

PULMONARY HYPERTENSION

Pathophysiology, Classification, and Hemodynamic Consequences

Mechanisms that connect pulmonary vascular disease to the bedside

Audience: RN | **Contact hrs:** 1.5 hrs | **Pharmacology hrs:** 0.0 hrs

Expected revision: September 30, 2028

Pharmacology hours are included in the total contact hours.

Activity purpose

Develop an RN-level understanding of pulmonary hypertension through its clinical classification, vascular and molecular mechanisms, tissue changes, right-ventricular consequences, and hemodynamic patterns. Connect these mechanisms with measured findings and practical clinical reasoning to support nursing assessment, patient education, and communication.

Learning outcomes

1. Classify pulmonary hypertension by its principal clinical mechanism and distinguish pulmonary arterial hypertension from the broader PH syndrome.
2. Explain how vasoconstriction, vascular remodeling, and thrombosis alter the pulmonary vascular bed.
3. Relate endothelial, smooth-muscle, adventitial, inflammatory, and genetic mechanisms to pulmonary vascular disease.
4. Distinguish historical histologic grading from contemporary clinical classification and functional assessment.
5. Calculate pulmonary vascular resistance and interpret pre-capillary, isolated post-capillary, and combined hemodynamic patterns.
6. Connect right-ventricular adaptation and failure with nursing assessment, communication, and escalation priorities.

Using this activity

Read the article, work through the numerical examples and fictional clinical cases, and then complete the separate assessment. The reading and assessment form one activity; their contact time is not additive. This is a pathophysiology activity, not a medication-selection or prescribing course.

Clinical perspective

Pulmonary hypertension is not a single disease. Elevated pressure can arise from increased vascular resistance,

high left-sided filling pressure, increased flow, or persistent arterial obstruction. Meaningful interpretation connects the underlying cause, measured hemodynamics, and right-ventricular response rather than treating pressure alone as the diagnosis. The clinical examples in this activity are fictional (Humbert et al., 2023; Kovacs et al., 2024; Vonk Noordegraaf et al., 2019).

01 | From a low-resistance circuit to disease

Normal flow and the meaning of pressure

The pulmonary circulation normally accepts the entire right-ventricular output through a relatively low-resistance, distensible vascular bed. Recruitment and distension of vessels help accommodate increases in flow. Pulmonary hypertension (PH) describes an abnormal pressure state, not a single disease. Understanding the cause of that state requires attention to the vessels, the downstream left heart, the amount of blood flowing through the circuit, and the response of the right ventricle. The pressure–flow–resistance relationship provides a foundation for understanding how these components interact (Humbert et al., 2023; Vonk Noordegraaf et al., 2019).

Pulmonary vascular resistance (PVR) is an important contributor, but not every person with PH begins with a primary pulmonary arteriolar disorder. Pressure may rise because resistance increases, because left-sided filling pressure is transmitted backward, or because pulmonary blood flow is unusually high. These mechanisms can coexist. Therefore, the statement “the pulmonary pressure is high” does not establish the clinical group or identify an appropriate treatment pathway (Humbert et al., 2023; Kovacs et al., 2024).

Hemodynamic definition

Resting PH is defined as mean pulmonary arterial pressure (mPAP) greater than 20 mmHg, measured by right-heart catheterization. Exercise PH is described by an mPAP/cardiac-output slope greater than 3 mmHg/L/min, rather than a single exercise pressure

