoid PDE5i. Diuretics not indicated — no fluid overload. Pulmonary rehab referral placed. **Home monitoring:** Daily weights (action: >2–3 lb gain/day), SpO_2 log. Vaccinations current. **Follow-up 8 weeks:** repeat 6MWD, NT-proBNP, reassess risk. MEAT documented."

Clinical Documentation Elements

- **Specify WHO group and underlying etiology:** "PH due to chronic obstructive pulmonary disease" (**I27.23 + J44.1**) — **NOT** "pulmonary hypertension" alone or **I27.20** unspecified
- **Include hemodynamic data with dates:** "mPAP 34 mm Hg, PAWP 12 mm Hg, PVR 4.8 WU (RHC 3/25/26)" — anchors classification and demonstrates active evaluation
- **Document functional class at every encounter:** "WHO Functional Class III" — strongest single predictor of survival at diagnosis and follow-up
- **Document risk stratification with all three variables:** "**Risk:** intermediate-high — WHO-FC III, 6MWD 285m, NT-proBNP 890 ng/L" — guides treatment intensity
- **Document treatment contraindications explicitly:** "PDE5i **NOT** recommended for non-severe Group 3 PH per ESC/ERS 2022 (Class III)" — prevents inappropriate therapy

Reframing Documentation Shortcuts

INSTEAD OF...	DOCUMENT...	WHY THIS SUPPORTS CLARITY
"Pulmonary hypertension" (unspecified)	"Pulmonary hypertension due to left heart disease **(I27.22)**, Group 2, confirmed on RHC: PAWP 22 mm Hg"	Specifies WHO group and hemodynamic basis; directs treatment to underlying HF, not PAH drugs
"PH, elevated echo pressures"	"PH, pre-capillary: mPAP 38 mm Hg, PAWP 12 mm Hg, PVR 6.2 WU (RHC 3/15/26)"	Hemodynamic data from RHC — **not echo estimates** — is required for WHO group classification
"PAH, on treatment"	"PAH **(I27.21)**, WHO-FC II, on dual combination therapy: tadalafil 40 mg daily + macitentan 10 mg daily. 6MWD 450 m, NT-proBNP 180 ng/L — low risk"	Records diagnosis, classification, current therapy, and risk status with supporting data
"Worsening PH symptoms"	"**PH progressing:** WHO-FC III (was II), 6MWD declined 285m → 220 m (22%), NT-proBNP rising 890 → 1350 ng/L over 3 months — **risk category escalated from intermediate-low to high**"	Quantifies trajectory with three-variable risk model; triggers treatment escalation

INSTEAD OF...	DOCUMENT...	WHY THIS SUPPORTS CLARITY
"PH, COPD related"	"PH due to COPD **(I27.23 + J44.1)**, Group 3, non-severe (PVR 4.8 WU). **COPD optimized**; PDE5i contraindicated per ESC/ERS 2022 (Class III for non-severe Group 3 PH)"	Links codes correctly; documents severity threshold; prevents inappropriate PAH therapy
"Stable PH"	"PH **(I27.24, CTEPH)**, post-BPA ×3, WHO-FC II, 6MWD 410 m, NT-proBNP 220 ng/L — intermediate-low risk; riociguat 2.5 mg TID continued; next reassessment 6/2026"	Specifies type, treatment history, current status with data, and plan

ABBREVIATIONS: PH = pulmonary hypertension; PAH = pulmonary arterial hypertension; RHC = right heart catheterization; PAWP = pulmonary artery wedge pressure; mPAP = mean pulmonary artery pressure; PVR = pulmonary vascular resistance; WU = Wood Units; WHO-FC = WHO Functional Class; 6MWD = 6-Minute Walk Distance; NT-proBNP = N-terminal pro-B-type natriuretic peptide; PDE5i = phosphodiesterase-5 inhibitor; CTEPH = chronic thromboembolic pulmonary hypertension; BPA = balloon pulmonary angioplasty

DOCUMENTATION

The single most impactful documentation improvement for PH is replacing **I27.20** (unspecified) with the group-specific code supported by hemodynamic data. When PAWP is documented from RHC, the WHO group classification — and therefore the correct treatment pathway — follows directly.

4 TREATMENT AND REFERRAL QUICK GUIDE

Therapy Escalation Criteria (ESC/ERS 2022)

Treatment escalation in pulmonary hypertension follows a structured, risk-driven approach with the critical distinction that targeted PAH therapies are reserved for Group 1, while Groups 2 and 3 require optimization of the underlying cardiac or pulmonary disease as the primary intervention.[1]

TRIGGER	ACTION	EXPECTED BENEFIT
All PH Groups — General measures first	Supervised exercise (**3–5×/week, 30–60 min**); vaccination; sodium restriction (2,400 mg/day); psychosocial support; O_2 for SpO_2 <90%; diuretics for fluid retention	Marked increased 6MWD from exercise; **reduces hospitalizations**
Groups 2/3 — Optimize underlying disease	**Group 2:** GDMT for HF; Group 3: bronchodilators, smoking cessation, LTOT, pulmonary rehab	**PAH drugs contraindicated** (Class III) in these groups; underlying disease optimization is treatment