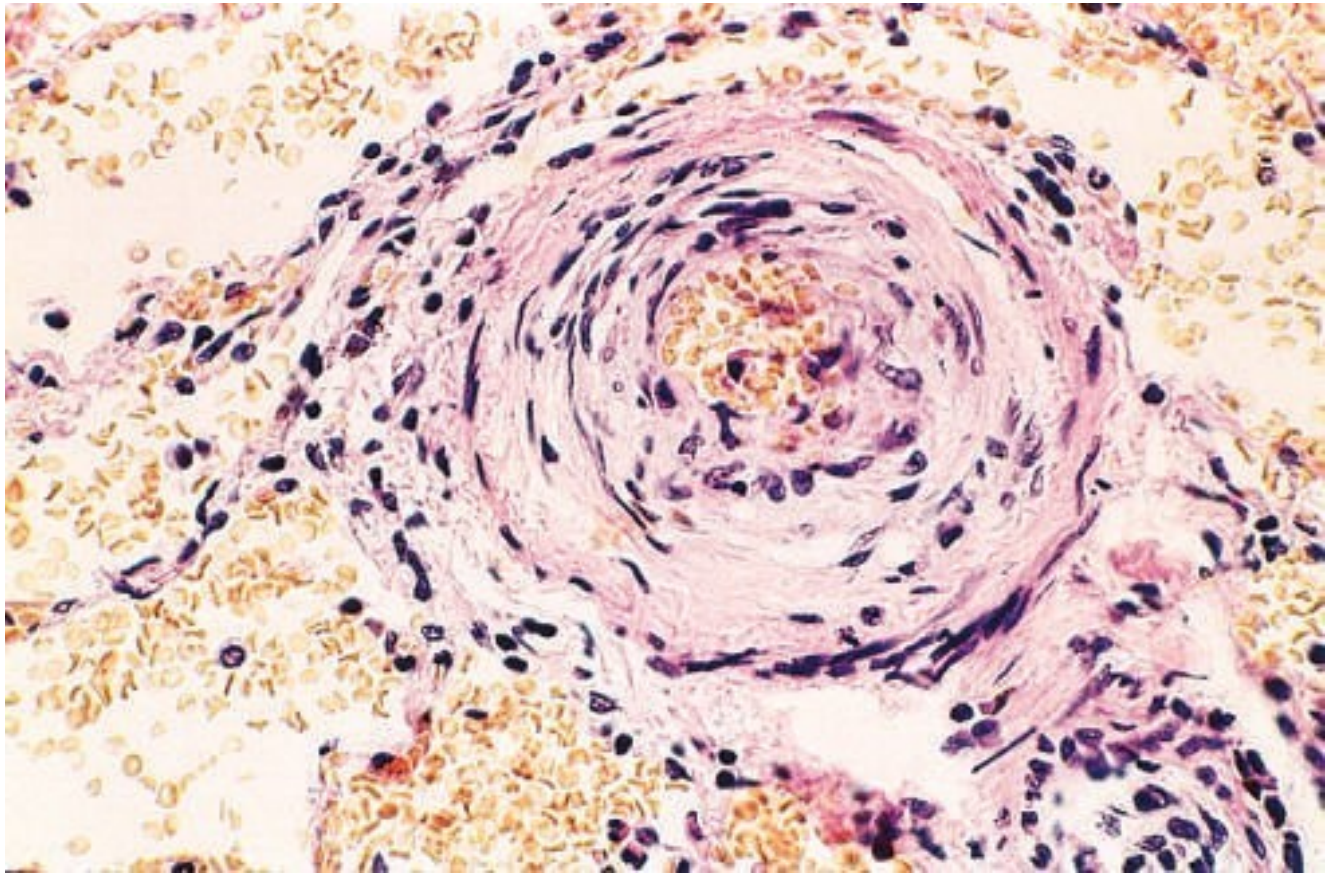


Figure 1. Medial hypertrophy. What to notice: The muscular wall is markedly thickened around a relatively small lumen. **Mechanistic link:** Remodeling reduces the effective cross-sectional area available for blood flow. **Interpretive limit:** A representative teaching image, not a diagnostic specimen from a current patient.

Image credit: Kumar, Abbas, and Aster, Robbins basic pathology (10th ed., p. 518). Elsevier, 2017 (Kumar et al., 2017).

above 30 mmHg. These definitions describe abnormal hemodynamics; they do not, by themselves, establish pulmonary arterial hypertension (PAH) or a medication indication (Humbert et al., 2023; Kovacs et al., 2024).

Read the image as structure, not a bedside test

The medial thickening in the image is a structural explanation for increased resistance, not a finding that can be inferred from one blood-pressure measurement. A narrow lumen leaves less space for flow, while a thickened wall reflects remodeling rather than transient constriction alone. Nurses do not need to diagnose a biopsy specimen to use this concept: persistent structural narrowing helps explain why an elevated pulmonary pressure may remain even after a temporary trigger has resolved (Guignabert et al., 2024; Kumar et al., 2017).

02 | Classification and etiology

Table 1. Clinical classification of pulmonary hypertension: the five-group framework.

Clinical group	Dominant organizing mechanism
1 · PAH	Pulmonary arteriolar disease; defined clinical associations

Clinical group	Dominant organizing mechanism
2 · Left-heart disease	Elevated left-sided filling pressure transmitted upstream
3 · Lung disease / hypoxia	Lung injury, hypoxic vasoconstriction, and vascular-bed changes
4 · Pulmonary artery obstructions	Chronic thromboembolic or other obstructive lesions
5 · Unclear / multifactorial	Several interacting mechanisms or incompletely defined pathways

Sources: Humbert et al. (2023) and Kovacs et al. (2024).

Keep three different questions separate

The clinical group asks why PH has developed. The hemodynamic pattern describes the measured relationship among mPAP, pulmonary arterial wedge pressure (PAWP), and PVR. Functional assessment asks how the disease affects the patient. These are complementary descriptions, not interchangeable labels. A person may have a pre-capillary pattern without having group 1 PAH. Likewise, a histologic grade is not a WHO func-

tional class (Humbert et al., 2023; Kovacs et al., 2024; Rabinovitch et al., 1984).

Group 1: pulmonary arterial hypertension

PAH includes idiopathic, heritable, drug- or toxin-associated, and selected disease-associated pulmonary arteriolar disorders. Associated conditions include connective tissue disease, HIV infection, portal hypertension, congenital heart disease, and schistosomiasis. Idiopathic PAH belongs in group 1. PAH with venous or capillary involvement also requires specialist recognition because involvement is not limited to the muscular arteries. An associated condition helps guide evaluation but does not replace assessment of the complete clinical and hemodynamic picture (Humbert et al., 2023; Kovacs et al., 2024).

Groups 2 and 3: downstream pressure or lung disease

In group 2, left-heart disease raises pulmonary venous and left-atrial pressure, which is transmitted upstream. Persistent loading may eventually be accompanied by pulmonary vascular remodeling. Left-heart disease is the most common cause of PH overall; chronic hypoxia is not a universal primary explanation. In group 3, parenchymal lung disease, hypoxia, and loss or alteration of the vascular bed contribute in varying proportions. The two mechanisms should not be merged simply because both can produce dyspnea and an elevated estimated pulmonary pressure (Maron et al., 2024; Shlobin et al., 2024).

Within group 3, hypoventilation syndromes are important considerations. Isolated nocturnal obstructive sleep apnea alone generally does not explain substantial PH. The clinical team should look for coexisting lung disease, daytime hypoventilation, left-heart disease, or another cause rather than stopping the evaluation after identifying sleep apnea (Humbert et al., 2023).

Groups 4 and 5: obstruction and mixed mechanisms

Group 4 includes pulmonary artery obstructions, especially chronic thromboembolic PH (CTEPH). Persistent organized material and associated small-vessel disease can impede flow long after an acute embolic event. Group 5 contains disorders with unclear or multifactorial mechanisms, including selected hematologic, systemic, and metabolic conditions. It is not a substitute label for an incomplete workup (Kim et al., 2024; Kovacs et al., 2024).

Clinical connection. A patient with systemic sclerosis may have PAH, interstitial-lung-disease-associated PH, left-heart involvement, or more than one contributor. The associated diagnosis is an impor-

tant clue, not a sufficient classification. Nursing handoff should preserve the documented clinical group and report uncertainty rather than relabeling every elevated pressure as PAH.

Phenotypes add detail; they do not replace the groups

American Thoracic Society research phenotypes, such as connective-tissue-disease-associated PH and maladaptive right-ventricular hypertrophy, help describe heterogeneity within and across clinical groups. These phenotypes complement rather than replace the five-group classification. A phenotype can refine the description of a patient while the clinical group organizes the principal cause. Both descriptions may be useful, provided they are not treated as interchangeable labels (Dweik et al., 2014; Kovacs et al., 2024).

03 | The pathogenic triad and the vessel wall

A multiple-hit process

The multiple-hit hypothesis describes how susceptibility interacts with acquired exposures, inflammation, mechanical stress, and other influences. A single abnormal pathway rarely explains the entire disease. For example, endothelial dysfunction may favor constriction and thrombosis, while inflammatory signaling promotes smooth-muscle and fibroblast activity. The resulting wall changes can then alter flow and further stress the endothelium. The model explains interaction; it does not prove that every patient has the same sequence of injury (Guignabert et al., 2024; Humbert et al., 2019).

Vasoconstriction: a change in tone

Vasoconstriction is narrowing caused by active contraction of vascular smooth muscle. An imbalance between relaxing and constricting signals can increase pulmonary vascular tone. Reduced nitric-oxide signaling and greater endothelin activity illustrate this imbalance. Hypoxia can also provoke pulmonary vasoconstriction. A response that redistributes flow locally may become a sustained load when hypoxia is widespread. Not all PH is predominantly vasoconstrictive, and a structural lesion does not disappear simply because smooth muscle relaxes (Guignabert et al., 2024; Humbert et al., 2019).

Remodeling: a change in structure

Remodeling involves altered cell growth, survival, migration, and extracellular-matrix deposition. Medial hypertrophy, muscularization of previously less-muscular distal vessels, intimal thickening, and adventitial expansion can reduce vascular caliber and distensibility. These processes may occur together. Remodeling is therefore more than a vessel “spasm”: it changes the

wall and the space available for blood to move through it. The extent and distribution differ among PH groups and between patients (Guignabert et al., 2024; Humbert et al., 2019).

Thrombosis: obstruction and biological signaling

In-situ thrombosis refers to thrombus developing within the affected vascular bed. Endothelial injury, altered antithrombotic function, and platelet activation can favor this process. Platelets also release vasoactive and growth-related mediators, linking thrombosis with constriction and remodeling. Increased tissue factor and reduced thrombomodulin favor a prothrombotic environment. This mechanism must not be equated with an embolus that traveled from elsewhere or with established CTEPH (Guignabert et al., 2024; Humbert et al., 2019; Kim et al., 2024).

Intima: an active endothelial interface

The intima contains the endothelial layer facing the bloodstream. Injury or dysfunction disturbs the balance among vasomotor regulation, cell survival, inflammation, and clotting. Reduced nitric-oxide availability, increased endothelin signaling, adhesion-molecule expression, and a prothrombotic surface illustrate these disturbances. Endothelium is therefore not an inert lining; it helps determine how the vessel responds to circulating cells and local signals (Guignabert et al., 2024; Humbert et al., 2019).

Media: contraction and muscularization

The media contains smooth-muscle cells that influence tone and wall thickness. Increased proliferation, reduced effective cell removal, and altered matrix turnover can produce thicker muscular walls. Remodeling can include extension of muscle into more peripheral, previously nonmuscular vessels and formation of a neointimal layer. These findings help connect microscopic disease to the rising resistance measured during catheterization. They are descriptions of vascular biology, not parameters that can be measured directly from a routine nursing assessment (Guignabert et al., 2024; Humbert et al., 2019).

Adventitia: the outer wall is involved too

Fibroblasts, inflammatory cells, and matrix proteins in the adventitia can participate in disease. Collagen deposition, cellular proliferation, and changes in the vasa vasorum thicken the outer layer. The “outside-in” concept emphasizes that disease signaling is not necessarily confined to a process beginning at the lumen. Inflammatory recruitment and adventitial activity can influence deeper layers. Both inside-out and outside-in interactions are

useful explanatory models, rather than competing diagnoses at the bedside (Guignabert et al., 2024; Humbert et al., 2019).

Teaching pearl. “Vasoconstriction,” “remodeling,” and “thrombosis” describe different processes. They often overlap, but none should be used as shorthand for all pulmonary hypertension. When reviewing a case, ask which process is supported and what evidence is still missing.

04 | Molecular signals and genetic susceptibility

The endothelium changes the balance

Pulmonary vascular disease involves an imbalance among vasodilator, vasoconstrictor, growth, and clotting signals. Nitric oxide normally supports relaxation through cyclic guanosine monophosphate (cGMP). Reduced availability weakens this relaxing influence. Prostacyclin also contributes to a vasodilatory, platelet-inhibitory environment. These are physiological pathways; discussing an endogenous mediator is not the same as teaching a drug regimen. This activity does not cover selection, dosing, or monitoring of PAH-targeted medications (Guignabert et al., 2024; Humbert et al., 2019).

Endothelin, serotonin, and thromboxane

Endothelin-1 is a potent constricting and growth-promoting signal. Activation of vascular smooth-muscle receptors can increase intracellular calcium and stimulate pathways involved in contraction and proliferation. Platelet-derived serotonin and thromboxane A₂ provide additional links between vascular tone and platelet biology. These mediators do not act independently: their effects intersect with endothelial dysfunction and structural change. A pathway may help explain disease without being a stand-alone measure of its severity (Guignabert et al., 2024; Humbert et al., 2019).

Inflammation and growth factors

Inflammatory cells, cytokines, adhesion molecules, and growth factors participate in remodeling. Macrophages and lymphocytes may be found around affected vessels, and fibroblast activity can expand matrix deposition. Vascular endothelial growth factor, platelet-derived growth factor, and related signals influence cell survival and growth. It is more accurate to describe a dysregulated network than to assign every cytokine the same role in every form of PH. Research findings do not make a single inflammatory laboratory result a diagnostic test for PAH (Guignabert et al., 2024; Humbert et al., 2019).