TRIGGER	ACTION	EXPECTED BENEFIT
Group 1 PAH, low/ intermediate risk, no comorbidities	**Dual oral combination:** PDE5i + ERA (sildenafil 20 mg TID or tadalafil 40 mg daily + ambrisentan 10 mg or macitentan 10 mg daily)	**HR 0.50 clinical failure vs. monotherapy** (AMBITION trial)
Group 1 PAH, elderly with comorbidities	**Monotherapy preferred:** PDE5i (sildenafil or tadalafil) at lowest effective dose; slower titration; tolerability focus	**Reduced AE rate** and discontinuation vs. dual therapy in comorbid elderly
Failure to achieve/ maintain low-risk at 3–6 months	**Add prostacyclin pathway:** selexipag 200 µg BID → max 1600 µg BID; or switch to triple therapy	Further risk reduction
High-risk or worsening despite dual therapy	**Triple therapy with parenteral prostacyclin**: IV epoprostenol (start 2 ng/kg/min) or IV/SC treprostinil (start 1.25 ng/kg/min)	Strongest hemodynamic effect; **reserve for high-risk or refractory disease**
Group 3 PH-ILD with non-severe PH	Inhaled treprostinil (Yutrepia) 72 µg QID	**Only approved targeted therapy for PH-ILD**; +31 m 6MWD
Group 4 CTEPH	Surgical PEA (treatment of choice); riociguat for inoperable; BPA for selected patients	Potentially curative (PEA); riociguat improves 6MWD and hemodynamics

ABBREVIATIONS: PDE5i = phosphodiesterase-5 inhibitor; ERA = endothelin receptor antagonist; HR = hazard ratio; AE = adverse event; PEA = pulmonary endarterectomy; BPA = balloon pulmonary angioplasty; CTEPH = chronic thromboembolic PH; GDMT = guideline-directed medical therapy; LTOT = long-term oxygen therapy; 6MWD = 6-Minute Walk Distance; PH-ILD = PH associated with interstitial lung disease
***Consult FDA labels for the most up-to-date dosage information, contraindications, and drug-drug interactions**

RED FLAG — TREATMENT SAFETY

PAH-specific vasodilator therapies (PDE5 inhibitors, ERAs) carry Class III (harm) recommendations for Group 2 (left heart disease) and non-severe Group 3 (lung disease) PH. Prescribing these drugs without RHC-confirmed group classification risks patient harm. The only targeted therapy approved for Group 3 PH-ILD is inhaled treprostinil.

Evidence-Based Treatment Protocols

Treatment selection in pulmonary hypertension is guided by WHO group classification and risk stratification, with distinct evidence-based protocols that differ fundamentally across groups — making accurate hemodynamic classification essential before initiating any targeted therapy.[1,2,15]

PH TYPE AND RISK CATEGORY	FIRST-LINE THERAPY	ESCALATION STRATEGY	MONITORING AND GOALS
Group 1 PAH, low/ intermediate-low risk (WHO-FC I–II, 6MWD >440 m, NT-proBNP <300)	**Monotherapy:** PDE5i (sildenafil 20 mg TID or tadalafil 40 mg daily)	**If worsening 6MWD or NT-proBNP rise:** Add ERA (ambrisentan 5–10 mg or macitentan 10 mg)	**q3-6 months:** assess WHO-FC, 6MWD, NT-proBNP, RHC every 12 months; target 6MWD ≥440 m, NT-proBNP <300
Group 1 PAH, intermediate-high/ high risk (WHO-FC III–IV, 6MWD <440 m, NT-proBNP >1400)	**Dual oral combo:** PDE5i + ERA (e.g., tadalafil 40 mg + macitentan 10 mg); consider prostacyclin (IV epoprostenol, SC treprostinil, or inhaled iloprost) for WHO-FC IV	**If still high-risk at 3–6 months:** Add prostacyclin; consider soluble guanylate cyclase stimulator (riociguat) per ESC/ERS; transplant evaluation	**q1-3 months:** full reassessment; RHC every 6 months; goal is risk reduction by 50% within 6 months
Group 1 PAH, elderly (≥70) with comorbidities	**Monotherapy preferred:** PDE5i (slow titration: sildenafil 20 mg daily or every other day for 2–4 weeks, then TID) OR tadalafil 20 mg daily for 4 weeks, then 40 mg daily	**Cautious escalation:** Assess tolerance at 4–6 weeks; dual oral only if persistently high-risk; monitor for hypotension and syncope	**q3 months:** BP, symptoms, drug tolerance; q6 months: 6MWD, NT-proBNP; lower RAF target: Risk reduction to intermediate-low acceptable
Group 2 PH (left heart disease)	**No PAH-specific drugs Optimize GDMT:** ACEi/ ARB/ARNI + BB + MRA + SGLT2i (HFrEF); loop diuretics + SGLT2i + careful IVF (HFpEF); cardiac rehab	**If PAWP remains elevated despite GDMT:** Optimize diuretics; consider increasing ACEi/ARNI; assess compliance; refer back to cardiology for advanced HF options	**q3-6 months:** Volume status (JVP, edema, weight), diuretic requirement, GDMT dose optimization; annual echo; avoid PAH drugs per Class III
Group 2 PH (left heart disease) — Age >65	**No PAH-specific drugs Optimize GDMT:** ACEi/ ARB/ARNI + BB + MRA + SGLT2i (HFrEF); loop diuretics + SGLT2i + careful volume management (HFpEF); cardiac rehab Start at low doses and titrate slowly; elderly patients at higher risk for hypotension and worsening renal function	**If PAWP remains elevated despite GDMT:** Optimize diuretics; consider increasing ACEi/ARNI; assess compliance; refer to cardiology for advanced HF options PAH-specific drugs (ERAs, PDE5i, prostacyclins) are contraindicated — Class III (harm)	**q3–6 months:** volume status (JVP, edema, daily weight), diuretic requirement, GDMT dose optimization, renal function, BP; annual echo; screen for AF (present in ~47% of PH hospitalizations >65); avoid PAH drugs

PH TYPE AND RISK CATEGORY	FIRST-LINE THERAPY	ESCALATION STRATEGY	MONITORING AND GOALS
Group 2 PH — SGLT2i considerations in elderly	**SGLT2i recommended** across age spectrum including ≥75 years (HR 0.75 for HF hospitalization/CV death) **Adjust diuretic doses** when initiating to avoid volume depletion	**If genital infections occur (RR 3.18)**: treat and continue if tolerated; if eGFR decline >30%: hold and reassess; coordinate ARNI + SGLT2i initiation timing to avoid compounded renal effects	**Monitor eGFR at 1 month, then q3–6 months**; genital hygiene counseling; weight and volume status; steeper eGFR decline expected in elderly with concurrent ARNI
Group 3 PH (lung disease)	**No PDE5i for non-severe (PVR ≤5 WU)** **Optimize underlying disease:** ICS/LABA/LAMA + pulmonary rehab + LTOT (SpO_2 >90%); if severe PVR (>5 WU): inhaled treprostinil only targeted option	**Reassess PVR at 3 months**; if PVR becomes severe (>5 WU) due to progression: add inhaled treprostinil; escalate lung disease management; consider transplant evaluation	**q3 months:** SpO_2, dyspnea severity, exercise capacity; q6 months: PFTs + DLCO; RHC q12 months if on targeted therapy; avoid worsening hypoxia
Group 3 PH (lung disease) — Non-severe (PVR ≤5 WU)	**No PDE5i** **Optimize underlying lung disease:** ICS/LABA/LAMA (COPD) or antifibrotics (IPF) + pulmonary rehab + LTOT targeting SpO_2 ≥89% for ≥15 hrs/day. LTOT qualifies when PaO_2 ≤55 or SpO_2 ≤88%, or PaO_2 56–59 with PH/cor pulmonale	**Reassess PVR at 3 months**; if PVR progresses to >5 WU: reclassify as severe and consider inhaled treprostinil; escalate lung disease management; evaluate transplant eligibility	**q3 months:** SpO_2, dyspnea severity, exercise capacity; q6 months: PFTs + DLCO; avoid systemic vasodilators (ambrisentan caused harm in ILD; riociguat associated with excess mortality in IIP)
Group 3 PH — Severe (PVR >5 WU)	**Inhaled treprostinil (Yutrepia/Tyvaso) — only approved targeted therapy for PH-ILD**; +31m 6MWD at 16 weeks in INCREASE trial. Cautious dose titration in elderly, insufficient data in ≥65; plasma clearance reduced up to 80% in hepatic insufficiency	Continue underlying lung disease optimization; **if functional decline persists on inhaled treprostinil:** consider transplant evaluation (age >70 limits candidacy at most centers)	**q3 months:** 6MWD, NT-proBNP, SpO_2, symptom assessment; RHC q12 months if on targeted therapy; monitor for cough, headache, jaw pain (common inhaled treprostinil side effects)

PH TYPE AND RISK CATEGORY	FIRST-LINE THERAPY	ESCALATION STRATEGY	MONITORING AND GOALS
Group 4 CTEPH	**Surgical pulmonary endarterectomy (PEA) is the gold standard for operable disease.** If inoperable or residual PH post-PEA: riociguat (soluble guanylate cyclase stimulator) 0.5–2.5 mg TID	**Riociguat escalation:** titrate from 0.5 mg TID by 0.5 mg increments at 2-week intervals per tolerance to max 2.5 mg TID; refer to CTEPH center for PEA assessment	**q3-6 months:** WHO-FC, 6MWD, NT-proBNP, RHC if on riociguat; assess for surgical candidacy at specialized CTEPH center; post-PEA surveillance annually
Group 5 PH (multifactorial)	Risk-stratified approach: clarify predominant mechanism (Group 1 vs. 2 vs. 3 pattern); **if Group 1-predominant:** trial of PDE5i; if Group 2/3-predominant: optimize underlying disease first	**Reassess at 4–6 weeks for response**; if PDE5i trial fails: discontinue and optimize comorbidities; refer to PH center for clarification of mechanism and targeted approach	**q3 months:** WHO-FC, 6MWD, NT-proBNP; RHC at 3–6 months to reassess classification; document mechanism clearly to guide escalation

ABBREVIATIONS: WHO-FC = WHO Functional Class; 6MWD = 6-Minute Walk Distance; NT-proBNP = N-terminal pro-B-type natriuretic peptide; PDE5i = phosphodiesterase-5 inhibitor; ERA = endothelin receptor antagonist; IV = intravenous; SC = subcutaneous; RHC = right heart catheterization; PAH = pulmonary arterial hypertension; GDMT = guideline-directed medical therapy; ACEi/ARB/ARNI = ACE inhibitor/angiotensin receptor blocker/angiotensin receptor-neprilysin inhibitor; BB = beta-blocker; MRA = mineralocorticoid receptor antagonist; SGLT2i = SGLT2 inhibitor; HFrEF/HFpEF = heart failure with reduced/preserved ejection fraction; IVF = intravenous fluid; PAWP = pulmonary artery wedge pressure; ICS/LABA/LAMA = inhaled corticosteroid/long-acting beta-agonist/long-acting muscarinic antagonist; LTOT = long-term oxygen therapy; PVR = pulmonary vascular resistance; WU = Wood Units; CTEPH = chronic thromboembolic pulmonary hypertension; PEA = pulmonary endarterectomy; TID = three times daily

***Consult FDA labels for the most up-to-date dosage information, contraindications, and drug-drug interactions**

CLINICAL PEARL — CLUES TO DIG DEEPER

Vasodilators have not proved effective for people with pulmonary hypertension due to chronic obstructive pulmonary disease (COPD). In contrast, vasodilators are often helpful for pulmonary hypertension that occurs in people with the following:

- Idiopathic or inherited pulmonary hypertension
- Autoimmune disorders
- Chronic liver disease
- HIV infection
- Some congenital heart disorders
- Pulmonary hypertension caused by medications or toxins
- Chronic thromboembolic pulmonary hypertension