Vasoactive intestinal peptide

Vasoactive intestinal peptide (VIP) is a vasodilatory peptide whose physiological and experimental effects include smooth-muscle relaxation. Its role contrasts with constricting mediators such as endothelin-1 and thromboxane A₂. Understanding a physiological mechanism does not, by itself, establish a routine treatment recommendation. The discussion here addresses vascular biology, not the selection or initiation of VIP therapy (Petkov et al., 2003).

BMPR2 and incomplete penetrance

BMPR2 encodes a receptor within the transforming-growth-factor-beta superfamily. Pathogenic variants can alter signaling relevant to vascular homeostasis and are an important cause of heritable PAH. However, inheritance of a susceptibility variant does not guarantee that a person will develop PAH. This incomplete penetrance is central to counseling. Genetic susceptibility should be discussed as an increased possibility of disease, not as certainty about an individual's future (Eichstaedt et al., 2023).

Idiopathic PAH means no associated cause has been identified after appropriate evaluation; heritable PAH describes a familial or genetically established context. A negative family history does not exclude genetic susceptibility. Conversely, the presence of PH from left-heart or lung disease does not automatically imply a heritable PAH disorder. Qualified genetic counseling helps explain what testing can establish, what a negative result cannot exclude, and what the result may mean for relatives (Eichstaedt et al., 2023).

Acquired exposures remain relevant

Anorexigen exposure, stimulants, connective tissue disease, HIV, portal hypertension, hemoglobinopathies, chronic hypoxia, and congenital shunts are potential contributors or associated contexts. These should be elicited without assuming that an exposure proves causation in an individual patient. A respectful, specific history is more informative than the vague question “Do you have a risk factor?” Record the exposure or condition and its timing; allow the specialist evaluation to establish the mechanism (Humbert et al., 2023; Kovacs et al., 2024).

Communication example. “The team is considering genetic counseling because some forms of pulmonary arterial hypertension are associated with inherited variants. A variant can increase susceptibility, but it does not mean every carrier will develop

the disease.” This avoids both false reassurance and deterministic language.

05 | Histology and historical grading

A spectrum, not a required checklist

Pulmonary vascular pathology may include intimal hyperplasia, medial hypertrophy, adventitial proliferation, inflammatory infiltrates, in-situ thrombosis, and plexiform arteriopathy. Different findings may coexist, but not every patient has every lesion. Histology also varies with the clinical group and the vascular compartment involved. Figure 2 illustrates a complex plexiform lesion; it should not be taught as a universal feature of all PH (Guignabert et al., 2024; Humbert et al., 2019).

Heath-Edwards grades: what the framework describes

The historical Heath-Edwards system was developed in the setting of pulmonary vascular disease associated with congenital heart disease. In broad terms, grades I–II describe muscularization, medial thickening, and early intimal change; grade III includes more substantial intimal fibrosis; and grades IV–VI encompass advanced lesions such as plexiform or dilatation changes and destructive vascular injury. This is a description of morphology in a specific historical framework, not a bedside staging system for every patient with PH (Kumar et al., 2017; Rabinovitch et al., 1984).

Do not promise reversibility from a grade

Histologic grade alone cannot establish that pulmonary vascular disease is reversible or irreversible in an individual patient. Follow-up after congenital-heart-defect repair has shown that outcome relates to factors beyond the grade alone, including age at repair and additional structural findings. A historical grade cannot independently determine whether an individual shunt should be closed or whether pulmonary vascular disease will resolve. Such decisions require expert assessment of the whole case (Rabinovitch et al., 1984).

Interpretation checkpoint. The phrases “group 1 PAH,” “WHO functional class III,” and “Heath-Edwards grade III” answer different questions. Group identifies the disease category; functional class describes clinical limitation; histologic grade describes tissue findings. Replacing one with another creates a false impression of severity or certainty.