Non-Rx Treatment and Patient Care

Non-pharmacologic management is the foundation of PH care across all WHO groups and is recommended alongside targeted pharmacotherapy by both the 2022 ESC/ERS guidelines and the ATS/CHEST guidelines.[1,16]

INTERVENTION	KEY DETAILS	CLINICAL IMPACT
Pulmonary rehabilitation	Supervised low-intensity aerobic exercise, breathing techniques, education; programs typically 8–15 weeks	**Improves 6MWD by average of 48.5 m (95% CI 33.4–63.6 m**; 11 RCTs, 418 participants); improves peak VO_2 and quality of life; Class I, Level A recommendation (ESC/ERS 2022)
Nutritional support	Sodium restriction 2,400 mg/day to reduce fluid retention; iron repletion if deficient; fluid restriction to 2 L/day in patients with RV failure and fluid retention	**Sodium restriction is particularly important in managing volume status in RV failure**; fluid restriction to 2 L/day associated with reduced RV area and significantly longer time to clinical worsening
Psychosocial support	Advance care planning, prognosis education, anxiety/depression screening and management	Class I, Level C recommendation (ESC/ERS 2022); disease-specific PROMs (emPHasis-10, CAMPHOR) **track functional status and provide independent prognostic information**
Combination pill simplification*	Opsynvi (macitentan 10 mg + tadalafil 40 mg): once-daily single tablet for Group 1 PAH, WHO-FC II–III	**A DUE trial:** superior PVR reduction vs. either monotherapy; ~50 m improvement in 6MWD at 16 weeks; reduces pill burden and supports adherence
Daily home weight monitoring	Patients on diuretics should regularly monitor body weight; alert threshold: weight gain triggers urgent evaluation	**Weight monitoring is recommended** by ESC/ERS 2022 for all PH patients on diuretics; ACC/AHA HF guidelines recommend daily weights with provider-specified action ranges for diuretic adjustment
Structured symptom tracking	Zone-based action plans (Green/Yellow/Red) using symptom diaries, SpO_2 logs	**Web-based symptom monitoring in pre-capillary PH reduced ED readmissions by 65%** (OR 0.35; p=0.04) and improved 6MWD by 9.8 m per 3-month interval

***Consult FDA labels for the most up-to-date dosage information, contraindications, and drug-drug interactions**

Follow-Up Timing

The follow-up frequencies in this table are derived from the 2022 ESC/ERS Guidelines for the Diagnosis and Treatment of Pulmonary Hypertension.[1]

RISK CATEGORY	FREQUENCY	KEY ASSESSMENT ELEMENTS
Low risk (score 1)	**Every 6 months**	WHO-FC, 6MWD, NT-proBNP; echo annually; maintain current therapy
Intermediate-low (score 2)	**Every 3–6 months**	WHO-FC, 6MWD, NT-proBNP; consider escalation if not improving
Intermediate-high (score 3)	**Every 3 months**	WHO-FC, 6MWD, NT-proBNP, echo; escalate therapy; PH center co-management
High risk (score 4)	**Every 1–3 months or per symptom burden**	Full reassessment; **urgent escalation to triple therapy**; transplant evaluation
Post-hospitalization	**Within 7 days of discharge**	Medication reconciliation; volume status; risk reassessment; care coordinator contact

ABBREVIATIONS: WHO-FC = WHO Functional Class; 6MWD = 6-Minute Walk Distance; NT-proBNP = N-terminal pro-B-type natriuretic peptide

Patient Education and Adherence

PH patients face uniquely complex adherence challenges: PAH-specific drugs are expensive (combination therapy $50,000–$80,000+/year), some require parenteral administration, and medication gaps >45 days are associated with worse outcomes. Shared decision-making should begin at diagnosis and include WHO group-specific education; patients must understand that PH has distinct types requiring different treatments, and that drugs appropriate for one group may harm patients in another.[1]

Documentation example: "Educated patient on recognizing PH warning signs: worsening dyspnea, new edema, lightheadedness, rapid weight gain. Reviewed Green/Yellow/Red zone action plan. Confirmed understanding of medication schedule and importance of continuous therapy. Provided written PH education materials and Pulmonary Hypertension Association resources. Discussed advance care planning. MEAT documented."

Comorbidity Management

CONDITION	PRIMARY TARGET/ GOAL	GUIDELINE MANAGEMENT[2]	CLINICAL NUANCES
Left heart disease (Group 2 PH)	Optimize GDMT; reduce PAWP	HFrEF: ACEi/ARB/ARNI + BB + MRA + SGLT2i; HFpEF: diuretics + SGLT2i; valvular: per indication	**No PAH drugs** (Class III). Volume management is primary intervention. ERAs worsen fluid retention
COPD (Group 3 PH)	Optimize bronchodilation; smoking cessation; LTOT	ICS/LABA + LAMA; pulmonary rehab; O_2 to maintain SpO_2 >90%	**No PDE5i** for non-severe (PVR ≤5 WU). Inhaled treprostinil only targeted option for PH-ILD

CONDITION	PRIMARY TARGET/ GOAL	GUIDELINE MANAGEMENT[2]	CLINICAL NUANCES
Diabetes	HbA1c <8.0% (individualize in frail elderly)	**SGLT2i preferred if HF comorbidity**; metformin if eGFR ≥30	HR 1.73 for death; **contributes to diastolic dysfunction and Group 2 phenotype**
Hypertension	BP <130/80 mm Hg	**ACEi/ARB as first-line**; avoid beta-blockers unless HF/rate control indication	**Common in left heart phenotype**; aggressive control reduces LV afterload
Atrial fibrillation	Rate or rhythm control + anticoagulation	**Rate control preferred** in elderly with comorbidities	~30% of elderly PH patients with left heart phenotype; worsens hemodynamics
Iron deficiency	Ferritin >100 ng/mL, TSAT >20%	**IV iron preferred** over oral (better absorption); correct even without anemia (Class IIb)	**Common in PAH**; contributes to exercise intolerance; correct before considering ESA