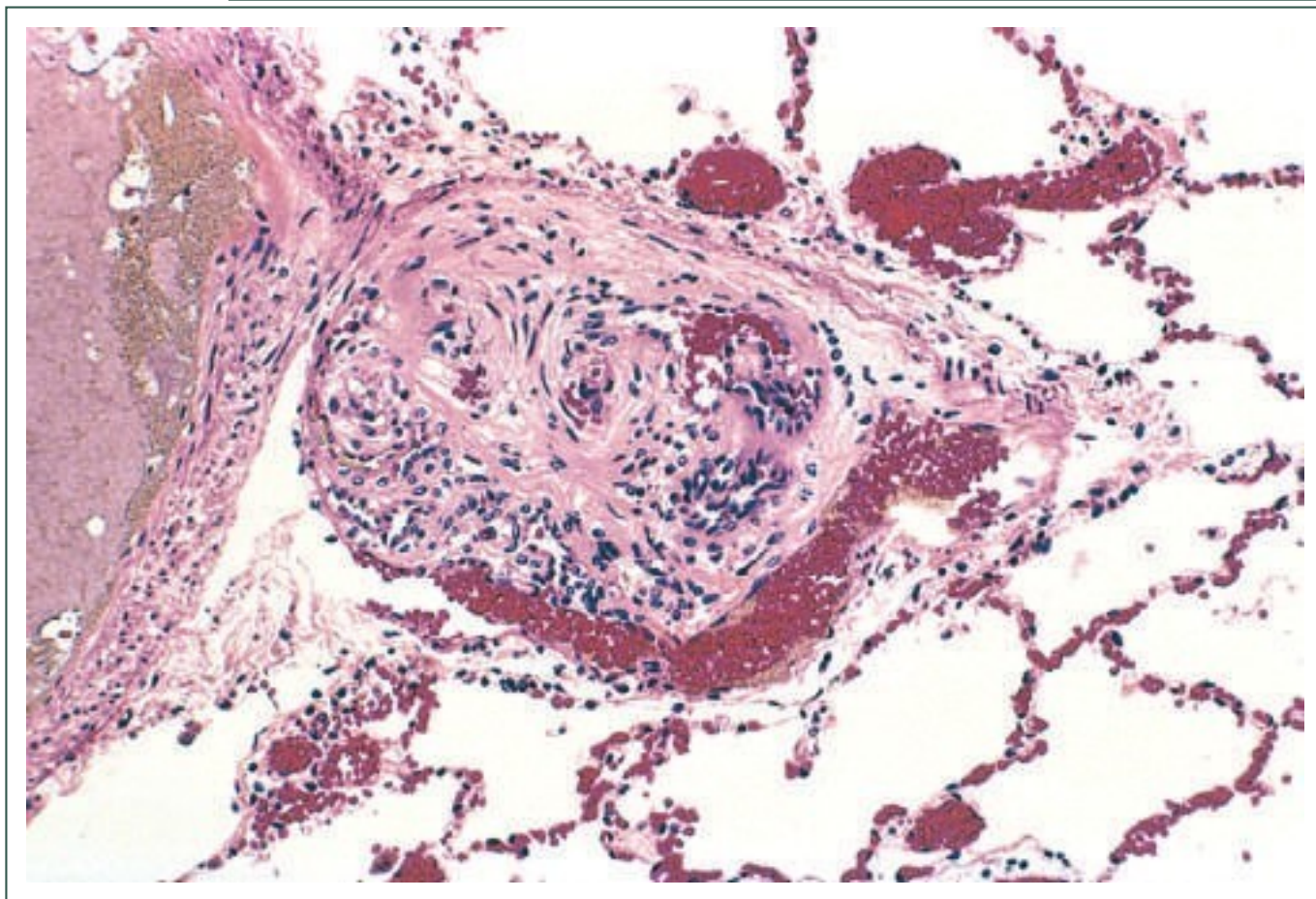


Figure 2. Plexiform vascular lesion. What to notice: A complex cluster of small vascular channels and cellular tissue alters the normal vessel architecture. **Mechanistic link:** Advanced remodeling may produce disorganized vascular structures as well as luminal narrowing. **Interpretive limit:** Plexiform lesions are associated particularly with PAH, not every clinical PH group.

Image credit: Kumar, Abbas, and Aster, Robbins basic pathology (10th ed., p. 518). Elsevier, 2017 (Kumar et al., 2017).

06 | Vascular compartments and persistent obstruction

Large arteries are not the entire story

Figure 3 demonstrates atheromatous change in a large pulmonary artery. This is distinct from muscular remodeling in small resistance vessels. Both findings may belong in a pathology discussion, but they do not establish the same mechanism. A large-vessel plaque should not be used to explain every case of raised PVR, nor should it be mistaken for proof of chronic thromboembolic disease. The appearance illustrates the importance of identifying which vascular compartment is involved (Guignabert et al., 2024; Kumar et al., 2017).

CTEPH: organized material and small-vessel disease

In CTEPH, persistent organized thromboembolic material can obstruct pulmonary arteries. Associated microvascular disease contributes additional resistance, including in regions beyond the obvious obstruction. The result is not simply a fresh clot that has remained unchanged. This distinction matters because evaluation at an expert center can identify options directed at the obstructive component as well as the wider disease

process. Persistent symptoms after pulmonary embolism warrant attention rather than an assumption that breathlessness is merely deconditioning (Kim et al., 2024).

Venous pressure can also load the circuit

In left-heart disease, elevated downstream pressure can raise mPAP even before PVR is increased. Over time, some patients develop an additional pre-capillary component. Thus, pulmonary venous pressure is not a trivial contributor, and “normal PVR” does not exclude PH when left-sided filling pressure is high. Interpretation requires both the wedge pressure and the resistance, rather than assuming that all pressure elevation begins in the pulmonary arterioles (Kovacs et al., 2024; Maron et al., 2024).

Applied reasoning. Three patients can have the same mPAP but different mechanisms: one has low wedge pressure and high PVR, another high wedge pressure with low PVR, and a third has chronic organized pulmonary arterial obstruction. The pressure value alone cannot distinguish these situations. The history, catheter data, and imaging must be interpreted together.

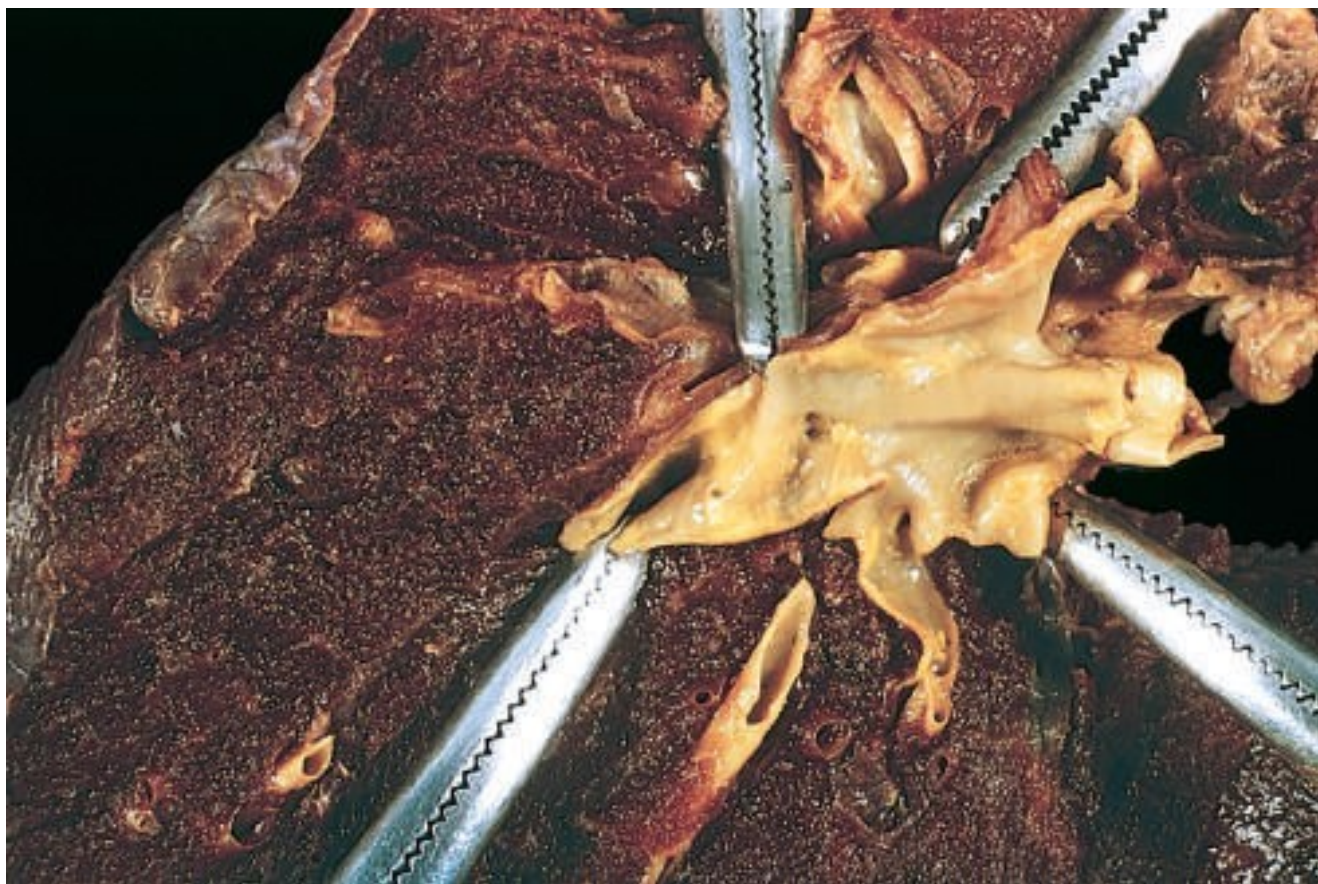


Figure 3. Large pulmonary artery atheromatous change. What to notice: The opened large pulmonary arterial branches show conspicuous pale intimal material. **Mechanistic link:** Macroscopic large-vessel changes differ from the distal arteriolar lesions in Figures 1 and 2. **Interpretive limit:** This pathology photograph does not diagnose CTEPH or determine treatment.

Image credit: Kumar, Abbas, and Aster, Robbins basic pathology (10th ed., p. 518). Elsevier, 2017 (Kumar et al., 2017).

07 | The right ventricle: adaptation and failure

A rising afterload changes the job of the ventricle

Increasing PVR contributes to greater right-ventricular (RV) afterload. The ventricle must generate more pressure to maintain flow. Initially, a thicker muscular wall may help compensate for the pressure load. This adaptation can preserve output for a time even while pulmonary vascular disease is progressing. The absence of obvious congestion therefore does not mean the circulation is normal or that the RV has unlimited reserve (Vonk Noordegraaf et al., 2019).

Afterload is not captured by PVR alone: pulmonary arterial compliance and pulsatile load also influence RV work. For nursing interpretation, the useful lesson is that pressure, resistance, flow, and ventricular performance describe different aspects of the same circulation. A single value cannot represent the entire burden on the ventricle. Catheter findings, imaging, symptoms, and serial assessments should be considered together (Vonk Noordegraaf et al., 2019).

When adaptation becomes maladaptation

Progressive loading may outstrip the ventricle's ability to compensate. Dilation, less effective contraction, and worsening forward flow can follow. Enlargement can contribute to functional tricuspid regurgitation and systemic venous congestion. RV performance is closely related to the patient's course; the ventricle's response is not just a secondary detail after measuring pulmonary artery pressure. The transition from compensated disease to failure should be understood as loss of circulatory reserve, not merely a larger chamber size (Vonk Noordegraaf et al., 2019).

Ventricular interdependence

Both ventricles occupy a shared pericardial space and are connected by the interventricular septum. RV pressure or volume overload can displace the septum toward the left ventricle, impairing left-ventricular filling. Less blood may then be available for systemic ejection even if the primary disorder is in the pulmonary circulation. Ventricular interdependence describes these shared mechanical consequences; the related historical term "reverse Bernheim effect" refers to the effect of right-sided

overload on left-sided filling (Vonk Noordegraaf et al., 2019).

A useful way to teach this is to follow the route of blood rather than memorizing chamber labels: an RV that cannot sustain adequate forward flow delivers less blood through the lungs to the left heart. Septal distortion can further limit left-ventricular filling. The clinical concern is therefore not only backward congestion but also reduced systemic perfusion. These two consequences can occur together rather than representing separate unrelated diseases (Vonk Noordegraaf et al., 2019).

Connect physiology to observation

Breathlessness and declining exertional tolerance may reflect limited ability to increase cardiac output. Edema or abdominal distension can suggest venous congestion. Presyncope or syncope, hypotension, worsening fatigue, and signs of impaired perfusion are particularly concerning when they represent a change from baseline. None of these findings alone establishes the PH group, and common competing causes remain possible. Their value is in the pattern, severity, timing, and trend that the nurse communicates to the clinical team (Humbert et al., 2023; Vonk Noordegraaf et al., 2019).

Why a lower pulmonary pressure may be misleading

From the equation $mPAP = (PVR \times \text{cardiac output}) + PAWP$, a fall in output can lower pulmonary pressure even when resistance remains high. A lower pressure is therefore not automatically evidence of vascular recovery. This is a mathematical inference from the pressure–flow relationship, not a separate diagnostic rule. It is most important when the patient is clinically worsening: a reassuring-looking pressure trend should not override deterioration in perfusion or function (Vonk Noordegraaf et al., 2019).