ABBREVIATIONS: GDMT = guideline-directed medical therapy; PAWP = pulmonary artery wedge pressure; HFrEF/HFpEF = heart failure with reduced/preserved ejection fraction; ACEi = ACE inhibitor; ARB = angiotensin receptor blocker; ARNI = angiotensin receptor-neprilysin inhibitor; BB = beta-blocker; MRA = mineralocorticoid receptor antagonist; SGLT2i = SGLT2 inhibitor; ICS/LABA/LAMA = inhaled corticosteroid/long-acting beta-agonist/long-acting muscarinic antagonist; LTOT = long-term oxygen therapy; PDE5i = phosphodiesterase-5 inhibitor; PVR = pulmonary vascular resistance; WU = Wood Units; PH-ILD = PH with interstitial lung disease; BP = blood pressure; ESA = erythropoiesis-stimulating agent; TSAT = transferrin saturation; IV = intravenous; HR = hazard ratio

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Cost-Smart Options

MEDICATION CLASS[2,3]	DRUG OPTIONS	ROUTE	ESTIMATED MONTHLY RETAIL COST (USD)
PDE-5 Inhibitors	sildenafil 20mg (generic Revatio), tadalafil 40mg (generic Adcirca)	Oral	**$20 – $100** (Generic) *Highly accessible at retail pharmacies.*
ERAs	bosentan, ambrisentan 10 mg (generic Letairis)	Oral	**$500 – $2,500** (Generic). Limited generic availability
	Macitentan	Oral	**$8,000 – $10,000** (Brand)
GC Stimulators	Riociguat	Oral	**$10,000 – $12,000** (Brand)
Prostacyclin Analogues	Selexipag	Oral	**$12,000 – $15,000** (Brand)
	Treprostinil	Inhaled	**$5,000 – $10,000** (Specialty)
	Treprostinil	SC/IV	**$10,000 – $20,000+** (Specialty)
	Epoprostenol	IV	**$3,000 – $10,000** (Varies by pump/supply)

MEDICATION CLASS[2,3]	DRUG OPTIONS	ROUTE	ESTIMATED MONTHLY RETAIL COST (USD)
Activin Inhibitors	Sotatercept	Injection	**$14,000+** (Brand/Specialty)
Kinase Inhibitors	Seralutinib	Inhaled	Investigational/Specialty pricing

When to Refer

SPECIALTY	URGENT (<2 WEEKS)	ROUTINE (4–6 WEEKS)[2]
PH Specialist Center	**Syncope or presyncope**; hemodynamic instability; signs of acute RV failure (JVP elevation + edema + ascites); WHO-FC IV; new poorly tolerated arrhythmias	Intermediate/high echo probability (TRV >2.8 m/s); risk factors for PAH (systemic sclerosis, portal HTN, HIV, family history); persistent dyspnea post-PE; any diagnostic uncertainty
Cardiology	**New arrhythmia** with hemodynamic compromise; suspected acute PE; chest pain with RV strain on echo	Diastolic dysfunction assessment for Group 2 classification; AV fistula evaluation if advanced disease
Pulmonology	**Acute respiratory failure with known PH;** rapidly worsening hypoxemia	PFT interpretation for Group 3 classification; ILD assessment; pulmonary rehabilitation referral
Palliative/ Supportive Care	Any patient declining targeted therapy or considering conservative management	Persistent high symptom burden; multiple hospitalizations; advance care planning; WHO-FC IV stable

ABBREVIATIONS: RV = right ventricle; JVP = jugular venous pressure; WHO-FC = WHO Functional Class; TRV = tricuspid regurgitation velocity; PAH = pulmonary arterial hypertension; HTN = hypertension; PE = pulmonary embolism; PFT = pulmonary function test; ILD = interstitial lung disease

Quality Metrics Tie-In

MEASURE (MY2026)[4]	STANDARD	APPLICABILITY TO PULMONARY HYPERTENSION
HEDIS COA: Care for Older Adults — Medication Review	**Denominator:** MA enrollees ≥66, continuously enrolled **Numerator:** 2 sub-measures documented annually: **(1)** functional status assessment, **(2)** medication review, **Exclusions:** hospice, ESRD	**Polypharmacy is common in PH patients on combination PAH-targeted therapy** (ERAs, PDE5i/riociguat, prostacyclins); medication review should reconcile: bosentan CYP3A4 interactions (reduces sildenafil levels, contraceptive efficacy, warfarin dosing), PDE5i/riociguat contraindication with nitrates (profound hypotension), selexipag/oral treprostinil CYP2C8 interactions with gemfibrozil and clopidogrel, and ERA-associated fluid retention risk in patients with left heart phenotype[2]
HEDIS COA: Care for Older Adults — Functional Status Assessment	**Denominator:** enrollees ≥66 **Numerator:** functional status assessment documented annually **Exclusions:** hospice, ESRD	**WHO Functional Class (I–IV) and 6-minute walk distance (6MWD) are the core functional assessments in PH** and directly satisfy this sub-measure; document at every visit to track functional trajectory and guide treatment intensity decisions. The ESC/ERS 2022 guidelines recommend multiparametric risk assessment using WHO-FC, 6MWD, and BNP/NT-proBNP at 3–6 month intervals[2]
HEDIS TSC-E: Tobacco Use Screening and Cessation Intervention	**Denominator 1:** enrollees ≥12; **Denominator 2:** positive tobacco users; Numerator 1: screened with documented tobacco status **Numerator 2:** received cessation counseling or pharmacotherapy; Exclusions: death, hospice	**Tobacco smoke is an independent risk factor for PAH development**; smoking history is significantly more common in PAH vs. CTEPH and controls. The "cardiopulmonary phenotype" of IPAH — elderly, predominantly male, heavy smoking history (>30 pack-years), low DLCO (<45%) — has the highest mortality risk and poorest treatment response. Document tobacco status at every visit
HEDIS TRC: Transitions of Care	**Denominator:** enrollees ≥18 with inpatient discharge; **Numerator:** 4 sub-measures within 30 days: (1) notification of inpatient admission, (2) receipt of discharge information, (3) patient engagement after discharge, (4) medication reconciliation post-discharge **Exclusions:** hospice	**PAH 30-day readmission rates reach ~20%, with up to 75% readmitted within 1 year**; mean cumulative cost of additional hospitalization per patient is $71,622. Medication reconciliation is critical for PAH-targeted therapy continuity (especially parenteral prostacyclins), ERA hepatotoxicity monitoring, diuretic adjustment, and anticoagulation management post-hospitalization[5]