Worked example. At an earlier assessment, mPAP is 50 mmHg, PAWP is 10 mmHg, and output is 5 L/min: $PVR = 8$ WU. Later, mPAP is 34 mmHg, PAWP remains 10 mmHg, and output is 3 L/min: PVR is still 8 WU. The pressure fell because flow fell in this constructed example. Resistance did not improve. The example illustrates why output and clinical condition must accompany interpretation of pressure.

Keep escalation linked to the patient

New syncope, rapidly progressive symptoms, or signs of RV failure warrant prompt clinical assessment and specialist involvement. An acutely unstable patient requires the local emergency response pathway rather than routine follow-up scheduling. The nurse's role is to

recognize the change, convey reliable findings, and act within the care setting's escalation process. This activity does not supply a resuscitation protocol or authorize independent changes to disease-specific therapy (Humbert et al., 2023; Kovacs et al., 2024).

08 | Hemodynamics: calculate, then interpret

Table 2. Resting definitions measured during right-heart catheterization.

Hemodynamic category	Resting criteria
PH	mPAP >20 mmHg
Pre-capillary PH	mPAP >20; PAWP ≤15 mmHg; PVR >2 WU
Isolated post-capillary PH	mPAP >20; PAWP >15 mmHg; PVR ≤2 WU
Combined post- and pre-capillary PH	mPAP >20; PAWP >15 mmHg; PVR >2 WU

Exact boundary values matter. Sources: Humbert et al. (2023) and Kovacs et al. (2024).

Use the measured variables

PVR is calculated as $(mPAP - PAWP) \div \text{cardiac output}$. Pressures are expressed in mmHg and output in L/min; the result is in Wood units (WU). One WU equals $80 \text{ dyn} \cdot \text{s} \cdot \text{cm}^{-5}$. Pulmonary arterial wedge pressure, historically termed pulmonary capillary wedge pressure (PCWP), estimates the downstream filling pressure in the appropriate measurement context, rather than directly sampling a pulmonary vein (Humbert et al., 2023; Kovacs et al., 2024; Vonk Noordegraaf et al., 2019).

Example A: a pre-capillary pattern

Suppose mPAP is 40 mmHg, PAWP is 10 mmHg, and output is 5 L/min. Subtracting the downstream pressure gives a 30-mmHg pressure gradient; dividing by flow gives $PVR = 6$ WU. With a wedge pressure at or below 15 mmHg and resistance above 2 WU, this is pre-capillary PH. It is not enough information to call the condition idiopathic PAH. Lung disease, chronic arterial obstruction, and other clinical possibilities still require evaluation.

Example B: isolated post-capillary PH

With mPAP 30 mmHg, PAWP 20 mmHg, and output 5 L/min, $PVR = (30 - 20) \div 5 = 2$ WU. The high wedge pressure and PVR at the 2-WU boundary fit an isolated post-capillary pattern. The mPAP remains abnormal even though resistance is not above the contemporary threshold. This constructed example shows how downstream pressure can generate PH without an additional elevated-resistance component.

Example C: combined post- and pre-capillary PH

With mPAP 40 mmHg, PAWP 20 mmHg, and output 4 L/min, PVR = 5 WU. Both downstream filling pressure and resistance are elevated. The combined hemodynamic pattern describes these measurements; it does not imply that two separate clinical diagnoses have already been established. The responsible clinical team must interpret the data in the context of the patient's heart, lungs, volume status, and measurement quality.

Avoid a pressure-only diagnosis

Echocardiography can raise suspicion and assess cardiac structure and function, but it does not replace catheterization for definitive hemodynamic classification. A reported estimated systolic pulmonary artery pressure is not the same variable as measured mPAP. A technically unreliable wedge measurement, a change in loading conditions, or an inaccurate output can alter the calculated pattern. Borderline findings deserve contextual interpretation rather than an automatic label (Humbert et al., 2023; Kovacs et al., 2024).

Calculation check. Record the values and units before calculating. Subtract PAWP from mPAP, then divide by output. Do not divide by a pressure, omit the wedge pressure, or compare a value expressed in $\text{dyn}\cdot\text{s}\cdot\text{cm}^{-5}$ directly with a WU threshold. A correct arithmetic answer cannot repair unreliable source measurements.

09 | Integrated clinical reasoning cases

These fictional cases illustrate the pressure–flow relationship and apply the clinical frameworks explained in the preceding sections. They are educational examples rather than patient records. For each case, separate the calculated hemodynamic pattern from the clinical diagnosis, then identify the observations and uncertainties that should guide nursing communication.

Case 1. A familial association is not the whole diagnosis

A 46-year-old with systemic sclerosis reports worsening exertional breathlessness. Catheterization shows mPAP 34 mmHg, PAWP 9 mmHg, and cardiac output 4 L/min. The chart notes that evaluation of lung disease and chronic thromboembolic obstruction is not yet complete.

Reasoning: $\text{PVR} = (34 - 9) \div 4 = 6.25$ WU, establishing a pre-capillary pattern. Systemic sclerosis is an important PAH association, but it can also coexist with other causes of PH. Calling the case confirmed group 1 PAH before the remaining evaluation would convert a plausible hypothesis into an unsupported

conclusion. The nursing handoff should state the measured pattern, associated condition, symptom trend, and pending studies (Humbert et al., 2023; Kovacs et al., 2024).

Case 2. High pressure without high resistance

A 74-year-old with documented left-heart disease has increased edema. Catheterization shows mPAP 32 mmHg, PAWP 24 mmHg, and output 5 L/min. A family member asks whether the high pulmonary pressure means that the patient has the same disease as a relative with idiopathic PAH.

Reasoning: $\text{PVR} = (32 - 24) \div 5 = 1.6$ WU. The pattern is isolated post-capillary PH, consistent with the documented left-heart context. A useful explanation is that elevated pressure on the left side can be transmitted backward into the lung circulation. The pressure is real, but its mechanism differs from an isolated pulmonary arteriolar disorder. The case does not support choosing a PAH-specific medication. It supports accurate classification and communication of congestion and its trend (Kovacs et al., 2024; Maron et al., 2024).