MEASURE (MY2026)[4]	STANDARD	APPLICABILITY TO PULMONARY HYPERTENSION
HEDIS DMS-E: Utilization of the PHQ-9 to Monitor Depression Symptoms	**Denominator:** enrollees ≥12 with diagnosis of major depression or dysthymia AND elevated PHQ-9 ≥10; **Numerator:** follow-up PHQ-9 documented within 4–8 months of index elevated score **Exclusions:** hospice	**Pooled depression prevalence in PH is 28.0% (95% CI 20.5–36.8%)**; **anxiety prevalence is 37.1%**. Despite ESC/ERS Class I recommendation for psychological support, less than one-fourth of affected patients receive treatment. PHQ-9/HADS at every visit with follow-up plan if positive[6]
HEDIS ACP: Advance Care Planning	**Denominator:** enrollees ≥66 **Numerator:** advance care plan or surrogate decision-maker documented, OR documentation that ACP was discussed but patient declined **Exclusions:** hospice	PAH is a progressive, life-limiting disease; **5-year survival is ~59%**. Patients may die from progressive right heart failure or sudden death — both scenarios limit communication capacity. ESC/ERS 2022 guidelines recommend ACP with referral to specialist palliative care at the right time; ATS/AAHPM 2022 policy statement recommends early ACP in serious respiratory illness. CPT 99497/99498 billable during AWV with no copay[2]
HEDIS PCR: Plan All-Cause Readmission	**Denominator:** enrollees ≥18 with acute inpatient discharge **Numerator:** unplanned readmission within 30 days (lower is better) **Exclusions:** planned readmissions, hospice, psychiatric	**PAH 30-day readmission rate is ~20–22%; SSc-PH readmission rate is 22–23% with higher in-hospital mortality** during readmission vs. index admission. Primary drivers include right heart failure decompensation, infection, fluid/electrolyte imbalance, and acute respiratory failure. Comorbidity burden (diabetes, CKD, atrial fibrillation, COPD) has increased significantly in PAH admissions[7]
MIPS #130: Documentation of Current Medications	**Denominator:** patients ≥18 with qualifying visit; **Numerator:** current medications documented or notation that no medications are prescribed **Exclusions:** none	**PAH combination therapy involves 2–3 pathway-targeted agents with complex drug-drug interactions:** bosentan is contraindicated with cyclosporine A and glyburide; PDE5i + riociguat combination is contraindicated; CYP2C8 inhibitors raise selexipag/oral treprostinil levels. Document all PAH-targeted medications (name, dose, route, frequency), anticoagulation status, diuretics, and supplemental oxygen at every encounter[8]
MIPS #431: Unhealthy Alcohol Use Screening	**Denominator:** patients ≥18 with qualifying visit; **Numerator:** screened for unhealthy alcohol use using validated tool (AUDIT-C) with brief counseling if positive; **Exclusions:** none	Alcohol use can exacerbate right heart failure, worsen fluid retention, and interact with PAH medications (particularly ERAs with hepatotoxicity risk — bosentan carries a boxed warning for liver injury). AUDIT-C at every visit; brief intervention + referral if positive

MEASURE (MY2026)[4]	STANDARD	APPLICABILITY TO PULMONARY HYPERTENSION

ABBREVIATIONS: 6MWD = 6-minute walk distance; AAHPM = American Academy of Hospice and Palliative Medicine; ACP = Advance Care Planning; ATS = American Thoracic Society; AWV = Annual Wellness Visit; BNP = brain natriuretic peptide; CI = confidence interval; CKD = chronic kidney disease; COA = Care for Older Adults; COPD = chronic obstructive pulmonary obstruction; CPT = Current Procedural Terminology; CTEPH = chronic thromboembolic pulmonary hypertension; CYP2C8 = cytochrome P450 family 2 subfamily C member 8; CYP3A4 = cytochrome P450 family 3 subfamily A member 4; DLCO = diffusing capacity of the lungs for carbon monoxide; DMS-E = Depression Monitoring and Follow-Up (Expanded); ERA = endothelin receptor antagonist; ERS = European Respiratory Society; ESC = European Society of Cardiology; ESRD = end stage renal disease; HADS = Hospital Anxiety and Depression Scale; HEDIS = Healthcare Effectiveness Data and Information Set; IPAH = idiopathic pulmonary arterial hypertension; MA = Medicare Advantage; NT-proBNP = N-terminal pro-B-type natriuretic peptide; PAH = pulmonary arterial hypertension; PCR = Plan All-Cause Readmission; PDE5i = phosphodiesterase type 5 inhibitor; PH = pulmonary hypertension; PHQ-9 = Patient Health Questionnaire-9; SSc-PH = systemic sclerosis-associated pulmonary hypertension; TSC-E = Tobacco Screening and Cessation (Expanded); WHO = World Health Organization; WHO-FC = World Health Organization Functional Class

QUALITY OUTCOME

No PH-specific HEDIS or CMS Star measures currently exist. However, preventing inappropriate PDE5i use in Groups 2 and 3, ensuring RHC-confirmed diagnosis, and reducing PH-related hospitalizations are high-impact VBC metrics that ACOs can track internally. Many PH patients also qualify for heart failure quality measures.