Case 3. Persistent symptoms after embolism

A 58-year-old remains short of breath months after a pulmonary embolism. Imaging identifies chronic organized pulmonary arterial obstruction. Catheterization shows mPAP 38 mmHg, PAWP 11 mmHg, and output 4.5 L/min. The patient believes that ongoing symptoms must mean the original clot is simply still “fresh.”

Reasoning: $\text{PVR} = (38 - 11) \div 4.5 = 6$ WU. The history and imaging point toward chronic thromboembolic disease rather than in-situ thrombosis as an isolated microscopic feature of PAH. Organized obstruction and additional small-vessel changes may both contribute. The team should arrange the appropriate expert evaluation; the nurse can reinforce the purpose of follow-up and report changing symptoms. The case does not determine operability or select a procedural treatment (Kim et al., 2024).

Case 4. A lower number with a sicker patient

A patient with established PH becomes light-headed with minimal activity and has cool extremities and less urine output than usual. The most recent measured mPAP is lower than at the previous assessment, but cardiac output has also fallen. Another staff member suggests that the pressure reduction proves improvement.

Reasoning: the equation allows mPAP to decrease when output falls. Clinical deterioration cannot be dismissed because one pressure value is lower. The priority is prompt evaluation of instability and communication of the complete hemodynamic and clinical picture. The nurse should not infer that medications can be reduced, wait for a routine appointment solely because pressure is lower, or assign a new PH group from the change. This is an application of the RV and pressure–flow principles, not a standalone treatment algorithm (Humbert et al., 2023; Vonk Noordegraaf et al., 2019).

Transfer the learning

Across all four cases, use the same sequence: identify the values that were actually measured; calculate the resistance when appropriate; name the hemodynamic pattern; distinguish that pattern from the clinical group; and relate the results to the patient’s current condition. State what remains uncertain. This makes a handoff more useful than repeating “pulmonary pressures are high” without a mechanism or trend.

10 | Nursing application and consolidation

Assessment that follows the physiology

The core nursing contribution is a reliable description of change. Compare current exertional capacity, breathlessness, edema, fatigue, and any presyncope or syncope with the person’s established baseline. Document the setting in which symptoms occur and whether recovery takes longer than usual. When appropriate in the care setting, integrate vital signs, oxygenation, weight trend, urine output, and signs of perfusion or congestion. These observations support the team’s interpretation; they do not independently establish a PH subtype (Humbert et al., 2023).

Make the handoff specific

A mechanism-focused handoff separates confirmed findings from inference. State the documented PH group, or note that classification is incomplete. Identify whether the reported pressure was estimated by echocardiography or measured during catheterization. Include the accompanying wedge pressure and output when discussing PVR. Report clinically important changes and the actions already taken. This structure connects clinical communication with the hemodynamic model and reduces the chance that a pressure estimate is misrepresented as a definitive diagnosis (Humbert et al., 2023; Kovacs et al., 2024).

Example handoff. “This patient has a documented pre-capillary pattern; the clinical group is still under evaluation. Since yesterday, exertion causes new presyncope. The latest catheter report lists mPAP, wedge pressure, and cardiac output; today’s vital signs and perfusion findings are a change from baseline. I have initiated our escalation process and am requesting immediate clinical assessment.” The wording intentionally distinguishes known data, new observations, and unresolved classification.

Education without overpromising

An understandable explanation starts with the circuit: “The right side of the heart pumps blood through the lungs. Several different problems can raise pressure in that circulation. The tests help the team decide whether the main problem is in the lung vessels, the left heart, the lungs themselves, or an obstruction.” Avoid saying that all PH is inherited, all disease is caused by low oxygen, or all patients need the same therapy. These statements overlook the different mechanisms and clinical groups (Humbert et al., 2023; Kovacs et al., 2024).

Stay within the scope of this activity

This reading develops mechanism-based understanding. It does not teach medication initiation, dose adjustment, vasoreactivity-test interpretation, shunt-closure eligibility, anticoagulation selection, or perioperative management. Recognizing a physiological pathway is not sufficient to choose a treatment. Use established orders, institutional procedures, and specialist recommendations for patient-specific care. The distinction is particularly important when a learner is familiar with medication names but has not yet established the clinical PH group.

Essential conclusions

PH is a pressure-defined syndrome with several clinical causes. PAH is one clinical group within it. Vasoconstriction, remodeling, and thrombosis may interact, but their contributions differ. The intima, media, and adventitia all participate in vascular biology. Histology describes structure and does not replace clinical classification. RV adaptation may maintain flow for a period, while RV failure can impair both forward perfusion and venous drainage. Hemodynamics should therefore be interpreted as a set, not as an isolated pressure (Guignabert et al., 2024; Humbert et al., 2023; Kovacs et al., 2024; Vonk Noordegraaf et al., 2019).

The practical endpoint is not memorization of every mediator. It is the ability to explain why a patient with high downstream pressure differs from a patient with high resistance, why an inherited susceptibility is not a guaran-

tee of disease, and why a falling pressure is not reassuring when output and clinical condition worsen. Apply that reasoning to the assessment questions and review any explanation that reveals a gap.

Terminology review

PH is pulmonary hypertension; PAH is pulmonary arterial hypertension. PVR is pulmonary vascular resistance. PAWP is pulmonary arterial wedge pressure, historically termed PCWP. CO is cardiac output; mPAP is mean pulmonary arterial pressure. RV and LV denote the right and left ventricles. CTEPH is chronic thromboembolic pulmonary hypertension. WU denotes Wood units. BMPR2 denotes bone morphogenetic protein receptor type 2. These abbreviations describe distinct measurements, diagnoses, and biological components; none should be substituted for another in documentation (Eichstaedt et al., 2023; Humbert et al., 2023; Kovacs et al., 2024; Vonk Noordegraaf et al., 2019).

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