5 CODING REMINDERS AND CASE EXAMPLES

Documentation Specificity

CRITICAL ELEMENT	CLINICAL REQUIREMENT	EXAMPLE DOCUMENTATION
WHO group classification	Specify the underlying cause of PH in every encounter; never default to **I27.20**	"PH due to left heart disease (**I27.22**), Group 2" **NOT** "pulmonary hypertension"
Hemodynamic confirmation	Document RHC values: mPAP, PAWP, PVR with date; these **determine the WHO group**	"RHC 3/25/26: mPAP 34, PAWP 12, PVR 4.8 WU pre-capillary PH"
Pre-capillary vs. post-capillary	PAWP ≤15 = pre-capillary **(Groups 1/3/4/5)**; PAWP >15 = post-capillary **(Group 2)**	"PAWP 22 mm Hg post-capillary PH, Group 2; underlying HFpEF"
Functional class	**WHO-FC I–IV at every encounter**; strongest single predictor of survival	"WHO Functional Class III less-than-ordinary activity causes dyspnea"

CRITICAL ELEMENT	CLINICAL REQUIREMENT	EXAMPLE DOCUMENTATION
Risk stratification	**Three-variable model:** WHO-FC + 6MWD + NT-proBNP; document category	"Four-stratum risk: intermediate-high (WHO-FC III, 6MWD 285m, NT-proBNP 890)"
Underlying etiology code	Code both PH type **AND** underlying condition	"**I27.22 + I50.32** (HFpEF)" or "**I27.23 + J44.1 (**COPD with exacerbation)"
PVR severity (Group 3)	Non-severe (≤5 WU) vs. severe (>5 WU) **determines treatment eligibility**	"PVR 4.8 WU non-severe; PDE5i contraindicated per Class III"
Current treatment and contraindications	**Document** active therapy, guideline basis, and explicit contraindications	"No PAH-specific therapy Group 2 PH; Class III per ESC/ERS 2022"

ABBREVIATIONS: RHC = right heart catheterization; mPAP = mean pulmonary artery pressure; PAWP = pulmonary artery wedge pressure; PVR = pulmonary vascular resistance; WU = Wood Units; WHO-FC = WHO Functional Class; 6MWD = 6-Minute Walk Distance; NT-proBNP = N-terminal pro-B-type natriuretic peptide; HFpEF = heart failure with preserved ejection fraction; PDE5i = phosphodiesterase-5 inhibitor

Annual Clinical Review and Confirmation

Confirm pulmonary hypertension remains **active, classified** by WHO group, and **clinically managed** at least once per calendar year:

- **Annual review:** PH must be reassessed at least once per calendar year via face-to-face or synchronous audio-video encounter, with MEAT documented by 12/31. This includes confirming the WHO group classification, current functional class, and current risk stratum
- **Visit modality:** In-person or video telehealth encounters qualify when they support meaningful evaluation of functional status, hemodynamic trends, and treatment response
- **HCC context:** All PH codes map to HCC 226 (RAF 0.36) under V28. Reconfirmation requires documenting the specific PH type (**I27.21–I27.24**, not **I27.20**), functional class, and current clinical status with objective data (6MWD, BNP/NT-proBNP, echo findings)
- **ICD-10 reconfirmation:** Verify the group-specific code at each annual review. If the underlying cause has evolved (e.g., new CTEPH diagnosis after prior Group 2 coding), update the ICD-10 to reflect the current classification
- **Avoid rollover documentation:** Do not copy forward a prior year's PH note without updating hemodynamic data, functional class, risk stratification, and treatment response. Undated or static documentation does not satisfy MEAT requirements

DOCUMENTATION REMINDER

Under HCC V28, all PH codes map to HCC 226 (RAF 0.36). However, the clinical complexity and cost of PH vary enormously by WHO group from optimizing underlying HF (Group 2) to $150,000+/year parenteral prostacyclin therapy (Group 1 high-risk). Clear, group-specific documentation supports accurate representation of clinical complexity and ensures care aligns with evidence.

Good Documentation vs. Insufficient

INSUFFICIENT	COMPREHENSIVE
"Pulmonary hypertension" (I27.20 — unspecified)	"PH due to chronic obstructive pulmonary disease **(I27.23 + J44.1)**, Group 3, pre-capillary, PVR 4.8 WU (RHC 3/25/26), WHO-FC III"
"Elevated PA pressures on echo"	"Echo sPAP 52 mm Hg (2/15/26); RHC confirmed mPAP 34, PAWP 12, PVR 4.8 WU (3/25/26) — pre-capillary PH, Group 3"
"PAH, stable"	"PAH **(I27.21)**, WHO-FC II, dual therapy: tadalafil 40 mg + macitentan 10 mg. 6MWD 460m, NT-proBNP 180 ng/L low risk. Next reassessment 9/2026"
"PH, on sildenafil"	"Group 1 PAH **(I27.21)**, WHO-FC III, on sildenafil 20 mg TID, escalation to dual therapy (adding macitentan 10 mg) per ESC/ERS 2022; intermediate-high risk"
"Worsening PH"	"PH **(I27.22)**, Group 2, worsening volume overload despite furosemide 80 mg BID. PAWP 28 mm Hg. HFpEF optimized. No PAH drugs per Class III. Diuretic adjustment + cardiology co-management"

ABBREVIATIONS: 6MWD = 6-Minute Walk Distance; BID = *bis in die* (twice daily); COPD = chronic obstructive pulmonary disease; Echo = echocardiography; ESC/ERS = European Society of Cardiology / European Respiratory Society; HFpEF = heart failure with preserved ejection fraction; mPAP = mean pulmonary artery pressure; NT-proBNP = N-terminal pro-B-type natriuretic peptide; PA = pulmonary artery; PAH = pulmonary arterial hypertension; PAWP = pulmonary artery wedge pressure; PH = pulmonary hypertension; PVR = pulmonary vascular resistance; RHC = right heart catheterization; sPAP = systolic pulmonary artery pressure; TID = *ter in die* (three times daily); WHO-FC = WHO Functional Class; WU = Wood Units

EHR Workflow Tips

- **[EHR TIP — Auto] Auto-populates a complete PH visit structure:** WHO group, underlying etiology code, current WHO-FC, last 6MWD and NT-proBNP values, current risk stratum (four-stratum model), current medication list with doses, and follow-up plan. Confirm mapped ICD-10 matches documented WHO group before signing
- **[EHR TIP — Auto] Maps directly to MEAT elements with PH-specific prompts:** MONITOR (functional class, 6MWD trend, NT-proBNP trend, O_2 status, weight log, current therapy); EVALUATE (RHC values with date, echo findings, PFTs, risk stratification calculation); ASSESS (WHO group + ICD-10 + stage + trajectory); TREAT (numbered action items with guideline anchor)

- **[EHR TIP — Alert]: PDE5i prescribing check** — flag active PDE5 inhibitor order when PH diagnosis is Group 2 (**I27.22**) or Group 3 (**I27.23**). Include hard stop or acknowledgment with Class III citation. This is the highest-impact utilization management intervention for PH in the VBC model
- **[EHR TIP — Workflow]: PH risk reassessment panel** — pre-visit worklist for PH patients: pull 6MWD, NT-proBNP, and WHO-FC assessment due within 2 weeks of scheduled visit. Schedule labs 1–2 weeks before encounter so results are available for real-time risk stratification
- **[EHR TIP — Best Practice]: Daily weight integration** — configure home weight data feed from Bluetooth-enabled scale or patient-reported log. Alert care coordinator when weight gain exceeds 2–3 lbs in 1 day or 5 lbs in 1 week. Reduces delayed recognition of decompensation
- **[EHR TIP — Alert]: I27.20** code flag — trigger "WHO group not specified" prompt when **I27.20** (PH unspecified) is added to the problem list or encounter diagnosis. Require clinician to select group-specific code (**I27.21–I27.24**) or document reason classification is not yet possible
- **[EHR TIP — Workflow]: Post-hospitalization PH protocol** — auto-generate follow-up visit within 7 days of discharge for PH-related admission. Include medication reconciliation, volume status assessment, risk restratification, and care coordinator contact in the visit template

Brief Case Examples

✓ SUCCESS — COMPREHENSIVE DOCUMENTATION

SCENARIO

68-year-old man with systemic sclerosis, diagnosed with pulmonary arterial hypertension (Group 1) 2 years ago. **WHO-FC II on dual combination therapy:** tadalafil 40 mg daily + macitentan 10 mg daily. **Current 6MWD:** 460m. **NT-proBNP:** 180 ng/L. **Last RHC (6 months ago):** mPAP 28, PAWP 10, PVR 3.8 WU.

Documentation: Secondary pulmonary arterial hypertension associated with systemic sclerosis (**I27.21 + M34.81**), Group 1, pre-capillary. **WHO Functional Class II:** ordinary activity without symptoms; moderate exertion causes mild dyspnea. **Four-stratum risk:** low (WHO-FC II, 6MWD 460m, NT-proBNP 180 ng/L — score 1.0). **On dual oral combination:** tadalafil 40 mg daily + macitentan 10 mg daily, tolerating well, LFTs normal. **RHC (10/2025):** mPAP 28, PAWP 10, PVR 3.8 WU,stable pre-capillary PAH. **Exercise training:** attending pulmonary rehab 3×/week, 6MWD improved from 420m to 460m over 3 months. Vaccinations current. Daily weights stable. Continue current regimen; reassess in 6 months. MEAT documented.

Outcome: HCC 226 supported. RAF 0.36 × $10,402.34 = ~$3,745/year. **Detailed MEAT:** hemodynamic data with date, functional class, three-variable risk stratification, current therapy with tolerance assessment, and documented improvement trajectory.

⚠ PITFALL — INSUFFICIENT DOCUMENTATION

SCENARIO

"Pulmonary hypertension. On sildenafil. Echo showed elevated pressures. Stable." Coded as **I27.20** (PH, unspecified). No WHO group specified. No RHC values documented. No functional class. No risk stratification.

Documentation: I27.20 maps to HCC 226 but undercodes SOI and ROM. Without WHO group classification, treatment appropriateness cannot be verified. This patient may have Group 2 PH where sildenafil is contraindicated (Class III, harm). No functional data means risk status is unknown and escalation triggers cannot be identified. Documentation does not support active clinical management or demonstrate MEAT.

Outcome: HCC 226 may be supported at 0.36, but undifferentiated documentation cannot demonstrate medical necessity for sildenafil or support the PH diagnosis. Potential clawback of ~$3,745/year if diagnosis is not supportable.

Fix: "Pulmonary arterial hypertension associated with systemic sclerosis (**I27.21 + M34.81**), Group 1, pre-capillary confirmed on RHC 9/2025: mPAP 32, PAWP 10, PVR 5.1 WU. WHO-FC II. On sildenafil 20 mg TID, tolerating well, no visual disturbances. 6MWD 390m, NT-proBNP 420 ng/L: intermediate-low risk. Consider adding ERA for dual combination therapy per ESC/ERS 2022. Reassess in 3 months." HCC 226 (RAF 0.36). Adds ~$3,745/year with documentation by specifying group, hemodynamics, functional class, risk stratum, and treatment rationale.

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